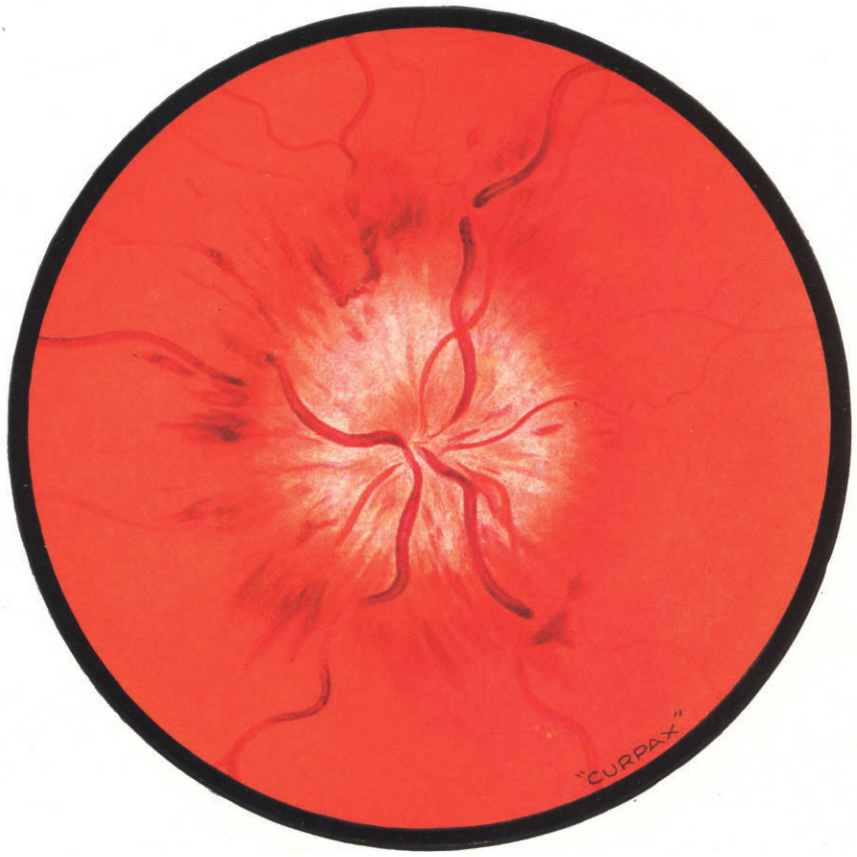

Papillœdema or " choked disc " due to raised intracranial tension. In this instance the papillœdema was due to a frontal lobe tumour. See also Plate III. (Case in the care of Dr. L. R. Yealland.)

NEURO-OPHTHALMOLOGY

BY

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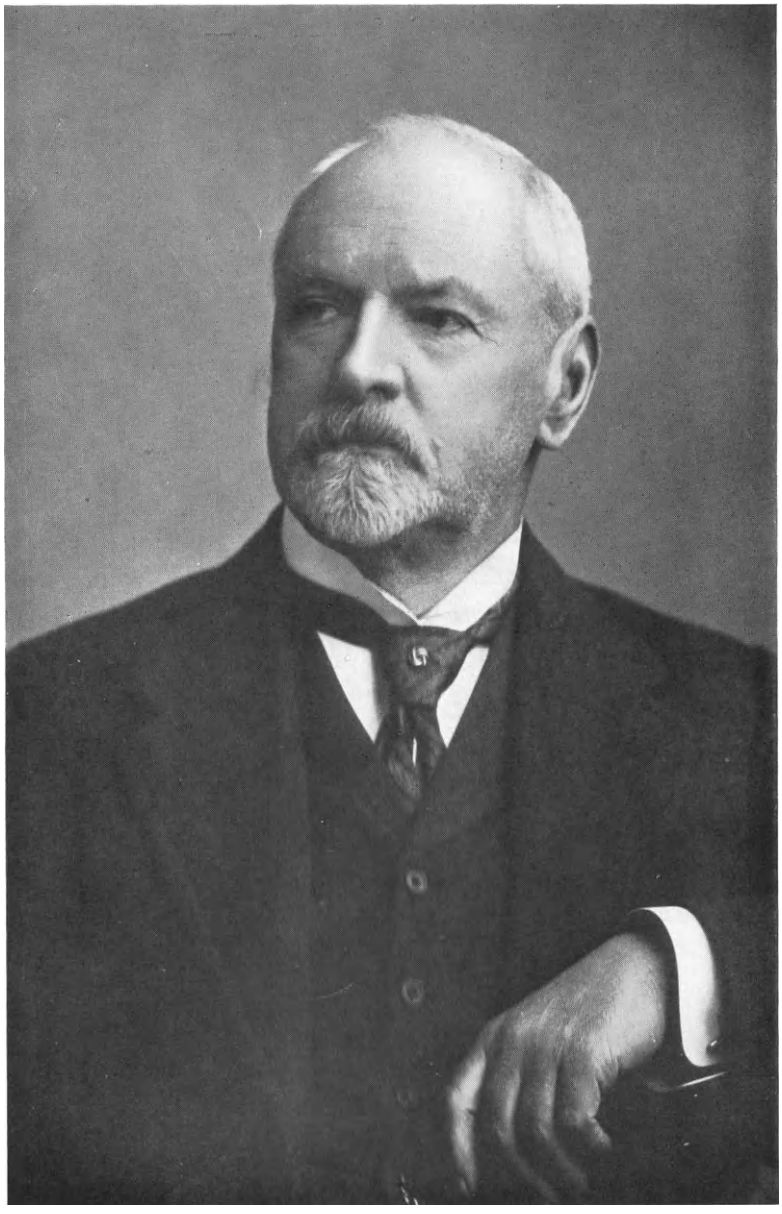

PROFESSOR JOHNSON SYMINGTON

THIS BOOK IS AFFECTIONATELY
DEDICATED TO THE MEMORY OF THE LATE
PROFESSOR JOHNSON SYMINGTON
M.D., F.R.S.,
WHO OCCUPIED THE CHAIR OF ANATOMY
IN THE QUEEN'S UNIVERSITY OF BELFAST

A GREAT ANATOMIST, A GREAT TEACHER
AND A TRUE FRIEND

PREFACE

At the request of the Fellowship of Medicine I have given from time to time demonstrations on the fundus oculi and lectures on neuro-ophthalmology at the West End Hospital for Nervous Diseases. Both from my own experience in preparation for these lectures and from questions put to me by the students, I have become aware of the need for a book which, while keeping to moderate proportions, will supply an answer to the queries regarding those subjects which form a connecting link between ophthalmology and neurology. As the subject-matter grew one learnt to understand why previous writers found it necessary to fill volumes or make large editions. In this book a bibliography has been appended containing the names of authors many of whom have added to their writings complete lists of references. The use of such a bibliography, I hope, will fill the want created by the necessity of keeping this volume within reasonable limits.

The invention of the reflecting ophthalmoscope by von Helmholz in 1851 brought in its train a considerable addition to our knowledge of medicine which was still further increased by the extensive observations of von Graefe and many others. It may be recalled that when von Graefe for the first time saw the back of the eye, with its nerve entrance and its blood vessels, his cheeks reddened and he called out excitedly " Helmholz has unfolded to us a new world." In justice it should be remembered that in 1847 Charles Babbage, a distinguished mathematician at Cambridge, invented an ophthalmoscope which consisted of a mirror with some spots of silver scraped off. Its worth was not then recognised. In Gowers' preface to the 4th Edition of " Medical Ophthalmology," he says : " When this book was written 25 years ago (1879), the subject with which it dealt was more familiar to physicians who constantly used the ophthalmoscope than to ophthalmic surgeons." The first reported case by A. H. Bennett of the removal of a brain tumour, on November 25th, 1884, which was performed by Sir Rickman J. Godlee, at the Hospital for Epilepsy and Paralysis, Maida Vale, contains a meticulous account

of the patient's signs and symptoms, including the state of the fundus oculi before and after the operation. Bennett mentions that optic neuritis was present in both eyes and after the operation the optic neuritis was distinctly improved. Hughes Bennett did not seem to have called in the aid of an ophthalmic surgeon. It was true, physicians did then depend on their own ophthalmic observations.

F. E. Batten, at Queen's Square Hospital for Epilepsy, in conjunction with Mayou, described " Familial Cerebral Degeneration with Macular Changes." This aroused in the mind of his brother Rayner Batten an interest in the ophthalmoscopic appearances of various macular conditions, and he in his turn has shown the value of an accurate appreciation of the state of the macula in fundus examinations.

Since the invention of the perimeter by Förster, in 1857, the quantitative method of estimating the field of vision has come to be widely known due to the efforts of Bjerrum, Roenne, Peter and Traquair. Knowledge of the anatomy, physiology and pathology of the visual paths has thus gradually been unfolded and has brought to light the fact that there must be co-operation between the ophthalmic surgeon and the neurologist ; the one cannot do without the other and although progress has reached the stage when the location of tumours of the brain may be discovered by the electro-encephalogram (Walter), yet this fundamental knowledge concerning visual paths can never be dispensed with. In every hospital for diseases of the nervous system an ophthalmic clinic has been established. The surgeon must be aware, although in a limited degree, of what is in the mind of the physician when a case is referred to him, and it is the personal experience in such a clinic that the author has drawn upon. It is his earnest hope that this book may be of value to many of the younger ophthalmic surgeons taking up work in such clinics as well as to students of neurology.

I desire to acknowledge the help which I have received from Dr. J. Purdon Martin, who has kindly read the book in proof stage. It was impossible to carry out all his suggestions but some have been incorporated in the text. The index has been carefully prepared by Mr. R. McIver Paton. The help provided by my secretary, Mrs. B. Pierce-Jones, M.A., in the work of translation, reference finding and as an amanuensis is deeply appreciated. Many of the illustrations, both in colour and black and white, have been executed by Mr. E. A. Place, whose services have been freely put at my disposal by Messrs.

Clement Clarke Ltd. Those authors who have kindly permitted me to use illustrations are acknowledged in the text. Standard text books on neurology such as those by Grinker, Brain, Wechsler, Thomson and Riddoch, Gehuchten, also " Recent Advances in Neurology " by Brain and Strauss, have been freely consulted. I would like to place on record the unfailing courtesy and helpfulness of those in charge of the Library of the Royal Society of Medicine. Finally, I wish to thank the publishers, Messrs. William Heinemann (Medical Books Ltd.), for their patience and encouragement in the preparation of this volume.

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HARLEY STREET, LONDON, W.

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"Transactions of Ophthalmology," Figs. 59, 100, 126, 128, 129, 141.

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Messrs. Longmans, Green & Co., Figs. 16, 16A, 21, 23, 58, 83 (Quain's "Anatomy"). Plate 1.

Oxford University Press, Figs. 18, 19, 26, 61, 62 (modified from Cunningham's "Manual of Practical Anatomy"), 69, 120, 121 (Harrison Butler's Slit-Lamp).

Miss M. E. Rea, Fig. 21.

Messrs. H. K. Lewis & Co., Figs. 24 (Wolff's "Anatomy of Eye and Orbit"), 51, 52, 112 (Rea's "Affections of Eye"), Plate 2, Plate 3, Plate 7, Fig. 1, Plate 14.

Masson & Cie., Fig. 28 (*Manuel de Neurologie oculaire*).

British Journal of Ophthalmology, Figs. 34, 43, 57, 59, 81, 139.

Proceedings of the Royal Society of Medicine, Figs. 35, 36, 72, 91, 93.

Brain, Figs. 40, 63, 64, 116.

Archives of Ophthalmology, Figs. 47, 95, 110.

Messrs. Burroughs Wellcome & Co., Plate 19.

Messrs. Lea & Febiger, Figs. 55, 117, 136 (Peter's "Perimetry").

Archives of Surgery, Fig. 66.

Edinburgh Medical Journal, Figs. 70, 73

Governors of the Hospital for Sick Children, Great Ormond Street, Fig. 71.

"Medical and Biological Research," Fig. 74.

Journal of Neurology and Psychopathology, Figs. 75, 76.

Kegan Paul, Trench, Trübner & Co., Fig. 77 (Crookshank's "Mongol in our Midst").

Post Graduate Medical Journal, Fig. 79.

Messrs. Wm. Heinemann (Medical Books) Ltd., Fig. 111 (Collins and Mayou's "Bacteriology"), Fig. 119 (Hewer & Sands' "Nervous System").

Lancet, Fig. 118.
Oxford Medical Press, Figs. 133, 135 (Rawling's " Surgery of the Skull ").
Julius Springer, Berlin, *Zeitschrift. f. d. ges. Neur. and Psych.*, Fig. 138.
Messrs. Henry Kimpton, London, Fig. 140 (Atkinson's " External Diseases of the Eye ").
Oxford Medical Publications, Plate 18 (E. Clarke's " Fundus of the Human Eye.")

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LIST OF ILLUSTRATIONS

NEURO-OPHTHALMOLOGY

CHAPTER I

EQUIPMENT NECESSARY FOR EXAMINATION OF THE EYE

(*a*) **Snellen's Test Type** (see Fig. 1) should be hung on the wall at a distance of 6 metres (or 20 feet) from the patient. Should the room not be long enough, reversed type may be obtained, and this, when hanging above the patient and seen in a mirror 10 feet away, will give the same data as regards visual acuity.

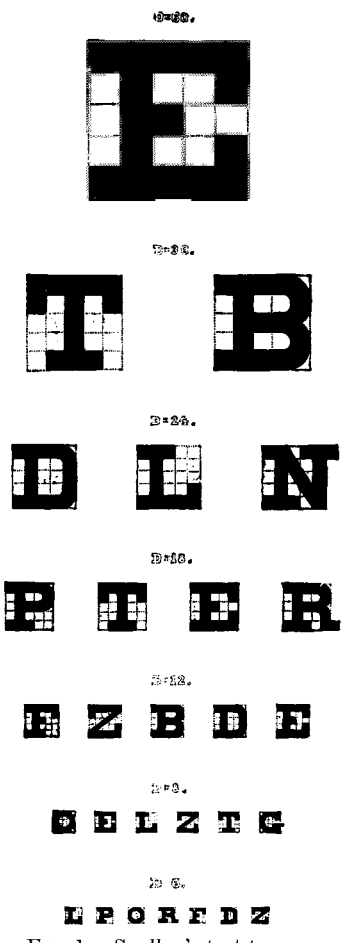


FIG. 1.—Snellen's test type.

Visual acuity is expressed by a fraction, the numerator of which denotes the distance at which the patient is sitting from the test type, while the denominator expresses the distance at which the smallest letters seen by the patient should be perceived by a normal eye. For example, if the patient, seated 6 metres away from the test type, can

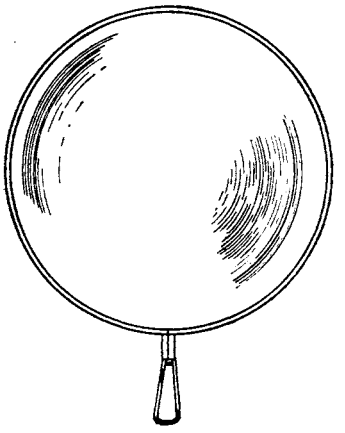


FIG. 2.—Magnifying glass in spectacle mount.

see the top line of the test type only (this letter should be seen by a normal eye at 60 metres), then the visual acuity is 6/60.

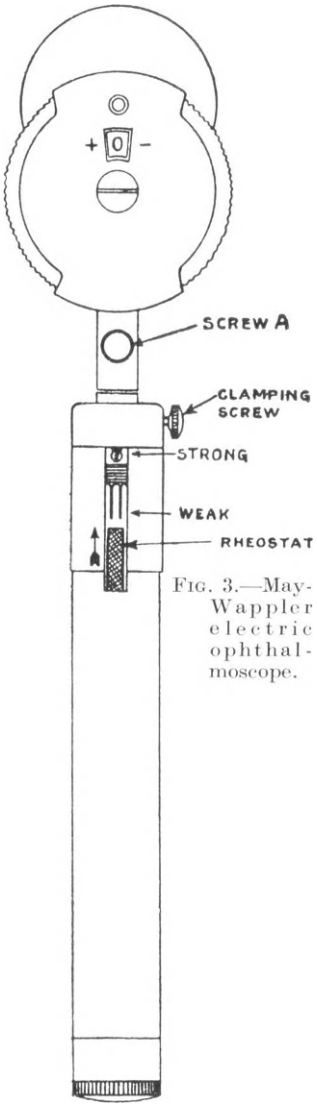


FIG. 3.—May-Wappler electric ophthalmoscope.

(*b*) **A 13 Dioptre Lens** 2 inches in diameter in a spectacle mount (see Fig. 2). This magnifying glass is of the greatest use. By its means light is thrown on the eye enabling one to observe the condition of the iris and its movements in response to light stimuli. When light is focussed on the eye by means of this lens the pupil normally contracts. This is known as the direct light reflex. If light is thrown on one eye and the pupil of the other be observed, the latter in normal cases should be seen to contract also. This is known as the consensual light reflex. Without throwing light on the eyes, if the patient directs his gaze to a point about 8 inches away, the pupils will be seen to contract; this contraction occurs simultaneously with the act of accommodation.

(*c*) **Electric Ophthalmoscope.**—The practitioner or student should also be in possession of an electric ophthalmoscope. It should be remembered that the most expensive is not always the most useful. The small electric ophthalmoscope with a prism reflector is infinitely better than a mirror with a slit. An ophthalmoscope, such as is illustrated in Fig. 3, is the one the writer often uses for hospital work. No time is wasted in endeavouring to see a fundus with this little instrument. Formerly it took months for a student to learn the use of the older type of ophthalmoscope, and, although it may be a desirable thing for the student to learn the indirect method of using the reflecting ophthalmoscope such as Morton's, he can examine an eye by the

direct method with little previous experience, using this small instrument which I have described. The time should come when every doctor will possess such an electric ophthalmoscope. The battery may be replaced for a few pence, and the same battery may be used in the Ever Ready pocket torch. It is a great saving of time and is most convenient, for by using this small prism-reflecting ophthalmoscope one is enabled to go round a hospital ward and examine fundi with or without dilatation of the pupils. A more expensive and exceedingly useful instrument is the new Hamblin ophthalmoscope.

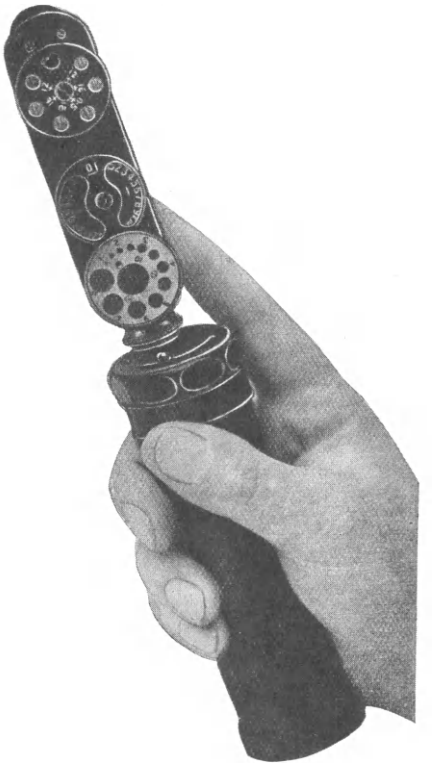


FIG. 4.—The Lister-Morton ophthalmoscope.

The view of the fundus oculi obtained by this instrument is incomparably superior to that of all previous instruments in point of diminished corneal reflex; absence of general glare from cornea; large field brilliantly and evenly illuminated with no trace of filament image or other irregularities, together with freedom from reflex at the edge of the sight hole. These features combine to give a clarity of view which permits direction examination of the macular region in a fully lighted room, without dilatation of the pupil.

At the same time the optical system which affords the above improvements permits the use of the instrument for the indirect method and as a self-luminous retinoscope. All these modes of use can be employed in a fully lighted room should occasion require. The method of adjustment is extremely simple, the only movement necessary being to slide the head of the instrument up or down. The best position for retinoscopy is found by first focussing a sharp image of the lamp filament on a wall at a metre distance and afterwards

bringing the head of the instrument towards the handle until the image is converted into a small homogeneous patch of light.

It is generally conceded that for both direct and indirect opthalmoscopy a good light is essential and one of the features of the Lister-Morton ophthalmoscope is its powerful concentration of light.

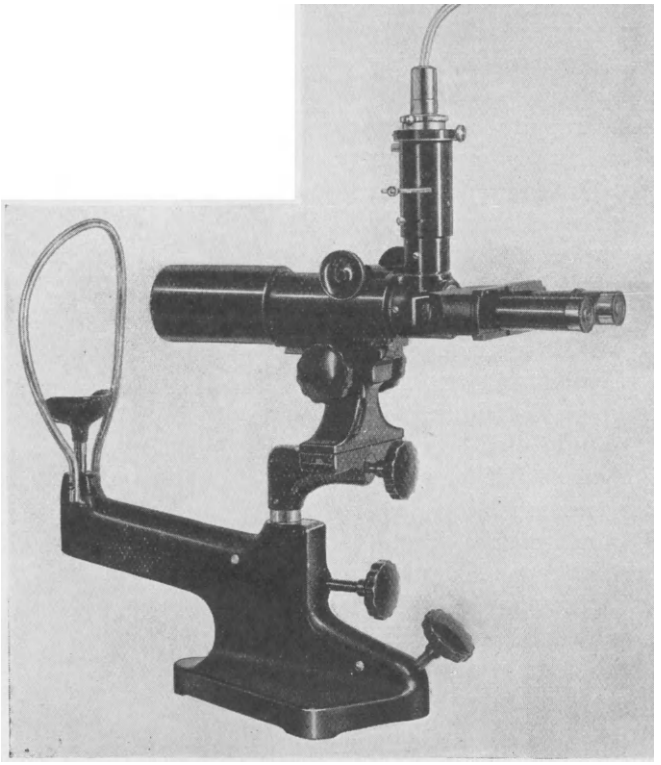


FIG. 5.—The Bausch and Lomb binocular ophthalmoscope.

For teaching purposes Messrs. Clifford Brown Ltd. have placed at the author's disposal the Bausch and Lomb binocular ophthalmoscope for stereoscopic fundus examination illustrated by Fig. 5. This instrument has many of the advantages of the large Gullstrand and is much less costly. The intensity of light is controlled by a resistance in the wiring circuit. The field of view is large, being equal to four discs' width so that disc and macula can be viewed simultaneously. The high illumination of the instrument makes red-free ophthalmoscopy possible.

(*d*) **Perimeter.**—Some form of perimeter is absolutely essential. One of the least expensive is that known as Bishop Harman's *scotometer* with a perimeter arm. One can record not only the patient's visual fields but also the scotomata or blind areas within them, and by its means may also measure the size of *the blind spot*—the area in the field of vision which corresponds to the optic nerve head.

Messrs. Rayner, of London, have recently put an exceedingly fine type of perimeter on the market (see Fig. 6). It is simple in construction and its movements perfectly sound. It consists of a solid aluminium turned arc carrying as a field point one of the series of coloured studs simply pressed into a spring holder in the carrier. The carrier itself is not seen, being enclosed in the arc. The standard chart used is the same as in the McHardy perimeter. The instrument is used in all the Egyptian Government ophthalmic hospitals.

FIG. 6.—Rayner's recording perimeter.

(*e*) Various forms of scotometers are in use, but that which has satisfied the writer most is the one known as the Armstrong recording scotometer, manufactured by Clement Clarke, of London.

This ingenious mechanical apparatus, designed by Dr. H. M. Armstrong, automatically records on a small chart the scotomata occurring in the central portion of the visual field outward to the 30 degrees circle.

The instrument consists of a strong upright wooden frame fitted

with an adaptable chin rest. The upper two-thirds of the frame is covered with black cloth and corresponds with the usual Bjerrum screen. The centre point is marked for fixation purposes, and each blind spot is indicated by a cross stitched on the cloth. A minute test object holder is suspended by fine phospho-bronze black wire and is moved in every direction over the cloth by a handle provided on the back of the screen. The chart holder is fitted into the top of the screen and records the movement of the test object over the screen in the ratio 1 : 5.

The chin rest, which folds back when the screen is not in use, is pulled out and falls into position two-thirds of a metre in front of the screen. The height of the chin rest from the floor is then such that a patient sitting on an ordinary chair will have the chin comfortably supported. A chart is inserted in the chart holder with its printed surface facing the patient. A small test object is selected and placed in the test object holder. The patient is then seated at the chin rest with one eye occluded and fixes the central point on the screen in the usual way. The operator stands at the side of the screen with one hand behind it holding the handle of the pantograph and trigger. On moving the handle over and in contact with the back of the screen the test object will follow its movements exactly over the front of the screen. The patient's answers as to when he sees the test object or when it disappears are recorded by sharply depressing the trigger on the pantograph handle with the thumb. This makes a prick on the chart.

The patient can be shown instantly the disappearance of the test object when it enters a scotomatous area by placing it on the blind spot marked by a cross on the cloth. No time is thus lost in explanations to the patient, who at once grasps the procedure and realises what is to be observed.

This instrument is so rapid and accurate in use that it is possible to chart each patient in the clinical routine. Test objects of graded sizes, commencing at 1 mm. in diameter and coloured white, red and green are supplied (see Fig. 7).

An extremely fine method of investigating central scotomata is by means of Bjerrum's screen, which is a plain black cloth, 1 or 2 metres square, mounted on a spring roller. With this screen test objects of 1 or 2 millimetres can be used at a distance of 2 metres. Slight defects in the visual field can thus be detected which would not be made manifest by the use of the perimeter. The

foveal area is the most sensitive to light, but this sensitivity decreases unevenly as the periphery of the retina is approached.

Bearing this in mind a much greater degree of accuracy, when

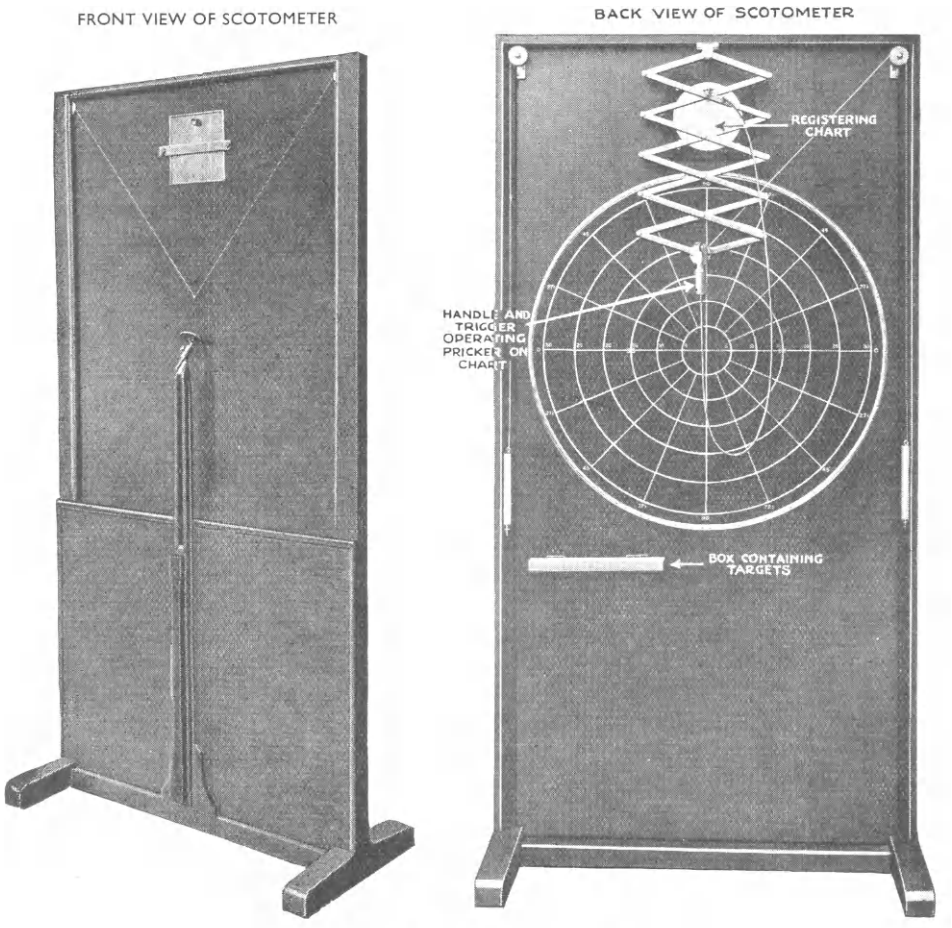


FIG. 7.—Armstrong's scotometer.

recording fields of vision, can be obtained by this method of quantitative perimetry than by ordinary perimetric examination with 5 or 10 mm. discs. Defects not detectable by ordinary methods and not even observed by the patient are clearly defined by the quantitative method as demonstrated by Bjerrum many years ago. Instead

of the perimeter arm a black screen is used and objects 1, 2, or 3 mm. in size are employed, which when viewed from a distance of 1 to 3 metres, a visual angle of one and a half to eight minutes is subtended, whereas by ordinary perimetric methods the angle is usually ten to fifteen times larger than this. By using a 1 mm. disc against the screen and varying the distance of the patient a large range of angles can be found and minute errors in the field of vision determined. The disc data is recorded as a fraction. If we take for example, the 1 mm. disc which is the most commonly used for 1-metre distance, the fraction is expressed as 1/1,000. Two metres or 2,000 mm. distance is also commonly employed with the 1-mm. disc ; then the fraction would be 1/2,000.

Traquair has shown that the field of vision may be conceived of as " an island of vision surrounded by a sea of blindness "—a remarkable analogy indeed. The surface of this island is uneven, the highest pinnacle representing the visual acuity of the fovea ; while at the periphery the surface becomes cliff-like, as here the visual acuity falls very rapidly. Surrounding the fovea is an area extending to 26 degrees, which is the normal field for a 1 millimetre object viewed 2 meters away, but outside this area the visual acuity again falls steeply. The blind spot would be represented by a deep hole extending down to the level of the horizon.

In many text-books the normal blind spot is referred to as the area cæca, while that portion of the retina between the area cæca and the fixation point including the latter is known as the centro-cæcal area.

Pitfalls.—During an ophthalmic examination one must never forget that there is a blind spot in every eye. It may easily be found. On a piece of writing paper mark a small cross No. 1, and on the right, 6 inches away from No. 1, mark a similar cross No. 2. If the paper is held 16 inches from the face cross No. 2 cannot be seen by the right eye, which is looking at cross No. 1, the left eye being covered ; the image of cross No. 2 has fallen on the optic disc. A patient who had already suffered from a detached retina suddenly discovered the blind spot in the other eye and came with fear and trembling, thinking that the retina of the healthy eye was also becoming detached.

A scotoma, if relative as in disseminated sclerosis, may easily be overlooked unless sought for by means of various coloured objects, red and green being the principal colours employed. The patient

should not be tired when his field of vision is being taken ; and from an hysterical patient it is almost impossible to obtain a consistent answer. The field of vision in the case of an hysterical patient varies from day to day.

An optic disc looks much whiter by contrast in a dark brown eye ; therefore, before taking the responsibility of declaring the presence of an optic atrophy, one should have had constant practice with the ophthalmoscope. Finally, in young children with hypermetropic eyes, the nerves proceeding to the optic disc from the retina show up so plainly that the mistake of calling such an appearance a papillœdema is not infrequent. Sometimes these fibres do not lose their medullary sheaths and show as a flame-shaped white patch resting on the edge of the disc (see Plate X). Students and medical practitioners should take the opportunity of examining cases of persistent medullated nerve fibres. These cases are most likely to be confused with papillœdema. They are to be differentiated by observing that in the former the vessels are not raised in the centre of the disc, and some portion of the edge of the disc is seen sharply defined, whereas in the latter the vessels in the centre of the disc are raised and the disc edge is blurred.

In patients suffering from severe anæmia or in those cases where there is a low hæmoglobin content of the blood the optic disc may appear quite pale ; also an overgrowth of glial fibres on the disc may simulate an optic atrophy.

The reader should reserve his judgment on any departure from the normal disc until his diagnosis has been confirmed by one whose daily work it is, both in hospital and private practice, to observe and understand what each small change seen by the ophthalmoscope signifies. He should examine as many normal eyes as possible, so that when the opportunity arises of examining an optic atrophy, papillœdema, etc., he will readily recognise the changed appearance of the optic disc.

If one eye appears more prominent than the other it should be ascertained how long this condition has been present. Remember also that retracted eyelids, as often seen in exophthalmic goitre, give the appearance of a slightly protruding eye. In *exophthalmic goitre* prominence of the eyes is not always equal ; on many occasions the writer has seen prominence of one eye only in the early stages of this disease. The previous appearance of the patient should always be inquired into from the patient himself or his

friends. Have the eyeballs always appeared prominent or have his friends noticed it but recently ? Has the squinting eye always squinted ? Has the patient always had a drooping eyelid ? Reference to old photographs is sometimes helpful.

With regard to the appearance of optic discs or pupillary changes, never attempt to examine such in an *artificial eye*, neither attempt to pull down the lower eyelid when an artificial eye is present. I have known of a physician who pulled down the lower eyelid, causing the artificial eye to fall out into his hand ; he did not realise such an eye was present and did not know that it is the lower lid which keeps an artificial eye in place.

If a patient is wearing glasses it would be well to inquire if strong cylinders are being used for the correction of astigmatism. If so, remember that on looking at the disc edge it may appear to be indistinct. Such a doubtful appearance will change as different lenses are rotated within the ophthalmoscope, the indistinct portion clearing up while that portion of the edge which originally was clear will now become blurred.

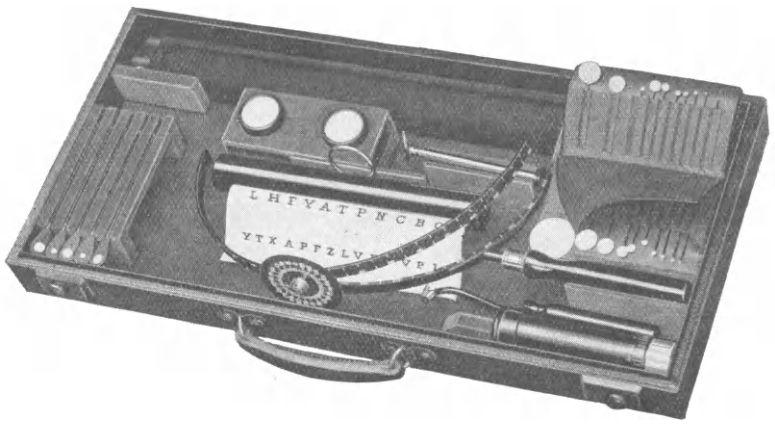


FIG. 7A.—Travelling case for use in neuro-ophthalmological examination. Containing test-type (window-blind pattern), axis-recording hand perimeter, Cardell optometer with two caps, set of Traquair's perimeter targets, small Bjerrum screen, in pigamoid-covered case. (Supplied by Finbars, London.)

CHAPTER II

THE PUPIL AND ITS REACTIONS

THE pupil is the central opening in the iris through which light passes to reach the retina. It acts in a similar manner to the stops of a camera. In order to obtain a clear image in dull light the pupil opens widely, and conversely, if the light is too abundant, the pupil becomes quite small.

In the correct formation of images on the retina it is necessary that both spherical and chromatic aberration should be absent. This is largely brought about by the contraction of the pupil.

The iris is a circular contractile diaphragm which at its circumference is continuous with the ciliary body. The anterior surface of the iris is striated in a radial direction. The markings appear more strongly in dark brown eyes. These striations seen through the slit-lamp show as cavities or crypts in which pigment and pigment cells lie. In an atrophic iris these depressions are seen to have lost the pigment, sometimes to such an extent that parts of the iris become quite transparent. Occasionally, in extreme forms of wasting disease, one may see with the ophthalmoscope in an undilated eye the red reflex of the pupil surrounded by red spokes of light, the transparent atrophic iris allowing the fundus reflex to be thus seen. The posterior surface of the iris lies immediately in front of the lens and normally is quite black in colour, due to being covered by the pars iridica retinæ.

The iris is seen through the transparent cornea and gives the tint to the eye, ranging from dark brown to light blue in different people. One iris may show different colours or one iris have a different colour from the other, both conditions being called *heterochromia iridis.*

The iris divides the space between the cornea and the lens into an anterior chamber and a posterior chamber, the two chambers communicating with each other through the pupil. The endothelial cells of the cornea lying on the membrane of Descemet are continued from its edge at the margin of the cornea over the front of the iris, but are absent over the crypts.

The stroma of the iris consists of cells and fibres of connective

tissue, the latter for the most part being directed radially towards the pupil. The colour of the eye depends partly on the pigment cells of the stroma, and partly on the pigment in the cells of the pars iridica retinæ. The muscular tissue of the iris consists of the sphincter pupillæ, which forms a ring round the pupil, and the dilator pupillæ, which radiates from the sphincter to the circumference, the latter being much less apparent than the former. The sphincter muscle is composed of bundles of plain muscle tissue. The dilator fibres form a continuous membrane close to the posterior surface. These fibres have the histological character of plain muscle cells but do not contain nuclei. In its development this membrane or layer never becomes vascularised nor does any mesoderm ever grow between it and the ectoderm. It thus differs from the sphincter by remaining in a more embryonic state. (Mann.) See Fig. 16A.

The long ciliary arteries, which are two in number, pierce the sclera a little to one side of the optic nerve and pass forward between the sclera and choroid until they reach the ciliary muscle. Behind the attachment of the iris each artery divides into an upper and lower branch, and these, anastomosing with the corresponding vessels on the opposite side and with the anterior ciliary arteries, form a vascular ring (circulus major). Within this is a smaller circle of anastomosing vessels (circulus minor) from which the capillaries spring.

The ciliary nerves, about fifteen in number, are derived from the ciliary ganglion and the nasal branch of the ophthalmic division of the fifth nerve ; they pierce the sclera posteriorly close to the entrance of the optic nerve and then pass forward in close contact with the choroid to the ciliary muscle ; within the ciliary muscle the nerves subdivide minutely and a few branches pass back into the choroid, but the greater number pass on into the iris. In the iris the nerves follow the course of the blood vessels.

It has been clearly proved by Langley and Anderson that the dilator fibres of the iris are stimulated by the cervical sympathetic. The sphincter muscle is supplied by the short ciliary nerves, while the dilator muscle of the iris is supplied by nerve fibres originating in nuclei situated near that part of the third nerve nucleus which supplies the sphincter fibres, their proximity explaining the reciprocal innervation of the two antagonistic sets of muscles. From these nuclei fibres pass down the cord as far as the eighth cervical and first thoracic ventral nerve roots, with which they leave the cord as

part of the white rami communicantes passing to the superior thoracic ganglion, and thence by the sympathetic chain to the superior cervical ganglion. The terminal nerve fibres for the dilator muscle pass by two distinct routes—first, from the superior cervical ganglion to the Gasserian ganglion of the fifth nerve, and then along the nasal branch of the first division, to pass with the two long ciliary nerves into the sclera and thence from the perichoroidal plexus to the dilator muscle. Other branches pass from the superior cervical ganglion to form plexuses on the various branches of the internal carotid artery; and from the cavernous plexus a fine branch passes to the ciliary ganglion which accompanies the short ciliary nerves.

Pressure by an aneurysm or from enlarged lymphatic glands in the thorax may cause a paralytic miosis or persistent contraction of the pupil. Paralytic mydriasis or a constant dilatation of the pupil follows paralysis of the minute branches of the short ciliary nerves which supply the constrictor pupillæ.

The following are the **principal reactions of the normal pupil** :—

(1) The direct light reflex.
(2) The consensual light reflex.

Also there are associated reflexes :—

(*a*) Reflex to accommodation.
(*b*) The orbicularis reflex.
(*c*) The oculo-sensory reflex.
(*d*) The psychical reflex.
(*e*) The spino-pupillary reflex.
(*f*) Reflexes associated with the ear.
(*g*) Reaction to drugs.

The direct light reflex is brought about by directing a beam of light into the eye. The reflex varies in amount according to the direction in which the beam of light is thrown. Using the indirect method of ophthalmoscopy, one can throw light on the surface of the optic disc, and this brings about a certain contraction of the pupil; but if the light is directed at the macula the contraction of the pupil is much greater.

If light falls upon the retina of one eye the pupil of the other is seen to contract. This is known as *the consensual light reflex.* It is the observation of these reactions that is so important in the diagnosis of diseases of the nervous system.

Of the *associated reflexes*, it must be borne in mind that when vision is directed to a near point accommodation takes place, and with this a narrowing of the pupillary aperture. It is the preservation of this reflex, but with the loss of the light reflex, that constitutes the Argyll Robertson pupillary reaction.

In sleep the pupils are always contracted (in hypnotic states frequently dilated). When the lids of one eye are closed the pupil on the same side contracts. This is called *the orbicularis reflex*, and is supposed to be caused by association with the movements of the eye upwards and inwards ; but, as Dr. Hall has pointed out, the eyes are not always directed upwards in sleep. We must therefore look in another direction for the explanation of this phenomenon.

The oculo-sensory reflex is brought about by sensory stimulation upon the eye or its adnexa.

The *psychical reflex* is seen in patients exhibiting extreme emotion or fear ; " the widely staring eye " of the terrified man is frequently mentioned in literature. Contracted pupils are found to accompany pain in the eye or in the extra-ocular tissue.

Small pupils which do not react to light are met with in certain cases. The writer once diagnosed Argyll Robertson pupils in a young woman but later found that she had suffered years previously from a severe injury to her spine through a fall. Her Wassermann reaction was completely negative.

Cochlear, vestibular and auro-sensory reflexes are produced by marked sensory stimuli, such as a tuning fork, catheterisation, etc.

Many *drugs* have the power of dilating the pupil ; such are atropine, hyoscine and homatropine. These are known as *mydriatics*. Others constrict the pupil, as eserine, opium, pilocarpine, etc., and are called *myotics*. The constricted pupil produced during the induction of anæsthesia is due not to the drug employed but to the excited condition of the central nervous system.

Cocaine when applied to the eye stimulates the sympathetic nerve endings in the dilator muscle of the iris. It does not paralyse the sphincter, so that only a moderate dilatation of the pupil is produced. Cocaine is employed in confirming the diagnosis of paralysis of the sympathetic nerve, for if this nerve be paralysed cocaine fails to produce dilatation of the pupil.

Dilatation of the pupils follows extreme exhaustion of the central nervous system or when the activity of all nerve centres is low, such as in alcoholic poisoning, or deep chloroform anæsthesia. In compres-

sion and severe concussion of the brain the dilated pupils produced are inactive to light. Hartridge mentions that in oxygen want dilated pupils form a characteristic sign.

The application of a mydriatic, especially atropine, to the eye should be made with due regard to the existing tension. In many elderly people the anterior chamber of the eye has become shallow through the closing of the angle between the cornea and the iris. In such eyes, if the pupils are dilated, the corneo-iridic angle becomes greatly lessened, so that there is a tendency for the tension of the eye to rise. After the application of homatropine an elderly patient should not be permitted to go away until a drop of $\frac{1}{2}$ per cent. eserine has been instilled into the conjunctival sac. If there is any doubt about the tension, and yet it is absolutely necessary to dilate the pupil, a second instillation of eserine should be made one hour after the application of homatropine. For the purposes of rapid dilatation homatropine and cocaine, 2 per cent. of each in a watery solution, should be used.

At this point a brief study should be made of the *afferent and efferent pupillary paths for light stimuli*. The afferent fibres are contained in the optic nerve. They undergo partial decussation in the chiasma like the visual fibres ; they then pass into the optic tracts, but do not pass into the external geniculate body. Their passage to the third nucleus is not definitely known (see Fig. 8). The constrictor centre is constantly sending out impulses which keep the pupil slightly contracted. It is owing to the afferent fibres forming a partial decussation in the chiasma that the consensual light reflex is obtainable. This reflex is brought about by throwing light into one eye and simultaneously observing the contraction of the pupil of the opposite side. If one observes the pupillary reflexes of a patient who has sustained an accident in which the optic nerve on one side is torn across, it will be seen that when light is thrown into the eye on the injured side there is no direct pupillary response, nor does the pupil of the opposite side show contraction ; but if light is directed into the eye on the uninjured side there will be a consensual light reflex observed in the eye on the side of the injured optic nerve. The reflex arc which brings about the consensual reaction is carried out by means of fibres which unite the two constrictor centres in the third nucleus.

In some of the lower mammals, where there is a complete decussation of the optic nerves, a consensual light reflex cannot be

elicited. It is in the course of these afferent fibres from the posterior end of the optic tract to the centres in the mid-brain that Parsons

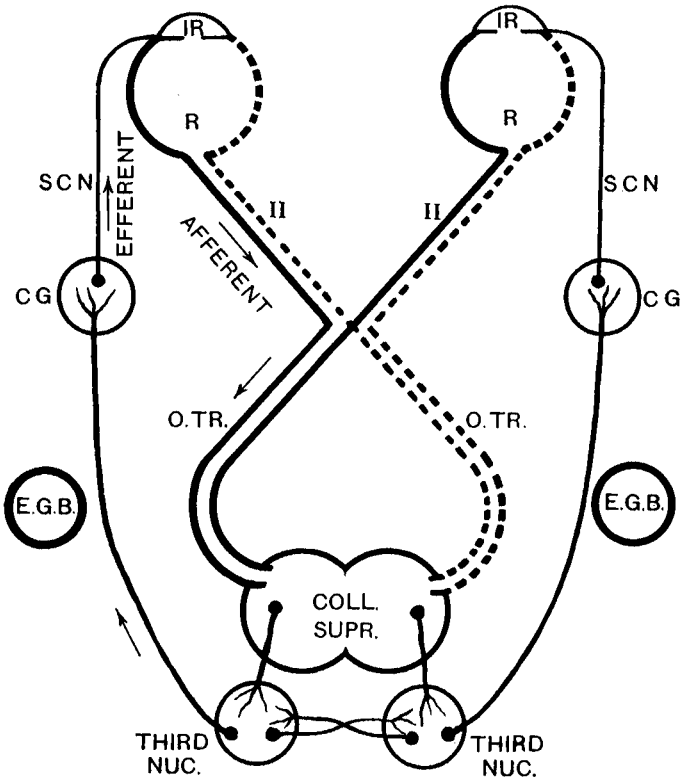


FIG. 8.—Diagram of the afferent and efferent pupillary paths for light stimuli. Afferent paths from left sides of retinæ, thick solid lines; afferent path from right sides of retinæ, thick dotted lines; efferent paths of left eye, thin solid lines; efferent paths of right eye, thin dotted lines; IR, iris; R, retina; II, optic nerve; O.TR., optic tract; COLL. SUPR., colliculus superior or anterior corpus quadrigeminum; THIRD NUC., nucleus of third nerve; E.G.B., external geniculate body; CG, ciliary ganglion; SCN, short ciliary nerves. (After Parsons.)

and others believe the lesion is situated which produces the Argyll Robertson pupil phenomenon.

Fig. 8 is a diagrammatic representation of the afferent and efferent pupillary paths of light stimuli. The end-organ for the pupillary stimulus is the neuro-epithelium of the retina, the same as for visual stimulus. This fact is established by observing that

in the photopic or light-adapted eye both the pupillary and visual responses are greatest from the foveal region and these responses vary *pari passu*, according to the degree of adaptation. The *afferent* pupillo-motor path is therefore contained in the optic nerve and these fibres like the visual fibres undergo partial decussation at the chiasma, from which they pass into the optic tract. They do not enter the geniculate bodies. The *efferent* (constrictor) path is by way of the oculo-motor nerve to the ciliary ganglion, and hence by means of the short ciliary nerves the fibres enter the eyeball and pass forward to reach the iris for the supply of the sphincter muscle. The following facts have been established both by experiment and pathological observation : (1) that section or avulsion of one optic nerve abolishes the direct light reaction but does not interfere with the consensual reaction due to stimulation of the opposite eye ; (2) that sagittal (antero-posterior) section of the chiasma abolishes neither direct nor consensual reaction ; (3) that section or destruction of one tract causes hemianopia with the hemianopic pupil reaction (Wernicke's) ; (4) that extirpation of the external geniculate body leaves the pupillary reactions unimpaired. From this we see that the afferent pupillary and visual fibres part company towards the posterior end of the optic tract.

The fate of the afferent fibres after they leave the optic tract has not been established. Many theories have been put forward, but it may be assumed that these fibres reach the pupillo-constrictor centre in the mid-brain. The exact position of this centre is unknown. The most probable hypothesis is that the fibres travel *via* the superior brachium to the superior colliculus ; there they are relayed passing to the third nerve nucleus. On sectioning the superior brachium, Karplus and Kreidl found that the light reflex was abolished. They also found in monkeys that on stimulating the tract or the brachium a pupillary contraction occurred, which was abolished on sectioning the brachium. Some authorities believe that the afferent fibres pass directly inwards to the optic tract, reaching the mesencephalic centre by way of the wall of the third ventricle. The importance of the exact knowledge of the course of the pupillo-motor path is understood when one realises that it must be in this vicinity such a lesion exists which results in the Argyll Robertson pupil. The work of Merrit and Moore seems to confirm the fact that such a lesion would have to be in the tegmentum ventral to the posterior commissure. v. Monakow believes that the

cells of origin of the constrictor fibres are scattered throughout the third nerve nucleus; this is supported by the clinical fact that frequently both ophthalmoplegia externa and interna occur combined as ophthalmoplegia totalis with comparatively localised lesions, indicating that the origins of the two sets of fibres are in close anatomical relationship.

Owing to the afferent pupillary fibres undergoing partial decussation in the chiasma, advantage is taken, clinically, of the use of *Wernicke's hemianopic pupil reaction*. This test is used for distinguishing a hemianopia due to a lesion in front of the mid-brain from one situated between this point and the visual cortex. There is no satisfactory means of carrying out this test, and observers differ in the value placed upon it, but many neurologists still put it to practical use. The method of testing this reaction is as follows: On throwing light on that half of the retina which corresponds to the blind half of the field of vision it is observed that the pupil fails to contract, but if the light is directed on the opposite half of the retina then the pupil will be seen to contract. This reaction indicates disease of the optic tract. It is extremely difficult to be sure that light which is directed on one half of the retina is not seen by any part of the other half, yet it should be remembered that a beam of light can be so directed on to the optic nerve that full stimulation to contraction will not occur as would be the case if a similar beam were directed in any other direction.

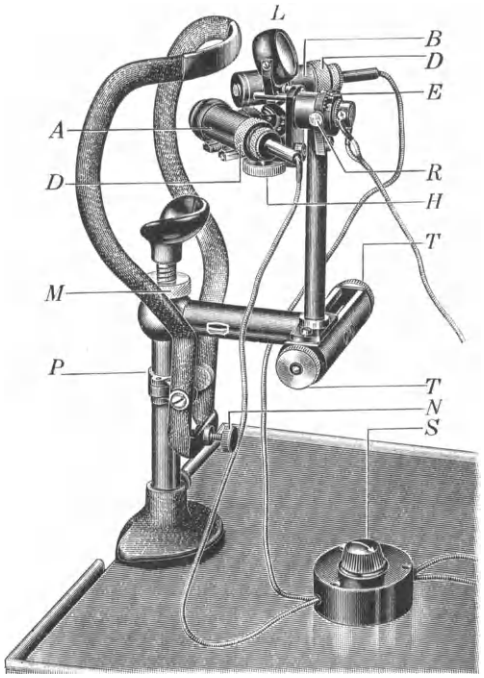


FIG. 9.—The Braun hemikinesimeter with chin and head rest. (Zeiss.)

A convenient method of testing for Wernicke's hemianopic pupil reaction is to use the fine sharp

slit-like focus of light formed by the removal of the "outer cap" or May condenser from the small May bulb of an ordinary electric ophthalmoscope. The sharpest focus of the slit light is obtained by a slight extension of the sliding focussing condenser tube which fits over the May lamp.

The hemikinesimeter (Fig. 9) invented by Dr. Braun at the instigation of Professor Elschnig provides an exact means of studying the hemianopic reaction of the pupil. This apparatus fulfils the conditions for exact investigation. There are two radiating pencils of

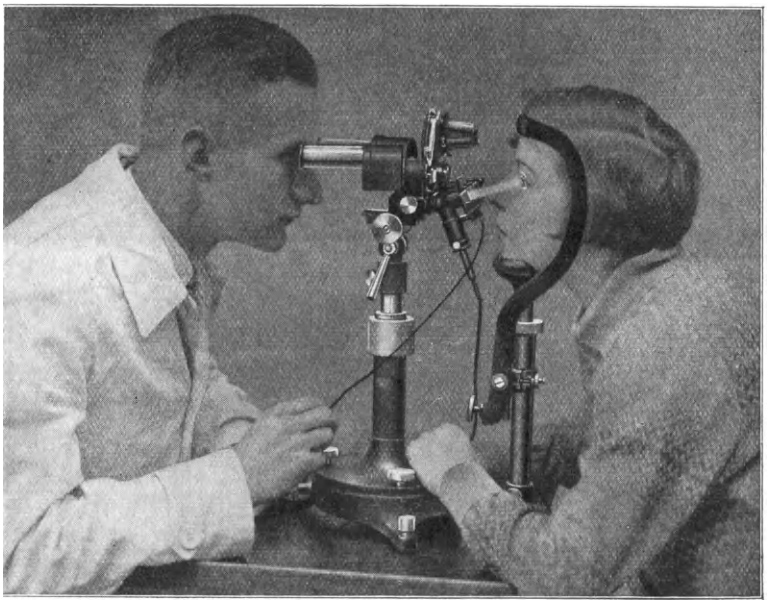


FIG. 10.

light of equal intensity; there are equal angles of incidence and equal distance of the two stimulating light sources with the shortest practicable interval of exposure to the light and with constant adaptation of the eye under examination. The scattering and reflection of the stimulating light on isolated peripheral elements of the retina is reduced to a minimum. The instrument is made by Zeiss.

The author is attempting to have the Sander supplementary pupilloscope (Fig. 10) and the hemikinesimeter combined in one instrument. The pupilloscope is used for determining the smallest

differences in the intensity of light which are just sufficient to induce a contraction of the pupil. The quotient of the two intensities necessary for the purpose expresses the motor sensibility to variation.

Another phenomenon associated with diseases of the nervous system is that known as *the paradoxical pupillary reaction* (Langendorff). This condition is seen in cases of general paralysis of the insane. The pupil is actually seen to dilate when a beam of light is thrown into the eye.

Hippus is the term applied to rhythmic contractions and dilations of the pupil. It has been noted in neurasthenia, epilepsy, hysteria and brain tumours. The pupil normally is in a state of constant movement, but hippus is a much more marked condition.

There is a condition of the pupil described as " cyclic contraction and dilatation of the pupil " which has been regarded as a congenital abnormality, but Greeves has demonstrated such a condition in a child who had just recovered from measles. In this case the pupil remained contracted for twenty seconds, and took twenty seconds to dilate and contract again, the whole cycle lasting forty seconds.

An important aid to the diagnosis of retrobulbar neuritis is the observance of a slowly dilating pupil while a continuous beam of light is cast upon the eye. In some syphilitic affections of the central nervous system there is a complete absence of all pupillary reflexes.

It should be remembered that paralysis of the sphincter pupillæ, together with that of the ciliary muscle, is termed *Ophthalmoplegia Interna*.

A blow on the eyeball may cause a paralytic dilatation of the pupil. A nurse while cleaning out a large syringe accidentally threw a column of water with some force against the eyeball. I observed the complete dilatation of her pupil for a period of over three weeks, after which time the pupil resumed its normal movements.

Raised intraocular pressure, as in glaucoma, will produce complete cessation of the movement of the pupil.

In complete paralysis of the third nerve the pupil is dilated, owing to the unopposed action of the sympathetic fibres.

If a pupil is seen to be very unevenly dilated, the student should not forget the possibility of the presence of a quiet irido-cyclitis. If a drop of homatropine is instilled into the eye, fine posterior synechiæ will be found binding down the edge of the iris to the

capsule of the lens. These must at once be ruptured, if possible, by the use of a mydriatic.

Tonic Pupils and Absent Tendon Reflexes.—There is a condition of the pupils, described by Adie, known as tonic pupils and absent tendon reflexes. It is characteristically unilateral and the pupil is almost always larger than its normal fellow. The condition is most commonly seen in young women. The pupil is never small, like the Argyll Robertson pupil. Examined rapidly, it appears neither to dilate nor contract, but if the patient is kept in a dark room for half an hour, and the pupils are then examined, they are found to be of equal size ; also on exposure to diffuse sunlight the pupil very slowly becomes normal again. While converging and accommodating the tonic pupil contracts slowly, but the contraction proceeds to such an extent that it becomes smaller than it normally should and the larger pupil is seen to become smaller than its fellow. The tonic pupil dilates easily and quickly to mydriatics such as atropine or homatropine. Foster Moore has described similar cases as " the non-luetic Argyll Robertson pupil," which have no relation whatever to syphilis, with knee jerks present and no evidence of nervous disease. But Adie states that loss of one or both ankle jerks is most frequent ; also loss of both knee and ankle jerks may be found. *Adie's syndrome*—tonic pupils and absent tendon reflexes—is much more likely to be discovered by the neurologist than by the ophthalmologist. This syndrome has never been observed in syphilis of the nervous system. The subjects are usually healthy young women whose serological tests prove to be completely negative. When compared with Argyll Robertson pupils the tonic pupil has little in common, *vide infra.* (Saenger, Markus, etc.)

Reflex Iridoplegia.—Cases apart from those described above are seen in which there is a sudden dilatation of one pupil, which remains so. In the case of one seen by the author the cause was obscure. Another case, that of a young man, was the result of a motor accident. The pupil would not contract to light or accommodation, but could be reduced in size by eserine. There were no other signs or symptoms of nervous disease, and accommodation was present.

Infrachiasmal, chiasmal and suprachiasmal lesions may produce reflex iridoplegia, while accommodation-convergence reflex is maintained, hence it has been loosely stated that the Argyll Robertson pupil is found in conditions other than syphilitic. Tonic pupils,

reflex iridoplegia and ophthalmoplegia interna (Hutchison) may be closely related, but several types stand out distinctly and must be looked upon as being quite separate from one another.

The Argyll Robertson Pupil is such a common phenomenon in diseases of the nervous system that it may be well to describe it at this point. Such a condition is demonstrated when light is thrown on the eye by means of the magnifying glass and the pupil fails to contract; but if light is still concentrated on the eye and the patient looks at a point 9 inches away, the pupil will now be seen to contract. There is then absence of reflex contraction to light but the presence of reflex contraction of the pupil to accommodation.

The best method of observing the Argyll Robertson pupil is as follows: If, in a darkened room and while directing the patient to look at a point 20 feet away (at this distance accommodation is not called into play), light is suddenly focussed on the eye by means of a lens or a small flash lamp, the pupil of a patient exhibiting this phenomenon fails to contract. While keeping the light focussed on the eye, if the patient is now asked to look at a finger tip 8 inches away the pupil will be seen to contract.

The Argyll Robertson pupil, therefore, consists of an absence or gross diminution of the reaction of the pupil to light, but the reaction to convergence and accommodation is preserved. Originally, it was described as being associated with tabetic cases. Typical pupillary reactions were described as the Argyll Robertson pupil in cases of spinal disease with contracted pupils (miosis). It is now a matter of common agreement not to restrict the term to such cases. Formerly the reaction was looked upon as specific evidence of syphilis of the nervous system, but many cases have been described that are not syphilitic, but which still show an Argyll Robertson pupil.

Wilson has enumerated such cases as follows:—

(1) Epidemic encephalitis.
(2) Disseminated sclerosis.
(3) Cerebral tumours in the vicinity of the third ventricle, aqueduct of Sylvius or anterior corpora quadrigemina.
(4) Syringomyelia or syringobulbia.
(5) Chronic alcoholism.
(6) Traumatic cases.

However, the minutest care should be taken to ascertain in the manner I have described if the Argyll Robertson pupil is present,

for I have noticed a tendency to describe this reaction in cases of contracted or unequally contracting pupils when such were not really typical of this reaction.

Several times I have examined the pupils of a lady who is now in her thirties. When she noticed that I was paying particular attention to the reaction of her pupils, she laughingly said : " Please don't call them Argyll Robertson pupils. They have always been small since birth and do not contract to light in a normal manner."

At the expense of repetition let the reaction be remembered as failure of the constrictor action of the pupil to the stimulus of light, while it is present to that of accommodation-convergence. Wilson says that clinical, experimental and pathological evidence combine to place the lesion on the afferent side of the light reflex arc, *i.e.*, anywhere up to the synapse of the pupillo-motor reflex fibres with pupillo-constrictor centre in the third nerve nucleus or its vicinity.

If no movement of the iris is seen by the naked eye in cases in which the Argyll Robertson pupil seems to be present, then a binocular magnifier should be used by the examiner, and thus minute movements of the iris can be detected. In a dark brown eye with a small pupil it is sometimes exceedingly difficult to tell if there is any contraction or dilatation of the pupil. The Argyll Robertson pupil is sometimes unilateral.

In the year 1869 there appeared a short paper in the *Edinburgh Medical Journal* entitled " Four Cases of Spinal Miosis, with Remarks on the Action of Light on the Pupil," by one named Argyll Robertson. He described how these pupils, small in size, in eyes with good vision—" sensitive retinæ "—contracted at once to the act of accommodation but remained immobile when light was directed on them. Such an unobtrusive paper has given to the medical world a sign of the greatest importance in syphilitic disease of the nervous system. Until quite recently the Argyll Robertson pupil was always associated with syphilis. Lately, many writers have apparently observed this sign in diseases other than that produced by syphilis, such as those already enumerated on p. 22, to which may be added cerebral hæmorrhage and thrombosis, arterio-sclerosis and senile dementias, diabetes and Friedreich's disease.

All four cases described by Argyll Robertson showed ptosis and symptoms of spinal disease.

Is it possible to distinguish the true Argyll Robertson pupil from

those which simulate it ? Contrast what is found in the true Argyll Robertson pupil with the tonic pupil and fixed pupil.

Argyll Robertson Pupil.	**Tonic Pupil.**	**Fixed Pupil.**
Miosis, generally bilateral and most commonly under 2·5 mm. in diameter.	Unilateral dilated pupil, generally over 2·5 mm. in diameter.	Unilateral dilated pupil.
Reacts slowly and unevenly to atropine or homatropine.	Reacts promptly to mydriatics and miotics (eserine).	Reacts promptly to miotics, but soon after dilates again.
Does not react to light either directly or consensually.	Reacts very slowly to light.	Does not react to light or accommodation.
Reacts briskly to accommodation - convergence.	Reacts very slowly to accommodation - convergence, the larger pupil becoming smaller than its fellow.	
Accommodation is instantaneous.	Accommodation is slow in returning to normal.	
Stroma of iris may show loss of pigment or actual atrophy.	Stroma of iris is normal in appearance.	Stroma of iris shows normal structure.

In the Argyll Robertson pupil, Adie, McGrath and others, including the author, agree that the consensual reflex to light is

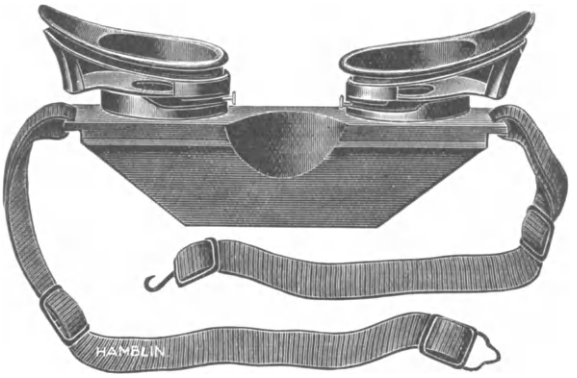


FIG. 11.—Zeiss' binocular corneal loupe.

absent as well as the direct reflex, but in the unilateral Argyll Robertson pupil, although both direct and consensual reflexes are

abolished on the affected side, the consensual reaction in the normal eye is obtained when the temporal portion of the retina of the affected eye is stimulated. If pupils are found in which reaction to light is lost, one has therefore many guides pointing out the true from the false. Observation should not be by naked vision alone, but a binocular magnifier, such as that supplied by Zeiss (Fig. 11), should be used to observe the action of the pupils and to examine the structure of the iris and shape of the pupillary aperture. Figs. 12, 13 and

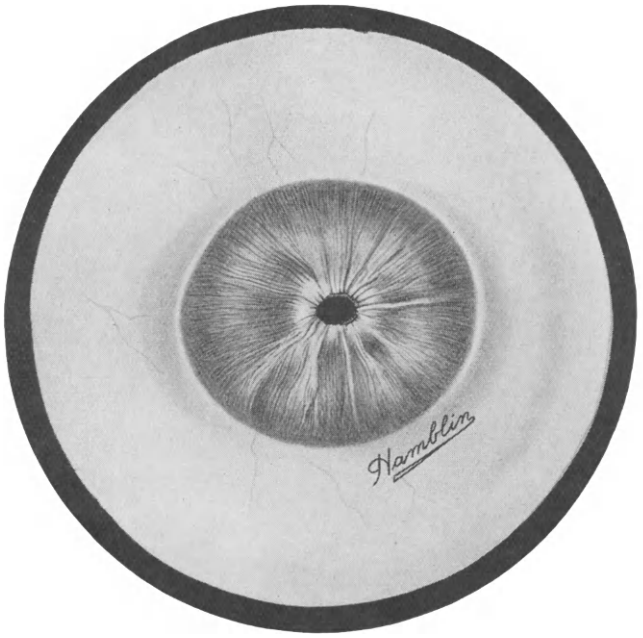


FIG. 12.—The iris in a case of advanced tabes. (McGrath.)

14, illustrating a paper recently published by W. M. McGrath, show very beautifully the condition found in many cases of true Argyll Robertson pupils. Note in Fig. 12 the stroma of the iris, in which the radial strands were abnormally thin, had lost their normal sinuosity, looking as if they had been combed out straight. The circulus minor and crypts had disappeared, while the pigmented posterior layer was irregularly visible, a true picture of a partially atrophic iris. Fig. 13 illustrates a case where atrophy had taken place only in two sectors, and here the pupillary edge was straightened

and uneven, remaining immobile on exposure to light, while the remainder of the iris contracted promptly with full amplitude to light —partial Argyll Robertson pupils. See also Figs. 14 and 15. These pictures of the right and left eye of a case of tabo-paresis, a woman aged fifty-four, show in both partial atrophy, the iris retaining its normal structure only in the temporal half. The nasal section reacts

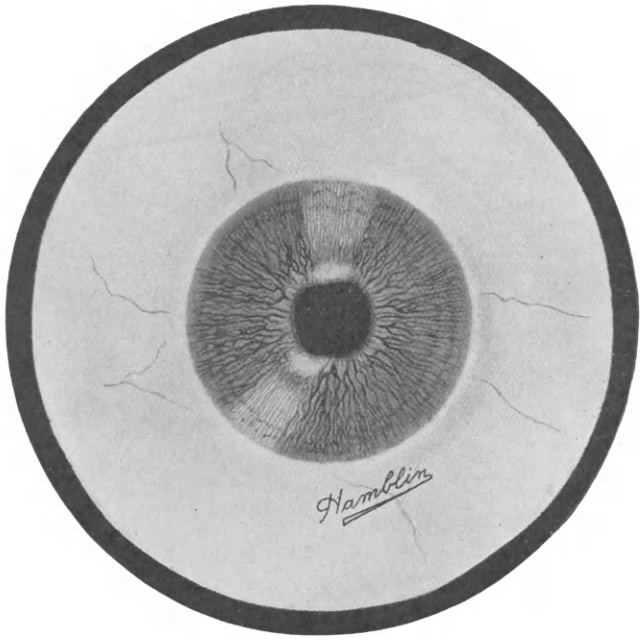


FIG. 13.—Partial Argyll Robertson Pupil. (McGrath.)

neither to light nor convergence, but in the temporal half a sluggish reaction on convergence remains.

Etiology.—McGrath most observantly notes that such irides exhibiting the Argyll Robertson phenomenon occur only in neuro-syphilis. I cannot, however, entirely agree with him regarding the atrophy of the iris. Elderly people often show such atrophy of the iris. Severely debilitated patients, as seen in a hospital for nervous diseases or in mental institutions, often show marked atrophy of the iris in such a manner that the retinal reflex to the mirror of an ophthalmoscope appears as a central red dot surrounded by faint

red lines like spokes of a wheel. Many of these cases show complete loss of pupillary reflexes. The author has seen cases of true Argyll Robertson pupils where neither pigmentary nor structural change of the iris could be observed. What does happen, however, is a decided atrophic change in an iris which has lost its innervation, either by long continued disease or the application of a mydriatic

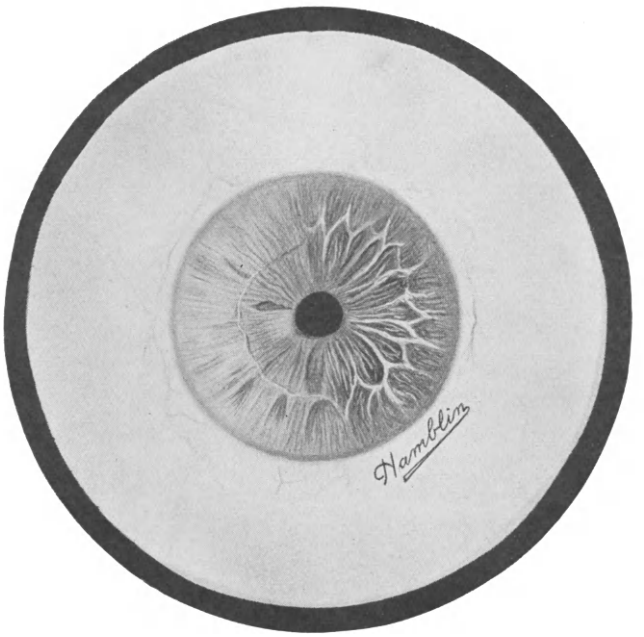


FIG. 14.—Partial atrophic (nasal side) iris. Left eye. (McGrath.)

or miotic for a considerable period. The latter is commonly seen in ophthalmic hospitals. McGrath, from his observations, argues that the lesion responsible for the Argyll Robertson pupil lies in the ciliary ganglion, noting that in Fig. 13 there is loss of sectorial contraction to light. He quotes Piltz, who found that stimulation of individual short ciliary nerves produced segmental contraction of the iris. Ferrier was of the same opinion.

As to whether there is a constant stimulus sent by the parasympathetic nerve supply which maintains the pupil in a condition of miosis, it is interesting to note that McGrath observed 15 cases of

miotic Argyll Robertson pupils immediately after death, and in all a wide dilatation of the pupils, remaining irregular in outline, was observed to occur within fifteen minutes.

Wilson, Edinger, Harris and others contend that the possible paths by which the light reflex passes are numerous, and are in the neighbourhood of the tegmentum, that the fountain-decussation of Meynert is one of the paths and that the lesion may be in the nerve

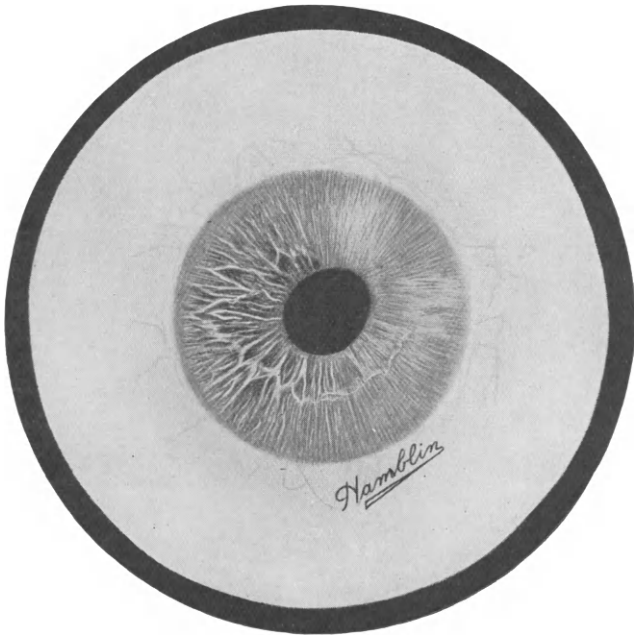


FIG. 15.—Partial atrophic (nasal side) iris. Right eye. (McGrath.)

fibres or cells. Reference to Cajal's drawing (see Fig. 16) of a coronal section of the tegmentum shows the complicated passage of the various nerve fibres, and at once indicates the difficulty of definitely assigning a particular function to certain structures. However, these writers are of the opinion that the lesion is to be found in the peri-aqueductal grey matter, where there is an interruption of conduction in the colliculo-nuclear fibres subserving the light reflex.

Paton and Mann " have adduced developmental evidence to show the relationship of the Edinger-Westphal nucleus with the

rest of the third nucleus and to show that its differentiation from the third nucleus synchronises with the development and functioning of the sphincter pupillæ musculature, both phylogenetically and ontogenetically." They state that the occurrence of the Argyll

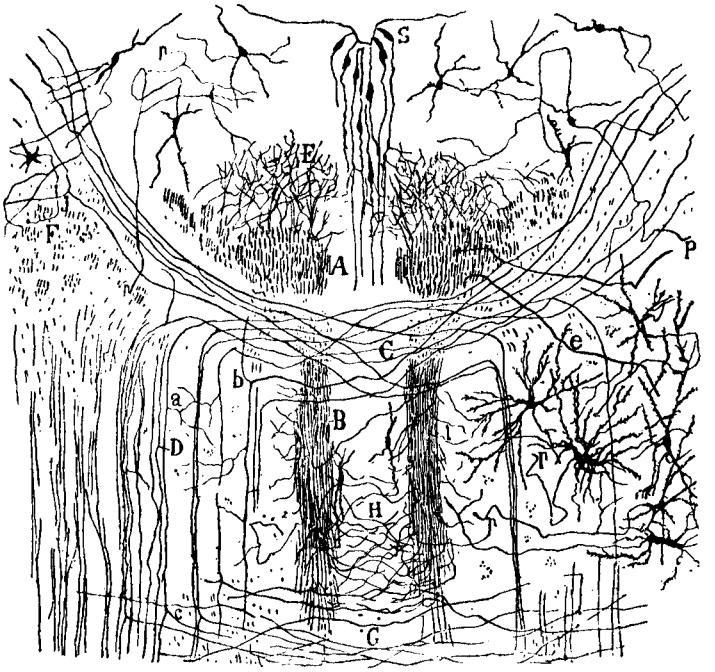


FIG. 16.—Coronal section of the tegmentum of a newly-born mouse. Golgi method. (Cajal.) A, dorsal longitudinal bundle; B, tegmental bundle of Gudden; C, Meynert's fountain-decussation, composed of fibres arising in the anterior corpora quadrigemina and passing into the ventral longitudinal bundle, D; E, collaterals of the fibres of the dorsal longitudinal bundle in the oculo-motor nucleus; F, longitudinal fibres of the tegmentum probably belonging to the main tract of the fillet; H, decussation of Forel, formed by the axons of cells of the red nucleus (some of its cells are seen on the right of the figure); p, axons of cells in the red nucleus and central grey matter; e, fibre passing from the tegmentum into the dorsal longitudinal bundle; S, ependyma-cells of the aqueduct.

Robertson pupil depends on a break in the reflex arc in that part consisting of the intercalated colliculo-nuclear neurons, and the nucleus of origin of the efferent irido-constrictor fibres.

It is generally accepted that the lesion responsible for the Argyll Robertson pupil is to be found in the afferent pupillary paths before they reach the constrictor centres.

Merritt and Moore have recently made a communication on the subject of the Argyll Robertson pupil. After reviewing recent anatomical and physiological studies, they have found an explanation for the Argyll Robertson pupil, quoting Beattie (*Proc. Roy. Soc.*, London, 106, 253, 1930, and *J. Anat.*, 66, 283, 1932), who showed that fibres running from this centre enter the anterior colliculus by passing ventrally to the posterior commissure. From there some of the fibres arch ventrally to enter the medial lateral portion of the midbrain and descend in the reticular formation of the pons and medulla. Others arch ventro-medially to run in the homolateral and contra-lateral posterior longitudinal bundles. They found that the pupillo-dilator fibres were uncrossed and were in connection only with the homolateral cervical sympathetic centre. In passing ventrally, therefore, these fibres are in close connection with the pupillo-motor fibres. Merritt and Moore have deduced from this that a single lesion can cause all the phenomena of the Argyll Robertson pupil. The lesion would have to be in the tegmentum ventral to the posterior commissure where the light reflex fibres are passing ventro-medially to the ocular motor nucleus and where the sympathetic fibres are on their way ventro-medially to the posterior longitudinal bundle and ventro-laterally to the reticular formation. They state that the Argyll Robertson pupil can be produced only by injury to both the afferent pathway and the sympathetic fibres, and must be at a point where the two pathways converge. They also quote Ranson ("The Pathway of the Pupillary Light Reflex," read at the Fifty-ninth Annual Meeting of the American Neurological Association, Washington, D.C., on May 9th, 1933), who found that constriction of the pupils was obtained when the brachium of the superior quadrigeminal bodies was stimulated and when the posterior commissure was stimulated. Except for a small tract in the pretectal region between the posterior commissure and the oculo-motor nucleus, where stimulation produces constriction of the homolateral pupil only, stimulation in the region of the anterior colliculus produced dilatation of the pupils. Ransom's work shows that the light reflex fibres which cross in the optic chiasm recross in the posterior commissure. Destruction of the pathway of the light reflex after these fibres have recrossed would cause unilateral Argyll Robertson pupils. They point out that the only other lesion which may cause this phenomenon is an infiltrating lesion such as a glioma of the anterior colliculus.

The Sympathetic and Parasympathetic Systems

In a later chapter reference will be made to the vegetative nervous system and its two great divisions. The dilator of the pupil is under the influence of the sympathetic system while the parasympathetic controls the sphincter of the pupil. Where these impulses to the iris originate is a question that is not yet settled. During the second half of the last century a prodigious amount of work was done in this direction. One has only to turn to the bibliography at the end of Parsons' article on the " Innervation of the Pupil " in the Royal London Ophthalmic Hospital Reports for 1904 to realise the interest then taken in elucidating the problems connected with pupillary movements.

Stimulation of the cortex cerebri, especially of those parts of the brain associated with eye movements, produces dilatation of the pupil, seen more readily in the monkey than in the cat or dog. (Parsons.) Paroxysmal movements of the iris (dilatation and contraction) were observed by Bianchi in two cases of prefrontal tumour, also Porot, Bardenat and Cor record the presence of unequally dilated pupils in the syndrome of frontal lobe tumours. The experimental data provided by Karplus and Kreidl show that pupillary dilatation may be elicited by cortical stimulation but that the efferent impulses are conveyed through the cervical sympathetic. Parsons, however, states that the dilator effect is diminished but by no means abolished by section of both sympathetic nerves in the neck. The diencephalon (hypothalamus and wall of third ventricle) also includes a centre through which reflex dilatation of the pupil takes place. This centre is located in the medial and rostral part of the subthalamic nucleus (Bard, Karplus and Kreidl), through which cortical impulses are mediated and are then conducted by the same pathways in the spinal cord as are those impulses arising from direct stimulation of the diencephalic centre. If the mesencephalon is sectioned pupillary dilatation can still be elicited by physiological stimuli showing that the sympathetic effect on the pupil is not wholly dependent on the functional integrity of centres above the level of the mid-brain (Keller). The sympathetic path is continued downwards in the lateral part of the pons, medulla and spinal cord as far as the inferior cilio-spinal centre of Budge, which is situated in the lower cervical and upper dorsal region of the cord. White rami communicantes leave the intermediolateral tract of the cord by the anterior roots of the seventh and eighth cervical and

first, second and third thoracic nerves. Cardozo has demonstrated that most of the preganglionic fibres involved in the innervation of the dilator muscle of the iris are found in the second thoracic nerve, but Gask and Ross operating on the human subject found that when they merely divided the sympathetic trunk below the stellate ganglion the eye fibres escaped injury, which shows that these fibres pass out from the cord in the white ramus from the first thoracic segment. The rami from the thoracic segments pass into their respective ganglia. The upper thoracic ganglion is fused frequently with the inferior cervical ganglion and is then known as the *stellate* ganglion. (In the dog and cat, however, the stellate ganglion corresponds to the united lower cervical and upper three or four thoracic ganglia of man.—Krause, Quain, Vol. 3.) The fibres pass to the inferior cervical ganglion which is connected to the middle cervical ganglion by the ansa subclavia (Vieussenii). Both anterior and posterior loops round the subclavian artery convey fibres upwards to the middle cervical ganglion and then to the superior cervical ganglion, round the cells of which they terminate. From the superior cervical ganglion the dilator impulses pass by way of fresh fibres constituting the nervus caroticus internus which accompany the internal carotid artery through the carotid canal into the skull. From this nerve two plexuses are formed, the internal carotid plexus lying on the lateral side of the artery and the cavernous plexus lying medial and inferior to the artery as it traverses the cavernous sinus. Many of the filaments arising from these plexuses pass around the circle of Willis and communicate with the opposite side. The filaments which enter the orbit are mainly from the cavernous plexus and pass by way of the third nerve, joining it at its division, to the sixth as it crosses the internal carotid artery in the cavernous sinus, to the Gasserian ganglion and the ophthalmic division of the fifth nerve and also a twig which is the sympathetic root of the ciliary ganglion. Small filaments are found on the ophthalmic artery and its branches. In addition to dilator effects, vasomotor impulses pass by way of the deep petrosal nerve and Vidian nerve of the pterygoid canal to the sphenopalatine ganglion (see Fig. 27) and then through the inferior orbital fissure. This latter path is the way of supply to the smooth muscle in the orbit. A sympathetic twig passes with the lacrimal nerve to the lacrimal gland.

During recent years both Dieters and Zernick confirmed what

was discovered as early as 1878 by François-Frank, that sympathetic oculo-pupillary fibres pass through the middle ear in man as well as in lower animals. They found the course of the sympathetic nerves to deviate from the internal carotid artery into the middle ear with the carotico-tympanic fibres. From the middle ear they pass through the base of the cranium lateral to the nerve of the pterygoid canal and become associated with the cavernous plexus. The majority of the sympathetic fibres do not actually pass through the ciliary ganglion. Some reach the eye through the long ciliary nerves while others traverse the sympathetic root of the ciliary ganglion and enter the short ciliary nerves distal to the ganglion. None effect synaptic connection with the neurons in the ciliary ganglion. The long ciliary nerves include two groups of fibres, viz., those which pass through the nasociliary nerve for the sensory innervation of the eye and those from the plexus on the ophthalmic artery. The latter are responsible for the innervation of the dilator pupillary muscle (Kuntz).

When the cervical sympathetic cord is divided the pupil contracts. There is also a fall in the intraocular pressue due to section of the vaso-constrictor fibres, but this slowly recovers as the vessels regain their inherent tonus. Karplus and Kreidl found that unilateral stimulation of the cervical cord elicited bilateral pupillary dilatation but this does not dispose of the existence of a cilio-spinal centre in the upper dorsal region of the cord.

The Parasympathetic System

The light reflex brings into play the parasympathetic innervation of the sphincter muscle of the eye. Light stimulation begins in the neuro-epithelium of the retina. From the retina afferent fibres pass through the optic nerve, the chiasma (where partial decussation takes place), the optic tract, the superior brachium to enter the superior colliculus. Here it is assumed by most authorities the fibres end and fresh fibres convey impulses through the fountain decussation (Fig. 16) to enter the oculo-motor nucleus. The path for the control of the constrictor pupillæ is therefore the afferent fibres in the optic nerve, tract, superior brachium, pretectal region, posterior commissure, fibres arching ventrally round the central grey matter at the upper end of the cerebral aqueduct (Karplus and Kreidl, Ransom and Magoun). Stimulation of the superior colliculus itself is not followed by constriction of the pupil. The efferent paths

are by way of the preganglionic fibres of the oculo-motor nerves and the post-ganglionic fibres arising in the ciliary ganglion.

It is believed that the function of pupillary movements and of accommodation are situated in the Edinger-Westphal nucleus lying anteriorly and medial to the lateral mass of the oculo-motor nucleus (see Fig. 25). When the amount of light entering the eye is suddenly increased the pupil contracts. A contraction also takes place when accommodation is brought into play. Observe this phenomenon in the eye of a young person and note that the same full action does not take place in an elderly subject due to local changes in the structure of the iris. The proximity of the cells in the oculo-motor nucleus governing accommodation and pupillary movements accounts for the reciprocal innervation of the two antagonistic sets of muscles. Stimulation of the oculo-motor nerves brings about contraction of the pupil through constriction of the sphincter muscle. Division of the oculo-motor nerve causes a moderate dilatation of the pupil due to removal of the tonus of the sphincter pupillæ, which effect is increased by cutting the short ciliary nerves or removal of the ciliary ganglion. Sympathetic stimulation after division of the third nerve brings about a further dilatation by activating the dilator muscle. Injury to either the afferent or the efferent path practically abolishes the constrictor reflex. Parsons divided the oculo-motor nerve after previous division of the cervical sympathetic cord, which caused the pupil to become immobile. Stimulation of any part of the cortex had then no effect on the pupil.

If the eye is directed to a point close at hand the ciliary muscle contracts, allowing the lens suspended by the zonule of Zinn to swell; the lens, becoming more convex, focusses light accurately on the retina. At the same time the pupil contracts, a reflex designed to cut off the rays from the peripheral part of the lens, so lessening the effects of spherical aberration. The constriction of the pupil also increases the depth of focus (Starling). The act of convergence is accompanied by constriction of the pupil and at the same time accommodation is brought into play, but these three actions, all under the influence of the oculo-motor nerve and their functions being closely related, are capable of functioning independently. The eye accommodated for distance in a bright light will show constriction of the pupil, while an object examined close at hand in a dim light is seen through a dilated pupil. Again, one may observe that not only on converging to a near point by means of the contraction

of the two internal recti the pupil diminishes in size but also on looking laterally at a near point by using the internal rectus of one eye and the external rectus of its fellow, constriction of the pupil takes place at the same time. A young person can exert accommodation at will, thereby making himself artificially myopic without bringing into play the external ocular muscles.

Examination of the pupil should include its size, shape, colour and position, both as regards lateral and antero-posterior displacement. In the normal state the pupil is often displaced slightly to the inner side and below the horizontal meridian ; in certain conditions of nervous disease the iris does not contract equally (*vide supra*). The pupil normally is largest in adolescence, but becomes smaller as age advances. The explanation, according to Fuchs, is that there is a gradual thickening of the connective tissue on the surface of the sphincter pupillæ. Also there is less elasticity of the iris due to increase of connective tissue in the walls of the blood vessels ; this is most marked in arterio-sclerotic conditions.

Anisocoria is a condition where there is inequality of the size of the pupils. Examining the eyes of children in the wards of a hospital reveals the fact that anisocoria is not at all uncommon. It occurs, according to Fuchs, in about 10 per cent. of normal cases. If one eye is myopic the pupil of this eye is usually larger than its fellow. In glaucoma the pupil is enlarged and somewhat oval and is immobile. In iritis, without any evidence of inflammation, the pupillary margin may become bound down by posterior synechiæ to the anterior surface of the lens. I have seen such adhesions placed so evenly at intervals that the physician in charge of the case asked if I knew of any disease or condition which would produce such contractions resulting in a crenated pupillary outline. One has but to instil a mydriatic to show the brown pigmented adhesions at the points of attachment. If the condition of anisocoria is present in an eye and is not associated with disease, the pupillary reactions will take place in the same proportion in each eye on the instillation of various drugs in the conjunctival sac, while the pupillary aperture maintains its circular outline.

The general mechanism of anisocoria, according to Byrne—apart from cases involving corneal scars, refraction, accidental instillation of some drug—involves the enhancement or impairment of function in some portion or portions of the mechanism which mediates (*a*) reflex dilatation, (*b*) inherent dilator tonus, (*c*) chemical

effector tonus, (*d*) reflex constriction, (*e*) inherent constrictor tonus, (*f*) chemical constrictor tonus.

One has observed anisocoria (just as Morax and others have done) when sudden dilatation of one pupil has taken place without any ocular lesion or any gross nervous lesion. Anisocoria may be observed in intracranial tumours, cerebral hæmorrhage, in meningitis either central or spinal, also in all those conditions which are associated with Horner's syndrome (*vide infra*). A most practical point, however, should never be forgotten when a case presents a unilateral dilated and immobile pupil—the intraocular tension should be examined, for glaucoma often appears in a most insidious fashion.

When at a loss to know which pupil is affected pathologically in anisocoria, *i.e.*, if the right is contracted, is it abnormally contracted or is the left abnormally dilated ? It should be borne in mind that when the lesion is confined to the efferent dilator or constrictor path the ensuing dilatation or contraction is confined to one pupil, leaving the other pupil unaffected. When, however, the lesion involves the afferent dilator or afferent constrictor path, the pupillary effector mechanisms in both eyes are affected (Byrne).

The paradoxical pupillary reaction was first observed by Budge and, later, by Langendorf, who described the phenomenon by this name. Piltz has described the paradoxical accommodation reaction which he observed in functional disorders of the nervous system where widening of the pupil occurs when the accommodation is adjusted for the near point and *vice versâ*. He has also described the true paradoxical light reactions of the pupil, namely, dilatation of the pupil in response to increased amount of light entering the eye. Usually, when the sympathetic innervation of the pupil is interrupted there is a miosis or contraction of the pupil, but Budge noticed that if the superior cervical ganglion were removed or the cervical sympathetic cord cut, the pupil on the same side under such conditions as asphyxia, excitement, intravenous injection of adrenalin or anæsthesia became larger than its fellow and widening of the palpebral fissure, together with some proptosis, takes place. Byrne showed that after a lesion of the sciatic nerve or of the brachial plexus, paradoxical pupillary dilatation could be readily demonstrated ; such pupillary movement can only be elicited by an intravenous injection of adrenalin twenty-four hours after removal of the superior cervical ganglion or as long as twelve to fourteen days after injury to one sciatic nerve. While observing such paradoxical

pupillary movement Byrne noticed the presence of a pseudo-paradoxical pupil dilatation which occurred mainly during the pre- and post-paradoxical period and is closely associated with pain and tenderness. " Looking at a patient's face it is an easy matter to determine whether he has pain or not, and if in pain from his injury the contralateral pupil is larger than its fellow if the injury be in the leg, whereas the homolateral is the larger of the two pupils if the injury be in the arm (positive phase of the pseudo-paradoxical phenomenon)."

Humoral Transmission of the Nerve Impulse

There has been a good deal of work done recently to show that humoral transmission of the nerve impulse in the autonomic nervous system is by means of a chemical transmitter—acetylcholin or sympathin (Cannon), hence the autonomic nerves are divided into cholinergic or adrenergic accordingly. Acetylcholin is liberated at the parasympathetic endings on stimulation of the system. This substance is unstable in alkaline solution but relatively stable in acid. It is inactivated by the esterase present in blood and protected from this enzyme by a small amount of eserine ; it is a prompt vasodilator and has been used recently with success in embolism of the retinal artery. Acetylcholin stimulates the parasympathetic nerve endings, an effect that is abolished by atropin. Duke-Elder discovered that acetylcholin produced contraction of the extrinsic muscles of the eyes, which action differentiates them from other skeletal muscles of the mammalia and resembles them only when they are deprived of their nerve supply. (See page 424.)

Acetylcholin is present in the iris and ciliary body of rabbits and is markedly increased after stimulation of the oculomotor nerve. It breaks down rapidly in the aqueous humour but is protected by eserin (Englehart). The stimulation to contraction of the iris by parasympathetic supply is abolished by the use of atropin, hence the pupillary dilatation which follows its instillation. Injection of acetylcholin into the ventricles of the brain gives results which seem to suggest a liberation of this substance at the various synapses (Henderson and Wilson). Dale suggests that acetylcholin acts as a sensitiser for the effector cells (shortens the chronaxie of the nerve cell or motor end plate so that it is momentarily attuned to the nerve itself).

For the action of atropin and physostigmine on the pupil, see Chapter X.

The action of such a drug as homatropin which dilates the pupil is much less effective in an eye which has become accustomed to tropical conditions of light. It may require two or three applications of an oily solution of homatropin to dilate the pupil of an Indian patient, whereas a drop of the watery solution of the same 2 per cent. strength will dilate the light-coloured pupil of the European in fifteen minutes. Also a brown pupil reacts more slowly to homatropin than does a grey or a blue one.

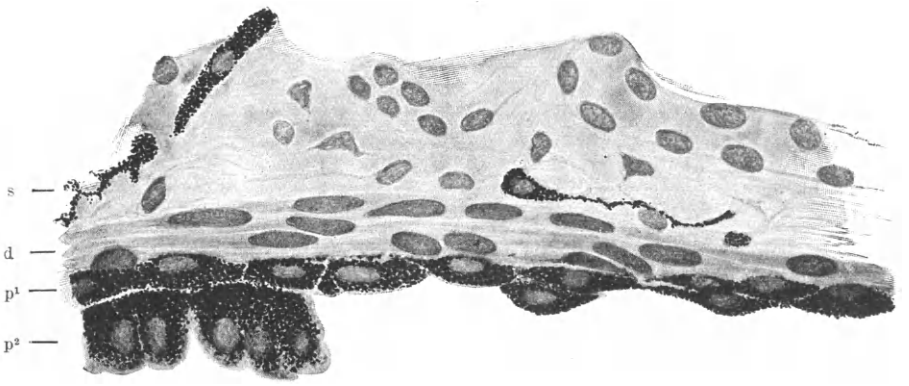


FIG. 16A. Section of posterior layers of iris, human, near its attachment to the choroid. (E. A. Schäfer from Quain, Vol. iii, Pt. 2.) s, iris stroma with connective tissue, branched pigment-cells and blood-vessels; d, layer of dilatator pupillæ; p^1, deeper layer of flattened pigment-cells appearing spindle-shaped in section; p^2, superficial layer of pigment cells of columnar shape. These cells are broken away from the larger part of the section.

In the frog and eel the iris musculature is directly influenced by light; in the human, when deprived of its nerve supply, the same phenomenon occurs. (See Chapter XI.)

CHAPTER III

THE MUSCLES AND NERVES OF THE EYE AND ITS ADNEXA

THE movements of the eyeball are produced by the actions of the *four recti muscles*—superior, inferior, internal and external; and the *two oblique muscles*—superior and inferior.

The superior rectus arises from the upper margin of the optic foramen and, passing forward above the optic nerve, ends in a thin delicate expanding tendon on the upper and anterior aspect of the

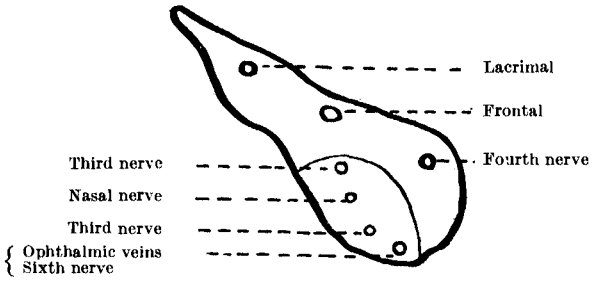


FIG. 17.—Left—Superior orbital fissure (Diagrammatic).

eyeball. It is supplied by the superior division of the third cranial or oculo-motor nerve. The inferior rectus takes origin from a fibrous band which bounds the inner extremity of the sphenoidal fissure. It is supplied by the inferior division of the third nerve. The internal rectus springs from the inner side of the optic foramen and is supplied by the inferior division of the third nerve, while the external rectus arises from a fibrous arch, the extremities of which form its two heads of origin. Between these two heads emerge the superior and inferior branches of the oculo-motor nerve, with the nasal branch of the ophthalmic division of the fifth lying between them. The sixth or abducent nerve also passes between the two heads of the external rectus and, with the ophthalmic veins, occupies the lowest position (see Fig. 17). The external rectus muscle is supplied by the sixth or abducent nerve.

The superior oblique muscle, arising from the roof of the orbit immediately in front of the upper and inner part of the optic fora-

men, extends forwards above the internal rectus. It ends in a slender tendon which passes through a pulley and then proceeds backwards and outwards to be attached to the eyeball. It is supplied by the fourth cranial nerve—the trochlear. The *inferior oblique muscle* arises from the orbital plate of the superior maxillary bone. It passes outwards below the inferior rectus muscle to gain insertion

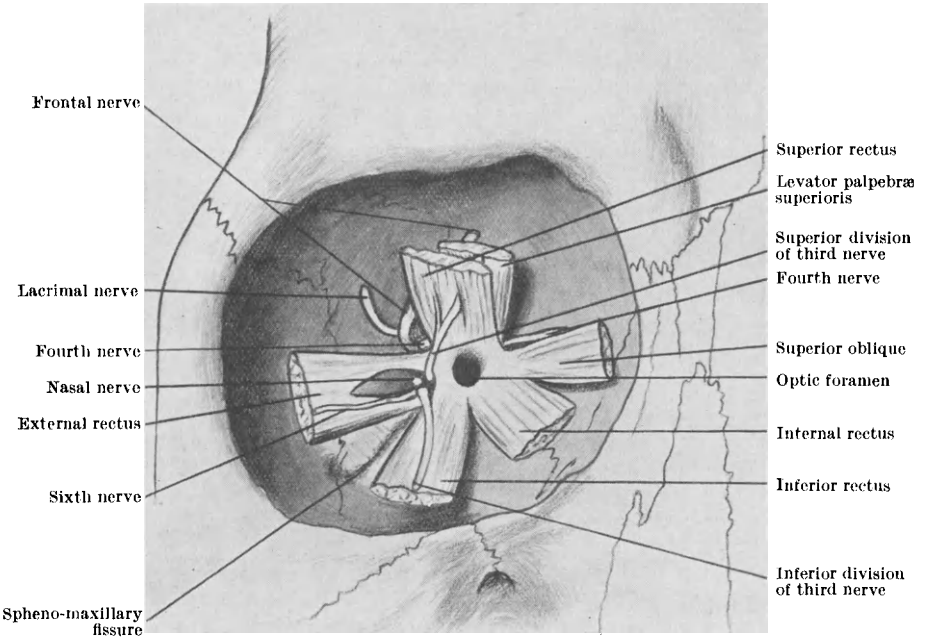


FIG. 18.—Diagram of the origin of the ocular muscles in relation to the optic foramen and the sphenoidal fissure, together with the relations of the various nerves entering the orbit. (After Cunningham.)

into the sclera of the eyeball under cover of the external rectus. It is supplied by the inferior division of the oculo-motor nerve.

Fig. 18 illustrates the origin of these muscles and shows the relationships of the nerves passing through the superior orbital fissure. Fig. 19 is an illustration of a dissection of the orbit and the middle cranial fossa. From the position and direction of the eyeball lying in the orbit it is seen that the antero-posterior axis of the orbit and the visual axis of the eye do not point in the same direction. The shape of the field of vision is also modified by the surrounding bony structure of the orbit.

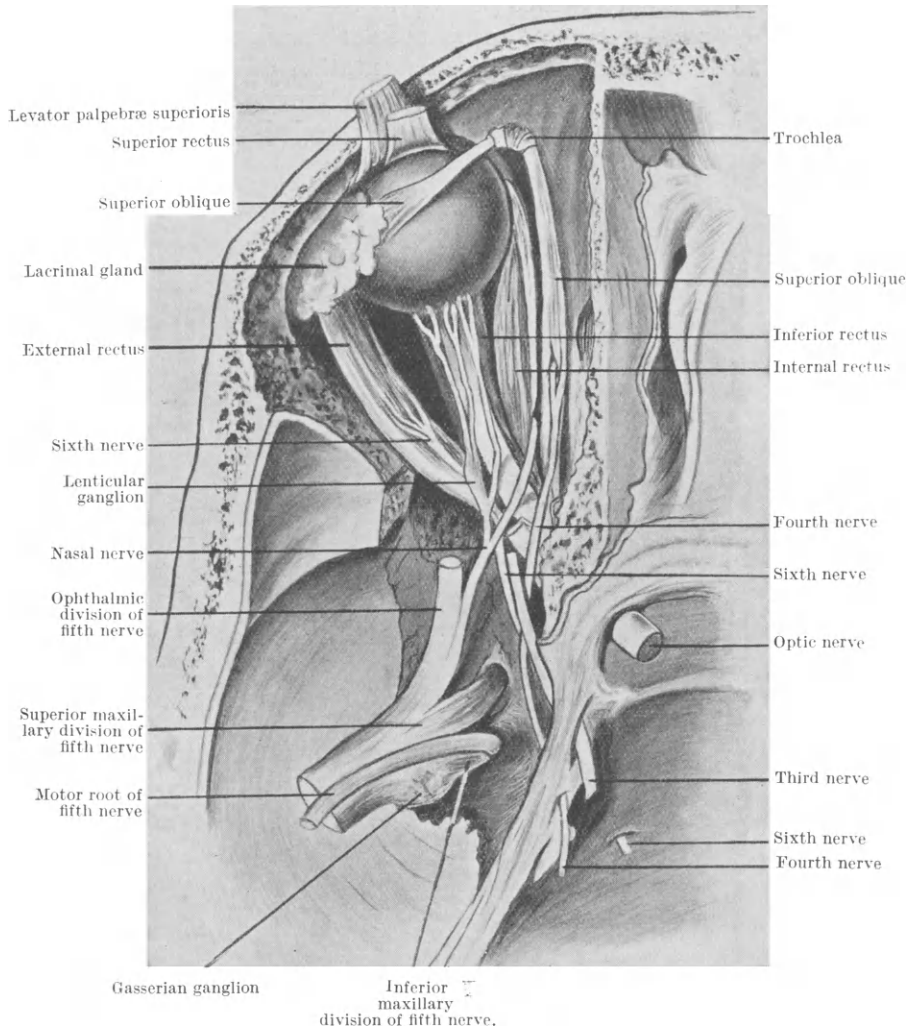


FIG. 19.—Dissection of the orbit and the middle cranial fossa. Both roots of the fifth nerve with Gasserian ganglion are turned outwards. (After Cunningham.)

In this illustration (see Fig. 19) both muscles, levator palpebræ superioris and the superior rectus, are reflected forwards exposing the tendinous attachment to the eyeball of the superior oblique muscle. A portion of the lacrimal gland has also been uncovered,

while the long ciliary nerves passing to the eyeball are seen lying within the cone formed by the extraocular muscles.

Fig. 20 is a diagrammatic representation of the extraocular muscles. A glance at this will indicate the movement produced by each muscle. The *superior rectus* turns the eyeball upwards and to a

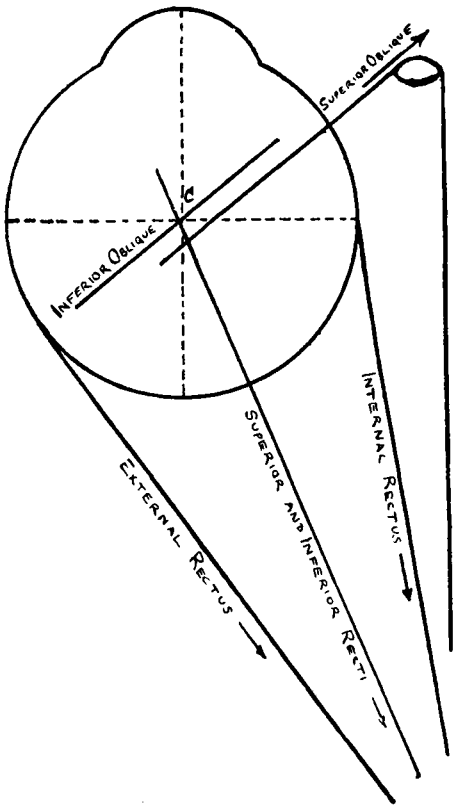


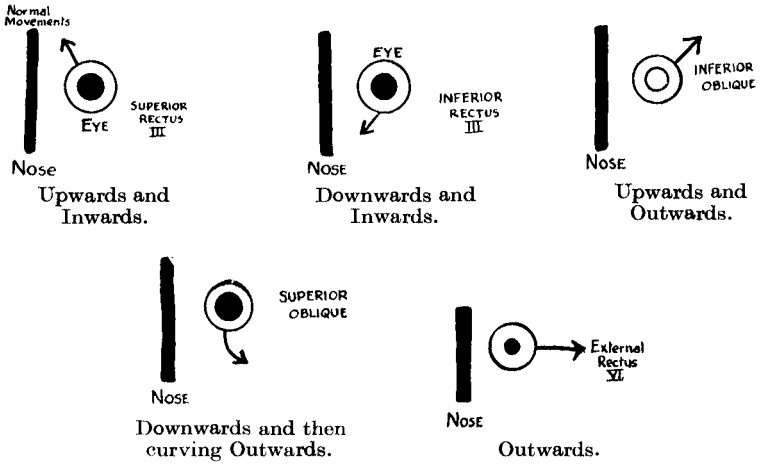
FIG. 20.—Diagram of left eyeball with attached muscles.

lesser extent inwards. The *inferior rectus* turns the eyeball downwards and inwards. The *internal rectus* turns the eyeball inwards, while the *external rectus* turns the eyeball outwards. The *superior oblique* muscle by its action rotates the eyeball downwards and outwards. The *inferior oblique* rotates the eyeball upwards and outwards. By the action of these muscles a perfect balance of both eyes is maintained so that the retinal images being accurately

superimposed are fused by the brain, true binocular vision resulting. The *superior rectus and inferior oblique* acting simultaneously turn the eyeball directly upwards. Each movement of the eyeball is a true synkinesis, the contraction of one set of muscles being accompanied by the relaxation of their antagonists.

When the head and body are erect and the eyes directed to the horizon straight in front, the visual axes thus being assumed to be parallel, the eye is said to be in the primary position. When the visual axis is directed elsewhere from its primary position it is said that the eye has assumed a secondary position. Listing's law may

NORMAL MOVEMENTS OF EYE MUSCLES



The Internal Rectus Muscle produces the exact opposite movement to the External Rectus, *i.e.*, eye turned directly *inwards*.

be stated thus : Any movement from a primary to a secondary position is one in which the eye rotates round an axis which lies in a plane vertical to the visual axis and passing through the centre of rotation of the eyeball. The vertical axis and the horizontal axis run vertically and horizontally in this plane. If we look upon the centre of rotation as a fixed point, a line passing through the macula and this point is known as the line of fixation, and is therefore sagittal to Listing's plane in the primary position. When the eyes look upwards they rotate round the horizontal axis, when looking from side to side the movement is around the vertical axis. A movement of rotation around the sagittal or perpendicular axis is

not possible, but when the eyes are directed obliquely away from the primary position there is a slight amount of torsion or rolling. We are all familiar with the altered position of after-images when the eyes are turned obliquely away from the primary position; for example, on looking at a red cross on a white wall, if the eyes are turned obliquely upwards the after-image of the cross is seen slightly rotated.

For a discussion on the laws of Donders, Tscherning, Helmholz and Listing, the reader is referred to the various works on practical optics, or for a simpler explanation to those by Maddox, Peter and Duke-Elder on the ocular muscles.

The movement of the eyes may be simply described as straight and oblique.

STRAIGHT MOVEMENTS

To nasal side	Internal rectus.
To temporal side	External rectus.
Upwards	Superior rectus and inferior oblique.
Downwards	Inferior rectus and superior oblique.

OBLIQUE MOVEMENTS

Upwards and to nasal side	Superior rectus, internal rectus and inferior oblique.
Downwards and to nasal side	Inferior rectus, internal rectus and superior oblique.
Upwards and to temporal side	Superior rectus, external rectus and inferior oblique.
Downwards and to temporal side	Inferior rectus, external rectus and superior obliquc.

Fig. 21 illustrates the manner in which the extra ocular muscles are inserted into the eyeball. The position and direction of these muscles are clearly indicated in Figs. 22 and 23, also the position of the eyes in relation to the orbital walls, nasal cavities and brain.

The muscles of the eyes are so beautifully adjusted that both eyes move equally, while no matter how fast they move there is no diplopia perceived. By careful measurement on a protractor the radius of which is equal to the diameter of the eyeball, and knowing the amount of imbalance present as measured by the Maddox rod, one can measure the distance the muscle should be advanced in order that single vision be exactly obtained, *e.g.*, if hyperphoria is present (one eye tending to have its visual axis directed higher

than its fellow) the superior rectus of the drooping eye can be accurately advanced to a new position in order to correct the error.

In some cases of squint and nystagmus it has been stated that these conditions have been due to disturbance of the proprioceptive

DIAGRAM SHOWING THE LINES OF INSERTION OF THE OCULAR MUSCLES INTO THE SCLEROTIC (FROM QUAIN AFTER MERKEL AND KALLIUS).

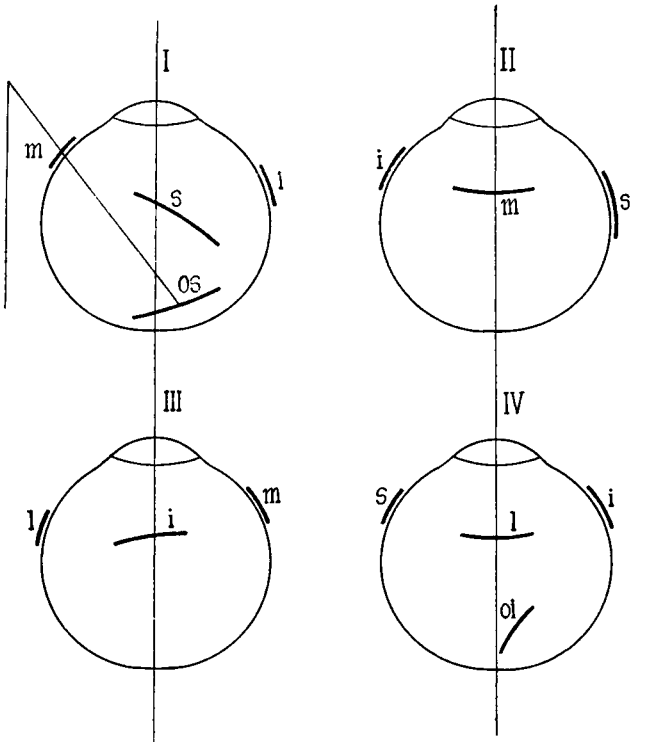


FIG. 21.—I, Globe from above; II, from the nasal side; III, from below; IV, from the temporal side; s, rectus superior; i, rectus inferior; m, rectus internus (s, mesialis); l, rectus externus (s, lateralis); os, obliquus superior; oi, obliquus inferior.

sense. We understand proprioceptive sense to mean that there are specialised sensory endings within the muscle fibres, surrounding connective tissue, tendons and joints from which impulses pass to the brain, conveying impressions of movement, position, stretching and vibration. Careful examinations and experiments have been

done by Irvine and Ludvigh which seem to prove that the proprioceptive sense in the extra-ocular muscles has been found wanting, and if present at all, plays no *rôle* in projection, stereopsis or the interpretation of motion on the retina.

The *third cranial* or *oculo-motor nerve* springs from the oculomotor nucleus in the grey matter of the floor of the aqueduct of Sylvius opposite the superior colliculus. The fibres pass forward through the tegmentum and emerge from the sulcus oculomotorius which lies on the inner side of the crus cerebri. Passing for about 20 mm. through the cisterna basilis, where it lies between the

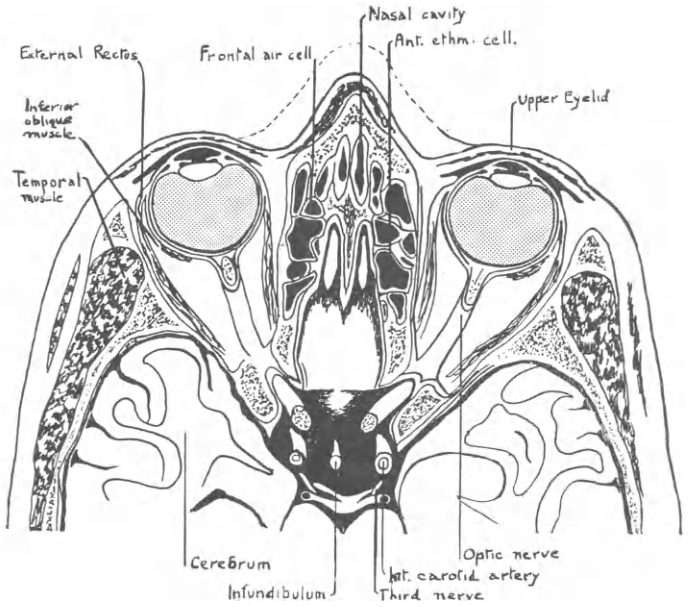
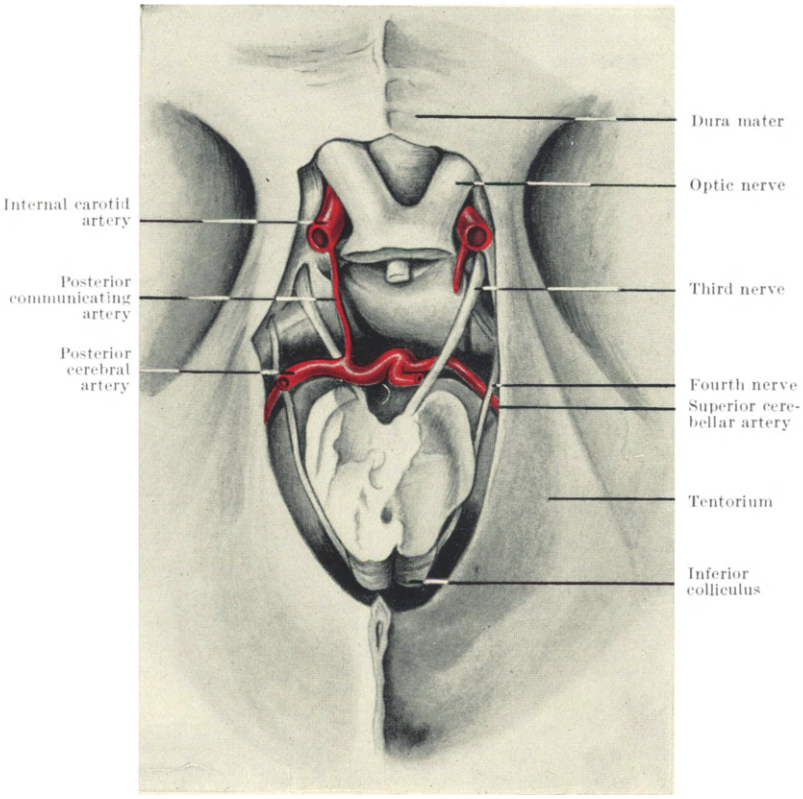


FIG. 22.—Horizontal section through middle of orbital cavities, eyeballs and optic nerves. Drawn by Miss M. E. Rea, B.A.

posterior cerebral and the superior cerebellar arteries, it lies to the outer side and anterior to the posterior clinoid processes, from whence it crosses the attached margin of the tentorium cerebelli to lie laterally to the pituitary fossa above the cavernous sinus. The nerve pierces the dura mater between the posterior and anterior clinoid processes and continues forward in the outer wall of the cavernous sinus to the inner end of the superior orbital fissure. For the arrangement of the structures in the superior orbital fissure

PLATE I



The third and fourth nerves in their intracranial course. (G. D. Thane, drawn by T. W. P. Lawrence from Quain's Anatomy, Vol. III, Part 2).

The cerebrum has been removed showing the mid-brain divided in the aperture of the tentorium. The third nerve is exposed on the right side by dividing the posterior cerebral and posterior communicating arteries. On the left side the tentorium and pedunculus cerebri are slightly separated so as to show the course of the fourth nerve more fully.

[*To face p.* 46.

see Fig. 17. While in the cavernous sinus the nerve receives sympathetic twigs from the cavernous plexus.

The oculo-motor or third cranial nerve supplies all the extrinsic muscles except the external rectus and superior oblique. The two divisions of this nerve enter the orbit between the two heads of the external rectus (see Fig. 18). The superior division supplies the superior rectus and the levator palpebræ superioris. The inferior

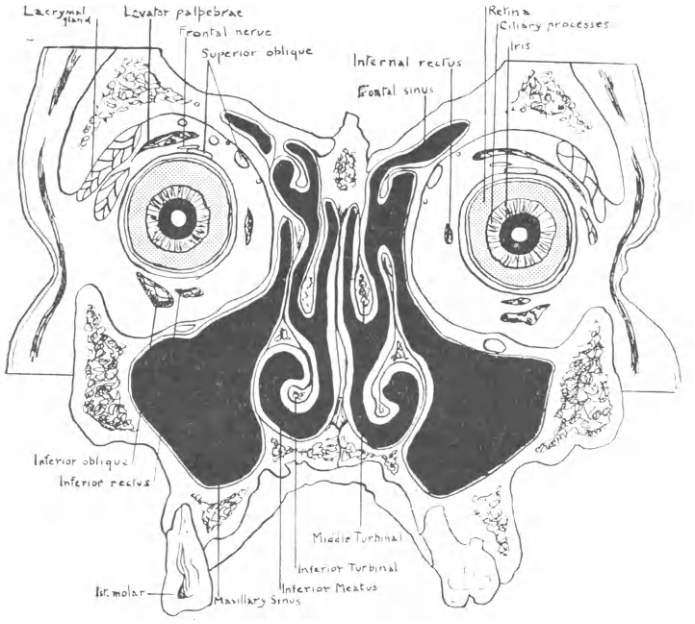


FIG. 23.—Coronal section of head of adult male, 2¾ inches in front of external auditory meatus, viewed from behind. (J. Symington.)

division, supplies the internal rectus, the inferior rectus and the inferior oblique. The nerves to the two recti enter the ocular surfaces of these muscles, while the nerve to the inferior oblique passes forwards between the inferior rectus and external rectus and enters the posterior border of the inferior oblique muscle.

Paralysis of the entire oculo-motor nerve, or any single branch of this nerve, may take place. Complete paralysis of this nerve produces ptosis, together with a downward and outward rotation of the eyeball, while the sphincter of the pupil which is supplied by this nerve is also paralysed, allowing dilatation of the pupil to take place.

The *fourth* or *trochlear nerve* is the most slender of the cranial

nerves, and has the longest course within the cranial cavity. It is distributed solely to the superior oblique muscle of the eye.

The nucleus of the fourth nerve, consisting of multipolar cells, continues downwards the column of cells which gives origin to the third cranial nerve ; it lies in the ventral grey matter of the aqueduct of Sylvius, opposite the upper part of the inferior colliculus. From the nucleus the fibres pass to the medial surface of the mesencephalic root of the fifth nerve, then downwards and inwards, entering the superior medullary vellum, where they almost completely decussate to the opposite side. After having crossed in the valve of Vieussens (superior medullary velum) and emerging from its upper end at the medial border of the brachium conjunctivum, the fourth nerve is directed at first outward across the superior cerebellar peduncle and then turns forwards round the pediculus cerebri (see Plate I), lying parallel to and between the posterior cerebral and superior cerebellar arteries. It enters the dura mater immediately beneath the free margin of the tentorium a little behind the posterior clinoid process and runs forward in the outer wall of the cavernous sinus, resting against the upper margin of the ophthalmic division of the fifth nerve and crossing the third nerve obliquely on its outer side from below upwards to the inner end of the sphenoidal fissure. Passing into the orbit above the external rectus muscle it inclines inwards over the levator palpebræ and superior rectus muscles and finally enters the upper surface of the superior oblique muscle. *Paralysis of the fourth nerve* is followed by limitation of downward movement. Diplopia occurs on looking down. Of the two images seen, the false one is lower and its upper end tilted towards the true image. Vertigo may be a prominent symptom.

Paralysis of ocular muscles may result from a lesion in any part of the nerves from the cerebral cortex to the muscles. Paralysis of conjugate movements is usually due to cortical or nuclear lesions.

When the extrinsic muscles of the eye are paralysed the condition is called *ophthalmoplegia externa*. If both extrinsic and intrinsic muscles of the eye are paralysed the term *ophthalmoplegia totalis* is applied. Ophthalmoplegia may be due to many causes, among these being *hæmorrhage* and *rheumatism*, the latter including many toxic conditions, which may act upon an ocular nerve alone or, by affecting surrounding structures also, may be associated with symptoms elsewhere. By hæmorrhage, thrombosis, embolism, etc., the intracranial portion of the ocular nerves may be damaged, pro-

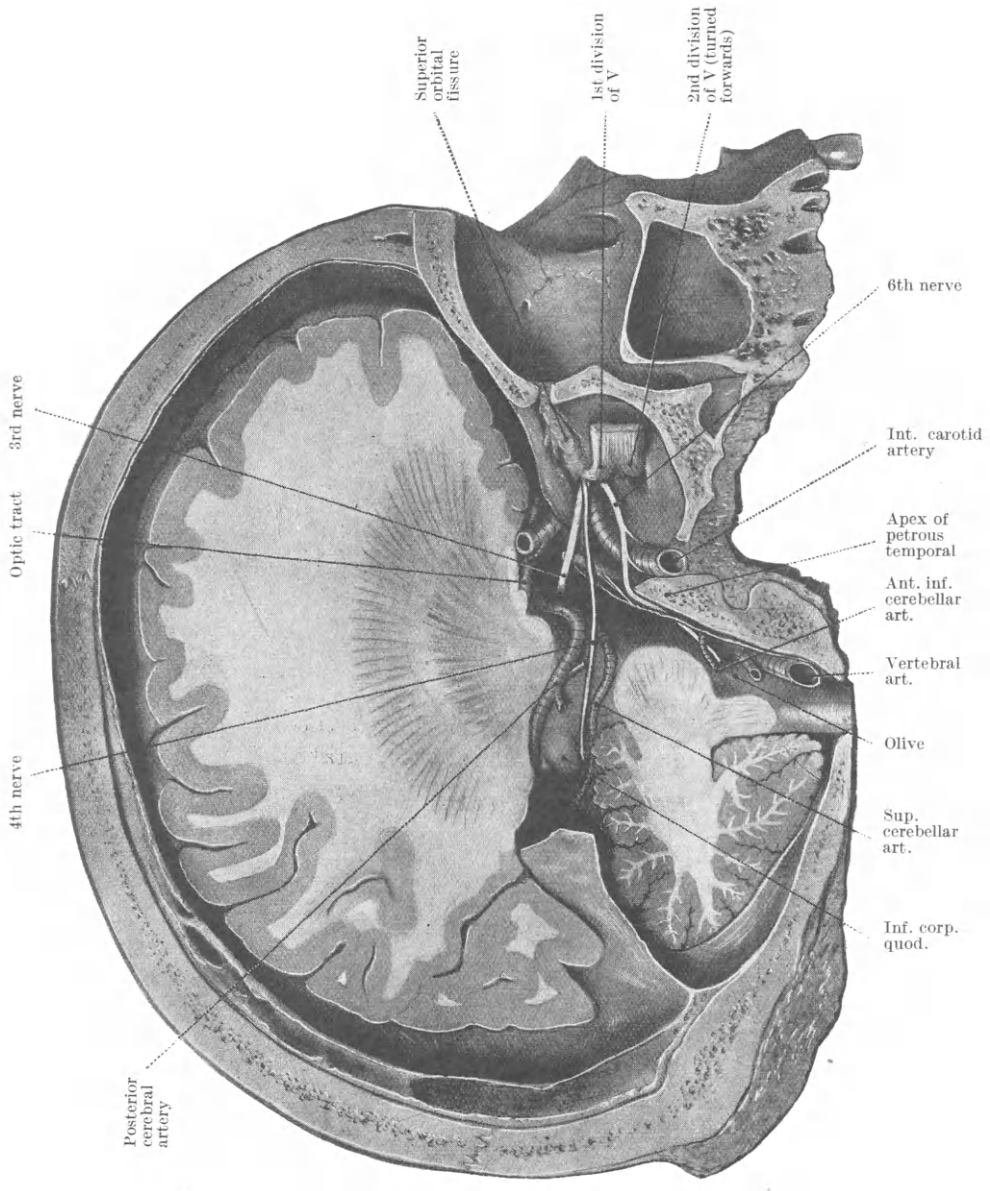


FIG. 24.—Dissection to illustrate the bend in the course of the sixth cranial nerve. (Kindly lent by Wolff.)

ducing various paralyses. The diphtheria toxin has a special predilection for both the external and internal muscles of the eyes, so

that paralysis of accommodation is frequently found due to the ciliary muscle of the eye being affected in this disease.

The *sixth nerve* (*nervus abducens*) has as its deep origin a nucleus in the floor of the fourth ventricle immediately above the striæ acusticæ, beneath the eminentiæ teres. From the inner side of the nucleus fibres pass forwards to the ventral part of the pons and there form a flattened band emerging from the lower edge of the pons external to the pyramid. At this superficial origin the basilar artery lies between the two sixth nerves and each is crossed by the anterior inferior cerebellar artery. About 15 mm. from the pons the nerve enters an aperture in the dura mater to the inner side and below the fifth nerve, passing posteriorly to the inferior petrosal sinus, crosses it and ascends almost vertically up the back of the petrous temporal near its apex. Here the nerve bends sharply forwards (Fig. 24) (Wolff's kink) under Gruber's ligament and the superior petrosal sinus and enters the cavernous sinus, where it passes on the outer side of the internal carotid artery, receiving sympathetic fibres from the carotid plexus to reach the orbit through the superior orbital (sphenoidal) fissure.

A paralysis of the abducent nerve alone has little or no localising value, it seems to be the structure most easily injured in any brain lesion. In young children a paralysis of the sixth cranial nerve is said to be the result of polio-encephalitis, but the author believes that in the great majority of children the paralysis is due to an injury at birth. Wolff has put forward the suggestion that owing to the relationship of the sixth nerve to the back of the apex of the petrous portion of the temporal bone, a position where the nerve in its course forwards turns abruptly at a right angle, pressure on the skull at birth could press the hind brain downwards, and so produce traction on the nerve at this point. Certainly pressure against bone is more likely to produce a lesion than pressure against a soft structure such as the antero-inferior cerebellar artery, as has been suggested by Cushing (see also p. 219).

When the refraction of these children, suffering from a paralysis of the sixth cranial nerve, is examined under atropine it is frequently found to be within normal limits. To ascertain if the squint in infants and young children is paralytic, it is only necessary to hold a small pocket torch before the child's face, when the eyes will at once be attracted by the light. Now ask the mother, while holding the baby in her arms, to turn away from the light so that the

baby's head is rotated from the paralysed side. As the mother keeps turning away from the light the baby endeavours to get his head back into the position in which it can still see the light. It can then be seen that the eye with the paralysed muscle attached cannot cross the middle line no matter how far the baby turns its head in the same direction. Such paralyses often recover, so that an operation for the squinting condition should not be hastily considered, but recovery is usually slow. Instead, a cover should be worn over the eye on the normal side to prevent amblyopia supervening in the squinting eye, and this also will help the recovery of the paralysed muscle.

Paralysis of an extraocular muscle has been found following on *spinal anæsthesia*. Cases have been described by Blatt, Moore, Levine and many others. Blatt found that in 64 per cent. stovain was used, in 26 per cent. procaine hydrochloride, in 7 per cent tropocaine and in 3 per cent. cocaine. Procaine hydrochloride and tropocaine were more quickly excreted from the body than stovain. Levine says the sixth, or abducent, nerve is the most commonly affected, but two cases each of the optic, facial and sympathetic have also been recorded. Schubens reported a case of bilateral abducent nerve palsy with choked disc after procaine hydrochloride —epinephrine lumbar anæsthesia.

No satisfactory explanation has yet been given to account for this condition.

The onset was found to vary from three days to three weeks, and while recovery took place from one week to several months, Popovici's case of the fourth nerve took nine months to recover.

Syphilis, disseminated sclerosis, etc., are responsible also for the various paralyses which occur. These will be described in detail in a later chapter. There is one form of paralysis which has not received adequate recognition, that is, a type of *diplopia produced by hæmorrhage*—whether into the muscle or the nerve I cannot state. It most commonly occurs as a vertical diplopia. An example is as follows : A lady about to leave her house suddenly saw that the stairs leading down from the doorway had become doubled, one flight being placed on a higher plane than the other. The sensation was both painful and nauseating. Double vision, however produced, will cause some degree of sickness, but this type of diplopia comes on with intense suddenness and is actually painful. Dr. A. F. Voelcker found the systolic blood pressure of this patient raised to

297 millimetres and the diastolic was abnormally high also. The patient was put to bed, the blood pressure was gradually reduced, and in three months' time the diplopia had entirely disappeared. A similar type is that found in patients who have met with severe accidents, most commonly motor-car accidents. Again, there is a sudden diplopia which may increase in extent for a week or so, remain stationary and then quickly disappear. The history of recovery is common to nearly all of these cases and is due to complete absorption of the hæmorrhage. At one time I thought that the hæmorrhage was in the muscle, but the last case of motor car injury which I have seen showed a paralysis of the superior and internal recti muscles. This therefore was due to a lesion of the oculo-motor nerve before dividing into the superior and inferior branches behind the orbit. It took exactly four months for complete recovery to take place. If this hæmorrhage had occurred at the third nerve nucleus there would have been accompanying symptoms from damage to neighbouring structures ; but it was not so in this particular case.

A type of nerve paralysis commonly seen in young women is produced by focal infection, more often than not such as is found at the roots of teeth. These cases are often suspected of suffering from early disseminated sclerosis, but the necessary attention to the dental condition, together with the administration of salicylates, brings about a cure.

Ocular palsies occurring in diabetes have been ascribed to neuritis, but are more probably due to vascular lesions involving the nerves. (Brain.)

Recurrent third nerve paralysis. This affection, which is rare, is seen in young females. It occurs on one side only and the recurrence takes place in the same nerve. The attacks, which last from days to months, are accompanied by headaches such as are found in migrainous subjects. The headaches may continue for several days. The intervals between the attacks last from ten days to a year. The lesion has been found in the trunk of the nerve (see p. 333).

Conjugate paralysis, which is an abolition of certain synkineses, may be produced by damage to the central nervous system ; the patient cannot direct both eyes upwards or downwards, to the right or to the left. Conjugate deviation of the eyes is brought about by means of fibres passing from the sixth nerve nucleus through the posterior longitudinal bundle to the opposite third nerve nucleus.

When one sixth nucleus is injured the patient is unable to look to the side, but other movements such as convergence remain unimpaired. The close association of the sixth nucleus with the seventh nerve nucleus may bring about in a common injury paralysis of the external rectus together with facial paralysis of the same side. Nuclear ophthalmoplegia is seen in cases of congenital ptosis, the innervation either to the superior rectus or to the levator palpebræ superioris or to both being absent. Congenital syphilis very rarely

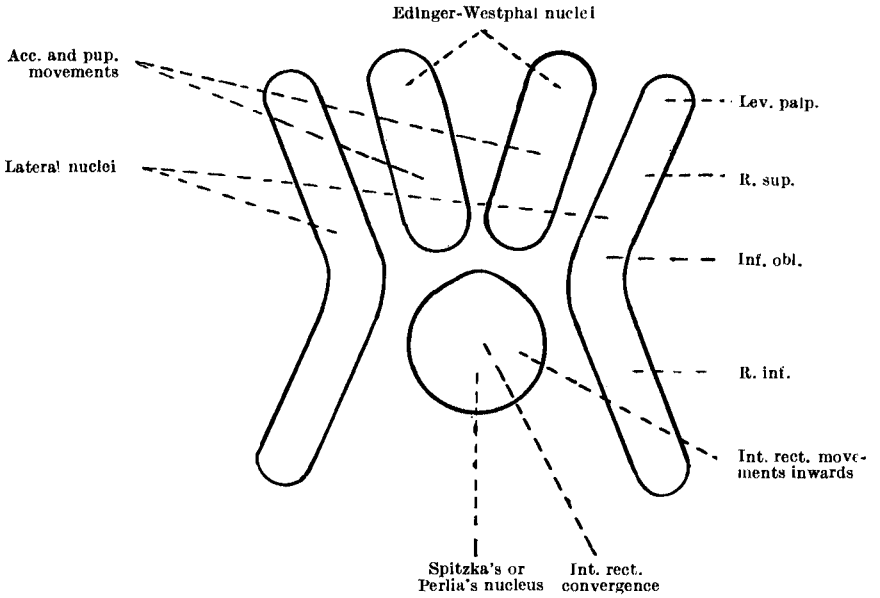


FIG. 25.—Diagram of the oculo-motor nerve nucleus.

produces a squint. In all the cases of interstitial keratitis which the writer has seen he has never once found an accompanying paralysis of an extra-ocular muscle; but a squint is common in acquired syphilis.

The *third and fourth cranial nerve nuclei* lie near the middle line on the floor of the Sylvian aqueduct beneath the superior colliculus. The nucleus of the oculo-motor nerve is divisible into several parts (see Fig. 25). The Edinger-Westphal nuclei consist of the paired group of small cells lying anteriorly between the diverging extremities of the lateral groups. The lateral nucleus consists of a paired group of large stellate cells somewhat reniform in shape with its

concavity outwards. The central nucleus (Perlia's or Spitzka's) is an unpaired group of large stellate cells lying between the lateral nuclei, this group is most probably concerned with convergence. It is probable that the lateral nuclei subserve the function of the extrinsic muscles, while the Edinger-Westphal that of accommodation and pupillary movements. Winkler says the cells of the Edinger-Westphal nuclei do not send fibres to the oculo-motor nerve roots, and consequently should not be considered as belonging to the oculo-motor nuclei, but are concerned with the synergic movements of the eye, especially for the control of the upward associated movements of the eyeballs. He thinks, however, that the Edinger-Westphal nuclei do regulate pupillary movements also.

The *central neuron of the oculo-motor nerve* is most probably found in the angular gyrus. Running through the base of the internal capsule it reappears as a part of Spitzka's bundle and decussates in the oculo-motor nucleus of the opposite side. From the ganglion cells of this nucleus the roots of the oculo-motor nerve pass, and after partial decussation run to the eye as the trunk of the third cranial nerve. The nucleus of the fourth cranial nerve also lies below the superior colliculus and behind the third nerve nucleus. There are *two cortical oculo-motor centres*, first in the frontal lobe, which control volitional movements, and secondly in the occipital lobe, controlling fixation reflex (see p. 80).

The nucleus of the third nerve, besides receiving fibres from the frontal cortex, obtains fibres from the superior colliculus and from the cerebellum *viâ* the superior peduncle. It sends fibres *viâ* the anterior and posterior longitudinal bundles to the fourth and sixth nerves of the same and opposite sides (see p. 122), also fibres which join the seventh nerve. These latter are thought to be the nervous supply to the orbicularis oculi and frontalis, hence in a supra-nuclear lesion of the seventh nerve these muscles escape.

The fifth or trigeminal nerve. Fig. 19 shows the Gasserian ganglion exposed by the removal of the dura mater. It lies in a space formed by separation of the two layers of the dura mater. This space is called the Cavum Meckelii. From the outer or convex part of the ganglion the three main divisions of the trigeminal nerve emerge—the ophthalmic division, the superior maxillary and the inferior maxillary division.

The interlacement of the fibres of the sensory root proceeding

from the antero-lateral aspect of the pons occupies a smooth depression which marks the anterior aspect of the apex of the petrous portion of the temporal bone, where it sinks into the ganglion. The motor root lies along the inner side of the large sensory root and is later found occupying a groove upon the deep surface of the ganglion. The ophthalmic division is the smallest of the three divisions of the trigeminal nerve. It is purely sensory ; proceeding forwards in the outer wall of the cavernous sinus it divides in the sphenoidal fissure into three terminal branches. In the cavernous sinus it lies on a lower level than the oculo-motor and trochlear nerves. The terminal branches of the ophthalmic division are the nasal, the lacrimal and the frontal. As is seen in Fig. 26, the nasal takes origin first ; then the lacrimal comes off the main stem and the latter is continued forwards as the frontal. The nasal nerve (naso-ciliary) having passed between the two heads of the external rectus muscle and between the two divisions of the third nerve, crosses the optic nerve obliquely as it runs forward between the internal rectus and superior oblique muscles. At the inner wall of the orbit it ends by dividing into two terminal branches, the infra-trochlear and the nasal nerve proper ; while in the orbit the nasal nerve gives off the long ciliary nerves and a long root to the lenticular ganglion. The two long ciliary nerves cross the optic nerve reaching the globe of the eye to the inner side where they pierce the sclera. Of the terminal branches the infra-trochlear, passing under cover of the superior oblique muscle and its trochlea, emerges from the orbit above the internal tarsal ligament and the inner canthus. Its twigs supply the lacrimal sac, the conjunctiva, the skin of the eyelids and the root of the nose. The nasal nerve proper is the larger of the two terminal branches. It leaves the orbit by the anterior ethmoidal foramen, through which it passes to the anterior part of the cranium. It runs in a canal at the outer margin of the cribriform plate. From this it turns forwards under the dura mater and disappears through a narrow aperture at the side of the crista galli to reach the nasal cavity. It emerges upon the face by passing between the lower margin of the nasal bone and the uppermost lateral cartilage of the nose and gives branches to the skin of the nose and communicates with the facial nerve.

Hutchinson states, in reference to *herpes ophthalmicus*, that the nutrition of the eyeball is in danger when this disease affects the oculo-nasal nerve (external nasal branch), which is made evident by

an irruption of the skin at or near the tip of the nose. Just before writing this paragraph I was asked to see an elderly patient suffering from herpes ophthalmicus. There was no sign of this disease on the skin of the nose but her eyeball was in a precarious condition, especi-

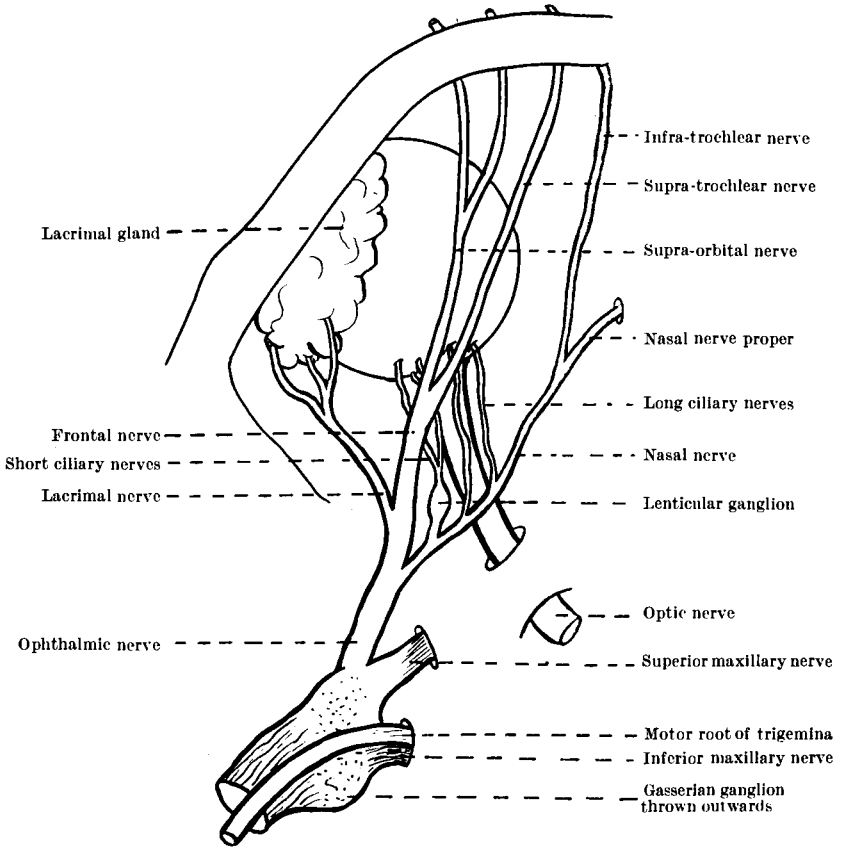


FIG. 26.—The ophthalmic nerve of left side. (After Cunningham.)

ally the cornea. Some of my colleagues agree with me that Hutchinson's statement cannot be fully accepted.

The lacrimal nerve is the smallest of the three branches of the ophthalmic division. It enters the orbit through the narrowest part of the superior orbital fissure and then runs along the upper border of the external rectus with the lacrimal artery. It enters the lacrimal gland, where it gives off several fine branches to the gland

and to the conjunctiva, and finally ends in the skin of the upper eyelid.

The frontal nerve is the largest branch of the ophthalmic. It enters the orbit through the spheno-maxillary fissure, runs forwards between the levator palpebræ superioris and the periostium and divides into two branches, the supra-trochlear and the supra-orbital; the latter, the larger of the two, passes through the supra-orbital foramen, giving off at this point several filaments to the upper eyelid. It then ascends upon the forehead, ending in two cutaneous branches for the supply of part of the scalp.

The supra-trochlear nerve passes above the trochlea or pulley of the superior oblique, escaping from the orbit between the trochlea and the supra-orbital foramen. It turns upwards on to the forehead and sends filaments to the conjunctiva and skin of the upper eyelid.

The lenticular ganglion (ciliary) (see Fig. 26) is a small quadrangular flattened ganglion about the size of a pin's head and of a pale reddish colour. It lies in the orbital fat between the optic nerve and the external rectus muscle on the lateral side of the ophthalmic artery. Its sensory root passes from the nasal nerve; the second or motor root is derived from the branch of the third nerve to the inferior oblique, while the sympathetic root is from the cavernous plexus of the sympathetic. These three roots enter its posterior border. Its branches are the short ciliary nerves, six to ten in number, arising from the fore part of the ganglion; the superior set passes forward above and the inferior branches below the optic nerve; they accompany the long ciliary nerves and pierce the sclera around the entrance to the optic nerve. They are distributed to the ciliary muscle, iris and cornea.

The Syndrome of the Nasal Nerve.—This symptom-complex, described by Charlin, is the evidence of a neuritis of the nasal branch of the trigeminal nerve. It is characterised by extreme pain quite out of porportion to the amount of ocular inflammation. There is nasal difficulty, especially marked hydrorrhœa. The cornea may be normal or it may show an ulcer or iritis may be present. Local treatment of the eye is of little value, and cocaine in the conjunctival sac does not give relief, but the application of epinephrine and cocaine to the mucous membrane of the nose anteriorly on the external wall brings rapid improvement in the ocular symptoms.

Neuroparalytic Keratitis.—This occurs sometimes when the fifth nerve is paralysed. The cornea becomes anæsthetic and the eyelids

are imperfectly closed, due to the loss of sensation in the eye. During an attack of herpes ophthalmicus the cornea becomes anæsthetic, and unless due care is taken ulceration will take place. Keratitis occurs in all cases of paralysis of both fifth nerves, but the consensual reflex apparently affords protection when only one nerve is affected. If the Gasserian ganglion is removed or the fifth nerve injected with alcohol for trigeminal neuralgia only a comparatively few cases get neuroparalytic keratitis (see Chap. 13).

Neuralgia of the Eye.—A large number of patients present themselves suffering from a condition which cannot be better termed than neuralgia of the eye. The severity of the pain varies greatly, from a slight aching in the eye to such pain that the ophthalmologist is forced to believe he may be dealing with a case of raised tension. The eyeball is painful to the touch. Turning the eyeball from side to side often intensifies the pain. It was not until the writer had personal experience of this condition that he learned to recognise and give it its true worth. The occasion was that of riding in a car, of which a cracked cylinder was admitting oil, so that dense clouds of smoke escaped from the engine. The windows and ventilators of the car had to be opened widely and the chilly sea air blew strongly through the car and across our faces. For two days and two nights the writer's eyes were painful, especially the right. It was impossible to sleep with the pain, and yet the eyes were not red nor was there any discharge. Constant hot applications, together with aspirin, brought about a cure.

Differential Diagnosis.—Such cases have to be examined most carefully. Eyelids are everted and calcium deposits in the conjunctiva looked for. The tension of the eye should be taken to exclude glaucoma. The absence of redness of the eyes excludes intra- and extraocular inflammation. The pain always disappears on the constant application of heat. Closely allied with this condition is an affection of the frontal nerves, *neuralgia of the forehead*, commonly brought about by exposure to draughts of air during sleep. Frequently the pain of the forehead is associated with some pain in one or both eyes, the fifth crainal nerve being the common source of supply to these parts. People who are liable to this type of neuralgia cannot sit at the open window of a train or motor car, while those who are martyrs to such an affection should remember that comfortable woollen nightcaps were not worn by our grandfathers without a purpose. I often prescribe the wearing of a light

Shetland wool shawl across the eyes and forehead during sleep. This simple remedy has brought relief to many of my patients who have suffered intermittently for years without knowing the cause.

Paralysis of the Seventh or Facial Nerve.—The seventh or facial nerve is composed of both motor and sensory fibres The nucleus of the motor portion arises in the formatio reticularis of the lower part of the pons, almost on the same level as the sixth nerve nucleus but deeper from the floor of the fourth ventricle. The sensory portion of this nerve is known as the pars intermedia, or nerve of Wrisberg. It arises from the geniculate ganglion of the facial, lying in the bony facial canal, and this sensory portion is attached to the brain stem between the eighth nerve and the motor portion of the seventh. The sensory portion proceeds peripherally from the geniculate ganglion as the great superficial protrosal nerve, while other nerve fibres pass on with the motor fibres to the stylo-mastoid foramen and there become the chorda tympani nerve. The facial nerve divides in the face into the larger temporo-facial division and the smaller cervico-facial division. The former supplies among other muscles the orbicularis palpebrarum. The temporal branches form communications with the supraorbital and lacrimal branches of the ophthalmic division of the trigeminal. It is possible, as Willbrand and Saenger have suggested, that the lacrimal gland has a double innervation. The sympathetic through the trigeminal nerve bringing about the normal secretion of tears, while the facial through the great superficial petrosal nerve, producing a copious flow of tears in emotional or psychic states. The great superficial petrosal nerve from the facial joins the great deep petrosal nerve from the carotid sympathetic plexus to form the Vidian nerve which passes to the spheno-palatine ganglion of the maxillary division of the trigeminal ; by blocking this ganglion Ruskin found it produced a diminution of the flow of tears when this was excessive (Fig. 27).

Vidian Neuralgia.—A case of sphenoidal disease has been reported by Vail, not only with symptoms hitherto ascribed to Meckel's ganglion neuralgia, but in addition to the typical distribution of the vidian nerve, viz. nose, eyeball, teeth, ear, mastoid and shoulder, the pain in the eye was accompanied by a contraction of the pupil with irregularity of its contour, reddening of the conjunctiva and pain in the upper canine, bicuspid and molar teeth.

As the orbicularis palpebrarum is supplied by twigs from the

facial nerve it follows that in paralysis of this nerve or Bell's palsy there is failure of the closing action of the eyelids wtih a consequent loss of winking. The lower eyelid sags, so that tears fall over the edge of the lids. There is therefore constant exposure of the eye, and as this leads to lack of moisture, then, when foreign material enters the eye, ulceration of the cornea readily follows, more so if

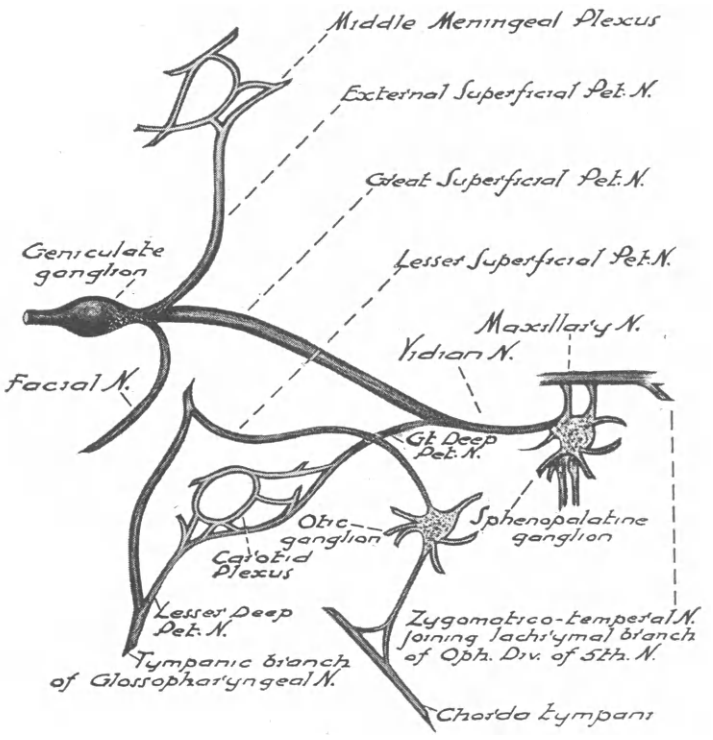


FIG. 27. The five petrosal nerves and secretory fibres to lacrimal gland.

by any chance paralysis of the ophthalmic portion of the trigeminal nerve be present.

The Lacrimal Gland.—The lacrimal gland, situated in the upper and outer part of the orbit, is shielded, partly by the orbital plate of the frontal bone and partly by the outer part of the upper eyelid. The gland is supplied by the lacrimal branch of the ophthalmic division of the trigeminal, the cervical sympathetic, and from the facial nerve through the great superficial petrosal nerve and sphenopalatine ganglion, and thence to the maxillary division of the

trigeminal which anastomoses with the lacrimal nerve by means of the temporo-malar nerve. In the operation for the cure of tic douloureux section of the fifth nerve by the temporal route is frequently followed by the loss of lacrimal secretion. Dandy has shown that this is due to injury of the great superficial petrosal nerve which occurs when the dura is stripped from the base of the skull; for when the posterior route is chosen this nerve is not interfered with and there is no diminution of tears.

Hypersecretion of the Lacrimal Gland.—In certain emotional people there is excess of lacrimination; also in old people this condition is seen. This state must not be confused with a blocking of the tear ducts leading into the nose. Excessive light will cause lacrimination in some people, as also will a retained foreign body or slight conjunctivitis or iritis. Infrequent winking on one side may cause hypersecretion on same side.

Diminution or Absence of Secretion.—In paralysis of the trigeminal or facial nerves there may be entire absence of the lacrimal secretion. If there is a lesion of the trunk of the facial nerve there is also paralysis of the soft palate and uvula. In some of the fevers, cholera and diabetes, tabes and exophthalmic goitre, the secretion may be diminished in amount. There is a psychical condition, too, in which patients state that they cannot weep as formerly.

The following résumé of the **chief causes of ocular palsies** may be of considerable value in aiding the reader's memory :—

(1) **Lesions situated in the orbit** such as are produced by abscess or cellulitis, sinusitis, involvement by growths, exophthalmic goitre, periostitis, the gummatous stage of syphilis, injuries, including punctures, fractures and hæmorrhage.

(2) **Basic lesions,** that is, lesions situated at the sphenoidal fissure, and those at the base of the skull between the sphenoidal fissure and the pons. Swanzy states that this second class—those due to basic lesions—provides by far the largest number of cases of paralysis of the orbit and muscles. These are chiefly of a rheumatic or syphilitic nature. The word rheumatic might well be exchanged for toxic. The sixth cranial nerve is especially prone to a toxic paralysis. The prompt termination on finding the source of the toxin or virus and its elimination should guard the reader from the common mistake of labelling the disease early disseminated sclerosis. Fracture of the base of the skull is sometimes indicated by paralysis of the fourth cranial nerve, due to involvement of the nerve as it

passes over the apex of the petrous portion of the temporal bone. Sixth nerve paralysis, however, is more commonly associated with this lesion.

Paralysis of the external rectus muscle in the course of inflammatory disease of the ear is known as Gradenigo's syndrome. It may be accompanied by tempero-parietal pain, usually in the ophthalmic branch of the trigeminal nerve.

I have just seen a case which has been published by Worster-Drought. It was that of a *pituitary adenoma* which had caused consecutively a hæmorrhage into the third, fifth, sixth and seventh cranial nerves ; each hæmorrhage had absorbed in turn except that of the third cranial nerve, and now for two years the patient has presented the appearance of a divergent squint due to paralysis of the internal rectus muscle. Also there are other facial acromegalic characteristics present due to the pituitary condition.

It must not be forgotten that the presence of primary optic atrophy or of a bitemporal hemianopia will sometimes furnish conclusive evidence of the basal origin of the extraocular muscle paralyses.

Ophthalmoplegic migraine or intermittent paralysis of the third nerve of one eye will be referred to later on (see Chap. 13).

(3) **Brain-stem lesions** involving the ocular nerve fibres in the substance of the mid-brain and pons are sometimes found ; these are called *fascicular ;* or the lesions may attack the nuclei of the nerve, which are then known as *nuclear.* The lesions producing such paralyses are cerebral syphilis, hæmorrhage, encephalitis lethargica, disseminated sclerosis, polioencephalitis and myasthenia gravis. Acute nuclear paralysis is due to an acute inflammatory process in the nuclei resembling that which produces poliomyelitis anterior acuta or to hæmorrhagic lesions. Alcoholic poisoning by producing an acute peripheral neuritis of the ocular nerve may be mistaken for acute nuclear palsy. *Nuclear palsies* are found in diabetes, influenza, and lead poisoning. Among the many cases of nicotine poisoning seen by the writer he has never found a case of ocular palsy due to this cause. *Ptomaine poisoning* can produce a nuclear palsy. Tumours in the region of the pons may cause paralyses, both fascicular and nuclear, but the presence of headache, vomiting and papillœdema will point to a true diagnosis. Internal hydrocephalus and aneurysms will produce pressure symptoms which cannot be regarded as true nuclear ophthalmoplegia.

Nuclear lesions may give rise to *conjugate lateral paralysis*, loss of power of motion of the eyes to one side or the other, while the power of convergence of the optical axes is retained. Cases of mid-brain tumours associated with paralysis of the upward vertical movements of the eyes, together with Argyll Robertson pupils, have been reported by Feiling and others.

(4) **Cerebral lesions,** *i.e.*, supranuclear in the internal capsule and corona radiata or cortex. Conjugate lateral paralysis is by far the commonest form of the symptoms produced by such cerebral paralysis. In paralysing lesions the deviation of the eyes is towards the lesion, while in irritating lesions the spasm of the affected muscles causes the deviation to be from the side of the lesion. These conditions are the reverse of what happens in conjugate lateral deviation due to pontine lesions. There are thus four possible cases (Landouzy and Grasset) (see p. 83) :—

Cerebral lesions	Paralytic	. Eyes turned away from paralysed side.
	Irritative	. Eyes turned towards convulsed side.
Pontine lesions	Paralytic	. Eyes turned towards paralytic side.
	Irritative	. Eyes turned away from convulsed side.

Gordon Holmes has stated that from the study of clinical and post-mortem cases it can be said that in the upper end of the mid-brain in the neighbourhood of the anterior quadrigeminal bodies are association centres which control the conjugate vertical movements of the eyes. He believes that a lesion of the anterior end of the mid-brain will involve upward movements, downward movements and convergence in this order as it progresses from before backwards ; also that the paralysis of the pupil is often associated with loss of upward movement, while loss of convergence is associated with paralysis of downward movement.

Finally, paralysis of the ocular muscles, both congenital and hereditary, is common and may vary from paralysis of a single muscle to complete ophthalmoplegia externa. Ptosis with or without defect of upward movement of the eyeballs is the commonest form. Ocular paralysis has followed mumps (Butler).

Summing up, we see that the various sites where a lesion may

interrupt the neuro-muscular chain and bring about a palsy are indicated in a concise manner by the following classification of oculo-motor palsies by Leslie Paton :

I. Paralyses of Central or Cerebral Origin, comprising Three Sub-groups :

(1) Supranuclear lesions :

(*a*) Cortical.

(*b*) Subcortical.

(2) Intranuclear lesions.

(3) Nuclear and nerve root lesions.

II. Paralyses of Extra-cerebral Origin, *i.e.*, of Nerve Trunks, again comprising two Sub-groups :

(1) Intracranial.

(2) Intraorbital.

III. Paralyses of Muscular Origin.

Again following Paton, we can classify the diseases which may cause an ocular palsy in the various parts of the neuro-muscular chain :

Classification of Ophthalmoplegias

I. Acute and Sub-acute Ophthalmoplegias.

A. Poisons :

Alcohol.
Lead.
Botulism.
(Carbon monoxide ; nicotine ; snake poison ; sulphuric acid.)

B. Infective agents :

Diphtheria.
Influenza.
Lethargic encephalitis.
Syphilis.
Tubercle.
(Measles, typhus, chicken-pox, septicæmia, chills, mumps.)

C. Trauma.

D. No etiological factor.

II. Chronic (Progressive) Ophthalmoplegias.

A. An isolated lesion.
B. Tabes and general paralysis.
C. Disseminated sclerosis.

Postero-lateral sclerosis (sub-acute combined).
Syringomyelia.
Paralysis agitans.

D. Bulbar palsies.

Progressive muscular atrophy.
Myasthenia gravis.

E. Graves' disease.
F. Diabetes.

III. Congenital Ophthalmoplegias of Nuclear Origin.

IV. Unilateral Ophthalmoplegia.

V. Recurrent and Alternating Ophthalmoplegia of Nuclear Origin.

Nystagmus.—Definition: Nystagmus is a condition in which the eyes are seen to move in a more or less rhythmic manner from side to side, up and down, or in a rotatory manner from the original point of fixation. Other types of movement have been observed, such as the see-saw nystagmus described by Maddox, in which one eye turned upwards while the other turned downwards; or the quivering nystagmus of Uhthoff, in which very small rapid and even movements occur when the eyes are being examined by oblique illumination or else at certain periods. In *spasmus nutans*, although the movements may be lateral, vertical or rotary, the eyes occasionally seem to converge or diverge rhythmically. It is possible in many people with normal eyes and sight to produce jerky movements when the eyes are moved to the extreme right or left. These nystagmoid jerks may be horizontal or rotary, and suggest that there is some difficulty in maintaining the eyes in these extreme positions. One must be careful not to include these cases among those of true nystagmus. Such pseudo-nystagmus is observed not only in persons with normal health, but in the debilitated and also in some cases of multiple sclerosis and hereditary ataxia.

Voluntary nystagmus has been described by Pyle and Ball and others who have seen this rare condition, where rapid oscillatory

movements can be produced at will. Ball describes a unilateral case, while Foster Moore has seen the movements produced bilaterally. Such cases have never suffered from true nystagmus.

True Nystagmus.—The movements in true nystagmus are nearly always bilateral, and when they are rolling, simultaneous, or parallel are described as *conjugate nystagmus*. If the movements are smooth it is called *pendular nystagmus*, such are seen in cases of amblyopia and miners' nystagmus. If the movements are slow in one direction and jerky in the opposite it is known as *resilient* or *jerky nystagmus*. This latter is typical of that seen in *vestibular nystagmus*.

Disjunctive and dissociated nystagmus are rare forms in which the eyes are moved in a converging and diverging manner or the see-saw nystagmus of Maddox. Uhthoff gave the name of *quivering nystagmus* to very small vibrations of a rapid symmetrical character which occurs when the eyes are examined by oblique illumination or which occurs only periodically. There is also a *unilateral nystagmus*. *Physiological nystagmus* is produced by looking at objects from a fast moving vehicle, such as from the window of a train. Finally, the movements may be seen in cases of ocular paralysis or in paralyses of latent gaze. It is then known as *paretic nystagmus*.

The movements in nystagmus are involuntary and show true reciprocal innervation of the muscles concerned. One sometimes observes these movements being carried out in the presence of severe strabismus. A concomitant squint of 45 degrees in an adult shows exactly the same movement as if the visual axes were parallel, but the functional integrity of the muscles of the eye is necessary in the production of nystagmus. Attempts to produce it are therefore an excellent method of detecting ocular paralysis in patients with head injuries who are in a state of coma (Hautant). Spicer says that of 200 cases 50 per cent. were horizontal, 15 per cent. were rotatory and 12 per cent. were vertical.

The movements in nystagmus vary greatly in amplitude. They may be coarse, *i.e.*, over 15 degrees; fine, under 5 degrees; or medium, which are between. In some cases the movements are so fine as not to be perceived by the naked eye, but when the fundus is examined by direct ophthalmoscopy, whereby the fundus of the eye is magnified greatly, the minute and rapid movements of the disc are seen.

I quote from de Lapersonne and Cantonnet's book—"Manuel de Neurologie Oculaire"—the following regarding nystagmus:—

"In his report to the Société Française d'Ophthalmologie in 1913, H. Coppez states that the mechanism of nystagmus may be as follows: Vestibular nystagmus is a disturbance in the function of the vestibular nuclei and their immediate connections. On the other

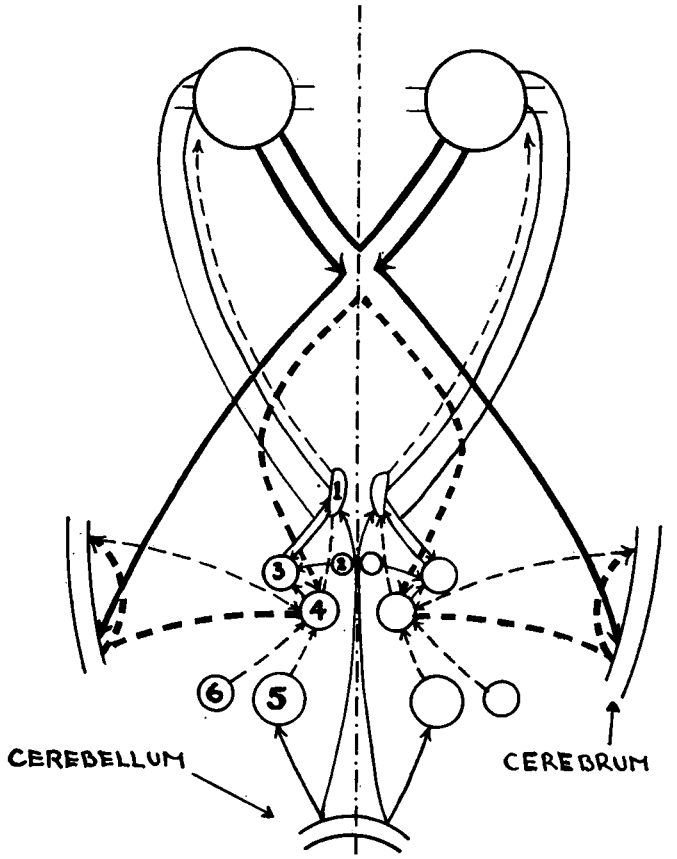


FIG. 28.—General scheme of nystagmus. (Coppez, from de Lapersonne and Cantonnet.) 1. Oculo-motor nucleus. 2. Trigeminal nucleus. 3. Clonic centre. 4. Tonic centre. 5. Vestibular apparatus. 6. Cochlear apparatus.

hand, nystagmus from ocular or auditory disturbances are due to effects produced on the nuclei or internuclear centres (that is on the co-ordinating relays between the peripheral nuclei and the real co-ordinating centres of vision). Cerebral nystagmus results from the suspension of the inhibiting action of the cortex on the centres

co-ordinating function. Cerebellar nystagmus results from an effect on the vestibular nuclei themselves : the vestibular nuclei can be regarded as aberrant cerebellar centres. The pathogenesis of nystagmus from weak vision may rest in the fact that the co-ordinating centres, escaping from the insufficient visual control, undergo stimuli which come to them from the various systems : strabismus, very often concomitant, would then be the result of the predominant action of one of the co-ordinating centres (Fig. 28).

" Nystagmus presupposes the existence of a centre of rhythmic co-ordination. If abnormal stimulation occurs, the usual associated deviations are no longer checked by this rhythm and continue their movement. The rapid phase of nystagmus is only a stratagem which causes the patient to think that the exaggerated deviation occurs always in the same direction, like animals which turn without ceasing in the same direction after lesions of the superior centres.

" Sauvineau has shown a close relationship between conjugate deviation and nystagmus, and H. Coppez was able to give this pretty definition : ' Many cases of nystagmus are merely conjugate deviation in motion.' A case (of disseminated sclerosis) observed by Rönne fits this definition well : when cold water was injected into the ear the patient showed, not nystagmus, but a sustained deviation of both eyes (conjugate deviation) ; this class of nystagmus therefore represents kinetic conjugate deviation. The others are only incomplete tetanus, a contraction, as in the nystagmiform twitches of paretic muscles or in miners' nystagmus.

" Here are some points in technique : to favour the appearance of nystagmus, raise the eyelid and tell the patient to look in the direction of the nystagmus sought for. Staying in the dark room for a time makes it easier. When the movements have begun, tilt the patient's head to the right, to the left or at 45 degrees backwards and forwards. The nystagmus changes in direction and form.

" There are often associated signs : vertigo and a tendency to fall, which occurs in the same plane as the nystagmus, but on the opposite side, except in cerebellar lesions where, for instance, it will be always backwards. In the gyratory test the patient must be made to turn in the direction of the ear which is being examined. The galvanic test is more sensitive. It is used mainly by neurologists, not by otologists who need to know if the labyrinth is completely destroyed " (see use of Tumarkin's nystagmograph).

" Hautant gives the following formula as normal :

" Caloric tests at 28 degrees : well-marked movements. Gyratory test : lasts forty seconds on the right and thirty on the left. Galvanic test at 4 milliamperes : head leans toward the pole.

" In pathological conditions : functional abolition of the vestibule is shown by a complete absence of nystagmus during the caloric test. The gyration test shows a diminution on both sides, especially on the side with the lesion. The galvanic test requires 15 milliamperes and the inclination of the head is abnormal.

" Vestibular hyperfunction causes caloric nystagmus which may last for three minutes. The same applies to the gyration test. Deficient function gives only short and feeble reaction to the caloric and gyratory tests. The galvanic test requires from eight to ten minutes to cause falling, which is in an abnormal direction.

" Professor Bard considers that the gyratory method of testing (ten turns in twenty seconds) is too violent. In addition, he does not separate tests of the different semicircular canals. He has therefore caused a chair to be made which can be transformed into a horizontal turning bed. Fixation of the eyes must be ensured by fixing an object which turns with the patient.

" Bard has described an inverted or backward form of nystagmus in hemiplegics.

" Lafon states that nystagmus is produced by lesions of the vestibular apparatus, of the cerebellar centres of balance, and of the paths joining them to the cortex. For him, voluntary nystagmus, miners' nystagmus and nystagmiform contractions are merely false nystagmus from paresis or contractions of the oculo-motor muscles.

" Finally, we must mention Brabant's opinion, according to which the sensitivity of the extrinsic muscles of the eye plays a predominant part in the genesis of phenomena of balance (real or illusory sensation of movement, of position, of immobility, balancing reactions, nystagmus, etc. . . .), even when these phenomena are produced by such means as rotation He goes further and assumes that the sense of balance in general, and the sense of movement of the body in particular, have generally no other peripheral organs than the extrinsic muscles of the eye."

The response of the eyes to a succession of moving objects is known as *optico-kinetic nystagmus*. The movement consists of two phases a slow deviation in the direction of movement and a rapid jerk in the opposite direction. The fundamental problems associated with optic nystagmus together with a list of the more important literature

on the subject are dealt with in a paper by Dodge and Fox. These writers believe that fixation and pursuit of an object may be determined by any sensitive part of the retina, even in the absence of central vision so that optic nystagmus can be evoked by peripheral retinal stimulation alone when central vision is completely excluded by an absolute central scotoma.

Etiology of Nystagmus.—The causes of nystagmus fall under the following heads: *Ocular nystagmus.* This condition appears in children who have been amblyopic since infancy or early childhood, the diminished vision being due to corneal opacities or nebulæ produced by *ophthalmia neonatorum*, congenital or infantile cataract, colour blindness, deformity of the macular area as in retinal abiotrophy or disease produced by congenital syphilis.

Accurate or minute vision is determined by the presence of the macula in the retina. Here, while the visual axis is directed towards an object, the image of such an object falls and is seen clearly and definitely to the minutest degree. If this area is not functioning then the eye movements are not fixed and the eye wanders from side to side trying to obtain the sharpest image, or, in other words, making an attempt to perceive as clearly as possible the object looked at. The writer has observed a very young child who, at the age of six months, lost its central vision, and now at two and a half years of age is seen trying to obtain the best image possible of objects looked at. He moves his head from side to side while the eyes wander in wide excursions as the child is asked to look at an object held in the hand of the surgeon. This stage could be looked upon as the beginning of nystagmus. The sensibility of the retina to form is greatest at the macula. It decreases rapidly from this point outwards towards the limit of the field of vision, so that images become more and more indistinct the farther they are removed from the macular area. It is due to loss of macular perception that many cases of nystagmus are developed.

The aimless wandering of the eyes in those who are congenitally blind or in those who have been blinded in later life should not be termed nystagmus.

Occupational Nystagmus.—This is typically seen in the disease known as miners' nystagmus. The movements in this type are chiefly rotary and often dissimilar. This is a distressing disease. The patient's health is poor, he has defective vision, which is worse at night, he suffers from headache, photophobia and complains of

movements of lights and objects. The symptoms are such as to prevent work being carried on. This type of nystagmus is common in mines where the illumination is feeble. The area surrounding the macula has slightly greater sensitiveness for light, so that macular vision in such dull light is not of the greatest importance. There is also a vestibular element in this disease, as the eye movements can be stopped by altering the position of the patient's head.

Llewellyn begins his book on " Miners' Nystagmus " by saying: " Miners' nystagmus is an occupational disease of the nervous system which is confined to workers in coal mines."

While he is personally convinced that the primary cause of nystagmus is deficient illumination, Parsons believes that the " chief cause of prolonged disablement and consequent compensation is the psychopathic condition associated with the disease and immeasurably increased by the boredom and anxieties associated with indefinitely prolonged unemployment."

Mackie says it takes thirty years for the average man to develop oscillations (a physiological adaptation but with potential risks in the event of loss of control). Caiger, quoting Healy, says: " At first the oscillations are a physiological psycho-optical reflex: they become eventually a pathological habit spasm." As nystagmus is unknown in India where the miners alternate mining with agriculture, so Mackie suggests that partial surface work should be carried out in a rotation of, say, one month on the surface to three underground. Also, he suggests that the name of this industrial disease should be changed.

Vestibular Nystagmus.—The movements of the eyes in this type of nystagmus are produced by disease of the labyrinth. If one labyrinth is injured or destroyed spontaneous rhythmic nystagmus towards the sound side is produced. If the other labyrinth is rendered functionless the movements now cease. In the normal individual vestibular nystagmus is elicited by rotating the subject on a revolving table, by syringing the ear with hot or cold water or by galvanisation of the ear. In all these cases the movements are rapid in one direction and slow in the opposite. The nystagmus is increased if the patient looks in the quick movement direction and diminished if he looks in the slow movement direction. The movements may be horizontal or rotary. It was Flourens who showed that extirpation of the labyrinth left the auditory sense intact but produced marked disorders of equilibration. He showed that

destruction of the horizontal canals produced continuous movement of the head from side to side in the plane of the injured canals. These reflex movements of head and eyes are due to the movements set up in the endolymph in the canals. Changes in the animal itself and not in its environment excite the nervous apparatus of the labyrinth. As Starling says, " We can assign it—the labyrinth—to the proprioceptive system. The resulting effect of the impulses arising in it is to maintain a reflex position of the head and eyes so that the optic axes in a position of rest are directed towards the horizon." Stimulation of the labyrinth therefore causes movements of the eyes which may or may not be associated with correlated movements of the head.

Nystagmus Due to Nervous Diseases.—Nystagmus is nearly always present in disease or tumours of the cerebellum ; nystagmus is also of assistance in the diagnosis of tumours of the cerebello-pontine angle. When the eyes are deviated from their normal position the nystagmoid movements are observed more especially if the deviation is towards the affected side.

In cerebellar nystagmus the movements show a quick jerk in the direction of the voluntary movements of the eyes and a slow return to the position of rest. The movements are horizontal, with a slight clockwise rotation on looking to the left and anti-clockwise on looking to the right. In bilateral lesions of the cerebellum there is more symmetry of the movements than in a unilateral lesion. In irritative lesions the slow motion is towards the affected side, but in destructive lesions they are away from it. Wechsler says, " While there are undoubted ocular, cortical and cerebellar complications of nystagmus the vestibular mechanism probably plays the ultimate roll in its production. The central vestibular nuclei mediate all the impulses, while the posterior longitudinal fasciculus is the final pathway to the ocular muscles."

In the early stages of cerebellar disease there may be a paresis of the ocular muscles, but this rapidly disappears, and the weakness, if present, allows a movement away from the affected side. I have not been struck by this symptom. Also " skew deviation," or a movement of the homolateral eye downwards and inwards, and of the contra-lateral eye upwards and outwards, is sometimes observed. A more constant symptom of cerebellar disease is the production of nystagmus when the eyes are deviated towards the affected side. The hypotonia of the muscles showing paresis may be the factor in

the production of the nystagmus, so that the nystagmoid movements are evidently due to an attempt by a strong nervous effort to maintain the normal steadiness of the muscles of the eye in the required direction. Cerebellar nystagmus can therefore be looked upon as a deficiency of tonus in the muscles which maintain the position of the eyes. Each side of the cerebellum is concerned with all the postures of the eyes, the movements towards that side being more strongly controlled than those which are away from the same side. The slow movement in the cerebellar nystagmus is probably controlled by the cerebellum, and the quick phase is most probably controlled by the mid-brain.

In multiple sclerosis a *conjugate nystagmus* is present in 12 per cent. of cases (Uhthoff). But Barany states that a *resilient or jerky nystagmus* is much more common. Nystagmus is a common symptom in encephalitis lethargica ; it is also present in syringomyelia, hereditary ataxia, Little's disease and idiocy. Uhthoff says nystagmus is present in about 100 per cent. of cases of cerebral abscess, but Taylor states that lesions of the cerebral hemispheres do not give rise to nystagmoid movements. Nystagmus is found in pontine and medullary lesions.

Acute nystagmus is found in certain intoxications such as alcohol. Other poisons, such as chloroform, ether, morphia, veronal, lead, etc., can produce nystagmus.

Hereditary and idiopathic nystagmus are not very common. The former occurs in males and may be associated with lack of pigment in the eyes and hair, while the latter may occur for apparently no reason in otherwise normal subjects.

The Course of Nystagmus.—In miners' nystagmus the apparent movement of things seen together with the vertigo which is present and the backward tilting of the head cause considerable discomfort. The condition clears up if the miner changes his occupation and environment. Many cases of ocular nystagmus persist through life, while the nystagmus due to nervous disease persists as long as the cause remains. In spasmus nutans and in hysterical forms nystagmus is not permanent, passing away in the former during the first two years of life.

Binocular Vision.—Cantonnet and Filliozat say that " binocular-vision is obtained by the exteriorisation in a single mental image of similar though not absolutely identical images formed on the retinæ of eyes symmetrical in attitude and acting synergetically

in their movements." When both visual axes are directed towards an object slightly dissimilar images of the object fall on corresponding points of the retinæ and both images are fused by the brain so that single vision results. Some have this fusion sense more fully developed than others. The writer, while testing the muscle balance of various patients, has seen cases where the eye movements have always been parallel, each eye possessing normal vision, and yet the maddox rod and the source of light or other double objects which are used in muscle testing could not be perceived simultaneously. Such cases with unequal errors of refraction, especially of the hypermetropic variety, readily become squinters unless the refraction is corrected early in life and training of the fusion sense resorted to.

Babies five or six months old learn to use their little spectacles which are tied round the head, and if from this early period the visual axes are maintained in parallelism fusion sense is gradually acquired.

A simple method of testing one's own binocular vision is to hold a pencil midway between a book which is being read and the face. If binocular vision is present the pencil then offers no obstruction to reading. If, however, words cannot be seen on account of the position of the pencil only one eye is being used.

In cases of squint which have not been corrected the image of the squinting eye is gradually ignored by the brain, and it therefore becomes what is termed an amblyopic eye. An ambylopic eye is therefore an acquired condition, and every effort should be made to prevent a child developing such. The sight of an amblyopic eye cannot be restored after seven years of age.

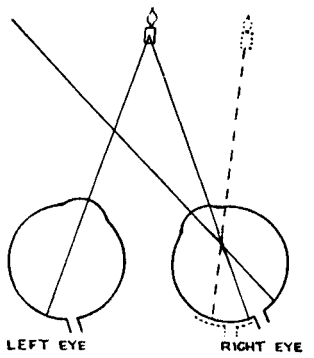


FIG. 29.—Paralysis of right external rectus.

Diplopia.—Should a sudden paralysis of an extraocular muscle take place in the case of a person with normal fusion sense two images will be seen. The image belonging to the eye with the attached paralysed muscle appears to be displaced or projected towards the paralysed side (see Fig. 29). For example, should there be paralysis of the right sixth cranial nerve then the right eye will converge, while the left eye is being directed straight forwards. The image belonging to the right eye will be on the right side of the

true image. This is known as homonymous double vision (see Fig. 29). Should, however, the eyes diverge as in a third nerve paralysis, then the images will be heteronymous or crossed (see Fig. 30).

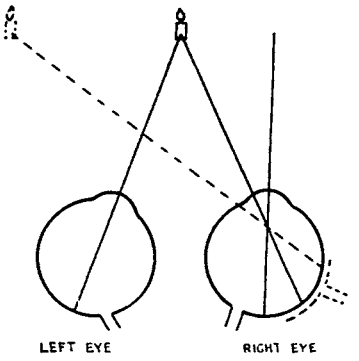


FIG. 30.—Paralysis of right internal rectus.

In order to determine which muscle is paralysed one can use the following method: Hold before the eyes an illuminated glass rod which is attached to an Ever Ready pocket torch (see Fig. 31), or a candle will do as well. At a distance of 16 or 20 inches from the face of the patient direct the patient's gaze first to the right then to the left, upwards and then downwards, asking him to say when he sees the rod or candle

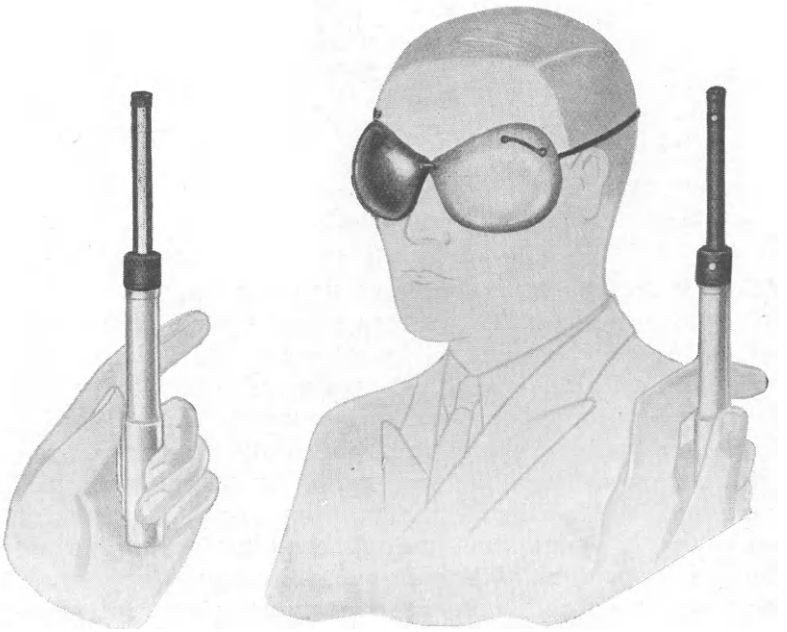


FIG. 31.—The Armstrong diplopia goggle and "Barlite." The eye-cups are coloured red and blue. The "Barlite" has two small illuminated holes opposite to and parallel with the line of light. The patient looks at the line of light and if diplopia is present the bar appears double, one bar coloured red and the other blue.

doubled. Suppose he is looking to the right and he says he sees two vertical images which are parallel to each other, at once cover the right eye asking him which image disappears. This will denote the image belonging to the right eye. If the image farthest to the right disappears, then there is paralysis of the right sixth cranial nerve, for the right eye is not rotated outwards. It should be remembered that the false image is always displaced towards the paralysed muscle. Many patients find it difficult to say which image is which ; therefore place before one eye, preferably the eye to which is attached the suspected muscle, a red disc of glass. The patient can now state quite easily which image disappears on covering one eye.

The following diagram is a modification of Werner's mnemonic scheme for demonstrating the diplopia produced by paralysis of the elevator or depressor muscles (see Fig. 32).

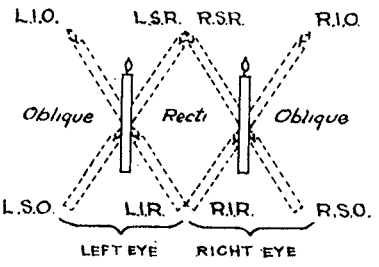


FIG. 32.—Werner's mnemonic for diplopia.

If the reader will keep in mind that the projection of the false image is in the opposite direction to the position of the eye he will then readily understand that the false image is displaced in the direction of the paralysed muscle. If the false image belonging to the right eye is higher than the image of the left then the superior rectus muscle of the right eye has failed to elevate the eyeball to its proper position. The case of the obliques, too, can be easily understood by reference to the above diagram. Suppose we wish to determine the nature of the diplopia produced by paralysis of the right superior oblique muscle, look at the diagram in its lower and outer part. There we see the false image is on the right and lower side of the true image ; it is homonymous. The upper end of the false image is inclined towards the lower end of the true image, this impression being obtained while looking downwards. The rotary movement of the oblique muscles is an outward one while the movements of the recti produce an inward rotation, therefore the false images in the diagram belonging to the recti form a lozenge-shape space between the two true images. As Swanzy has said : " The figures can be called to mind either as consisting of the four recti in the centre and the four obliques at the outsides, or as being made

up of a cross for each eye with the two recti on the inside and the two obliques on the outside."

The greatest displacement of the false images is obtained by looking most strongly in the direction of the paralysed muscle.

General Symptoms of Paralysis of an Extraocular Muscle.—When one of the muscles attached to the eyeball is paralysed strabismus or squint is produced. Then, secondly, the patient suffers from diplopia or double vision, which in the beginning is very nauseating, causing giddiness or unsteady gait. Patients quickly discover the relief given by covering one eye, especially the eye with the attached paralysed muscle. Thirdly, the secondary deviation is seen to be greater than the primary deviation ; as this is the important diagnostic point between a paralytic squint and a concomitant squint—a squint associated with an error of refraction and non-paralytic—it should be perfectly understood. Take, for example, a paralysis of the left external rectus. Cover the right eye with a lens, the lower half of which consists of ground glass. Movements of the eye can be observed from above through the upper plane, but the eye itself cannot see the object through the ground surface of the lens. Move the object to the left, requesting the patient to follow the movement. The left eye will not cross the middle line, but the right eye will be turned strongly towards the inner canthus, into such a position that if its visual axes were prolonged outwards it would pass the object on its left side. The attempt to move the left eye outwards requires such a considerable nervous impulse to be imparted to the paralysed muscle, the left external rectus, that the accompanying nervous impulse to its associated muscle, the right internal rectus, is greatly in excess of the natural amount required, and the right eye is moved too far inwards, or, in other words, an exaggerated movement inwards results.

In the case of a concomitant squint the primary and secondary deviations are equal.

Special Symptoms :—

Paralysis of the External Rectus :

Deviation of eye is inwards.
Outward movement limited.
Head is turned towards affected muscle.
Images are homonymous and in the same level.

Paralysis of the Internal Rectus :

Deviation of eye is outwards.
Inward movement limited.
Head turned towards affected muscle.
Images are heteronymous and in the same level.

Paralysis of Superior Rectus :

Deviation of eye is downwards and slightly outwards.
Upward movement is limited.
Head is inclined and rotated towards sound side.
False image is above, vertical separation increases in the upper field, and is greatest up and out.

Paralysis of Inferior Rectus :

Deviation of eye is upwards and slightly outwards.
Downward movement is limited.
Head is inclined and rotated towards affected side.
False image is above, vertical separation increases downwards and is greatest down and out.

Paralysis of Superior Oblique :

Deviation of eye is upwards and inwards.
Downward and outward movements limited.
Head is inclined downwards and outwards towards affected side.
False image is below and on the outward side, the images are homonymous.

Paralysis of the Inferior Oblique :

Deviation of eye is downwards and inwards.
Movement of eyes limited upwards and outwards.
Head is inclined upwards and outwards towards the affected side.
False image is above and on the outer side, the vertical separation increases upwards and inwards and the images are homonymous.

In addition to the various paralyses mentioned there are cases where one finds instead of a completely paralysed muscle a paresis or partial paralysis which may disappear or may develop into a complete paralysis. These cases are commonly seen as the result of accidents where a hæmorrhage has taken place into or around the

nerve supplying a muscle or a group of muscles. They are also seen in disseminated sclerosis and exophthalmic goitre.

Heterophoria.—A further division of muscular anomalies is that known as heterophoria. Part of the routine of a thorough examination of the refraction should include an examination of the muscle balance. Normally both eyes are held in a position of parallelism when looking at a point beyond 20 feet—this distance being commonly referred to as infinity—and the optic axes are parallel to each other. When the muscle balance is such that there is practically no effort required to maintain this position a condition of orthophoria exists. The tendency for the visual axes to deviate from such parallelism is known as heterophoria.

The heterophorias include *esophoria*, which is a tendency of the visual axes to converge ; *exophoria*, where the visual axes tend to diverge ; *hyperphoria*, where the visual axis of one eye tends to be directed higher than its fellow. Rarely the condition of *cyclophoria* is found, in which there is a rotary tendency due to insufficiency of the oblique muscles.

Heterophoria may be produced by the condition of the refraction ; esophoria is commonly present in hypermetropia ; while exophoria is seen most commonly in myopia. Other causes of this condition may be congenital. While operating to remedy the condition of squint the writer frequently finds a muscle showing such marked hyper-development that the only conclusion he can come to is that many of these conditions have existed since birth.

Examination of the Muscle Balance.—The simplest method of demonstrating the presence of heterophoria is to place before the right eye a Maddox multiple rod while the patient looks at a light, such as a candle flame, placed 20 feet away. When the Maddox rod is horizontal the image of the flame is converted into a vertical red bar of light and, as fusion sense cannot in any wise combine this image with that of the candle as seen by the left eye, the eyes at once tend to move into a position of rest. If the red bar of light moves to the right of the candle flame it indicates that the right eye has moved towards the left—this is the condition known as esophoria. Whereas, if the bar moves to the left of the candle flame the eyes are in a position of divergence—exophoria. Now turn the Maddox rod through 90 degrees so that the glass rods are vertical, and the patient will see the red bar of light on a horizontal plane. If the bar is lying below the candle flame it indicates that the right eye

has been rotated upwards—right hyperphoria. If the red bar has moved to a position above the flame it indicates left hyperphoria.

The method of measuring the amount of the heterophoria is to find that prism which will restore the red bar of light to the position it should normally occupy, namely, the centre of the candle flame.

There is a small margin of error of the muscle balance present in most people. Therefore, when a slight muscle imbalance is discovered it need not be thought that prisms must be prescribed. Experience alone teaches the oculist how much of the heterophoria should be corrected.

Worth has laid great stress on the lack of development of fusion sense as a contributory cause of heterophoria.

Conjugate Ocular Movements.—The evidence for the location of the centres controlling conjugate ocular movements has not been conclusively proved. Sherrington, Leyton and Risian Russell obtained conjugate movements on stimulation of the frontal lobes, Sir David Ferrier and Bernheimer on stimulation of the angular gyrus which lies behind the supra-marginal convolution. Conjugate movements have also been obtained by stimulation of the visual area in the occipital lobe (Schäfer). It is probable, however, that the movements produced by excitation of the occipital cortex are not willed and purposeful but are due to excitation by visual impressions reaching the cortex. Fibres may be traced from the occipital cortex to the mid-brain, also eye movements may be elicited by stimulation of the calcarine region after removal of the frontal centres.

It is, however, due to disease of the brain-stem that the most complete and permanent disturbance of conjugate movements of the eyes occurs (Holmes), limitation or loss of lateral movements to one side is the commonest form.

The suggestion that the lateral conjugate eye movements are carried out by means of fibres from the abducens nucleus was put forward by Duval and Laborde in 1880, but as Leonard Kidd points out, neither he nor many others, such as Van der Schueren, can accept the Duval-Laborde hypothesis. Van der Schueren, experimenting on rabbits, sectioned the third, fourth and sixth nerves of one side ; a few days later he found chromatolysis of all the cells of the homolateral sixth nucleus and none whatever in the contralateral sixth nucleus. If, as Leonard Kidd says, the sixth nucleus sent any fibres to the contralateral third nucleus or root, Schueren

would have found chromatolysis of some of the cells of the sixth nerve on the side opposite to his section of the third nerve.

As the nuclei of the abducens and oculo-motor nerves lie some distance from each other, it is necessary for them to be connected in order that a harmonious conjugate movement be brought about; this is done through the posterior longitudinal bundle and all its connections. There is evidence of the presence of degenerate fibres in the posterior longitudinal bundle in all those cases examined pathologically, of loss of lateral conjugate eye movements which were accompanied by preservation of the action of the contra-lateral internal rectus muscle in convergence.

But there is a further suggestion that lying in front of and ventral to the abducens nucleus there is a supranuclear centre, and into it come all the impulses that can excite conjugate movements of the eyes to the same side, voluntary impulses that come from the opposite frontal centre, impulses from the occipital and temporal lobes which effect reflex adjustments of the eyes to visual and auditory stimuli, others from the lower visual and auditory reflex system in the roof of the mid-brain, impressions of vestibular origin from Deiters' nucleus and probably proprioceptive impulses from the muscles of the neck. It is probable that through this centre, too, the antagonists of the contracting muscles are reciprocally inhibited. This supranuclear mechanism has the physiological property common to all motor mechanisms above the level of the final common paths in that it is concerned in the production of movements and not in exciting the contraction of individual muscles (Holmes). An example of a lesion in this region which can dissociate lateral conjugate movements is seen when a patient cannot obey a command to look to one side but can follow a slowly moving object in this direction.

Collier states: "There does not seem to me to be any adequate reason for postulating 'supranuclear centres' situated in the brain stem for the bilateral co-relation of eye movements, since interruption in the downcoming path from the cerebrum would have a precisely similar effect as would the destruction of the hypothetical supranuclear centres" (see also Mushen's work).

There are four possible modes (Collier) in which lesions of the brain stem can produce ophthalmoplegia. (1) By interrupting only the supranuclear path by which incitations descend from the cerebrum to the oculo-motor nuclei as in lesions situated at the junctions of the thalamus with the superior colliculus, or when a lesion involves

the posterior commissure which is probably an important decussation of the supranuclear path, since in one of Collier's cases a lesion of one-half of the posterior commissure caused complete loss of upward and downward movements of both eyes ; (2) by involving the cells of the oculo-motor nuclei alone, as in chronic progressive neuronic degenerations akin to progressive muscular atrophy, and also the acute neuronic affections which have been named peripheral neuritis of the eye muscles, of which diphtherial ophthalmoplegia, tetanus ophthalmoplegia and ophthalmoplegia met with in polyneuritis, myasthenia and in botulism, are examples. Collier also says : " The ocular paralysis of poliomyelitis should be placed in this group, since like the facial paralysis in this disease it seems invariably to recover." (3) By involving the intramedullary portion of the peripheral oculo-motor nerve. (4) By a gross lesion, involving any combination of the supranuclear path, nuclear cells and intramedullary peripheral nerve.

Gordon Holmes has stated that from the study of clinical and post-mortem cases it can be said that in the upper end of the mid-brain in the neighbourhood of the anterior quadrigeminal bodies are association centres which control the conjugate vertical movements of the eyes. He believes that a lesion of the anterior end of the mid-brain will involve upward movements, downward movements and convergence in this order as it progresses from before backwards ; also that the paralysis of the pupil is often associated with loss of upward movement, while loss of convergence is associated with paralysis of downward movement.

This agrees with Winkler's statements regarding the function of the Edinger-Westphal nuclei (see p. 54).

In supranuclear paralysis of the ocular muscles the ocular axes remain parallel in contradistinction to nuclear paralysis, which is usually accompanied by diplopia owing to convergence or divergence of the ocular axes produced by paralysis of an individual muscle or muscles. But it must be remembered that ptosis may be met with as an isolated phenomenon in certain supranuclear lesions.

A brief review of oculogyric spasm, better known as spasm of lateral conjugate deviation of the eyes, is given by Davison and Goodhart in which they correlate oculogyric with cephalogyric movements. They say the entire corticomesencephalic mechanism of binocular turning of the eyes to the right was called by Grasset the *dextro-oculogyric apparatus*, and that of turning the eyes to the

left the *lævo-oculogyric apparatus*, so that "each hemisphere sees and looks to the opposite side."

The following table after Bielschowsky is a comparison of the features of conjugate deviation in cases of lesions of the hemispheres and in cases of pontine lesions :—

Comparison of the Features of Conjugate Deviation in Cases of Lesions of the Hemispheres and in Cases of Pontine Lesions.
(Bielschowsky—"Motor Anomalies of the Eyes".)

Lesions of the Hemispheres	Pontine Lesions
1. In the first stage, regular and of considerable magnitude.	1. Relatively rare and as a rule of small magnitude.
2. Deviation usually of short duration.	2. Deviation, if present, permanent.
3. Deviation toward the side of the lesion.	3. Deviation towards the opposite side.
4. Deviation frequently a symptom of stimulation.	4. Deviation usually a paralytic symptom, only rarely a stimulation symptom.
5. Head turned (as a regular symptom) in the same direction as the deviation of the eyes.	5. Abnormal position of the head not a typical symptom; if present, the head usually turned in the opposite direction to the deviation of the eyes.
6. Associated paralysis of the muscles for contralateral movement, usually slight and transient.	6. Associated paralysis in the direction of the lesion, nearly always severe and permanent.
7. Invariably symmetrical functional disturbance of the associated muscles.	7. Frequently asymmetrical paralysis of the associated muscles in consequence of the extension of the supranuclear lesion to the nucleus or the nuclei.
8. Paralysis of the extremities and of the facial nerve collateral with the associated eye muscle paralysis.	8. Paralysis of the extremities, if present, opposite to the side of the eye muscle paralysis; paralysis of the facial nerve, if present, usually collateral with the eye muscle paralysis.
9. In lesions of both hemispheres all eye movements (including the vertical) restricted or impossible.	9. In pontine lesions of both sides, paralysis of side-to-side movements without disturbance of the vertical movements.

Bielschowsky described a form of strabismus which resembles a trochlear paresis where, as he pointed out, there is no difference in the angle of primary and secondary deviation and no reduction in the ability of the upward-squinting eye for motion downwards and there is an extension of the fields of vision (Vestergaard). It is most probably due to some co-ordination disturbance of a supra-nuclear nature.

CHAPTER IV

PAPILLŒDEMA

MEDICAL practitioners and neurological students should become familiar with the ophthalmoscopic appearance of the retina. Plate II (Fundus of the Eye) illustrates the normal appearance of an average eye as seen in the white races of mankind. The long axons originating from the ganglion cells of the retina, after running in the nerve fibre layer, proceed along the optic nerve to the brain. As these fibres converge from the retina they are heaped up slightly at the margin of the optic disc to form the optic papilla. In the centre of the nerve head is a depression of varying depth known as the physiological pit, at the bottom of which can usually be seen the fibres of the sclera constituting the lamina cribrosa.

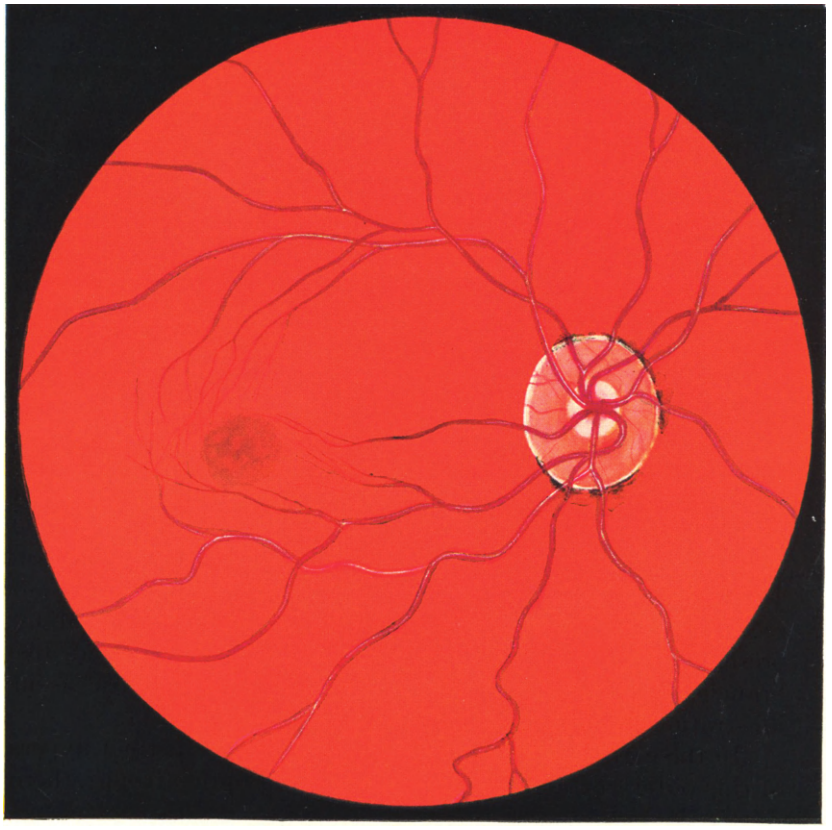
From the illustration it can be observed that the outer portion of the optic disc is not so deeply coloured as the adjacent part of the fundus, but not so pale as the physiological pit in its centre. The colour of this portion of the nerve head or optic disc should receive the utmost attention. The colouring is produced by the presence of minute blood vessels. Several small but distinct branches may often be observed passing over the disc as in the illustration.

In the early stages of papillœdema there is a distinct hyperæmia of this outer portion of the nerve, while in optic atrophy there is a decided pallor. It is largely due to the presence or absence of these minute vessels that a diagnosis from the normal condition is made.

On the right side of the optic disc on Plate II, is some choroidal pigment, which may, however, form a complete ring round the disc or may be entirely absent. The arteries emerging from the disc are lighter in colour than the veins passing into it. Where the arteries and veins cross each other there is no kinking, as is commonly found in arterio-sclerosis. To the left of the optic disc and situated at the posterior pole is the normally pigmented macula.

The old term *optic neuritis* ought to be largely discarded. Where there is a swelling of the disc due to raised intracranial pressure the term *papillœdema* should be employed. This differs

PLATE II (R. Lindsay Rea)



THE FUNDUS OF THE NORMAL EYE

On the right is the optic disc with some choroidal pigment around its edge. The surface of the optic papilla is paler in colour than the surrounding retina. In the centre of the papilla is the normal physiological pit which is much paler in colour than the surrounding surface and, at the bottom of the papilla, the lamina cribrosa is faintly seen. The arteries are lighter in colour and are seen emerging from the depths of the physiological pit. The veins are darker in colour. To the left is the normally pigmented macula. Note that where the arteries cross the veins there is no kinking.

[*To face p.* 84.

essentially from the true neuritis due to retrobulbar disease and from that seen in neuro-retinitis. In papillœdema the changes are primarily in the disc itself; in true optic neuritis the swelling of the disc is due to inflammation behind the disc and the vessels involved at the nerve head, while in neuro-retinitis the primary cause is in the retina, and the inflammatory changes resulting produce the œdema of the disc. In papillœdema there is a simple non-inflammatory œdema of the nerve head.

It must be clearly understood that by the term papillœdema a pressure symptom alone is meant; neither the optic nerve nor the retina is in a state of inflammation, when the disc is swollen as a result of such inflammation then the term *true optic neuritis* must be employed.

Should a papillœdema due to raised intracranial pressure persist, the optic nerve will slowly atrophy, with consequent loss of vision. Therein lies the greatest danger of delaying a decompression operation on the skull. If the pressure produced by papillœdema has been allowed to continue, say, for two or three months, a consecutive optic atrophy is almost inevitable. Frontispiece illustrates a case of papillœdema produced by a tumour of the the frontal lobe.

The appearance presented by such a fundus when seen for the first time is extremely puzzling. The vessels appear disconnected; disc surface and retina are confused, but gradually the whole picture dawns on the observer. The interruptions in the appearance of the continuity of the vessels are due to exudates covering the latter. The edge of the optic disc is completely obscured; the vessels come forward in a swollen manner from the centre of this broad area; the pressure on the nerve is transmitted to the veins, so that the blood does not freely pass through them and they therefore present a swollen appearance. Over and around the disc are fine exudates and small hæmorrhages. These are confined to the surrounding region of the optic disc, and do not follow the vessels for any distance from the nerve head.

Papillœdema occurs in 80 *per cent. of intracranial tumours* (Paton). In tumours of the medulla oblongata and pons papillœdema is not commonly found, nor if the tumour is confined to the white matter of the brain, the highest percentage being found with tumours of the mid-brain, parieto-occipital region and cerebellum. The depth of the papillœdema varies from a slight swelling to that equal to 5 or 6

dioptres. I have seen this amount exceeded. Others have observed as much as 8 dioptres in extent. Three dioptres are equivalent to a swelling of one millimetre of the nerve head.

Frontal tumours produce a severe papillœdema, as shown in the frontispiece. *Tumours of the parietal region* usually show a mild papillœdema which does not, as a rule, persist for any length of time. *Tempero-sphenoidal* tumours, like those of the frontal region, show papillœdema of marked amount, as also do tumours of the *optic thalamus and mid-brain.* I have always been impressed by the appearance of a severe papillœdema in the early stages of tumours of the *cerebellum.* Owing to the anatomical structure found in the posterior portion of the cranium *subtentorial tumours* produce a severe form of papillœdema, while those occurring above the tentorium do not produce such marked swelling of the disc.

The state of the vision in papillœdema varies greatly. In the early stages there may be no disturbance of function, full central visual acuity, full fields of vision, while measurement of the blind spot by the scotometer may show no departure from the normal. If secondary changes do not take place while the papillœdema is subsiding vision will not be affected. Unfortunately, however, great destruction of vision may ensue by the pressure on the nerve fibres at the optic disc. Loss of central vision or the gradual loss of the whole field of vision may take place. In a subject showing papillœdema produced by basal gummatous meningitis, it is quite possible to bring about resolution of the disease by treatment; yet in many instances, in order to preserve intact the visual pathway, it is absolutely necessary to decompress the skull for the relief of intracranial pressure while treatment is still being vigorously carried on during and after the operation.

De Schweinitz drew attention to the fact that sometimes, due to the œdematous state of the nerve head, the blind spot is found to be enlarged before the swelling of the disc is evident. Gowers already had noted the increase in the size of the blind spot in some cases. Yet one has observed that even in the presence of a well-marked macular fan there may be no interference with central vision although the œdema has spread as far as the perimacular region.

Concentric contraction of the field of vision begins to take place as the result of atrophy of the nerve head from pressure. In the majority of cases the contraction proceeds in a regularly concentric manner until central vision is finally involved. This is the direct

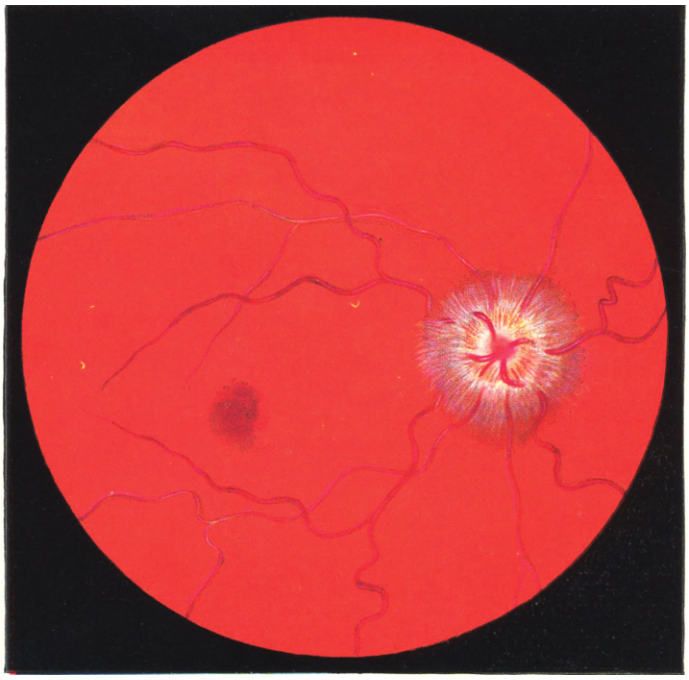


FIG. 1. PAPILLŒDEMA

The optic disc red and swollen is seen protruding into the vitreous. The edge of the disc is obscured by œdema and fine hæmorrhages; these remain in the vicinity of the disc and do not follow the vessels in their course across the retina. The veins are swollen and the arteries are reduced in calibre and at the disc edge both arteries and veins are obscured thus losing their continuity. The physiological pit is filled in; the remainder of the fundus is normal.

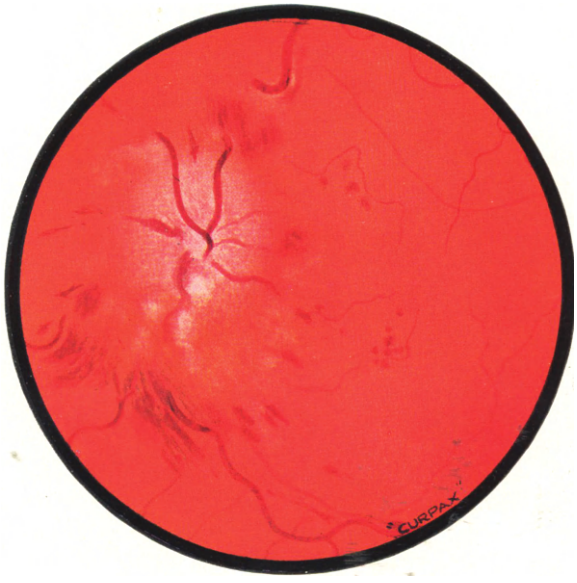


FIG. 2. PAPILLŒDEMA

Papillœdema due to a tumour of the cerebellum. The disc edge cannot be recognised, the continuity of the vessels is lost, hæmorrhages and white exudates obscure the disc and surrounding retina. The condition indicates a more acute stage than Fig. 1. (Case in charge of the late Dr. E. Macnamara).

[*To face p.* 86.

opposite to what occurs when a local nerve lesion is present. In the latter case central vision is usually the first to disappear.

It must be borne in mind that as Hughlings Jackson first pointed out, " even when papillœdema is marked, vision may be unimpaired —acuity and colour vision being perfect and the field unrestricted."

During the state of raised intracranial pressure the papillœdema produced is nearly always bilateral, but need not be of equal amounts. A papillœdema on one side with a slowly developing optic atrophy on the other is indicative of a frontal lobe tumour, also of tumours in the pituitary region.

When papillœdema is found in one eye only it may indicate the early stages of raised intracranial pressure or the appearance may be due to changes within the orbit such as tumours or cellulitis of the orbit, inflammatory or hæmorrhagic changes.

If there is the slightest suspicion that vision may suffer from the presence of a papillœdema, decompression should be proceeded with immediately. One sees swelling of the nerve head from 4 to 6 dioptres becoming greatly reduced during the few days following the operation. One need not fear severe deformity of the head as the result of this operation. Many patients who are in perfect health and have had this operation performed upon them have a perfectly normal appearance, together with full vision.

The student of neurology may find a pitfall lying in his pathway while attempting to see the fundus through a pupil which is only moderately dilated. To him the edge of the disc appears indistinct and he cannot see the vessels on the surface of the disc clearly. He has probably forgotten to look at the pupil itself with a + 20 lens, for if he had done so he would have noticed there the presence of early lens opacities or, in other words, incipient cataract through which the whole fundus appears indistinct.

In order to measure accurately the amount of swelling of the nerve head the student must learn to relax voluntarily his own accommodation. To prove that he can do so he should be able to see instantly the double image of a pencil held 8 inches from his face while looking at a wall or a ceiling beyond. The same effort must be made while looking at the surface of the disc. The examiner, keeping his accommodation in abeyance and focussing the light on the vessels of the surface of the disc, rotates the convex lenses to the highest number with which he can still see distinctly the highest vessel. Suppose he then rotates a higher convex lens before his eye

the edge of the vessel will appear slightly blurred. He will now return to the lens which gave him a distinct view of the highest vessel on the surface. Noting the number of this lens he will now direct his attention to the vessels of the fundus situated some distance from the edge of the disc. To see these sharply he must reduce the number of the lens in the ophthalmoscope and stop at that number which gives the clearest view of these vessels. Subtracting the latter number from the former gives the height of the papillœdema in dioptres. It should be remembered that *three dioptres are equal to* 1 *millimetre of swelling.*

Plate III is inserted in order to show that papillœdema affects only the nerve head and the immediate surrounding portion of the the retina. There may be both hæmorrhages and exudates found on the surface of the disc and on the retina around the disc, but these are not found accompanying the vessels beyond this circumscribed area, so that when examining such a fundus with the ophthalmoscope it is not difficult to see clearly a fine vessel situated, say, above or below the macula. If the beam from the ophthalmoscope is turned from the disc to the fine vessels just mentioned they will appear indistinct until the convex lenses are reduced in strength. Say that in the first instance the surface of the disc has been clearly seen with a + 8D lens and then the fine vessels above or below the macula are seen with a + 2D lens, subtraction shows that the swelling due to the papillœdema is equivalent to + 6D. It takes considerable practice to do this accurately. In every case examined the height of the swelling should be measured as carefully as possible.

The following changes take place in the development of papillœdema: (*a*) There is an increase in the calibre of the veins passing to the nerve head. Sometimes large and even tortuous veins are seen in a normal eye, but when these are associated with any symptom of disease, either of the nervous system or of the health generally, they should be carefully observed. (*b*) The veins continue to show dilatation while the disc head shows a deepening of the red colour, together with slight blurring of the upper or lower nasal margin. (*c*) The œdema on and around the nerve head begins to appear and the nasal and temporal edges of the disc become blurred, so that the entire sharp edge of the disc is lost. (*d*) Now there is a decided increase in the elevation of the surface of the disc. The œdema increases and minute hæmorrhages form, while the physiological pit

PLATE IV (R. Lindsay Rea)

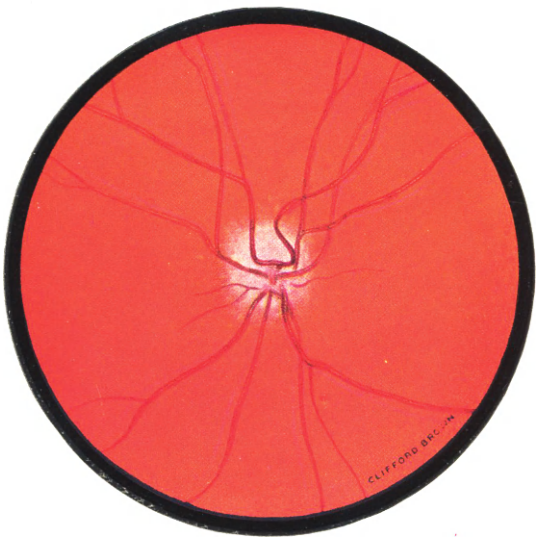


FIG. 1. SECONDARY OPTIC ATROPHY

The disc is pale in colour but not so white as in a primary optic atrophy. The disc edge is blurred. The physiological pit has disappeared. In this particular case the state of the papilla was secondary to a retrobulbar neuritis of the optic nerve close behind the eyeball. At no time was the swelling of the disc severe.

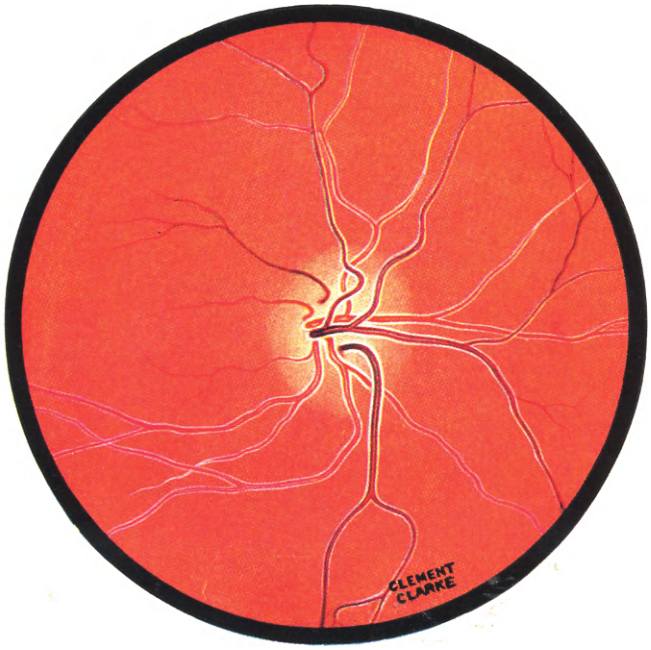


FIG. 2. CONSECUTIVE OPTIC ATROPHY

This case of a blind woman who suffered from an intracranial tumour (probably a tuberculoma) in childhood, shows a bilateral consecutive optic atrophy. The fundus of the right eye demonstrates most beautifully the white lines accompanying the vessels peripheralwards. These are not seen in primary optic atrophy.

[*To face p.* 88.

is beginning to be filled in. (*e*) In the most acute stage, such as is shown in Plates III and IV, all semblance of the nerve head is lost, and to a beginner the picture is most puzzling, only portions of the vessels lying on a whitish background, together with minute hæmorrhages on this area, being seen. However, by changing the lens in the ophthalmoscope from the higher to the lower numbers, the levels of the various vessels can be perceived. The optic disc has protruded itself forward into the vitreous and, as the vessels pass from the disc and turn backwards over its edge, they are observed to be partly obscured by the œdema and hæmorrhages.

Above is the picture of the development of a papillœdema. It may not always pass to an acute stage ; there are conditions in which a papillœdema in both eyes has never gone beyond stages (*c*) and (*d*) and has remained so for years. These are commonly seen in mental hospitals, and are due to permanent changes in the disc.

If the pressure in the nerve head is not reduced atrophy of the optic nerve follows. The appearance presented by a consecutive optic atrophy is illustrated by Plates VII and VIII. It will be seen that the edge of the disc has lost its sharp appearance. The physiological pit has been filled in. White lines, which are thickened perivascular sheaths, are found accompanying the vessels for a varying distance, while slight pigmentary change of a diffuse nature is seen around the disc. These are the sequelæ to the inflammatory disturbance produced by the pressure at the nerve head.

Primary Optic Atrophy	**Consecutive Optic Atrophy**
(*a*) Sharp edges.	(*a*) Blurred edges.
(*b*) Physiological pit present.	(*b*) Physiological pit filled in.
(*c*) Normal choroidal pigment as in Plate II.	(*c*) Finely scattered irregular pigment.
(*d*) Sheaths of vessels invisible.	(*d*) Sheaths of vessels visible as white lines accompanying the vessels as far as the equator of the eyeball (see Plate VIII).

Plate IV, Fig. 2 (Consecutive Optic Atrophy), is the drawing of the right fundus of a woman who has been blind since childhood. It shows the presence of the perivascular sheaths to a remarkable degree. More commonly Plate IV, Fig. 1, is the appearance found after the entire subsidence of a papillœdema.

Causation of Papillœdema.—Originally it was believed by von Graefe that papillœdema was due to pressure on the cavernous sinus, causing venous status in the central vein of the retina and the consequent condition of swollen optic disc he termed "Stauungspapille," a term translated by Clifford Allbutt as "choked disc." But Sesemann demonstrated that the anastomosis between the orbital and facial veins was so free that the effect of pressure on the cavernous sinus was at once relieved and did not cause more than a transient dilatation of the retinal veins, and other observers such as Hutchinson and Uhthoff have pointed out the extreme rarity of papillœdema in cavernous sinus thrombosis. After Schwalbe's discovery that the sheath of the optic nerve could be injected and distended by injection into the subdural space around the brain, many theories arose to account for the presence and appearance of papillitis. Schmidt-Rimpler suggested that the intracranial pressure may distend the optic sheath, thus causing a swelling of the intraocular portion of the optic nerve. Further, he injected a coloured liquid into the nerve sheath and found it passed into the lymph spaces in the nerve at the lamina cribrosa. This led him to believe that the irritation of fluids passing into these lymph spaces was responsible for the neuritis which followed. Manz supported Schmidt-Rimpler's theory, but Leber and Deutschmann opposed the theory of the Stauungspapille from the distention of the nerve sheath. They believed the swelling of the nerve head was due to pathogenic material being conveyed to the optic nerve behind the eye. Benedikt formulated the theory, which was held also by Hughlings-Jackson, that intracranial tumour causes optic neuritis by acting as a foreign body in the brain, ascribing the mechanism to the vasomotor nerves, a mechanism, however, that is not known to exist.

Parinaud thought the optic neuritis and swollen nerve sheath were part of the cerebral œdema produced by the distention of the ventricles of the brain. Gowers, Edmunds and Lawford believed that optic neuritis was a condition which followed a descending inflammation of the optic nerve or sheath, although Gowers admitted that distention of the sheath of the nerve alone is probably sufficient to cause papillary œdema by its mechanical effect and may intensify the processes otherwise set up, especially if the fluid is of an irritating quality. Cushing and Bordley, upholding the mechanical theory, believed that acute œdematous swelling of the disc followed the injection of fluid under considerable pressure into the cranial

subdural space of dogs, but Wolff and Davies could not confirm the experiments. The latter injected dyes into the cranial subarachnoid space at pressures which are compatible with life, and found they did not enter the optic nerve, also the theory of the production of papillœdema by injecting fluid into the cranial subarachnoid space at pressures compatible with life is not upheld by their investigations. Dupuy-Dutemps and Paton and Holmes have shown that when papillœdema occurs the vein within the subarachnoid space of the optic nerve is flattened and thus much narrowed.

Based upon the various measurements of the intraocular venous and arterial pressures, Lauber concludes that the determining factor in the development of swelling of the optic discs and the accompanying phenomena is the raised diastolic venous pressure and its increase in relationship to the diastolic retinal arterial pressure. Levy records a case where papillœdema occurred in an eye as the result of low intraorbital pressure alone.

Pathology.—The pathology of this condition has been described by Paton and Gordon Holmes. They have shown that papillœdema is a venous congestion of the optic disc and surrounding retina caused by compression of the central vein as it passes from the optic nerve across the subarachnoid space of the optic nerve sheath. Cerebrospinal fluid passing down the subarachnoid space under increased pressure from the cerebral subarachnoid space is accountable for the pressure on the vein. There is little change in the artery, but the outflow of blood and lymph is affected and the œdema of the nerve head is thus produced (see Fig. 81, p. 215).

Parsons, the first to introduce the term papillœdema, has also contributed to the pathology of this condition. The blurring and swelling of the disc is due to an œdema of its tissues and the anterior layers of the lamina cribrosa. In a microscopical section new formed neuroglia can be seen. It is this which gives the chalk-like appearance in the post-neuritic atrophy. The proliferation of neuroglia finally squeezes the nerve fibres out of existence. Contraction of the neuroglia flattens the disc head and may leave an irregular edge to the disc. It is in this region that newly formed pigment is sometimes found. The physiological pit is filled in by the œdematous fluid under the internal limiting membrane. The swelling of the disc head has pushed the retina slightly aside. The nerve fibres of the temporal part of the retina are gathered into masses which are clumped together above and below the disc, due

to the internal limiting membrane being lifted up into small vesicles, and it is here that the first blurring of the disc edge takes place. The fibres forming the papillo-macula bundle are short and curved and cannot accommodate the œdema. In some cases, therefore, a macular fan, consisting of fine white lines running in a semi-curved direction between disc and macula, is produced, but this is not obvious in the majority of cases. The fundus generally, apart from the area involved, presents a normal appearance. The changes taking place produce enlarged veins with distended capillaries. The hæmorrhages are mainly found at the edge of the disc (see Plate III), and are flame-shaped or drop-like.

Although the changes taking place in papillœdema are not truly inflammatory, Paton and Gordon Holmes occasionally found small-celled infiltration in the connective tissue sheath of the vessels. Possibly this was merely a reaction to the intense and destructive changes produced. There is a slight congestion of the anastomosis between the choroidal and retinal vessels at the edge of the disc. They have also pointed out that the inflammatory process is secondary to the degeneration, but the papillœdema is not primarily an inflammation. In long-continued œdema the nerve fibres swell and become varicose, breaking up into degenerative masses called cytoid bodies. The perivascular tissue becomes thickened and finally is changed into connective tissue. This change produces the white lines accompanying the vessels as seen in Plate IV, Fig. 2.

Illustrated in the " Histopathology of the Eye," by Adalbert Fuchs, are two microscopic sections, one of optic neuritis and the other of papillœdema. In the former, which was the result of an endophthalmitis following an injury to the eye, the vessels in the optic papilla are engorged with blood and ensheathed with lymphocytes. The papilla itself contains numerous cells and the granular layers of the retina remain at the edge of the scleral canal. In the latter illustration, that of a papillœdema, the result of a cerebral tumour, the nerve head is extremely swollen and its vessels dilated. The nerve in front of the canal stains lighter than that portion of the nerve behind the lamina cribosa, and the retina is pushed away from the side of the canal by the swelling of the nerve fibres (hence the enlarged blind spot observed in the field of vision in many of these cases of papillœdema). The lymphocytic infiltration seen in the former is absent in the latter, also in the latter the cerebro-spinal fluid is indicated as a faintly staining liquid lying in the

PLATE V

FIG. 1. OPTIC NEURITIS. (Magnified 20 times)

Hypopyon followed an injury to the eye twelve days previously. Perception and projection of light had become defective.

The vessels in the optic papilla are engorged with blood and ensheathed by lymphocytes (periphlebitis). The papilla itself contains numerous cells. The infiltration (J) accounts for the ill-defined edge of the optic disc in the ophthalmoscopic picture of optic neuritis.

The swelling of the disc surface is insignificant; the two granular layers of the retina begin strictly at the scleral canal.

In this case the neuritis forms a part of the endophthalmitis and is caused by the toxins of the bacteria in the vitreous body.

FIG. 2. PAPILLŒDEMA OR "CHOKED DISC." (Magnified 20 times)

The patient, a young man, was suffering for some length of time from attacks of sudden obscuring of vision when stooping rapidly or turning the head. The ophthalmoscope revealed a marked papillœdema. The patient died from a cerebral tumour.

The swelling of the disc surface is extreme, and its vessels are dilated. The central vein is filled with blood but the branches of the central artery are empty. The thick optic nerve passes through the narrow sclerotico-choroidal canal changing its colour at the lamina cribrosa. In front of it the nerve stains much lighter since the nerves are here devoid of their medullary sheaths. The particularly light staining of the nerves is due to their separation by œdema. The nerve fibres push the margin of the retina (R) away from the scleral canal, forming the neuritic swelling (N) upon the choroid. In contrast with optic neuritis (Fig. 1) the vessels are not ensheathed with cells.

The vaginal space is filled with a faintly red staining liquid (H) which pushes the dural sheath away from the pial sheath revealing the trabeculæ of the arachnoid sheath. The vaginal liquid being the cerebrospinal fluid, contains little albumen hence its poorly staining property. The increased intracranial pressure forces the cerebrospinal fluid along the vaginal space and in addition to stemming back the blood in the central vein may also help to push the optic nerve into the scleral canal.

The swelling of the nerve head has removed the margin of the retina from the scleral canal, accounting thus for the enlargement of the blind spot in cases of papillœdema. (Both Figs. 1 and 2 are kindly lent by Adelbert Fuchs and are published in his "Atlas of the Histopathology of the Eye.")

[*To face p.* 92.

PLATE V

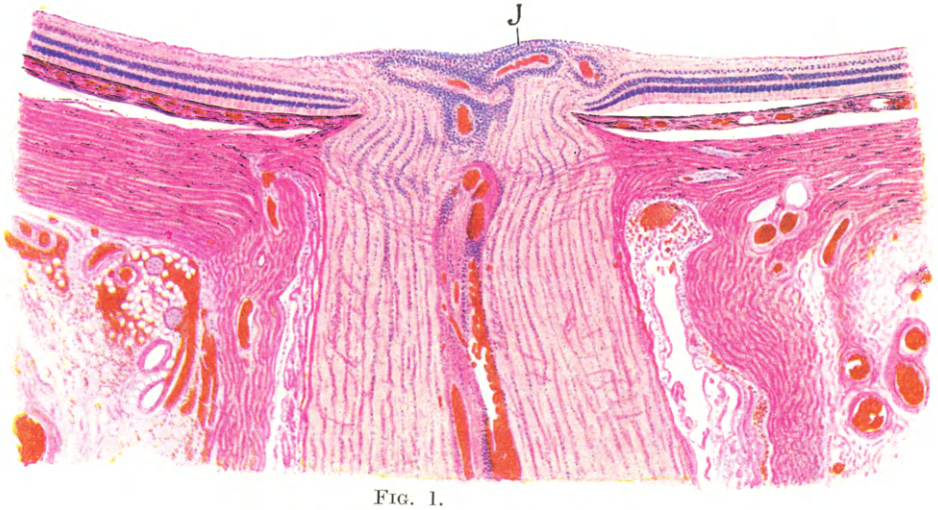


FIG. 1.

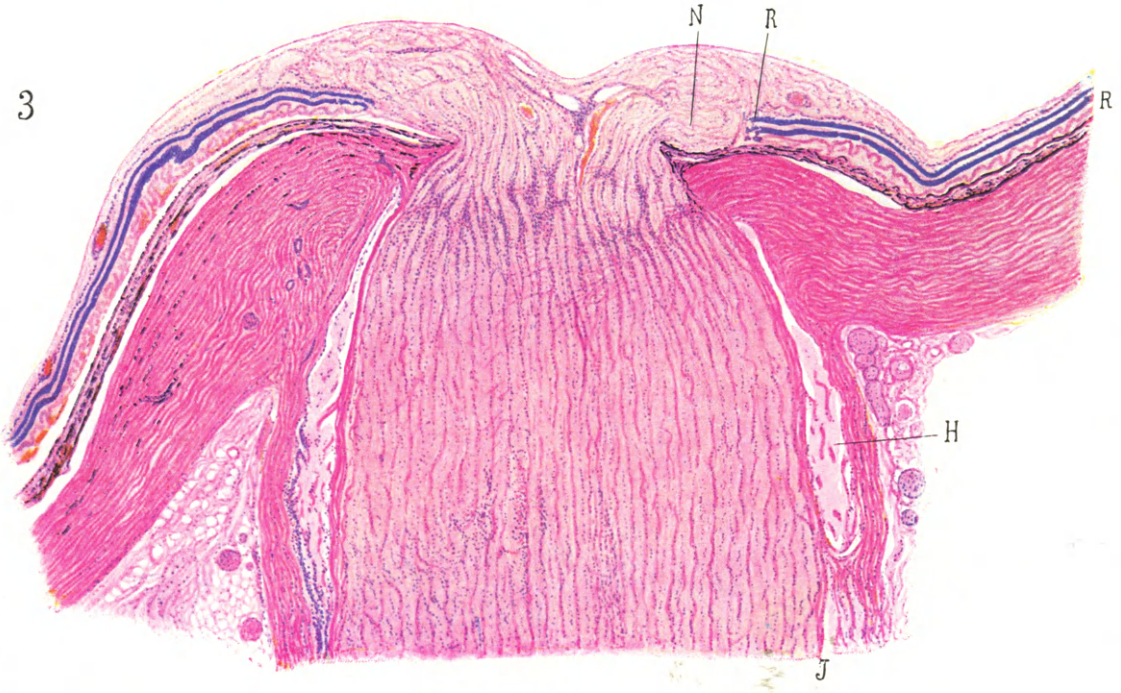


FIG. 2.

intervaginal space, being pressed there by the raised intracranial pressure (see Plate V, Figs. 1 and 2).

Etiology.—Intracranial disease is the commonest cause of papillœdema. The remaining causes are extremely numerous. The factors concerned, as will be shown in the following chapters, include those due to heredity, congenital malformation of the skull, meningitis, especially the tuberculous form of this disease, (posterior basal meningitis is seldom accompanied by papillœdema), inflammation produced by syphilis, etc., aneurysms and hæmorrhages. Papillœdema is sometimes found in the acute and infectious diseases, including the exanthemata. It has been seen in malaria (Maxwell).

It should be remembered that after a severe hæmorrhage swelling of the disc may be found. Also chronic papillitis may be seen in certain individuals, particularly in the insane, although the writer has seen similar cases in individuals who appear perfectly normal both physically and mentally. Toxic conditions, whether endogenous or exogenous, may sometimes be found to be the cause of this condition. Renal disease and severe generalised arteriosclerosis can produce well-marked forms of papillœdema. Nettleship and Priestly-Smith have recorded cases in which a papillœdema has coincided with persistent discharge of watery fluid from one nostril. This fluid by itself might indicate inflammatory disease in the superior meatus or adjacent bony cavities, but if this discharge should prove to be cerebrospinal fluid then raised intracranial pressure or hydrocephalus is present.

Uhthoff has given the following table of statistics which he has observed of papillœdema :—

	Per cent.
Brain, tumour of	71
Tuberculosis	3·6
Cerebral syphilis	12
Tower skull	2·2
Brain abscess	2·2
Hydrocephalus and serous meningitis	2·2
Meningitis	1·1
Nephritis	1·1
Nephritis and lead poisoning	0·3
Anæmia	0·9
Lead poisoning	0·3
Retrobulbar optic processes	0·3

Both Marcus Gunn and Gowers believed that the state of the refraction of the eye could affect the incidence of a papillitis. Gunn thought that myopia had a retarding effect, while Gowers believed that papillœdema occurred more frequently in hypermetropic than in emmetropic eyes. Observations by many observers, including those of the writer, do not agree that such is the case.

A problem presented itself to the author in dealing with the following case. The patient, a woman sixty years of age, was suffering from intense headache which seemed to be associated with her eyes. Upon examination of the latter, extremely raised intraocular tension was found in the right eye and both pupils were so small that it was difficult even in the left eye to see the state of the discs. One dared not dilate them in the presence of such raised tension; the next day the right eye was trephined with immediate relief of headaches, and now through the dilated pupil of the right eye the edge of the disc was seen to be indistinct. On the seventh day the patient was up and, free from all pain, was preparing to leave the nursing home the next day, but in the morning she awakened with severe headache and gradually passed into coma, remaining in that state all day. Dr. Worster-Drought, on examination, diagnosed the condition as a hæmorrhage into a metastatic carcinoma in the brain; the primary growth was a breast tumour which had been removed a year previously, this removal had been somewhat tardily undertaken. The following day the patient died. The problem was—could the raised intracranial pressure produce raised tension in the eye? We know from experiments conducted by Henderson, Hill, Fremont-Smith, Forbes and Duke-Elder that whenever absorption does not keep pace with filtration there will be an accumulation of fluid in the tissue spaces recognised in the elastic skin and subcutaneous tissues as œdema. In the cranium and eye, however, which are rigidly enclosed, this results in an increase in pressure—hydrocephalus or glaucoma—for intraocular and intracranial pressure may be said to be the result of a delicate balance between the hydrostatic and osmotic pressure of the capillary and venous plasms within the eye and cranium. Could blood suddenly liberated and mixing freely with the cerebrospinal fluid raise the intracranial pressure and also the intraocular, at the same time accompanied by such intense cephalagia as was seen in this patient?

The *pseudo-papillœdema* of some hypermetropic eyes may cause difficulty in diagnosing true papillœdema. Although in pseudo-papillitis the appearance of blurring of the edge of the disc is seen, the disc edge is sharply defined. The surface of the disc is not raised and, if due to a variation of the depth and position of the physiological pit the vessels are found to curve over as if the edge of the disc were raised, yet the vessels passing over the remainder of the disc edge are seen to be on the same level. However, there are cases where the most competent observer finds difficulty in deciding whether the appearance is that of a true or pseudo-papillœdema. He will carefully watch from time to time for any increase in the calibre of the veins, for any reddening of the colour of the disc or for an increase in its elevation or enlargement of blind spot.

What seems to present the greatest difficulty to many students and practitioners is the appearance produced by the presence of optic nerve fibres which have preserved their medullary sheath. Plate VI (Opaque Nerve Fibres) illustrates such a case. Where these medulated fibres are present the disc loses its sharp edge and the vessels disappear and reappear from the mass of white tissue, so that there is a very definite resemblance to papillœdema. It will be noticed, however, that the surface of the nerve head is not elevated, that the white area around the disc is not uniform, and that there is an entire absence of œdematous or hemorrhagic changes on or around the disc. One must readily admit the student's difficulty in looking at such a fundus. When such a case as that shown in Plate VI, occurring in a myope, was shown to a large post-graduate class, not one of the nineteen members of the class diagnosed the condition. Therefore in demonstrations of the fundus oculi it is wise, if at all possible, to include a case showing opaque *nerve fibres.*

Recurrence of Papillœdema.—It is indeed rare to see a case of recurrence of papillœdema. Various writers have reported cases from time to time. The writer can recall only one case of recurrence in ten years' experience at a large hospital for nervous diseases. In Gower's "Medical Ophthalmology," p. 55, 4th edition, a case of Hughlings Jackson's is quoted in which a boy aged twelve years had double optic atrophy following intracranial disease some years previously and now was found to have distinct double papillœdema with symptoms of internal tumour. De Schweinitz and Thomson, in relating their case, point out that the second attack may

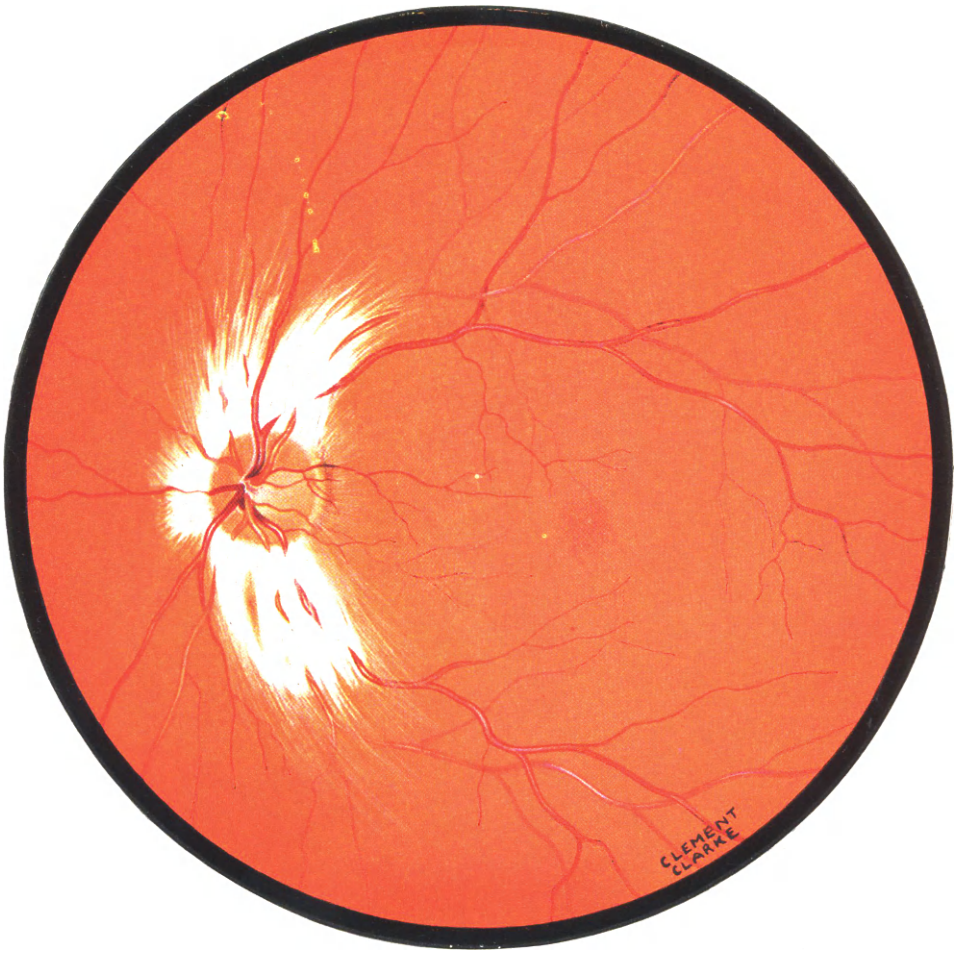
destroy any vision left after the primary papillœdema has passed away.

Papillœdema or Consecutive Optic Atrophy associated with persistent Escape of Cerebrospinal Fluid.—Some cases have been reported where there has been a persistent or intermittent dripping of fluid from the nostril, associated with diminished vision (due to swelling of the optic disc or a consecutive atrophy), epileptiform fits, vomiting and drowsiness, delirium and weakness of lower extremities, but headache was the most common symptom. The fluid proved identical with the cerebrospinal fluid. If the fluid should occasionally cease to flow, the symptoms increased. In 1899, St. Clair Thomson related a case of this kind and collected reports on twenty others. Lieber's case, which he quotes, proved to be one of internal hydrocephalus. The fluid probably escapes from the sub-dural space in contiguity with the olfactory nerve-endings. In many of these optic atrophy was present, and in some double optic neuritis. The cerebral symptoms varied from headache to giddiness, drowsiness, vomiting, delirium and convulsions. Post-mortem examination of four cases pointed to hydrocephalus and not cerebral tumour. In one the fluid escaped through an almost imperceptible hole in the cribriform plate beside the crista galli. Schwalbe (also quoted) demonstrated a physiological communication between the subarachnoid space and the lymph channels of the nose.

The amount of discharge varies greatly. In one case it actually amounted to three quarts a day. The affection commences in early adult life and may persist for a long period or may eventually cease. Meningitis has been a terminal cause in some cases.

Prognosis in Papillœdema.—When raised intracranial pressure is responsible for the presence of papillœdema, the condition should not be allowed to remain longer than a few months. Good vision is compatible with a marked papillœdema, but by perimetric examination it is found that after the papillœdema has subsided considerable injury has been done to the optic nerve. Paton has stated that if during the stage of the papillœdema a diminution in the calibre of the arteries is observed, then atrophic changes are taking place in the nerve, but this cannot be applied universally. The main point is not to delay decompression as soon as such is indicated. Those cases in which vision has been lost after a decompression were probably due to a further rise of intracranial pressure following severe secondary hæmorrhage.

PLATE VI (R. Lindsay Rea)



OPAQUE NERVE FIBRES

This peculiar appearance which in many cases strongly suggests a papill-œdema is due to the presence of the myelin sheath of the nerves in the retina which normally ends behind the lamina cribrosa. Rarely these white patches are seen separated some distance from the optic disc, but when, as in this picture, the white area more or less surrounds the optic edge the retinal vessels are seen to disappear and appear again at the edge of this area, giving the impression that the vessels are obscured by œdematous exudates. The medullary nerve-sheath areas are always characterised by the brushlike extremities.

[*To face p.* 96.

CHAPTER V

OPTIC ATROPHY

THE term **Optic Atrophy** is applied to the condition of the nerve head when degeneration has taken place in the fibres of the optic nerve. The natural colour of the disc becomes lighter in tone as age advances, but this does not signify in any wise that a degenerative change has taken place ; but when increasing pallor of the disc is associated with a defect of central vision or of the visual field it may then be looked upon as a true atrophy of some of the optic nerve fibres. In *disseminated sclerosis* it is the fibres of the papillo-macular bundle which suffer most, so that pallor due to degeneration of these fibres is found frequently on the temporal side of the disc. The pallor may be due to empty capillaries, or narrow vessels, most of which normally cannot be seen by the naked eye.

In a partial optic atrophy of disseminated or multiple sclerosis the temporal half of the optic disc which contains the papillo-macula bundle is markedly white, due to absence of these small vessels. It is often difficult to decide whether the temporal pallor in these cases is real or whether it is due to an enlarged and elongated physiological pit. Even in this latter condition the small vessels on the temporal side of the disc should be looked for. If atrophy is present these minute vessels will not be seen.

In both literature and practice it will be found more convenient to employ the terms *primary optic atrophy* and *consecutive optic atrophy*. The former condition has not been preceded by any inflammatory or pressure changes, while the latter is consequent upon a papillœdema. *Secondary or post-neuritic atrophy* is the term reserved for those cases of optic atrophy following a true optic neuritis, *e.g.*, inflammation of the retina and optic nerve or inflammation of the optic nerve alone.

Primary Optic Atrophy	Consecutive Optic Atrophy
(*a*) Sharp edges.	(*a*) Blurred edges.
(*b*) Physiological pit present.	(*b*) Physiological pit filled in.
(*c*) Normal choroidal pigment as in Plate II.	(*c*) Finely scattered irregular pigment.
(*d*) Sheaths of vessels invisible.	(*d*) Sheaths of vessels visible as white lines accompanying the vessels as far as the equator of the eyeball (see Plate VIII).

In primary optic atrophy there is always loss of vision in a greater or lesser degree, and as there cannot be regeneration of the nerve fibres the state of the vision cannot improve. In secondary (post-neuritic) optic atrophy this statement does not hold good. One has seen a case showing an atrophied disc with entire absence of vision, yet, as the result of anti-syphilitic treatment, central vision rose to 6/9 and the extreme limits of the field of vision returned, but between the two was a large horseshoe-shaped scotoma. This recovery has persisted now over six years. It was not obvious in this case that the atrophy was secondary, and so the diagnosis of primary optic atrophy might readily have been made, the implication being that no measure of sight would be recovered. These cases are seen most frequently in syphilitic subjects. It has happened that a quack who has been shrewd enough to know the value of mercurial treatment in syphilitic disease has brought about what he calls a cure to the discomfiture of those who had stated that a cure was impossible.

The case just described, together with a similar one recorded by Adler, of recovery of the central visual acuity might be placed in the category described by Cushing as being caused by " physiologic block." When this was relieved the function of the fibres which were not destroyed recovered.

A comparison should be made of Plate II (Fundus of the Eye) and Plate VII (Primary Optic Atrophy). In the former note the presence of the vessels on the surface of the disc, while in the latter they are completely absent or invisible. The disc in the normal fundus has a soft pink tint and may contrast with the whiteness of the central physiological pit ; whereas in primary optic atrophy the disc surrounding the physiological excavation may appear equally white or

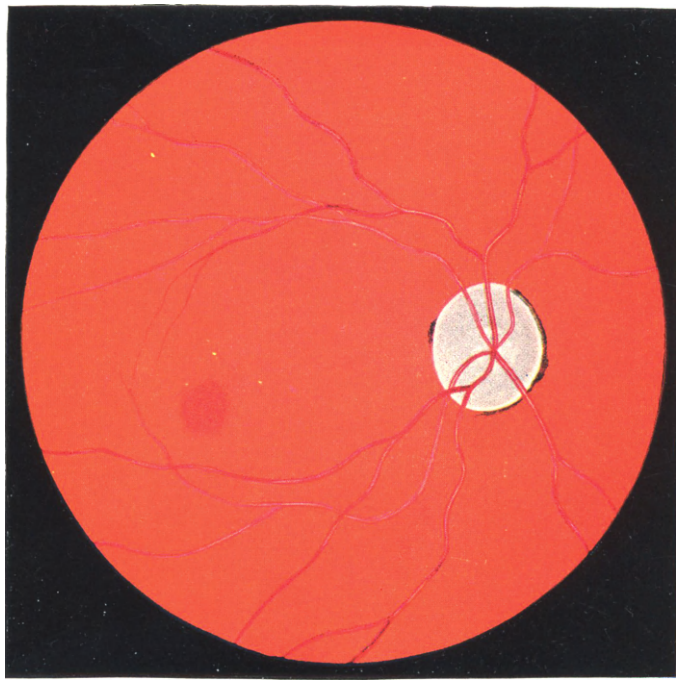


FIG. 1. PRIMARY OPTIC ATROPHY

The optic disc is greyish-white with sharp edge and a normal choroidal ring. Frequently the colour of the disc in primary optic atrophy is of a marble whiteness. The physiological pit is present and the size of the vessels is normal or slightly reduced in calibre. The remainder of the fundus appears normal. When pigment is present in the retina surrounding the disc due to inflammatory changes it is never so black nor definitely outlined as is the normal choroidal pigment.

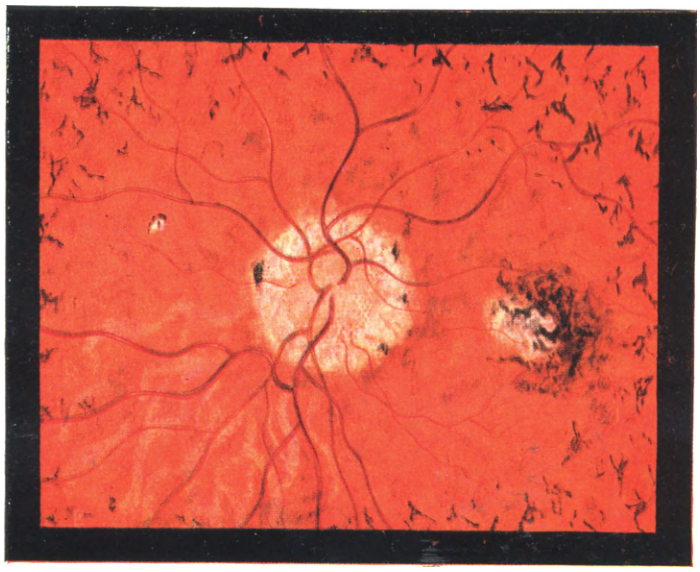


FIG. 2. RETINITIS PIGMENTOSA

The retinal vessels are diminished in size and the disc is slowly becoming atrophied, remaining yellow in colour and waxy-looking. Towards the equator the characteristic "bone-corpuscle"-shaped pigment is present, which is superficial and is observed to lie on the vessels. In this particular case some macular degeneration was present also, but is not usual.

[*To face p.* 98.

even grey. At the bottom of the pit, if such is present, the lamine cribosa will be seen (see Fig. 36).

McIntosh, Fildes and others say that primary optic atrophy can occur as a sign of parasyphilis alone or combined with primary degeneration of certain columns of the spinal cord.

In disseminated sclerosis there is destruction of the nerve elements, due to constriction produced by the proliferation of the supporting structures. Other diseases, such as tabes, may show a similar destruction, while a certain degree of sclerosis in the optic nerve, even some distance removed from the ball, can produce a degeneration of this nerve with a consequent optic atrophy. If a lesion takes place at any point between the optic disc and the external geniculate body degeneration proceeds not only from this point on the cerebral side, but also on the ocular side. If the ganglion cells of the retina are destroyed optic atrophy follows.

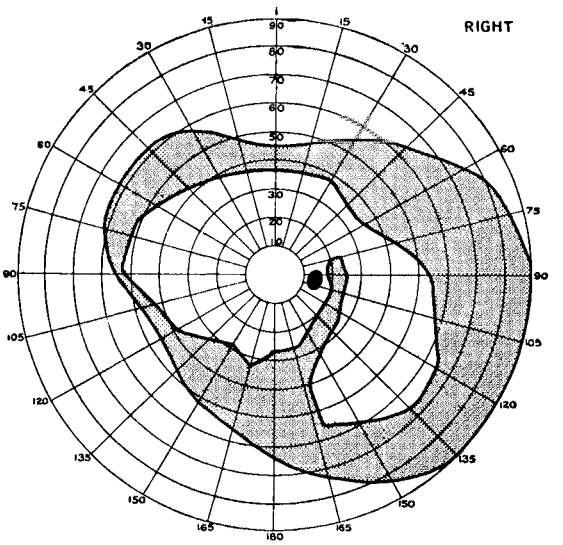


FIG. 33.—Field of vision in a case of tabo-paresis. One year after onset of optic atrophy.

It has been shown experimentally by Usher and Dean that a wound of the retina results in the degeneration of a corresponding tract of fibres in the optic nerve.

Etiology.—One of the chief causes of primary optic atrophy is "*locomotor ataxia*" (*tabes dorsalis*), in which both eyes are affected, but often in an unequal degree. In this disease the first symptom of the onset of optic atrophy is disturbance of dark adaptation, while colour sense and normal visual fields are still present. Later the disc begins to whiten. There is now a concentric diminution of the visual fields and a partial loss of colour vision. At this stage patients may ask how long will it be before the onset of complete blindness. It

usually takes about two years for complete optic atrophy to ensue.

It should be noted that the red and green fields disappear before the blue. In simple or non-congestive glaucoma where the disc sometimes strongly resembles primary optic atrophy the loss of colour fields does not occur until late in the disease.

The contraction of the field of vision in tabes dorsalis usually begins on the outer side, and the narrowing of the field is by no means uniform (see Figs. 33 and 34). There is both a parenchymatous and an interstitial change in the optic nerve. If the parenchymatous change predominates there is great loss of visual

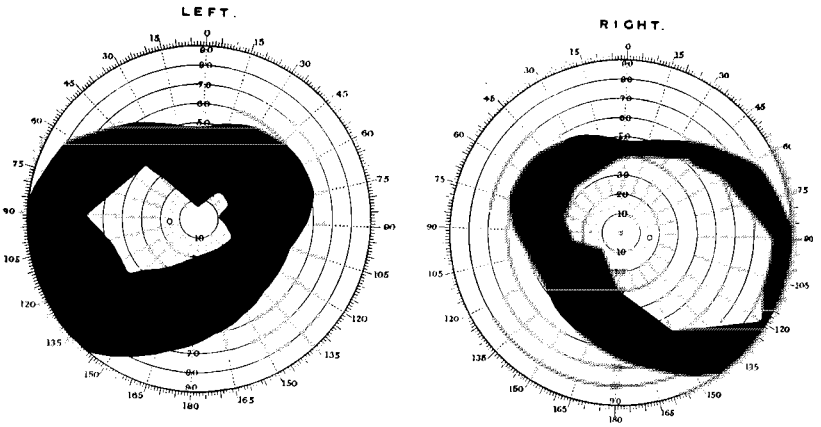


FIG. 34.—Fields of vision in a case of tabes. Pupils were unequal and neither pupil reacted well to light. The left optic disc showed a further degree of primary optic atrophy than the right. Central vision in left eye = 6/9, while that in the right = 6/5. (From Leslie Paton's paper on Tabes and Optic Atrophy.)

function and the reaction in the nerve is trivial while the degeneration is extensive; but where the interstitial changes predominate the reaction—proliferation and infiltration—is marked while the degeneration is slight. The parenchymatous tissue has little or no recuperative power, and therefore degeneration in it means loss of function.

Uhthoff says that the process producing optic atrophy commences in a fatty degeneration and absorption of the myelin sheaths, followed by changes in the axis cylinders. These first show varicosities and subsequently also degenerate, leaving only the fine threads of the collapsed nerve fibre sheaths. In the later stages all

that is left is fatty drops, myelin drops and amyloid bodies. The changes in the interstitial tissue follow on these parenchymatous degenerations, the finer septa and processes of supporting tissue disappear, and the larger septa become more swollen. It is only in the later stages that glial proliferation comes into evidence. For a full discussion on this subject I would refer the student to Paton's paper in the *British Journal of Ophthalmology*, 1922. In this paper he concludes from clinical examinations that in the case of tabes dorsalis, especially by the evidence of the visual fields, there are indications that the primary lesion is in the neighbourhood of the chiasma and is both parenchymatous and interstitial, and does not begin in the ganglion cells of the retina.

Primary optic atrophy occurs also in disseminated sclerosis, diffuse cerebral sclerosis, general paralysis, amaurotic family idiocy and acute myelitis optica of Devic. Occasionally it is found in Friedreich's ataxia, Schilders's disease, congenital diplegia, cerebellar atrophy, perineal muscular atrophy, paralysis agitans, pituitary disease and herpes ophthalmicus. Optic atrophy may be post-influenzal or it may follow Malta fever and blackwater fever. (Paton.)

Primary optic atrophy may also be due to menstrual disorders, exposure to cold and the toxic action of certain drugs. Chesterman has seen many cases of primary optic atrophy caused by the use of arsenic compounds in the treatment of sleeping sickness ; while in the earlier days of the treatment of syphilis by arsenic some cases were reported. To-day both the chemical composition of these arsenical compounds and the method of their administration are so much better understood that in the London Lock hospitals primary optic atrophy, as an evidence of arsenical poisoning, has not been seen during the past ten years. Lead poisoning may show either primary optic atrophy or an optic neuritis. Severe hæmorrhages and syphilis are also causes (Wright states that in India secondary optic atrophy produced by syphilis is the commonest variety of optic atrophy met with) ; diabetes, congenital deformities of the skull, alcohol and tobacco—any one of these may be the ætiological factor in the production of primary optic atrophy.

Cases have been reported by Gowers, Terrien, Renard, Singer, Hawthorne, Whiting and others where optic atrophy has followed severe hæmorrhage from the alimentary canal or uterus. It is rare to see such a sequel to the extensive hæmorrhages of the battlefield

or of everyday accidents, but many have been recorded after hæmatemesis. Singer states that blindness appears :—

During the hæmorrhage in . .	8·3	per cent	of cases
Immediately after hæmorrhage .	11·6	,,	,,
In 12 hours	14·2	,,	,,
Two days after	19·2	,,	,,
Between 3rd and 16th days . .	39·2	,,	,,
After the 7th day	7·5	,,	,,

In some cases the sight appears to improve slightly but usually the vision becomes worse. Treacher Collins thought the consequent optic atrophy followed on a primary ischæmia of the retina of profound degree but Terrien adds a toxæmic cause, the action of which prevents the recovery of the cells affected by severe loss of blood.

Optic atrophy may be the result of fracture of the base of the skull. Rawling has shown that the region including the optic foramen may suffer from fracture of the skull in any and every direction. Of von Hoelder's 88 cases of fractures of the base 90 per cent. involved the orbit and 61 per cent. the optic canal. (Quoted by Margoline.) The nerve may be torn, pressed upon, or an extravasation of blood into its sheath may take place. A common form of injury to-day is seen resulting from a fall on the frontal region from off a motor cycle. There is instantaneous blindness of one eye. The patient may not show severe concussion and fracture may not be suspected while often the X-ray plate is negative. Six weeks later the optic disc in the blind eye may be noticed becoming white.

Margoline compiled statistics of optic atrophy observed in Prof. Haltenhoff's clinic, and in 44 the lesion resulted from trauma. Of these, 12 were due to direct lesion of the nerve, 19 were due to indirect lesion with immediate loss of sight, 8 cases due to indirect lesion with late loss of sight, while the remaining 5 could not be classified. These delayed cases may be due to contusion of the nerve, formation of callus, hæmorrhage, traumatic meningitis in the neighbourhood of the optic foramina, atrophy consecutive to a traumatic papillitis, which is rare, but what are termed late post-traumatic atrophy of the optic nerve are, in all probability, due to a serous meningitis with the formation of a chronic arachnoiditis (see p. 416).

Hereditary Optic Atrophy (Leber's Disease).—This disease com-

mences usually at the age of twenty and occurs more frequently in men than in women. It is a form of retrobulbar neuritis and is transmitted usually through an unaffected mother to the child, generally a male. Direct inheritance is less common than collateral. Both eyes are always affected, the affection coming on either rapidly or gradually, with failure of central vision. The central scotoma may be partial for colours and later becomes absolute for white and colours, and generally one eye is attacked days or months before the other. The peripheral field as a rule is normal, but occasionally contraction defects of either a sector or concentric shape may occur. After the central scotoma has become absolute the condition usually remains the same. Occasionally there is an improvement in the sight, but more rarely complete blindness ensues. At first there are no changes seen in the retina ; then a slight blurring of the edge of the disc is observed, followed by optic atrophy. The atrophy of the temporal half of the whole disc may slowly develop without any blurring of the edges of the disc. It has been suggested that Leber's disease can be regarded as an abiotrophy due to deficient vital energy of the structures involved, so that they are unable to maintain their nutrition beyond full development and therefore gradually degenerate. On the other hand, Fisher has suggested that Leber's disease is due to transitory changes in the pituitary body, resulting in pressure upon the chiasma about the age of puberty.

When Leber's disease occurs in women it develops not infrequently at the menopausal period.

This disease was first described in 1871 by Leber, who collected 55 cases. According to Favier, the youngest patient hitherto seen was a child, aged six years, and the oldest were aged fifty-two and sixty-seven. The visual disturbance occurs suddenly and develops rapidly in four or five weeks, when it becomes more or less stationary. General disturbance may occur in the form of migraine, vertigo, vomiting and sometimes epileptic attacks. Up to the present all treatment has proved ineffective.

A pedigree of congenital optic atrophy embracing sixteen affected cases in six generations has been described by Thompson and Cashell. The opinion was expressed by Nettleship that the exceptional cases of congenital familial optic atrophy recorded in literature were probably instances of a typical Leber's disease, but Thompson points out certain differences between those he describes and the typical cases described by Leber. Thompson's cases are : (1) Con-

genital. (2) Sexes are equal whereas in Leber's cases the males largely predominate. (3) Inheritance is through an affected

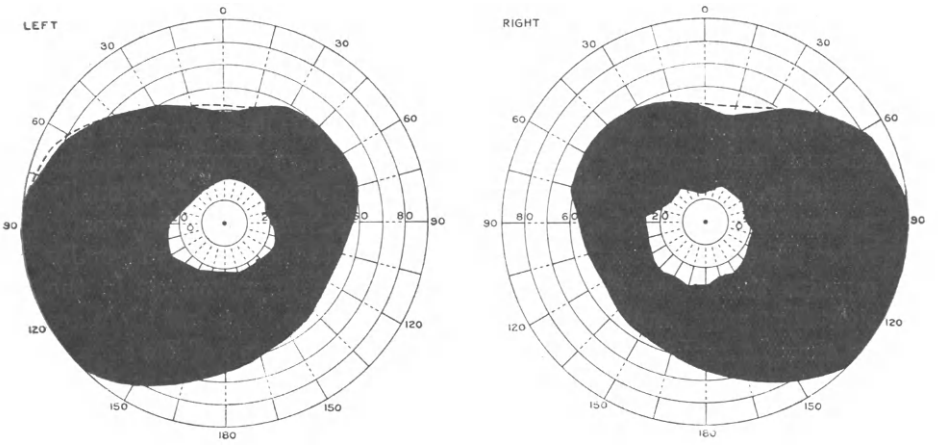


FIG. 35.—Reduced fields of vision in a case of congenital optic atrophy. Central vision—6/24 in each eye. (Willoughby Cashell.)

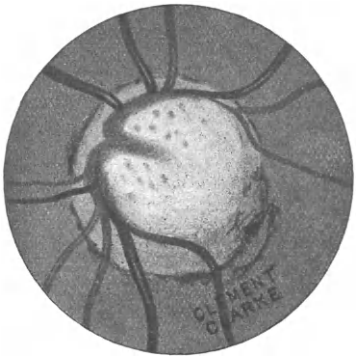


FIG. 36.—Primary optic atrophy showing a white disc with a deep, glaucomatous-like pit at bottom of which are seen the markings of the lamina cribosa. (Thompson and Cashell.)

mother, never through an unaffected mother or through the father; in Leber's cases it is through an unaffected mother. Therefore, in the former, inheritance is dominant, in Leber's cases it is recessive and sex-linked. (4) Prognosis. In Thompson's cases the condition appears to be stationary, similar to Leber's disease. (5) Optic discs are white with clearly defined physiological pitting, but instead of a scotoma being present as in Leber's disease, there is peripheral contraction (see Figs. 35 and 36).

In all of these cases except the two youngest in the sixth generation, nystagmus has been either constant or intermittent. Cashell said he could find no other abnormality in the central nervous system. Thompson mentioned Griscom's cases, where the disease began in early childhood, but the inheritance was through an affected father.

Amaurotic Family Idiocy (Tay-Sachs' Disease) (see Chap. VIII).—In this disease the primary change is in the nerve cells of the nervous system and retina. It will be more fully described in a later chapter ; it is but necessary to mention here and now the ophthalmoscopic appearance of the fundus. The cherry red macula, surrounded by a circular white opaque area of the retina, is characteristic of this disease. The cherry red macula, however, observed in embolism of the central artery is not so deep in colour as that of amaurotic family idiocy. In the later stages of this disease the optic disc gradually loses its normal colour, becoming quite white while the retinal arteries show marked constriction (see Plate IX).

Microscopical observation of the neurons of the body shows progressive loss of the Nissl granules and an increase of the neuroglial material to an abnormal degree. There is a decrease in the nucleoprotein of the brain, together with an increase of simple protein. The change taking place in the ganglion cells of the retina is similar to that which takes place in the ganglion cells elsewhere. It is this change which causes the opacity in the retina and finally leads to atrophy of the nerve fibres (see p. 148).

Retinitis Pigmentosa.—The first symptom of which a patient suffering from this disease complains is *night blindness*, that is, difficulty in seeing in a dim light. This night blindness, or *hemeralopia*, is found in two hereditary diseases of the retina—*retinitis pigmentosa* and *retinitis punctata albescens.* The disease is an extremely chronic and progressive degeneration of the choroid, beginning in childhood and often ending in blindness in middle or advanced life. The night blindness precedes the appearance of pigmentation in the retina by several years. The nutrition of the outer layers of the retina fails early, so that when atrophy of the rods and cones has taken place, and they have disappeared, the pigment epithelial cells of the retina pass through gaps left in the membrana limitans interna. The pigment cells become grouped in the perivascular sheaths, giving rise to the branching appearance of these cells as seen through the ophthalmoscope. The degeneration commences near the equator of the eye and spreads both anteriorly and posteriorly. The macula is not affected until very late in the disease. The fields of vision gradually contract and, although central vision remains normal for a long time, patients find it difficult to move about owing to the fact that they have what is called tubular vision, that is, the effect on vision produced by looking down two long cylinders, one being

placed before each eye. Such a person standing near a chair will feel first for the seat before sitting down. Examination of the fundus shows the typical spider-like black spots lying near the retinal vessels, often anterior to them, so that the vessels may be partly obscured in places. As the retina becomes more and more atrophic the pigmentation increases, the ganglion cells are destroyed, and this leads to degeneration of the nerve fibres passing from the ganglion cells to the optic disc. Optic atrophy, therefore, follows, but the disc does not show the typical whiteness of a primary atrophy ; it is of a waxy or yellowish appearance due to an overgrowth of neuroglia in its vicinity, while the retinal blood vessels become very small (see Plate VII, Fig. 2).

Finally, a posterior cortical opacity appears in the lens which increases slowly in density. The opacity is central and is therefore an obstacle to light entering the eye, further diminishing the already reduced visual acuity. Doyne believed the removal of the lens improved the general condition of the eye, but in reality the improvement is due to the greater amount of light now entering the eye.

Retinitis pigmentosa, typical and atypical, is found in the Laurence-Moon-Biedl syndrome (see p. 197).

Two types of retinitis pigmentosa were differentiated by Wibaut—one, a dominant type in which there are no associated lesions ; the other, the recessively inherited variety, is usually accompanied by deafness or other nervous lesions.

In the Laurence-Moon-Biedl syndrome macular vision is affected in some cases, while others show the ring scotoma typical of the state of the field of vision in early retinitis pigmentosa.

The retinal condition is sometimes associated (Zondek) with changes in the diencephalon such as are found in juvenile amaurotic idiocy and the Laurence-Moon-Biedl syndrome.

The cause of retinitis pigmentosa is not known. Although not a congenital affection, there is a marked tendency to it (familial), as is evidenced by the large number of cases which are hereditary. The writer has lately examined a boy showing the commencement of this disease. His father and uncle became blind from it, as did his grandfather. This disease occurs frequently in the offspring of consanguinous marriages. It is not syphilitic, although syphilitic retinitis may closely resemble it. It occurs in families where there is a history of nervous disease. Nettleship wrote of three of his cases, many of whose relatives were epileptics, insane or mentally deficient.

Gowers, to whom we are indebted for the term abiotrophy, has said that the optic nerves frequently suffer from abiotic wasting, and it is to this class of disease that retinitis pigmentosa belongs. De Schweinitz describes several cases where, in addition to polydactylism and pigmentary degeneration of the retina, there were definite symptoms of pituitary dysfunction.

When Horner first described the syndrome now known by his name, he drew attention to the presence of dilatation of the retinal veins on the affected side. This has brought forth the suggestion that in such cases as those of retinitis pigmentosa, where the vessels are contracted, it might be possible to effect a certain degree of dilatation by the operation of cervico-thoracic ganglionectomy. Wagener, in 1931, studied 20 cases of Raynaud's disease, 10 with Buerger's disease (thrombo-angeitis obliterans) and several others, all of which had the operation of bilateral ganglionectomy done for relief of their symptoms. The retinal vessels were measured by Morgan's retinal graticule both before and after operation. Of these 35, dilatation of the arteries occurred in 34, bilaterally in 18 and unilaterally in 16. The report of his cases has suggested to some surgeons that by the performance of such an operation as cervico-thoracic ganglionectomy retinitis pigmentosa could be cured.

Royle, in 1932, did report some improvement in the condition of 6 cases, stating that in some the field of vision was found to be enlarged. In the " Surgery of the Sympathetic Nervous System " Abel quotes Royle and even mentions that night blindness disappears. A perusal of the pathology of the condition of retinitis pigmentosa will easily convince the reader that mere dilatation of the retinal vessels could not replace destroyed rods and cones, dissolve new-formed interstitial neuroglia and replace migrated pigment cells. The structure of the vessels in retinitis pigmentosa shows hyaline degeneration and their lumen tends to become obliterated. Such changes are secondary, not primary (Collins, Mayo, Wolff and Verhœff). Night blindness precedes the appearance of pigment in the retina by some years. If any improvement could possibly be obtained by such an operation it would have to be done in the most initial stages of the disease. However, there are cases of amaurosis fugax (temporary blindness) associated with Raynaud's disease, also cases of hemeralopia (night blindness) associated with vitamin A deficiency, arterio-sclerosis, acrocyanosis (White), sun exposure and such cases as mentioned by Barondes and

Abel where there is a spastic condition of the vessels and, finally, temporary attacks of blindness—the prodromal stage of thrombosis or embolism of the central artery—in all those states where definite improvement takes place by treatment with nitro-glycerine (Imre by personal communication and Barondes) or by acetylcholine (Corrado) improvement would most probably be derived from the operation of cervico-thoracic ganglionectomy (see Chap. XIV).

Types of Optic Atrophy

It is sometimes convenient to classify optic atrophy according to the level at which the lesion of the optic nerve has taken place. I append a table taken from Wright's paper on Optic Atrophy in the *British Medical Journal* of 1923 :—

(1) Optic atrophy following direct damage to neurons by pressure or selective poisons, including sub-inflammatory increase in interstitial elements, excluding inflammation and œdema.
   At the level of the retina : methyl alcohol, alcohol and tobacco ; diabetes. (Axial atrophy ; partial atrophy.)
   At the level of the papilla and pars arteria centralis : glaucoma ; tabes dorsalis ; disseminated sclerosis. (Simple atrophy ; spinal atrophy ; primary atrophy ; genuine atrophy ; tabetic atrophy ; grey atrophy ; non-inflammatory atrophy.)
   At the level of the orbito-cranial portion of nerve and tract : pituitary disease ; disseminated sclerosis ; tabes dorsalis ; fracture ; hydrocephalus ; carotid pressure. (Simple atrophy ; secondary atrophy ; descending atrophy.)

(2) Optic atrophy following damage to neurons through post-inflammatory or post-œdematous change in supporting tissues.
   At the level of the retina : retinitis pigmentosa ; retinitis (syphilitic) and choroiditis (renal). (Secondary optic atrophy ; ascending atrophy.)
   At the level of the papilla and pars arteria centralis : syphilis ; renal disease ; brain tumour ; oxycephaly, etc. (Post-neuritic optic atrophy ; post-papillitic atrophy or consecutive optic atrophy.)
   At the level of the orbito-cranial portion of nerve and tract : acute true retrobulbar neuritis ; periostitis ; orbital cellulitis and abscess ; nasal sinusitis ; influenza ; encephalitis ; acute infective diseases. (Secondary optic atrophy.)

(3) Optic atrophy following damage to neurons and supporting tissues by deprivation of blood supply.
   At the level of the retina : arterio-sclerosis (with and without high blood pressure) ; embolism of central artery ; quinine. (Retinitic atrophy.)
   At the level of the papilla and pars arteria centralis : retrobulbar

growth (?); hæmorrhage into sheath; disease of central vessels (?).

At the level of the orbito-cranial portion of nerve and tract: vascular disease.

Atrophy in the optic tractsis not immediately followed by atrophy of the optic nerves. The fibres of the latter, by their connection with the ganglion cells of the retina, may be kept in a normal state for several years after complete blindness has taken place.

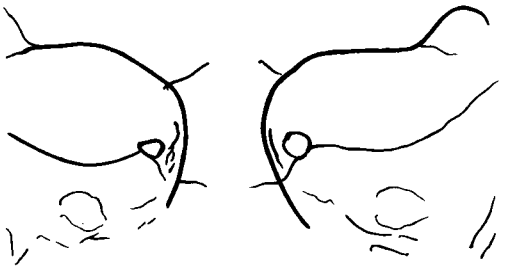


FIG. 37.—Tracing of radiogram showing the unequal size of optic foramina. The right is triangular in shape, smaller than the left. This is due to atrophy of the right optic nerve since birth. (Rhese and Pfahler.)

Treacher Collins and Mayou state that if atrophy of the optic nerve should occur in a case where there is an abnormal development of medullary sheaths (see Plate VI) the medullary substance breaks up and is removed; and the white opaque area in the retina becomes transparent, allowing the red reflex of the choroid to be seen through it.

The pathological processes at work in consecutive and secondary optic atrophy are quite different. In the former, that is the atrophy following a papillœdema, there is at first little change, although the fibres are somewhat disturbed by the presence of œdema. Later, the fibres bathed in this œdematous fluid swell up and show moniliform varicosities of which the central area or core stains differently from the rest; this apparent nucleus suggests the presence of cytoid bodies. The swelling affects the cement substance, binding together the individual fibrillæ which compose the nerve fibres. As the swelling increases disintegration progresses until the whole fibre degenerates into fatty *débris*. Finally, neuroglial proliferation in the disc replaces the degenerated fibres and kills out the nerve fibres left intact. The glial and connective-tissue sclerosis, due to shrinking, pulls back the retina into position, leaving the disc surface an opaque white colour and sometimes surrounded by irregular and indeterminate pigmentation.

In secondary optic atrophy, that is atrophy following a state of inflammation of the retina, optic nerve or both, the inflammation in

the nerve below the entrance of the retinal vessels causes pressure on the retinal vein, where it lies in the scleral foramen ; the engorgement and tortuosity of the vein so produced is followed by œdema of the nerve head and surrounding retina, and hence hæmorrhages in the retina are frequently present. The œdematous disc head pushes out the retina, the fluid accumulating between the nerve fibres forms cystic spaces and contains granular *débris*. If the exudate in these spaces undergoes a hyaline change, it may give rise to a permanent swelling of the optic disc. The inflammatory exudate fills in the physiological pit obscuring the lamina cribosa, and a fine membrane may even form over the disc surface. The exudate may even spread over the surrounding retina and pass along the perivascular lymph spaces, leading to the formation of white lines along the vessels (see Plate IV, Fig. 2).

In primary optic atrophy there is no newly-formed tissue on the disc surface and the lamina cribrosa is usually exposed due to the atrophy and shrinking of the nerve fibres (see Fig. 36).

Primary optic atrophy includes the atrophy associated with disseminated sclerosis, with tabes and general paralysis, and with hereditary optic atrophy. The lesions present in disseminated sclerosis are discreet, they are rose-pink coloured and may affect any part of the optic pathway. Observing a case in progress, where the lesion was close to the eyeball, the disc was slightly swollen and red-coloured. A few months later the dark-red colour had disappeared leaving the temporal marginal portion of the disc quite pale. Some of the neurofibrils disappear and although the myelin sheath degenerates the neurofibrils can still function, which differs from what occurs in tabetic optic atrophy where the fibres are soon destroyed. In disseminated sclerosis sight may be lost temporarily, but in general some vision finally remains.

In tabetic optic atrophy grey patches are seen on section of the nerve resembling the degeneration in the posterior columns of the spinal cord. For a long time the view was held that optic atrophy in tabes began in the ganglion cells of the retina. Erb, Charcot, Gowers and Coppez believed the primary neuronic degneration began thus. Léri in 1904, however, stated that the changes in the ganglion cells were secondary to the changes in the optic nerve fibres, while Stargardt pointed out that even when complete optic atrophy had taken place normal ganglion cells can still be found in the retina. Generally, one cannot detect any ophthalmoscopic

change in the retina even in the presence of a completely white atrophic disc.

In tabetic atrophy the nerve fibres lose their white medullary sheath, becoming transformed into fine fibrillæ which gives the lesion its grey translucent appearance. Between the fibres are cells filled with fat and later, owing to sclerotic change, thickened septa and increase of glial nuclei take place. Léri, from an examination of 84 cases, states that the lesion is an interstitial neuritis, a syphilitic cirrhosis of vascular origin and a syphilitic meningitis, the nerve being secondarily affected by the meningitis. The peripheral loss of field indicates that the lesion is first found at the periphery of the nerve. According to Stargardt there is no fibre degeneration unless there is an exudative process in some part of the course of the nerve. The exudative change is found, he says, in the intracranial and foraminal portions of the nerve, next the chiasma and more rarely in the orbital portion. In tabes this exudative process is an isolated one in the optic nerve and may even spread to the brain, while in general paralysis the process spreads from the brain to the optic nerve.

In the light of the work of Noguchi, Levaditi, Mott, Head, Fearnsides, McIntosh and Fildes, Paton states that it must now be allowed that all the manifestations of syphilis are due to the local production of toxins in the presence of the spirochæte and that the reaction between the spirochætes and the tissue varies at different periods and in different tissue. The question is still unsettled, he says, as to whether this locally produced toxin acts directly on nerve tissue, producing a parenchymatous degeneration or primarily on connective tissue, vascular and lymphatic tissues, with a consequent secondary nerve degeneration, or that the nerve degeneration and the connective tissue degeneration are co-ordinate results due to the presence of the spirochætes in both tissues.

CHAPTER VI

VISUAL TRACTS AND CORTICAL REPRESENTATION OF VISION

THE **Optic Nerves** spring from the fore-part of the chiasma, and as they diverge from one another each becomes rounded in form and of firmer texture. Each nerve is enclosed in a sheath derived from the pia mater and the arachnoid. Passing beneath the anterior cerebral artery it enters the optic foramen, where it receives a sheath from the dura mater. The bony canal through which the optic nerve passes on its way to the orbit is in relation on its medial side with the sphenoidal sinus and sometimes a posterior ethmoidal sinus, from which the optic nerve may be separated by an extremely thin plate of bone. The nerve on reaching the orbit shows this sheath dividing into two layers. One becomes continuous with the periosteum of the orbit, while the other forms the sheath of the optic nerve as far as the eyeball. The nerve pierces the sclera about 3 millimetres to the nasal side of the posterior pole and very slightly below it. Here the outer fibre sheath of the optic nerve derived from the dura mater blends with the sclera, while the bundle of nerve fibres are carried forwards through a series of small apertures. This perforated portion of the sclera is called the *lamina cribosa*. It is at this point that the fibres of the optic nerve lose their medullary sheaths. During its passage through the orbit the nerve is accompanied by its ophthalmic artery which crosses the nerve on its upper surface, together with the ophthalmic vein and the naso-ciliary nerve. The ophthalmic artery comes to lie on its outer and lower aspect.

The central artery of the retina is the first of the smallest branches of the ophthalmic artery. It runs for a short distance within the dural sheath of the optic nerve and at a point about 12 millimetres behind the eyeball it pierces the nerve obliquely and runs forward in the centre of the nerve to the retina. The optic nerve is closely accompanied by the minute ciliary nerves and vessels. It is slightly longer than the distance which it has to traverse from the optic foramen to the globe of the eye, so that the movements of the eyeball may not be impeded.

From the structure and mode of development of the optic nerve it can be looked upon as a prolongation of the brain substance rather than as an ordinary cerebrospinal nerve. The formation of its sheath, showing spaces which communicate with those of the subdural and subarachnoid membranes of the brain, permits the extension of inflammatory affections of the meninges and the passage of blood from hæmorrhage due to disease or accident along the nerve. The fibres of the optic nerve are not uniform in size. *The larger fibres* are thought to subserve the function of *sight*, while the *smaller* are supposed to be concerned with the *afferent pupillary light reflex*.

The axons composing the optic nerve begin in the ganglion cells of the retina. These nerves, on their passage to the optic disc, are non-medullated and are therefore invisible. Occasionally, however, bundles of these nerves close to the optic disc have preserved their medullary sheath and are known as opaque nerve fibres (see Plate VI). Occasionally white plaques of opaque nerve fibres are found in other parts of the retina far removed from the disc. The importance thus attached to their appearance is given on account of the common error made by the inexperienced when examining discs with the ophthalmoscope. A class of nineteen post-graduates were examining two such cases at a hospital for diseases of the nervous system and not one of them gave a correct diagnosis. The majority suggested that they were cases of papillœdema.

The two optic nerves meet behind to form the optic chiasma, there many of the fibres intermingle. Those coming from the temporal side of each retina do not decussate but pass directly to each corresponding tract. The fibres from the inner or nasal half of each retina do decussate ; those from the right eye pass to join the left optic tract ; while those from the nasal side of the left eye pass to the right optic tract. The fibres which pass from each macular area enter the temporal side of the optic nerve, there to form what is called the papillo-macular bundle. This bundle as it passes backwards gradually assumes a central position in each optic nerve. Many of these fibres cross in the chiasma ; indeed, the posterior edge of the chiasma consists entirely of crossing central fibres. It is this bundle which suffers so severely in disseminated sclerosis. (Fig. 38).

The fibres of the optic nerves form a complete decussation in many mammals, and in all vertebrates below mammals, so that all

the fibres pass from one optic nerve into the opposite optic tract ; but in some of the higher mammals, such as the rabbit, monkey and in man, the decussation is incomplete.

According to Salzer's enumeration there are nearly half a million fibres in each optic nerve ; they vary in size and, according to Güdden, are divisible according to their calibre into finer and

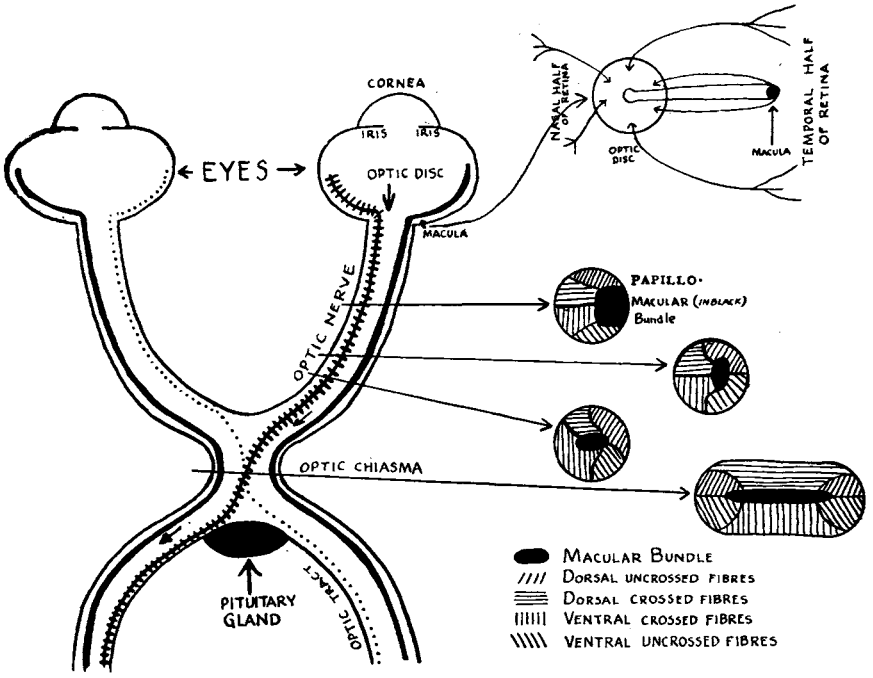


FIG. 38.—Various positions of the papillo-macular bundle. (After Henschen.)

coarser fibres. He regarded the former as afferent fibres concerned in the production of the pupillary reflexes, and the latter as conveying visual impressions. The afferent pupillary fibres part company with the visual fibres before the latter enter the so-called primary visual centres—the lateral geniculate body, pulvinar and superior colliculus. When the superior corpora quadrigemina in young animals are destroyed the finer fibres undergo atrophy. In addition to the optic nerve fibres, there is a bundle at the posterior part of the chiasma running along the mesial side of the optic tracts to join the medial geniculate bodies of the two sides. This is known as

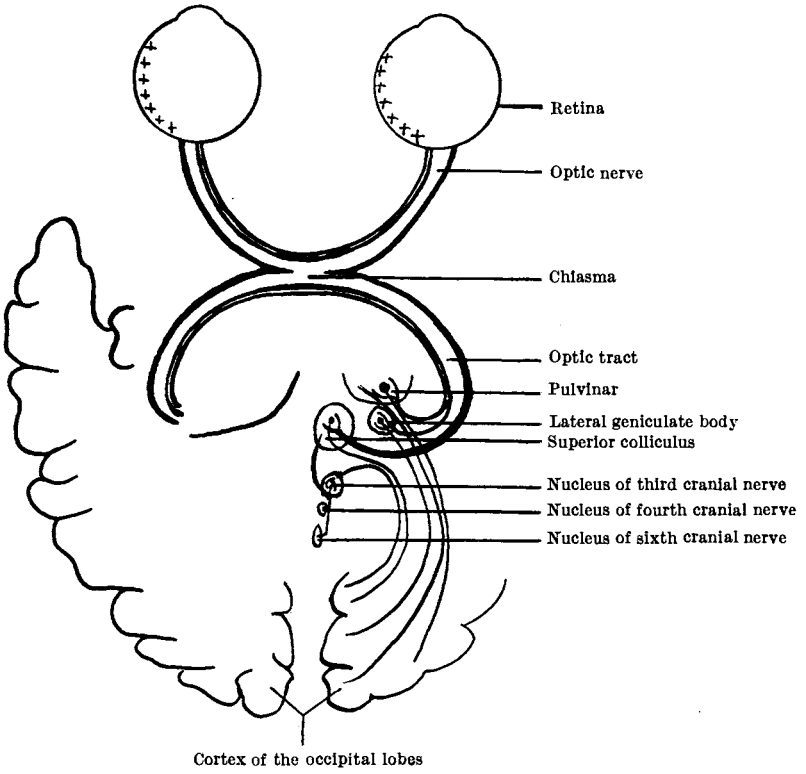


FIG. 39.—Diagrammatic appearance of visual nerve paths and their connections. On the left side of each retina are markings corresponding with the right side of the field of vision. Fibres from the left half of each retina together with some crossed and uncrossed macular fibres pass in the left optic nerve and tract to reach the primary visual centres. The superior colliculus is connected with the nuclei of the 3rd, 4th and 6th cranial nerves. The optic radiations are seen to spread from the pulvinar and lateral geniculate body into the occipital lobe.

The effect of complete lesions at various sites are as follows :—

1. Through the optic nerves, amaurosis or blindness of eye on same side. No pupillary response to direct light stimulation of ipso-lateral eye, but contraction upon light stimulation of the contra-lateral eye.
2. Chiasma. Bi-temporal hemianopia.
3. Optic tract. Contra-lateral hemianopia.
4. External or lateral geniculate body. Contra-lateral hemianopia. (Lesions of pulvinar and superior colliculus do not result in hemianopia.)
5. Lesions in optic radiations or occipital cortex show hemianopia with normal pupillary reactions. A lesion in the lower wall of the calcarine fissure is followed by a homonymous (upper) quadrant hemianopia.

the *Commissure of Güdden.* It appears to have no relation to the visual function, as it undergoes no change after removal of the eyes of young animals, while atrophy of the optic tracts follows removal. This commissure forms part of the auditory mechanism.

Each optic tract passes from the chiasma backwards around the crura cerebri and gives off a mesial and lateral root. The mesial root connects the two medial geniculate bodies, and is referred to above as the commissure of Güdden.

From what has been said it can be gathered that each optic tract is composed of uncrossed temporal fibres, crossed nasal fibres and uncrossed and crossed macular fibres. (Fig. 40.)

The optic tracts pass backwards from the chiasma and end in

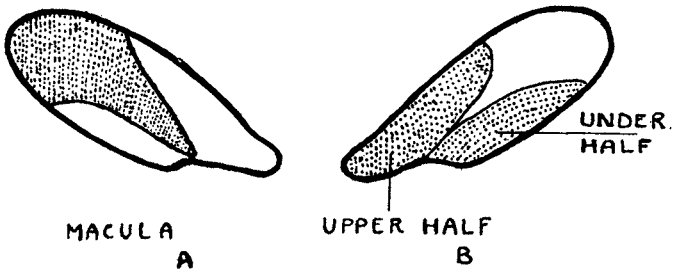


FIG. 40.—Transverse sections of both optic tracts. (*a*) Shows the maximal degeneration occurring in the optic tract after lesions of the macula. (*b*) Shows the maximal degeneration occurring in the optic tracts after lesions of the upper and lower halves of the retina. The fibres from the upper portions of the retina lie dorso-medially. (From Brouwer.)

what are known as the primary visual centres: these are the lateral geniculate body, superior colliculus of the corpora quadrigemina, and the pulvinar of the optic thalamus (see Fig. 39).

The primary optic centres are also connected with the nuclei of the third, fourth and sixth cranial nerves by means of the posterior longitudinal bundle. Also there is a connection between the superior colliculus and the medulla and cord by means of the mesial fillet.

It is stated (Brouwer) that 90 per cent. of the retinal fibres of the optic tract pass into the lateral geniculate body so that a comparatively few retinal fibres pass into the superior colliculus of the quadrigeminal body. Certain reflex movements and adjustments of the eyes are probably carried out by means of various fibres entering the superior colliculus; these come from the retina, from

the lemniscus (thus making connection with the sensory tracts of the medulla and spinal cord), from the cerebral cortex through the lateral root or superior brachium and from the opposite superior quadrigeminal body. From each superior colliculus a bundle of fibres passes on each side sweeping round the central grey matter of the aqueduct to the ventral part of the dorsal longitudinal bundle and decussates with its fellow in the raphe, where the two bundles form the fountain-decussation of Meynert to become longitudinal in the part of the tegmentum ventral to the dorsal longitudinal bundle and as the tectospinal tract passes partly through the red nucleus giving off to it collaterals, and then downwards in the formatio reticularis of the mid-brain, pons and medulla and finally intermingling with the fibres of the dorsal longitudinal bundle in the spinal cord. Thus, as Whitnall remarks, "The superior colliculus may be regarded as a ganglionic centre for the co-ordination of the visual impressions with those of other regions of the body influencing its movements."

There is a certain amount of doubt regarding fibres ending in the pulvinar, but the relation of the optic thalamus to the cerebellum and red nucleus, as well as its relationship to the precentral convolution, makes it most probable that it is intimately related to the movements of the eye and the higher visual functions of stereoscopic vision. The fibres which pass to the superior corpora quadrigemina represent those seen in the lower animals and therefore are most primitive phylogenetically. The largest number of optic fibres terminate in the lateral geniculate body. Many fibres from the macular region end here also, so that this area is most important as regards central vision.

From the lateral geniculate body and the pulvinar spring the optic radiations or fibres of Gratiolet, which lying in the retro-lenticular part of the posterior limb of the internal capsule and internal to the auditory bundle sweep backwards into the occipital lobe on the outer side of the posterior horn of the lateral ventricle to the cortex of the cuneus and the occipito-temporal convolutions which is continuous into the temporal lobe (see Fig. 41). The fibres of the optic radiations occupy a larger part of the posterior hemisphere than was originally thought. They form a large flat band, the medullary optic lamina, whose ventral border extends into the temporal lobe, the posterior portion of which turns backwards towards the anterior end of the calcarine fissure, while the dorsal

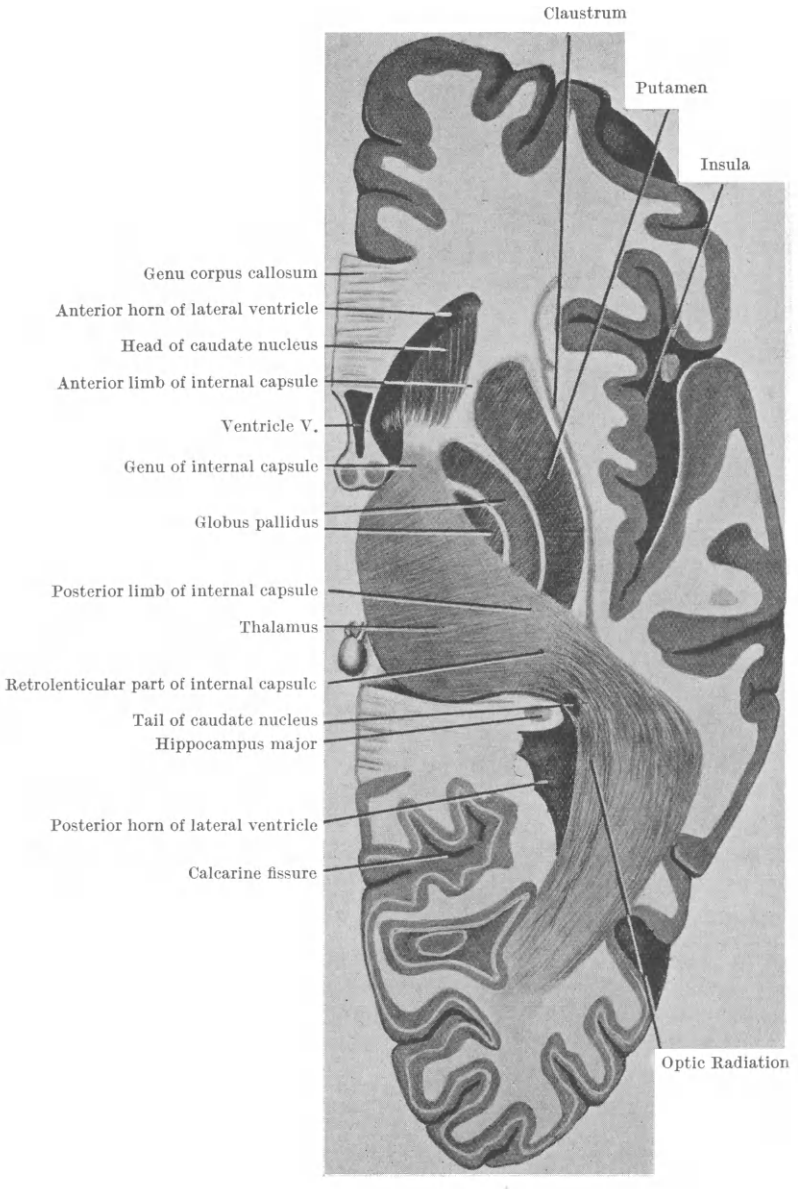


FIG. 41.—Drawing of horizontal section of cerebral hemisphere at the level of the putamen and globus pallidus (lenticular nucleus), showing the sweep backwards of the optic radiations (medullary optic lamina) into the temporal and occipital lobes.

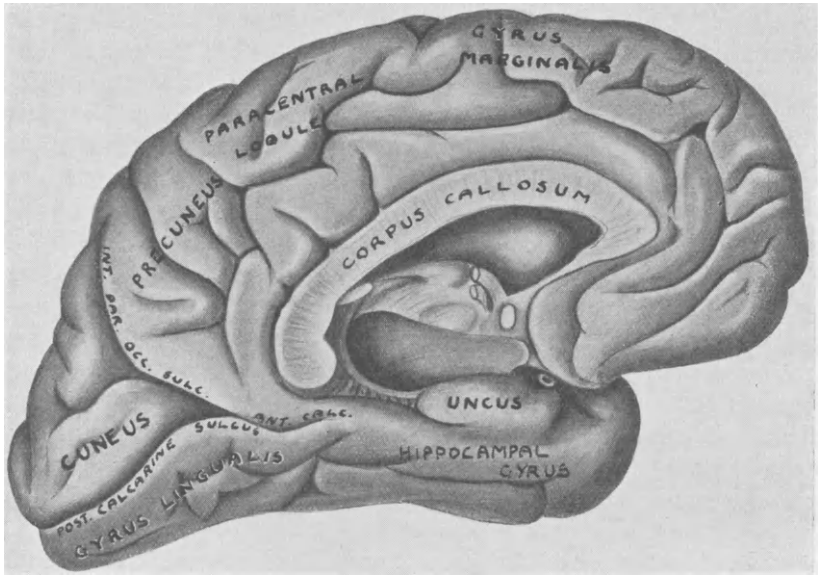
border runs upwards through the white matter of the parieto-occipital lobe as far as the level of the insula from which it spreads downwards towards the posterior portion of the calcarine fissure. Bundles of fibres are given off from this lamina turning into that part of the sensory visual area known as the area striata situated mainly on the mesial aspect of the occipital lobe but also at the occipital pole. The structure of the cortex of the brain in this area shows a white line, the line of Gennari. The greater part of the calcarine fissure lies within an area where the cortex is distinguished by the presence of this white line which is visible to the naked eye in the grey matter. The area striata occupies the bottom and sides of the posterior calcarine fissure and extends into the adjacent superficial cortex but it does not reach to the anterior end of the anterior calcarine fissure. At this part the area is confined to the lower lip of the fissure. The limits of this area have been defined by Bolton, Elliot Smith, and Campbell (see also J. Purdon Martin).

The optic radiations or medullary optic lamina is composed mainly of ascending fibres arising from cells of the lateral geniculate body and partly from the pulvinar. The superior colliculus does not supply any fibres to the lamina. It contains commissural and association fibres, also descending fibres from the visual cortex to the lower visual centres. Like the optic tract its fibres are so arranged that impressions from corresponding halves of the retinæ pass through each lamina and from the macula of each eye also.

The visual cortex of the brain is probably limited in the occipital lobe to the area between the sulcus lingualis below and the sulcus cunei above, including the walls of the posterior calcarine sulcus and the ventral wall of the anterior calcarine sulcus (see Fig. 42). The area extends posteriorly to the occipital poles and anteriorly to the precuneus. The greater part of the calcarine fissure lies within an area where the cortex is distinguished by having a white line—the line of Gennari—visible to the naked eye. This area—the striate area—is the chief end station of the optic radiations, and is termed the visual sensory area. Meyer and Cushing have shown how the ventral part of the optic radiation passes forwards into the temporal lobe and then sweeps backwards in a wide curve to the calcarine fissure. Tumours, therefore, in the tempero-sphenoidal lobe can cause defects in the visual field, quadrantic in shape, together with visual hallucinations and subjective

sensations of smell, described as the "*uncinate fits*" *of Hughlings Jackson* from the involvement of the uncinate process of the hippocampal gyrus.

The macular fibres lie closely along the connection of the tract with the brain, gradually rotating outwards until they enter the geniculate body dorso-laterally. These fibres then pass in the radiations between the dorsal and peripheral fibres and terminate



Mesial surface.

FIG. 42.—The gyri and sulci on the mesial aspect of the left cerebral hemisphere, the calcarine sulcus indicating the visuo-sensory area.

in the most posterior part of the occipital lobe, where they occupy a relatively large share of the visual cortex. Some observers have put forward the view that both sides of each macular area in the eye are represented in each cortical macular centre. This is most probably not the case, but that each cortical macular centre represents only the lateral halves of the two maculæ. It must be remembered that each cortical macular centre by spreading over the occipital pole has a double blood supply.

Duke-Elder says that "the ventral visual field, or the dorsal half of the retina, is projected from the mesial part of the geniculate

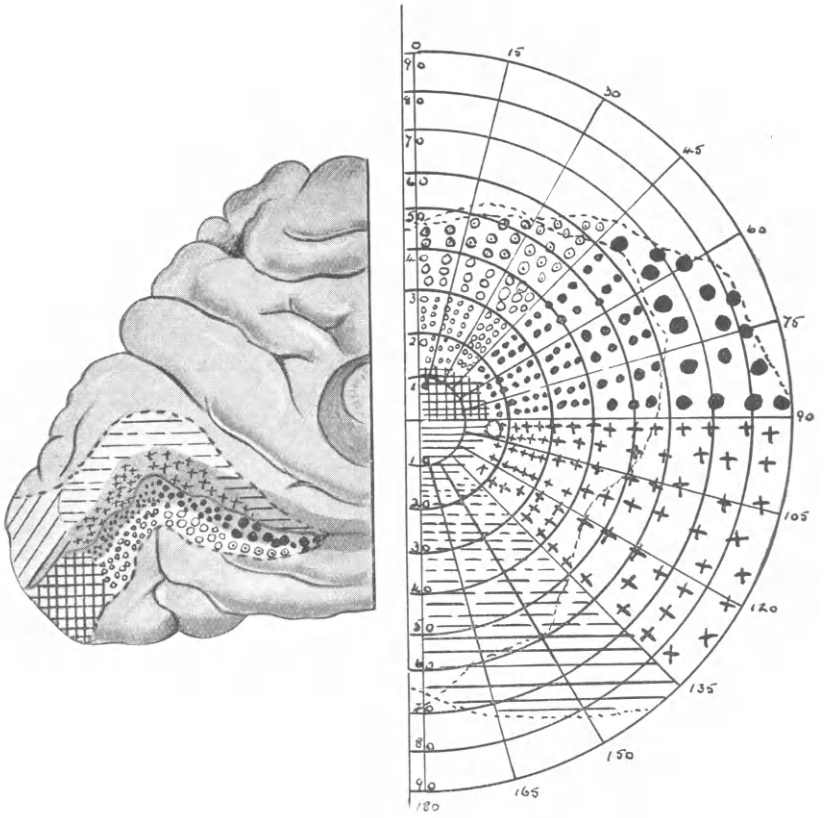


FIG. 43.—Mesial surface of left occipital lobe together with the right half of the corresponding field of vision.

The lips of the calcarine fissure are pulled apart showing the probable location in the striate area of points corresponding to those of the right visual field. Fibres passing from the upper portion of the retina pass in the dorso-medial portion of the optic tract to the lateral geniculate body in its medial part. From there fresh fibres arise and pass through the dorsal portions of the optic radiation to reach the dorsal wall of the calcarine fissure. Hence on the right side of the field of vision those areas represented by the various markings correspond to the areas of the calcarine fissure similarly marked. It will be noted that the peripheral areas of the field of vision correspond to the most anterior parts of the visual cortex, while the macular area is very largely represented at the occipital pole. (Gordon Holmes.)

body along the dorsal portion of the radiations to the dorsal lip of the calcarine fissure ; and the dorsal field or the ventral half of the retina is projected from the lateral part of the geniculate body up

the ventral portion of the radiations to the ventral lip of the calcarine fissure."

(Fig. 43.) Gordon Holmes has kindly permitted me to use the diagrams which demonstrate the cortical representation of the various parts of each retina. It will be noticed that the black dots, representing the fibres of the optic nerve beginning in the lower and inner quadrant of the retina, terminate in the anterior portion of the lower lip of the calcarine fissure. The crosses representing the fibres passing from the upper and inner quadrant of the retina terminate in the anterior portion of the upper lip of the calcarine fissure ; while the lines representing the macular fibres pass to the most occipital part of the fissure and are continued on the outer surface of each occipital pole. This at once explains why the macular area of vision is so often spared in complete hemianopias. The calcarine fissure is supplied by branches of the posterior cerebral artery, while the occipital pole is supplied both by the posterior and middle cerebral arteries, so that if one of these vessels is blocked by embolus or thrombosis the other will supply blood to this area of the cortex (see paper by Fox and German).

The Central Connections of the Oculo-Motor System and the Posterior Longitudinal Bundle.—For a long time it has been known that electrical stimulation of the posterior part of the second frontal convolution anterior to the precentral gyrus induces a conjugate movement of both eyes to the opposite side. Stimulation higher up on this convolution results in a conjugate downward movement of the eyes. These are relatively known as conjugate lateral deviation and conjugate vertical deviation. Behind these points of stimulation lie the centres for the eyelids. The path of these supranuclear fibres is through the corona radiata, the internal capsule and the cerebral peduncle of the mid-brain and pons, where the fibres decussate and pass into the pons. The fibres divide at this level just above the sixth nerve nucleus, some entering this nucleus while others pass into the longitudinal bundle, and running upwards reach the contralateral oculo-motor nucleus. It is thus that the movements of the external rectus muscle of one eye are brought into harmony with the movements of the internal rectus of the opposite eye. (Gordon Holmes suggests there may be a separate centre for such a function lying ventral to the nucleus of the sixth cranial nerve.)

It has been known that stimulation of the occipital lobe also produces conjugate deviation of the eyes ; that, in fact, the calcarine

area is a centre not only for perception, but has connections with the ocular nuclei. Stimulation of this area produces conjugate deviation of the eyes, but such deviation is overcome if the frontal lobe is stimulated simultaneously, the former always predominating. Fibres from the peristriate areas pass through the posterior part of the internal capsule to the oculo-motor nuclei.

The oculo-motor nuclei are connected with each other especially by means of the posterior longitudinal bundle, so that reflex deviation of the eyes is brought about by stimulation of the retinæ and any part of the visual field.

By means of the posterior longitudinal bundle impulses pass from the auditory portion of the eighth cranial nerve, which affect the ocular centres so that there is a direct ocular response to sound. Similarly, labyrinthine impressions pass through this bundle to the oculo-motor nuclei. In the subthalamic region the posterior longitudinal bundle receives fibres from the nucleus of Darkschewitsch and the interstitial nucleus of Cajal, the former bringing it into relationship with the posterior commissure. The third, fourth and sixth nuclei in the mid-brain are united by the passage of this bundle. Deiter's nucleus supplies connecting fibres close to the upper part of the pons. While passing through the pons the posterior longitudinal bundle receives fibres from the fifth and eighth nerves. The bundle now passes through the medulla, giving off fibres to the spinal accessory nucleus to become part of the anterior column of the cord. The posterior longitudinal bundle therefore connects up both oculo-motor nuclei, the superior colliculus, and auditory and vestibular nerves and probably the facial nerve.

In Muskens's recent work his physiological analysis closely relates conjugate movement of the eyes and body posture. He links up the oculo-motor nuclei, the vestibular nuclei and the basal ganglia and so extends the co-ordinating mechanism forwards through the region of the posterior commissure to the globus pallidus in the basal ganglia. But we must still believe that willed lateral deviation of the eyes enters the posterior longitudinal bundle from below.

Vascular Supply to the Visual Pathway.—The ophthalmic artery, a branch of the internal carotid, supplies the optic nerve ; it is derived from the internal carotid just as that vessel is emerging from the cavernous sinus on the mesial side of the anterior clinoid process, and it enters the orbital cavity through the optic foramen below and lateral to the optic nerve. A branch of the ophthalmic,

called the *central artery of the retina*, passes between the bundle of fibres of the optic nerve to the inner surface of the retina at the middle of the optic disc. Emerging at the disc the central vessel divides into branches, usually two, each of these again dividing into further branches. Small twigs from the superior and inferior temporal branches of the central artery supply the macular region (see Plate II) but the fovea itself is entirely free from blood vessels. Beneath this region there is a marked thickening of the chorio-capillaris. An embolism of the entire central artery of the retina or any one of its branches may take place.

The chiasm receives branches from the internal carotid, the anterior cerebral and the anterior and posterior communicating arteries. Plate I illustrates the relations between the carotid arteries and the chiasm and optic nerves.

The optic tracts are supplied by small vessels derived from the posterior communicating artery in front and the anterior choroidal branch of the internal carotid behind. The posterior cerebral artery supplies the lateral geniculate body and the corpora quadrigemina, which are therefore nourished independently of the tract. Part of the optic radiations are supplied by the anterior choroidal artery, but mainly from the calcarine branch of the posterior cerebral artery.

The pulvinar, which is the prominence on the posterior end of the thalamus, is also supplied by basal branches of the posterior cerebral artery.

The visual cortex is supplied by the calcarine artery and other branches of the posterior cerebral artery. Small branches from the middle cerebral artery reach this area. The vessels are so arranged that the grey and white matter are supplied independently. The terminal branches of the arteries do not anastomose, so that injection into one arterial stem cannot be conveyed through the terminal arteries into another main branch. The veins, however, open into the pial venous network and intercommunicate freely, so that arterial obstruction is likely to do more damage than would a venous thrombosis.

Branches from the anterior spinal, the vertebral, the basilar and the posterior cerebral arteries supply the pons and medulla oblongata. These branches are arranged in two sets, the lateral or radicular which follow the roots of the nerves, and the median which, passing into the raphe, reach the grey matter on the posterior surface.

CHAPTER VII

THE INTERPRETATION AND LOCATION OF LESIONS IN THE VISUAL PATHWAY AS SHOWN BY THE PERIMETER

It is difficult to describe fully in a text-book such as this all the diseases which affect vision, but it is hoped that the reader will find sufficient guidance to enable him from what is written to interpret his own findings. Among other things he must learn some accurate method of recording fields of vision to which he can refer from time to time, noting the progression or retrogression of the effects of the disease and its treatment. Fig. 44 illustrates one of the commonest perimeters in use. It is the McHardy self-recording perimeter. A more elaborate instrument and probably one of the best on the market is the Lister self-recording instrument. The perimeter consists of an arc marked in the degrees of a circle. This arc is capable of rotation around a point on which the patient fixes his gaze. A few demonstrations will teach the reader all that is necessary. It should be remembered that the small white objects which are looked at should be well illuminated and should not be allowed to get soiled. Two, five and ten millimetre objects are the most commonly used. The patient's chin is placed on the chin rest while he holds in his hand an

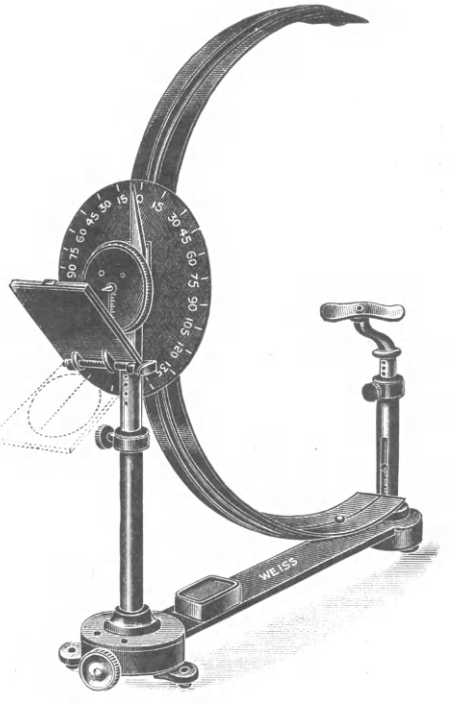


Fig. 44.—The McHardy self-recording perimeter.

operculum before the eye which is not being examined. The head must be placed vertically and kept still. The illumination should come from behind the patient (see first Chapter).

The student should become familiar with some convenient method of measuring the field of vision without the aid of an instrument. He cannot always have a perimeter at hand or time may not permit of an examination by one of the standard instruments mentioned. At such times hand perimetry (confrontation perimetry) is the best method to employ. The examiner stands directly opposite the patient, with the eyes on the same level. Now, for example, it may be assumed that the field of vision of the patient is the same as that of the examiner ; therefore, when a patient places his left hand over his left eye and the examiner places his right hand over his right eye objects seen at the limits of the field of vision by the examiner should be seen by the patient also. The distance between the examiner and the patient should be 18 inches. The patient's eye is directed forward to the examiner's exposed eye. The examiner's left hand is passed outwards until his fingers are just seen at the limit of his field of vision. If he moves his fingers the patient says whether he sees any movement or not. With this distance as a radius, and if a circle is described with the patient's eye as the centre, at 90 degrees (9 o'clock or 3 o'clock to the patient), both patient and examiner should see the fingers moving. The hand is then lowered through an arc of 45 degrees on the same circle where movements of the fingers can still be observed by both. The hand is then brought directly below the patient's eye, but will have to be approached to a point on a circle of 65 degrees from the centre point of vision before movement is seen. The hand is still continued counter-clockwise until it is somewhat to the left of the eye, where roughly 45 degrees is the normal limit of the field of vision ; while, if the examiner's hand is held horizontally to the left of the patient's right eye, the patient will see movement of the fingers at a radius approximating 55 degrees. Held above the horizontal line and to the inner side of the eye the radius should have increased slightly to about 60 degrees. Vertically above the eye the field of vision again is 45 degrees, but when travelling through an arc of the circle midway between the vertical point and the extreme right of the patient the field of vision should approximate 70 degrees. By this method of hand perimetry an obvious defect in the field of vision is easily detected.

The chart supplied for the use of Lister's perimeter is illustrated by Fig. 46. It will be understood from this diagram that the field of vision is not a continuous circle, but is unevenly constricted owing to the contour of the surrounding portions of the face. As faces are so dissimilar two fields of vision are never quite alike.

Lesions of the Optic Nerve.—Reference has already been made to the blind spot of Mariotte, which is that area in the field of vision

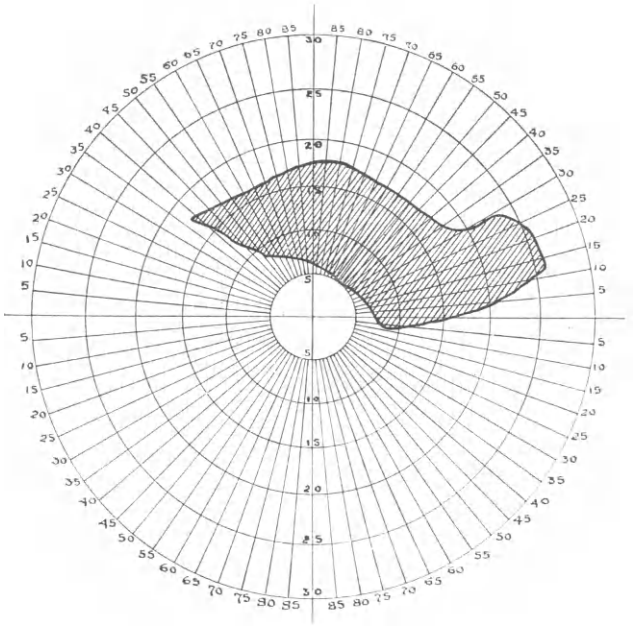


FIG. 45.—An absolute scotoma in the field of vision of the right eye due to an isolated patch of retinitis. Recorded with Armstrong's scotometer.

represented by the optic disc of each fundus. This blind spot is situated about 17 degrees from the point of fixation. If the white movable object is carried outwards along the arc in a horizontal direction, while the patient fixes his gaze on the central white spot, the moving object will come to the inner edge of this blind spot, disappear and again reappear at the outer edge of the blind spot. If such an area were found elsewhere in the field of vision it would be termed an *absolute* scotoma. The presence of chronic glaucoma and of papillœdema cause an enlargement of the blind spot in the visual field. Concentric diminution of the visual field may be

brought about by changes in the optic nerve due to tabes, retro-bulbar neuritis or progressive consecutive optic atrophy. Scotomata, relative or absolute, are found in the visual field in disseminated sclerosis, in inflammatory affections of retina, choroid, optic nerve and orbit. Fig. 45 is the field of vision in the case of a young woman suffering from an isolated patch of retinitis. The shaded area is that of an absolute scotoma. All that can be observed in the eye with the ophthalmoscope is a fine change in the colour of that part of

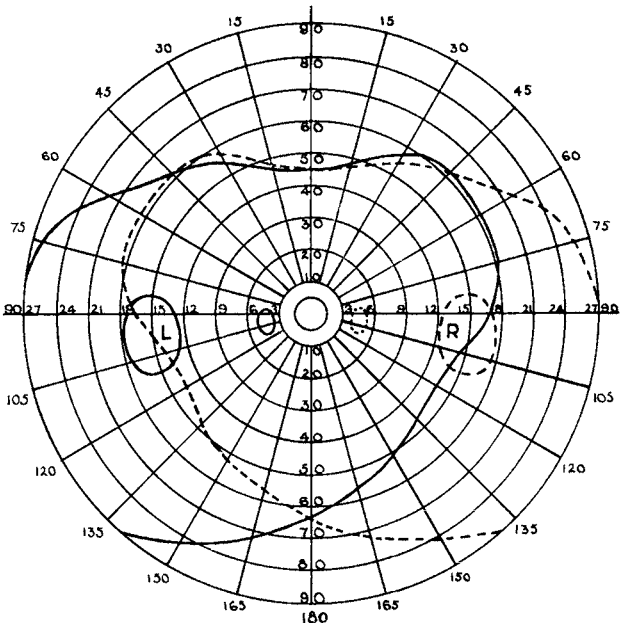


FIG. 46.—The Lister perimeter chart.

the retina corresponding to this blind area. Later some pigmentary change will manifest itself indicating the termination of the disease.

It should be observed how little the field of vision is affected, even in marked cases of papillœdema, while in retro-bulbar neuritis, where there is little or no change of the disc, there is marked decrease of vision, both central and peripheral. Bilateral macular degeneration, whether caused by toxins or vascular conditions, leave permanent central scotomata of varying density (see Chapter VIII).

In contrast to Fig. 45 is that of Fig. 33, which illustrates the diminishing field in a case of tabo-paresis. (The chart shown in

Fig. 33 is the form employed in the McHardy perimeter, whereas the chart illustrated in Fig. 45 is the one used in Armstrong's scotometer, one of the finest instruments of its kind (see Chapter I). Fig. 33 shows the field of vision gradually diminishing from without inwards at the same time as the optic disc is slowly becoming paler in colour.

Doyne recently demonstrated fields of vision showing scotomata chiefly in cases of tobacco amblyopia. The blindness in these cases was central. The area extends frequently from the macula to the disc. In the lesser forms of amblyopia this area is not blind, but colours such as red and green are not perceived by this portion of the retina. Such partially blind spots are known as *relative* scotomata. They are nearly always central, and are commonly seen in retro-bulbar neuritis and in tobacco amblyopia (see Chapter XVI).

In addition to white objects being used on the perimeter arm, red, blue and green are also employed. The colour field is smaller than that for white. Next to the white field the blue is largest, then comes red and green. The colour fields vary, not only with the density of the light, but also with the saturation of the colour and the size of the object. In cases of hemianopia which are due to disease of the cortex, investigation with the perimeter shows that the colour fields are often lost before the field for white.

Reversal of the colour fields (the blue field lying within the red) or intermingling of the boundaries is found occasionally in functional cases. In cases of pituitary disease the loss of colour perception is always in advance of that for form.

Traumatism of the Optic Nerve.—In a previous chapter (Chapter V) it was stated that slight blows or falls on the head may be followed months or years later by optic atrophy. An injury which affects the optic nerve may or may not be accompanied by a fracture of the orbital walls or optic canal. When the nerve is severely damaged there is instantaneous and permanent blindness of the homolateral eye. In a few weeks the onset of primary optic atrophy is observed. Gradually the disc is seen to become white.

The optic nerve being an intercentral nerve degeneration takes place in both directions, thus differing from a peripheral nerve where degeneration occurs only in the downward direction. When the optic nerve is divided the fibres of the optic portion slowly atrophy and the optic disc becomes white. The atrophy, however, proceeds much quicker away from the eyeball. The ganglion cells

of the retina begin to show chromatolysis and finally degeneration of the retinal nerve fibres takes place. (Friedenwald.)

Occasionally the visual pathway is not completely interrupted as shown by Fig. 47. In this particular case a central and annular scotoma with an almost complete peripheral field followed on an injury to the right optic nerve due to compression by callus resulting from a fracture of the sphenoid bone which extended into the optic canal. This is by no means a common condition.

Retro-bulbar Neuritis and other inflammatory conditions are described in a later chapter.

Vascular Lesions.—The pathogenesis of arterio-sclerotic visual

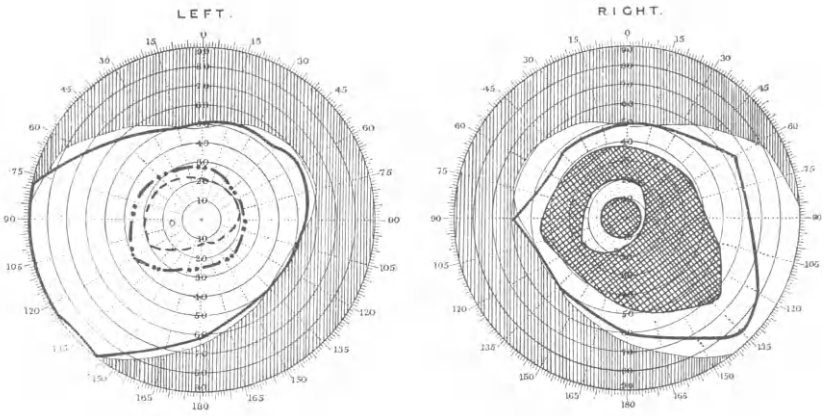


FIG. 47.—A central and annular scotoma in the right optic field due to compression by a callus which followed a fracture of the sphenoid bone : the fracture extended into the optic canal. The left field (including red and green) is normal. (Lillie and Adson.)

disturbance as connected with the optic nerve is not always the same. There may be pressure alone from a sclerosed internal carotid artery or a change in the nerve or chiasm from arterio-sclerosis of the minute vessels supplying their substance. (Alpers and Wolman). Optic disturbances from arterio-sclerosis suddenly develop in one eye and then remain stationary, the progress is uneven. (Rönne.) In a case under the author's care, that of a man sixty years of age suffering from arterio-sclerosis, almost complete blindness suddenly ensued. The resulting field of vision is that illustrated by Fig. 48. Only a small portion remained below to the outer side of the central area. Three months later the sight had improved and had included the macular area (Fig. 49). The

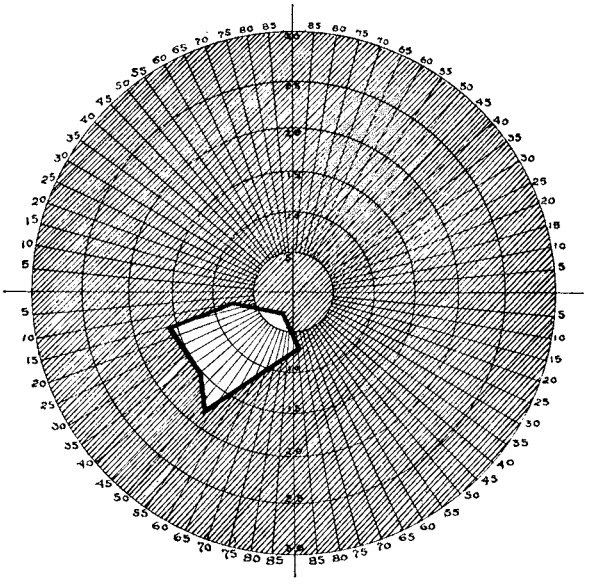


FIG. 48.—Field showing small area of vision remaining due to thrombus of central artery.

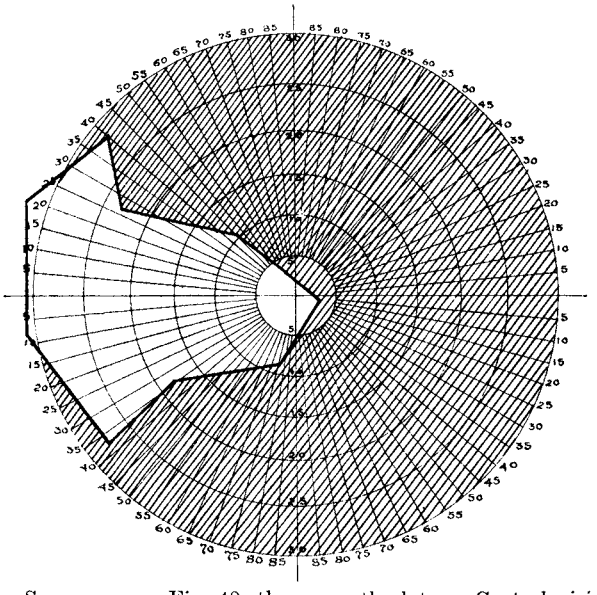


FIG. 49.—Same case as Fig. 48, three months later. Central vision had returned and total area of sight enlarged.

fundus revealed a small central artery strongly resembling the appearance of embolism of this vessel. In another case, that of a woman, aged forty-five, who did not realise she was suffering from hyperpiesis, while playing tennis suddenly found the vision of one eye greatly reduced. Examination showed a normal fundus but the field of vision presented a large oval scotoma above the macular area. Gradually this blind area became smaller in size until in four months' time only a vertical line remained in the position of the scotoma and finally this disappeared altogether. The diagnosis was a hæmorrhage into the optic nerve.

In advanced arterio-sclerosis sudden blindness may ensue: the fundus shows the presence everywhere of minute hæmorrhages and

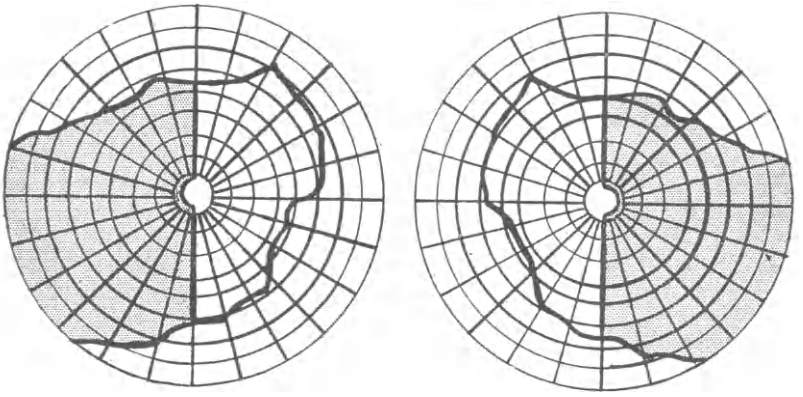


FIG. 50.—Fields of vision showing bitemporal hemianopia. The macula is spared.

the disc head is swollen. The prognosis in such cases regarding both sight and life is extremely grave.

Lesions of the Chiasma.—Lesions in the neighbourhood of the chiasma cause defects of the field of vision such as in Fig. 50. This is a bitemporal hemianopia. Other defects are also found, such as homonymous hemianopia, blindness of one eye, irregular defects in the field. The work of Cushing and Walker shows that the homonymous type of hemianopia is by no means as rare as Uhthoff believed. As stated above, the loss of colour perception is in advance of that for form. In the majority of cases the defect commences in the upper temporal field and progresses more rapidly from this point towards the centre than from the remaining periphery, so that a gourd or cup-shaped field is produced. The temporal field is

gradually lost, and finally the nasal field is reduced to a small island in the lower nasal quadrant (see Fig. 102 (1), (2), (3) and (4).)

The lesions causing such visual defects are due to many causes. Walker and Cushing believe that pressure and traction go hand in hand, while Fisher suggests that the distortion of the field is the

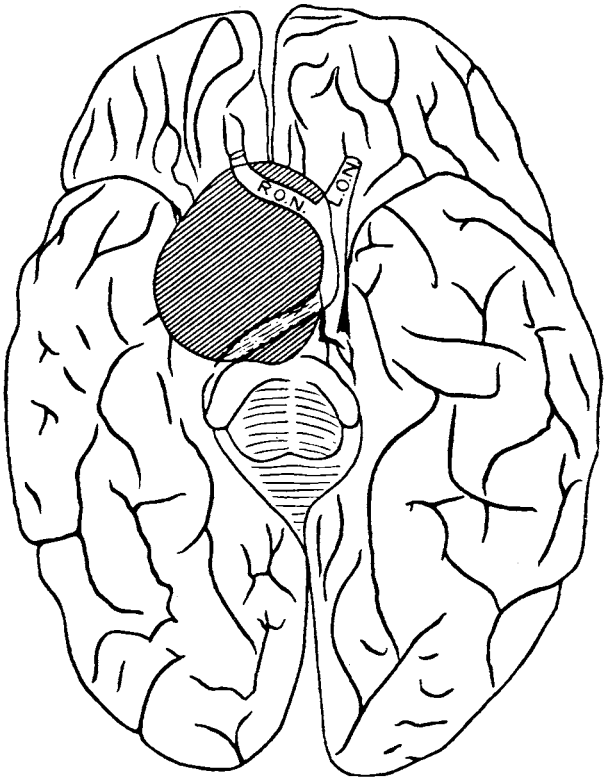


FIG. 51.—Tumour in region of pituitary fossa. R.O.N. Right optic nerve, stretched and flattened, is pushed over to the left side of the tumour. L.O.N. Left optic nerve was normal in appearance. (The autopsy was performed by Carnegie Dickson.)

result of traction upon the nerve and chiasma, rather than of direct pressure upon the spot. A case of the writer will show that both pressure and traction can be present, but with results totally unlike what is commonly found. Fig. 51 depicts the condition of a brain which was removed recently at an autopsy. In the neighbourhood of the pituitary body a large endothelioma was found which had

eroded the pituitary fossa, completely destroying the dorsum sellæ. The patient appeared to be extremely well one week before death although the right eye had become blind from the gradual onset of primary optic atrophy a few months previously. Quite suddenly a papillœdema appeared in the left eye and a few days later the patient was admitted to hospital. The post-mortem examination showed a large tumour 40 millimetres across occupying the pituitary region. On section it was found to be an endothelioma intimately connected with the wall of the cavernous sinus. The tumour had flattened out the fossa (see Fig. 52), and, extending upwards on the outer side of the right optic tract, had pushed this tract over to the left side. The right optic tract and optic nerve were stretched and flattened by the pressure of the tumour. Hence the blindness of the right eye. The left optic nerve and tract were not stretched nor pressed upon. The tumour was pressing not only on the right optic nerve but also on the right optic foramen, so that there was not the slightest possibility of cerebrospinal fluid passing down within the sheath of the right optic nerve to the eye, hence the absence of a papillœdema. It was only when the tumour continued growing and the intracranial pressure increased that a papillœdema was found in the left eye. The entire absence of symptoms suggestive of cerebral lesion was remarkable. The patient sang a solo in church on the Sunday before his admission to hospital.

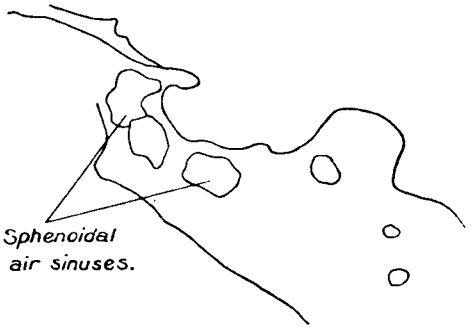


FIG. 52.—Drawing from radiogram of base of skull, showing enlarged and flattened pituitary fossa due to tumour shown in Fig. 51.

Quadrantic hemianopia, being a quadrantic defect in the field of vision, is sometimes found in lesions of the chiasma, but it is not usually so regular as Fig. 53 would seem to show. The lesion may produce a decided quadrantic defect in one field while the other eye may be almost devoid of visual perception. It is interesting to know that the recovery of the affected part of the field may be rapid after operation, and most cases improve if operated on before the stage of complete loss of temporal field. If, however, the optic disc

shows definite atrophy there is little hope of recovery of any portion of the field. In a case of the writer's showing progressive pituitary cystic disease one optic disc was completely white, this eye being practically blind. There was definite quadrantic loss in the field of the other eye, yet the application of deep X-ray therapy produced a complete cessation of further increase of signs and symptoms. Vision in the eye with the quadrantic defect improved so much that

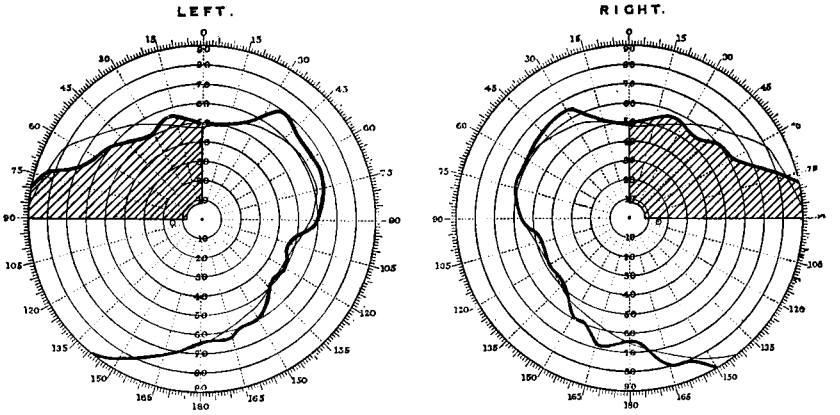


FIG. 53.—Fields of vision illustrating bitemporal quadrantic loss of vision.

the patient was able to return to his normal work—that of a sailor on the high seas.

In addition to the causes mentioned above are suprasellar cysts, meningiomas, gliomas, aneurysms and pressure from the third ventricle (see p. 285).

It is commonly recognised that a compression or other diffuse lesion of an optic nerve usually produces a disturbance of central vision in the form of a partial or complete scotoma, but Gordon Holmes has pointed out that such scotomata do not occupy an important place in the symptomatology of suprasellar endotheliomata, which compress the optic nerves before they involve the chiasma. In only four of his fifteen cases of suprasellar tumours central or paracentral scotomata were present when the patients came under observation. In connection with these suprasellar tumours Gordon Holmes states that a bitemporal hemianopia is relatively rare and that it never includes the whole and yet remains limited to the temporal halves of the fields of vision and is rarely symmetrical.

Altitudinal Hemianopia.—Meningitis affecting the base of the brain, especially the gummatous variety, may cause a lesion of the chiasm. If there is pressure on or disease of the upper or lower

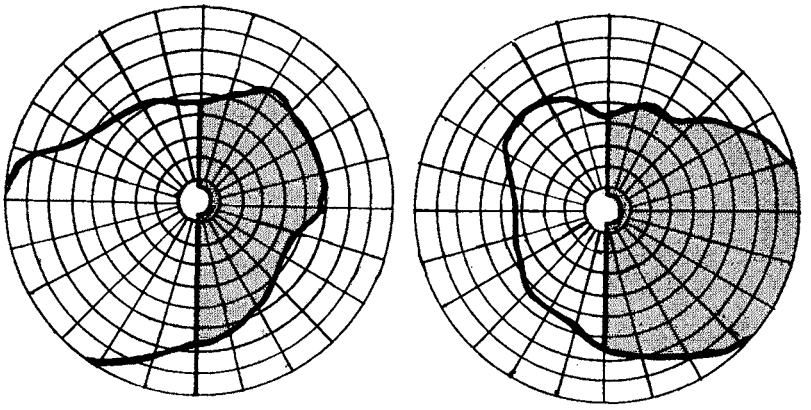


FIG. 54.—Right homonymous hemianopia.

surface of the chiasm there will follow respectively an inferior or superior altitudinal hemianopia. The dividing line for colour in this type falls short of the dividing line for form, where in the binasal or bitemporal forms these coincide (see Fig. 55).

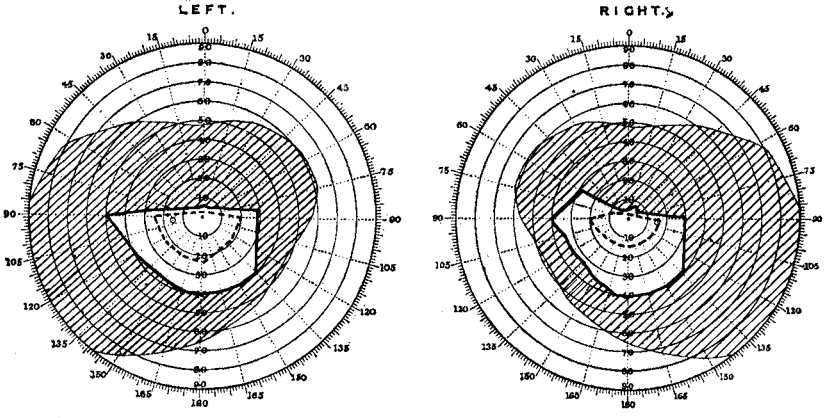


FIG. 55.—Altitudinal hemianopia due to gumma of the chiasm. (Peter.)

In cases of thrombosis of the Sylvian artery (middle cerebral) opposite the posterior segment of the decussation of the optic nerves and of thrombosis of the posterior cerebral artery, diagnosis may be suggested by the former producing a hemianopia of the

inferior quadrant and the latter a hemianopia of the superior quadrant.

Lesions of the Optic Tract.—In addition to the changes mentioned in the field of vision there are other forms of defect, as shown in Fig. 54, which illustrates the condition known as right hemianopia—the field of vision is deficient on the right-hand side, the macula in each eye having escaped. The nasal half of the right retina is not functioning, therefore its temporal field is absent. The temporal half of the left retina is also blind so that its nasal field is absent. The loss of half of the visual field is known as hemianopia. Homonymous hemianopia denotes the loss of the right or left half of the binocular field of vision. Fig. 54 illustrates a case of right homonymous hemianopia. A lesion producing this condition may be situated in any part of the visual tracts from the occipital lobe to the chiasma. In a right-sided homonymous hemianopia the opposite sides of the retina would be affected as indicated by the cross markings in each eye of Fig. 39. The optic tracts from the chiasma to the lower visual centres are composed of crossed and uncrossed fibres, the crossing having taken place at the chiasma. Therefore it will be seen that the lesion producing a right-sided homonymous hemianopia, as in Fig. 54, may be situated in the left optic tract (see Wernicke's hemianopic pupil reaction, p. 18).

Lesions of the lateral geniculate body cause homonymous hemianopia, while those of the **pulvinar and superior colliculus** (corpora quadrigemina) do not. As the macula is so extensively represented in the lateral geniculate body a lesion here would most probably cause not only an homonymous hemianopia but a central hemianopia or scotoma as well. The work of Rönne clearly demonstrates the degeneration in this area in connection with macular degeneration. This is referred to on p. 429. As the majority of people read from left to right a right-sided hemianopia is therefore much more quickly discovered than a left.

Lesions of the Geniculo-Calcarine Pathway.—Many cases of hemianopia are due to lesions in the occipital lobe or in the optic radiation. Both the grey matter and the subcortical white matter are invariably involved. Gunshot wounds, tumours, syphilitic changes or other diseases of the blood vessels are the commonest causes. Recently an elderly doctor wishing to discover what was wrong with his motor car got beneath it. On rising he suddenly became aware of the loss of one-half of his visual field. The diagnosis was that of a

thrombus in a branch of the posterior cerebral artery on the opposite side of the blindness. A lady whose blood pressure had risen to 250 millimetres suddenly lost one-half of her visual field. This was accompanied by speech defect which helped to locate the lesion in that part of the optic radiation contained in the internal capsule.

In the majority of cases of hemianopia the macular area in each field is present owing to each macula having escaped. The probable reason is the widespread distribution of the macular representation at the occipital pole. Before Lister and Holmes suggested this explanation it was believed that each macula had a bitemporal representation. Of this we are not at all certain.

Brouwer and Zeeman, by following up degenerative changes in animals, have shown that the fibres from the optic tract pass into the geniculate body in a very definite manner. The upper middle third of the geniculate body is composed of macular fibres, while the medial and lateral thirds are formed of fibres coming from the upper and lower peripheral parts of the retinæ respectively.

From the lateral geniculate body the fibres enter the posterior limit of the internal capsule emerging as the optic radiations (see Fig. 41), and ending eventually in the area striata of the occipital lobe. As mentioned in the previous chapter, the fibres from the dorsal half of the retina are projected from the medial part of the geniculate body along the dorsal portion of the optic radiations and enter the dorsal lip of the calcarine fissure, while the fibres from the ventral half of the retina remaining below, as in the optic nerves and tracts, sweep round the tip of the descending horn of the lateral ventricle and, passing below the ventricle, reach the ventral lip of the calcarine fissure.

Referring to Fig. 41, it is readily perceived that a lesion such as a tempero-sphenoidal abscess may damage the lower portion of the optic radiation and so cause a superior hemianopia due to interference with the fibres from the ventral half of the retina. Vascular lesions and tumours are among the commonest causes of injury to the optic radiation. Traquair illustrates the fields of vision produced by a glioma of the right parietal lobe which completely destroyed the optic radiations on the right side.

Migrainous patients sometimes describe an homonymous hemianopia due probably to vascular spasm in the vessels of the brain. In such cases Wilson says the blind area extends right up to the centre of the macula. Extreme fatigue will sometimes produce the

same result. At the end of a wearisome day, spent in X-raying large numbers of wounded at a casualty clearing station in France, the writer, upon sitting at the mess table, suddenly became aware of the fact that he could not see anything on the right side of his field of vision. The sensation brought with it slight nausea. After a few hours' rest the full field of vision returned; headache was not present. Reference will be made later to amaurosis fugax, which is a somewhat similar phenomenon (see Chap. XVII).

Prosper Veil suggests a classification of hemianopias to be conveniently remembered as basilar, pre-ganglionary and post-ganglionary cerebral hemianopias.

Fig. 56 illustrates left hemianopic fields which resulted immedi-

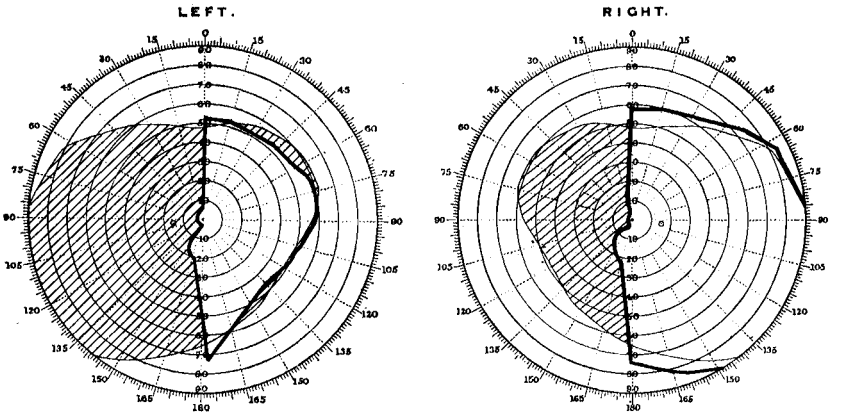


FIG. 56.—Left homonymous hemianopia due to severe loss of blood.

ately from extensive loss of blood in an attempt to commit suicide. For several years repeated observations were made but change in outline did not take place. Note the sparing of the macula and the peculiar bulge below the macula both of which remained unchanged.

During the Great War Holmes, Lister, Marie, Chatelin and many others collected material which demonstrated the effect of injuries in the region of the occipital lobes. From his investigations Holmes came to the conclusions embodied in the illustration (Fig. 43). A destructive injury of the optic radiations generally produced a complete hemianopia in which the blindness may or may not reach the point of fixation. Instead of a hemianopia there may be a large area of blindness. Fig. 57 illustrates a case described by Holmes, where a small penetrating missile entered the skull 7 cm. above the inion

and 5 cm. to the right of the sagittal suture. The exit wound was 10 cm. vertically above the tip of the left mastoid. At autopsy examination of the brain showed the entrance wound to be in the

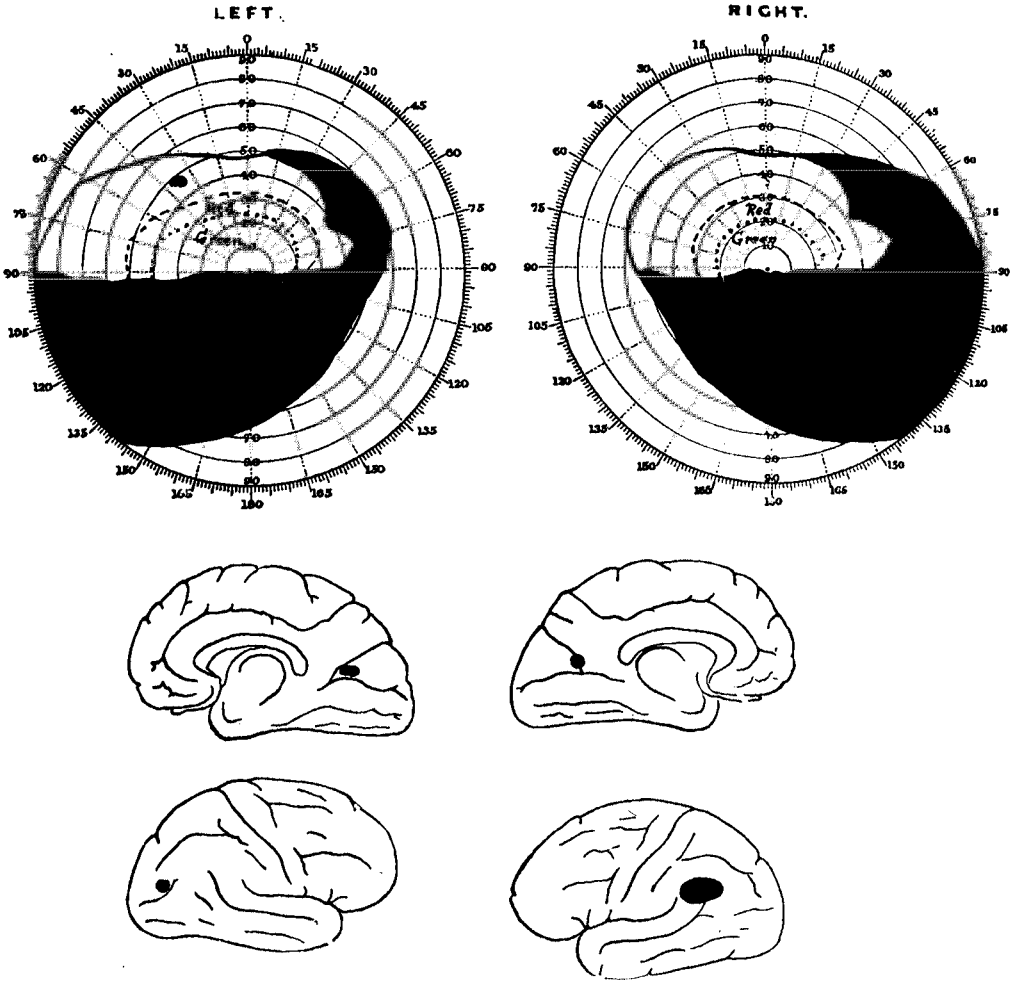


FIG. 57.—Passage of penetrating missile through both hemispheres producing destruction of dorsal portions of optic radiations in each. Fields of vision show the resulting loss in each lower half. (Holmes.)

middle of the lateral surface of the right occipital lobe some distance behind the parieto-occipital notch. From here the missile passed through the dorsal part of the optic radiations and emerged on the

mesial surface of the hemisphere in the angle between the calcarine and the parieto-occipital fissures. The missile then entered the mesial surface of the left hemisphere in the parieto-occipital fissure, the destruction it produced reaching to 0·5 cm. of the calcarine fissure. It passed through the dorsal portions of the optic radiations and made its exit through the anterior portion of the angular gyrus. When this patient was admitted to the Base Hospital he had marked loss of visual orientation in space; he was unable to recognise accurately the relative and absolute positions and distances of objects by sight alone. Central vision in each eye was 6/12. There was complete blindness in both lower quadrants. The loss of the inferior portion of the field of vision was due to the destruction of the dorsal portion of the optic radiations. The defect in the upper right quadrant was due to softening under the gyrus angularis.

Homonymous hemianopia when due to vascular disease does not so frequently involve the fixation area (see Fig. 56). This is due to the overlapping of the arterial supply by the posterior and middle cerebral arteries.

We have seen that hemianopia results from lesions both of the optic radiations and the occipital cortex. A pure hemianopia without other associated symptoms will be due most probably to a lesion of the occipital cortex and subcortical white matter, whereas a hemianopia due to a lesion of the optic radiations may be accompanied by sensori-motor phenomena. This is due to the proximity of the internal capsule. In a case under Peter's care a left-sided hemiplegia preceded by left hemianæsthesia, left homonymous hemianopia and conjugate deviation of the eyes to the right with marked pain in both arm and leg, indicated the lesion to be in the neighbourhood of the optic thalamus (see p. 169).

The suggestion that the fibres in the optic radiation have a clear-cut anatomical arrangement has been put forth by Holmes and is accountable therefore for the sharp horizontal margins of the resulting hemianopias found in injuries of this region. Long previously Meyer had shown that three bundles of filaments which make up the geniculo-calcarine path remain of fairly uniform thickness throughout.

Some cases of occipital injury which have resulted in a hemianopia have gradually shown recovery. Hine has studied such cases. Recovery, he found, took place from the centre to the periphery, but he noted that in the case of cerebral loss of substance the scotoma

gradually contracted from the periphery to the centre as the surrounding œdema of the brain subsided. The field usually returns in the upper segment before the lower. Colour perception follows the perception for white. These are cases of concussion injury.

" Lesions of the lateral surfaces of the hemispheres, particularly of the posterior parietal regions, may cause certain disturbances of the higher visual perceptual functions with intact visual sensibility, as loss of visual orientation and localisation in space, disturbance of the perception of depth and distance, visual attention loss and visual agnosia." (Holmes.)

As hemianopic fields of vision occur in lesions both of the supranuclear and infranuclear paths it is necessary to differentiate them by some means. In the first place, in lesions below the primary visual centres the Wernicke's pupillary reaction is present. Secondly, optic atrophy is present or will in time supervene. Thirdly, complete forms of hemianopia are found in lesions of the optic tract, but those occurring in the geniculo-calcarine path may be small, gradually becoming larger until even the macula may become involved. Fourthly, in tract lesions the macula is usually involved, but is generally spared in lesions of the geniculo-calcarine path.

A more thorough study of perimetry may be made by reading Peter's " Hand-book of Perimetry " or the magnificent volume by Traquair.

Valuable contributions to the study of perimetry have also been made by C. B. Walker, Professor Roenne, Mayer, and many others.

CHAPTER VIII

THE MACULA

Owing to its peculiar structure and position, the macula is the most sensitive spot in the retina. The development of such an area coincides with the higher development of the nervous system. This area is first noticed in fishes, is much better developed in birds and approaches the more normal macula in the primates. In *homo sapiens* the macula reaches its highest development in order that it

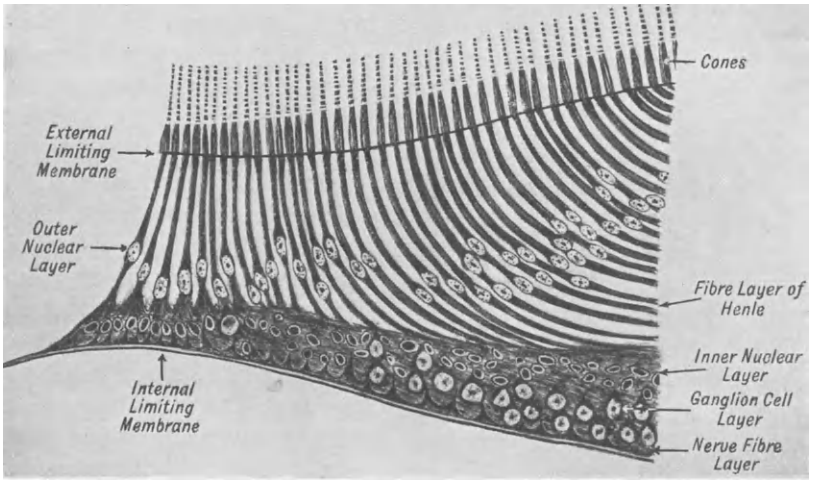


Fig. 58.—Diagram of a section through the fovea centralis. Tracing from a photograph. (Preparation by Golding-Bird.) (From Quain.)

may subserve the function of accurate vision. Here the finest detail is perceived with extraordinary acuteness. Fritsch, who studied the development and structure of the macular area in apes and man found great variation in the depth and extent of the fovea in different individuals. Treacher Collins and the author examined several cases where the central visual acuity was subnormal and had been since infancy, yet the appearance of the fundus in these cases was normal, as was the refraction of the eye. The conclusion

was that the macula had never developed and that the area in these cases was not differentiated from the remainder of the retina.

Structure of the Macular Region.—This area, 1 to 3 millimetres in diameter, situated 3·5 millimetres from the temporal edge of the optic disc, and almost 1 millimetre below the horizontal meridian is the thinnest part of the retina. It consists of the macula lutea (yellow spot) and the fovea centralis (central depression) in its centre. The yellow tint of the macula is not absent in the centre of the fovea, but is merely less conspicuous owing to the thinness of the layers. This yellow colour is peculiar to the primates ; in man it develops after birth. The pigment may be seen by red free light, especially in dark people. There are no rods in the fovea, which is the central depression of the yellow spot, while the cones (4,000 in number) are packed so closely together that they are flattened where they are in contact, each cone having its own nerve fibre. These fibres passing to the brain in the optic nerve are gathered together as the papillo-macular bundle (see Fig. 38) and constitute more than one-third of the whole number of fibres in the optic nerve. (Elliott Smith.) (Fig. 58.)

The effect of light on the neuro-epithelial elements (rods and cones) is to produce a swelling of the rods and a contraction of the cones, while the pigment granules of the retinal pigment wander from the bodies of the cells into processes between the rods and cones ; but in the dark the converse takes place, the rods become thinner and the cones lengthen. The presence only of cones in the fovea has two effects, acuteness of vision here is extreme, while the appreciation of light of low intensity is poor. The sensitivity of the retina to light, whereby such a condition as dazzle is gradually reduced to clear vision, is brought about by light adaptation, while conversely, invisible objects gradually become visible in the dark by dark adaptation. In the light-adapted eye the fovea is the most sensitive part of the retina, while in the dark-adapted eye the para-central region, that is the region consisting of both rods and cones, becomes the most sensitive area. Hence the physiological night blindness of the fovea, as described by the older astronomers who discovered they could see a minute star when their eyes were directed to an adjacent point, but not to the star itself. It was believed at one time that visual purple did not exist in the cones, but this is now disputed ; the visual purple is bleached by light and regenerates on the withdrawal of light. The lack of Vitamin A

FIG. 1. THE NORMAL MACULA

At this particular area a fine pigmentation is observed which is free from the presence of vessels. As in the illustration a bright shining spot is frequently observed in the centre of the pigmented area. This is the fovea.

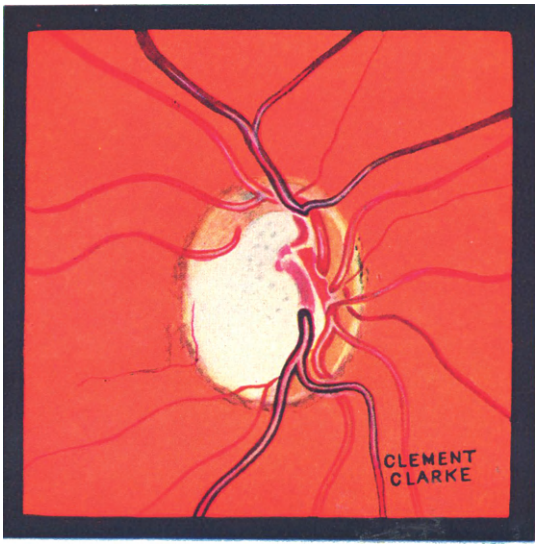


FIG. 2. PARTIAL PRIMARY OPTIC ATROPHY OF DISSEMINATED SCLEROSIS

Compare the pink colour of the inner band of nerve substance with the pale appearance of the outer half. The pallor extends to the edge of the disc.

[*To face p.* 144.

hinders the process of regeneration and so may be a factor in night blindness. The amplititude in sensitivity of the retina is small at the fovea, but is much greater at the periphery. Visual acuity of the retina shows a much smaller amplitude, especially at the fovea.

The macula is devoid of blood vessels. The perimacular region is supplied by small twigs from the superior and inferior temporal divisions of the central artery and also by small macular branches proceeding directly from the temporal side of the optic disc (see Plate II). The central area around the fovea devoid of vessels measures about 0·5 millimetre in diameter. The nerve fibres from the macula pass in an arched manner to the temporal side of the disc forming the papillo-macular bundle. This bundle soon passes to the interior of the optic nerve (see Fig. 38). The position of the macular fibres disturbs the orderly arrangement of the retinal fibres generally, so that on the temporal side of the optic disc both above and below the macular fibres, are the arcuate fibres (arcuate juxta-papillary bundle). It is due to this arrangement of the fibres that in some cases of papilloedema the appearance of a macular fan is presented. The œdema passes through the limiting membrane, finding its path along the curved junction of the macular and arcuate fibres.

The macular area is the most responsive portion of the retina to light stimulus. Its fibres are not only for the conduction of visual impulses, but also contain afferent pupillomotor fibres which pass to the constrictor centres after decussation.

Ophthalmoscopic Appearance.—In order to view the macular region with an ophthalmoscope, the examiner should employ the direct method so as to obtain the greatest magnification possible. It is necessary to have the pupil dilated, but a skilled ophthalmologist can see this area through a normal pupil, especially by means of an electric ophthalmoscope, the beam of which is not too powerful, such as the May electric instrument. The method of examination is as follows : Look for the temporal side of the optic disc, gradually make the light traverse a path outwards until a finely pigmented area is reached lying free from vessels which are above and below (see Plate XIII, which illustrates a normal macular area). Frequently in the centre of this area—the fovea—a bright shining spot is seen surrounded by a reddish coloured area. The yellow pigment is not seen, but when the macula is examined by red free light the pigment appears black, with a faint halo reflex around the macula.

Students are taught to examine optic discs, but seldom does one find a search being made for this minute but particularly important area of the retina.

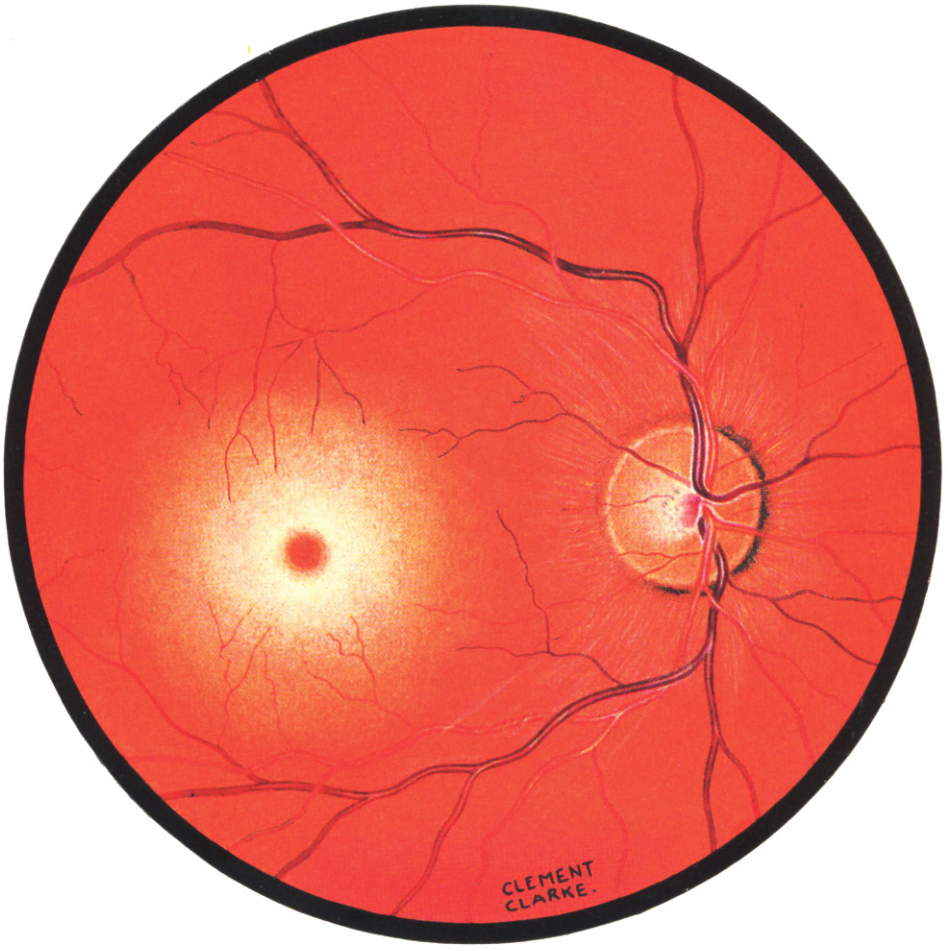
Macular Diseases related to Diseases of the Nervous System.—There are many conditions which result in degeneration of the tissues of the human body. Such degeneration may be physiological, deficient or toxic in nature. But there is a degeneration which is due to a premature failure in the vitality of the cells affected. This is termed *abiotrophy*. As Collins and Mayou have stated, "there may be a precociousness in degeneration just as there may be a precociousness in development. These changes are not produced by inflammation nor disturbance in nutrient supply, they are always bilateral and usually hereditary."

Under the title of abiotrophy may be grouped *hereditary ocular palsies, retinitis pigmentosa, infantile and juvenile amaurotic family idiocy, symmetrical macular pigmentary degeneration, Leber's optic atrophy* and *Niemann-Pick disease.*

Infantile and Juvenile Amaurotic Family Idiocy, or Cerebro-Macular Degeneration.—Amaurotic family idiocy, as described by Warren Tay in 1881, and more fully by Sachs in 1887 (also by Kingdon and Russell in 1897), is a familial disease characterised among other things by changes in the macular region and by atrophy of the optic nerve. It frequently occurs in several members of the same family. Epstein records four cases of this condition in one family. The parents were Russian Jews who had eight children. Of these the first child, a girl, was normal in every way until about seven months of age, when she became helpless and blind, had many convulsions and died when she was eighteen months old. The third child, a boy, was well during the first few months of life, became blind when about six months old, could not support his head or sit up, had many convulsions and died at two years of age. The sixth child, a girl, was normal up to six or seven months, then had frequent attacks of convulsions, became blind and died at twenty-two months of age. The eighth child, a boy, aged eight months, was apparently well until six months of age, when the critical symptoms began. Examination of the retina by the ophthalmoscope revealed a cherry red spot in each eye. The Wassermann, tuberculin and spinal fluid tests were negative.

Pathology.—The pathology characteristic of amaurotic family idiocy is widespread degeneration of the ganglion cells of the entire

PLATE IX



AMAUROTIC FAMILY IDIOCY. (Tay-Sachs' Disease)

The ophthalmoscopic appearance of the retina in this disease shows the typical cherry-red spot situated in a greyish-white area. The macula, which is of a dark red colour, is much deeper than the cherry-red macula seen in embolism of the central artery.

[*To face p.* 146.

nervous system. The ganglion cells of the retina are involved, but otherwise the retina is absolutely normal. The ganglion cells show progressive loss of the Nissl substance in all the neurons of the body and an increase of the neuroglia fibril substance to an abnormal degree. The loss of the Nissl substance is followed by the formation of vacuoles in the cytoplasm, and finally shrinkage or disappearance of the cells. The cherry red spot seen at the macula in embolism of the central artery resembles that seen in amaurotic family idiocy, but the pathology is quite different. In the former there is a coagulative necrosis of the ganglion cells, which cells rapidly become absorbed and disappear, whereas in the latter the long continuance of the opacity of the retina is due to the long time the degenerative changes in the cells go on before they finally disintegrate. The appearance of the cherry red spot therefore in amaurotic family idiocy persists until death, whereas in the case of embolism of the central artery it disappears in two or three weeks' time.

Etiology.—The infantile form of amaurotic family idiocy is confined to the Jewish race. The disease seems to be more common in Polish Jews, but is not rare in Hebrews of other nationalities. The causes of the pathological changes are unknown.

Symptoms.—The child is normal at birth, the symptoms usually appearing between the third and sixth month. The child becomes listless and shows weakness of the muscles of the neck and back. It loses interest in surrounding objects. The muscle tone of the whole body is greatly lowered and the child begins to lose its sight. It is at this stage of the disease that ophthalmoscopic examination of the retina shows the typical cherry red spot which is found at the macula (see Plate IX), the macula itself being situated in a greyish white area which is about twice the size of the optic disc. Although these appearances are not present at birth, they remain until death. The optic nerve becomes atrophied, and complete blindness ensues. The limbs become weak and ultimately paralysed. The paralysis may be either flaccid or spastic. The infant becomes markedly sensitive to various stimuli, especially to sound, and starts or exhibits epileptiform twitchings on the sudden approach of the sound of noises. The final stage of the child shows the development of marasmus, the optic disc has become pale and at about two years of age death ensues, either by marasmus or possibly by some intercurrent illness, such as pneumonia.

Diagnosis.—The characteristic appearance of the child, the pre-

sence of the cherry red spot at the macula and occasionally nystagmus, together with the familial nature of the affection, point out the diagnosis.

Juvenile Type of Amaurotic Idiocy.—The juvenile form of amaurotic family idiocy described by Batten, Batten and Mayou, Spielmeyer and others, closely resembles the infantile type, although it may not have the same spectacular features. It begins in the second half of childhood, between the ages of six and twelve years, usually running a course of two years, when the child dies in blindness.

Sjögren has recently published a volume in which his main object has been to make a detailed clinical study and investigation of the hereditary features of the disease. He carefully selected 120 cases. From his book it is shown that the condition of juvenile amaurotic idiocy is relatively more frequent in Sweden than elsewhere. In most of the cases he quotes the affected children have been quite normal in their development up to about the sixth year, when a rapidly progressive loss of vision sets in together with mental deterioration and, in some cases, epileptic seizures. The psychical symptoms are constant. These little patients, after normal development, become stupid and lose interest. They cannot manage their school lessons, become irritable and are easily made to cry. They find difficulty in speaking and finally all intelligence vanishes. Sjögren comes to the conclusion that the condition is inherited, with a very slight degree of probability, as a recessive single gene character. He points out that it must be sharply defined from the infantile type of the disease. In examining the history of the parents and other relatives in the cases analysed in his book he found that dementia præcox, oligophrenia and epilepsy occurred in an extraordinarily large number of cases.

In 1925 the histology of juvenile amaurotic idiocy was described very thoroughly by Greenfield and Gordon Holmes. They found changes in the nerve cells in the cerebral cortex, optic thalamus, the putamen and globus pallidus, the brain stem and the spinal cord. The changes in the cerebellum were of special interest. In this disease the primary changes are obviously confined to the neurons, the proliferation of the neuroglia seen in the most affected parts as in the cerebral cortex, the cerebellum and the retina being evidently secondary thereto. The essential lesion of the disease is a special form of degeneration of the cell body associated with the deposition

of a peculiar form of lipoid. In the prelipoid phase Schaeffer describes it as " lecitinoid degeneration." It is at this stage that the infantile cases usually prove fatal, but in cerebro-macular degeneration a further conversion of the prelipoid substance causes scharlach-staining granules to appear, while in the family idiocy without blindness of Erwachsen a still more advanced osmio-reductive phase is reached (Hurst). Greenfield and Gordon Holmes state that the most striking feature in the histological changes in the retina was undoubtedly the degeneration of its outer layers and their replacement by proliferated neuroglia, while the inner layers, that is the nerve fibre, ganglion cells, inner reticular and inner nuclear layers, remained intact or were affected much less severely. There was an affection of the nerve cells of the ganglion layer similar to that which was found in the cells of the central nervous system.

The retinal changes in the juvenile type of amaurotic idiocy may be therefore defined as a primary change in the ganglion cells combined with an independent degeneration of the outer neuronic element of the retina, which in some cases extends to the inner nuclear and inner reticular layers. It is this affection of the outer layer that distinguishes the retinal lesions of the juvenile from those of the infantile type of family amaurotic idiocy. In the latter the essential changes are confined to the ganglion cell layer. Ophthalmoscopic changes are observed to commence with a small white area at the macula surrounded by a reddish ring. As these macular changes become more pronounced the retina becomes more and more atrophied. The retina shows a streaky white appearance with some black pigment scattered finely over it. In the late infantile and juvenile forms the cherry red spot is not seen, although the macular area may be pigmented. In practically all the cases described the optic discs were abnormally pale, though actual atrophy became evident only some time after loss of vision was noted.

Torrence from his investigations concludes that there are connecting links in regard to age incidence, race proclivity and eye symptoms between the infantile and juvenile forms. Nystagmus and optic atrophy occur in both. The cherry red spot may change and come to resemble the pigmented macula of the juvenile form. Also, he says, some juvenile forms show the cherry red spot. In some families with juvenile types of amaurosis there is a mixture of type, some with cherry red spots and others with little or no retinal change. Sjögren, however, states they must be sharply defined.

Differential Diagnosis.—As spasticity may be present it is not always easy to differentiate the disease from ordinary diplegias and paraplegias. The retinal changes, however, are not present in the latter. The onset of blindness, too, serves to distinguish cerebro-macular degeneration from rickets, a somewhat common error in diagnosis.

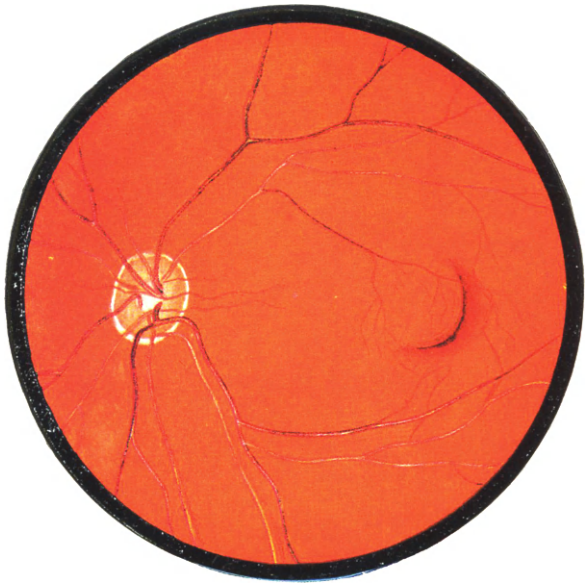
Niemann-Pick Disease.—There is yet another disease which strongly resembles the Tay-Sachs' disease. It is known as the Niemann-Pick disease, which also occurs in infants, beginning about the third or fifth month. It is also familial, found mainly among Jews, and terminates life at the end of the second year. The characteristic appearance of the fundus in Tay-Sachs' disease may or may not be found in the Niemann-Pick disease. This syndrome indicates an enlarged liver and spleen, together with a brown tint of the skin. The patients are anæmic in appearance. There is widespread lipoid degeneration of the bodily organs, including the brain.

Familial Occurrence of Macular Coloboma associated with Apical Dystrophy of Hands and Feet.—Arnold Sorsby recently showed a number of cases illustrating this group. The colobomata of the macula are of the pigmented variety. The parents were not consanguineous and no macular coloboma could be traced in any members of the mother's family. The apical dystrophy of hands and feet consists in the absence of nails, especially on the index fingers and big toes, stunting and even absence of phalanges, tendency to bifurcation of the terminal phalanx of the thumb and big toe. The intelligence of this group is normal and there is no history of mental deficiency.

Sorsby has also grouped together under the term " central and para-central tapeto-retinal degeneration " three distinct types of familial retinal degeneration : first, hereditary optic atrophy, of two types—recessant and dominant ; second, hereditary pigmentary degeneration—so-called retinitis pigmentosa ; third, hereditary macular degeneration—in which the lesion is confined to the macula and makes its appearance at different ages in different families, but always at the same time in the same family. Sometimes the lesion is present at birth, at other times it occurs at the second dentition and sometimes in adult or later life.

Cocayne, commenting on Sorsby's group, said that he did not think that it was necessary to believe that the defect in the eye was in any way associated with the skeleton one ; he could not see any

PLATE X (R. Lindsay Rea)



ACUTE PRIMARY MACULAR DISEASE

FIG. 1.—The earliest sign of any change taking place is a crescentic hæmorrhage which later comes to surround the macular area, the blood changing to pigment and enclosing a yellowish area, resembling an acute choroiditis. Central vision is rapidly lost. Two years previously the right eye was similarly affected.

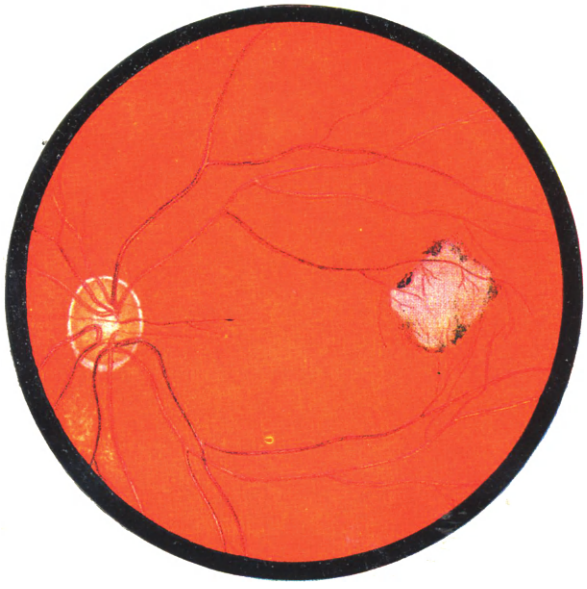


FIG. 2. -A year later the macula has assumed this appearance, its edges being marked by heaped-up choroidal pigment. During the succeeding five years a further atrophy took place around this area. Central vision was entirely lost.

[*To face p.* 150.

objection to regarding this as an instance of linkage of two unrelated defects, both appearing first in the mother. It was possible that one of that mother's parents, instead of having one mutation in a germ cell, had two mutations affecting two independent genes in the same chromosome. If so, both the defects were bound to be transferred together to every affected member since they were carried in the same chromosome, unless crossing-over took place. In man few examples of linkage were on record. He said that in the Laurence-Moon-Biedl syndrome there was recessive polydactyly which occurred fairly commonly without the retinal defect, which he thought was probably due to another gene altogether.

In dealing with the retinal abiotrophies, Arnold Sorsby has associated the following conditions :

Retinal Abiotrophy

With central nervous system.	With skin.	With skeleton.	With lipoid metabolism.
Tay-Sachs' disease.	Gronblad's syndrome.	The S. family. (Sorsby's cases).	Niemann-Pick disease.
Batten-Mayou disease.			
Laurence-Moon-Biedl syndrome.			

Among the cases seen at an ophthalmic hospital, dystrophy conditions of the macula are not so commonly met with as inflammatory conditions. Pagenstecher and Genth in 1875 described a macular disease which was of an inflammatory type. Elschnig in 1919 described a discoid disease of the macula, while later Junius and Kuhnt published a work in 1926 on Disciform Degeneration of the Macula. Rayner Batten, brother of F. E. Batten, wrote in the " Transactions of the Ophthalmological Society " in 1921 classifying macular disease under two headings, familial and non-familial. We have already dealt with the familial types, but mention must be made of the non-familial as classified by Batten.

NON-FAMILIAL

(1) *Congenital :*

(*a*) Developmental.—True coloboma, *i.e.*, congenital defect in the choroid and retina at this spot.

(*b*) Non-developmental.—False coloboma, possibly due to intrauterine inflammatory process.

(2) *Juvenile :*
(*a*) Primary macular disease, followed by secondary optic atrophy.
(*b*) Secondary macular disease.—Following optic neuritis. Coincident with secondary optic atrophy.
(3) *Adult :*
(*a*) Progressive.—Extending to choroido-retinitis beyond macular area.
(*b*) Non-progressive.—Macular area only involved. Permanent macular changes, but partial, or complete recovery of sight.
(4) *Senile :*
(*a*) Senile degeneration
(*b*) Tay's choroiditis.
(*c*) Senile macular choroiditis.
(*d*) ? Retinitis circinata.

Rayner Batten established the condition of acute primary macular disease as a distinct entity from the familial group. The author had the privilege of being closely associated with Rayner Batten for many years at hospital, and was continually in consultation with him about cases of this kind which presented themselves. Many cases appeared as slight forms of choroido-retinitis in the macular region, with central vision affected in a more or less degree. There is no family tendency to this condition. It is a disease of young adult and middle life. It sometimes runs an extremely acute course with marked change at the macula, together with a slight vitreous haze in this region. It affects both eyes. The milder type does not always begin simultaneously in both eyes ; sometimes months intervene between the onset in each eye. The disease runs a definite course and then becomes quiescent. Plate X (Fig. 1) illustrates the beginning of a disease in an adult aged sixty-three. The right eye was affected two years previously. Now a crescentic-shaped hæmorrhage was visible on the outer circumference of the macula of the left eye ; gradually the hæmorrhagic area changed and the macula itself became yellowish in colour, until after a period of two years the disease finally abated, leaving this condition (Plate X, Fig. 2), a picture almost identical with the macula of the right eye.

A great many cases merely show a pigmentary change such as

illustrated by Plate XI (Fig. 1). One can watch with the ophthalmoscope the changes slowly taking place. Sometimes among the pigment are found minute hæmorrhages, and while these hæmorrhages persist central vision steadily deteriorates. The visual acuity of the case, illustrated by Plate X, fell from 6/5 to 3/60 in two years and then slowly improved until the vision became 6/60. The causes of this disease are various ; they are undoubtedly of a toxic nature, the focus being dental sepsis, nasal sinuses, gonorrhœa,

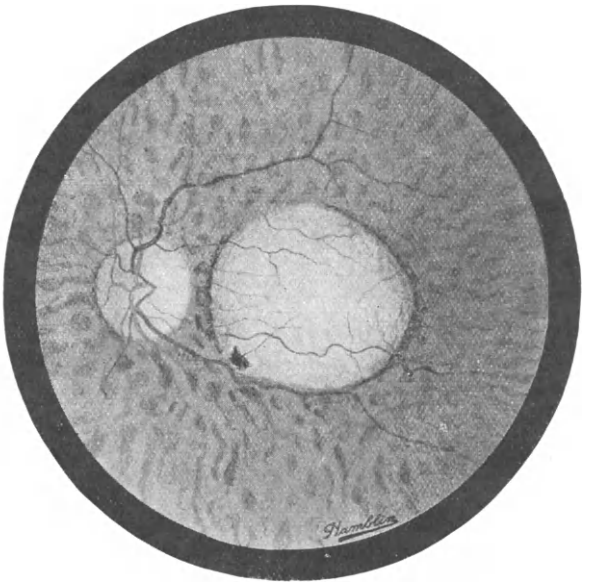


FIG. 59.—Typical raised mass at macula. (Davenport.)

influenza and pregnancy (Batten). Neither Batten nor the author ever saw a case which could be described as syphilitic in origin.

A marked symptom of macular change of which a patient may complain is distortion of objects ; *e.g.*, a telegraph pole appears to be twisted or bent at the part looked at, while down below it is straight (retinal metamorphopsia) ; red and green shades of colour are difficult to distinguish and a scotoma for blue develops before that for red and green. Visual acuity fails.

In elderly people there is a definite macular degeneration associated principally with arterio-sclerosis, indeed I believe it is quite

common in patients over sixty-five years of age. Both hæmorrhages and pigment are found plentifully scattered over and around the macula. Gradually the hæmorrhages are replaced by exudates, frequently leaving a condition known as retinitis circinata (Plate XI, Fig. 2). We have observed many cases of this type ; sometimes the retinitis circinata is found in one eye only, the other eye showing macular change of a pigmented character, but the disease is always bilateral and sometimes occurs a year or so before death. The central vision becomes very poor indeed, especially when retinitis circinata is present. It is interesting to note that Plate XI, Fig. 2, is the actual picture of a complete macular degeneration with surrounding retinitis circinata, which resulted from the patient, a young woman, being struck by lightning. There is complete central blindness. This case, together with several others, has helped to establish in the mind of the author the fact that certain macular conditions may be traumatic in origin.

Occasionally a large hæmorrhage takes place in the choroid behind the macula. R. C. Davenport has described several of these cases. The illustration (Fig. 59) was kindly lent to me by him ; the white mass at the macula was organised matter produced by a deep hæmorrhage near which cholesterin crystals may be found. The white ring of exudate, known as retinitis circinata, is made up of fat cells and deposits which have been concerned in the disintegration of red blood cells present as the result of a deep retinal hæmorrhage. Nettleship in 1902 described similar cases.

There are many border-line cases where it is difficult to say whether they are maculo-cerebral types of heredo-degeneration or not. Braun, Behr and others have described such cases where there was a definite onset of macular change in young adults which produced diminished vision, sometimes only for colours, others for white. Close examination of the nervous system may reveal slight weaknesses such as of the facial nerve, etc., while the associated history may reveal definite disease of the nervous system in close relatives.

Waldstein has demonstrated two cases, that of a young man and his sister, both in the early twenties; one had a macular degeneration, the other an aplasia of the macular region with lack of a macular and foveolar reflex and the presence of nystagmus. So that two absolutely different kinds of macular affection (degeneration and aplasia) may occur in the same family.

PLATE XI (R. Lindsay Rea)

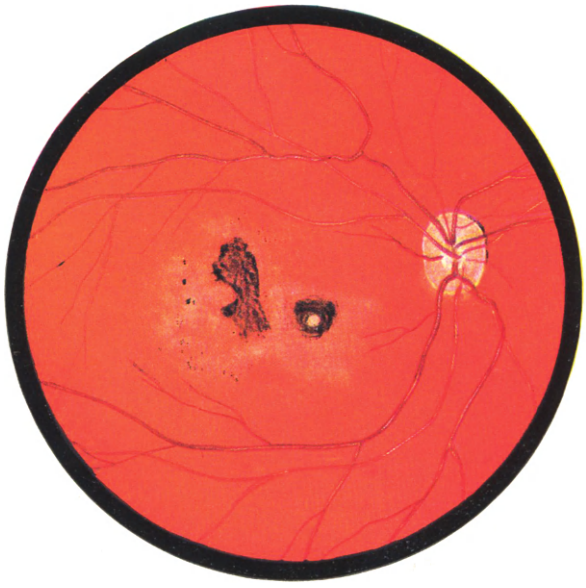


FIG. 1. BILATERAL PIGMENTARY DEGENERATION OF THE MACULA

Occasionally this is seen in one eye only but frequently it is bilateral. It is found not only in adolescence but in middle-aged and elderly people also. A similar macular change is found in the Juvenile Type of Amaurotic Family Idiocy.

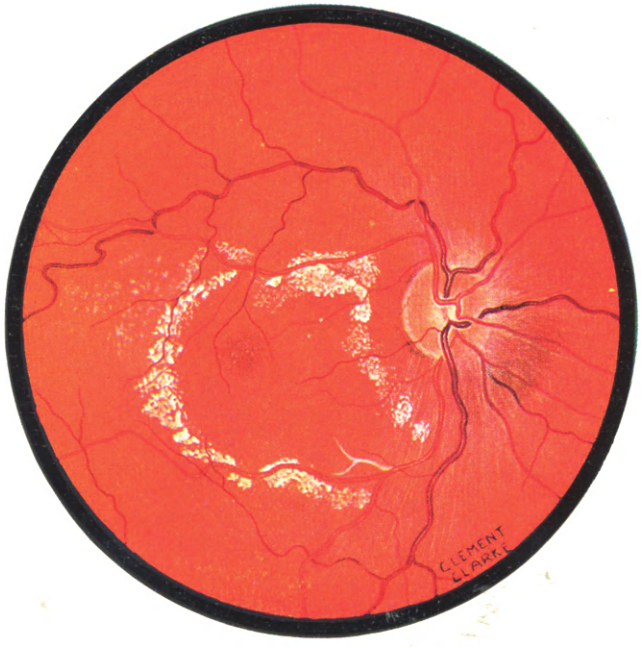


FIG. 2. RETINITIS CIRCINATA

This condition is observed in elderly arteriosclerotic subjects. A wide distribution of fine hæmorrhages is at first found giving way to the presence of exudates arranged in the typical manner seen in the picture. The macula and perimacular area become functionless. This ophthalmoscopic picture was of a middle-aged woman who was in normal health but had central blindness in this eye. The only history obtainable that might have led to such a condition was that once she had been struck by lightning.

[*To face p.* 154.

Although many consider macular changes of senility as familial I agree with Ralph Lloyd that senile macular degeneration is but one of a group of symptoms resulting from vascular changes in the brain.

Macular colobomas, like other colobomas of the choroid, may be familial.

A report on bilateral macular coloboma in monozygotic twins has just been published by Gesell and Blake. They include a full bibliography of the literature on ocular correspondence in twins.

Diplopia without extraocular palsies has been observed by Kubie and Beckmann. They think this condition is due to impaired macular vision, the patient attempting to use the adjacent areas of the retina in which visual acuity is greater.

In case of loss of macular perception in one eye the scotoma may be delineated by the stereoscopic method, using Lloyd's stereo-campimeter slate.

The macular region can be observed through an undilated pupil by using the Cardell polarized ophthalmoscope. The use of this instrument eliminates the surface reflex from the cornea.

CHAPTER IX

LOCALISING VALUE OF OCULAR SYMPTOMS IN THE DIAGNOSIS OF DISEASES OF THE BRAIN

Mid-brain Lesions.—A brief reference may be made to the anatomy of the mesencephalon or middle portion of the brain, tumours of which usually involve some of the nerves connected with the eye. Fig. 60 shows the front view of the mid-brain, pons and the medulla (the brain-stem), the mid-brain consisting mainly of the crura-cerebri and resembling two large rope-like strands which emerge together from the upper portion of the pons varolii and diverge as they proceed upwards to enter the cerebrum.

At the point where each crus disappears into the corresponding side of the cerebral hemisphere, it is embraced on its outer side by the optic tract. From the interpeduncular space emerge the third cranial or oculo-motor nerves ; this little space is therefore called the sulcus oculo-motorius. Winding round the outer side of each crus and just above the pons is found the fourth cranial or trochlear nerve. From the ventro-lateral surface of the pons the fifth cranial or trigeminal nerve proceeds, while from the area between the lower part of the pons and the upper part of the medulla the sixth, seventh and eighth cranial nerves are seen emerging closely together. It will be observed, therefore, that if a tumour or hæmorrhage implicates one nucleus, most probably its neighbour and surrounding structures will also be involved.

Encircling the outer side of the crus is seen the fourth nerve, which is a thin, fine structure. It comes from the region of the corpora quadrigemina. If this area is the seat of a lesion there will be loss of up and down head movements, together with a diplopia produced by paralysis of the superior oblique muscle.

While it is usual with lesions of the mesencephalon in the region of the posterior commissure to meet with bilateral ptosis of the so-called " sympathetic type " often associated with small pupils and sometimes complete loss of upward movements of the eyes, yet sometimes with lesions in the same region one may meet with a

corresponding opposite (Collier) in that the lids are unduly retracted or tucked back and that the pupils may be large, also that retracted lids may be the sole sign of a lesion in this region. Probably the

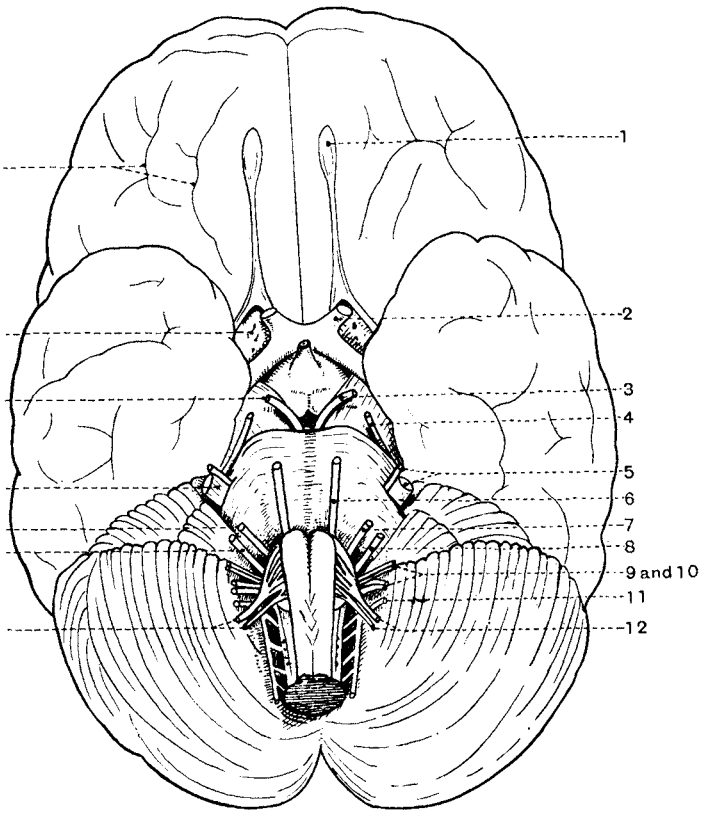


FIG. 60.—The base of the brain with cranial nerves showing their relationship to the brain stem.

1. Olfactory bulb.
2. Chiasma (with pituitary body lying behind) formed by the union of the optic nerves in front and giving off the optic tracts behind.
3. Third or oculo-motor nerve.
4. Fourth (patheticus) nerve winding round the crus cerebri.
5. Fifth (trigeminal) nerve emerging from antero-lateral aspect of pons.
6. Sixth nerve.
7. Seventh facial nerve.
8. Eighth (auditory) nerve.

9 and 10. Vagus and glosso-pharyngeal nerves.

11 and 12. Spinal accessory and hypoglossal nerves.

Sixth, seventh and eighth nerves emerge between the pons and medulla. (T. Walmsley and M. E. Rea.)

lesion responsible for the tucked lids and large pupils is situated farther forwards than is the lesion productive of ptosis and small pupils. The clinical points which may serve to distinguish ophthalmoplegia of the supra-nuclear origin from that due to a nuclear lesion are tucked lids with unvarying parallelism of the eyes and therefore no diplopia, contrasted with loss of parallelism, diplopia, ptosis and internal ophthalmoplegia in the nuclear lesions.

Tumours of the Mid-brain, if in the region of the crus cerebri, give rise to hemiplegia and hemianæsthesia of the opposite side. The third cranial nerve may be affected, and if the tumour is large it may press upon the fourth, fifth and sixth cranial nerves. If the tumour involves the tegmental region of the mid-brain an Argyll Robertson pupil with defective conjugate movements of the eyes will be found. Alpers and Watts found quadrantic fields.

Tumours of the Pineal Gland.—The pineal gland or epiphysis projects backwards and downwards from the posterior wall of the third ventricle above and between the superior corpora quadrigemina; it conceals the posterior commissure and the median groove.

Tumours of the pineal gland are rare. From its position a tumour may press upon and close the aqueduct of Sylvius, producing internal hydrocephalus, leading to hypopituitarism. There may be a disturbance of growth if the tumour develops in young boys.

The following signs and symptoms of a tumour (consisting of two kinds of cells lying between cellular tissue septa) of the pineal gland are given by De Monchy :—

(*a*) Increased intracranial pressure, producing a papillœdema of four dioptres in each eye.

(*b*) Apparent Argyll Robertson pupils.

(*c*) Rhythmical spasm of convergence.

(*d*) Almost complete paralysis of upward and downward movement of the eyes.

(*e*) Disturbance of internal secretions are the same as those found in dystrophia adiposo-genitalis.

Some years ago Kinnier Wilson drew attention to a condition of the pupils known as ectopia pupillæ, or corectopia, which he observed in certain mesencephalic lesions, such as tumours of the third ventricle, etc. The pupils are seen to be displaced upwards and inwards, and sometimes the corectopia remained permanently to the end, in one it was seen only during congestive attacks, and then the

pupils returned to their normal positions. Tumours of the pineal gland or neighbourhood may show the same abnormality.

Dr. Worster-Drought has forwarded to me his notes on three cases of tumour of the pineal body.

One of these cases was a young man aged twenty-three years, with a history of three months' occipital headache, occasional dizziness and drowsiness, who suffered attacks of " mental vacancy " during which he would sit or stand motionless for periods varying from a few minutes to half an hour. This patient was somewhat obese and his figure showed some trace of a feminine type. The genitalia and secondary sexual characteristics were well developed. There was a papillœdema of 5 dioptres in the right eye and 4 dioptres in the left, with slight concentric diminution of both visual fields. His blood pressure was low, 100 systolic and 80 diastolic. X-ray examination showed an enlarged sella turcica and some erosion of the posterior clinoid processes. The pineal body was calcified and mesial. The cerebro-spinal fluid showed normal protein content. A ventriculography showed moderate dilation more marked on the left side. The patient died two weeks after a decompression operation.

Dr. Carnegie Dickson gave his summary of the autopsy as follows :—

Congenital epidermoid tumour (pearly tumour or cholesteatoma) of the pineal anlage, compressing the corpora quadrigemina and the lamina (valve of Vieussens) and subjacent aqueduct of Sylvius, producing marked hydrocephalus, including dilation and bulging of infundibulum, and this probably accounting for provisional diagnosis of " pituitary tumour."

In a summary of 113 cases of tumour of the pineal gland macro-genitosomia præcox, a syndrome consisting of mental precocity, abnormal growth of the skeleton and premature development of the genitalia and secondary sexual characteristics, was stated by Haldeman to be absent in all males between the ages of three and sixteen. Blindness or impairment of vision occurred in forty-five cases, while the most important of the eye signs are paralysis of the upward movements, diplopia, abducens paralysis, nystagmus, ptosis and absence of pupillary light reflex.

Pontine Lesions.—The fibres of the pyramidal tract, which in the pons form scattered bundles, are gradually gathered together and cross to the opposite side at the lower part of the pons and

upper part of the medulla ; this is known as the decussation of the pyramidal fibres. A lesion, therefore, affecting these pyramidal fibres in the upper part of the right side of the pons will produce a paralysis on the left side of the body and *vice versâ*. Pontine lesions are suggested by a paralysis of the conjugate lateral movements of the eye, especially if associated with a paralysis of the fifth and seventh nerve together with a crossed hemiplegia.

Looking at the illustrations (Figs. 60 and 61), it is evident that a lesion which affects the nucleus of the third, sixth, seventh or eighth

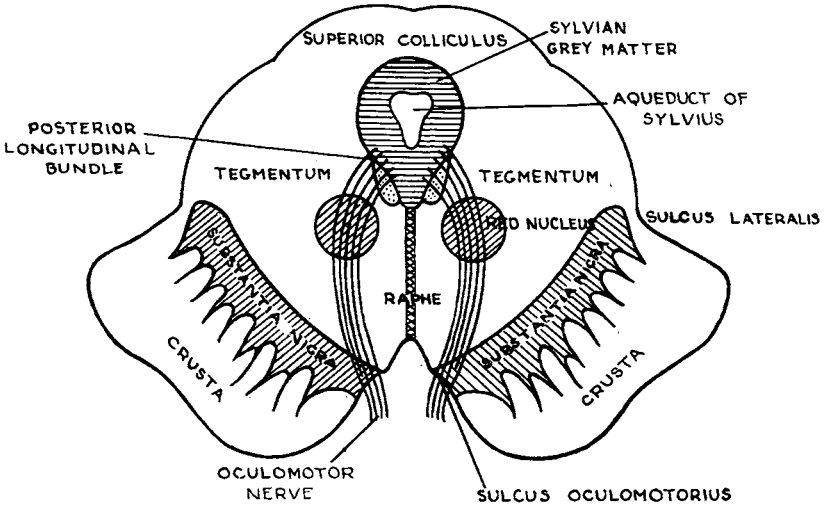


FIG. 61.—Diagram showing a section of the mid-brain taken at the level of the superior corpora quadrigemina. (After Cunningham.)

nerves will most probably also affect the pyramidal bundles in the neighbourhood, so that not only will a squint be produced, but a paralysis of the limbs or of the face and limbs as well.

A brief recapitulation of the various paralyses of the lateral conjugate movements due to lesions of the posterior longitudinal bundle has been given by Moncreiff.

There are several **syndromes** which, as they are related to ocular conditions, should be borne in mind :—

(1) The first is known as **Weber's Syndrome,** which is a paralysis of the third nerve together with a hemiplegia of the opposite side ; there will also be some paralysis of the face and tongue of the supranuclear type, as the decussation of fibres to the facial nuclei is

taking place at this level. Where would be the situation of such a lesion ? A glance at Fig. 60 shows that the lesion is situated at the junction of the pons and the crura of the mid-brain.

(2) **Parinaud's Syndrome.**—A lesion in the mid-brain, probably in the colliculi, it may cause a paralysis of conjugate movement upwards with paralysis of convergence.

(3) **Benedikt's Syndrome.**—This is a paralysis of the third nerve, together with tremor of the opposite limbs. It is a lesion of the red tegmental nucleus, which is situated at the upper part of the

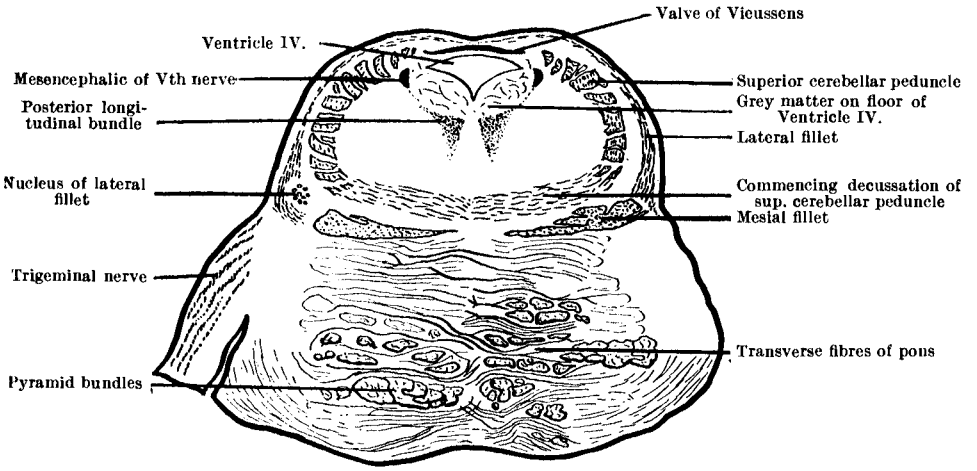


FIG. 62.—Transverse section through the upper part of the pons varolii of the orang. (After Cunningham.)

mid-brain, the part which receives the fibres from the superior cerebellar peduncle (see Figs. 61 and 62).

(4) **Millard Gubler's Syndrome.**—This is a lesion in the lower part of the pons, resulting in a paralysis of the face of the lower motor neurone type, internal strabismus due to involvement of the sixth or abducent nerve, together with a hemiplegia of the opposite side. Supposing the lesion is on the right side, the right eye will converge, as the internal rectus muscle will be unopposed by the paralysed external rectus muscle ; also the eye cannot turn to the right beyond the middle line.

(5) **Foville's Syndrome** (Grasset).—The sixth nerve nucleus is involved, but instead of a strabismus there is conjugate deviation to the healthy side. The seventh cranial nerve is involved, so that

there is paralysis of the same side of the face and there will be a hemiplegia of the limbs on the opposite side. The lesion is in the pons.

(6) **Horner's Syndrome.**—Paralysis of the ocular sympathetic may result from a lesion in the lateral part of the pons. Horner's name is usually associated with the ocular signs of paralysis of the cervical sympathetic. Such are miosis due to paralysis of the dilator pupillæ and unopposed actions of the sphincter pupillæ. The eye appears to be slightly shrunken—*enophthalmos*—and a slight ptosis due to paralysis of the tarsal muscle, but not of the levator palpebræ, is seen. In addition are found vasodilation and anhidrosis on the same side of neck and face. Horner's syndrome is present in thrombosis of the posterior inferior cerebellar artery.

It was in 1869 that Horner described the typical symptoms of a lesion of the cervical sympathetic trunk, which symptoms included hypotonia (lowered tension of the eyeball) (Fulton). The lesion is commonly described in connection with disease of the lower cervical segment of the spinal cord. Lesions of the lateral column of the cord above this level may produce Horner's syndrome. Even within the skull a lesion may exist which may produce paralysis of the oculo-sympathetic. Raeder has described such symptoms which were due to a tumour lying in close proximity to the Gasserian ganglion, which he termed " paratrigeminal paralysis of the oculo-pupillary sympathetic."

While in Munich the author was shown some cases of what there is also known as Horner's syndrome. These cases showed as their chief features abnormally long arms, a hollow retracted infrasternal region, while portions of the trigeminal distribution of the face on one or both sides showed definite anæsthesia. The pupils in some cases were unequal also. Such a condition is undoubtedly embryological in origin (shown by Passow).

(7) **Gradenigo's Syndrome.**—Paralysis of the external rectus due to extension of infection from the middle ear to the brain membranes in the neighbourhood of the tip of the petrous portion of the temporal bone, causing paralysis of the sixth nerve.

Medullary Lesions.—Lesions involving the lateral part of the medulla, as of the pons, may show Horner's syndrome, together with analgesia of the trigeminal nerve.

The lateral syndrome of the medulla, the result of thrombosis of the posterior inferior cerebellar artery, has been described by

Anderson and other writers. It consists of complete inability to swallow solids or fluids, pain or paræsthesiæ such as tingling over the distribution of the first and second divisions of the trigeminal nerve, Horner's syndrome—miosis, enophthalmos and ptosis—present on the affected side and in which the contraction of the pupil, however, is not so marked as in sympathetic paralysis, paralysis of the palate and defective sensation of taste on the same side, abolition of the nausea reflex and defective sensibility to pain and temperature on the face on the same side as the lesion, and of the trunk and limbs on the opposite side extending upwards as far as the umbilicus. In one of Dr. Yelland's cases there was, in addition to the above symptoms, complete corneal anæsthesia on the same side. The eye was covered for three months, the patient merely instilling a little boric ointment after each daily washing with weak boric lotion. For over four years the eye has remained perfectly sound, but the patient takes the utmost care to keep the conjunctival sac clean.

Lesions of the Frontal Lobe.—Tumours of the frontal lobes of the brain are notoriously difficult to diagnose. J. Purdon Martin, in the *British Medical Journal*, writes of several cases as follows : " Three cases of frontal lobe tumour are described, in two of which the growths involved both frontal lobes." Only one of his three cases showed ocular signs and symptoms, a partial optic atrophy on the temporal side of the discs with a large central scotoma. The only observed clinical signs which were common to all three cases were mental changes and tremor. In two cases severe generalised convulsive attacks constituted the first definite symptom. Attention is drawn to a peculiar exaggeration of the plantar reflexes encountered in two of the cases.

Purdon Martin also states that a unilateral anosmia is of more precise value than almost any other signs of frontal tumour, but it does not occur early in the case. Leslie Paton has drawn attention to the fact that a tumour situated in the frontal lobe may not produce a papillœdema. This agrees with the case quoted above and illustrated by Fig. 51.

In a study of 22 cases of tumour of the frontal lobe Kubitschek found symptoms of raised intracranial pressure—headache, vomiting and failure of vision. In 8 of his cases there were psychological disturbances and unilateral failure of vision in 5. There was bilateral disturbance of smell and unilateral exophthalmos. The

visual fields showed concentric contraction. Ataxic symptoms were not constant.

In many cases of gunshot injuries to the brain, remarkable recovery of vision occurs. This is due to recovery from concussion of the surrounding brain. It is somewhat analogous to a case the writer recently saw where a small leaden pellet fired from a gun passed through the upper eyelid and came to rest close to the sphenoid bone at the posterior part of the orbit. Although no nerves were cut, the movement of the upper eyelid was absent ; yet in a few days complete restoration of movement occurred.

Temporal Lobe Lesions.—In order to show the optic tracts in Fig. 60, the temporal lobes have been pulled apart. Tumours in this region may press on the optic tracts. They produce loss of visual field and sometimes one-half of the entire field is obliterated. This is known as complete hemianopia. Jackson described uncinate attacks or dreamy states in such cases ; sometimes there are visual hallucinations, together with generalised convulsions which form a complete symptom-complex of temporal lobe tumours. Cushing has shown that hemianopia in the upper quadrants or simply homonymous quadrantic contraction of the upper part of the visual field indicates a lesion of the anterior and lower portion of the opposite temporal lobe, while a lesion of the upper and posterior part of the lobe causes a similar condition in the lower visual field. The field defect of the eye on the side of the lesion is usually greater than that of the other eye. It is owing to the spreading out ventrally and laterally of the optic radiations in the temporal lobe while these fibres are passing from the external geniculate body to the calcarine area that such quadrantic defects are produced (see Fig. 41).

From the study of the anatomical relationships of the optic tract, it is seen that such diagnosis is applicable only in commencing tumours. The uncinate form of attacks described by Jackson are associated with temporal tumours bordering on the hippocampal uncus. They consist of olfactory or gustatory sensations often disagreeable and commonly associated with visual hallucinations, the latter being referred by the patient to the opposite side of the lesion. The type of the hallucination is of great localising significance. Visual hallucinations which are complex in character and form definite pictures are indicative of a lesion in the temporal region ; while simple visual hallucinations, flashes of light, often coloured and suggesting fireworks, are more often associated with

an occipital lesion. *Wernicke's aphasia* is present in lesions of the left temporal lobe; although articulating easily, the patient is unable to express himself and understands commands imperfectly and executes them badly. Cushing lays great emphasis on the use of the perimeter in the early recognition of temporal lobe tumours. Walker, who has made a great many perimeter drawings, found that quadrantic defects only became obvious when using the smaller test objects on the perimeter. The partial field defects short of hemianopia are characteristic of the involvement of the optic radiation in this region. Cushing quotes the following thirty-nine cases of intracranial tumours situated in or which had pressed upon the temporal sphenoidal lobe :—

No field defects	6
Homonymous hemianopia, *i.e.*, median vertical separation of blind and seeing halves . .	8
Partial hemianopia, *i.e.*, more or less quadrantic defects at some time during the period they were under observation	25

It is interesting to note that an inferior quadrantic hemianopia, as Gordon Holmes has pointed out, may be produced by damage involving the more dorsal fibres of the optic radiations (see Fig. 57).

McCarten has sent me notes of a case which had been admitted to a mental hospital. The case a week previously had been diagnosed by a clinic of a large teaching hospital as mental. He observed slight papillœdema in each eye, and this was the only neurological sign present of a gliosarcoma of the left temporal convolution. The tumour was removed one week after admission to hospital. Minute perimetric observations would have assisted in the diagnosis of this case.

Lesions of the Occipital Lobes.—Fits beginning with visual disturbances may be the early signs of tumours of the occipital region. There may be an hemianopia which is homonymous, but if the tumour presses upon a part of the optic radiations a quadrantic hemianopia may be found. When tumours of the parietal region extend backwards they may impinge upon the occipital region and an hemianopia may be found, while visual aphasia may result when the tumours are on the left side. De Schweinitz thought that a relative hemianopia called homonymous hemiachromatopsia is caused by a lesion of less intensity than one producing an absolute

hemianopia. Such a lesion has been found in the cortex and in the right internal capsule.

Epileptiform attacks, which may be preceded by a visual aura, occur in 50 per cent. of cases. If there is a pupillary reflex obtained by throwing the light on the blind half of the retina in cases of hemianopsia (Wernicke's pupillary inaction sign) the lesion is within the pupillary reflex arc, not higher than the external geniculate body, the pulvinar or the corpora quadrigemina. Therefore a lesion involving the optic radiations does not interrupt the light reflex. If nystagmus is present it is due to pressure on the cerebellum.

When writing on optic nystagmus and its value in the localisation of cerebral lesions Fox and Holmes say : " Our observations tend to support Stenvers' hypothesis that reflex centres for optic nystagmus lie in the occipital lobes and in the second frontal convolution and that these are connected by a reflex path which runs through the white matter of the hemisphere."

Allen, summarising his paper on occipital lobe tumours, says that, of 40 cases, the initial symptoms were epileptiform attacks in 30 per cent. ; visual hallucinations in 12·5 per cent. ; general mental impairment in 17 per cent. ; headache in 35 per cent. ; transient or progressive failure of vision in 15 per cent. ; and strabismus in 2·5 per cent. In only 12·5 per cent. did the initial symptoms suggest that the visual paths were involved. He found on examination of the patients, mental changes, 60 per cent. ; contralateral homonymous defects of the visual field, 94 per cent. ; papillœdema or optic atrophy, 70 per cent. ; inequality of the pupils, 35 per cent. (contralateral pupil larger in 25 per cent.) ; ocular paresis, usually of the external recti, 30 per cent. ; nystagmus and nystagmoid jerkings, 35 per cent. ; disturbances of speech function, 30 per cent. (50 per cent. of left-sided tumours) ; disturbances of sensation of the supra-thalamic type, 55 per cent. ; and minor degrees of motor disturbance, usually in the contralateral limbs, 90 per cent. The majority of the visual fields show changes in the contralateral halves only (see Figs. 63 and 64), but in some individual cases there were quadrantic and peripheral crescentic defects.

It may be difficult to differentiate tumours occurring in the temporal and occipital lobes, especially when the tumour is on the right side of the brain. Hemianopic fields of vision are exceedingly common with tumours in either situation. Clinical observations,

according to Horrax and Putnam, afford the only criterion available. They found that intracranial tumours confined to the occipital lobe are relatively rare, occurring in only forty instances in a series of

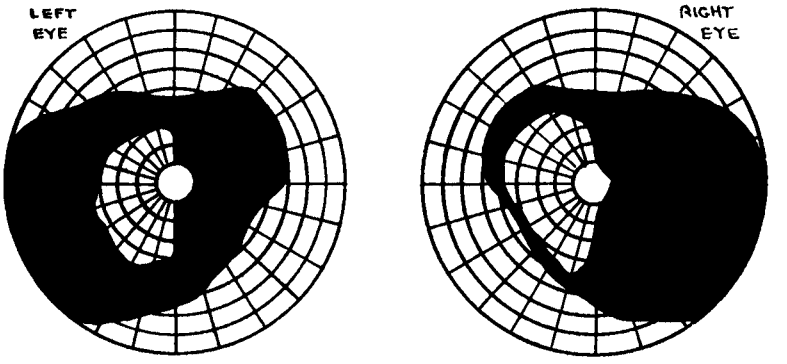


FIG. 63.—Contralateral homonymous hemianopia with sparing of the field for central vision, due to a tumour invading the mesial aspect of the left occipital lobe. (Allen.)

1,181 cases of verified intracranial growths. Of these forty instances, 73·6 per cent. show fields of vision exhibiting contralateral homonymous hemianopsia. An upper quadrant homonymous defect of the

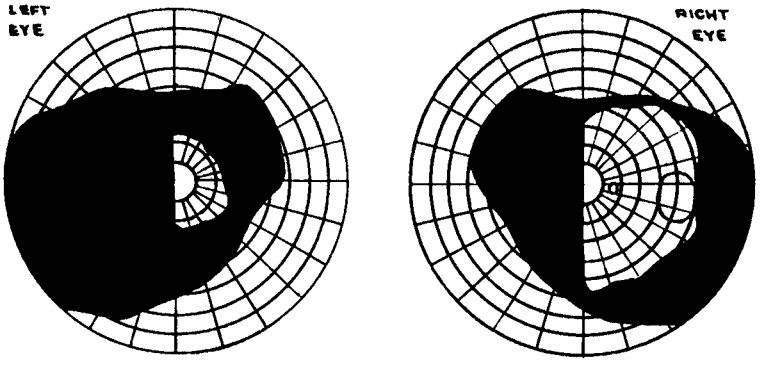


FIG. 64.—Contralateral homonymous hemianopia up to the fixation point due to an endothelioma weighing 135 grammes invading the right occipital lobe. (Allen.)

field of vision was not found in any case of the series. Visual hallucinations were present in 15 per cent. before operation for removal of the tumour. They found that complex "formed" images are not provoked by occipital growths, and that the high

incidence of spared macular fibres together with the absence of upper quadrantal defects in occipital growths is helpful, since bi-section of the macula and upper quadrantal lesions is frequent in temporal lesions, although the differential diagnosis between temporal and occipital tumours may be impossible without ventriculography.

Tumours in the Region of the Third Ventricle.—In 1911 Weisenburg published the results of his study of twenty-seven cases of tumour of the third ventricle. His three groups referred to are (1) those situated on the floor of the third ventricle ; (2) obstruction of the foramen of Munro ; and (3) extension into the aqueduct of Sylvius. Tumours of the third ventricle, Dandy states, are easily, safely and unequivocally diagnosed and localised by ventriculography.

It is remarkable that lesions or tumours arising from the base of the brain are especially associated with vegetative functions. Tumours extending into the third ventricle from the roof are usually lacking in symptoms.

A colloid cyst of the third ventricle together with the operation for its removal has been described by Paterson and Leslie. In this case a typical severe attack was characterised by a gradually increasing headache becoming so severe that at length unconsciousness supervened. During the early mild seizures there was a very noticeable deterioration of vision, between attacks the vision gradually became less. Papillœdema was present (4D) in both eyes, but the visual fields were normal. Dandy and others have mentioned the fact of the headache becoming worse with certain changes of position of the head, this being due apparently to the intermittent blocking of the foramen of Munro.

In association with tumours of the third ventricle Penfield has described what he calls *diencephalic autonomic epilepsy*, a term used to indicate the attacks produced by disturbance of the function of the autonomic nervous system. The case described by Penfield—tumour of the third ventricle with internal hydrocephalus—showed after a period of restlessness flushing of the face, rise of blood pressure, contraction or dilatation of the pupils, protrusion of eyeballs, lacrimation and profuse sweating, increase in pulse rate, slowing of the respiratory rate, and, towards the end of an attack, loss of consciousness.

From a study of cases of tumours in the region of the third ventricle Fulton and Bailey have enumerated symptoms as follows :

(1) Hypersomnia or uncontrollable drowsiness. "The patient would fall asleep while waiting to be photographed." Such cases resembled narcolepsy, a term employed to denote a form of periodic somnolence. (2) Diabetes insipidus. Cases have been recorded where due to injury of the tuber cinerium the patient has passed excessive quantities of urine daily, accompanied by marked thirst. (Pascheff's case of fracture of the sella turcica and left petrous bone due to bomb explosion showed a bitemporal hemianopia, loss of reflex of right cornea together with polyuria, symptoms which remained unchanged for three years.) (3) Adiposity, genital dystrophy. (4) Pyramidal and extra-pyramidal symptoms due to pressure on the neighbouring internal capsule. (5) Cerebellar ataxia. Pressure may reach the red nuclei or the cerebro-cerebellar association tracts. (6) Thermic disturbance. (7) Sensory symptoms (thalamic syndrome). (8) Ophthalmic symptoms. Optic atrophy is common and is slowly progressive. The field of vision does not show the quadrantic loss so commonly as that due to hypophyseal adenomas. Paralysis of conjugate eye movements has been noted by Weisenburg. (9) Mental and emotional symptoms. "This fatuous serenity of mind with complete failure to appreciate the gravity of his own physical condition is a mental concomitant not observed with other tumours except occasionally with those of the frontal lobes and here usually associated with marked intellectual deficiency." (Fulton and Bailey.)

Lesions of the Optic Thalamus.—Tumours of the optic thalamus, like those in many other parts of the brain, are difficult to locate. If the pressure is mainly on the internal capsule, the symptoms consist of hemiplegia, hemianæsthesia and hemianopia; in this condition also loss of control over the emotional movements may be found. Spontaneous pains, often extreme in their severity, are referred to anæsthetic or absent limbs. Choreiform movements have been noted in lesions of the optic thalamus, and hemianopia may be caused by invasion of the optic radiations.

Lesions of the Cerebellum.—Tumours pressing on the cerebellum give rise to nystagmus, ataxia, or falling over in one or other direction, while the papillœdema produced by tumours in this region is generally severe. A papillœdema is more frequently met with when the cerebellum is involved than any other part of the brain. Frequently one measures 6D with the ophthalmoscope, indicating the intensity of the papillœdema, which has protruded

the surface of the optic disc 2 millimetres forward into the vitreous. In addition to this severe condition of choked disc Marcus Gunn has emphasised the fact that the macular area may in some cases show markings resembling albuminuric retinitis in tumours of the cerebellum (the so-called macular fan, see p. 92). Nystagmus is very frequently present, except in mid-line tumours, and may be from side to side or of a rotary character. The side to side movements vary according to whether the patient is looking to the right or to the left, being of brief excursion to one side, while longer and slower to the other; the latter generally indicate the side of the tumour. *Skew deviation* is described, where one eye is turned downwards and inwards and the other upwards and outwards (see Fig. 65). If there is slight pressure on the sixth nerve in these cerebellar tumours, then weakness of the external rectus will result. This indirect effect tends to deviate the eyes from the affected side.

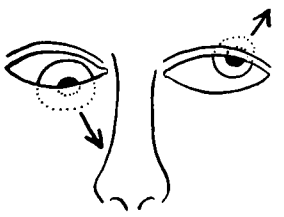


FIG. 65.—Skew deviation of the eyes seen in certain cerebellar tumours. (Hertwig-Magendie Phenomenon.)

Harvey Cushing, in a recent review of seventy-six cases of cerebellar astrocytomas—gliomatous tumours having a neoplastic basis composed of highly differentiated neuroglial cells—says that the symptoms usually date from childhood, with periodical morning headache and vomiting. Slowly vision becomes impaired, due to insidious onset of papillœdema following secondary hydrocephalus, which invariably results from their mid-cerebellar situation. The headache is more often frontal than occipital. Nystagmus is frequently absent, and the hydrocephalus is out of all proportion to the determinable cerebellar signs. The lower extremities are mainly affected by the ataxia.

The investigation of the visual fields is extremely important in every case of intracranial tumour. Cairns quotes a case of a student aged nineteen years who showed both signs and symptoms sufficient to suggest tumour of the cerebellum. But examination of the visual fields disclosed a left homonymous hemianopsia which threw an entirely different light on the case. At operation a highly vascular tumour was found on the internal aspect of the temporal horn of the ventricle. But for the perimetrical findings, this case might have been diagnosed as a cerebellar tumour, with disastrous consequences.

There is indeed much to be said for an early investigation of ophthalmic conditions in an intracranial neoplasm. Patients have been brought to hospital already blind, or with vision limited to perception of light. The field of vision being impossible to measure, a mistaken diagnosis is unavoidable. Cairns mentions such a case where the cerebellum was exposed, but necropsy showed a huge glioma of the left parietal and occipital lobes. If this patient could have been seen earlier a right homonymous hemianopsia would have indicated the site of the lesion. A papillœdema which has been allowed to remain in existence say for three months may pass into consecutive optic atrophy with total, or almost total, loss of vision, and the taking of fields in such cases is impossible.

Bailey and Cushing state that an abducens palsy, often bilateral, is of common occurrence in many cerebellar tumours, possibly more common, it would appear, in mid-line tumours than in any others. They say this is the one outstanding symptom in addition to the general pressure symptoms brought about by hydrocephalus and the instability and cerebellar ataxia which the patients show. Such a condition as strangulation of the abducentes may be merely mechanical (Critchley).

Cerebello-pontine Tumours.—At a recent autopsy the writer found a cerebello-pontine tumour the size of a marble pushing its way along the eighth cranial or auditory nerve and gradually invading the internal auditory meatus. Its progress had practically severed the sixth, seventh and eighth nerves, and had pushed the cerebellum aside. The symptoms of such a condition are as follows ; paralysis of the sixth nerve supplying the external rectus allows the eye on that side to squint inward. The seventh nerve is paralysed, therefore that side of the face will be immobile, while, on account of the destruction of the eighth cranial nerve, there is deafness on the same side. There will be pressure on the cerebellum, producing nystagmus, with coarse inco-ordination of movement.

Bilateral acoustic tumours are usually associated with von Recklinghausen's disease, but Gardner and Frazier have described thirty-eight cases of bilateral deafness in one family extending over five generations and in which the accompanying symptoms such as blindness, headache and vomiting in many of these cases proved the prevailing lesion to be bilateral acoustic nerve tumours. No other symptoms were present which would suggest these were cases of von Recklinghausen's disease.

Unilateral acoustic tumours, according to Penfield, are not true neurofibromas, but those associated with von Recklinghausen's disease are, for in the tumours associated with von Recklinghausen's disease nerve fibres will be found penetrating them.

Tumours of the Fourth Ventricle.—According to Lereboullet and Cushing the most common and the most malignant types of tumour of the fourth ventricle are the medullo-blastoma and the astrocytoma, both highly cellular and most liable to metastasis and especially after incomplete removal. There are vascular tumours capable of treatment by electro-coagulation methods, also epithelial tumours of the choroid plexus.

Headache is an early and frequent symptom. Vomiting alone or accompanied by headache and vertigo may be present. Vision is not early affected, but papillœdema is constantly found and is slowly progressive. Primary optic atrophy does not occur, but a consecutive optic atrophy may follow the papillœdema. Pressure on the chiasm by a distended infundibulum may lead to hemianopic fields for colour. (Lereboullet.) Vision varies from a transient amblyopia to complete blindness ; the latter may supervene quickly in a child, eight days in one case is recorded. The pupils dilate in proportion to the lowering of vision, nystagmus is frequent, and is usually in a horizontal plane, but where the roof of the fourth ventricle is involved the nystagmus may be vertical.

Cerebral Abscess.—Many surgeons, including the late Sir Victor Horsley, have believed that papillœdema by its early appearance or greater severity on one side is diagnostic of the side on which the tumour is, but this is extremely doubtful. This sign, however, is of much greater value in the case of intracranial abscess. There may be papillœdema on the side of the abscess only with partial paralysis of the third nerve. A ptosis with a dilated pupil on the same side is almost pathognomonic of an ipso lateral, cerebral or cerebellar abscess. The papillœdema persists longer after an operation for abscess than for tumour, and, indeed, may only then begin. Nystagmus is commoner in cerebellar abscess than in a cerebral abscess.

Meyers records a study of twenty-four cases of abscess of the brain which he has observed. In each case the diagnosis was confirmed either by operation or by post-mortem examination. Conjugate deviation of the eyes was a most valuable sign in the diagnosis. He found that in cerebral abscess this deviation was to the side of the lesion, while the head was turned to the same side.

Lesions of the angular gyrus produced more marked symptoms than those of the Rolandic area, while such symptoms were absent in lesions of the frontal region. In abscess of the cerebellum conjugate deviation is always present, regardless of the position of the head, and is directed to the side opposite to the lesion. The position of the head may show flexion to the side of the lesion. Nystagmus may be present in a slight degree, or even absent in abscess of the cerebellum ; if present, it is towards the side of the lesion if the labyrinth is functioning and to the opposite side if the labyrinth is destroyed.

It should be remembered that papillœdema does not usually appear in time to give the necessary indication for surgical inter-

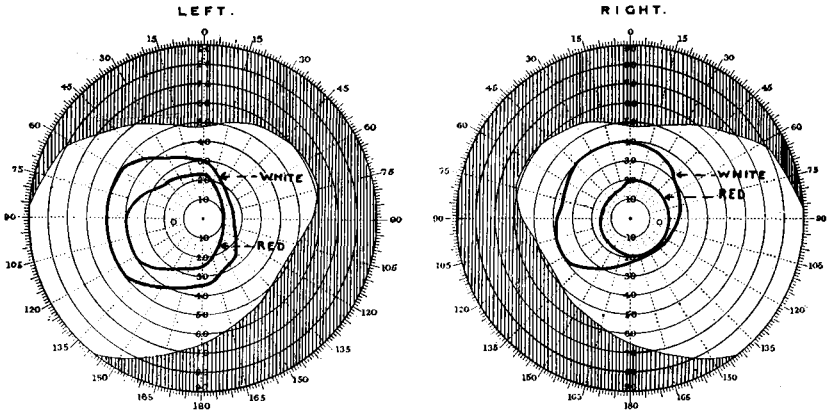


FIG. 66.—Extensive defect in both visual fields, but especially of the right, due to abscess of the left temporal lobe. (Coleman.)

vention, but it is always present in the later stages, particularly in large abscesses which have become quiescent.

In dealing with the method of treatment of abscess of the brain Coleman described a case where papillœdema was not present, the only one in the series, but considerable interference with the field of vision of the right eye was the result of an abscess of the left temporal lobe. (See Fig. 66.) The left pupil was dilated also ; he stresses the difficulty of obtaining reliable perimetric data.

Foster Moore points out that in a case of intracranial abscess occurring as a complication of otitis media, perimetric investigation may help to decide between a temporo-sphenoidal or a cerebellar location, the reason for this being that the ventral fibres of the optic

radiations as they leave the external geniculate body extend forward into the temporal lobe before turning backwards. Therefore pressure or damage to their fibres will produce changes in the field of vision.

Tuberculous Meningitis.—In tuberculous meningitis one-fourth of the cases shows a slight papillœdema, but the appearance of tubercle of the choroid is much more frequent. Parsons believes that tubercle of the choroid is not so uncommonly found in tuberculous meningitis as is usually believed, but it is often found only a day or two before death. Round yellow spots are seen near the optic disc, or less frequently scattered over the choroid. The pupils are usually contracted at first, but later are dilated and fixed. There is often partial ocular paresis, especially of the third cranial nerve, generally shown as a ptosis, but bilateral third cranial nerve paralysis is almost unknown—a point of distinction from syphilitic basal meningitis. Conjugate movements may also be affected. Prosper Veil, of Paris, says that in the course of syphilis or tuberculous meningitis and after cerebral hæmorrhage we sometimes get a hemianopia, but it is more particularly brain softening due to arterial thrombosis that in daily practice is the most frequent cause of homonymous hemianopia. This is also the writer's experience. It occurs in elderly high tension or renal subjects and in the young whose arteries have been damaged by syphilis.

Ripka rightly lays stress on the sudden appearance of a squint in an apparently healthy child as it may be the only evidence of the onset of tuberculous meningitis.

Pachymeningitis Interna Hæmorrhagica of Infancy.—The onset of symptoms of pachymeningitis interna hæmorrhagica occurs during the first year. The head may show enlargement with bulging of the anterior fontanelle. Sight may eventually be lost, due to an optic atrophy succeeding optic neuritis.

Acute Leptomeningitis.—Children suffering from this disease may show a severe form of retro-bulbar neuritis. There is rapid complete blindness followed by recovery. During the stage of blindness the child may be extremely irritable, which may be the result of the disease on the brain, or may be caused by retro-bulbar pain or by the blindness itself.

Photophobia is frequently present. The pupils may be unequal and show sluggish reactions, while the fundus often presents engorged vessels with papillœdema, or the fundus may be normal. The upper eyelids may droop, and, due to an extra-ocular paralysis, both squint

and diplopia may occur. The sixth or abducent nerve is commonly affected. As a sequel to meningitis primary optic atrophy may follow with complete permanent blindness. When temporary blindness is present the fundus may appear quite normal ; the blindness may last from a few days to six months. This condition has been called *acute cerebral amaurosis of infancy* (Gay). The optic atrophy found in young children suffering from congenital syphilis and which has followed a syphilitic meningitis has the appearance of a consecutive optic atrophy rather than a primary optic atrophy. The author has attempted treatment in several of these cases, but always without any return of vision.

Epidemic Cerebrospinal Meningitis.—In epidemic cerebrospinal meningitis there may be a descending neuritis causing a swelling of the head of the optic nerve with a little œdema. Other eye symptoms are sometimes found, *e.g.*, conjunctivitis, swelling of the lids, photophobia, pupils unequally dilated or contracted, keratitis or ulceration of the cornea, or even uveitis may be present. Paresis and paralysis of the ocular muscles are seen, especially of the external rectus, and diplopia is often present (see p. 485).

In *posterior basic meningitis*, which occurs in infants from a few months old to two and a half years of age, blindness may follow without any change being present in the fundus oculi. The eyelids are usually retracted, which gives a staring appearance. Strabismus often occurs. The blindness often passes away, but in some cases permanent blindness with optic atrophy forms a sequel to this disease (see p. 487).

According to Steegmann not only in severe infections and toxæmias, but also in meningitis do the ganglion cells of the external geniculate body undergo swelling, chromatolysis and pigment atrophy.

Encephalitis Lethargica.—During the past ten years the epidemic disease known as *encephalitis lethargica* has been brought very prominently before the mind of the medical profession. At one time it was stated that lethargica or epidemic encephalitis was characterised by the motor apparatus of the eye alone being affected, while vision was left intact. Subsequent investigation showed this view to be incorrect. Papillœdema and retro-bulbar neuritis have since been observed. Waardenburg has described one case of retro-bulbar neuritis which produced a definite scotoma, while another showed narrowing of the visual field. Agnello has described

the case of a girl aged thirteen years who, when the temperature commenced to fall and at the beginning of convalescence, complained that her eyes were quite blind. He found that the retinæ were normal, and suggested that the lesion in this case was in the sub-cortical white matter of the occipital lobes.

Although it has been definitely stated in the Report of the Local Government Board on Encephalitis Lethargica, London, 1918, No. 121, that papillitis has been observed in this disease, it has been pointed out to me by J. Purdon Martin that a lethargic patient with a papillœdema is a thousand times more likely to have a cerebral tumour (probably frontal) than to be suffering from encephalitis lethargica.

Affections of the ocular muscles are more common in the acute stage of this disease. The sequel shows them to be less common. Finally, however, weakness of the ciliary muscle of the eye, together with inequality of the pupils, are found. There may be a loss or a defect in the power of accommodation and of convergence. The writer has seen several cases showing as a sequel to this disease deficient power of convergence, although each patient could direct both eyes normally to right or left. This defect of convergence has remained permanently.

What has sometimes been described and termed " the reversed Argyll Robertson pupil " is simply the loss of the pupillary reaction to accommodation associated with defective convergence, while the light reflex remains intact.

Nuclear and supranuclear ophthalmoplegias are not at all common, but conjugate paralyses are frequent.

Early in 1918 Economo's disease appeared almost simultaneously at several centres in England. Professor A. J. Hall, of Sheffield, reported that in one week three patients presented themselves with bilateral facial paralysis of spontaneous origin, the advance heralds of encephalitis lethargica.

During investigations by the Ministry of Health, it was found that 75 per cent. of cases of encephalitis lethargica showed some ocular palsy, ptosis being the most common defect. In 25 per cent. it was unilateral. Foster Moore did not find any fundus change in seventeen cases which he examined ophthalmoscopically. The ocular paralysis passed away soon, but some lasted as long as nine weeks after the onset of the disease.

When examining chronic cases of encephalitis lethargica, students

should always bear in mind that the dilated pupils or inequality of pupils may be the result of hyoscine, belladonna or stramonium medication.

A persistent phase of this disease is shown by the presence of *oculogyral crises;* the eyes may be suddenly turned upwards and the patient is powerless to look down or close his eyes. He can often tell when these crises are about to come on. A description of these attacks was given by Professor A. J. Hall in the Schorstein Memorial Lecture on Chronic Epidemic Encephalitis. He stated

FIG. 67.—An oculogyral spasm. (Hall.)

that about 15 per cent. of his cases had these attacks—mostly young people of both sexes. After a definite warning, the eyes are suddenly turned up, although in a few cases a downward movement may occur at first. More rarely the movements may be entirely lateral, but the result is the same. The eyes become fixed and cannot be got back into the proper position. Strong suggestion may cause the attacks to pass off, but once the eyes become fixed sleep usually follows. The attacks may occur at regular or irregular intervals from two or three a day to perhaps only one a week. They may last only a few minutes or several hours, during which time the patient is helpless, and in most cases the attack ends in sleep and the patient wakes up quite free from it. In a certain

number of cases the attack ceases spontaneously. It is only in a few cases that the attacks do not end in sleep. (Figs. 67 and 68.)

Emotional excitement, suggestion or " having to do something they do not like " is sure to bring on an attack. It has been suggested that these attacks are hysterical or hysteriform, but against this there is considerable evidence. Although hysteria has been known throughout the ages, this type of attack was unknown until epidemic encephalitis appeared. Such attacks cannot be made by any voluntary effort. I have seen Professor Hall's cinematographic

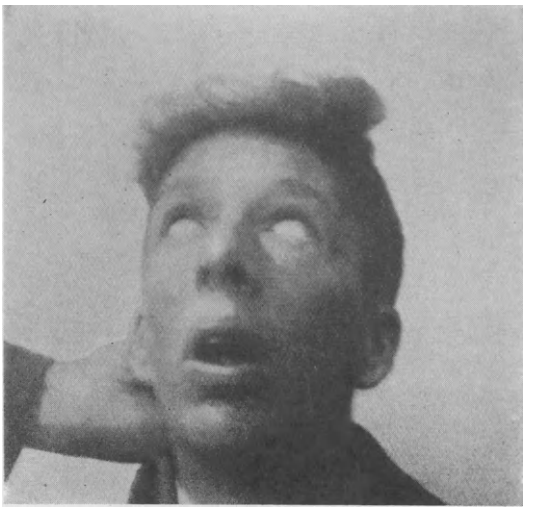


FIG. 68.—An oculogyral spasm. (Hall.)

reproduction of these cases during an attack. They are totally unlike anything seen in hysteria. His theory is that when the lids are closed, as in sleep, the tone of the ocular musculature is relaxed, the eyes are no longer adjusted for binocular vision, and they usually wander upwards behind the lids. The patient is unconscious of this and has no control over it.

Professor Hall further proved that the position of the eyes in sleep is not so constant as one supposes. He follows up his analogy by regarding these oculogyral attacks as being produced by the sleep of the nerve centres controlling only the muscles of the eyeball. In the oculogyral attack no other part is resting, not even the eyelids. The absence of strong action of the occipito-frontalis in

oculogyral attacks has been noted in all his cases. Other oculogyral attacks which have been described include those in which the lids close and cannot be opened. These are uncommon.

That oculogyral spasms indicate a condition which, Bogaert states, extends far beyond the ocular localisation, presenting a whole extrapyramidal, psychic and sympathetic symptomatology. In his study of three cases of Parkinsonian patients Bogaert gives details of them having in common speech difficulties in connection with ocular paroxysms. One case showed true palilalia beginning and ending with the ocular movements. He says that anxiety is the state most conducive to such crises and that the oculogyral effect is an inhibitory crisis revealing an innate antagonism between a general and a localised inhibition.

In describing the ocular disturbances in encephalitis Kennedy attributed the chronic contractions of the eyelids, the ocular spasms and the oculogyral crises to a flood of impulses pouring down from the basal ganglia producing disturbances in the oculomotor and associated centres, disturbances similar to other abnormal involuntary movements, also the hypotonia so characteristically found in post-encephalitic disease and attributed to the uncontrolled activity of the striatum, although Reilly says the essential causative factors have been postulated to exist in other regions also. Jelliffe gives a digest of the available literature until recent times.

In the Mott Memorial (1929) there appeared a chapter entitled "*The Ocular Syndrome of Epidemic Encephalitis*," by McGowan and Cook, both of whom had considerable experience in the epidemic of encephalitis lethargica. They published the following table :—

Nature of Sequel.	Ministry of Health Series.	Lancashire Series.
Indefinite palsies . . .	6·8	4·1
Squint	6·4	10·4
Impaired vision	6·4	18·7
Diplopia	3·2	6·2
Nystagmus	2·4	4·1
Ptosis	1·2	5·2

They mention that although encephalitic Parkinsonianism may exist with complete freedom from ocular lesions, yet when due to

chronic epidemic encephalititis it is rarely unaccompanied by signs of ocular defect. On the other hand, idiopathic paralysis agitans and most types of Parkinsonianism due to causes other than epidemic encephalitis are seldom accompanied by ocular symptoms. Commenting on the pupil reflexes and convergence, they state that the most common lesion found in chronic epidemic encephalitis is that affecting the accommodation reflex, some impairment being noted in 103 of their cases, and a total absence of this reflex in 44. The reaction to light was impaired in 44 cases and totally absent in 6. A milder degree of dilatation was more commonly observed, but one patient showed pin-point pupils. They found that double vision, as compared with other eye symptoms, is relatively much rarer in the chronic than in the acute stage. The question of operation has arisen in connection with divergent strabismus found later in some of these cases. From the author's experience there is not the slightest reason why such a divergent strabismus should not be corrected by means of an orthodox squint operation.

Two patients complained of seeing red at times ; this condition McGowan and Cook termed erythropia.

In discussing the oculogyral crises, these writers state that they vary considerably in duration, some lasting only a few seconds, while others may continue practically all day. Sleep abolishes them, but they may recur immediately on waking. In their opinion the cause of the crises is to be found in the lesion of the nervous mechanism subserving conjugate movements of the eyes, this lesion making tonic seizures possible owing to the impairment of cortical inhibition, which they consider a fundamental characteristic of the chronic encephalitic, as evidenced by his facile childish temperament, his apache tendencies, and many of his abnormal mental reactions. They conclude by referring to the frequency of ocular defects in the encephalitic and their consequent importance in differential diagnosis, and state that Parkinsonianism plus ocular symptoms means chronic epidemic encephalitis, whereas Parkinsonianism without ocular symptoms points to arterio-sclerosis or one of the less usual causes of Parkinsonianism.

The beneficial effects of prominal (Bayer) in the treatment of oculogyral crises have been described by Leak, who gave a 3-grain tablet each morning, later adding half a tablet at night.

Acute Serous Encephalitis.—This disease, known also as acute meningo-encephalo-myelitis of childhood, was described by Brown

and Symmers in 1925. It occurs in children under seven years of age, and is rapidly fatal. There is severe headache, vomiting and convulsions. There may be papillœdema present. When papillœdema occurs in post-vaccinal encephalitis it is generally transient. Occasionally pupillary abnormalities and ocular paralyses have been present in these conditions. Nystagmus may be present also.

Superior Encephalitis.—*Synonym :* Polioencephalitis acuta hæmorrhagica superior of Wernicke. What has been described as *Wernicke's periaqueductal hæmorrhagic syndrome* is a toxi-inflammatory process analogous to poliomyelitis, and, as a rule, is fatal in from ten to fourteen days. The disease is more or less limited to the region about the aqueduct of Sylvius. Wernicke ascribed chronic alcohol as the cause, although the condition is found in influenza, ptomaine poisoning (Holden), carbon monoxide gas poisoning and in the hæmorrhagic diathesis (Wechsler). As the region chiefly affected is in the neighbourhood of the nuclei of the third and fourth cranial nerves there follows an ophthalmoplegia of rapid onset, paralysis of conjugate movements, pupillary reflexes and ptosis. The intrinsic muscles may escape although both internal and external ophthalmoplegias are commonly seen. This condition must not be confounded with acute ophthalmoplegia, due to acute peripheral neuritis, sometimes seen in alcoholic poisoning.

Optic atrophy and nystagmus may be present. Extension of the disease may result in ataxia, loss of tendon reflexes, and delirium. Although recovery may take place, permanent paralysis of the ocular muscles, mental symptoms and ataxia may remain.

After death hæmorrhages are found in the floor of the grey matter of the Sylvian aqueduct.

Acute Hæmorrhagic Encephalitis of Strümpell.—Young children are chiefly affected by this disease. It may be primary or follow the infectious fevers. There are widespread hæmorrhagic foci throughout the cerebrum, mid-brain, pons and medulla. If the disease is rapidly fatal, as it usually is, the ocular involvement does not become manifest, but should the patient show signs of recovery or the disease become chronic, then various paralyses appear: there may be diplopia or squint due to ocular paralysis, nystagmus and hemianopia.

Diseases of the Basal Ganglia or Extrapyramidal Syndromes.—It is not the purpose of this book to attempt an explanation of neurological problems, so sheltering behind such a statement the author

will briefly refer to what are known as disorders of motility, extra-pyramidal syndromes or diseases of the basal ganglia. As an aid to the memory of the student, I have inserted Fig. 41, which is a drawing of a horizontal section through the right cerebral hemisphere at the level of the widest part of the lenticular nucleus. The optic radiations are somewhat diagrammatically drawn to show the wide sweep they take into the temporal lobe. The basal ganglia of the cerebral hemispheres are the lenticular and caudate nuclei, (which together form the corpus striatum) the claustrum and the amygdaloid nucleus. Some include the thalamus as well. The caudate nucleus is a highly arched mass of grey matter with a thick swollen head or anterior extremity. Posteriorly it presents the appearance of an attenuated tail. In the horizontal section, therefore (Fig. 41), part of the head is shown and a small portion of the tail. The caudate nucleus is seen therefore to form an arch over the thalamus, the extreme end of the tail turning downwards and forwards into the descending horn of the lateral ventricle on the roof of which it finally joins the amygdaloid nucleus. The lenticular nucleus lies completely embedded within the white medullary substance of the cerebral hemisphere on the outer side of the caudate nucleus and the optic thalamus. It is divided into external and internal medullary laminæ. The outer lamina is called the putamen. The two inner laminæ are termed the globus pallidus. The claustrum, a thin layer of grey matter, lies in the white substance between the putamen on the inner side and the island of Reil on the outer.

The caudate and lenticular nuclei together constitute the corpus striatum. Fibres pass from the cerebral cortex, from the thalamus and from the mid-brain to the corpus striatum. Connecting fibres pass between the caudate and the putamen and the globus pallidus, while a special bundle (ansa lenticularis) of efferent fibres passes from the globus pallidus to the thalamus, the red nucleus, the substantia nigra and the hypothalamic nucleus or corpus Luysii.

Morgan traces fibres from the corpus striatum to the oculomotor nucleus and the nucleus of Edinger-Westphal, and also fibres by a strio-bulbar fasiculus to the trochlear and abducens nuclei and to the nuclei of most of the cranial nerves. He found that bilateral lesions of the corpus striatum led to the death of the animal (cat), but unilateral lesions led to circus movements, difficulty in chewing and swallowing, disturbance of voice production, hypertonicity, tremor and constriction of the pupils.

The minute anatomy of this region must always be associated with the name of Kinnier Wilson. There are probably many other inter-connections than those mentioned above, so that it is exceedingly difficult for neurologists to place the blame for a particular syndrome on any one particular structure. It is acknowledged that since the epidemic of encephalitis with its disseminated lesions and protean symptomatology its sequels may simulate practically every basal ganglion syndrome (Wechsler). Brain and Strauss state that the anatomical arrangements suggest that the striatum possesses a subordinate reinforcing rather than an initiating function. Disease of the corpus striatum is characterised by symptoms of rigidity, tremor, loss of voluntary movement, hypokinesia, impaired associated movements and absence of true paralysis, together with absence of sensory disturbances. This is really the Parkinsonian syndrome, while chorea, athetosis and torsion spasm form the hyperkinetic syndromes usually associated with the basal ganglia. All these are grouped under the title of extrapyramidal syndromes.

The Various Forms of Parkinsonianism

Paralysis Agitans.—In many cases this disease is entirely devoid of ocular symptoms. However, occasionally tremor of the eyelids is observed, or the eyelids may be drooping, producing a sleepy expression. Wilson states that the eyeballs move slowly, while the winking is less frequent, but if the patient is asked to look quickly to one side, winking may occasionally be observed. The pupillary reflexes are not affected in this disease.

Parkinsonianism as a Sequel to Encephalitis Lethargica.—In a paper on the anatomo-pathological basis of the Parkinsonian syndrome, McAlpine described his histological findings in eight cases of this disease. He agrees with other writers that the only group of cells which is constantly affected in this disease is that of the substantia nigra. He found that the basal ganglia and in particular the globus pallidus showed no noteworthy changes.

The above-mentioned diseases when closely investigated exhibit pathological changes which extend far beyond the corpus striatum. Rademaker concludes that it is the pathological changes in the red nucleus which are responsible for alteration of muscle tone. Hence we may include lesions in such parts of the mid-brain as the

substantia nigra and red nucleus as being responsible in some degree for the so-called extrapyramidal syndromes. (Bahr.)

Parkinsonianism usually develops in an insidious manner during the twelve months following an acute attack of encephalitis. Young adults up to the age of forty are peculiarly liable.

Oculogyral Spasm.—Attacks consisting of spasm of the conjugate ocular muscles are commonly seen. The eyes are usually turned upwards, occasionally in a lateral direction, and rarely downwards. The eyes are usually described as " suddenly shooting up into the head." A most excellent description of the oculogyral crises is given by McGowan and Cooke, who stated that they found this phenomenon confined to Parkinsonians. They state that these crises are sometimes only part of a more generalised tonic seizure, the muscles of the neck often being involved. In other cases the associated phenomena were feelings of discomfort in varying degrees. Slight headache or aching of the eyeballs or a feeling that the eyes will " stop up for ever " are complained of. Some cases are accompanied by screaming, stamping of the feet, or even outbursts of obscenity. They state that these outbursts are usually well controlled by hyoscine. Sleep abolishes the attacks, but they may recur immediately on wakening. The time of day, fatigue and emotion are factors in precipitating attacks. Some of the patients would lie down with their heads beneath the blankets which successfully assisted in closing of the eyes. It was found that psychical factors, both voluntary and suggestive, were capable of preventing the onset of attacks. Some patients who had an attack each evening, when told that this condition would prevent their attending dances, found the attacks did not come on.

The movement of convergence is nearly always impaired, although on looking to right or left the synergic movements of the internal recti are normal. In advanced cases of Parkinsonianism conjugate movements, both horizontal and vertical, may show some paresis and may also exhibit jerky movements associated with blinking. (Wilson.)

Arterio-sclerotic Parkinsonianism.—Cerebral arterio-sclerosis is responsible for a further form of Parkinsonianism. These cases are usually found in late middle and old age. Such cases may show arterio-sclerotic retinitis. Parkinsonianism plus ocular symptoms means chronic epidemic encephalitis, whereas Parkinsonianism without ocular symptoms points to arterio-sclerosis or one of the

other less usual causes of Parkinsonianism. Parkinsonianism very rarely becomes a manifestation of cerebral syphilis, although it is definitely established that the extra-pyramidal syndrome is frequently associated with general paralysis (Pardee). The treatment, however, seems to be hopeless as restoration of the function of the corpus striatum is beyond the curative power of any specific treatment.

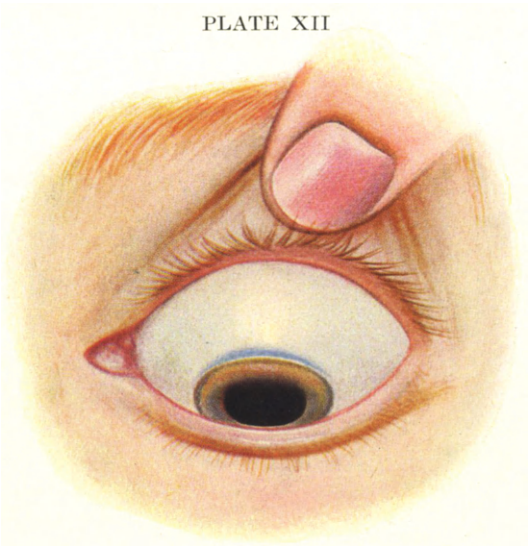
In dealing with patients exhibiting the Parkinsonian syndrome it should be remembered that loss of accommodation which necessitates stronger convex glasses for reading than is usual for the age of such a patient may be due to hyoscine medication.

There is a condition—encephalo-myelitis—which may be associated with vaccination or measles, and is usually designated acute disseminated encephalo-myelitis which is characterised pathologically by small patches of demyelination round the blood vessels. (Marsden and Hurst have called it perivascular myelinoclasis). A condition clinically identical occurs without any association with an exanthem. Involvement of the optic nerve in these cases is unusual, but, by personal communication, J. Purdon Martin has stated that in two patients under his own care such has been observed. The first in the acute stage showed a condition of retrobulbar neuritis and papillitis. The second was observed two to three months after the acute attack when partial primary optic atrophy was present. The field of vision showed a large central scotoma with serious impairment of vision which became permanent.

CHAPTER X

ABNORMALITIES : CONGENITAL AND DEGENERATIVE. SUBARACHNOID HÆMORRHAGE

Hepato-lenticular Degeneration or Pseudo-sclerosis of Westphal.—The diseases known by the names hepato-lenticular degeneration and pseudo-sclerosis of Westphal are closely allied to each other. Although at one time thought to be different diseases, they, together with progressive lenticular degeneration described by Wilson, are now usually considered to be identical. They all may be included under the title hepato-lenticular degeneration. It is a chronic progressive and fatal disease of early adolescence. It is frequently familial, but neither congenital nor hereditary. Degeneration of the corpus striatum, together with cirrhosis of the liver, are the chief pathological features of this disease. As in other extrapyramidal syndromes, tremor, rigidity, impairment of voluntary movement, loss of emotional expression, together with slight mental deterioration, are present. In 1902 Kayser described an unusual form of pigmentation of the cornea in a man suffering from multiple sclerosis. He described the cornea as showing at its periphery a zone of clouding which made the cornea partly opaque and of a dark brownish-green colour. With a corneal microscope the opacity was seen to be caused by a collection of fine golden particles lying in the deeper layers of the cornea. He did not, however, refer to the condition of the liver. It was left to Fleischer to draw attention to the combination of corneal pigmentation and symptoms of cirrhosis of the liver as pathognomonic of this disease. This peculiar pigmentation of the cornea is mentioned by Wilson, and the name of Kayser-Fleischer ring has come to be associated with Kinnier Wilson's disease. Williamson Noble has had a colour drawing made of this condition, and I am indebted to him for permission to reproduce the colour plate, No. XII. It is the consensus of most observers who have tried to see the Kayser-Fleischer ring that without the slit lamp and corneal microscope it is practically impossible to see the condition with the naked eye, yet in one case at least Kinnier Wilson stated that he noticed the golden glow of the peripheral portion of the



THE KAYSER-FLEISCHER RING PRESENT IN WILSON'S DISEASE

FIG. 1. Naked-eye Appearance

Observe the ring of golden-brown pigment near the limbus of the cornea, most dense in the upper and lower parts and gradually fading off between.

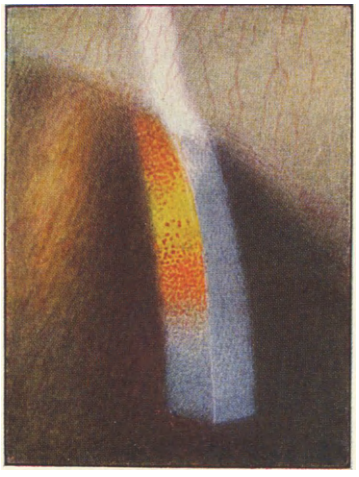


FIG. 2. Slit-lamp Appearance

Mr. Williamson-Noble reported that the slit-lamp examination revealed " the brown pigment forms a definite ring about 5 mm. wide, starting 1 mm. from the corneal margin. Between the outer edge of the ring of pigment and the margin of the cornea there is a ring of greenish, opalescent dots very closely set together. All the pigmentary deposit is in the posterior layer of the cornea ; with retro-illumination the cornea looks almost clear." J. G. Greenfield examined in the laboratory of the National Hospital, Queen Square, two cases showing the Kayser-Fleischer zone in the cornea. He said that pigment was found to be confined to the outer part of Descemet's membrane ; it did not reach the edge of the cornea and only came inwards to a distance of 2 mm. It consisted of very fine granules less than 1 microm in size. They did not correspond exactly to any of the known pigments derived from blood but were closely allied, particularly in the matter of solubility (*e.g.*, insolubility in weak acid and solubility in alkalis) to the malarial pigment which was derived from blood. Krause quoting Gerlach said that even after the injection of silver salts no silver was found present in a cornea showing a Kayser-Fleischer ring. Rohrschneider believes the pigment is a derivative of hæmoglobin (Case under the care of Dr. F. M. R. Walshe. Drawings by Hamblin of naked eye and slit-lamp appearance kindly lent by F. Williamson-Noble).

[*To face p.* 186.

iris. Harrison Butler says that the golden glow was seen in the lower part of the iris in the case which he examined in June, 1925. Examination by means of the corneal microscope revealed in the corneal prism that the limbus had gold-brown granular deposits which became less dense as the prism was moved towards the centre. Hepato-lenticular degeneration is one of the diseases of the nervous system in which the aid of the ophthalmologist is of the utmost importance in diagnosis, just as in many other conditions the neurologist would be wisely advised to seek the early advice of the ophthalmologist, especially where this disease is suspected in one member of a family. All the siblings should be examined also for corneal pigmentation,

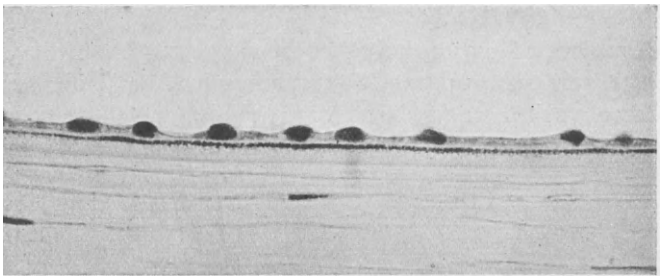


FIG. 69.—Section of cornea from a case of hepato-lenticular degeneration (Wilson's Disease). The pigment is confined to the outer part of Descemet's membrane ; it did not reach the edge of the cornea. The pigment consisted of fine granules each less than 1 microm in size.

cirrhosis of the liver and symptoms of nervous disease. The microscopical section of the cornea shows that the granules are actually nodules in Descemet's membrane which gradually become smaller and more sparsely scattered towards the centre of the cornea. These granules do not show any iron reaction. With hydrogen peroxide complete bleaching occurred in three days while the melanin pigment of the iris was only partly decolourised in eight days. The granules were not coloured by Scharlach R. They were insoluble in alcohol, ether and xylol (Barnes and Hurst) (Fig. 69).

Torsion Spasm.—Torsion spasm, or dystonia musculorum deformans, is one of the hypokinetic divisions of the extrapyramidal syndromes, and although closely allied to hepato-lenticular degeneration neither corneal pigmentation nor other ocular signs and symptoms have been described.

Chorea.—In Sydenham's chorea the eyes may participate in the irregular jerky motions, and these may produce transient diplopia,

or the eyes may be rolled from side to side, the head turning in the same direction. The pupils may be dilated or unequal and even hippus has been noticed. Fundus changes are not constant. It is remarkable that many children affected by chorea suffer from errors of refraction ; therefore, it is important to remember that each child suffering from this disease should have a careful estimation of its refraction done.

On the treatment of chorea by nirvanol Wetschler mentions that in addition to various symptoms produced by this drug other symptoms appear such as headache, dizziness and conjunctivitis, all of which disappear with the cessation of the initial reaction.

Oxycephaly

The subject of oxycephaly has been most fully dealt with in a paper by Greig, who quotes 136 references to the same. He recognises three varieties : (1) True oxycephaly, which is recognisable at birth and is due to general craniofacial synostosis usually associated with syndactyly ; (2) Delayed oxycephaly, which is a general synostosis of the cranium manifesting itself in childhood and not associated with other deformities of the extremities ; (3) False oxycephaly, a localised cranial synostosis occurring during cerebral growth. It is not congenital and is often of definite origin.

In the great majority of cases oxycephaly is not hereditary but occurs as an isolated affection in one family although many instances have been recorded where the deformity occurred in more than one member of the same family as stated by Harman, Chatelin and Crouzon.

Although the first variety is what has been described as the oxycephalic syndrome, yet the outstanding feature is the pointed or tower skull. As in Mongolism or microcephaly the cephalic condition is the dominant one, but the primary defect is cerebral not cranial.

The oxycephalic skull, including the face bones, is absolutely sutureless, every suture has undergone obliteration. The orbital index averages 108. The size of these apertures is exaggerated by the lack of development of the maxillary bones. The striking feature of these orbits is their shallowness (see Fig. 70) and the great diminution of their containing capacity brought about by the orbital surface of the great wing of the sphenoid being displaced mesially and forward. Examination of the middle fossa of the skull reveals the extreme narrowness of the pituitary fossa. The mesial

extremities of the petrous bones approach to within 10 mm. of each other while the dorsum sellæ is represented by an insignificant ridge of bone fused within these two points. The floor of the fossa measures 3 mm. transversally and consists only of two lateral spicules of bone on either side of two small foramina which are not

FIG. 70.—The orbital fossa of an oxycephalic skull. Note the shallowness and great diminution of the containing capacity of the orbits due to the mesially displaced great wing of the sphenoid. The poorly developed maxillæ contain no trace of sinuses. (Greig.)

in connection with the carotid grooves. Sometimes the floor is narrowed to a knife edge. The sulcus chiasmatus is very small. The cause of the premature synostosis in oxycephaly is not known ; rickets and syphilis are not responsible.

The radiogram of the skull gives a false impression of thickness. The consolidation is due to want of diploë but is not thick. At no part of the cranium are " digital impressions " visible (Greig).

Ocular Symptoms.—The eyelids are heavy and exophthalmos is commonly present, probably 50 per cent. according to Beaumont, also actual dislocation of the eyeball has been recorded several times (Stephenson). Associated with oxycephaly is strabismus or squint which is invariably divergent, the ocular movements are commonly restricted and nystagmus may be present (Fig. 71).

Tortuous veins are seen on the retina (Coates and Patry). Optic atrophy is present in a great number of cases and is not solely due to

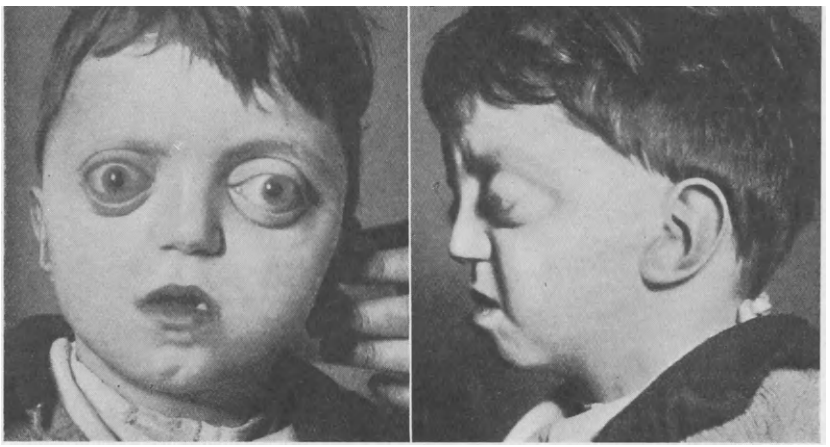


FIG. 71.—A case of oxycephaly observed from the age of seven months. The radiogram of the skull shows "thumb markings" due to increased intracranial pressure. For some months past the right eye occasionally became dislocated and each time the boy was able to push the eyeball back again. (E. A. Cockayne, in charge of this case at Great Ormond Street Hospital.)

intracranial pressure or narrowed foramina. According to Greig there is neither osteophytic growth nor thickening to narrow the optic canal. He says the most probable explanation of the optic atrophy is the upward deflection of the nerve with the displacement of the brain, the defective arterial supply and the restriction of the venous return. On the other hand papillœdema, due probably to the pressure exerted by the growing brain, has been observed very early in life and is followed by consecutive optic atrophy before the age of seven years (Uhthoff).

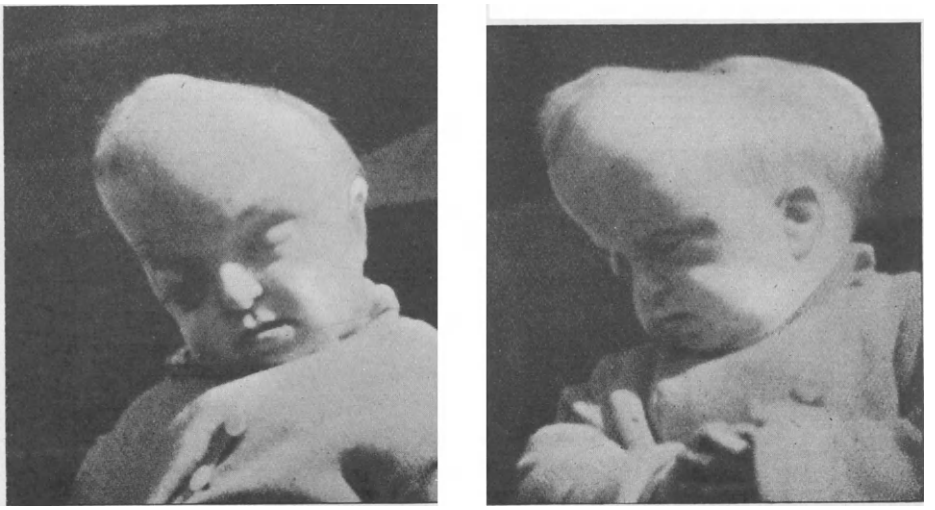
Writing on the pathology of optic atrophy in oxycephaly Nordmann believes that the atrophy is consequent on papillary stasis (papillœdema) following on cranial hypertension, evidence of

which is commonly present, especially in the young. There is great diminution of vision in nearly all the cases.

Regarding the mental condition of true oxycephaly I would again quote Greig : " In true oxycephaly average mental capacity may be claimed, genius never, but the great majority are below the average."

The condition is one for which a decompression operation with ventricular puncture according to Osler and McCrae, is indicated, but they do not state at what age. According to Greig when the brain has obtained its maximum bulk amelioration is not to be expected.

Ida Mann states that she believes oxycephaly to be a true developmental anomaly of genic origin.



(*a*) (*b*)

FIG. 72.—A case of Crouzon's dysostosis cranio-facialis. Note the drooping position of the head. (*a*) Viewed from the anterior aspect the frontal region of the skull rises high with some degree of asymmetry, the summit of the eminence being displaced to the right. The right eye in addition to proptosis has a divergent position. (*b*) The lateral view shows the characteristic formation of the head which distinguishes it clearly from that of oxycephaly. There is a notable depression between the frontal and occipital masses. The anterior fontanelle which is still unossified and pulsates is situated at the apex of the frontal prominence. The nose suggests somewhat the description " parrot beak." The patient is a boy aged two years, whose mental capacity is that of an imbecile ; he cannot walk. There are no deformities of the limbs, fingers or toes. The right eye can be readily dislocated in front of the eyelids. When four months old the left eye was found dislocated in front of the eyelids and could not be replaced. It was then removed. (Under the care of A. J. Ballantyne.)

Dysostosis Cranio-Facialis

In 1912 Crouzon described two cases, a woman aged twenty-nine years, and her child of three years, both of whom showed a condition of the head allied to oxycephaly, but in addition to the exophthalmos striking facial characteristics were present. Each case presented a marked boss in the line of the frontal suture, the forehead receding to a high pointed vertex (Fig. 72). The nose was hooked, resembling a parrot's beak. There was marked prognathism which carried the teeth of the lower jaw 3 or 4 cm. in front of those of the upper jaw. Crouzon says that whereas oxycephaly is characterised by cranial deformities and associated with exophthalmos, yet facial deformities do not otherwise occur. Also, that his cases differ from the acrocephalo-syndactily of Apert in that syndactily is not present, and further, that dysostosis cranio-facialis differs from all other types of skull deformity in that it is both hereditary and familial in character.

In this deformity there is marked exophthalmos and a divergent squint, also optic atrophy has been described in similar cases.

Corneal Opacities associated with Bony Changes and Mental Deficiency

A syndrome characterised by corneal opacities associated with bony changes, mental deficiency and hepato-splenomegaly has been described by Helmholz and Harrington. The most striking feature in their cases is the uniformity of cloudiness of the cornea. They summarise the characteristics of the syndrome as follows : cloudiness of the cornea, restricted motion of the joints of the extremities, short thick club-like hands and feet, with limited extension, lumbar kyphosis, scaphocephalic head and mental retardation. Hurler described somewhat similar cases, while Cockayne showed a case, a female child, aged four and a half years, whose corneæ were examined by Penman. A slit-lamp examination revealed the opacities as ill-defined spots scattered throughout the substantia propria of the cornea most thickly in the anterior portion. The corneal epithelium was normal. The condition is a corneal dystrophy. The cases of Helmholz are not syphilitic but the one shown by Cockayne undoubtedly is.

Ocular Hypertelorism

This rare deformity of the skull has been described by Greig in 1924. The original of this study was a girl named Mary Macdougal. The thorough anatomical description, by Greig, of her deformed

skull is only equalled by his kindly appreciation of her gentle and affectionate nature when alive.

A face that might suggest oxycephaly and which has been described as such in medical literature, is the one now under consideration, but neither in oxycephaly nor rhinoplasty is " far-apartness " of the eyes a necessary attribute, while in hyper-

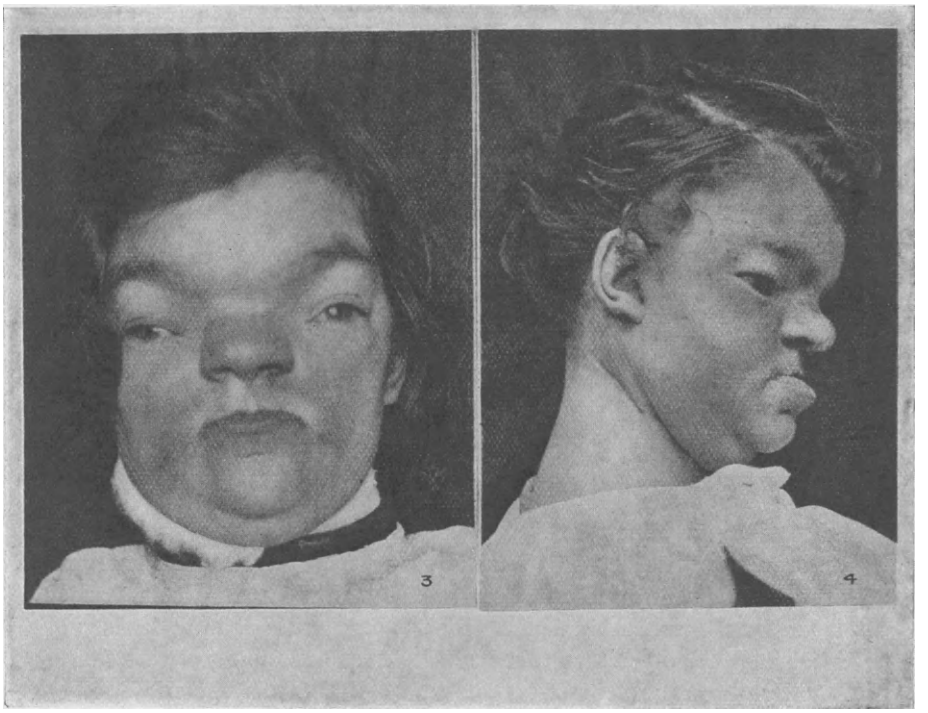


FIG. 73.—Hypertelorism in which " far-apartness " of the eyes is the most marked feature. (D. M. Greig.)

telorism it is essential, also in true oxycephaly sutures are not present but in hypertelorism they are. Reference to Fig. 73 shows at a glance the facial characteristics of hypertelorism. The eyes are set very far apart and, as Greig says, " the patient, like a hare, can see objects directly in front so readily or so well as when placed laterally or when turning the head away from them." There is bilateral strabismus present and probably optic atrophy supervenes, for sight gradually fails.

An interference in development, going back early in embryonic life, present certainly in the chondro-cranium, is laid down, which ultimately leads to an abnormal development of the lesser wings of the sphenoid, a broadening out of the nasal bones with an interstitial bone between them. As Ogilvie and Posel note, the enlargement of the lesser wings will push the whole frontal bone upwards, forwards and outwards and with it the corresponding maxillæ. These observers thought that scaphocephaly, oxycephaly and hypertelorism bear a definite relationship to one another and this relationship consists in a common origin, namely, an abnormal development of the sphenoid bone.

Some cases are familial and mental deficiency has been recorded in the non-familial cases—as in Greig's case.

Amongst the associated defects are syndactyly and acrocyanosis (cyanosis of the extremities), high arched palate, enlarged terminal phalanges of the thumbs and undescended testicles.

A further type of acrodysplasia—*syndactylic oxycephaly*—has been lately described by Greig, in which the skull does not conform anatomically to true oxycephaly and the condition is not clinically the same. In true oxycephaly both the bones of the cranium and the face are affected; in the type under discussion the cranium alone is affected. There is normal mentality, vision is unaffected, but there is syndactyly of both hands and feet, restriction of movement of various joints and scoliosis.

Morquio's Disease.—A familial osseous dystrophy involving the whole skeleton has been described by Morquio. In this disease it is unusual for the head and face to be involved, but Davis and Currier have reported two cases where extensive changes were found in the skulls resembling oxycephaly, together with the presence of optic atrophy and mental deficiency.

Osteitis Deformans.—In this disease there is a primary absorption and rarefaction of bone, followed by an excessive formation of fibro-osteoid tissue. The bones of the skull may share in this disease and become exceedingly thick, so that it may increase in size and show bossing on the vertex together with bulging of the side of the skull. The *juga cerebralia et impressiones digitati* of the frontal bone become more uneven and exostoses may also appear. The sella turcica and the sphenoidal sinus are altered in shape and size, so that it is not to be wondered at that there are ocular syndromes due to involvement of the optic nerves passing through deformed optic

foramina. Wylie has described two cases of optic atrophy in patients affected with this disease. There is diplopia present, indicating damage to ocular nerves, and hearing was impaired in one ear. Paget found that of twenty-three cases examined, four had become blind. Retinal hæmorrhages are found in some cases of osteitis deformans and degeneration of the macular area has resulted in central blindness. Leslie Paton has stated that he had seen five cases of osteitis deformans with optic atrophy. In most of these cases there was evidence of pressure on other cranial nerves, especially the auditory and the trigeminal. The visual fields will vary according to the damage done by the contracted optic foramina compressing the optic nerves.

Ormond has pointed out that the two groups of ocular symptoms in osteitis deformans may be summarised as follows :

(1) Those due to pressure on the nerve as it passes through the optic foramen leading to changes in the field of vision and visual acuity ; also to optic atrophy.

(2) Those due to definite widespread disease of the vascular tissues resulting in retinal and choroidal disturbance with hæmorrhages not primarily affecting the nerve head at all.

Diabetic Exophthalmic Dysostosis

(Synonyms : Hand-Schueller-Christian's disease—dysostosis hypophysaria.

Diabetic exophthalmic dysostosis is an exceedingly rare disease. It was described by Dr. Henry Christian of Boston, in 1919. Schueller also described two cases in 1915. The disease is characterised by progressive decalcification of the cranial and other flat bones, exophthalmos and the presence of diabetes insipidus. The Wassermann reaction is negative. Although the cause of this disorder is unknown, a disturbance in the secretion of the pituitary gland as being responsible for the defects in the flat bones is an hypothesis that has considerable support. Schueller thought the disease was due to pituitary underfunction and hence proposed the name *dysostosis hypophysaria*. It is now known that the tuber and tuberal region are connected with the pituitary and its stalk by definite nerve tracts and channels. Lesions of these parts produce diabetes insipidus and this dysfunction can be controlled by extract of the posterior lobe of the pituitary. In some cases there is a deposit of foam cells in the pituitary gland and tuber cinereum, also

lipoid degeneration may be present, in this respect resembling amaurotic family idiocy and Niemann-Pick disease.

Bach and Middleton have reported the presence of multiple myelomata, associated with diabetes insipidus.

The most recent reference to the Schueller-Christian syndrome is that by Horsfall and Russell Smith in the *Quarterly Journal of*

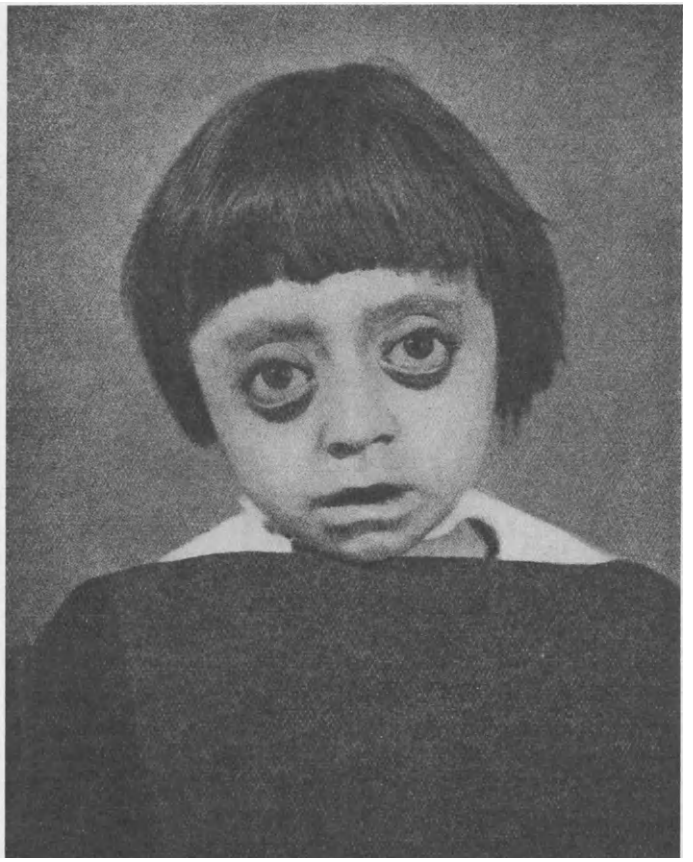


FIG. 74.—Diabetic exophthalmic dysostosis. A girl three and a half years of age. This was the original case described by H. A. Christian of Boston.

Medicine, 1935, vol. 4, No. 13, in which they state that sixty cases have been recorded and they themselves describe a case most fully in which the roentgenograms reveal areas of almost complete

decalcification in the bones of the skull, ribs, vertebræ and femur. In this case there were present polydipsia, polyuria, exophthalmos and defects in the bones. The pupillary reflexes were normal, the disc showed some pallor but no papillœdema, while the retinal vessels exhibited tortuosity without engorgement. They point out that in thirty-six cases recorded exophthalmos was present in twenty-eight. Necropsies on cases with exophthalmos have shown uniform intra-orbital xanthomata. Chester concluded that the basic lesion was a " chronic, non-infective, abacterial, inflammatory granuloma due to the deposition of lipoid substance in the involved tissues." Horsfall and Russell Smith suggest that the pathological condition might be termed " cholesterol granulomatosis."

In children affected by this disease there is a progressive bilateral exophthalmos, as the softening of the cranial bones increases, although the cause of the exophthalmos may be of nervous origin and not by pressure. From a perusal of the literature one gathers that various authors regard the pituitary body, the tuber cinereum and the hypothalamus as being collectively responsible for such an abnormal metabolism. Hausman and Bromberg have emphasised the high incidence of antecedent infection in their list of cases quoted. In the case (Fig. 74) quoted by Christian the little girl, at the age of three and a half years, began to drink more water and pass urine frequently. At the same time the right eye became prominent. In this particular case there was no disturbance of vision, but in other cases reported there is diplopia on looking to either side, the fundus oculi appears normal, no evidence whatever of increased intracranial pressure being found. Radiograms demonstrated the condition of the skull bones. The mentality of these children is normal, and there are no other symptoms of central nervous system involvement. The disease is progressive and may terminate fatally.

THE LAURENCE-MOON-BIEDL SYNDROME

When Nettleship, in 1908, reviewed the literature on retinitis pigmentosa he drew attention to the resemblance of certain cases described by Laurence and Moon with those reported by Jonathon Hutchinson in 1900. These cases belonged apparently to the same family, but Hutchinson had seen them ten years after Laurence. Their descriptions were similar with minor discrepancies.

It was in 1866 when J. Z. Laurence and R. C. Moon, surgeon and

house-surgeon respectively of the Ophthalmic Hospital, Southwark, described four cases of retinitis pigmentosa occurring in the same family and accompanied by general imperfections of development. There were eight members in this family, the two eldest being quite healthy, but the third, fourth and fifth children were " afflicted and had bad sight," while the eighth, the only girl of the family, was " quiet and slow and had bad sight." In all four cases there was night blindness, but instead of contracted fields of vision there did not appear to be any limitation of the normal field in any one case.

In 1901 Fröhlich described a case of tumour of the hypophysis cerebri without acromegaly, in which there was the typical hypophyseal adiposity with genital dystrophy. Gradually from the observation and writings of both ophthalmologists and endocrinologists the bizarre combination of obesity, hypogenitalism, polydactyly and mental retardation came to be fully recognised as a familial syndrome. This full complement did not emerge until Biedl described it in 1922. He showed a brother and sister in which there were no changes of the hypophysis cerebri, no signs of cerebral tumour or abnormal pressure, but they had all the characteristic symptoms of what has come to be known as the Laurence-Moon-Biedl syndrome. It was at the suggestion of Solis-Cohn and Weiss in 1925 that this strange combination of disorders has passed under the name of the Laurence-Moon-Biedl syndrome.

In 1933 Hain reported developmental anomalies in the albino rat having some resemblance to those found in the Laurence-Moon-Biedl syndrome in man. These consisted of microphthalmia, lens opacities and blindness, while in families related to these rats other peculiarities, such as skull deformities and genital malformations, presented themselves.

Since Laurence and Moon described their cases some seventy-nine have been reported. In 1935 Cockayne, Krestin and Sorsby while describing their cases, which were shown at the Royal Society of Medicine, exhaustively surveyed and analysed the literature on the subject.

(*a*) They point out that in all the cases recorded with one doubtful exception there was *retinal degeneration.* This was chiefly of two kinds, one in which the appearance of the fundus strongly resembled the typical retinitis pigmentosa (see Plate VII), the other showed the appearance of the typical cerebro-macular dystrophy—

stippled maculæ (see Plate XI), with in some cases partial optic atrophy and slight peripheral degeneration of the retina. Here and there cases of atypical retinitis pigmentosa have been described, but the typical form is the fundus appearance characteristic of the Laurence-Moon-Biedl syndrome. Other visual defects were constant—nystagmus was frequent while strabismus and optic atrophy were occasional. Apart from ophthalmoscopic appearances the eye symptoms differ in one important respect from the typical retinitis pigmentosa—the juvenile onset of a severe fundus lesion is uncommon in classical retinitis pigmentosa (Sorsby).

(*b*) *Polydactyly* or *syndactyly* were almost invariably present, but in the Laurence-Moon-Biedl syndrome there was never more than one extra digit.

(*c*) *Mental development* shows retardation in every instance. As this syndrome comes to be commonly recognised many cases like that of Fig. 75 will undoubtedly be discovered in the various mental hospitals. The skull radiograms show a sella smaller than normal, but in adult cases Biedl has drawn attention to the unusually high dorsum.

(*d*) *Adiposity.*—Obesity was present in practically all the cases reported and is of the Fröhlich type, that is, the fat is increased in the proximal parts of the limbs, the normal contours in the female are exaggerated, while it produces a feminine outline in the male. Although Laurence and Moon described the presence of hypo-genitalism in their cases this is not invariably present.

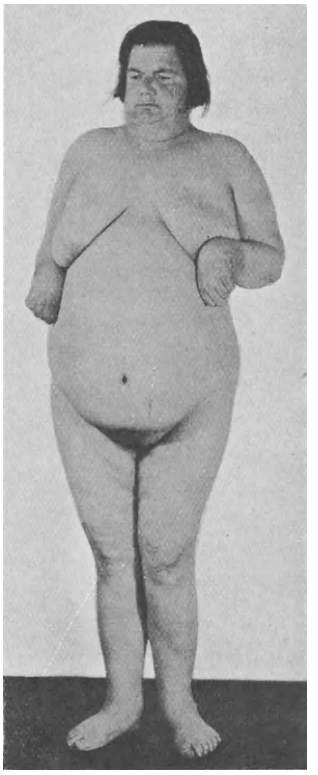


FIG. 75.—A case of Laurence-Moon-Biedl syndrome. Note obesity of abdomen and mammæ. Patient exhibits constant choreiform movements of the face as well as rotatory movements of the eyes. Right hand possesses six digits with syndactyly of middle and double ring fingers. (Ring finger presents two distinct nails). The left foot exhibits six well-formed toes. The right foot is normal. (Case under R. M. Stewart and published by P. Klenerman.)

The sexes appear to be equally affected, and although several

cases may occur in the same family there are no records of the occurrence of the syndrome in their ancestral relatives. Consanguinity has been noted in two cases only.

Cockayne suggested that the meso- and epi-blastic parts of the syndrome are recessive and are due to mutations of two genes in the same chromosome.

Some attribute the skeletal manifestations to embryonic pressure, thus resembling in its ætiology the congenital defects found in achondroplasia, Mongolian idiocy, cleidocranial dysostosis and club

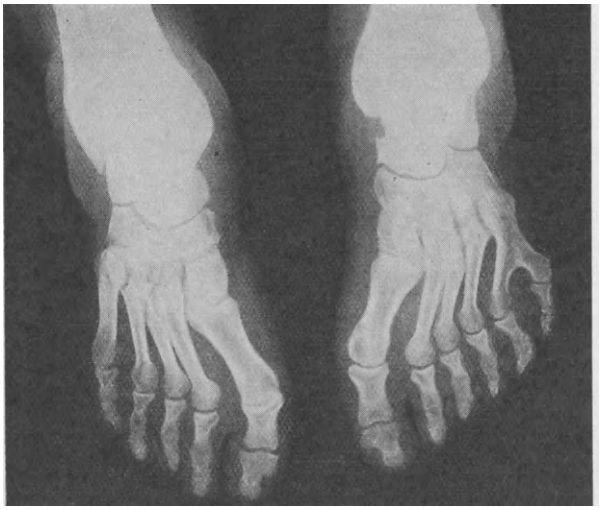


FIG. 76.—Radiogram of feet of case illustrated by Fig. 75. The little toe of left foot presents two terminal phalanges, two middle phalanges and two heads to a common metatarsal bone.

foot. Internal hydrocephalus has been suggested by Raab as a cause of the skeletal deformities and the inborn tendency to retinal changes which usually develop in childhood. Biedl considers the disease of cerebral rather than hypophyseal origin. In one of Biedl's cases treated with thyroid and pituitary extracts vision improved, the menses were re-established and loss of weight to the extent of 13 lbs. occurred.

The illustration (Fig. 75) was published by Pauline Klenerman, the original of which was kindly sent to the author by Dr. R. M. Stewart. This particular case under Dr. Stewart's care was a woman aged forty years. She was extremely obese, particularly on the

abdomen and mammæ. The pupils were normal, there was definite nystagmus and lenticular opacities were present. Ophthalmoscopic examination showed irregular pigment all over the retinæ. Her right hand possessed six digits, with syndactyly of middle and ring fingers (ring finger had two distinct nails). The left foot exhibits six well-formed toes and no webbing. The illustration of the radiogram of the feet shows the bifurcating fifth metatarsal of the left foot (Fig. 76). The patient is an imbecile and is disorientated, remaining all day in a wheeled chair.

When shown an ophthalmic drawing of the fundus of a patient under the care of Dr. W. M. McGrath I said the picture he held in his hand was that of a typical retinitis pigmentosa with distinct atrophy of the disc. The drawing had been taken from the eye of a woman who appeared to be a definite case of the Laurence-Moon-Biedl syndrome.

Leontiasis Ossea.—In this disease there is an overgrowth of the bones of the face and skull, sometimes producing deformity of the cranium and the various cavities of the face. If there is increased intracranial pressure, papillœdema followed by post-neuritic atrophy, and sometimes primary optic atrophy, is found.

Mongolian Idiocy.—This is a condition found in quite young children, producing imbecility together with marked physical abnormalities. The appearance of the face is characteristic in this disease (see Fig. 77). The uneven and oblique attachment of ligaments in the deformed orbit produces an oblique direction of the palpebral fissures. The eyes do not appear symmetrically placed, the mouth is usually kept open and the tongue seems too large for the mouth ; speech is difficult. In spite of repeated reproof, the child will not keep his mouth shut. There are other deformities, such as small hands with malformed fingers. These children show a varying amount of mental deficiency. Ormond states that some form of lens opacity is often present, producing a cataract of the lamellar type. Many of these children squint, but operation is not readily advised, as the tissues of such children appear extremely unhealthy and react badly to surgical interference. The eyelids are often affected by blepharitis of a very chronic type.

To those who are specially interested in the subject of Mongolism I would suggest the perusal of the late Dr. Crookshanks' work, " The Mongol in Our Midst," in which he discusses the homologies of mongols, imbecile mongoloids and orangs ; the rarity of Mongolism

among the Jews; the lowered state of development of the external organs of sight and the corresponding lack of development in the occipital lobes of the brain. Also in this volume reference will be found to the work of Kurz on the brain of the imbecile mongoloid as compared with the brain of the adult European. Crookshanks says

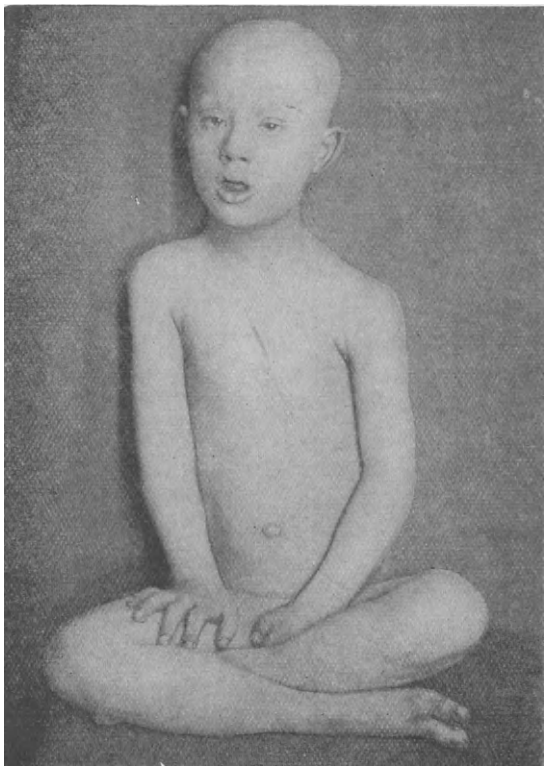


FIG. 77.—A mongol in typical attitude. (From F. G. Crookshank's book "The Mongol in Our Midst.)

that in a certain number of cases it has been found that the mother of imbecile mongoloids has suffered from hypothyroidism during the carrying. He showed that in Mongolism there is not only a general deficiency of thyroidal internal secretion but a general quantitative deficiency affecting the pituitary body and pineal gland, the gonads and the thymus if not the suprarenal gland. In the Belgrave Hospital for Children he employed as treatment a combination of

thyroid, thymus, suprarenal, gonad and pituitary substances and was convinced that decided improvement took place in imbecile mongoloids. Reporting on the examination of the brains of ten cases of Mongolian-idiocy Davidoff says the morphologic changes that are constant are small cerebrum and brain stems, the embryonic convolutional pattern and small content of the ganglionic cells of the third cortical layer.

A general description of 177 mongols has been given by Brushfield, together with the theories connected with the ætiology of such a condition. Brushfield and Berry cannot find that syphilis is in any way the cause, but Berry points out the fact that the fathers of a great many of her patients have been shell-shocked, and so it appears that it is the father and not the mother (as hitherto thought) who is sexually responsible.

Hydrocephalus.—Hydrocephalus may be congenital or acquired. The former is due to maldevelopment of the brain and the condition may be associated with meningocele and spina bifida. Congenital syphilis may be a causal factor. In congenital hydrocephalus nervous symptoms follow the enlargement of the head and become marked as the cranial sutures close. Papillœdema appears at this stage, but this is not so commonly found as optic atrophy. Papillœdema may even be superimposed on an optic atrophy.

Acquired hydrocephalus may be due to intracranial neoplasm, meningitis or ependymitis. If papillœdema follows and is long continued, it may be necessary to perform a decompression operation to avoid consecutive optic atrophy and blindness.

In the congenital and early acquired forms of hydrocephalus the downward pressure on the orbits sometimes causes the eyeballs to be rotated downwards, which may interfere with vision. If there is pressure on the optic chiasma or tracts, optic atrophy may ensue. This, indeed, is frequently found to be the case. Bitemporal hemianopia is sometimes discovered, but rarely papillœdema. In acquired hydrocephalus of later life there may be the three cardinal signs of raised intracranial pressure—papillœdema, vomiting and headache—and in addition to these an ataxic gait may be present. If the bitemporal hemianopia is present, it will help in arriving at a true diagnosis of the case.

In long-standing cases of internal hydrocephalus the floor of the third ventricle becomes distended and may press upon and flatten the chiasma so that the lateral fibres may be indented by

the internal carotids. In this way binasal hemianopia may be produced.

Cerebral Diplegia or Little's Disease.—This disease, sometimes known as congenital spastic paralysis, is characterised by spasticity of both sides of the body together with weakness, but the rigidity is out of proportion to the weakness. Mental impairment of all degrees is found.

There are many theories put forward to explain this disease. Little, by whose name the disease is called, attributed it to asphyxia or meningeal hæmorrhage resulting from difficult labour. Maternal syphilis has been suggested, but Collier stated that this condition was due to agenesia of the nerve cells supplying the pyramidal system. Degeneration and disappearance of neurones are the anatomical lesions constantly found with resulting atrophy, sclerosis and arrested cerebral development.

Patten concludes that "the frequent occurrence of bilateral motor involvement together with defect in intelligence indicate something more than the effects of trauma or vascular accidents in the neurologic conditions of the new-born infant." He further states that a developmental defect or an arrest of development exists which affects the integrity of the cortical cells or the proper myelinisation of the corticospinal tracts and association fibres.

This is not a hereditary disease. At first the child appears normal, but after some weeks or months rigidity of the legs may be noticed; that of the arms may follow, but it is not so greatly marked. There is ataxy, mental dullness, nystagmus, squint and even optic atrophy may appear.

Otitic Hydrocephalus.—A form of this disease, known as otitic hydrocephalus, has been described by Symonds. He believes that the not very uncommon complication of otitis media may be a state of increased intracranial pressure due to the presence of an excess of normal cerebrospinal fluid. In this disease there is either an excessive secretion from the choroid plexus or a defective absorption through the arachnoid villi. The term otitic hydrocephalus implies, according to Symonds, no active process of inflammation. In the fully developed state there is intermittent headache, together with papillœdema. A sixth nerve paralysis on the side of the discharging ear has been noticed in some cases, while the temperature and pulse rate are normal. In a case which lately came under the author's care, at the first examination of the eyes a papillœdema

of two diopters was found in the left eye of a woman aged forty-nine. She was recommended for admission to hospital, but no other neurological signs or symptoms were found. This patient had suffered somewhat previously from otitis media. Pus had appeared discharging from the left ear ; this had now ceased, the swelling of the disc had disappeared, but the disc edge still remained indistinct. There is no impairment of vision. The disease occurs almost always in children and adolescents, but, as seen above, it may occur in older people. The differential diagnosis from cerebral abscess in the early stages of otitic hydrocephalus may be a matter of considerable difficulty, especially if an early phase of meningitis has caused for the time being an excess of cells and protein in the cerebrospinal fluid.

In a recent publication Bourgeois, who has made an extensive study of otitic hydrocephalus as a sequel to otitis media, a condition first probably recognised by Bárány about twenty-six years ago, states that this condition must be distinguished from meningitis serosa and from rapidly fatal cases of meningitis. He insists repeatedly upon lumbar puncture and the examination of the cerebrospinal fluid. He describes three forms : an external diffuse form over the cortex, an internal form in which the cerebrospinal fluid becomes encysted in the ventricles, and a localised form in which the fluid is encysted in the posterior fossa. The third form is not so uncommon as the other two.

Fractures of the Base of the Skull.—The sixth cranial nerve is frequently affected by an injury to the base of the skull, less commonly the third and infrequently the fourth nerve (see p. 407).

One sees a large number of accidents produced by motor cycling, and in many of these cases, whether there is a fracture present or not, the optic nerve has been irreparably damaged. There is instantaneous blindness on the side of the injury, and not until many weeks have elapsed does the optic disc on that side become white. On the side of the injury the direct light reflex is lost, but the consensual light reflex on this side is still present. If the optic nerve is found to be injured or if a paralysis of an extra-ocular muscle is discovered, the conjunctiva beneath the upper lid should be examined, as signs of a hæmorrhage filtering from above through the bulbar conjunctiva will indicate a fracture of the roof of the orbit. In this situation it is extremely difficult to detect a fracture by means of radiography. In one of the writer's cases where there

was no paralysis present, it was this hæmorrhagic sign which indicated such a fracture. The injury followed the slamming of the door of a car against a young girl's head.

Hæmorrhage into the sheath of the optic nerve will produce swelling of the nerve head.

Pulsating exophthalmus may occur as the result of a fracture of the base of the skull, an aneurysmal varix being formed between the internal carotid and the cavernous sinus. Both the conjunctival veins and those of the fundus are dilated, the movements of the eyeball are impaired, and the whole globe may become disorganised.

A fracture of the base of the skull may involve the chiasma, so that on examining the patient subsequently a bitemporal hemianopia, possibly quadrantic in form, may be found (see Chapter X).

Dermoid and Hydatid Cysts of the Cranial Bones

Various cysts are found both in the bones of the skull and within the meninges. Some, as Cushing suggests, originate in the diplœtic spaces, and as they grow the tables of bone are pushed apart, the inner being that which suffers most from erosion. These cysts—" tumeurs perlées " of Curveilhier—have a characteristic mother-of-pearl appearance. Their point of origin may be in the region of Rathke's pouch, third or fourth ventricle or extra-durally, especially in the tempero-parietal region.

Hydatid cysts are also found within the skull. Multiple cysts within the cranium have been found in groups of soldiers who have partaken of the same polluted food in tropical countries.

There are two chief cestode larvæ found in man—the *Cysticircus cellulosæ*, the larva of *Tænia solium*, and the *Echinococcus*, the larva of *Tænia echinococcus*.

MacArthur says : " Cysticircosis has been brought into prominence by the discovery that epilepsy developing in British soldiers during or after service abroad is commonly due to this disease." When the eggs enter the alimentary tract of man the liberated embryos penetrate the intestinal mucosa and are borne by the blood stream to their first habitat, the situation of which is determined by the active movements of the parasites and not by mechanical means. In man, MacArthur says, the embryos have a predilection for the brain and musculature—in fact, in any organ except the bone. The parasites may lodge in any part of the brain—meninges, cortex, basal ganglia, ventricles, white substance—but in general, grey

matter is invaded more commonly than white. The eye, too, may become involved. The parasite lodges most frequently beneath the retina and with growth may burst through the retina and may be seen moving actively in the vitreous, as was formerly described by von Graefe, or may succeed in invading the anterior chamber. Sometimes the embryos invade the brain in such large numbers that the intensity of the reaction they provoke is incompatible with life, but in less acute forms when papillœdema is present, with irregular fever and headache, there are motor and sensory changes and deepening mental stupor ending in death.

Calcification sufficient for detection in a skiagram points to the death of a parasite at least three years before. As a rule, the cysticirci in the brain do not calcify and so cannot be seen in the skiagram. The numerous parasites commonly present in the brain and their wide distribution there, contra-indicate a general resort to surgery. A temporary amelioration by the removal of one or more cysts has been followed by a recurrence of symptoms ending in death.

Radiography, in the case of dermoid cysts, will often demonstrate an area of calcification within the cyst, together with an area of rarefaction of the skull bones in its neighbourhood, while in the case of hydatid cysts growing within the skull bones the appearance of rarefaction is presented. If these cysts are in the neighbourhood of the orbit, vision may be diminished by pressure on the optic nerve as it passes through the optic foramen, or pressure on a nerve passing into the orbit may cause paralysis of one or more ocular muscles. Rand demonstrated the change in the fields of vision in a case of an intracranial dermoid cyst which contained hair follicles, sweat glands and stratified squamous epithelium. The cyst filled the left middle temporal fossa. There was severe papillœdema present in each eye. The radiograms showed an area of calcification in the cyst wall.

Intracranial Aneurysms, Subarachnoid and Cerebral Hæmorrhage

Recently the subject of subarachnoid hæmorrhage has claimed a good deal of attention. It has been pointed out that ophthalmoscopic examination may render considerable assistance in forming a correct diagnosis. In addition to the grosser ophthalmoscopic appearances there may be found changes in the visual field due to pressure on the visual pathways or pressure upon the ocular nerves,

producing a paralytic squint. A case which the writer recently observed was that of a young woman aged thirty-three who was admitted to hospital. She had suddenly become unconscious, and upon regaining consciousness discovered that she was practically blind. The appearance as seen by the ophthalmoscope in the left eye is represented by Plate XIII. A large amount of blood was lying in the vitreous in front of the left optic disc, the latter being completely obscured by the mass of blood. On the retina of the right eye and below the optic disc a large subhyaloid hæmorrhage was seen, but no blood had escaped into the vitreous. There was a complete paralysis of the left oculomotor nerve. The diagnosis of subarachnoid hæmorrhage was made and confirmed by the examination of the cerebrospinal fluid; the latter was found to be evenly mixed with blood. That was four years ago.

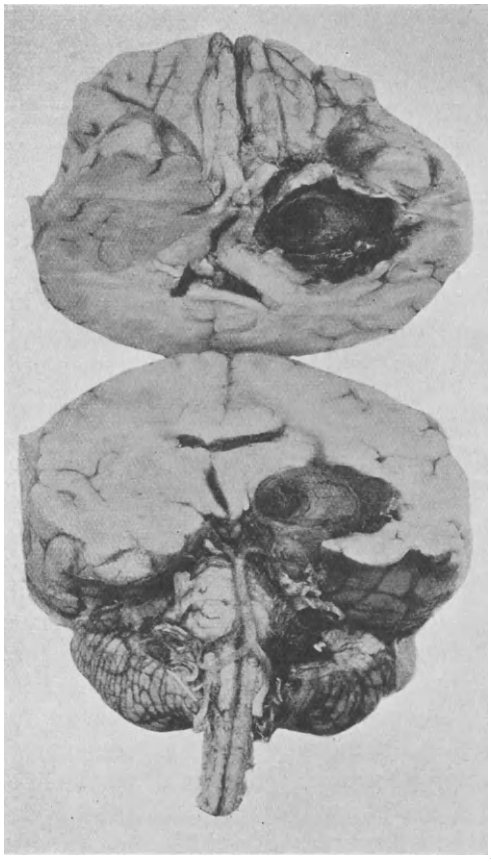


FIG. 78.—Coronal section of brain showing aneurysm involving the whole of the left side of the circle of Willis. (Carnegie Dickson.)

On going to press news of the death of this patient has just come to hand. This case was admitted to hospital under the care of the late Dr. E. D. Macnamara, but was finally placed under Dr. L. R. Yealland's supervision. It was with the latter's kind permission that the report of the autopsy was received by the author, and, Dr. Carnegie Dickson, with his usual generosity, has supplied the photograph of the brain (see Fig. 78) cut

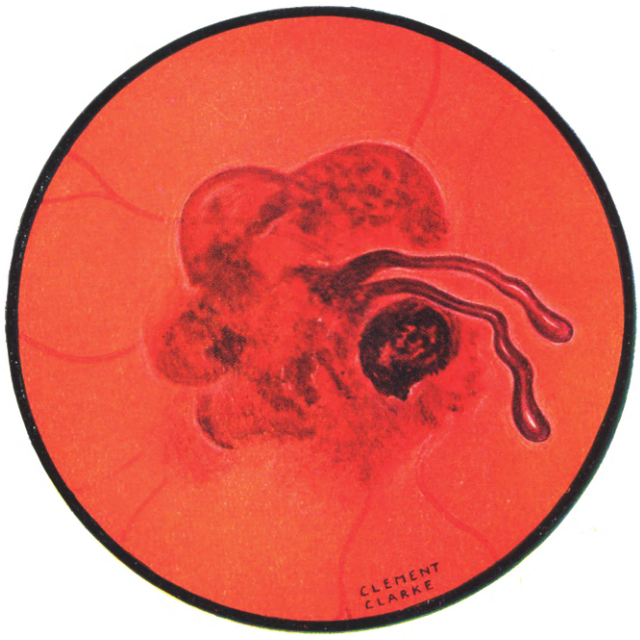


FIG. 1. FUNDUS IN SUBARACHNOID HÆMORRHAGE

A large mass of blood has made its way into the vitreous, coming forward in cleft-like spaces towards the back of the lens. Paton has described a case where the hæmorrhage burst into the vitreous in both eyes. Such hæmorrhages, he believed, were due to direct leakage through the lamina from the hæmorrhage in the sheath of the optic nerve.

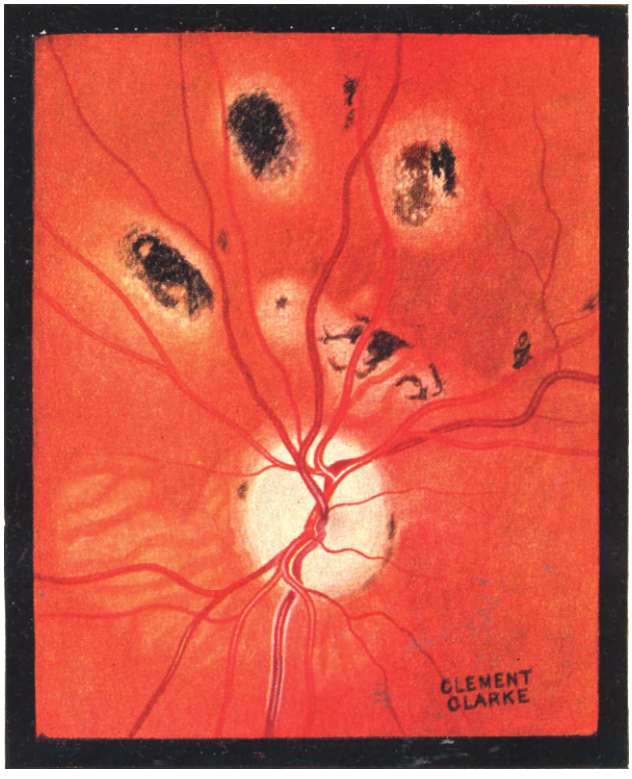


FIG. 2. DISSEMINATED CHOROIDITIS

Bilateral disseminated choroiditis seen in untreated congenital subjects of syphilitic disease; in acquired cases the condition is unilateral. Should a patch of choroiditis fall on the macula central vision becomes greatly diminished.

[*To face p.* 208.

across in a coronal manner. Dr. Carnegie Dickson writes saying, " Herewith a copy of a photograph of the very interesting case of aneurysm of the circle of Willis. You can take your choice as to which blood-vessel it has arisen from, as it involves the left posterior cerebral, which, as you can see in the photograph, passes directly into it, or the left internal carotid or middle cerebral, as the aneurysm involves all the left side of the circle of Willis."

Another case, one under the care of C. A. Birch, was that of a

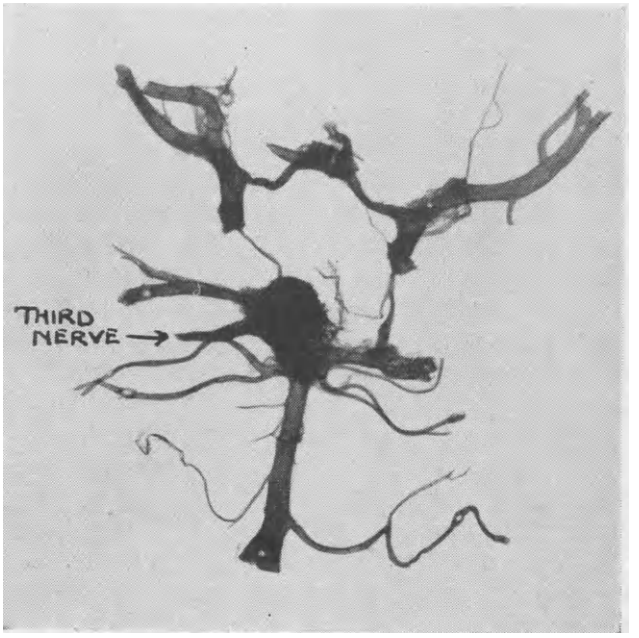


FIG. 79.—Grenz ray photograph showing aneurysm at junction of right middle cerebral and right posterior communicating arteries. The third nerve is adherent to the aneurysm. (C. A. Birch.)

married woman, aged thirty-seven years, who suddenly developed a severe occipital headache and became stuperose. On admission to hospital she was found to be disorientated and restless. Her symptoms were neck rigidity ; Kernig's sign absent ; pupils, fundi and cranial nerves normal ; blood pressure 185/105. The cerebrospinal fluid was uniformly bloodstained. Lumbar puncture improved her condition. In a fortnight the cerebrospinal fluid was clear, but three weeks later she suddenly became comatose and

again the cerebrospinal fluid was bloodstained. There was a complete right oculo-motor paralysis and bilateral papillœdema with retinal hæmorrhages. At autopsy a large subarachnoid hæmorrhage was found at the base of the brain and also an aneurysm, the size of a pea, at the junction of the right middle cerebral and right posterior communicating arteries to which the third nerve was attached. (See Fig. 79.)

The commonest cause of intracerebral arterial hæmorrhage is rupture of an atheromatous artery in a case of high blood pressure. Other causes are birth injuries, hæmorrhagic pachymeningitis, encephalitis, poliomyelitis, tuberculous meningitis, sinus thrombosis, leukæmia, anthrax infection, neoplasms, aneurysms occurring in coarctation of the aorta and in polyscleritis acuta nodosa, congenital and syphilitic aneurysms.

The commonest cause of direct rupture of vessels according to Turnbull is medial degeneration associated with abnormally high blood pressure. The arteries which rupture under these circumstances are with few exceptions the central branches of the cerebrals. Of thirty-three examples in twenty-nine cases he found medial degeneration following upon congenital developmental deficiency.

Of the various causes of cerebral hæmorrhage, apart from arteriosclerosis, Collier considers the three most important to be (1) encephalitis, (2) neoplasms and (3) aneurysms. He says that many cases of encephalitis lethargica have been complicated by subarachnoid or cerebral hæmorrhage.

Small berry-like multiple aneurysms have been discovered and classified as congenital, but such are not syphilitic. The lesions produced by the rupture of such aneurysms are the commonest cause of cerebral hæmorrhage in younger subjects. It has been pointed out that the term congenital aneurysm, as applied to many cases, is inaccurate and does not express what is meant, and that the assumption that there is an aneurysm in all these cases, whether found or not, overlooks the fact that a rupture may occur without necessarily being preceded by an aneurysmal dilatation. Substantial evidence is given to such a statement by the fact that hæmorrhages in the vitreous are commonly seen, and close examination of the fundus before the rupture and after absorption has shown neither dilatation of the vessels nor other nævoid angiomatous conditions. As already mentioned, there are cases where the retinal veins show a certain amount of varicosity, and such cases occasionally suffer

from mild epileptiform attacks. It is possible, therefore, as Hall has suggested, that a detailed examination of the whole brain might show definite evidence of widespread vascular change.

Of 5,432 consecutive post-mortem examinations of the brain carried out at the London Hospital, congenital intracranial aneurysms were present in the proportion of 1 to every 125 cases (Turnbull). Schmidt states that the true intracranial aneurysms are almost exclusively localised on the greater extracerebral arteries ; they are seldom found in the brain substance. The place of predilection for an aneurysm is a bifurcation of one of the great arteries or the place where smaller branches arise, especially the origin of a communicating artery. Greenfield examined the cerebral arteries in five non-atheromatous cases of cerebral aneurysm and found a common defect present in all. This defect was in the muscular coat of the artery, and was found, not only at the point where the aneurysm formed, but at practically every bifurcation of the cerebral vessels in these subjects. Wyley Forbus describes the absence of the tunica media in some cases. Aneurysms are most frequently to be found in the middle cerebral artery, then in decreasing order in the basilar artery, the internal carotid artery, the anterior cerebral artery, the posterior communicating artery, the anterior communicating artery, the vertebral artery, the posterior cerebral artery and the inferior cerebral artery. Small aneurysmal dilatations on the anterior communicating artery occur in 10 per cent. of all post-mortem specimens. Busse, in discussing some cases of aneurysm of the anterior communicating artery, mentions that in all the living cases he found little or no pupillary change, hæmorrhages were present on the fundus, papillœdema in some, no constriction of fields and no ocular paralysis. In some cases dizziness and severe headache radiating even to the spine, and in others complete unconsciousness.

Intracranial aneurysms most frequently cause symptoms between the ages of twenty-five and forty. Birch, therefore, points out that subarachnoid hæmorrhage in patients over fifty is more likely to be due to arterio-sclerosis rather than to the " congenital " variety of aneurysm. Foster Moore quotes and beautifully illustrates a case of unruptured aneurysm of the ophthalmic artery in infective endocarditis which had produced complete rupture of the optic nerve. Schmidt considered the ætiology of his 23 cases as follows : congenital anomalies, local diseases in the arterial wall, traumata,

arterio-sclerosis ; but he considered syphilis as unimportant, although in four cases syphilis of the central nervous system was found. Fearnsides estimated that while 96·2 per cent. of aneurysms of the aorta were due to syphilis, only 6·25 per cent. of aneurysms of small arteries such as those of the circle of Willis were due to the *Spirochæta pallida.*

Diagnosis before and after Rupture.—The symptoms of subarachnoid aneurysm before rupture are somewhat vague, but patients who have bled into the subarachnoid space from a ruptured basal aneurysm have sometimes been in remarkably good health until a further rupture takes place. Adie has pointed out that the condition known as ophthalmoplegic migraine must always give rise to the suspicion that the symptoms were really due to the presence of a leaking basal aneurysm (see also Sjöqvist's paper).

Paroxysmal headache is sometimes complained of, and this too resembles migraine. One of Riddoch and Goulden's cases suffered from recurrent headaches over a period of four years, brought on by exertion or prolonged stooping and culminated in vomiting.

Tinnitus.—One of the symptoms mentioned in Case 4 of Riddoch's is the presence of noises in the head and partial deafness.

Epistaxis.—Epistaxis may occur periodically and gives decided relief to the patient. A patient under the author's care suffered from a blood pressure of 220. A hæmorrhage took place into the vitreous of the left eye. Somewhat later, periods of unconsciousness, for half an hour at a time, followed. During these periods her maid could not rouse her as she could easily do from sleep. The patient would then suddenly become conscious, feeling in her normal state again. One day a severe epistaxis came on, lasting more or less intermittently for a week and causing the blood pressure to fall to 157. Since then there have been no unconscious fits. Immediately before this attack of epistaxis the patient called at an optician's shop and said she had arranged to meet a friend there. She walked up and down outside the shop for half an hour, and as the friend did not appear she went back into the shop and left her card, saying if her friend called she was to come to her flat for lunch. She took a taxi home. Later in the day my patient realised that she had been waiting for a friend who she knew perfectly well had died ten years previously. This was a period not of unconsciousness but of disorientation in time.

Paralysis.—One of Jefferson's cases when shopping, found she had

diplopia and was suffering from a severe headache. Then for six months the patient suffered from a partial ptosis and was actually recovering from a complete right-sided ophthalmoplegia when first seen. There was no doubt that she had had paralysis of all the

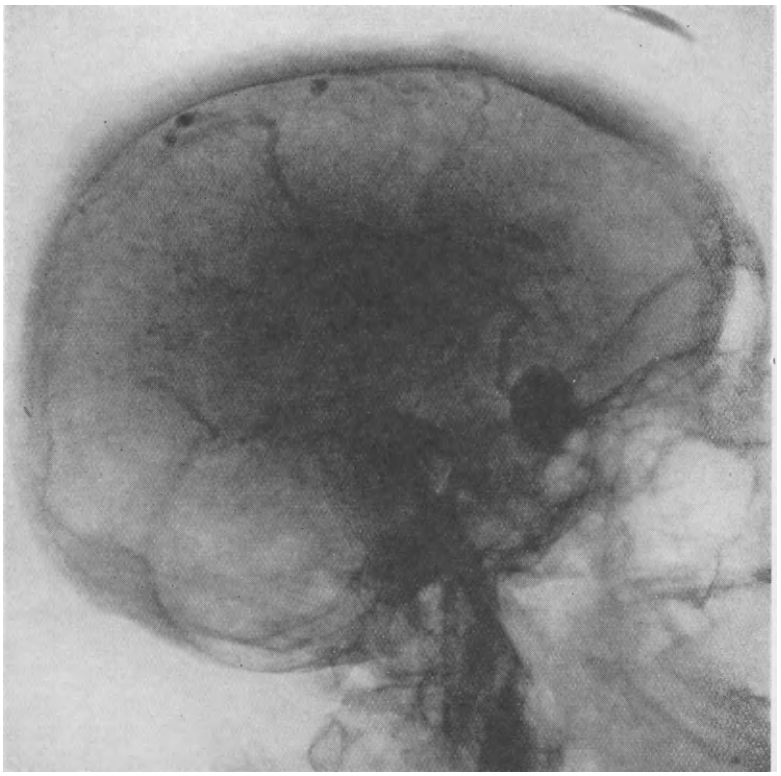


FIG. 80.—Arteriogram showing aneurysm of the right internal carotid artery at the cavernous sinus. Jefferson points out that this particular arteriogram is also partly a phlebogram, for at the top of the picture are seen the issuing cerebral veins. (Arteriogram of a case under G. Jefferson.)

ocular muscles on the right side at some former date. Both discs were normal and visual acuity was normal also. There was depression of the right corneal reflex and hypo-æsthesia of the first division of the trigeminal nerve. The arteriogram (Fig. 80) revealed an aneurysm of the internal carotid artery in its intracavernous portion.

The *diagnosis of actual rupture* into the subarachnoid space in the

case of a large effusion is indicated by meningeal irritation of dramatically sudden onset. Headache, usually occipital, is severe and stiffness of the neck is often marked. In Riddoch's case of recurrent epistaxis the bleeding was always preceded by sleepiness, yawning and sensations of heat on the top of the head. One evening the patient went to bed thinking his nose would soon begin to bleed, but he awoke in the morning with severe occipital headache and rapidly became unconscious. If the hæmorrhage is extensive coma deepening into death may supervene. Such a hæmorrhage from a large tear in the wall of the aneurysm is indistinguishable from that of a fatal intracerebral hæmorrhage. However, in less severe cases many unconscious patients survive. The patient may then become partly conscious, he may vomit, there may be disorientation and delirium with severe headache. It is said that transient and variable extensor plantar responses may be present. Focal signs may show themselves by the blood impinging on certain cranial nerves producing paresis or paralysis. Loss of corneal sensibility and strabismus may occur.

The diagnosis may be confirmed by (1) ophthalmoscopy and (2) lumbar puncture. As is shown by Plate XIII, blood may be found in the vitreous or on the retina either in minute flame-shaped patches or large subhyaloid diffusions. On Plate XIII the hæmorrhage has followed clefts in the vitreous, hence the large tongue-like projections. The main factors determining the occurrence of an intraocular hæmorrhage in cases of bleeding into the cerebral subarachnoid space from a ruptured aneurysm are the severity of the leak, the distance of the aneurysm from the optic nerves and the presence or absence of arachnoid adhesions in the cisterna basalis. (Goulden.) The nerve head may be swollen (see Fig. 81). Lumbar puncture shortly after rupture will, in the great majority of cases, reveal blood in the cerebrospinal fluid. Such blood must be disguished from that which might result from the puncture of a vessel by the needle. It has been suggested by Froin that three successive samples be taken and in all the same complete admixture of blood and spinal fluid should be found. No coagulum appears when the specimen is allowed to stand and the supernatant fluid should be yellow in appearance. Concentration of blood cells may be very high or the fluid may appear clear to the naked eye.

Symonds describes how the blood in various conditions may reach the subarachnoid space. He points out that in diagnosing sub-

arachnoid hæmorrhage from cerebro-meningeal hæmorrhage, the bloodstained fluid originating from an intracerebral hæmorrhage may rupture internally into one of the ventricles or externally into the subarachnoid space producing permanent cerebral damage (if indeed any such case recovers).

Birch points out the interesting fact that in the case where the blood pressure had risen to 250/150 the cerebrospinal fluid pressure

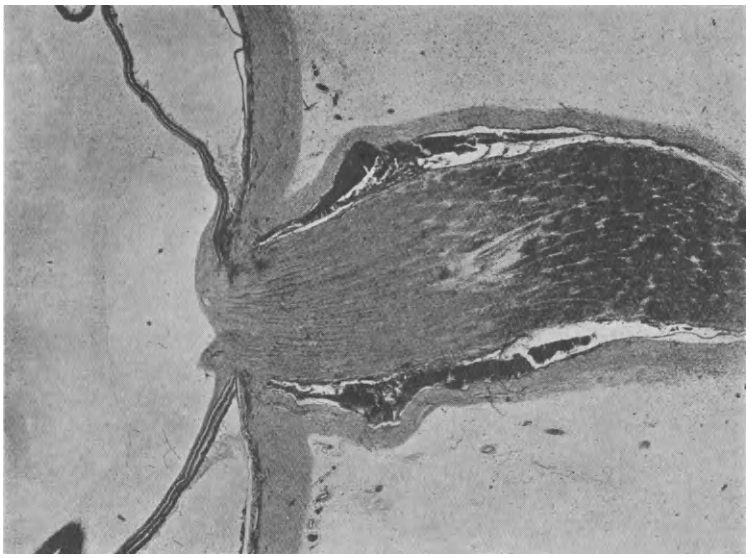


FIG. 81.—Longitudinal section of optic nerve. The vaginal sheath of the nerve distended with blood. No trace of blood in the nerve itself. Note the swelling of the nerve head into the vitreous due to compression of the central vein by pressure from distension of the subarachnoid space and traction on the vein in the vaginal space caused by separating the dural and pial sheaths. (From a section by Professor Turnbull and published by G. Riddoch and C. Goulden.)

was 230. Removal of 12 c.c. of the fluid caused the blood pressure to fall at once to 150/110. He adds that in the present state of our knowledge lumbar puncture should be performed to make a diagnosis, but should not be repeated while bleeding is going on unless there is evidence of a dangerous rise of intracranial tension.

It is but rare that an aneurysm of an intracranial artery is diagnosed before rupture. Reference to Plate I (mid-brain, third and fourth nerves) will indicate the nature of the evidence of the

situation of an aneurysm by its pressure. Pressure symptoms include primary optic atrophy in one eye, defects of the visual field, paralysis of the third, fourth and sixth cranial nerves, exophthalmos, pain and anæsthesia in the cutaneous area supplied by the first division of the trigeminal nerve. It will be readily understood that these symptoms will be produced by anteriorly situated aneurysms, while those of the basilar, vertebral or cerebellar arteries produce symptoms of crossed hemiplegia with possibly facial weakness on the side of the lesion.

Radiography can be of great assistance in the diagnosis of cranial aneurysms. Both Meadows and Purdon Martin mention the appearance of a calcified ring in the area of the aneurysm. In Meadows' case the presence of the aneurysm was further confirmed by injecting thorotrast into the common carotid. The radiogram of the skull which he illustrates, clearly indicates the site and its nature, most probably an aneurysm of the internal carotid artery (Loman).

In addition to erosion of the skull and calcification of the sac a most convincing diagnosis of aneurysmal dilatation is shown by injecting thorotrast into the internal carotid artery as described and illustrated by Moniz, Urban Löhr and Sai. Such an arteriogram has been sent to me by Mr. G. Jefferson. His patient, a woman fifty-seven years of age, suffered severe headaches, pain on right side of head (second division of the trigeminal), diplopia and ptosis of right eyelid. There was also paralysis of the right abducent nerve, while the corneal reflex was almost absent. Fig. 80 illustrates the arteriogram obtained after injection of thorotrast. An aneurysm of the right internal carotid artery was present at the cavernous sinus. Jefferson's treatment consisted of occlusion of two-thirds of the right internal carotid artery by three wide ligatures.

When the hæmorrhage has occurred, papillœdema may be present; it is usually bilateral and not of great amount. Both retinal and vitreous hæmorrhages may be present (see Plate XIII). These hæmorrhages, according to Goulden and Riddock, are due, not to the passage of blood from the subarachnoid space of the optic sheath, but to pressure on the central vein of the retina where it crosses the optic sheath, hence the venous congestion of the retina (Fig. 81).

The third nerve is most frequently affected, after that the fourth and sixth and the first division of the trigeminal. Aneurysms situated at the posterior part of the base of the brain may produce

Weber's syndrome or the Millard-Guber syndrome. Rupture of an aneurysm may prove rapidly fatal, although in most cases the patient lives for a few days, while some appear to make a complete recovery, but ultimately the prognosis is bad. Many die from subsequent leakage.

The treatment of subarachnoid aneurysmal hæmorrhage is well described by Birch and includes treatment before rupture, immediately after and during the stage of recovery.

The prognosis however in all cases is serious. According to Ohler and Hurwitz in a group of twenty-four cases of subarachnoid hæmorrhage, of all causes there was a 50 per cent. mortality.

Intracranial Hæmorrhage in Infancy and Childhood.—It is not uncommon for intracranial hæmorrhage to take place in young children. Sheldon has published a paper on fifty cases where the diseases suffered by such children are mentioned, *e.g.*, intracranial trauma during birth, congenital syphilis, chronic nephritis, anæmia, blows on the head, whooping cough, septic meningitis, etc. He says "the commonest situation for an intracranial hæmorrhage in the adult is in the substance of the brain whereas in children the hæmorrhage results from thrombosis of meningeal vessels. The hæmorrhage from birth injuries are usually extra-cerebral." It is stated by McCullagh that death in the new-born may occur from tentorial tears produced by pulling the occiput too far forward against the pubic arch and so injuring the tentorium. In the majority of the cases quoted by Sheldon the intracranial hæmorrhage was not diagnosed before death, but in some cases where days and weeks intervened convulsions were present (probably these occurred at the time of the hæmorrhage), also facial paralysis and rigid limbs, while examination of the fundus oculi showed swelling of the optic disc.

Pulsating Exophthalmos

Since 1809, 588 cases of pulsating exophthalmos have been recorded (Locke), and of these 23 per cent. were spontaneous, while 77 per cent. were traumatic in origin. Of the latter 94 per cent. had arterio-venous communication. Witham states that there is a latent period after the date of the reception of the injury before the cardinal signs of proptosis, pulsation, thrill, objective and subjective noises and vertigo are fully established. The condition, which is usually uniocular (Fig. 135), may be brought about by the formation of a communication between the internal carotid artery and the

cavernous sinus (angioma arteriale); on the other hand the cause may be an aneurysm of the internal carotid or the ophthalmic artery. Aneurysm is more likely to be the condition found in elderly people, but when the exophthalmos is marked the condition is more likely to be an arterio-venous aneurysm (Locke).

Pulsating exophthalmos must be differentiated from the proptosis of Graves' disease, orbital cellulitis, ethmoidal mucocele and orbital tumour. It must also be differentiated from intermittent exophthalmos due to hyperplasia of the jugular vein and its tributaries (angioma venosum).

Vision may not be affected, but on the other hand there may be dilated pulsating veins seen in the fundus, together with papill-œdema or even primary optic atrophy. The third and sixth cranial nerves may become involved or pain may be produced by the stretching of the fifth cranial nerve, or trophic changes of the cornea may follow the disturbance of the fifth nerve (see Chapter XV).

Whitham says that the ligation of the internal carotid on the same side offers more certain results than any other operation, especially if the collateral circulation is previously prepared by daily compression of the carotid artery for several weeks. Over 50 per cent. are cured and the mortality of operation is 10 per cent.

Thrombosis of the Cavernous Sinus.—If we consider the anatomy of the cavernous sinus and its communicating venous channels, we see that thrombosis of the cavernous sinus may be due to extension from many sources. The causes are (*a*) infective, either locally, such as orbital cellulitis, erysipelas of the face or a metastasis in infectious diseases and septic conditions; (*b*) thrombosis, possibly traumatic; and (*c*) marasmic (Shore).

As the symptoms of orbital cellulitis and cavernous thrombosis are somewhat alike, it is important to remember that the former are very rarely bilateral, while in half the number of recorded cases of the latter symptoms are referable to both sides. If in addition to symptoms of orbital cellulitis there is œdema in the mastoid region behind the ear, the diagnosis of cavernous sinus thrombosis is certain. The ophthalmic changes are due to obstruction of the venous circulation and to involvement of the various nerves which pass through the sinus. There are proptosis, beginning on one side and later spreading to the other side, chemosed conjunctiva with swollen lids, one or more ocular muscles paralysed, and in the later stages immobility of the eyeball. The cornea also becomes anæsthetic and

the pupil dilated. Rarely is marked papillœdema seen, but often dilated veins with œdema of the retina is found. In the last case which the author saw, it was impossible to examine the fundi owing to the swollen eyelids. Infection had taken place from a small abscess resulting from epilation of the hairs of the eyebrow four or five days before death. The previous week, however, an abortion had been brought about by an unqualified person.

In septic conditions of the face, such as carbuncle in the region of the mouth, there is great danger of thrombo-phlebitis of the cavernous sinus taking place. Hamilton Bailey suggests that almost immediately the angular vein should be ligatured just below the inner canthus of the eye and the vein divided between sutures; if thrombosis of the cavernous sinus has actually commenced he says we should at once carry out Eagleton's combined operation, which consists first of the ligature of the common carotid and, secondly, the removal of the eyeball. If the ophthalmic vein is filled with purulent clot, then by enlarging the sphenoidal fissure the cavernous sinus itself may be drained.

The general symptoms accompanying thrombosis of the cavernous sinus are profound. They are septic temperature, rigors, vomiting and severe cerebral symptoms. The accepted mortality is almost 100 per cent.

Thrombosis of the Superior Longitudinal and Lateral Sinuses.—Papillœdema may be found in these conditions and in both strabismus may occur. There is marked supraorbital neuralgia. A case of bilateral paralysis of the abducent nerves has been described by S. D. Greenfield. The paralsyis was due to acute purulent thrombophlebitis of the jugular vein and of the lateral sinus. In an article in which he describes most clearly the anatomy of the cranial sinuses, he draws attention to the proximity of the origin of the abducent nerves and says that it is due to this close association that bilateral involvement is so often found in fracture of the base of the skull and in basilar meningitis. Eagleton is of the opinion that bilateral involvement of the abducent nerves is due to venous stasis and is not the result of acute inflammation at the site of the nerve trunks. Walsh also refers to such paralyses.

WORD-BLINDNESS

What is *word-blindness?* Write on a piece of paper "Sit down," and place it before the patient standing at your desk. His visual

apparatus is intact, yet he cannot understand the written command, and so what is written conveys no meaning to him. He remains standing. He is *word-blind*.

Many years ago Gall, the founder of phrenology, conceived the idea that the cerebral hemispheres must be regarded as the material basis of consciousness. Broca, in 1861, stated that aphasia, that is, loss of power of speech, when it occurred in right-handed people, was always associated with a lesion of the third frontal convolution of the left hemisphere. This became known as Broca's area. Ferrier, in 1876, carried out and confirmed Fritsch and Hitzig's experiments which demonstrated that the cortex of the brain was excitable and that certain areas on excitation produced definite movements. Thus a motor area was defined situated in front of the fissure of Rolando, stimulation of which from above downwards was followed by movements of the leg, body, arm and face. Stimulation of that part of the cerebral cortex on the left side governing eye movements was followed by deviation of the eyes to the right. Thus gradually the areas of the cortex were discovered and described as the seat of strictly limited " centres " of preordained function. Experiments performed by Sherrington and Layton, published in 1917, helped to confirm what many others had already thought concerning the subject of the mechanisms whereby co-ordination of the various centres brought about the performance of speech, reading and writing. Marie already had said that the idea of separate centres for the function of speech could no longer be upheld although he considered the zone of Wernicke to be the seat of the intellectual processes involved, not because they are preformed centres for language but brain structures adapted by training to that function. This zone comprised the supramarginal convolution, the angular gyrus and the base of the first and second temporal convolutions (Fig. 83). After the ablation of the cortical area concerned in movements, say, of the hand, and when the neural shock has passed off, the animal still possesses the power of relearning an action which has been abolished by the anatomical destruction. Head, writing of speech and cerebral localisation, says: " The loss of function is manifested in terms of the process itself and does not reveal the elements out of which it has been built up. Moreover, the various mental activities interrupted during acts of symbolic formulation and expression can be exercised in a normal manner during some other behaviour."

We see, therefore, that local ablation or the presence of disease will produce no loss of function provided there is an alternative path whereby another portion of the cortex can be called into activity. Only if a secondary path is not available can the re-education of function become impossible. The latter is the case of the visual function. There is no alternative path when destruction has inter-

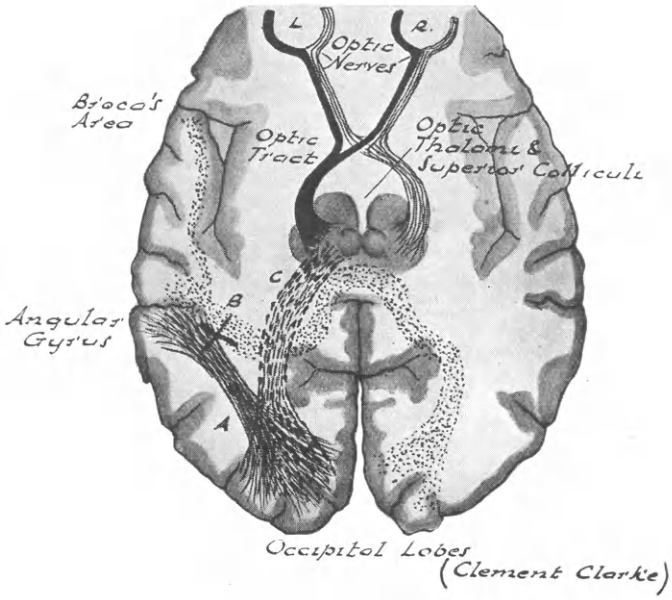


FIG. 82.—Diagram adapted from the drawing by Déjérine showing the position X of the lesion causing word blindness.

C, optic radiations passing from the external geniculate body to the occipital lobe ; *A*, fibres connecting the occipital lobe with the angular gyrus ; *B*, fibres passing from the right occipital lobe to the left angular gyrus (dotted band).

fered with the visual fibres in the occipital lobe. In Déjerine's diagram (Fig. 82) showing the position of the lesion causing pure word-blindness the various connections of the optic radiations are shown. Some fibres pass upwards and outwards to the angular gyrus, others proceed from the left thalamic region to the left calcarine fissure, while others are seen passing from the right calcarine fissure to the left angular gyrus. There are fibres which bifurcate in the ventral part of the radiations (medullary optic lamina) near the peduncle of the cuneus, which run upwards towards the splenium

forming an optic commissure and may, Pfeifer states, be associated with a bilateral representation of the macula.

A lesion where marked X in Fig. 82 results in the condition known as word-blindness, in which awareness of objectivity and recognition of the concrete meaning of objects are both retained but in which the abstract or associative meaning of the printed word is lost. Destruction of both occipital poles is necessary to destroy the respective functions, but a unilateral lesion such as X is sufficient, but only when it occurs in the dominant or lead hemisphere—the left hemisphere in right-handed persons or *vice versâ*.

Acquired Word-Blindness.—In 1877 Kussmaul showed that a condition existed in which clinically a patient showed complete text-blindness although the power of sight, the intellect and the power of speech were intact. For such a condition in which the patient, although not blind, is unable to read visible words he suggested the term *word-blindness*. In 1904 a case of word-blindness with right homonymous hemianopia was described by Hinshelwood, MacPhail and Ferguson. Briefly the facts were as follows : A teacher of French and German, aged fifty-eight years, suddenly discovered one morning he could not read a French exercise which had been submitted to him. He then to his astonishment found he could not read any printed book. On examination his visual acuity was normal. He could read figures on the same scale as Jaeger No. 1, but a right homonymous hemianopia was found to be present. He spoke fluently, he could write easily but could not read what he himself had written. Death occurred nine years later. During this interval he still could count up to millions, could write to dictation, but could not read a single word of what he had written. Autopsy revealed the presence of an enormously dilated lateral ventricle extending backwards and downwards to the tip of the occipital lobe. The lower half of the left occipital lobe was replaced by the dilated ventricle. Broca's convolution and the left angular gyrus were intact. The interruption there had affected the optic radiations of the left occipital lobe and their extension upwards to the cerebral cortex. This, as Hinshelwood remarks, gives strong support to Henschen's theories of the position of the cortical centres of vision. Hinshelwood, quoting Déjerine and Sérieux, described their two interesting classes of cases of word-blindness. The first class comprises the cases where the patient was word-blind and agraphic (unable to write). In the second class

the patient was word-blind and could still write to dictation although he could not read a single word he had written. Déjerine and Sérieux contended that before a person can write he must have acquired the power of reviving visual memories of words and letters. Destruction of the cortex of the angular gyrus will render it impossible to revive these images, hence writing is impossible. In the second class (that is, those cases which are word-blind but can write) there is a lesion in the region of the fibres of Gratiolet (see Fig. 41) which interrupts the path of such fibres as pass to the angular cortex.

There are many varieties of word-blindness ; some cannot read musical notes (amusia) and this may be an isolated symptom of the condition present. One case was described where the notes of music could be read and interpreted but the accompanying words were unrecognised. Another retained the power of reading Greek but was partially word-blind for English, French and Latin. In all these groups of visual memories it seems clear that there is complete functional independence. The visual memory centres in the left angular gyrus are connected with the primary visual centres in both occipital lobes. Hence destruction of the left occipital lobe is followed by right homonymous hemianopia but not word-blindness, since the right occipital lobe is still connected with the visual memory centre in the left angular gyrus. As Hinshelwood says : " To have complete word-blindness this centre (left angular gyrus) must itself be destroyed or its connections with both right and left occipital lobes must be cut off." An interesting case of transitory word-blindness associated with right homonymous hemianopia has been described by Gjessing, but unfortunately in this case an autopsy was not performed.

Congenital Word-blindness.—It was through the discovery of acquired word-blindness that congenital word-blindness came to be recognised. The first reference in medical literature to this condition was by Pringle Morgan in 1896. The case referred to was that of a boy of fourteen years who, normally intelligent in other ways, experienced the greatest difficulty in learning to read. After seven years of school and tutors he could only spell with difficulty words of one syllable. Although the boy could read figures fluently, yet he could not read letters. Only by spelling aloud could he recognise letters. According to Thomas, congenital word-blindness cases are of very common occurrence. According to Helene Frank, cases of

reading difficulty, which is sometimes termed word-blindness, are met in every school and in every rank of society. She found backward readers were children from seven to eleven and a half years of age, most of whom were of average intelligence. The children at infant schools were four to seven years of age. One of the cases described by Hinshelwood could rarely read by sight more than two or three consecutive words ; "he could proceed only by spelling aloud the word or spelling it silently with his lips—*glosso-kinesthetic memory of Bastian.*" The memory for sounds is developed before that for written words. Individual words are first stored and then by reading aloud letter by letter the meaning of short words is gradually comprehended. The auditory centre is called upon to aid the visual centre to function regarding the comprehension of words. In the *British Journal of Ophthalmology* for February, 1936, p. 73, a most interesting account is given by a lady of her experiences as a sufferer from word-blindness. Her father and his only brother were stammerers. To this lady the book was a blank and conveyed nothing to her. She was neglected by her English teacher, punished by a German governess and bullied by her mother to read, but all was useless. Years later she served on several committees in London during her married life, yet only at this time could she read the short lines of *The Times* very slowly. Although fond of choir music, the words had to be learnt at home by heart. Her eldest brother said : " Considering how slow she was in reading and backward, it is wonderful how clever she is now and that she always had originality and efficiency and did her job specially well."

Congenital word-blind children can learn to write by copying but not to dictation. The graphic motor centre is not stimulated directly from the auditory, but the intervention of the visual memory centre is necessary whereby the visual images of words are revived. If the visual memory centre has not acquired a store of images, then stimulation of the graphic centres is impossible.

Orton describes three distinct types of visual cortical mechanisms. The first is the " visual perceptive," the arrival platform for visual stimuli, but none have as yet passed to consciousness. The second is the " visual recognitive," destruction of which produces mind-blindness. Munk's dogs could get about without colliding, but to them the whip no longer had terrors, nor did the sight of food evoke any response. The third type—the " visual associative "—is the great posterior associative zone where " associative interlinking of

the data garnered in various sensory fields probably occurs." Lesions at this level result in word-blindness.

The term " strephosymbolia " instead of congenital word-blindness has been suggested by Orton as a descriptive name for the whole group of children who show unusual difficulty in learning to read.

The term " pure word-blindness " has been given several meanings. To such cases as congenital word-blindness without any visual defect (such as hemianopia) Hinshelwood applies this term. He uses as criteria for diagnosis three degrees : (1) Pure word-blindness, noting the gravity of the defect and the purity of the symptoms, that is the inability to read in an otherwise healthy child. (2) Congenital dyslexia (a term first introduced by Berlin), in which symptoms are not so grave as the first. (3) Congenital alexia, a term reserved for those cases where the defect in the visual memory centre is only part of a general cerebral deterioration. Dana, speaking of " pure alexia," says it is always accompanied by hemianopia, while on the other hand Wallin says : " We find no evidence whatever for any such distinction between word-blindness and dyslexia on the one hand and alexia on the other. We have discovered no qualitative difference between word-blindness in a moron, in a border-liner and in a normal," and " The highest grades of feeble-minded (the so-called low-grade morons) are subject to the same reading disability as pupils who are merely backward or normal in intelligence. Like the latter they do distinctly poorer work in reading and spelling than in many other school subjects."

Recognising the occurrence and importance of dyslexia and also the difficulties of diagnosis to which it gives rise, Ronne has devised a test which within the time at the physicians's disposal in his consulting rooms permits of comparing the reading abilities of the individual child with the average reading ability corresponding to its age. The use of the test is as follows : The child is requested to read the exercise aloud. During this procedure the time in seconds required for reading it, and the number and type of reading mistakes are recorded on a special copy. The test consists of four reading exercises presenting the greatest possible variety of difficulties. The principle of these tests is the measurement of the reading power of the children without consideration of their comprehension of the reading matter.

The following is an account of an interesting case. A young

man, eighteen years of age, was seen by the author. A warning was given beforehand not on any account to ask the patient to read. Previously he had been described as somewhat mentally defective and requiring careful handling. The young man nervously entered the room. He was well built, had a good face which apart from a marked inward squint betokened normal intelligence. Using the object chart (animals and toys) the examination of the visual acuity proved the vision to be 6/5 in each eye. He had normal peripheral fields and his refraction showed lenses were not necessary. He had an alternating converging strabismus of 45 degrees. When asked to name the objects on the chart he did so without hesitation. By this time confidence had been established so the reading test types were produced, and now it was clearly revealed that the case was one of pure word-blindness. By the glosso-kinesthetic method he laboriously spelled letter after letter and then hesitatingly pronounced the word such as " tribe " or " feel." This young man could discuss politics, history and geography in a normal manner but he could not read either letters or figures except in the manner just described. It was evident therefore that such a combination as inability to read or do " sums " and a repulsive squint plus an absolute inferiority complex (he had normal brothers) had given the impression of mental deficiency to all his friends. To this young man the " look and say " method was utterly useless. He said he had to spell each letter individually at all times before he could recognise the word, no matter how small. The treatment of this patient will be to correct the squint as soon as possible so as to remove the consciousness that he is not normal in appearance. Work will be suggested which will not entail reading and which will provide scope for an otherwise normal brain and body.

Goldstein mentions a case of visual agnosia, the result of a cerebral lesion, who learned to read by the kinesthetic experiences gained through tracing the letters with movements of the eyes.

Treatment.—There appears to be two schools of thought concerning the treatment of the word-blind. Again I quote the one outstanding classic. Hinshelwood advocates the use of the most elementary primers, beginning by learning the letters of the alphabet by speaking them aloud. Most of the cases quoted in this work had acquired after considerable effort the visual memories of the letters of the alphabet. The short reading lessons are carefully graded from day to day so as never to produce anything approaching cere-

bral exhaustion. One Sunday the author's youngest child, aged five years, was heard stumbling over the first few letters of the alphabet. A small award was offered if he could say all the letters correctly by Friday. But on Tuesday morning he repeated the alphabet from beginning to end without a mistake. Yet some parents of the cases quoted by Hinshelwood have patiently persisted for nine months teaching their children to repeat these twenty-six letters. Special classes and special teachers are required to work with and teach these unfortunate children. Many have advocated the " look and say " method—that is, to recognise printed words as a whole, as a series of pictures like Chinese or Egyptian hieroglyphics. The word " dog " to be taken as a whole and not as three separate letters d-o-g. Orton thinks this method exhibits logical absurdity, for reading whole words, he says, is a later acquisition than reading letter by letter. On the other hand, Duguid says that on the whole the " look and say " method has proved to be the most successful in his cases. It is clear according to Thomas that the best system of teaching reading to the word-blind is the education in pronunciation of the phonic system, the auditory spelling of the ordinary analytic system and the fixing of all words by the " look and say " methods. " All have a part in the ideal system."

MIRROR WRITING AND LEFT-HANDEDNESS

The examination of children in ordinary elementary schools, physical defective and mental defective schools has brought to light that a certain proportion, relatively 0·48, 1·1 and 8 per cent. (Gordon), are mirror writers. By mirror writing we mean that variety of script which runs in an opposite direction to the normal, individual letters being reversed. In Lucy Fildes' study of mirror writing she has noticed, just as in word-blindness, that the defect may vary in magnitude from an occasional reversal of single letters to complete reversal of all letters and entire words. When children are brought to the oculist the parents will occasionally vouchsafe the information that the boy or girl when learning to write persisted in writing backwards, but eventually this was given up and writing was performed in the usual manner. The earliest reference in literature to mirror writing, so Critchley states, was made by Rosinus Lentilius in 1698. In his " Miscellanea medico-practica Tripartita " he mentions a left-handed epileptic girl who used to write with her left hand " inveris literis," which could be read only

when held before a mirror. Cases have been recorded where as a result of a lesion in the neighbourhood of the left Sylvian fissure, alexia, anarthria (difficulty in articulating words) and left-handed mirror writing have followed. Brain has known of a case of injury to the occipital region which resulted in mirror writing with the left hand. In many of the earliest writings letters were found to be reversed and in some a line written to the right was followed by a line written to the left. This back and forth writing was likened to an ox ploughing a field and was therefore called "bustrophedon" or "ox-turns."

There is no definite theory established as to the pathogenesis of mirror writing. The dextral orientation of letters of various shapes became more fixed as the development of language took place, so that, as Orton states, orientation of letters and direction of writing are intimately connected. "But the bustrophedon would indicate that exclusively dextral orientation is by no means determined by brain capacities, but has been established on a purely arbitrary limitation fixed by custom and education." If, as Orton suggests, the visual image impinging on one occipital lobe is the mirrored opposite of the image in the other hemisphere, then it is possible that there is actually one writing centre in each hemisphere—the right centre being latent in right-handed people but is called forth into use in left-handed people. Yet in some cases under the influence of hypnosis or hysteria mirror writing has appeared, which means that there is a disturbance of the constant suppression governing the activities of the right hemisphere.

If the question is asked "Can the mirror writer read his own writing?" the answer is that it entirely depends on the person and the condition of the central nervous system. Leonardo da Vinci apparently could read with ease his own diary composed of mirror writing, but those cases where a right-sided hemiplegia has developed and is followed by mirror writing with the left hand, find the greatest difficulty in thus reading.

It is interesting to learn that left-handed children are found more commonly in mental defective schools; Gordon gives the figures in ordinary schools 7·3 per cent., and in mental defective schools 18·2 per cent. Ambidexterity is also found much more commonly among mental defectives. Among normal children the left-handed are frequently the most efficient and capable; among the mental defectives it is exactly the reverse.

It has been observed by Ballard, Fildes and others that in the

attempt to change a left-handed child into a right-handed one stammering has supervened, and also that stammering is more prevalent among left-handed children. This supports the suggestion by Gordon that something has occurred which has interfered with the proper functioning of the dominant hemisphere—usually

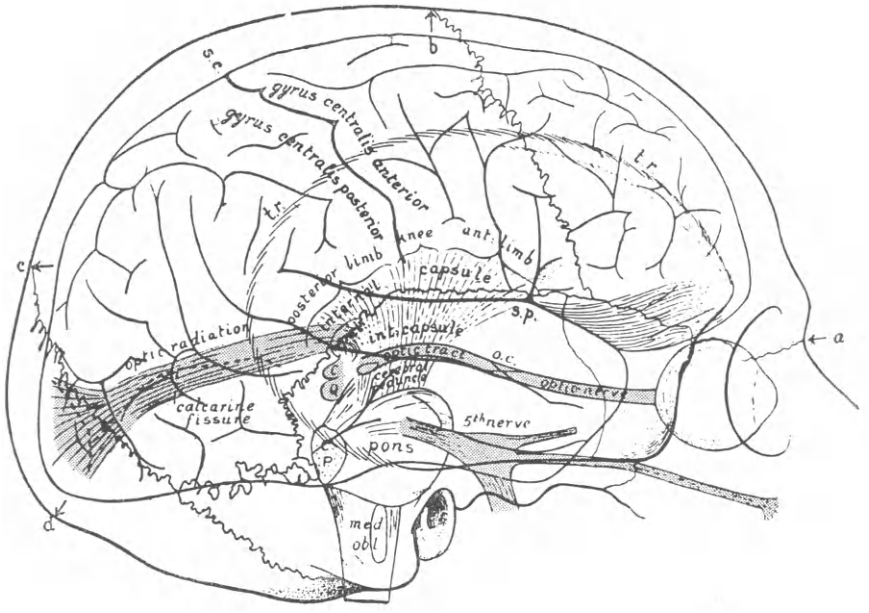


FIG. 83.—Right lateral aspect of the skull and cerebral hemisphere with orthognal projection of the medulla oblongata, pons, mid-brain, internal capsule, visual paths and fifth cranial nerve. The supramarginal gyrus surrounds the upturned end of the posterior extremity of the posterior ramus of the Sylvian fissure at the position marked by the lettering *t.r.* Immediately behind the supramarginal gyrus is the angular gyrus. (J. Symington from Quain by permission of Longmans, Green & Co.)

a, nasion ; *b*, bregma ; *c*. lambda ; *d*, inion ; *t.r.*, temporal ridge ; *s.c.*, sulcus centralis ; *s.p.*, Sylvian point.

the left—and will explain why there are so many left-handed children in mentally defective schools and why in the case of the left- and right-handed twins the left-handed twin and not the right-handed is found in these special schools.

A magnificent example of mirror-writing is illustrated in " Life," March 22nd, 1937, page 80. A nine-year-old schoolgirl is shown having just written on the blackboard. The illustration shows this writing and its reflection in a mirror.

MIND-BLINDNESS

Another variety of visual aphasia is mind-blindness. As in word-blindness two considerations must be borne in mind: the complex machinery necessary for "symbolic formulation and expression" may never have been fully developed or through some acquired lesion such machinery has developed a breakdown. We turn again to a great dissention. Broca, after an autopsy on a case which had suffered from a clinical condition of "aphasia," believed he had localised the seat of articulate speech in the third frontal convolution. Marie later denied the existence of the classical motor aphasia of Broca. "Cette zone ne constitue pas un centre préformé, mais seulement un centre adapté." From extensive studies Marie believed that articulate speech was destroyed by a lesion in a quadrilateral area which included the Island of Reil and the sub-adjacent structures, namely, the claustrum, internal and external capsule, the caudate and lenticular nuclei. Henschen and Déjerine agree with Broca and have accepted his doctrines as opposed to Marie, Niessl v. Mayendorf and many others.

Mind-blindness or optic agnosia is found in patients in whom the visual paths from the retina to the occipital cortex are intact, yet they cannot recognise common objects which they see. "They see men as trees walking." They have lost the power of recognising objects while the ability of seeing them remains. Kussmaul, who many years ago wrote a classical article, "Disturbances of Speech," introduced the term "apraxia," meaning the loss of the memory of the uses of things and the understanding of the sign by which things are expressed. This is more than mind-blindness; for instance, the patient may be unable to recall through the faculty of hearing the nature of objects. In the loss of power to imitate movements apraxia is seen. Mind-blindness is sometimes associated with a lesion of the parieto-occipital region, the left in right-handed people, and *vice versâ*. It is found in elderly people suffering from arterio-sclerosis.

To-day we must follow such men as Head, Lashley and Herrick who have tackled the problem of aphasia from a modern standpoint. They believe that language or thought cannot be localised. The cerebrum functions as a whole yet the destruction of a part interferes with the completed processes of the brain. Quoting Head, "These diverse defects of speech do not reveal the elements out of which the use of language is built up, on the contrary they show the way in which a highly complex series of acts can be dis-

turbed by lesions of certain portions of the brain." Grinker says the cerebral cortex is concerned in analysis of many various afferent systems and their regrouping in a wide range of efferent pathways. Diseases such as tuberculosis by means of circulating toxin may affect the cerebrum and may exhibit themselves in an interference with speech. In such cases Kollants has described the relationship between inward speech and mistakes in writing.

It is difficult therefore to locate the area or areas, lesions of which are responsible for mind-blindness. In Déjerine's article on " Différentes Variétés de Cécité Verbale," he quotes a case of mind-blindness which was examined by Dr. Landolt in 1887. This case not only showed mind-blindness but a hemianopia and a hemichromatopsia of the right side. Niessl v. Mayendorf, writing on mind-blindness (Seelenblindheit), says there is both a psychological sense and a pathological-anatomical mechanism underlying this defect. In true mind-blindness there must be no defect in the optical system and the patient must be of normal mentality. He says the agnosia may be limited to the inability to understand objects perceived with central vision (macular agnosia) or the surroundings seen with the retinal periphery (peripheral agnosia). " In examining mind-blindness as regards its psychical nature we must proceed to find out whether we have before us a defect of the content of the conscious or of its activity. We must clear up the question of whether we cannot recognise the normally perceived object because we cannot excite the optical memory picture of it from the periphery, or because we have lost it or whether our optical agnosia is the consequence of a mental lesion through which we are unable to carry through the intellectual process of identification, because we cannot concentrate our attention on the perception, because we are not able to retain long enough the impression perceived to compare it with the corresponding memory picture, or because we have lost the psychical ability to add from perceptions and memories other sensory fields to our cosmic picture."

" These psychological considerations which Kuelpe in the year 1912 set up against the theory of the clinician Wilbrand, Wernicke, Nothagel and others (the theory which traced mind-blindness entirely back to a loss of or inability to call up optical memory pictures) are unfounded and untenable, as we must postulate a loss of the above-mentioned categories of the activity of the conscious only for the optical mental field, and because above all the normal power of

thought, the activity of the conscious, is powerless when the content of the conscious (which is constituted from the optical memory pictures) is no longer at its disposal. The activity of the conscious, however, is an activity of the whole cerebrum which is only 'snuffed out' in a particular region of the mind when the requisite mental material is lacking. This loss then is the primary one, that of the power of identification the secondary one. For this reason we must regard mind-blindness as a mnestic disturbance. The different types of weakness of memory from what they have in common may be regarded as a loss of power to use effectively, if unconsciously, the material in the mind, if indeed this function with its material vehicle has not for ever been lost."

Niessl v. Mayendorf states that a case of mind-blindness has never been observed in isolated disease of the occipital cortical convexity, there must be an interruption in the optic radiations at the same time. The singleness of binocular vision, according to Sherrington, is the product of a synthesis that works with already elaborated sensations contemporaneously proceeding. It is at the level of the recognitive zone (Orton) that this fusion probably takes place and that mind-blindness occurs only when there are bilateral lesions of this zone. This is stated more clearly by Niessl v. Mayendorf, who bases his theory on three facts : (1) On the bihemispherical presence of the macula lutea ; (2) on the central projection of the retina in the calcarine cortex and thus on a limited localisation—also the point of sharpest sight in the occipital pole ; (3) on the existence of a macular bundle of the optic radiations arising from the outer corpus geniculatum, descending obliquely in a large bend through the parietal lobe which connects the latter with the occipital pole. He believes mind-blindness is the clinical symptom of an interruption of the left macular bundle of the optic radiations or left central projection of the macula in the cortex of the occipital pole. He also points out that we cannot constantly prove the presence of a central scotoma in mind blindness as the right brain macula takes up the optic impressions of both forvæ. *The image perceived may thus remain perfectly normal.* It is, however, at the same time, cut off from all associations which generally occur in the left hemisphere in the organ so accustomed to receive them. From this arises a disturbance of association without the necessity of anatomical paths of association being cut off by a central disease.

Thus mind-blindness would appear as a purely functional dis-

turbance, orginally conditioned by an organic lesion, which can be partially restored by the right hemisphere.

Amnesic Colour-blindness.—This is a symptom which has always been accompanied by right homonymous hemianopia. The patient sees colours and can recognise them but he is unable to name each colour. This condition is most probably due to a lesion in the occipital lobe.

Visual hallucinations may occur in the blind field in cases of hemianopsia. They are not to be mistaken for the visual hallucinations of insanity and are probably due to irritation of the visual memory centre. The hallucinations due to a tumour of the temporal lobe consist of definite images of people, animals or definitely formed scenes. Those due to occipital lesions are simple, such as flashes of light, sparks, etc., of brief duration or persistent.

Albers-Schönberg's Disease. We may conclude this chapter with a brief reference to two comparatively rare diseases, Albers-Schönberg's disease and Morgagni's endocraniosis. Both diseases have many aspects in common. In Albers-Schönberg's disease the characteristic feature is extremely dense changes in the bones, sometimes called *marble bones*. There is a pronounced anæmia with enlargement of liver and spleen. A recent case has been described by Kretzmar and Roberts, whose radiological investigations showed the bones of the skull to be so dense that the X-rays employed in the usual routine technique could not penetrate them. They found excessive deposits of calcium in the skeleton.

Due to pressure on cranial nerves, optic atrophy and nystagmus are found, but in Kretzmar's case there was also exophthalmos with a wide bridge of the nose and a left-sided facial paralysis. The outline of the pituitary fossa was scarcely defined, due probaby to calcification within the fossa. Anæmia was not obvious in this case.

Morgagni's Intracranial Hyperostosis is a definite lesion on the internal surface of the cranial bones more frequently found at the frontal and basal regions, consisting of new formation of bone tissue forming patches or bands which protrude from the cranial bones, causing compression or irritation of the brain. These patients suffer from intractible frontal headaches, apathy, asthenic and psychic depression. Cranial nerves are affected so that progressive blindness, loss of smell or deafness may follow (Pende). Owing to disturbance at the base of the brain, diabetes insipidus, irritability, disorders of sleep, psychic changes and even dementia may follow.

CHAPTER XI

TUMOURS OF THE OPTIC NERVE

It is commonly stated that primary tumours of the optic nerve are rare, yet one must admit that the appearance of unilateral exophthalmos which suggests such a condition is not at all infrequent, being seen both in ophthalmic and neurological practice.

Verhœff states that during the past sixty-four years about 300 cases of tumour of the optic nerve have been described in the literature. Leber in 1877 was the first to attempt a classification of tumours of the optic nerve. Those he called essential tumours which developed from within the dural sheath and he separated these from those which arose in the outer sheath. Wishart, of Edinburgh, in 1833 was the first, according to Byers, to describe clearly a tumour of the optic nerve. Byers himself in 1901 wrote an extensive article entitled " The Primary Intradural Tumours of the Optic Nerve." A full bibliography accompanies this paper. Parsons in 1903 write on " The Primary Extradural Tumours of the Optic Nerve," while in 1910 Hudson wrote on 154 cases, a paper which has indeed become classical and has been referred to by almost every writer on this subject since that time.

For the purpose of this chapter we will divide the optic nerve into its *intraocular*, *intraorbital* and *intracranial* portions. Tumours have extended along the optic nerve and have penetrated to within the globe. Coston's case of *intraocular* extension was observed by the ophthalmoscope. The disc, which had dilated capillary loops on its surface, was elevated to 6D with blurring of the edge of the disc. The pathological report on this case showed a true extension of the tumour, a dural endothelioma, along the nerve invading the disc. The nerve was atrophied. In describing the phakomatosis of Bourneville or tuberose sclerosis of the brain Van der Hoeve described a cystic mass situated over the optic disc, which discharged itself regularly into the vitreous. " So we proved that the presence of tumours in the retina and optic disc is a frequent sign in the syndrome tuberose sclerosis." The author has in his possession a

section showing the presence of a melanotic sarcoma situated around the optic disc. An interfascicular endothelioma of the choroid has been described by Mackay, which tumour produced a swelling of the disc, giving it a grey appearance. Small nodules were found on the optic nerve as far as 14 mm. from the eyeball. Extension from a tumour of the optic nerve and its appearance within the eyeball has been illustrated by Wolff, who says the invasion took place through the opening for a posterior ciliary artery and came to lie alongside the disc. This also could be observed by the ophthalmoscope. Endotheliomata, according to Hudson, show a pronounced tendency to invade the globe. Of 29 cases he mentions 8 which penetrated the eyeball, 4 of which involved the choroid, 3 affected the choroid and sclera, while the remaining tumours involved the nerve head, sclera and choroid.

The appearance of such a tumour within the eyeball must be carefully distinguished from tubercle of the nerve head, druses—crystalloid outgrowths from the lamina vitrea of the choroid—which sometimes spread over the disc, and hyaline bodies which cluster like sago grains over the disc surface elevating the nerve head 10 or 12D.

The so-called tumours of the optic nerve belong in the main to the *intraorbital* portion of the nerve. Tumours in this situation are usually classified as extradural or intradural, the former arising from the nerve sheaths being generally described as endotheliomata or meningiomata, while the latter are usually considered as arising from within the dural sheath, from the subarachnoid trabaculæ, pial sheath, septa or glial tissue of the nerve.

The intradural growths have been classified by Favalro, quoted by Oberling and Nordmann as fibroma, myxoma, myxofibroma, endothelioma, neuroma, glioma, neuroglioma, sarcoma, etc.

Probably a better conception of the tumours of the optic nerve might be gained as Hudson suggested by a subdivision of the whole of the available cases based not only on a critical examination of their records but more especially on the histological features. The majority of tumours, according to Hudson, should have as the most suitable designation degenerative gliomatosis, implying a generalised overgrowth of neuroglial tissue of infiltrative character dependent on some degenerative change in the tissue of unknown ætiology. Since these primary optic nerve tumours are composed essentially of neuroglia, invade the connective tissue stroma, penetrate the pia,

grow exuberantly in the subdural space and are histologically similar to many gliomas of the brain, Verhœff says in Penfield's Book : " These facts constitute sufficient grounds for regarding them as true gliomas." (Fig. 84). Verhœff has classified the tissue of these tumours into three main types. The first, the finely reticulated type, is similar to the neuroglia of a normal optic nerve. In the interspace of a delicate reticulated syncytium is a matrix of fine neuroglia fibrils embedded in which are small round or ovoid nuclei. The second type shows the cell reticulum to be much coarser with marked vacuolization of the cytoplasm. This type resembles that of myxoma. The third type of glioma is the spindle-cell or coarsely fibrilated type. The cytoid bodies found in these glial tumours which Verhœff describes are not peculiar to this type of tumour but are found in pituitary adenomas, chordomas, meningiomas, gummas, etc. (Kiehle). A series of beautiful histological photographs of a glioma of the optic nerve has been included in Kiehle's paper.

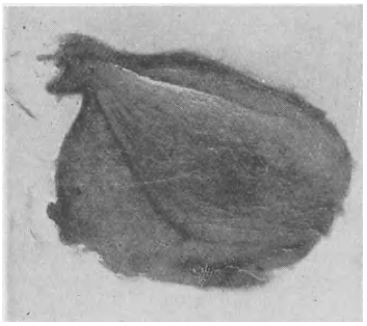


FIG. 84.—Glioma of optic nerve showing small pedicle of nerve (on left) between tumour and eyeball × 2. (Kindly lent by Montague Hine).

Lest confusion should arise between glioma of the optic nerve and glioma (retinoblastoma) of the retina, we should recall to mind the embryology of the central nervous system. In the very early embryonic stages the nervous system originates from a tube arising from the ectoderm. This is the medullary groove, the walls of which are formed of neurospongia while the inside of the tube is lined with an epithelial sheath, the neuro-epithelium. From the widened anterior end of the neural tube the cerebral vesicles are developed, anterior, middle and posterior. From the lateral walls of the anterior vesicle the two optic vesicles arise. The neuro-epithelial layer lines all the vesicles and in the optic vesicle becomes the pigmented sheet and the ten layers of the retina. From the neuro-epithelium originate those elements which are the potential functional centres of the nervous system, while from the neurospongia develop those cells which ultimately become the meninges, the

Schwann cells, the sheaths of Schwann which envelop the nerve fibres and the neuroglial cells which in their ramifications give support to the ganglion cells and to the nerve fibres. This brief description has been suggested to me by Weskamp, who points out that in speaking of glioma one must refer to the blastoma derived from the neuroglia, a tissue formed by astrocytes, obligodendrocytes and microgliacytes derived from the neurospongia. The retinoblastoma (glioma of the retina) originates in the retinal cells which are derived from the neuro-epithelium.

Two facts should be borne in mind regarding the malignancy of tumours. First, the more rapid the growth of a tumour the greater the malignancy. And, again, the malignancy of a tumour follows the fact well established by Bailey and Cushing that the earlier the cell type in the histogenic series the more malignant is its nature. *Glioma of the optic nerve* is benign, its growth is slow and is usually towards the cerebrum. Recurrence, according to Hudson, has not been recorded. *Glioma of the retina* has a most malignant evolution. It usually springs from the most posterior part of the retina, including the disc. The epithelial-like cells soon break through the tumour nodule and infiltrate the surrounding tissues spreading into the orbit, base of brain and even distant metastases arise. These retinoblastomas are the only known neoplasms of glial origin which metastasize to distant parts (Verhœff). Writing of gliomas of the optic nerve Grinker says that tumours of the optic nerve are probably all of the spongioblastic unipolar or bipolar type. And since the optic nerve arises from an evagination of the primary cerebral vesicle and contains all the adult forms of neuroglia, any or all of the gliomatous tumours described in the brain should be found arising from the optic nerve.

The second group of Hudson's cases consists of six cases of fibromatoses of the nerve sheath. In this type of tumour formation there is an enormous growth of fibrous tissue involving the outer sheath of the nerve but not the vaginal space nor nerve stem. Hence the explanation of several cases where exophthalmos preceded the loss of vision by a considerable extent of time.

The third group consists of endothelial tumours of the nerve sheath. These tumours resemble the endothelial tumours of the brain membranes; some authorities say they are one and the same and that the endothelioma begins primarily in the intracranial space and extends downwards into the orbit. But there is no reason to

doubt that such tumours can arise within the orbit for, as Coston remarks, the meningeal sheaths about the optic nerve do not differ from those about the brain. However, these tumours tend to grow forwards towards the globe and finally form a cup-shaped depression around it (see Fig. 85). They thus differ from the gliomas, where in many instances there has been a necklike pedicle of unthickened nerve intervening between the tumour and the globe as in Fig. 84.

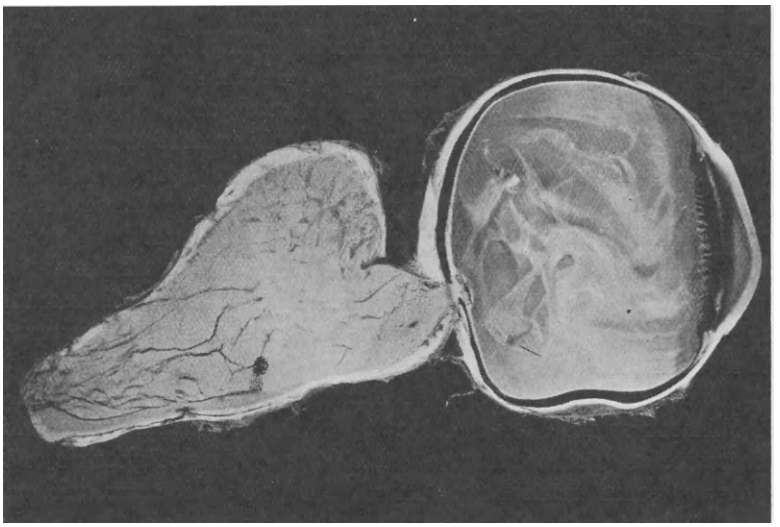


FIG. 85.—Endothelioma of optic nerve showing tendency to form cup-shaped depression around the globe. The elevation (papillœdema) of the optic nerve head is evident, while the nerve is seen running along the base of the tumour. (Section prepared by E. Wolff. Case under care of Montague Hine.)

In Parsons' description of the histopathology of this type of tumour he says "the arachnoid is very evident owing to its great infiltration and the proliferation of its cells," while "the dural sheath, apart from some infiltration, is normal in its inner layers"; peripherally it fades off insensibly into the tumour. Mallory states that the histological term endothelioma is fundamentally incorrect and that arachnoid fibroblastoma would be a better appelation as the endothelial cells are merely differentiated connective tissue cells arising from those cells covering the fine connective tissue and elastic fissure strands which form the arachnoid membrane. The microscopic structure of these endotheliomas is illustrated by the

description of such a tumour by Stallard. " The neoplasm probably arose from the arachnoid mater. It extended forwards as far as the most anterior limit of the arachnoid space where it infiltrated some of the posterior layers of the sclera, several small groups of cells splitting these and spreading between the lamellæ. Posteriorly it extended as far as the line of excision. There was no histological evidence of intra-ocular extension.

"The point of maximum distension of the dura was forwards in the vicinity of the posterior surface of the sclera. The neoplasm was composed of oval-shaped endothelial cells, such as are found covering the elastic and connective tissue trabeculæ of the arachnoid mater. These cells had a granular cytoplasm and a nucleus with a well-defined chromatin network and nucleoli. Clear clefts were evident among some of the cells, the nuclei lying adjacent to these. For the most part these cells had an irregular arrangement but some were disposed in definite groups around blood vessels, some in columns and others in whorls. The cell masses were broken up by vascularized connective tissue trabeculæ and collagenous fibrils. The neoplasm had infiltrated the deeper layers of the dura mater but had not perforated it. The pia mater was thickened and the optic nerve surrounded and sharply defined by several layers of newly formed condensed fibrous tissue and collagen fibres : it was infiltrated at several points by the cells of the neoplasm. There were areas of hyaline degeneration at some sites in the connective tissue trabeculæ and in the walls of blood vessels. A few areas of cell degeneration with lymphocytic and phagocytic infiltration were present. At some points there were attempts to form concentrically laminated psammoma bodies. The optic nerve was atrophic and hyaline degeneration was present in some parts of the supporting neuroglial tissue. The central vessels were very dilated and engorged with blood just behind the lamina cribrosa and the capillaries traversing the optic nerve were distended with blood. A moderate degree of perivascular lymphocytic infiltration was present around the central retinal vessels at the optic disc. The dural sheath was thickened and the vessels of the dura and posterior ciliary vessels were dilated and engorged with blood, particularly the veins."

Age Incidence.—In the first group, namely, the gliomatous tumours, the onset, as evidenced by exophthalmos in 45 per cent. of the cases, occurs between birth and the fifth year and 49 per cent. between the sixth and twentieth years of life. But Hudson recorded

one case only after the fiftieth year. These tumours therefore belong chiefly to the age of childhood and youth.

Of the six cases of fibromatoses of the nerve sheath recorded by Hudson four arose before ten years of age, the fifth at twenty-one and the sixth at thirty-one years of age. While in the fourteen cases of endotheliomata which have been recorded by Parsons, five occurred between birth and ten years of age, four between eleven

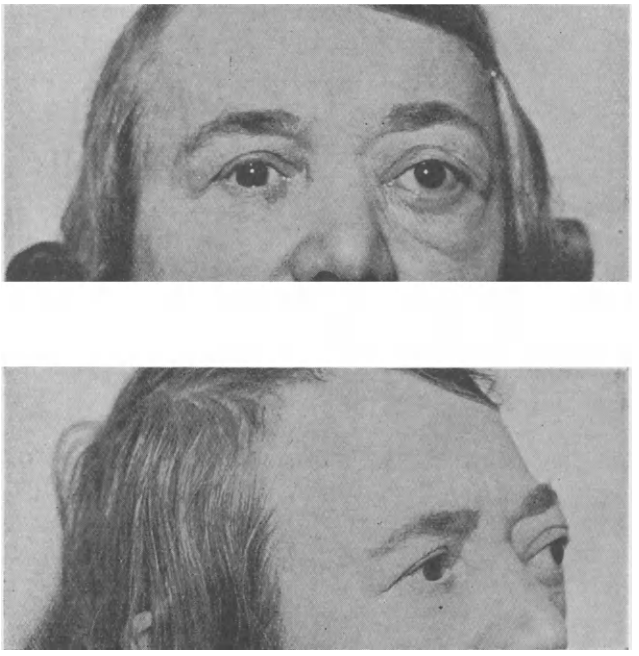


FIG. 86.—Exophthalmos due to tumour of left optic nerve. Four years' history. Left eye blind, optic disc showing grey atrophy but swollen to extent of 4 D. Vision of right eye is normal.

and twenty, then one for each decade until seventy years of age. Hudson, however, found 50 per cent. of his third group occurred after thirty years of age. Speaking generally of tumours of the optic nerve Byers has noticed the tendency for females to be affected more than males and the left nerve more frequently than the right. He, too, like so many others, has been struck by the part played apparently by traumatism and inflammation, not only local but general such as typhoid, in a large number of the cases recorded.

SYMPTOMATOLOGY

Tumours of the optic nerve usually grow slowly. Within a fortnight the author has examined two cases where in one (Fig. 85) the growth as evidenced by exophthalmos has taken just over a year to grow, the other, however (Fig. 86), has been present for four years. In the latter case the eye is still capable of somewhat limited movement in all directions. Hudson mentions two cases where considerable growth took place in six months. Wishart had observed his case, which apparently was a glioma, for over two years.

Unilateral Exophthalmos.—It is this symptom which in many cases has been the first to call the attention of the patient or his friends to something being amiss. In a few cases squint has preceded the exophthalmos, but in a large number both exophthalmos and loss of vision have occurred simultaneously. According to Parsons these two symptoms alone, unilateral and early exophthalmos with early amaurosis, are almost in themselves proof of the presence of the condition under consideration.

In addition to tumours of the optic nerve *unilateral exophthalmos* may result from many conditions.

(1) Congenital unilateral exophthalmos.
(2) Unilateral myopia of high degree.
(3) Early Graves' disease.
(4) Orbital periostitis.
(5) Orbital cellulitis.
(6) Sinusitis.
(7) Tumours of the orbit.
(8) Intracranial tumours in—
    (*a*) the anterior fossa above the roof of the orbit ;
    (*b*) the chiasmal region ;
    (*c*) the pituitary region ;
    (*d*) the middle fossa.
(9) Aneurysm of the ophthalmic artery and arterio-venous aneurysms of the orbit.
(10) Trauma.

No. 1 is illustrated by the case of a young girl under the author's care whose mother was the first to notice the exophthalmos, but repeated examinations over several years have not revealed any increase of exophthalmos nor disturbance of vision or muscle balance.

No. 2 is exemplified by the case of a child not yet two years of age who has 23 D of myopia in one eye, the other eye being normal.

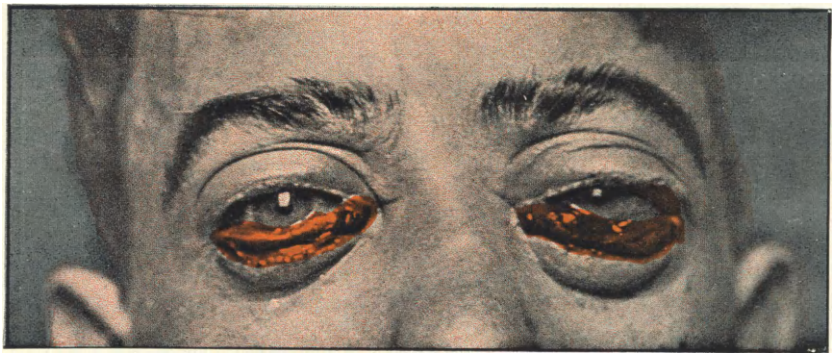
No 3. Cases are brought to the oculist with the complaint that one eye seems to be growing larger than its fellow. Often these turn out to be early cases of primary thyrotoxicosis or Graves' disease. The vision, however, is unaffected although some muscle imbalance is early observed. (See Chapter XIII, p. 340.)

No. 4. Any form of *localised periostitis* of the orbit may result in protrusion of the eye. Plate XIV illustrates the type of exophthalmos resulting from syphilitic periostitis which in this case began in the left antrum and spread to the left orbit, then through the ethmoidal sinuses, finally reaching the right orbit. The infection was blood-borne and was spread by means of the anastomoses between the alveolar branches of the internal maxillary artery and the ethmoidal branches of the ophthalmic artery. Within seven days the left eye became proptosed until the lids could not meet over the cornea, while the sight of this eye diminished just as rapidly. The eye, of course, was immobile. Such a case could be included among those labelled pseudo-tumour of the orbit. (Williamson-Noble, Benedict and Knight.) With intensive anti-syphilitic treatment at the end of six months both eyes were in their normal place and full vision was restored. At the end of two years' treatment, consisting of arsenical intravenous injections, mercury and iodide, the Wasserman reaction of the blood became completely negative. The onset of this specific periostitis took place four years after infection, for which no adequate treatment had been given. (See " Affections of the Eye in General Practice.")

No. 5. Orbital cellulitis is indicated by unilateral exophthalmos, the eyeball is fixed and the lids and conjunctiva are swollen and chemosed. There is pain, together with raised temperature.

No. 6. When a *chronic infection* of the ethmoidal bone has taken place and the cell or cells have become swollen, a radiogram of the part taken in an antero-posterior direction will reveal the eggshell-like swollen ethmoidal sinus. Not infrequently in frontal or antral sinusitis there is slight proptosis with limitation of movement of the eyeball and perhaps diminished vision, but the localised pain over those areas and the absence of chemosed conjunctiva helped to decide the diagnosis. Exophthalmos due to sphenoidal sinusitis is more difficult to diagnose; as in the other forms of sinusitis the rhinologist's opinion must be sought. In connection with sinus

PLATE XIV (R. Lindsay Rea)



PSEUDO-TUMOUR OF THE ORBIT

Appearance when first seen. The eyes are protruding so that the lids will not meet. The conjunctivæ are swollen and chemosed ; the vision of the left eye nil.

To face p. 242.

disease it is worth while perusing the record of Jessop's two cases, also Cohen's paper on " Inflammatory Exophthalmos."

No. 7. *Tumours of the orbit* include dermoid cysts. These when intraorbital are usually situated above the inner canthus or more superficially at the outer canthus, where they must be diag-

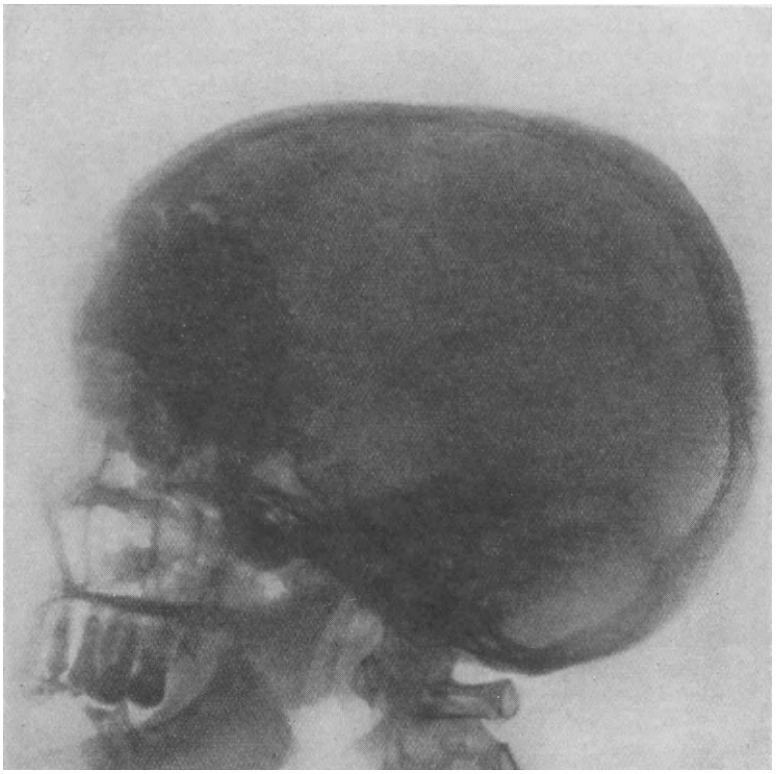


FIG. 87.—Skiagram of osteoma of right frontal region which produced proptosis of right eye. There was pain over this region following a blow three years previously. (T. R. Hill's case.)

nosed from meningo-encephaloceles. (Fisher.) Lediard described the case of a dermoid cyst which caused complete dislocation of the eyeball, while in Knapp's case the cyst extended from the conjunctiva back to the apex of the orbit. Hydatid cysts are much less common than dermoid cysts. Angiomata, osteomata (Fig. 87), and neuromata are found. Fig. 124 illustrates a case of von Recklinghausen's disease in which Rayner Batten removed the eyeball for

severe proptosis due to orbital growths. The swollen eyelid was left and continued to increase in size. Sarcoma and carcinoma occur, also their metastases from distant parts may be found in the orbit.

No. 8. *Intracranial tumours* situated in the anterior fossa may cause proptosis of one eye. Such a case is illustrated by Fig. 88 (*a*). When the case first came under the author's care the left eye was markedly proptosed, but in spite of the forward and downward position of the eye binocular vision was still present, and the vision

Fig. 88.—(*a*) Appearance of patient before operation. Marked left-sided proptosis. No diminution of vision. (*b*) Normal position of left eye after operation with normal movements.

of each eye was normal. The condition had lasted over eight years. Stereoscopic radiograms revealed an erosion of the roof of the orbit and a similar erosion of the frontal bone on the vault of the skull. Hugh Cairns removed a cholesteatoma, the size of a bantam's egg, through a fronto-parietal osteoplastic flap, and in so doing he exposed the orbital contents through the erosion of the roof of the orbit. I had taken the precaution to fill the conjunctival sac with boric ointment and sewed the lids together. At the conclusion of the operation the eyeball had fallen back into its place, but six days later, due to effusion of blood the proptosis was worse than ever, the stitches gave way, the cornea became anæsthetic and the lower

half opaque. The conjunctiva was enormously swollen but uninfected. I, therefore, under cocaine, snipped with scissors the conjunctiva allowing serum to flow freely and again filled the sac with soft 5 per cent. boric cream. The lids were strapped over the cornea with plaster. Cairns boldly opened the head wound, removed considerable clot, then cauterised some bleeding points which had not been in evidence previously. The result a few months after is shown by Fig. 88 (*b*). The eye is in its normal position, the cornea has regained its sensitivity, but a thin corneal scar is present below the centre of the pupil, so that vision when last taken was 6/18.

Dr. T. R. Hill has kindly permitted me to publish the radiogram, Fig. 87, of a case under his care. Three years previously the patient had sustained a blow on the head. Now pain was present over the forehead with proptosis of the right eye. A large osteoma was revealed, which is well shown on the skiagram.

Tumours of the chiasma usually erode the optic foramen and push forward the orbital contents. Growths in this situation will be referred to presently. Also pituitary tumours expanding the sella turcica may be responsible for uni- or bilateral proptosis (see Fig. 103).

Tumours of the middle fossa may be the cause of proptosis. It was probably this type of tumour to which Sir A. Lawson referred when demonstrating a case of neoplasm of the temporal fossa with proptosis of the corresponding eye.

No. 9. *Vascular Derangements within the Orbit.*—Vascular derangements within the orbit may bring about not only proptosis but the form usually described as pulsating exophthalmos (see Fig. 135). One form results from a traumatic or spontaneous rupture of the internal carotid artery into the cavernous sinus. (Trotter.) Hæmangiomata of the orbit are usually cavernous (MacGillivray, Ewing), the proptosis resulting being influenced by posture, crying or other emotional disturbances. Aneurysm of the ophthalmic artery, although repeatedly referred to in literature, is really most uncommon according to Pfingst. "Search of the literature has failed to reveal a single case of ophthalmic aneurysm in which association of the lesion with disturbance of the larger intracranial vessels could definitely be excluded at post-mortem examination." However, it seems that in Foster Moore's case the aneurysm of the ophthalmic artery was not associated with any

other intracranial lesion. In addition to Pfingst's case another has been described by Sanford, Craig and Wagener. In the former case there was concentric contraction of the field of vision, while in the latter there was an inferior altitudinal anopsia present. In Pfingst's case the retinal veins were distended and tortuous and the disc was raised 3–4 D, while in Sanford's case the left disc was slightly pale. In Foster Moore's case he said, " The fundi are essentially normal." In those cases of pulsating exophthalmos pressure on the carotid diminishes the bruit heard over the orbit and also the feeling of thrill. Relief was given in Sanford's case by removing the roof of the optic canal allowing the optic nerve to bulge with considerable restoration of vision. The condition was bilateral.

No. 10. *Trauma.*—Hæmorrhage into the orbit from trauma or general causes, such as whooping cough, scurvy, hæmophilia and arterio-sclerosis may cause displacement of the eyeball forwards.

Fracture of the surrounding bony walls may result in proptosis. Air escaping from fractured ethmoidal sinuses may be detected by palpation for crepitation.

Foreign bodies lodged in or behind the orbit can result in exophthalmos. These were commonly seen during the Great War. (See Plate X, Chest Radiography, Rea.)

The direction of the displacement of the globe usually coincides with the direction of the orbital axis, but exceptions occur owing to uneven growth of the tumour. The chief exception is found in those cases where the displacement is forwards, downwards and outwards. There is resistance to backward movement of the eyeball, while palpation with the tips of the fingers around the globe may reveal a hard mass. The proptosis, too, is usually painless until pressure symptoms take place from the enlarging growth pressing on the ophthalmic branches of the trigeminal within the orbit. Malignant orbital growths are much more painful than the primary growths of the optic nerve. Variations of slight degree may take place in the proptosis of the eyeball. In addition to the growth there may be congestive changes. In Brailey's case changes took place at the menstrual period. A third factor in the cause of proptosis is mentioned by Byers, who observed that effusion into Tenon's capsule and the supravaginal lymph space of the optic nerve brought about a sudden increase of the exophthalmos.

Limitation of movement is less likely to occur in the case of glioma than in endothelioma or fibroma.

A rapid estimation of the degree of exophthalmos can be made by the observer standing behind the seated patient and looking down his forehead. The patient's head is slowly retracted until the upper lid of the proptosed eye is seen. On further retraction of the head the lid on the opposite side comes into view. If at this stage both upper lids are raised by the tips of the forefingers the corneæ are seen, one being more prominent than the other. At the best this is but a rough and ready measure.

To estimate accurately the degree of exophthalmos one can employ an instrument as illustrated by Fig. 89. This is Hamblin's

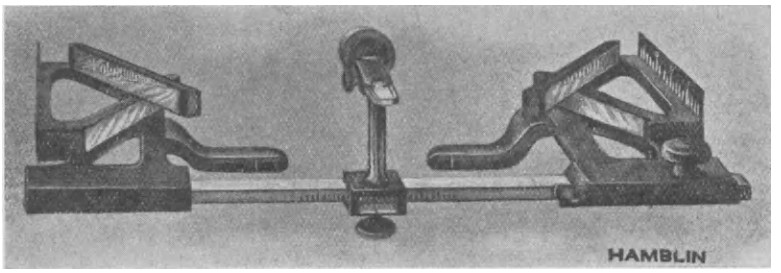


FIG. 89.—Hamblin's (improved) Hertel exophthalmometer. This instrument is designed on principles suggested by Professor Hertel, but improved to avoid the parallax of older instruments. It records with great accuracy any degree of exophthalmos. The principle is that of reflection of the corneal apices in reflection at right angles to the outer orbital margins. Two pairs of accurately set mirrors are made so as to be adjustable to the outer margins of the patient's orbital cavities. The observer is able to see in the one mirror of the pair a reflection of the vertical profile of the cornea, and in the other mirror of the pair the reflection of a scale apparently on the same plane. As the two images are coincident and apparently in contact, a reading may be taken (in millimetres) of the amount of the exophthalmos.

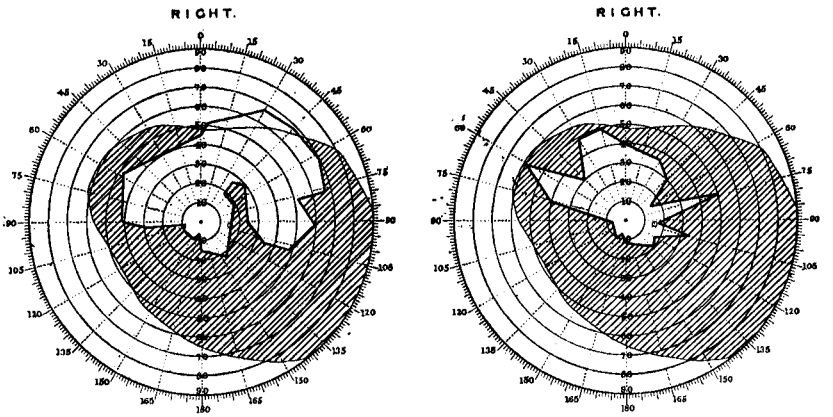
modification of Hertel's exophthalmometer. The observer looking into the mirrors can take an accurate reading in millimetres of the amount of proptosis.

Changes in Refraction.—Owing to the pressure of the tumour behind the globe, as in Fig. 85, the eyeball is flattened and therefore the eye becomes more hypermetropic. In the case just quoted there was an increase of 2 D of hypermetropia in the eye on the side of the tumour, while in Neame and Wolff's case the hypermetropia amounted to 7·5 D. In those cases where the exophthalmos is due to a tumour outside the orbit the whole of the orbital contents are pressed forward so that the globe is not deformed as in the case

illustrated by Fig. 88 (*a*), where the refraction remained constant for many years.

Secondary glaucoma was noticed in six of Byer's cases.

Loss of Vision. It has already been noted that proptosis accompanied by loss of vision is practically always pathognomonic of tumour of the optic nerve. Of Hudson's cases of glioma defect of vision was observed to precede exophthalmos in 14 cases. Defect of vision and exophthalmos were noted at the same time in 24 cases, while only in 9 was the proptosis noted first and on subsequent examination each eye was found to be blind. Therefore, "a



(*a*) (*b*)

FIG. 90.—(*a*) Field of vision result of endothelioma of optic nerve confined to orbit. (*b*) Field of same case five months later.

primary defect of vision with subsequent obvious tumour formation evidenced by exophthalmos is by far the commonest sequence." (Hudson.)

In both fibromatosis and endothelioma the exophthalmos generally preceded loss of vision, sometimes by a considerable time.

Visual Fields.—Of great importance in the diagnosis of tumours of the optic nerve and their situation is the state of the visual field. Through the kindness of Montague Hine I was able to obtain the field of vision of the case represented by Fig. 85. (Endothelioma.) On December 19th, 1935, the field of vision of the right eye is shown by Fig. 90 (*a*), but by May 7th, 1936, the field had contracted to what is seen in Fig. 90 (*b*). The central vision at this later date was 6/9, but by September 24th the patient could only count fingers

at one metre. The direct pupillary light reaction was absent, but the consensual reaction and reaction to accommodation were still present. The vision of the left eye was normal. By the way, the first symptom this patient noticed was the presence of halos around lights.

Tumours situated at the optic foramen or chiasma usually show hemianopic type of visual fields. In Mehney's case there was bitemporal hemianopia with islands of vision in the temporal field. At operation a glioma involving the intracranial portion of the optic nerve was discovered. So many cases of removal of intracranial tumour of the optic nerve and chiasm have ended fatally, usually by hyperpyrexia, that it is refreshing to read of Stammers' case, where removal of a tumour from within the right optic nerve in its intracranial portion was followed by recovery. The fields in this case showed complete anopsia on the right side, while there was general constriction on the left. In a case described by Grinker there was a right homonymous hemianopia with its margin through the fixed point, while the upper left quadrant showed a marked contraction for red. There was a distinct dilatation of the optic canal. A transfrontal craniotomy revealed a tumour of the chiasm involving both optic nerves and extending as far back as could be seen. Further interference could easily have led to the death of the patient from disturbance of the heat centres in the brain behind the chiasm.

While discussing fields of vision in connection with proptosis it has been mentioned that altitudinal changes are to be associated with pressure of an aneurysm. A superior hemianopia was described in such a case by Martel, François and Guillaume. In Pfingst's case of an anomalous ophthalmic artery the field of vision was more or less concentrically contracted similarly to what results from pressure by a tumour on the optic nerve before the intracranial portion has been affected by extension.

Optic Disc Changes.—In the early cases papillœdema is generally observed, but in those of long standing consecutive optic atrophy has appeared and the eye is usually blind. It is remarkable that in Hudson's collected cases the vision in the vast majority was almost or completely abolished. With all the advantages of modern methods of perimetry these cases should be observed long before vision has been severely affected.

It has been pointed out by Byers that in those reported cases

where ophthalmic changes were not visible pathological examination showed a full half-inch of normal nerve was present between the eyeball and the tumour. It seems, therefore, that the situation of the newly formed tumour is a factor in the varying ophthalmoscopic picture. If the tumour is situated at the central end of the optic nerve then most probably only a simple descending optic atrophy from pressure and nutritional changes would result. If a glioma extends forwards in the substance of the nerve the atrophy may precede the papillœdema and vision is lost early.

The author has a case at present under observation, the history of which is that for the past year the patient, a young man of twenty-seven years of age, has noticed some diminution of vision of the right eye. He was admitted to a hospital for nervous diseases and thoroughly examined. All neurological and serological tests were negative. Now at the end of a year there is (1) proptosis of the right eye to the extent of 3 mm. ; (2) unilateral papillœdema of 3 D ; (3) slowly contracting field of vision with considerable limitation of nasal field in the small isopters, with a colour scotoma ; (4) central vision reduced to 6/12 ; (5) movements of the eyeball are normal, and (6) the optic foramina are normal and equal in appearance. A tentative diagnosis has been made of a glioma of the optic nerve.

Defective Muscular Movements.—For a considerable time during the slow growth of the tumour within the cone of the extraocular muscles defective movement may not be observed in any direction. In thirty-five instances the movements of the globe were normal, while in forty-two cases there was marked impairment of eyeball movements in one or more directions. (Byers.) The pressure produced by the tumour on the muscles, the extension of the growth to between the globe and the insertion of the muscles, also congestive changes in the orbit are responsible for the defective muscular movement. The patient therefore may complain of diplopia immediately before, during or after the time of the appearance of the exophthalmos.

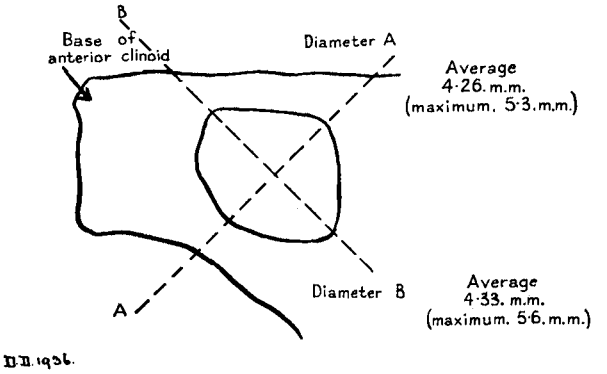
Palpation.—If the globe is markedly proptosed it may be possible to palpate the tumour. It may even be necessary to do this under a general anæsthetic, but when an exploratory palpation is to be done it is essential that the state of the Wassermann reaction of the blood be known. The only drawback in the cure of the case illustrated by Plate XIV is somewhat defective external movement

of the left eyeball due to the left external rectus having been divided in order that the presence of a tumour might be felt. Such a procedure should never have been done without knowing the state of the blood, which in this case showed a positive Wassermann reaction.

Corneal Sensation.—In many cases corneal sensibility is diminished or is found to be entirely absent, especially in severely proptosed cases. Inability to close the eyelids combined with corneal anæsthesia soon brings on some form of keratitis. If the optic nerve has been removed and the globe left *in situ* with care the cornea can be preserved for years, yet in some of the cases recorded the globe has been destroyed by suppurative keratitis.

Pupillary Movements.—These are normal except when complete blindness on one side is present. The pupil then only reacts consensually.

Enlargement of the Optic Canal.—While examining radiograms of the ethmoidal sinuses and surrounding areas Rhese, in 1911, frequently noticed that the shadow of the optic foramen appeared on the plate. Having pointed this out it quickly became apparent that such would be of considerable assistance in the diagnosis not only of intraorbital tumours extending towards the brain but of those intracranial growths which tend to grow into the orbit. Hartmann describes twenty-nine methods which have been devised for obtaining the best skiagrams of the optic canal. The aperture through which the optic nerve passes into the orbit has generally been described as the optic foramen. It is really a canal varying



Dimensions of normal foramen (R) (based on Goalwin).

FIG. 91.

from 4–9 mm. in length. (Whitnall.) Sometimes it is longer. Goalwin stated, " Its minimum cross section is at the posterior end," but Keyes from his study of 2,000 skulls, or as he prefers to put it, 4,000 optic foramina, came to the conclusion that the smallest cross section of the canal is not constant. It was not always at the posterior orifice that the canal was narrowest, neither was the shortest diameter at any constant point. Also, he stated that variations in the calibre of the foramen were so frequent that its diameters at any given point did not present a complete picture.

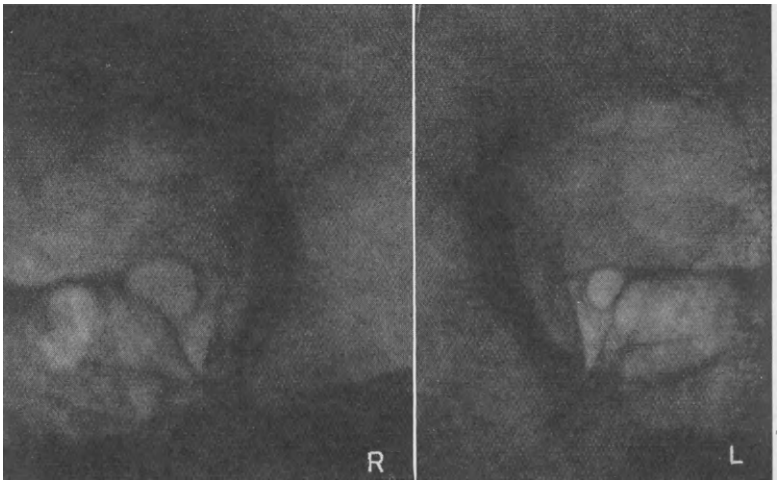
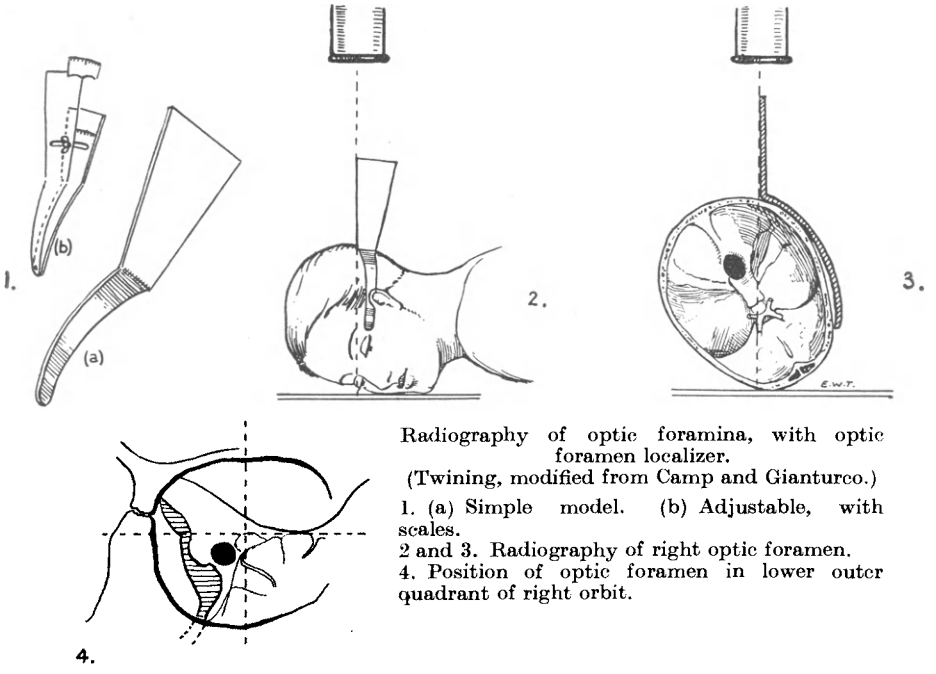


FIG. 92.—Skiagram of R. and L. orbits showing enlarged optic foramen on right side through which a glioma had extended intracranially. (Jefferson.)

Finally, he said, " the rœntgenographic outline was not always larger than the average or even larger than other diameters of the foramen, that variations in the calibre of the optic foramen are frequent, so that interpretations of rœntgenograms of the optic foramen should be expressed as possibilities and probabilities and not certainties." However, it is well to bear in mind some average such as given by Goalwin and illustrated by Fig. 91, but he states that a canal measuring less than 2·8 mm. in one of its diameters cannot contain a normal optic nerve. The important thing to remember in making a decision regarding the possibility of an abnormal optic canal being present is that the skiagrams of both right and left optic canals be compared (see Fig. 92).

A graphic record prepared by Keyes shows that the projected long axis of the canal falls in the great majority of cases along the horizontal midline at the junction of its upper and lower outer quadrants of the orbital entrance. In not a single case that he examined was the shadow of the optic canal thrown on the mesial half of the plane of the aditus orbitæ. 79 per cent. of projected foramina were found within the orbital aperture and within the outer half of the orbit. The remainder were tangential to or behind the outer and lower orbital margins.

The various methods employed in the radiography of the optic



Radiography of optic foramina, with optic foramen localizer.
(Twining, modified from Camp and Gianturco.)
1. (a) Simple model. (b) Adjustable, with scales.
2 and 3. Radiography of right optic foramen.
4. Position of optic foramen in lower outer quadrant of right orbit.

FIG. 93.

canal are discussed and illustrated in Hartmann's magnificent volume on ophthalmic radiography. Those with which we have become most familiar are the methods of Rhese, Hartmann, Goalwin, Lysholm, Camp and Gianturco and Pfeiffer. The main points in the technique of the radiography of the optic canal, as shown by Hartmann, are that the central ray should coincide with the axis of the optic canal and that the central ray must be perpendicular to

the plane of the sensitive plate. It is in the attempt to obtain such correctly that so many methods have been devised. To meet these requirements Hartmann has given the following data : the angle formed by the axis which passes through the optic canal and the sagittal or anterior-posterior axis of the skull should be 35·5 degrees and the angle between the axis of the optic canal below and the nasion-inion line above in the sagittal plane should be 31°. A convenient method of obtaining skiagrams of the optic canal has been devised by Camp and Gianturco. Fig. 93 illustrates an optic canal localiser which Twining in following these authors has had constructed. The position of the head is obtained by placing a sheet metal guide on one side of the head, say the left, No. 2 of Fig. 93. This guide is so shaped that its front edge points through the axis of the right optic foramen when accurately adapted to the left side of the head. For this purpose only two landmarks are necessary, the outer canthus of the eye and the upper junction of the pinna with the scalp. The head is now placed in such a position that the guiding edge is vertical and in the axis of the central ray. The guide is removed and an exposure made. The operation is repeated with the guide on the right side of the head, when it points to the left optic foramen. No. 4 of Fig. 93 shows the position of the optic canal projected in the plane of the aditus orbitæ. The base of the clinoid process is well shown at the same time.

The various anatomical structures that are thus defined by making a radiogram of the optic canal in this fashion are diagrammatically shown by Fig. 94.

Van der Hœve, who early recognised the work of Rhese, wrote a paper in which he showed that enlargement of the optic canal was present in—

(1) Hydrocephalus.

(2) Glioma of the orbit.

(3) Glioma of the intracranial portion of the optic nerve and chiasm.

(4) Neurofibromatosis diffusa of the optic nerve.

There is no doubt that the radiological demonstration of the optic canal will throw considerable light on some cases of retrobulbar neuritis, fractures of the orbit, skull deformities and medico-legal cases where loss of sight is alleged to have been caused by injury. In cases of intracranial aneurysm with monocular ophthalmoplegia two points have been observed by Jefferson and Twining which have

proved of great value. These are (*a*) enlargement of the inner end of the sphenoid fissure and (*b*) thinning of that strut of bone which passes from the body of the sphenoid to its lesser wing and forms the lower and outer boundary of the optic canal. When well marked the one is certain to accompany the other. In a case of Jefferson's which showed the absence of the shadow of the optic foramen due to a dural endothelioma (meningioma) an enlarged foramen rotundum was found. This apparently explained the reason for the neuralgic pain in the upper jaw accompanied by hyperæsthesia there.

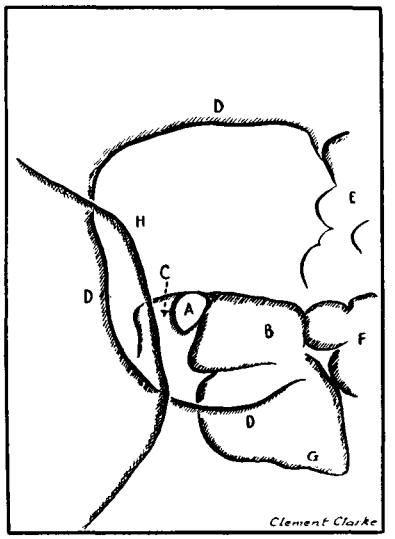


FIG. 94.—Explanatory diagram of the various shadows seen on left side of Fig. 92.

A. Optic canal.
B. Sphenoidal sinus.
C. Sphenomaxillary fossa.
D. Orbital margin.
E. Frontal sinuses.
F. Ethmoid sinuses.
G. Maxillary sinus.
H. Greater wing of sphenoid.

Malignant disease at the apex of the orbit tends to erase the foramen. Without deformity of the canal hyperostosis associated with an intracranial tumour at the end of the sphenoidal wing might show up in the skiagram situated above the canal. Although not deforming the canal such hyperostosis projecting backwards could press upon and destroy the intracranial portion of the optic nerve.

Diminution of the optic canal may follow the absence of growth of the optic nerve in anophthalmia —failure of growth of eyeball (Hartmann), or following birth injury. Sedan and Aurientis described the case of a girl aged nine years, who was completely blind in the right eye. Unilateral optic atrophy had been present since birth, and it was evident that the smaller and irregular size of the optic foramen on the side of the blind eye had followed the diminished size of the optic nerve due to atrophy (see Fig. 37).

Fractures through the foramen may cause its partial or complete obliteration.

It is in cases of monocular blindness primarily that radiology of the optic canal will yield its greatest results. Small optic canals,

associated with optic nerve disturbance, have been described by L. E. White, who shows that extensive pneumatisation around the canal tends to produce narrowing and leads to greater injury by infection. White states: "While it is possible surgically to enlarge the optic canal cases are rarely seen at the stage when it would be beneficial." Van der Hœve has described the operation for the removal of the roof of the optic canal which was followed by beneficial and lasting results to vision by removing pressure from the optic nerve.

The differential diagnosis of glioma and endothelioma may generally be summed up as follows :—

	Glioma.	Endothelioma.
Age	Occurs in young people.	Occurs more commonly in old people.
Visual disturbance .	Occurs before proptosis.	Proptosis generally precedes loss of vision.
Eye movements . .	Less disturbance of normal movements.	More pronounced limitation of movement.
Eyelids and conjunctiva	Circulatory disturbance may be absent.	Marked evidence of circulatory obstruction.
Pain . . .	Not frequent.	Frequently present.
Intraocular extension .	Generally absent.	May be present.
Intracranial extension.	Commonly occurs.	Less common.

TUMOURS OF THE INTRACRANIAL PORTION OF THE OPTIC NERVE

From a perusal of Hudson's paper it is clear that the growth of many of the tumours of the optic nerve which he described was not confined to the orbit. Post-mortem examination showed extension in many cases through the optic canal into the cranial cavity. Such growth is slow or almost stationary, so that the patient eventually succumbs to some other ailment. Of the seven cases of endotheliomata which were examined post-mortem, five revealed the presence of direct intracranial extension. In Heuer's list of twenty-eight chiasmal lesions there was a sarcoma arising from the optic nerve sheath, also a glioma which was a primary chiasmal tumour extending beyond these confines.

As the intracranial portion of the optic nerve is not covered with

dura mater an endothelioma can arise only at the foramen and may grow forwards through the optic canal or extend backwards. Gliomas can arise in any part of the optic nerves or chiasma or may extend into the tracts.

In one of Dandy's cases the intracranial portion of each optic nerve was surrounded in a collar-like manner by a small tumour. The tumours were pale, greyish pink, resembling granulation tissue. The diameter of each tumour, including the optic nerve, was 1·5 cm., while posteriorly it extended along the nerve 0·75 cm. It had grown through the optic canal for a distance of 1·5 cm. but extended into the orbit only as a linear strand. Both tumours were dural endotheliomata. In order to remove the right tumour it was necessary to strip the dura from the optic nerve. The removal of the portion of tumour which had grown into the optic canal necessitated the removal of part of the roof of the canal and orbit. Partial removal of the left tumour restored vision considerably including colour fields. In Dandy's second case a small tumour, the size of a cherry, was found beneath the left optic nerve causing the latter to arch sharply. A band of fibrous tissue over the optic foramen and anterior clinoid process held the optic nerve tightly against the tumour constricting the nerve. Severance of this band released the nerve from the clutches of the tumour, and although the tumour was not removed owing to the operation being conducted from the right side yet remarkable restoration of vision resulted from division of the fibrous band.

Adding Byers' and Hudson's cases (without duplication) Dandy points out that considerably more than the obvious 21 per cent. of tumours of the optic nerve showed intracranial extension, and indeed in twenty-three post-mortem examinations twenty-one showed an intracranial growth, and in only one instance did the cranial cavity contain no tumour.

In a series of 233 tumours arising from or near the hypophysis and which had affected the chiasm by pressure, thirteen interpeduncular gliomas were described, of which seven were true chiasmal tumours ; these latter have been described by Martin and Cushing. Four of their cases originated during the first ten years of life, but one case occurred at fifty-six years of age. All seven cases were gliomatous and involved one or both optic nerves. The lateral view of the pituitary fossa was deformed anteriorly beneath the anterior clinoid processes, this being undoubtedly associated with

the dilated optic foramen. During the operation on Martin and Cushing's second case a drawing was made of the appearance of the tumour (Fig. 95). A symmetrical tumefaction involving both chiasm and nerves up to the optic foramen is described. The wedge-shaped marking is due to removal of a piece of the tumour for pathological investigation; this showed dense neuroglial tissue surrounded by an outer fibrous capsule, the neuroglia consisting of masses of fibrils. Cytoid bodies, globular and rod-like masses of

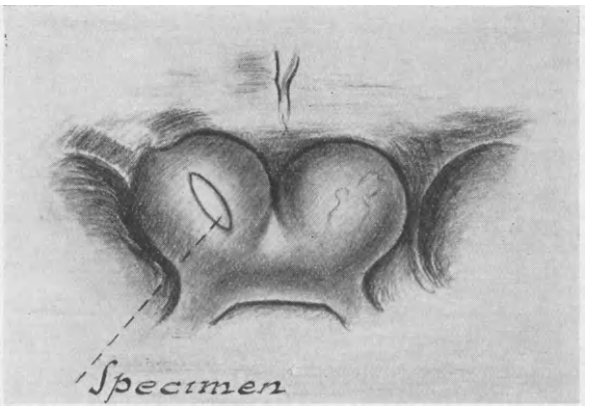


FIG. 95.—A symmetrical tumefaction involving chiasm and optic nerves up to optic foramina. Skiagram showed apparent extension of the sella which is often associated with enlarged optic canals. (Martin & Cushing.)

hyalin, such as Verhœff has described in some of his cases of glioma were present.

In 1936 Meheny described three cases of primary tumour (glioma) of the optic nerve. In the first case it was necessary at operation to remove the roof both of the orbit and the optic canal. In the second case the tumour was found to extend from a point 0·5 cm. from the globe to the chiasm on the left side, enlarging the optic canal on its way, such enlargement having previously been demonstrated by radiography. Again the roof of the orbit and optic canal were removed, the nerve was divided behind the globe, the chiasm divided close to the right optic nerve and the tract divided to include as much of the tumour as possible. The tumour in the third case arose from the left optic nerve, which was entirely replaced by neoplastic tissue and pressed the right optic nerve laterally. Only in Meheny's third case did a fatality occur. Of the

seven cases reported by Martin and Cushing five died immediately or soon after operation. Apparently where death occurred at operation the cause was hyperthermia. No wonder Martin and Cushing took a depressing view of tumours of the intracranial portion of the optic nerve and chiasm. Cairns removed a glioma from a boy who lived one year after operation. Fig. 96 shows the enlarged optic foramen in Cairns's case through which the tumour had extended backwards to the intracranial cavity. Cairns also removed a meningioma from a woman in her late forties, the radiogram of whom is illustrated by Fig. 97. The tumour extended from the suprasellar region through the right optic canal, which is seen to

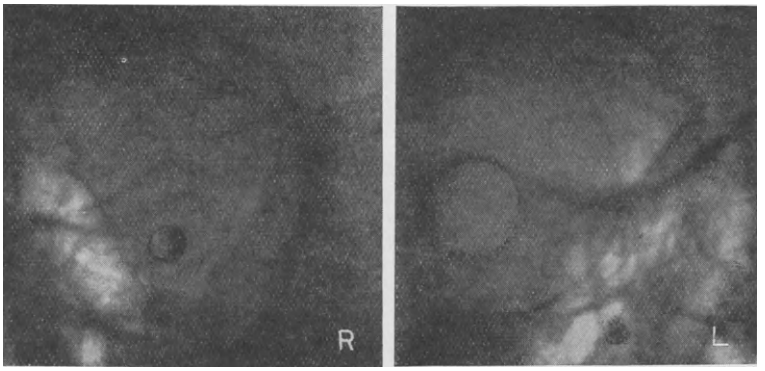


FIG. 96.—Skiagram of R. and L. orbits showing enlarged optic foramen on left side through which a glioma had extended intracranially. (Cairns.)

be enlarged in the skiagram. The eyeball was left *in situ*. Fifteen months later I examined this eye. It was immovable and divergent in position. The inner half of the cornea was anæsthetic ; touching the outer half with a wisp of cotton wool induced blinking, which is remarkable, for of a surety the ciliary nerves were divided along with the optic nerve. Something else equally remarkable presented itself ; the pupil was dilated and fixed as would be expected and when light was thrown on it or the fundus examined there was no pupillary reaction ; the consensual reaction was absent also, but when the arc light of the fundus camera (Nordenson) illuminated the eye, the pupil slowly contracted down and became smaller than its fellow, and as the fundus could not be seen the artist had to send the patient away for tea. On her return the pupil had enlarged again

and remained so while a drawing of the retina was being executed (see Plate XV).

The subject of tumours of the optic nerve associated with von Recklinghausen's disease will be dealt with later. Such a case under Jefferson's care exhibited a tumour of the right optic nerve which had extended intracranially. The enlarged optic canal is shown in Fig. 92. (Histologically the tumour was a polar spongioblastoma). The boy is still alive three years after operation. The left side is normal. The diagram, Fig. 94, provides an explanation of the

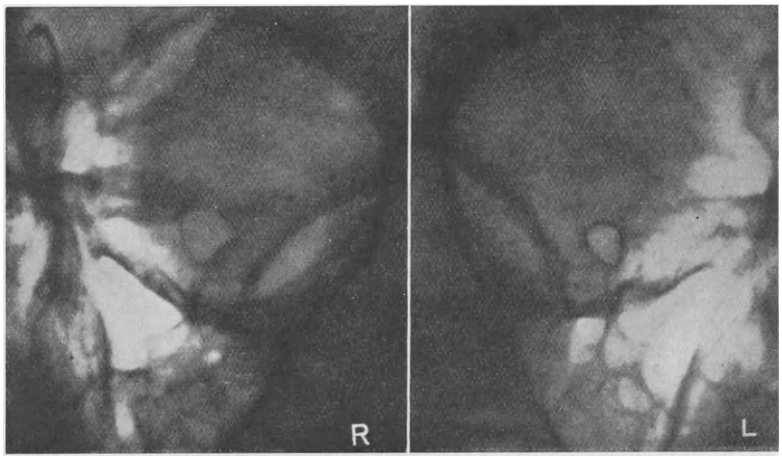


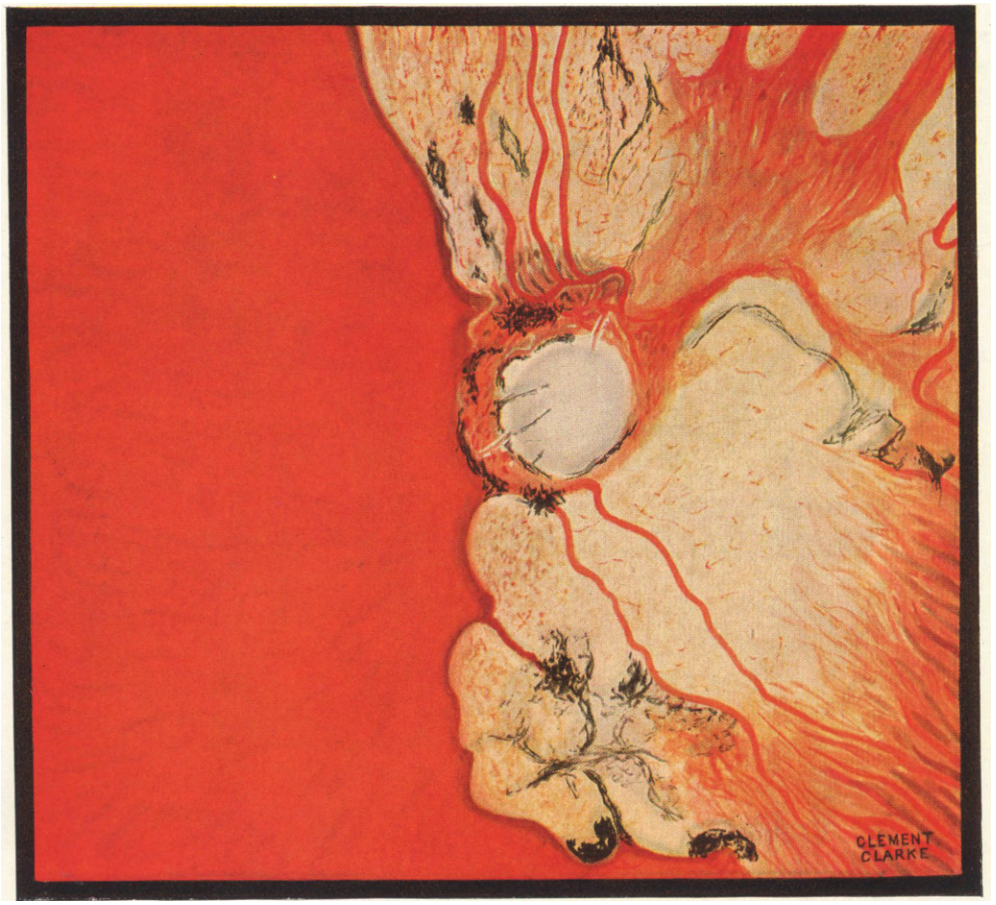
FIG. 97.—Skiagram of R. and L. orbits showing enlarged optic foramen on right side through which a meningioma extended intraorbitally. (Cairns.)

shadows shown on the left side of Fig. 92, which is normal and well defined.

Diagnosis.—*Exophthalmos.* In the first place there may or may not be exophthalmos present. If present it may be uni- or bilateral.

We must depend mainly on the ophthalmoscope, fields of vision and radiography to assist us in our diagnosis. The majority of cases recorded show the presence of optic atrophy, many of which are of the consecutive type while some are primary. In one of Martin and Cushing's cases evidently a papillœdema was superimposed on an optic atrophy attributed to a presumed internal hydrocephalus. It is pointed out, however, by Dandy that as most of the prechiasmal tumours are small, signs of raised intra-

PLATE XV



FUNDUS OF EYE WHICH HAS BEEN LEFT IN SITU AFTER REMOVAL OF AN ORBITAL TUMOUR INVOLVING THE OPTIC NERVE

The disc is white and devoid of blood vessels except for faint remnants that are left on edge of disc. The nasal half of the retina has degenerated but the temporal half is much more normal in appearance. (Case in charge of H. Cairns.)

[*To face p.* 260.

cranial pressure are therefore absent, and that there is a progressive loss of vision in one or both eyes. In each of his cases there was a primary optic atrophy. In Meheny's first case, that of a large (15 mm.) tumour of the left optic nerve, there was a left papillœdema of 3-4 D, the right fundus being normal. In the second case, a tumour of the left optic nerve, the left fundus showed a papillœdema of 1 D while the veins of the right fundus were full. Meheny's third case showed diffuse (consecutive) atrophy of the right disc while the left disc was more atropic than the right. It seems probable, therefore, that such tumours may produce a papillœdema or that primary optic atrophy may become slowly evident. In the former case vision may remain good through a lengthy period, but in the latter vision slowly and steadily diminishes.

Fields of Vision.—In Grinker's case he states, "The finding of a homonymous hemianopia with its margin through the fixation point, marked disturbance in the pupillary reactions, the combination of partial optic atrophy and partial mild papillœdema fixes the location of the tumour about the chiasm." If the tumour affects the intracranial portion of the optic nerve the fields of vision show bitemporal or homonymous hemianopia or total blindness of one eye and hemianopia in the other. Fig. 90 illustrates the irregularly concentric diminution of the peripheral field in the case of an intraorbital growth generally accompanied by a central scotoma for form and colour. The field of one eye only is affected if the growth is confined to the orbit.

Radiography.—For the diagnosis of chiasmal and prechiasmal tumours it is necessary to obtain skiagrams both of the sella turcica and of the optic canal. The anterior wall beneath the anterior clinoid processes may be eroded, resembling that which is seen in Fig. 103. Enlargement of the optic canal is positive proof of the presence of a tumour involving the optic nerve in this region.

The pupillary reactions are constant in only one respect—the reactions are sluggish on the side directly affected. However, owing to atrophy of the optic nerve the reaction to light in many cases is entirely absent.

Quantitative perimetry will indicate most probably the earliest signs of those tumours now under consideration. Such changes in the field of vision will be slight and slowly progressive in contradistinction to the sudden and rapid changes found in typical retrobulbar neuritis.

INFLAMMATORY PSEUDO-TUMOURS OF THE ORBIT

At this stage it may not be out of place to discuss briefly such a condition to which the name—inflammatory pseudo-tumour of the orbit has been given.

Since Birch-Hirschfeld's description of such a case many have been recorded. They have all the same characteristic; they all cause proptosis of one or both eyes. They arise suddenly or with slow onset, remaining stationary for a long time, or they get worse or may resolve. Vision in some cases is scarcely affected, while in others blindness rapidly supervenes.

A useful classification according to Birch-Hirschfeld is as follows :—

(1) Those cases of pseudo-tumour, where recovery is spontaneous following the administration of mercury, iodies, etc.

(2) Those cases where exploratory operation has been done but no tumour found.

(3) Those where the macroscopic examination has suggested a tumour but microscopical examination has discovered the tumour to be an inflammatory mass.

The diagnosis in many cases is not easy, for, as Ewing remarks, the diagnosis between lymphocytic proliferation due to inflammation and that due to neoplastic activity is difficult, while an intermediate condition is sometimes present.

In one of Williamson Noble's cases the masses of lymphocytes apparently proliferated long after the infecting organisms had been removed.

Focal infection is responsible for some of the cases recorded by Benedict and Knight. Possibly the three cases described by Williamson Noble are illustrative of the various causes responsible for such a condition. His first case was undoubtedly gummatous. Such is the case illustrated by Plate XIV, which resolved completely by the active administration of anti-syphilitic remedies. The onset of proptosis and blindness was extremely rapid in this latter case.

The second case showed lymphocytic infiltration which followed rapidly on an attack of influenza, while the third case showed proliferation of connective tissue cells consequent on their irritation by products of hæmorrhage.

Where lymphocytic infiltration has been described the process has usually been confined to one orbit, whereas, according to

Lagrange and Gayet, both orbits are symmetrically involved in cases of lymph-adenoma.

In Williamson Noble's paper he suggests that in coming to a conclusion regarding the diagnosis of inflammatory pseudo-tumour of the orbit one should—

(1) examine the urine for sugar ;
(2) perform the Wasserman reaction ;
(3) exclude tuberculosis ;
(4) a differential blood test and estimation of blood coagulation time should be done ;
(5) search for focal infection, including dental radiography ;
(6) exclude Graves's disease ;
(7) possibly do an exploratory operation (but not until the Wasserman reaction is known).

TREATMENT OF TUMOURS OF THE OPTIC NERVE

Until recent times tumours of the optic nerve have been dealt with either through the aditus orbitæ or by removal of the lateral wall. The discovery that the optic foramen could be delineated easily by radiography has brought about quite a new aspect of the methods of treatment of these tumours. Formerly, the method of removal was to excise the tumour, removing it and the globe together. Occasionally, exenteration was done, but this in the majority of cases was unjustifiable, as these tumours show little tendency to invade the orbital tissues. After Knapp, in 1874, showed a case where the eyball was left *in situ* it became the object of most surgeons to emulate his example. In Knapp's own description of this operation he stated that he first made an opening through the conjunctiva and Tenon's capsule between the superior rectus, internal rectus, and superior oblique muscles until he could palpate the tumour with the tip of his index finger. Separating the tumour from the sclera he divided the optic nerve behind the globe and then at its orbital end, without cutting any of the ocular muscles. Finally, with flat scissors he pried out the tumour, leaving the globe in place. Following this operation its repetition frequently occurred with various modifications, such as tenotomising muscles, external canthotomy, etc.

A somewhat more efficient mode of gaining access to the orbit was described by Lagrange as " De la conservation du globe

oculaire dans l'ablation des tumeurs du nerf optique. Description d'un procédé nouveau." The external canthus was divided, the lids held apart by sutures. The conjunctiva on the ball was dissected back to the external rectus, which was tenotomised, the end of the muscle held by a suture for replacement later. The tumour was isolated by dissection and the eyeball turned inwards. A ligature was passed around the tumour by a Cooper's needle, the ophthalmic artery was avoided and the nerve sectioned near the optic foramen. By pulling on the suture the tumour was withdrawn from the globe. The external rectus was re-united and the lids and conjunctiva were sutured.

Rollet, in 1914, described a method of approach to tumours behind the eye. He sutured the lids to prevent ulceration, then a long curved incision, 3 cm. in length, was made along the outer margin of the orbit—" subaponeurotic orbitotomy." The orbital fascia was then separated from the bony margin by a concave rugine which opened a space between the contents of the orbit and the bony wall, the periosteum being on its outer side and the orbital contents on the inner. The finger was used for palpation and dissection. The eyeball can be retained by this method also.

In 1887 a Swiss surgeon named Krönlein introduced an operation for the removal of dermoid cysts from the orbit, but this operation was not regarded as an ophthalmic procedure until Braunschweig, in 1893, introduced it into ophthalmology. Byers' translation of Krönlein's original description is as follows :—

" *Skin Incision.*—The incision begins in the temporal region at the point where the linea semicircularis of the frontal bone is distinctly felt through the skin, in other words, about 1 cm. above the margo supra-orbitalis, and extends downwards along the outer orbital margin as a slight curve with the convexity forwards, to the level of the upper margin of the xygomatic process of the malar bone, where it turns backwards and ends in the middle of this structure.

" The skin incision is then carried down to the bone along the outer orbital margin, and through the periosteal incision the whole of the periosteum is separated from the lateral orbital wall by means of a raspatory, a proceeding easy of accomplishment. The point of the raspatory is then passed downwards to the inferior orbital fissure in order to fix the spot where the converging bony incisions presently described meet one another.

" *Bony Incisions.*—The osteoplastic resection includes the whole of the outer orbital margin (process. zygomaticus oss. front. and process. front. oss. zygomatic), and that part of the outer wall which lies between

this limit and the inferior orbital fissure (pars orbitalis oss. zygomatici and anterior part of the ala temporalis oss. sphenoid.). The piece of bone to be temporarily removed has therefore the shape of a wedge whose base is formed by the outer orbital margin (process. zygom. oss. front. and process. front. oss. zyg.), and whose apex ends at the anterior part of the inferior orbital fissure.

" The bony incisions are best made with a sharp chisel without any further ' Præparation,' and especially without loosening the natural connections to which, together with the skin flaps, falls the work of nourishing the separated piece of bone during the early period of healing. First of all, the process zygomaticus of the frontal bone is chiselled transversely through above the distinctly visible and palpable sutura zygomatica-frontalis, and the bony incision continued forward in a direct though oblique line through the lateral orbital wall to the raspatory in the inferior orbital foramen. Following this the processus frontalis of the malar bone is chiselled horizontally through close to its base and the incision likewise continued into the fissure.

" When this has been done the bony piece thus made free, together with the skin, fasciæ and muscle flaps of the temporal region, can be so far turned outwards that the entrance to the orbit in its lateral part appears free. The operation is usually easy and quickly performed ; after successful removal of the tumour, the skin and soft parts are replaced and held in position by sutures and healing follows without difficulties or disfigurement.

A modification of the Krönlein operation has been devised by Angelucci which seems simpler. He makes an incision through the soft parts starting at the external angle of the orbit and descending vertically to the base of the orbital process of the malar ; this incision is about 4 cm. long. From the lower end of this a second incision runs for 20 mm. along the upper border of the zygoma. He divides the orbital process of the malar with a saw, below for a depth of about 6 mm. and above, following the line of the fronto-malar suture, for a depth of 2 or 3 mm. Separating the periosteum from the internal surface of the bone and grasping the bone in cutting bone forceps he dislocates it outwards, thus producing a fracture along the line of the spheno-malar suture. This procedure with regard to the division of the bone seems simpler than Krönlein's method.

Studying these operations we are forced to ask ourselves two things : Firstly, what is the ultimate fate of the globe, and secondly, has the whole of the neoplasm been removed ?

It has been shown (Byers) that probably 55 per cent. of the globes left *in situ* proved to be successful, but ultimately this figure would

be greatly reduced. Some of the globes shrank, some orbits suppurated and the globe had to be removed, the cornea in others became opaque and very few had normal movements. Ptosis was a common disfigurement.

Knapp's method and Krönlein's have been compared with regard to their results. Byers thinks the evidence is in favour of Krönlein's operation, but Hudson leans to the former method. (See Hudson's table of comparison.)

Many ophthalmic surgeons do not attempt to leave the eyeball *in situ*. No doubt by opening Tenon's capsule and removing the eyeball first, then dissecting back, a tumour confined to the orbit could be removed. A glass or metal globe could be inserted and covered in by the purse-string suture combined with double-breasting the overlying tissues. This would result in a much more presentable appearance than a deep hollow orbital pit. An artificial eye, even though its movements may not be very free, if it is on a level with the opposite eye and holds the upper eyelid in place, is yet a much better cosmetic result than the original eye incapable of movement—divergent or otherwise—with ptosis and with the possibility of shrinkage, etc.

We have already seen how radiography of the optic canals has opened up a new field in neuro-ophthalmic surgery. If indications are such that the tumour extends through the optic canal then those operative procedures already mentioned fail in the attempt to remove the tumour in its entirety. The approach to intracranial tumours has brought into being many and varied operations. Formerly the lateral route was used to approach the region of the pituitary and the chiasm, but gradually it became clear that the so-called transfrontal route was the best method. Those cases—3, 4, 6 and 7—of Martin and Cushing's, had a transfrontal operation performed. Cairns (by personal communication) says there is no doubt that the transfrontal or, as he prefers to call it, the subfrontal route gives the best access to that region where the roof of the orbit and optic canal may have to be removed and the tumour involving the intracranial portion of the optic nerve dealt with.

Lysholm and Olivecrona describe the operation done for a case where the tumour involved the optic nerves intracranially :—

" Under local anæsthesia a right-sided transfrontal flap was turned down. There was considerable increase of the intracranial pressure necessitating ventricular puncture. After the pressure had been

relieved it was possible to elevate the frontal lobe and open the dura along the lesser wing of the sphenoid. The chiasm and both optic nerves were elevated by a bluish-red tumour bulging beneath these structures. Between the legs of the chiasm a small nubbin of tumour tissue projected upwards. The tumour could be seen to extend far to the right, pushing the carotid artery to the right. When the artery was touched the patient complained of the same pain at the top of his head which he had been having for the last few months." They, however, did not find it necessary to open the optic canal in this case.

When the optic foramina appear equal and normal in size and the fields of vision do not present a hemianopic appearance, an intra-orbital operation may be considered as in Costin's case. The weakness of the operative method employed in one case is shown by the statement: "After an orbital operation had been performed, in view of the possibility of extension to the cerebrum it was deemed advisable to give the child post-operative roentgen treatments." In another case, although the skiagram showed the left optic foramen to be twice as large as the right, yet a Krönlein's operation was done. Such an operation should not be performed if an optic canal shows enlargement.

To expose the "anomalous" ophthalmic artery described by Pfingst Dr. Spurling turned down a left frontal flap. The frontal lobe covered with dura was reflected from the floor of the anterior fossa. The roof of the orbit was removed piecemeal with rongures. All the bone was widely removed from the roof of the orbit down to and including the bone over the optic foramen. Then the dura was opened in line with the superior limb of the bone flap in order that the optic nerve and foramen might be directly inspected. Such a method of approach has the approval of Cushing, Heuer, Dandy, Cairns and others. Therefore, when a tumour is found to have extended intracranially or intra-orbitally, surely both the ophthalmic surgeon and the neuro-surgeon should combine to obtain the best result. One principle, however, should always be observed. Both craniotomy and enucleation of the globe must not be done at the same time else meningitis will ensue.

CHAPTER XII

THE REGION OF THE OPTIC CHIASMA AND PITUITARY BODY

THE optic chiasma is formed by the junction of the two optic nerves as they pass upwards and backwards from the optic foramina. The chiasma is of oblong form which, measured transversely, averages 13 millimetres, while antro-posteriorly it measures 8 millimetres and is 3 to 5 millimetres in thickness. It is enveloped by pia mater and lies in the cisterna basalis of the subarachnoid space on the sphenoid bone, but it does not occupy the sulcus chiasmatis. In 1909 Symington pointed out that the optic chiasma was usually placed behind the sulcus and formed a prominence on the anterior part of the floor of the third ventricle; behind lies the tuber cinereum, from which the hypophyseal stalk or infundibulum springs to pass into the posterior lobe of the pituitary body. The internal carotid artery forms a very important relation to the chiasm. It lies close to its outer side (see Plate I); one of its branches, the anterior cerebral artery, passes in front of the chiasm to reach the longitudinal fissure, while another branch, the posterior communicating artery, passes backwards to join the posterior cerebral artery, forming part of the circle of Willis. The right posterior communicating artery is usually larger than the left. If the chiasm is gently raised with forceps a thin lamina will be seen passing upwards into the great longitudinal fissure to join the rostrum of the corpus collosum, closing the third ventricle in front; it is continuous on either side with the anterior perforated spot, through which pass the basal branches of the anterior and middle cerebral arteries.

In the Bowman Lecture, 1923, de Schweinitz quoted Professor Schaeffer's detailed study and report on the position of the optic chiasma in relation to the diaphragma sellæ and hypophysis. In 79 per cent. of the bodies examined the chiasm was placed partly or wholly over the diaphragma sellæ and underlying hypophysis, 5 per cent. showed the anterior part of the chiasm resting on the sulcus chiasmatis, while in only 4 per cent. the chiasm rested on the dorsum sellæ behind the diaphragma sellæ (see Fig. 98).

The pituitary body, a small reddish-grey mass of a somewhat flattened oval shape, about the size of the kernel of a cob-nut (Sharpey-Schaefer), and widest transversely, lies within and occupies the sella turcica of the sphenoid bone. It is enclosed above by a small circular fold of the inner layer of the dura mater which forms a roof for the sella turcica and is termed the diaphragma sellæ. Through this diaphragm the infundibulum passes from the tuber cinereum above to the posterior lobe of the pituitary below. The circular sinus, consists of two transverse channels, one passing in front and the other behind the infundibulum, connecting the two cavernous sinuses. The pituitary body consists of two lobes and a pars intermedia, which can be seen by the naked eye on section. The larger anterior lobe is hollowed out behind to receive the

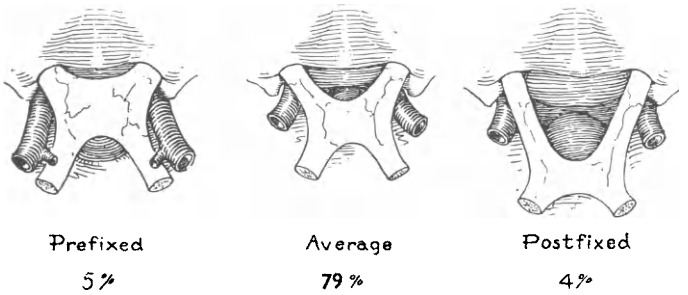


FIG. 98.—To show the three principal types of chiasm. (After Schaeffer.)

smaller posterior lobe. The infundibulum passes into the posterior lobe, and is not structurally connected with the anterior lobe. The *pars anterior* is extremely vascular, and its cells, as shown by staining with erythrosin-orange G-toluidin blue, consist of two kinds, clear and granular. The clear cells are known as the chromophobe cells, the granular as chromophil cells. The staining further shows that the granular cells are both oxyphil and basiphil. The chromophobe cell has the potentiality of developing into one or other of the chromophil cells (Biggart). The pars anterior is usually enlarged in acromegaly and gigantism. The *pars intermedia*, clearly seen in the cat, is not so clearly separated from the pars anterior in man. It is well marked off from the posterior lobe, the pars nervosa, but at certain places the cells pass from the pars intermedia to the pars nervosa, where they undergo a hyalin and granular colloid change ; these bodies pass into the third ventricle. The *pars nervosa*

consists mainly of neuroglia cells and ependymal fibres. It receives nerve fibres from large cells in the grey matter of the brain lying behind the optic chiasma, and some of these fibres pass into the anterior lobe. In addition, there is a portion of the pituitary body known as the *pars tuberalis*, consisting of strands of epithelial cells, and is extremely vascular. Its function is unknown.

The posterior lobe is derived from the floor of a hollow downgrowth from that portion of the developing brain which afterwards becomes the third ventricle, while the anterior lobe is formed by an ectodermal offshoot from the primitive buccal cavity (Rathke's pouch). A canal is sometimes found passing from the anterior part of the hypophyseal fossa of the sphenoid bone to the under surface of the skull and marks the original position of Rathke's pouch ; this is termed the cranio-pharyngeal canal.

The tumours which are found in the region of the chiasm and the pituitary body are adenomas, endotheliomas (meningiomas) gliomas, tumours of the hypophyseal duct (craniopharyngiomas or adamantinomas), adeno-carcinomas and the rare cordoma. Aneurysms also may be the cause of lesions in this neighbourhood, while chronic basal arachnoiditis must also be mentioned.

Cushing says that of all the lesions that produce bitemporal loss of the fields of vision with primary optic atrophy in adults and middle-aged persons the adenomas of the hypophyseal anterior lobe are the most common.

In the region of the chiasma and hypophysis a meeting-place has been provided anatomically for the ophthalmologist, neurosurgeon and neurologist, whose united labours for the past score of years have revealed a considerable amount of knowledge concerning the visual pathways and the derangements thereof which formerly were but imperfectly understood. We read with admiration the works of Harvey Cushing and C. B. Walker ; we follow with extreme interest the descriptions of cases by Gordon Holmes and Percy Sargent, yet we cannot forget that in the year 1904, and even before this date, Sir William Gowers wrote on " chiasmal neuritis." His words are worth quoting : " In a group of cases, rare but well defined, there is failure of sight in the temporal half of each field of vision, indicative of damage to the fibres that cross at the chiasma, probably at the seat of their decussation. The temporal hemianopia may be irregular in form and incomplete in extent, but often extends up to the middle line. There may be complete loss of the half

fields for colour when perception of white is only lessened, or when there is a paracentral scotoma, on the defective side. The optic nerves within the eye gradually become pale, presenting the aspect of simple atrophy." Nettleship in 1897 described similar symptoms produced by a growth from the pituitary body.

The tumours may be *intrasellar* or *suprasellar* ; that is, they may arise from within the sella turcica and from beneath the diaphragma sellæ, or their point of origin may be above this level, yet the symptoms which on first thoughts one would imagine to be of a somewhat opposite character are practically identical.

The *adenomas* which arise from the anterior lobe of the pituitary are of four kinds—the chromophil, the chromophobe, basophil and mixed cell adenomas. Of the first two Cushing stated that in a series quoted there were 219 examples of the chromophobe variety as against eighty-nine chromophil. The endocrine disturbance associated with the chromophil variety is marked if the tumour occurs before the cessation of skeletal growth. There is pathological hyperpituitarism, so that gigantism occurs, or if the lesion takes place during later years acromegaly develops. Hyperglycæmia and glycosuria may occur, this condition not being so responsive to insulin as that of diabetes mellitus. In the case of a chromophobe adenoma hypopituitarism occurs, such as impotence in the male or cessation of menstruation in the female. They grow more rapidly, too, than the chromophil tumours. Basophil and mixed cell adenomas are much rarer than those just described. In 1932 Cushing suggested that the multiglandular syndrome—obesity, hirsutism, hyperpiesia, osteoporosis, polycythæmia and amenorrhœa —was probably due to the excessive secretion of the basophil cells of the pituitary gland. This condition has come to be known as *pituitary basophilism.*

Endotheliomata or meningiomas are suprasellar tumours. They arise from the meninges in the neighbourhood of the cavernous sinus (see Fig. 99), diaphragma sellæ, tuberculum sellæ and the chiasmal sulcus. They occur in adult life usually ; due to their irregular manner of growth they involve the optic nerve or the optic tract before the chiasma. In many cases of this kind the patient presents himself with a blind eye, primary optic atrophy being present. Headache is not severe and endocrine disturbances are practically never present (Holmes). In view of the frequency of this type of tumour among those of the chiasmal region the possibility

of the presence of a meningioma in adults should be borne in mind when symptoms of slowly progressive damage to one or both optic nerves, as evidenced by central or temporal blindness associated with primary optic atrophy without a rise of intracranial pressure, are present, with absence of endocrine disturbances and negative radiological findings.

In the differential diagnosis between an adenoma and a meningioma (Cushing) the following points may be emphasised:

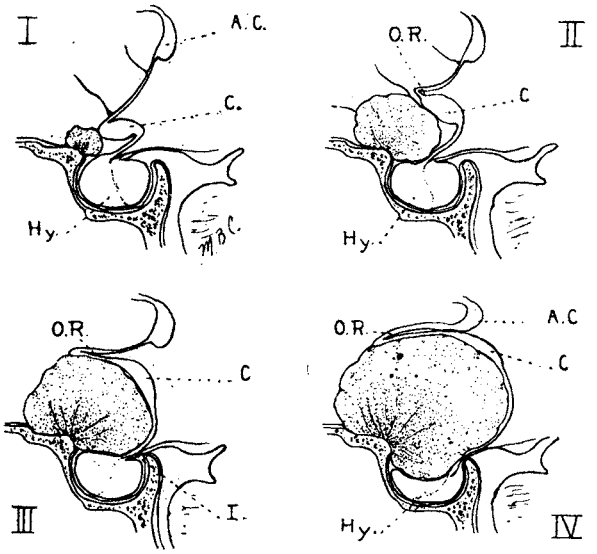


FIG. 99.—To illustrate on sagittal section four stages of the advancing deformation of chiasm and third ventricle produced by a meningioma. (Cushing.)

(1) greater evidence of pressure atrophy in the former; (2) co-existence of mild hypopituitary signs; and (3) the more rapid evidence of symptoms, for the pituitary adenoma is doubtless a more rapidly extending tumour than a meningioma.

In the syndrome of meningeal fibroblastoma arising from the lesser wing of the sphenoid bone, Groff says the most striking and significant features are the ocular signs. Eight out of 9 cases showed atrophy of one or both discs, 5 were bilateral and 3 were unilateral. One of the latter had papillœdema on the opposite side. The visual field showed homonymous hemianopia. Exophthalmos occurred in 4 of these cases; it was unilateral and on the side of the lesion.

The oculo-motor nerve was paralysed in 3 cases and the abducent in one. The trigeminal nerve was involved, there was pain in 1 case over the ophthalmic division, another had paræsthesia and pain over the ophthalmic and maxillary divisions. Unilateral loss of sense of smell was recorded in 2 cases. In some there was unilateral erosion of the pituitary fossa with enlargement of the optic canal.

Tumours of the hypophyseal duct or cranio-pharyngiomas, also called *tumours of Rathke's pouch* or *adamantinomas*, being congenital, usually give first evidence of their presence in childhood. The radiologist is often the first to diagnose the condition, as calcification in the suprasellar region generally indicates such a tumour. The calcification may consist only of a few flakes or may be the size of a tennis ball. These tumours are not at all uncommon. The anlage of the tumour may be at the anterior angle of the anterior lobe or may be within the sella. If in the latter situation the growth may expand the sella and may resemble an intrasellar adenoma. Headache and vomiting may be marked and papillœdema rather than optic atrophy may occur. If the chiasma is pressed upon from above the bitemporal hemianopia may begin in the lower quadrant. When the symptoms of this tumour appear in childhood, the result may be sexual and skeletal infantilism, but if they occur during adolescence there may be signs of hypopituitarism. Indeed, if the tumour presses on the tuber cinereum hypothalamic symptoms present themselves, such as polyuria, adiposity and hypersomnia (Bogaert).

It was stated by Holmes and Sargent that when the optic nerves are compressed by an endothelioma simple primary optic atrophy occurs, but papillœdema or consecutive optic atrophy is present in a considerable proportion of stalk tumours. Critchley and Ironside found papillœdema in 8 cases of adamantinoma and primary optic atrophy in five.

The differential diagnosis, according to Martin and Cushing, between a chiasmal glioma and the more common variety of suprasellar tumour arising from Rathke's pouch, both frequently recognised in childhood, might be presented as follows :—

Tumour of Cranio-pharyngeal Pouch	Tumour of Chiasm
Primary optic atrophy. In late stages, owing to hydrocephalus, œdema may be superimposed.	Primary optic atrophy occasionally with tumour involvement of nerve head. Unilateral exophthalmos in advanced cases.

Bitemporal hemianopsia, or, if vision is lost in one eye, fairly acute vision retained in the seeing half of the other eye.

Process slow, often remaining stationary for long periods.

Sella variously deformed, enlarged or normal. Posterior clinoids more affected than anterior. Suprasellar shadows common.

Secondary pituitary manifestations common with adiposo-genital dystrophy and infantilism.

Acuity low in both eyes with fields showing less typical hemianopic defects.

Process on the whole more rapid and progressive.

Sella in advanced cases shows apparent extension under anterior clinoids from distention of optic foramina. No suprasellar shadows.

Secondary pituitary manifestations inconspicuous. Cutaneous indications of von Recklinghausen's disease to be looked for.

Gliomas of the chiasm arise from the chiasm or the adjacent wall of the third ventricle, spreading forwards sometimes as far as the bulbar end of the optic nerve. They are found in childhood and often associated with generalised neuro-fibromatosis (von Recklinghausen's disease). The situation of the tumour causes blindness, and this may be its first symptom. The field defect does not show the typical vertical meridian, but they are generally bitemporal, while primary optic atrophy is the rule, although a bulging of the optic nerve head may be observed, due to the spread of the glioma down the length of the nerve. This tumour produces a bulging or erosion beneath the anterior clinoid process, which can be seen roentgenologically. The enlargement of the optic foramen can sometimes be observed in the radiograms. We see then that cranio-pharyngiomas and gliomas of the optic nerves and chiasm occur most commonly in children, whereas adenomas rarely do, and, moreover, the former do not cause such regular defects in the visual fields as are produced by the adenomas. Hypothalamic symptoms, too, are commonly associated with the cranio-pharyngiomas and gliomas, but not with the adenomas.

The *chordoma* is a rare tumour (1 in 1,700—Cushing ; Grinker states that thirty have been described in the entire neurological literature). It arises from the remnant of the notochord at the junction of the dorsum sellæ and the occipital bone. In Cushing's case there was a progressive loss of temporal field with primary optic atrophy. These tumours may spread backwards into the brain stem or forwards into the naso-pharynx.

Aneurysms in the region of the chiasm. It is quite possible for

an aneurysm to arise from the circle of Willis, internal carotid, anterior cerebral or anterior communicating artery, or even as far back as the basilar artery (Hutchinson), which by its growth may press upon the chiasm and produce the syndrome of a suprasellar tumour. They may be differentiated by a history of diplopia, even years before visual field defects are evident. In the case reported by Harris there was a spontaneous return of vision after the patient had been blind for some weeks. The aneurysm in this case arose from the left anterior cerebral artery compressing both the chiasma and left optic tract. The variability of vision or even the recovery from blindness is not diagnostic of tumours in this region, although Holmes has quoted cases of endotheliomata where vision did return for a short period, but this is much more common in the case of aneurysms. Further, symmetrical bitemporal field defects are rarely seen in the case of aneurysms. Radiography may present nothing abnormal, but, on the other hand, it may show notching or erosion or shadows may be formed in advanced arterio-sclerosis.

Chronic Cisternal or Basal Arachnoiditis. This is a condition in which the chiasmal syndrome may be present in the absence of a tumour. Holmes and Cushing both describe thickening of the soft membranes at the base of the brain with cystic formation containing fluid under pressure, the release of which may bring about improvement in vision or may fail entirely to do so. The fields in these cases vary from time to time in such a way that a retrobulbar or toxic neuritis might be suspected. Papillœdema is usually present, but in Sargent's two cases quoted by Holmes the discs were pale. In a series of thirty-three cases of cisternal arachnoiditis Horrax described several as having resulted from the extension inwards of an otitis media ; some have followed trauma, infected contusion of the scalp, ocular and naso-pharyngeal infections. Months after an unsuspected fracture of the base of the skull had taken place bilateral optic atrophy may supervene from the development of chronic basal arachnoiditis.

According to Hinds Howell, the first good description of arachnoiditis was given by Horsley in the *British Medical Journal* in 1909. Hinds Howell also agrees that head injury, with or without fracture of the skull, is almost certainly an established antecedent, as also is recovered meningitis.

It is well known that the pituitary gland enlarges during pregnancy, and may even project from the sella turcica, but it is only the

anterior lobe that is found to take part in this enlargement. Histologically Erdheim and Stumme found that the chromophobes or "Hauptzellen" were considerably increased; they were also enlarged and became the "Schwangerschaftszellen," or pregnancy cells. In spite of this enlargement the field of vision is rarely found to be contracted. McCurry examined the fields of vision in 70 cases of women during their last month of pregnancy. His results led him to believe that bitemporal contraction of the field of vision in pregnancy does not occur. On the other hand, Lohlein states that in 78 per cent. of all pregnant women bitemporal hemianopia is constant towards the end of pregnancy. He further states that

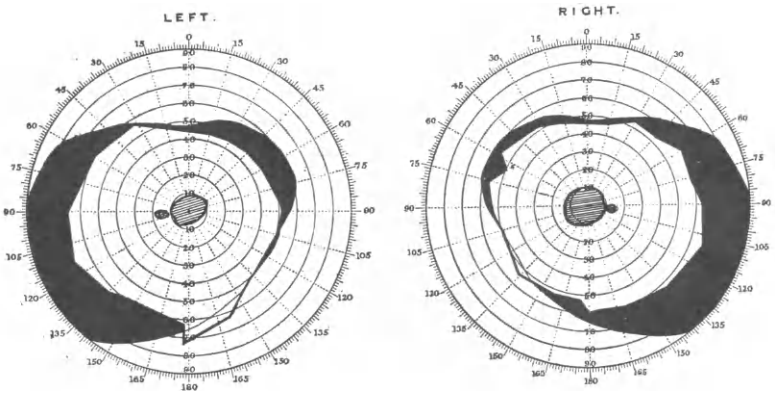


FIG. 100.—Fields of vision in a case of Leber's optic atrophy. (Burroughs.)

the increase in volume of the pituitary gland is chiefly from above downwards, that is, the pressure is on the posterior angle of the optic chiasma. This coincides with his "field of vision" findings. However, Shimkin, in his "Contribution à l'étude de l'hemianopsie bitemporale gravidique," says that after detailed study of the literature and on the basis of his own examination he has come to the conclusion that the bilateral hemianopia in pregnancy has to be attributed to a compression of the chiasma by the pituitary body hypertrophied during pregnancy, provided that the pituitary body of pregnancy is associated with a tumour of the hypophysis, which existed already before pregnancy in primipara, or has developed during and in connection with the pregnancy in the multipara. Where hemianopia was present, it was found that central vision was considerably lowered.

It will be remembered that in Chapter V it was stated that Herbert Fisher believed Leber's hereditary optic atrophy was probably due to swelling of the pituitary gland; this is strongly supported by radiograms of the pituitary fossa shown and described by Burroughs, who demonstrated the fields of vision of one case (Fig. 100) in which marked bitemporal contraction of the fields is present, together with a partial central scotoma in the field for each eye also. Since that time Lagrange has reported the

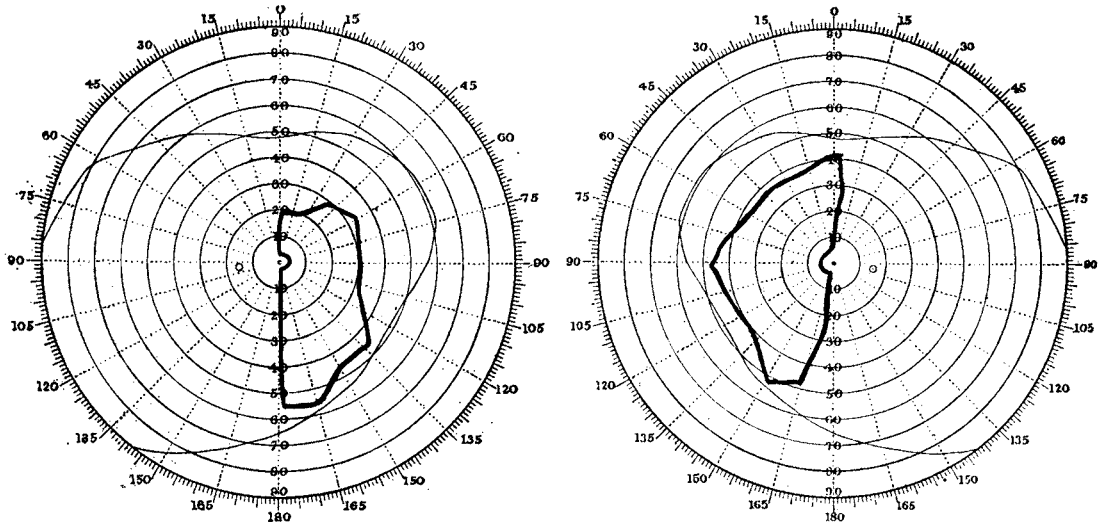


FIG. 101.—Fields of vision in a young woman, twenty-five years of age, small in stature, who had never menstruated and had normal mentality. In May, 1934, there was a partial primary optic atrophy in each eye with fields of vision as indicated: R.V. 6/60, L.V. 6/9. But by February, 1936, R.V. was 2/60 and L.V. *Nil*. (Case in charge of Neill Hobhouse.)

result of treatment of this disease by thyroid, and as the result of some success counsels the early adoption of organic therapy.

Signs and Symptoms of Pituitary Disease.—There are three theories which have been enumerated to explain the chiasmal syndrome. Fisher believed that traction exerted upon the optic nerve, chiasm and tract would account for the production of field changes. Cushing and Walker state that pressure and traction go hand in hand. It is probable that the essential factor is the production of arterial ischæmia and venous stasis. The return of vision has been demonstrated in the presence of a white optic disc, a

condition which would be impossible if neuronic degeneration had taken place. A case is mentioned by Cushing where, on exposure of the stretched chiasm and optic nerve, one optic nerve was severed at the optic foramen; there followed an immediate improvement in vision of the other eye. It is the removal of what Cushing calls physiological block which permits recovery of vision even in the presence of marked pallor of the disc. Toxins also can act upon the chiasm, producing similar changes to what are found when the optic nerve is affected by the presence of a plaque as in disseminated sclerosis—a chiasmal retrobulbar neuritis resulting.

The field defects found in lesions of the chiasm are of four types :—

(*a*) Bitemporal hemianopia with or without a scotomatous area. (Fig. 101.)

(*b*) Homonymous hemianopia.

(*c*) Blindness on one side with temporal hemianopia on the other.

(*d*) Irregular defects.

Fig. 102 shows the progress of the contraction of the field of vision in a typical case of bilateral hemianopia. A depression takes place in the upper and outer quadrant in one field, both for form and colour, the latter always being in advance of the former (1). As the depression increases the lower and outer quadrant becomes affected, and the field of the opposite eye begins to show a similar change (2). Soon half the temporal field is lost, and may or may not include the fixation spot (3). The temporal field of the other eye shows increasing contraction, but is not quite so advanced as the first; indeed, seldom are the fields symmetrical. In some cases a scotoma becomes evident just as the upper outer temporal quadrant has shown a depression. The scotoma lies outside the central point of fixation at the apex of the upper and outer quadrant, and extends upwards and outwards to meet the peripheral depression, finally breaking through. The scotomatous fields usually indicate that rapid changes are taking place.

The colour fields show a similar change, and always precede that for form. It is this gradual change for form and colour presenting a steadily enlarging hemianopia which contrasts sharply with the symmetrical and complete hemianopia produced suddenly by a suprachiasmal or cerebral lesion. Close examination of the field of vision in these types may show a small temporal island of vision

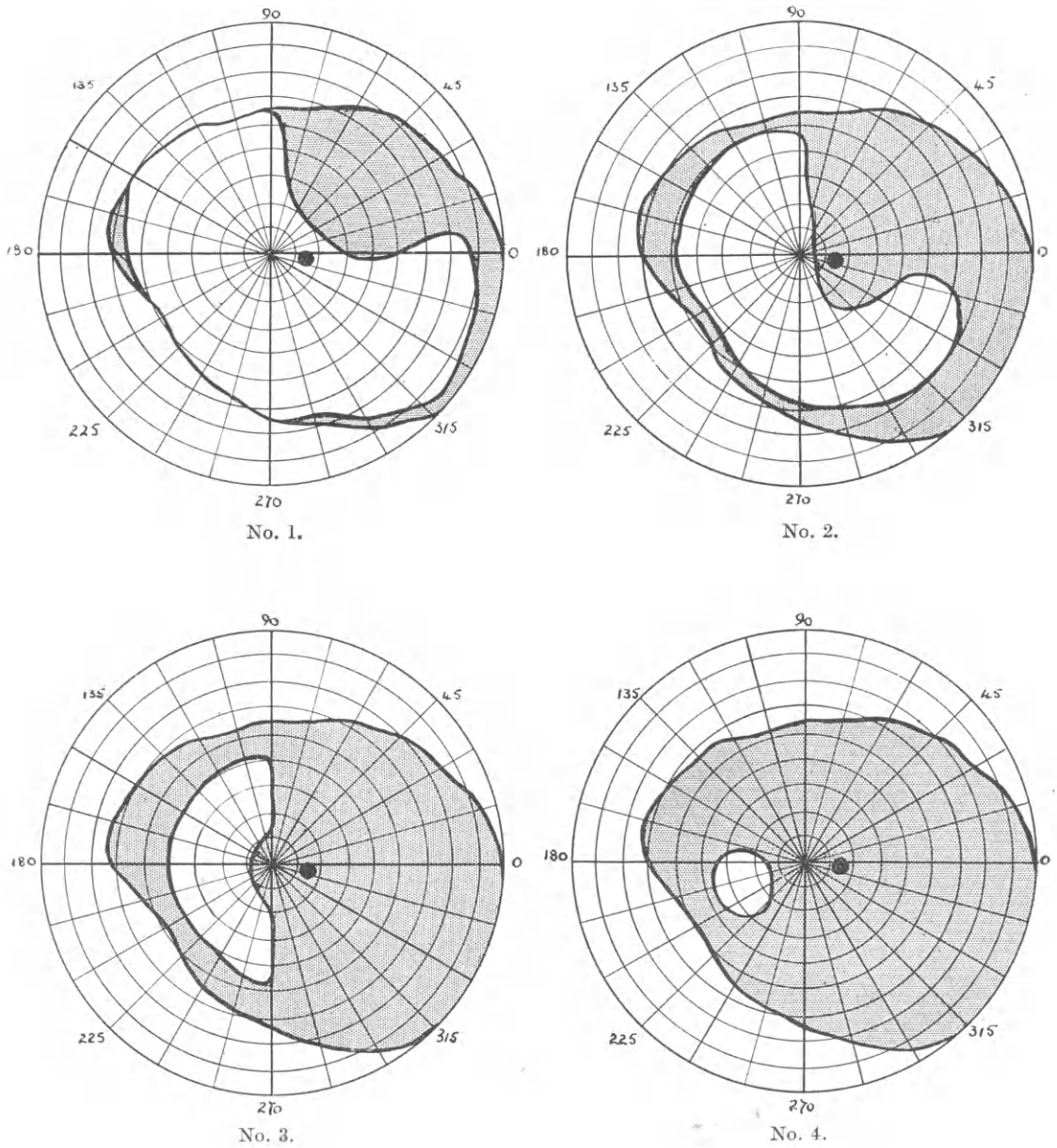


FIG. 102.—Four stages of advancement of a right temporal field defect.
(After Cushing and Walker.)

left at 50 degrees on the horizontal meridian (4). Its presence indicates the chiasm as the site of interference, and is not found when the lesion is prechiasmal or when the tumour is of slow growth in acromegaly. It is remarkable how such lesions spare the large bundles which proceed from the macular area. Often central vision is practically normal on one or both sides, although, as Holmes points out, testing with small coloured objects will often reveal a definite defect in central vision. There may be such colour defects without temporal loss, and so cases have been treated as retrobulbar neuritis when the possibility of a suprasellar endothelioma should have been borne in mind.

Homonymous defects were observed by Beckman and Kubie, but they were produced by hypophyseal stalk tumours. In some cases the typical bilateral temporal hemianopia has been observed to change to a homonymous form, a change which may indicate a halt in progress of the lesion with an improvement in the vision of one or both eyes (Traquair).

A ganglion neuroma of the third ventricle with diabetes insipidus and hypopituitarism has been described by Doyle and Kernohan. This tumour had protruded through the floor of the third ventricle and had invaded the optic chiasma, the intracranial portion of the optic nerves and the optic tracts, particularly the left. The girl, aged thirteen years, first complained of headache and progressive loss of vision, the field of vision showed a complete right hemianopia, due, no doubt, to the destruction of the left optic tract by the neoplastic process.

Many cases come before the neurologist with one eye blind and the other showing a temporal defect. Temporal defects also may begin in the lower quadrant and less commonly in the nasal quadrants.

Irregular defects frequently occur, such as a blind eye on one side with normal vision on the other, as in the case illustrated by Fig. 51. Remembering the anatomical arrangement of the fibres in the chiasm, it can easily be imagined that pressure on one part of the chiasm not in the middle line may produce almost any type of field. The macular fibres cross posteriorly in the chiasm, and so may be affected by a retro-chiasmal growth passing upwards and forwards, resulting in a central scotoma. If the lesion is more to one side than the other, then the temporal defect will be larger on the affected side. Again examining Fig. 51, one would have thought the resulting condition of the fields would be a temporal hemianopia

in the left eye and the right eye blind, but actually between the onset of the blindness in the right eye and the appearance of a papillœdema in the left several months intervened without change.

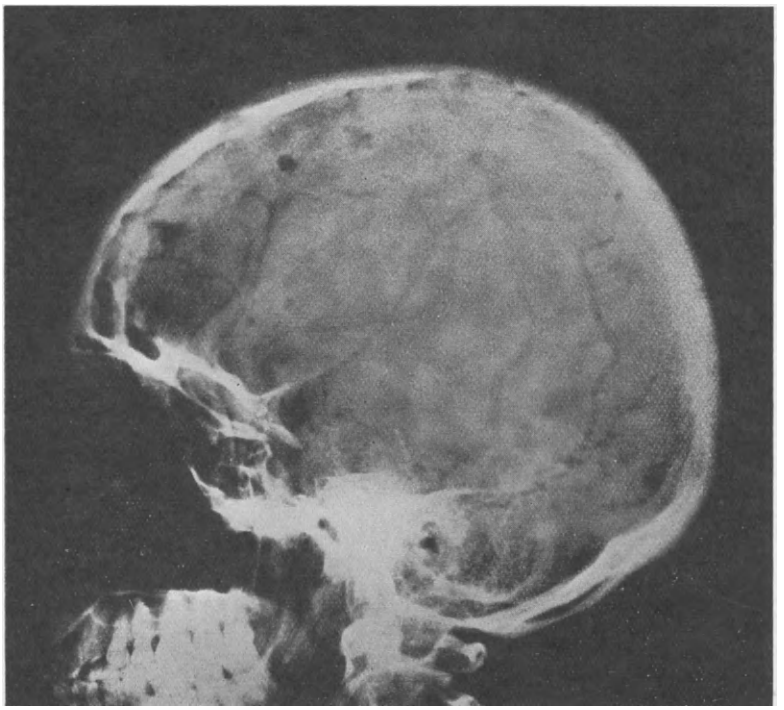


FIG. 103.—Skiagram showing enlarged pituitary fossa. Erosion has taken place beneath the anterior clinoid processes and at the apex of the petrous bone. Case of a young woman thirty years of age who showed symptoms nine days after the birth of her third child. There was a paralysis of the right sixth cranial nerve with paresis of right arm and leg, accompanied by frontal headache. One year later vision began to fail, first in the right eye and then in the left. After a further six months bilateral exophthalmos was observed and continued to increase. Recent examination showed patient to be absolutely blind. A consecutive optic atrophy was present in each eye and the surface of each disc was raised one diopter. (Skiagram taken by H. M. Worth.)

Binasal hemianopia is rarely seen. It is a possibility in the case of bilateral pressure on the chiasm by atheromatous internal carotid arteries. The condition is indeed rare. Zentmayer has recorded a case.

Recovery of the field has been recorded in many cases, the

recovery taking place in the reverse manner to the onset of the field defect. Blindness has given way to sight on release of pressure in many instances, the recovery on the whole being rapid. It may appear unnecessary to remind the reader that syphilis can simulate any growth in the brain, and a necessary procedure in the investigation of a case of raised intracranial pressure or change in the field of vision is to have the serological tests done before any operative interference is attempted (see Fig. 55, p. 136).

The optic disc usually shows a condition of atrophy, and may vary from the slightest shade of pallor to a marble whiteness. Cushing has seen tumefaction of the nerve head while in a state of optic atrophy, and Uhthoff enumerates 24 per cent. of cases showing a papillœdema without endocrine disturbances, and 11 per cent. with such change as acromegaly or distrophia adiposo-genitalis.

Exophthalmos may be produced by a downgrowth of the tumour, as in glioma of the chiasm invading the orbit, obstruction in the cavernous sinus to the venous return flow from the orbit, and also in sellar tumours there may be an exophthalmos resulting from expansion disturbances (see Fig. 103).

Involvement of the ocular nerves may take place, especially in lateral growths. De Lapersonne and Cantonnet suggest that the symptoms of hypophyseal lesions may be divided into (1) the chiasmatic syndrome—hemianopia, optic atrophy and diminishing vision ; (2) the syndrome of the external wall of the cavernous sinus in which the ocular nerves are mainly involved, beginning with the sixth, then the third, finally the fourth, together with neuralgic pain disturbances over the region supplied by the ophthalmic branch of the trigeminal. They suggest that in some of the latter cases the surgical approach might be endo-nasal. In the hands of Hirsch the mortality of the endo-nasal operation is 5 per cent.

In pituitary enlargement the third and fourth nerves are more commonly affected than the sixth nerve, as the latter is protected in this position by the internal carotid artery.

Atrophy of the optic nerves is usually accompanied by some degree of shrinking, but in the " glistening pallor " (Cushing and Walker) of the atrophy produced by pressure in pituitary tumour there is a suggestion that operative procedures may restore some degree of vision, whereas if left too late when true atrophy has set in, recovery of vision is impossible.

Among the many causes of headache should be remembered the type known as pituitary headache. It differs from the headache caused by brain tumours in that it is less intense and is less often accompanied by nausea and vomiting. The character of the pain is boring and continuous. The patient indicates that the pain may be occipital, frontal, bitemporal, or holding his head will say that the pain is in the centre of the head. The pain may simulate migraine accompanied by teichopsia (flashes of light) or scotomata. It was the latter type of headache which suggested to J. Herbert Fisher " that a periodic temporary swelling accompanying functional over-activity of the pituitary body explains migraine better than any other hypothesis."

Slight mistiness of vision, not an actual lowering of the normal (6/6) standard, but just a haze on looking at the test types, has been observed by de Schweinitz in the early stages. Also " blue mist " has been noticed by Fisher in the later stages of pituitary disease.

The centro-cæcal scotoma seen in chiasmal amblyopia resembles that caused by toxic amblyopia (Nettleship), but Traquair points out that the latter is not divisible into quadrants when examined quantitatively, as is possible in the former.

When the records of patients suffering from pituitary disorders are examined, it is found that quite a large proportion come before the surgeon when irremediable damage is done to the sight of one or both eyes. It therefore behoves the physician to bear in mind the early persistent headache, the occasional misty vision and the loss of temporal fields which may be discovered merely by the simple confrontation method of perimetry.

The roentgenology of the sella turcica is indispensable in the diagnosis of a pituitary tumour. Although the swelling of the pituitary body, such as occurs in adolescence, menstruation, and pregnancy, does not show on a skiagram, yet enlargement of the sella when it occurs is easily seen. The normal limits of the sella turcica and how to obtain them, according to Dr. Pancoast (quoted by de Schweinitz), are defined as follows : " The operator is careful to direct the rays through and on a line with the pituitary fossa, and to hold the plate at a distance of 36 inches, whereby the distortion is decreased to so small a minimum as to be of no moment. The operator next draws a line between the lower aspect of the clinoid processes and measures the greatest depth to the floor of

the sella and also the greatest antero-posterior diameter. In normal circumstances the measurement of the pituitary fossa varies from 8 to 9 millimetres in depth and 9 to 10 millimetres antero-posterior diameter in an average individual. A fossa depth of 6 to 7 millimetres and an antero-posterior of 7 to 8 millimetres represents the average measurement in the normal child from six to ten years of age. Skiagram of a normal adult pituitary fossa is shown by Fig. 104.

Hamby and Gardner describe the encephalographic characteristics in nine cases of suprasellar tumours confirmed at

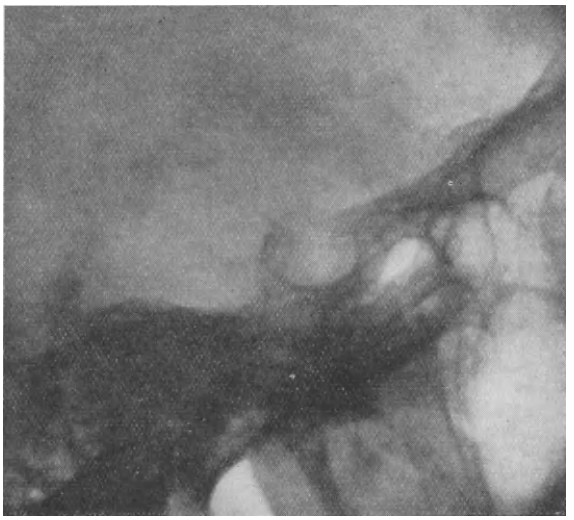


FIG. 104.—A normal adult pituitary fossa.

operation. The craniopharyngiomas showed an antero-inferior filling defect in the third ventricle with more or less obstruction of the foramina of Monro, with resulting hydrocephalus. The meningiomas were characterised by an outlying of the superior border of the tumour, with less compression of the third ventricle, but with filling defects in the floors of the anterior horns of the lateral ventricles. See also Sosman's paper (Bibliography).

Ocular Phenomena in Acromegaly.—Discussing his findings in 50 cases of acromegaly, Lillie could not find any departure from the normal X-ray picture of the sella turcica or the normal ocular conditions in 13 of their cases. In 21 cases X-ray examination

showed enlargement of the sella turcica, but with no ocular changes characteristic of chiasmal involvement. In 16 cases, however, definite ocular changes of this kind were found with enlargement of the sella turcica and erosion of the posterior clinoid. Sixty-eight per cent. therefore of all this series of acromegalic patients were normal from the ophthalmological point of view. Of 81 cases of pituitary disease exhibiting field defects, 14 were acromegalics.

Binasal Hemianopia.—Before concluding this chapter something should be said of the possibility of the production of binasal hemianopia, due to a lesion of the optic nerves as they join the

Differential Diagnosis of Pituitary Lesions
(After Walshe, Grinker and Cushing)

	Adenomas.	Endotheliomas.	Cranio-pharyngiomas.	Glioma of Optic Nerves and Chiasm.	Aneurysms.	Chronic Basal Arachnoiditis.
	Chromophobe. Chromophile Mixed.					
Age incidence	Adolescents and adults.	Middle-aged adults.	Childhood to middle age.	Childhood.	Adults (may occur at any age).	All ages depending on cause.
Fundus oculi	Primary optic atrophy.	Primary optic atrophy.	Papilloedema in early life, later generally optic atrophy.	Primary optic atrophy with or without swelling of nerve head.	Papilloedema slight or marked.	Papilloedema and primary optic atrophy.
Visual fields	Bitemporal hemianopia.	Bitemporal hemianopia.	Bitemporal hemianopia and irregular.	Irregular field defects.	Irregular field defects.	Bitemporal hemianopia and scotomata defects.
Pressure symptoms	Early headaches, later raised intracranial pressure.	Absent at first, later pressure on chiasm and optic nerves.	Early symptoms in childhood, severe in character. Hypothalamic symptoms.	Absent or late.	Absent or may suddenly become marked.	Slight.
Endocrine disturbances	Hypopituitarism. Hyperpituitarism (acromegaly). Mixed.	None.	Hypopituitarism.	None or hypopituitarism.	None.	None.
Situation	Sellar.	Suprasellar.	Suprasellar.	Suprasellar.	Suprasellar.	Suprasellar.
Radiogram	Enlarged sella.	Negative finding or slight flattening or absorption of clinoid processes.	80 per cent. are calcified and are well marked. May dilate sella.	Erosion beneath anterior clinoids and dilatation of optic foramina.	Erosion or none. Arteriogram.	None.

chiasm, or, indeed, of the chiasm itself. Such a condition is extremely rare. Reading over Johnson's paper on homonymous hemianopia, as seen in 49 cases of brain tumour, one does not come across any mention of binasal hemianopia.

In 1912 Cushing and Walker wrote a paper on this subject. They found 5–6 per cent. in a series of 300 cases of brain tumour. They stated that binasal hemianopia occurred as a late sequel of an advanced choked disc in the stage of recession of the œdema,

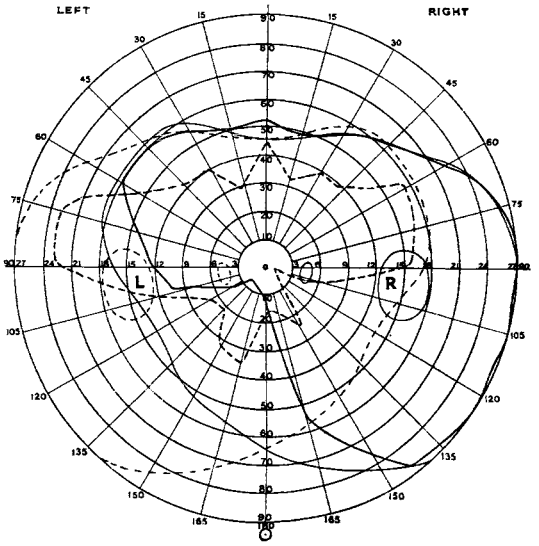


FIG. 105.—Fields of vision in a case of pituitary tumour which yielded to deep X-ray therapy. Considerable central vision returned, allowing patient to return to work.

affecting the fibres from the temporal portion of the retina more than those from the nasal side. They further stated that equal degrees of bilateral involvement imply, as a rule, a distant, often cerebellar lesion, with secondary hydrocephalus. An internal hydrocephalus with distortion of the third ventricle crowds the optic nerves downwards and outwards against the carotid vessels, which transversely indent the outer aspects of the optic nerves. Quoting Knapp, these writers also refer to those cases where there is the possibility of binasal hemianopia by the pressure on the lateral aspect of the optic nerves or chiasm by carotid arteries in a state of arterio-sclerosis.

Traquair states that unilateral nasal hemianopia without other defects in either field depends on a lesion of the uncrossed fasciculus where it is mixed with crossed fibres, and therefore not in the body of the chiasm.

Treatment of Pituitary Tumours by Deep X-ray Therapy.—The reports on the effect of radium or deep X-ray therapy on gliomatous tumours have not been encouraging (Roussy, Laborde and Levy), but there is a form of pituitary tumour that responds well to deep X-ray therapy. Two cases illustrative of this may be quoted. The first case, under the joint care of Worster-Drought and the author, presented himself in November, 1926. The vision of the

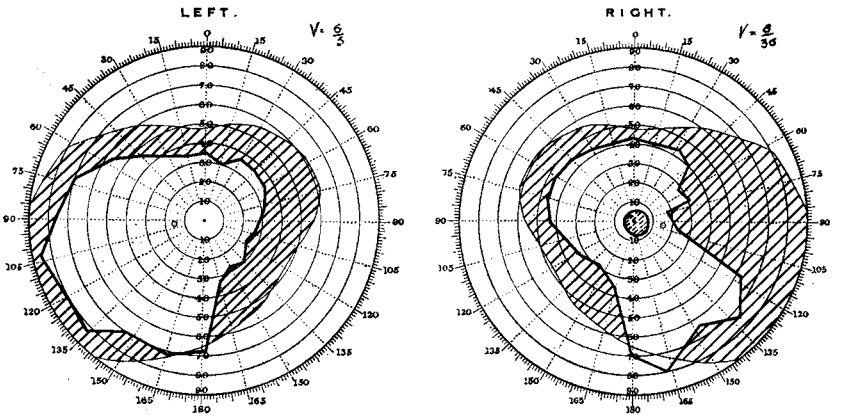


FIG. 106.—Fields of vision in a case of pituitary tumour at the beginning of treatment by deep X-ray therapy. R.V. 6/36, L.V. 6/5.

right eye was 6/5, and the vision of the left 6/36. In the right eye the disc was becoming pale in colour, while in the left advanced optic atrophy had taken place. The field of vision, as illustrated by Fig. 105, showing the presence of a bilateral inferior quadrantic defect, seemed to indicate some pressure on the chiasm. The skiagram of the pituitary fossa revealed a normal sella turcica. During the next few months eight deep X-ray exposures were given with the happy result that the fields of vision became stationary (the right was rapidly contracting). The central vision of the right remained 6/5, while that of the left rose to 6/9. One year had elapsed from the beginning of treatment, and for several years since the patient has been observed and has been found to show no further deterioration of vision.

The second case came under the joint care of Neill Hobhouse and the author. The case, that of a man aged thirty-nine, was first examined on December 1st, 1934. The optic discs appeared normal, but the central vision of the right eye showed a scotoma for form

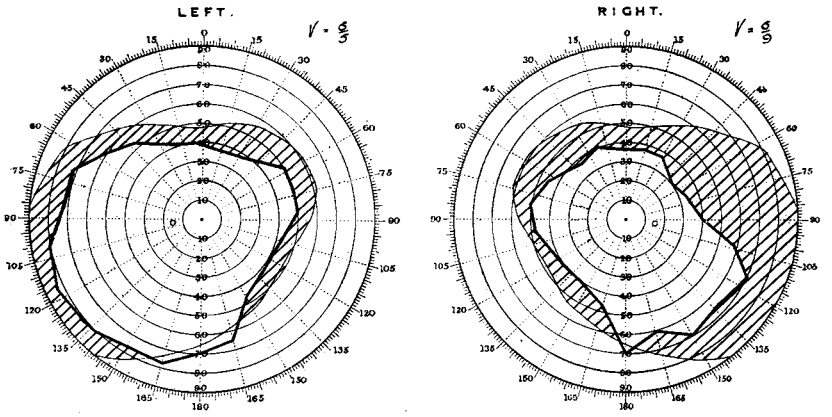


FIG. 107.—Fields of vision (same case as Fig. 106) at termination of treatment.

and colours, vision being equal to 6/36 ; that of the left eye was 6/5. The peripheral fields were contracted (Fig. 106), especially that of the left eye. Deep X-ray therapy was begun, and by July 11th, 1936, the date of the last examination, the central vision of the right eye had returned to 6/9, while both peripheral fields had expanded considerably (Fig. 107). This was probably a pituitary adenoma.

CHAPTER XIII

OCULAR MANIFESTATIONS IN DISEASES OF THE NERVOUS SYSTEM

Syphilis of the Nervous System.—Syphilis is one of the commonest causes of organic nervous disease. Possibly 15 per cent. of all diseases of the nervous system are due to the *spirochæta pallida*. The invasion of the nervous system takes place most probably by the blood stream, although some claim that the spirochætes pass by the lymphatic and perivascular lymph spaces. The dissemination by the cerebrospinal fluid must not be forgotten, as changes in the cerebrospinal fluid have been found as early as the first six weeks after infection. When early syphilitic eruptions make their appearance, rapid changes in the cerebrospinal fluid are detected, while in the late secondary syphilitic eruptions the highest percentage of involvement of the cerebrospinal fluid is found. Sixty per cent. show definite findings and 25 per cent. less marked changes. (The cell count is 15 or above; globulin, two plus; Wassermann reaction, four plus; and Lange's gold a definite syphilitic curve) (Dennie, Ueda).

The affinity shown by the spirochætes for the nervous system has awakened the suggestion that there is a special strain of the organism which is neurotropic in nature, while, on the other hand such forms of syphilis as meningo-vascular lesions are caused by the dermotropic variety. Since from day to day one sees cases of congenital syphilis showing gummatous ulcers, bilateral serosynovitis of the knee joints (Clutton's joints), gummata of the iris, etc., and knowing these cases to be the children of parents now suffering from tabes dorsalis, or having died from general paralysis, it is difficult to give credence to such a theory. What one and all know who have had extensive experience in dealing with syphilitic subjects is that deficient treatment in the early days of the infection will allow neurosyphilis to develop. Healey, from an examination of 169 cases, concludes that cardiovascular syphilitic lesions are the rule rather than the exception in the so-called general paralysis. Tabetics and general paretics, therefore, are not free from the liability of possible involvement with tertiary syphilis. Many of the

cases of general paralysis referred to had acquired their infection while on war service and had received a short course of " 606." Within ten years these cases relapsed badly with cardiac and nervous syphilis.

It has been demonstrated that when patients with the allergic gummatous type of late syphilis are inoculated subcutaneously with the syphilitic virus a gummatous-like lesion develops at the point of inoculation, but when patients suffering from parenchymatous neurosyphilis are similarly inoculated, no lesion of any kind results (Queyrat and Pinard).

Although it has been stated that allergy has no relation to immunity in syphilis, yet clinical studies have shown that the course of late syphilis may be influenced by the character of the secondary outbreak. Fournier found that late syphilis occurred when the secondary stage was mild, insignificant or absent, while severe secondary lesions appeared to protect the patient against the subsequent development of the late lesions, specially those of the central nervous system. A severe inflammatory reaction with relatively few spirochætes resulting in the formation of gummata may involve any part of the body ; such are termed allergic lesions. The patients who develop gummata of bones, skin or other organs are less likely to develop neurosyphilis or cardiovascular syphilis than are patients who do not show the allergic or gummatous type of reaction (Woods).

Neurosyphilis in third generation syphilis has been described by Nabarro. Among the cases he quotes one has shown local meningitis with changes in the liver and the spleen, together with a positive Wassermann reaction in the cerebrospinal fluid. Another showed Henebert's syndrome, *i.e.*, attacks of giddiness or nystagmus, either spontaneously induced by compression or aspiration of the air in the external auditory meatus, etc.

To satisfy critical investigation Elliott, while quoting a case, says : " For the diagnosis of third-generation syphilis Sams has given the following criteria :—

(1) The diagnosis of syphilis in the third generation should be based upon a specific lesion, definite stigmata or a repeatedly positive blood test.

(2) Acquired syphilis in one of the grandparents (first generation), preferably the grandmother, must be established.

(3) Congenital syphilis in one of the parents (second generation), and again preferably the mother.

(4) Exclusion of acquired syphilis in both parents and children so far as is possible.

(5) Certainty of parentage of the children in question."

The question has often been asked, "When is a syphilitic patient cured?" If we cannot look into the brain or visually examine the spinal cord we can at least see into the eye. The rate of progress of recovery of a syphilitic disease of the eye can be observed and recorded. Langendorff said: "The most reliable test of the severity of a disease of the eye is to note the degree of vision left after the culmination of this disease, and judged by this test, the value of consistent specific treatment stands out plainly."

I would refer my reader to the following two tables taken from

TABLE I

Untreated Cases

Cases.	Treatment.	Result.
4. R. and L.	Nil	R.V. 6/60, L.V. 6/60.
9. R. and L.	Atropine and Syrup Ferri Iod.	R.V. 6/12 partly, L.V. counts fingers.
15. R. and L.	Iron tonics	R.V. 6/36, L.V. 6/60.
22. R. and L.	Atropine	R.V. 6/24, L.V. 6/36 (one letter).
27. R. and L.	Atropine	R.V. 6/60, L.V. 6/24.
28. R. and L.	Hyoscine	R.V. 6/18 partly, L.V. 6/24 part
33. R. and L.	Atropine and tonics	R.V. 6/18 part, L.V. 6/60.
36. R. and L.	Atropine	R.V. 6/24 part, L.V. hand movements.
47. R. and L.	Nil	R.V. counts fingers, L.V. 6/18.
48. R. and L.	Nil	R.V. less 6/60, L.V. 6/24.
50. R. and L.	Nil	R.V. nil, L.V. counts fingers.
51. L.	Nil	L.V. 6/24. Symptoms of G.P.I.
54. R. and L.	Nil	R.V. poor perception of light, L.V. 6/18.
57. R. and L.	Nil	R.V. 6/60, L.V. 6/24.
59. R. and L.	Nil	R.V. 6/24, L.V. 2/60.
63. R. and L.	Nil	R.V. 3/60, L.V. no perception of light.
64. R.	Nil	Three years ago, now left is affected for first time.
66. R. and L.	Nil	Practically blind.
67. R. and L.	Nil	R.V. 6/18, L.V. 6/24.
68. R. and L.	?Hg.	R.V. 6/12, L.V. 6/9.
70. R. and L.	Hg. and Atropine	R.V. 6/9, L.V. 6/9 but scleritis supervened.
71. R. and L.	"Drops"	R.V. 6/9, L.V. became myopic—10D.
74. R. and L.	Nil	R.V. 3/60, L.V. 6/12.
77. R. and L.	Unknown	R.V. 6/9, L.V. less 6/60.
79. R. and L.	Unknown	R.V. less 6/60, L.V. 6/24.

TABLE II

Treated Cases

Cases.	Result.
6. R. and L.I.K. . .	Slight corneal haze in L.E.
7. R. and L.I.K. . .	R.V. 6/6, L.V. 6/6.
8. R.I.K. . . .	No report.
11. R. and L.I.K. . .	R. cornea clear, L. central haze.
12. R. and L.I.K. . .	No report.
17. R. and L.I.K. . .	R. cornea clear, L. central haze.
19. R. and L.I.K. . .	R.V. 6/9, L.V. 6/9.
20. R.I.K. . . .	Four months later V. 6/24.
21. L.I.K. . . .	No report.
23. R. and L.I.K. . .	R. cornea clear, L. almost clear.
24. R. and L.I.K. . .	R.V. 6/12, L.V. 6/18 post. synechiæ. This boy refused atropine.
25. R. and L.I.K. . .	R.V. 6/6, L.V. 6/6. This girl had recurrent I.K.
29. L.I.K. . . .	Cornea clear.
30. L.I.K. . . .	No report.
32. R. and L.I.K. . .	R.V. 6/6, L.V. 6/9.
34. R. and L.I.K. . .	R.V. 6/9 part, L.V. 6/6.
37. R. and L.I.K. . .	R. corneal haze, L. cornea clear.
38. R. and L.I.K. . .	No report.
39. R. and L.I.K. with ulcer	No report.
41. R. and L.I.K. . .	R. corneal haze, L. cornea clear.
42. R. and L.I.K. with L. recurrent . .	R.V. 6/12, L.V. 6/36 and still clearing.
43. R. and L.I.K. . .	No report.
44. R.I.K. . . .	R.V. 6/6 part.
45. R.I.K. . . .	R.V. 6/6
49. R. and L.I.K. . .	Continuing to improve.
52. L.I.K. . . .	Cornea clearing.
55. R.I.K. . . .	R.V. 6/6.
56. R. and L.I.K. . .	R.V. 6/6, L.V. 6/6.
58. R. and L.I.K. . .	Both corneæ *almost* clear.
60. R.I.K. . . .	R.V. 6/6.
65. R. and L.I.K. . .	Mercury improved the condition.
72. R. and L.I.K. . .	Referred to own doctor for treatment.
73. R.I.K. . . .	Recent case.
75. R. and L.I.K. . .	In two months vision of each eye was 6/24ths.
78. New case.	
80. New case.	
And so on to Case 91.	

the author's book on "Interstitial Keratitis." The first table shows the resulting vision of cases of untreated syphilis of the eye, the second the result of treatment given promptly and thoroughly from the beginning of the disease. In untreated syphilis of the eye

blindness is common, whereas in treated syphilitic cases the result is usually most satisfactory. Hutchinson and Jackson say 10 per cent. of congenital syphilitics acquire labyrinthine deafness, this condition supervening as the interstitial keratitis is disappearing. Of 300 cases of interstitial keratitis which I have treated, not one has acquired deafness ; also choroiditis, which is commonly present in untreated cases, is singularly absent in those who have received both arsenical treatment combined with mercury, bismuth and iodides. Those who are responsible for the treatment of either acquired or congenital syphilis should not be discouraged by refractory cases, but should persevere even over a term of years.

The initial doses of arsenical injections should be small and frequently repeated. Cases have been seen where treatment has been vigorously carried on for a time and then stopped ; many of these have shown the Jarisch-Herxheimer reaction. These cases are called neuro-recidides and depend upon suppression of the infection and defence mechanism by therapeutic interference in primary or early secondary syphilis, with a subsequent intensive reaction at the site of foci of surviving spirochætes in the central nervous system (Mackie).

Since writing the chapter on optic atrophy the author was requested to examine the eyes of a patient to whom the doctor in charge intended giving a further course of arsenical injections in view of a stubborn positive Wassermann reaction. The eyes were found to be normal in every respect, as also were the fields of vision. Twenty days later the patient presented himself again. In the meantime he had received two injections of tryparsamide with an interval of one week between each. Immediately after the second injection the patient discovered he had some peculiar change in his vision. Ophthalmoscopically the fundi presented the same appearance as at the previous examination, but the fields of vision were found considerably reduced (see Fig. 108). Johnston Abraham, who subsequently examined the case, said he had often seen reduced fields of vision following practically every kind of arsenical injection, but they generally recovered. He pointed out that what appeared to be a large gummatous ulcer on one tonsil was in reality due to mercurial poisoning. In the days before " 606 " was invented these mercurial ulcers were commonly found to be present. They were due to overdoses of mercury. Juler has reported a case of bilateral optic atrophy which rapidly

supervened after several arsenical injections, but, as I have stated in an earlier chapter, such a result has not been found once during the past ten years at the London Lock Hospitals.

Secondary neurosyphilis does not, as a rule, produce any ophthalmic signs or symptoms. I have seen the development of an Argyll Robertson pupil within a year from the date of infection, but this must be extremely rare. It has been stated that iritis occurring in secondary syphilis is an assurance against the eventual development of tabes or general paralysis, but from an extensive study of syphilis in the Austrian Army by Mattauschek and Pilcz it was concluded that an uveitis is of no prognostic significance, favourable

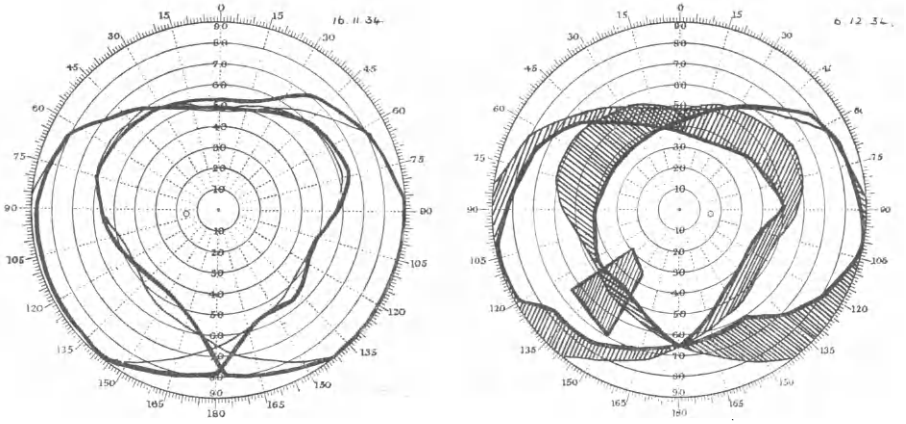


FIG. 108.—Fields of vision taken on Nov. 16th, 1934, and on Dec. 6th, 1934, respectively. Note reduction in peripheral fields due to two injections of tryparsamide given with one week between each.

or unfavourable, in regard to the subsequent development of parasyphilis.

However, according to Kauders, cases are found in which less than two years after primary infection basal meningitis caused cranial nerve palsy or choked disc with simulation of tumour and which responded promptly and satisfactorily to malarial therapy.

Of 4,134 officers suffering from syphilis and whose histories were followed from ten to thirty years 10 to 25 per cent. developed paresis, tabes or cerebrospinal symptoms (Mattauschek and Pilcz).

Tertiary Meningo-Vascular Syphilis

Syphilitic meningitis may be gummatous in character, and it may be the pachy- or lepto-meninges that are involved, either of

the base or convexity of the brain. Cranial nerve palsies are common, especially when the base of the brain is involved. The optic and oculo-motor nerves suffer most. The fifth, sixth and seventh nerves are liable to be attacked. Paralysis of the intrinsic and extrinsic ocular muscles may occur and vision may be diminished or lost. Papillœdema as well as headache is present. The pupils may become fixed rather than Argyll Robertson in type. The atrophic change in the optic nerve may result in the appearance of contracted fields, scotomata, hemianopia or blindness. The onset is usually rapid.

Hemiplegia

Syphilitic hemiplegia, described as early as 1497 by Leoniceno, generally comes on in young adults or in early middle age, generally about ten years after infection. The middle cerebral artery or any of its lenticular branches may be thrombosed, so that if the mid-brain is involved Weber's syndrome may follow, that is, paralysis of the oculo-motor nerve on the same side with hemiplegia on the opposite side (see p. 160). Pupillary changes may be present also. Pontine and medullary lesions may also be seen.

As papillœdema is a general accompaniment of brain syphilis, it is necessary to distinguish such a condition from raised intracranial pressure due to a brain tumour. The difficulty is that both may occur in the same patient at the same time. The symptoms of syphilis are generally opposite to those caused by a single focal lesion. Uhthoff states that in cerebral syphilis showing papillœdema 65 per cent. are due to a gummatous syphilitic tumour of the brain or meninges and 23 per cent. are due to gummatous basal meningitis.

Parkinsonianism as a syphilitic manifestation is seldom seen, but, if present, Argyll Robertson pupils may be found. In encephalitic Parkinsonianism the pupillary reflex defect is seen on convergence, and not to light as in the syphilitic condition.

Syphilitic hydrocephalus in the adult (Greenfield and Stern) is a fairly common result of basal gummatous meningitis. More often it is of the communicating type, although complete obstruction of the foramen of Majendie is not uncommon, and the foramina of Luschka may also be closed up by plastic meningitis. It appears that syphilitic hydrocephalus is definitely more common at the present day than gummatous meningitis associated with the presence of isolated large gummata. Greenfield and Stern point out that

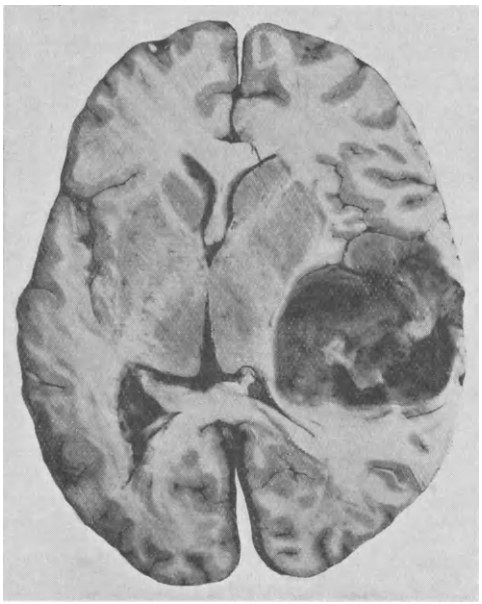


FIG. 109.—A large gliomatous tumour (glioblastoma multiforme) supervening upon an old partially encapsulated gumma in the right temporo-sphenoidal region and spreading extensively in right hemisphere. The case was that of a male aged fifty-one diagnosed two years previous to admission as a case of general paralysis. Admitted under Dr. Worster-Drought with the history of "obsessional neurosis." After a fit six months previously the patient became drowsy, gradually lost power in the left arm and leg for several weeks. Pupils were moderate in size, right less than left, and both irregular in outline. Both failed to react to light and consensual stimulation but were normal to accommodation. Left corneal reflex diminished. Patient died two days after admission with hypostatic pneumonia of both lungs.

The microscopical appearances vary extremely in different parts of the sections cut. Though extensive areas of necrosis are also present in the tumour itself, the appearances may be explained by the supervention of an actively growing glioblastoma multiforme in a "capsule" composed of proliferating neuroglia, consisting at some parts largely of fibrillary astrocytes. Chronic and recent vascular and perivascular changes—(small round-celled infiltration), chronic and subacute inflammatory (granulomatous) processes, all intermingled make up a very complex lesion. (Section prepared and description given by W. E. Carnegie Dickson.)

the loss of consciousness, attacks of stupor and reduction of intelligence suggest a more general affection of the brain than is found in diffuse gummatous meningitis, that headache and papillœdema are also probably caused by hydrocephalus, but that the cause of the cranial nerve palsies, on the other hand, is due to basal meningitis.

In syphilitic hydrocephalus ocular motor palsies are commonly found, and diplopia, defective upward movement of the eyes, inequality of the pupils and nystagmus are seen. Papillœdema was present in all their cases except one, and some had progressed to optic atrophy. Both papillœdema and optic atrophy were due rather to the general increase of intracranial pressure than to meningeal inflammation around the optic nerves.

Gumma of the Brain.—It is surprising to find that of the various neoplasms of the brain gummata form a very small proportion (Fig. 109). Sachs states that 0·5 per cent. of 800 intracranial operations were for gumma of the brain, while Cushing has stated that of his 2,000 patients with verified intracranial neoplasms only twelve cases of gumma were found. Looking back over a period of years, I have been impressed by the fewness in number of this type of neoplasm, and yet those I have seen stand out prominently in my memory. Probably it is due to the phenomenal result of treatment. In one case—a woman taken to hospital in an unconscious state—severely raised intracranial pressure was diagnosed, and before the result of the Wassermann reaction was known a large temporal decompression was performed. I saw the patient the next day. Severe papillœdema was observed in each eye, also a paralytic squint was present, while it was now revealed that the Wassermann reaction was strongly positive. Intensive treatment by means of neoarsenobillon injections, together with mercury and iodides, brought about a complete cure. The operation area healed well, vision returned to its normal standard, and in six months' time the woman was back to the factory able to earn a living, both for herself and her children. This case, however, is the exact antithesis of one described recently by Paterson and Leslie in the *British Medical Journal*. In their case there was a definite history of syphilitic infection followed by energetic treatment until the Wassermann reaction had become negative. Fourteen years later the patient had his first Jacksonian seizure, so again thorough antisyphilitic treatment was carried out, but the fits continued and the mental state deteriorated. X-ray examination of the skull was negative. A decompression operation was performed, and on opening the dura a breaking-down gumma was found at the upper part of the post-central gyrus. The gumma was enclosed in a firm capsule, and this was considered the probable reason why the antisyphilitic treatment had no effect on the growth.

Horsley was interested in the treatment of gumma of the brain, and thought that after a short period of antisyphilitic treatment, say six weeks, the case should be handed over to the surgeon.

It is possible for a patient to have a positive Wassermann and yet to be suffering from a non-syphilitic neoplasm of the brain. One case of the author's, that of a young married woman who showed the onset of optic atrophy and whose blood indicated the presence of the spirochæte, finally became blind. In spite of anti-syphilitic treatment the patient died. At the post-mortem examination a tumour which was not syphilitic, the size of a hen's egg, was found at the base of the brain.

Sometimes gummata are multiple and attached to the large vessels at the base of the brain. Operation in such a case would be useless, while medicinal treatment would be most probably followed by a cure.

Tabes Dorsalis.—The two commonest diseases of the nervous system met with in general practice are tabes dorsalis and disseminated sclerosis. Both are infections of the nervous system. There is no doubt of the nature of the infection in tabes dorsalis. Fournier, Noguchi and others have established the relationship of the spirochæte to this disease, while in the case of disseminated sclerosis there is still a doubt as to whether it is toxic or infective in nature. Both diseases exhibit numerous ocular signs and symptoms.

The Optic Nerve in Tabes.—In tabes we find 20 per cent. show primary optic atrophy. This atrophy, generally beginning in the pre-ataxic stage, is steadily progressive, the progress being indicated not only by the whitening of the disc, but by the gradual decrease of the field of vision. The atrophy may appear fifteen or twenty years before the locomotor symptoms begin. The field of vision shrinks until finally the macula is involved and total blindness ensues. Fig. 33 illustrates a typical contraction of the field of vision in an eye which had suffered from the gradual onset of primary optic atrophy for almost a year. The optic atrophy appears in both eyes, but, like cataract, the state of one is usually well in advance of the other. The author has had several tabetic patients whose first complaint was that of failing vision. The optic atrophy is always of the primary variety. The edge of the disc is quite sharp, the physiological pit, if present, is not obliterated, and the lamina cribrosa is distinctly seen. The surface of the disc is pale, sometimes quite white, but in others it is grey in colour. The vessels are

usually reduced in calibre. In juvenile tabes exactly the same condition may be seen. Indeed, optic atrophy is very common in juvenile tabes, just as ataxia is comparatively infrequent in these cases. It does seem a remarkable coincidence that if in tabes dorsalis primary optic atrophy supervenes early, the patient does not suffer so severely from motor paralysis, while in many cases where there is ataxia vision remains normal for a considerable time. If the atrophy commences after the onset of the ataxic gait there seems to be an improvement in co-ordination as the optic atrophy progresses.

The appearance of optic atrophy is shown by Plate VII. Irregular pigment is never found around the disc, as in secondary forms, nor are whitened perivascular sheaths ever found around the vessels proceeding to and from the disc. The student should endeavour to become familiar with the appearance of the normal optic disc by examining the eyes of as many healthy individuals as possible. To miss the discovery of the slightest pallor of the disc is indeed a serious error. It is often a recognition of such a slight change which brings about the discovery of such a disease as tabes. Both the ophthalmologist and the neurologist commonly see tabetic patients who have been treated for months for various ill-defined symptoms, and yet one glance at the disc would have straightway indicated the disease.

The extent of the field of vision is governed by the amount of atrophy of the optic nerve which is present. The commonest form of field of vision is that showing general concentric shrinkage, the colour fields for red and green being early lost. Sometimes irregular sectorial defects, which gradually spread, are found, but the former, in the writer's experience, are much more numerous. A more minute account of the changes in the field of vision in tabes dorsalis is given by Leslie Paton in the *British Journal of Ophthalmology*. He quotes Uhthoff, who distinguishes between two main classes of defect. (1) The whole field seems to suffer simultaneously with an early loss of colour fields and early loss of visual acuity. There is a relative peripheral contraction of the fields for white, but a full field may still be obtained in the course of an ordinary investigation, *e.g.*, with 10 millimetres white at $\frac{1}{2}$ metre distance. (2) The area of defective visual fields is sharply delimited from unaffected areas which show full normal vision, and central acuity may be quite good.

Stargardt's more elaborate classification includes :—

(1) Peripheral loss of white and colours with simultaneous loss of function in other parts of the field. With advancing peripheral loss the visual acuity falls and at the same time the distinction between red and green and later blue and yellow.

(2) Peripheral loss for white and colours with good function in the untouched fields.

(3) Peripheral loss of colours with full fields for white. Visual acuity may be normal or diminished.

(4) Partial (sectorial) loss with more or less perfect functioning in the rest of the field.

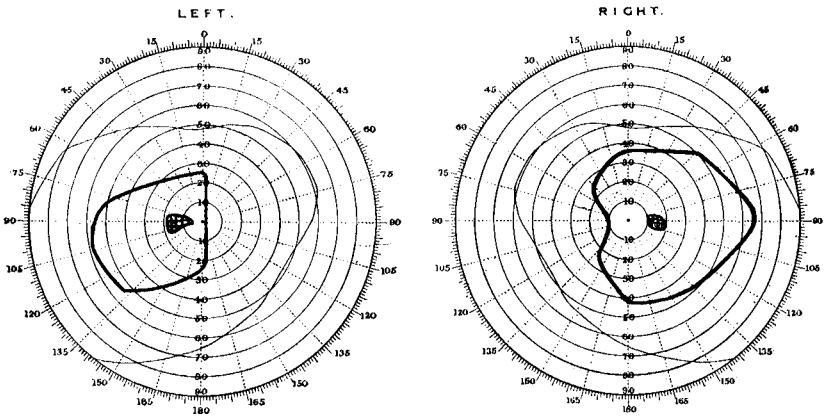


FIG. 110.—Binasal hemianopia associated with primary optic atrophy in a case of tabes dorsalis (Drake).

(5) Normal limits to the peripheral fields with diminished visual acuity and colour sensation.

(6) Central scotomata.

(7) Hemianopic defects (Fig. 110).

From the author's experience scotomata are very rarely found. It is stated that they are found in 2 per cent. of cases (Uhthoff). As mentioned in Chapter VII, neither Batten nor the author has ever found degeneration of the macula present as the result of syphilis. The papillomacular bundle is not a special site of election for those processes which produce atrophy of the optic nerve as a whole in tabes dorsalis (Fig. 111).

Although both homonymous and bitemporal forms of hemianopia have been described, these are rarely seen ; indeed, a good many

observers, including Uhthoff, do not regard hemianopia as a pure tabetic manifestation. Traquair says that "in the rare cases in which indications suggestive of hemianopia are found careful examination by quantitative perimetry is necessary to show whether signs of tract or chiasmal interference are really present, or whether the condition is merely one of homonymous or bitemporal depression simulating true hemianopia only in position and shape."

Cases of primary optic atrophy associated with binasal hemianopia show rapid progression of the atrophic process to complete blindness (Drake).

There have been many theories put forward as to the cause of optic atrophy in tabes dorsalis. Gowers and others thought that

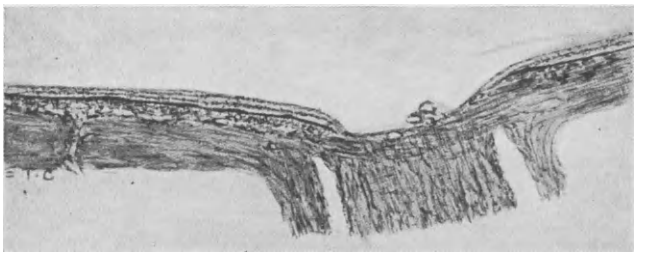


FIG. 111.—Microscopical section through the head of the optic nerve in a case of simple atrophy such as occurs in tabes or hereditary optic atrophy. The size of the nerve is reduced and a depression has formed on the inner surface of the lamina cribrosa in the position normally occupied by the optic papilla. (Collins and Mayou.)

the atrophy began in the ganglion cells, while Uhthoff stated that the atrophy began in the nerve fibres or axones. Such a theory would be defined as an atrophy produced by primary neuronic degeneration with secondary fibrosis, while others, such as Elschnig, Willband and Saenger, have thought the changes began at the papilla, or optic disc—atrophy caused by a primary peripheral and interstitial neuritis with secondary degeneration of nerve fibres. Leslie Paton has shown that interstitial changes and the parenchymatous changes in the nerves are co-ordinate results of the activity of the tabetic virus and not mutually dependent on one another.

Optic atrophy is often the first sign of tabes before the loss of tendon reflexes, the Romberg sign or other tabetic symptoms. The only other symptom which is likely to precede atrophy is the Argyll Robertson pupil.

The *pupillary changes* consist of the Argyll Robertson reaction, miosis or narrowing of the pupils, irregularity of pupillary outline and unequal pupils. The Argyll Robertson pupillary reaction, however, is by far the commonest ; in 70 to 90 per cent. of tabetics this reaction is found. It is generally bilateral, but occasionally unilateral (3 per cent.). Gowers states that 15 per cent. of tabetics showed loss of both light and accommodation-convergence reflexes, but this figure is undoubtedly too high. I have discussed the Argyll Robertson reaction in Chapter II, where I have pointed out McGrath's valuable contribution to this subject. It is worth while in every case examining the stroma of the iris by means of a magnifying glass.

Paralyses of both intrinsic and extrinsic ocular muscles occur. The ocular paralyses found in tabes dorsalis show the third cranial nerve to be chiefly affected—20 per cent. according to Parsons, while 13 per cent. show the sixth cranial nerve and 3 per cent. the fourth to be involved, while there is a total ophthalmoplegia in 2 per cent. The patients suddenly complain of seeing double. This is the first and earliest sign of the onset of paralysis of one branch of the third nerve. It may recover temporarily and, as a rule, does not spread to the other branches until a complete nerve paralysis occurs. If the third cranial nerve is completely paralysed the upper lid hangs down—ptosis. When the lid is raised the eyeball is seen to be rotated downwards and outwards. Examination of the movements of the eyeball shows that only two muscles are capable of movement—the external rectus, which is supplied by the sixth cranial nerve and which rotates the eyeball outwards, and the superior oblique muscle supplied by the fourth cranial nerve, which rotates the eyeball downwards and outwards, while the pupil is dilated owing to the unopposed action of the sympathetic. Fleeting diplopias have been described in the early stages of tabes. Sherrington has suggested that these may be due to involvement of the afferent proprioceptive fibres from the oculo-motor muscles and the muscles of the head and neck. Gordon Holmes lays emphasis on the appearance of ptosis, or drooping of the lids, as an isolated symptom apart from palsy of any of the external ocular muscles. He says there are few cases which do not present it to some degree. The upper lids droop equally, or the one perhaps more than the other, but at first they rise to the full extent or almost so when the patient looks up. In order to compensate the ptosis

the frontalis muscle is permanently contracted and the forehead is therefore wrinkled. Largely to this is due the " tabetic facies," by which many cases can be recognised at the first glance.

Paralysis of a nerve supplying an extra-ocular muscle may be so slight that the paresis can only be discovered by examining the muscle balance by means of the Maddox multiple rod. The degree of paresis can be measured accurately by looking at a lighted candle placed 6 metres away. The flame of the candle is seen through the Maddox rod held before the right eye, while the candle is seen naturally with the left eye. If the red bar of light moves to the left it means that there is a slight divergence of the visual axes while in a position of rest—exophoria. But if the bar moves to the right then the visual axes are converging—esophoria. The prism required to superimpose the red bar on the light of the candle is the measure of the paresis of the muscle affected. It is less frequently found that the sixth cranial nerve is paralysed. Where such is the case it will be found that the external rectus muscle cannot rotate the eyeball beyond the middle line. It is at this point that the patient complains of seeing double when a pencil held vertically before the eye is passed to the outer side of the field of vision. The farther away from the middle line the greater the distance between the two images of the pencil.

It must be remembered that syphilis can cause atrophy, both primary and secondary, of the optic nerve, so that optic atrophy must not be looked upon as an invariable sign of tabes. Syphilis can produce degeneration of the optic nerve analogous to the destruction of the posterior columns of the spinal cord, while meningo-vascular syphilis may cause an optic neuritis ending in atrophy.

As Adler has pointed out, vision may remain normal for months or years after the appearance of primary optic atrophy, yet a patient being aware of loss of vision will often ask, " How long will it be before I am blind ? " It is generally believed that loss of sight will take place in a year or two, yet the " two-year " rule does not always hold. I am quite certain from experience that by the diligent use of mercurial inunction, combined with the iodides and intravenous bismuth, loss of vision can be delayed for at least a further twelve months.

Suker and Jacobsen have stated that in tabetic optic atrophy intracranial injections of mercuric chloride have given better results than any of the other methods used. The method of choice

is the intraventricular injection as early as possible. They suggest, therefore, that every patient with syphilis of any number of years' standing should be critically examined in order to determine the absence or presence of involvement of the central nervous system.

MacCormac has recently stated that it is now well known that in all forms of recent syphilitic infection involvement of the central nervous system, as shown by the changes in the cerebrospinal fluid, is relatively common, the central nervous system participating in the general infection. This accepted fact permits of two conclusions, first that the spirochætes in the central nervous system can be and are reached and destroyed by intramuscular and intravenous treatment ; and secondly, as one is entitled to infer from the above, that the specialised technique which claims to influence syphilis of the central nervous system by introducing therapeutic substances directly into the cerebrospinal fluid is unnecessary, and presumably much less efficacious than the intramuscular and intravenous routes.

In the treatment of neurosyphilis Lees, in his Presidential Address to the Society of Venereal Diseases, states that the use of iodides in commencing doses of 15 grains, in conjunction with mercury, for the first few weeks should be employed. As stated above, the author believes the postponement of complete primary optic atrophy for a considerable time can be accomplished by the diligent use of both these drugs. Bismuth given by the intramuscular route is favoured by some who find it easier to handle than mercury. When pain is a prominent symptom, the intraspinal route is indicated, although few neurologists at the present day advocate the Swift-Ellis treatment. Reece still employs this method in rapidly advancing meningeal involvement with severe crises and optic atrophy. I have seen Sir Charles Ballance insert a cannula into the lateral ventricle and wash out the cerebrospinal fluid through a lumbar puncture, followed by the replacement of the fluid by N.A.B. This was in an extremely acute case of central nerve-system involvement. As in ophthalmology, so in neurology, novarsenobillon is probably the most popular arsenical preparation used. In the case of children suffering from congenital syphilis, if the state of the veins makes the injection of N.A.B. difficult, sulpharsenol can be safely injected into the buttock. From an extensive experience of the use of this drug, the author would readily commend its use.

The use of tryparsamide by neurologists is growing in popularity.

It has been found that the pentavalent group of arsenicals, to which tryparsamide belongs, penetrates the nervous system much more easily than does the trivalent. Stovarsol, better known as acetarsone, belonging also to the pentavalent group, has been in extensive use in France and has been recommended by various authorities. Some believe that a previous course of N.A.B. enhances the effect of tryparsamide. The weekly dose of the latter is usually three grams, also it should not be forgotten to include in the treatment both mercury and iodides at the same time as the arsenical injections. From the ophthalmological point of view, to rely on mercury and iodides alone is to allow the spirochete time to destroy the vision, whereas a rapid result is obtained by the immediate use of N.A.B. It is well to follow up the malarial treatment of general paralysis by tryparsamide also. The greatest danger in the use of the latter drug is the production of optic atrophy. The field of vision is at first concentrically reduced (Lazar), but this will recover if the optic nerve fibres are not damaged. Lees found from 3 to 10 per cent. of cases showed visual impairment upon full doses of tryparsamide being given. If there is any sign of visual impairment due to a recognisable optic atrophy, then tryparsamide is contra-indicated. As optic atrophy is less common in general paralysis than in tabes this drug may be more safely employed in the treatment of the former.

It may be a help to those who wish to decide between the use of mercury or bismuth to read Cannon and Robertson's paper in which these authors suggest that both metals have their place in anti-syphilitic treatment. Mercury can be given advantageously to robust patients. To the less healthy ones bismuth offers a valuable substitute. They think arsphenamine delivers a strong initial attack on the spirochæta pallida and should be reinforced by one of the heavy metals. Finally to prevent the parasite becoming drug-fast alternating the two metals is of decided advantage.

Those who are specially interested in the general treatment of syphilis may be referred to the *League of Nations Quarterly Bulletin of the Health Organisation*, Vol. IV, 1935. Here, however, we are considering especially neurosyphilis. Fever therapy has been used since Wagner-Jauregg in 1918 introduced malaria as a treatment for dementia paralytica. Since then vaccines, foreign proteins and sulphur compounds have been used for the production of fever.

The results of the treatment of tabetic optic atrophy by sulphur

have been recorded by Winkler. Acting with Dr. Fried they gave firstly an injection of bismuth (10 per cent. suspension of subsalicylate of bismuth) and the preparation " Pentabi " twice a week, the first bismuth injection being half the dose of the following one. With the second bismuth injection sulphur is administered also, using 0·2 c.c. colloidal sulphur preparation in a weak concentration (5 mgm. sulphur in 1 c.c.). This preparation is called " Schwefeldiasporal intravenös," manufactured by Chemisch Fabrik Dr. Klopfer, Dresden, and is injected intramuscularly at the same time with bismuth. The next injection is due four days later and is continued once a week. After the third injection it is advisable to use 1 per cent. suspension of flowers of sulphur powder (sulph. præcipit.), taking care not to exceed the elementary sulphur quantity of the last injection. Four to six fever reactions with a temperature of 101° to 103° are absolutely necessary. The treatment must not be repeated in the same year.

Clark has used malaria in the attempt to retard or control the atrophic process in the optic nerve. Of twelve cases he says eight have showed improvement.

By using a simplified, air-conditioned cabinet (the Kettering hypertherm) Culler and Simpson have been treating ocular syphilis with marked success. They maintain that the one factor common to all forms of fever therapy is the production of sustained fever and this, they say, is satisfactorily produced by the above apparatus.

Whatever treatment is used for syphilis none is adequate unless it brings about a change from positive to negative in the Wassermann reaction of the blood.

Congenital Neurosyphilis

Congenital neurosyphilis occurs both in meningovascular and parenchymatous forms. Transmission is probably by way of the placental circulation although cases have been described where the child was syphilitic and the mother non-syphilitic. It has been the author's practice for years to ascertain the serological reactions of the mother when a case of interstitial keratitis in one of her children has appeared before him. In not one single instance has it been found that the mother's Wassermann reaction was negative and so, whenever possible, the mother has had specific treatment too, along with the child. The Wassermann reaction, however, of the children varies enormously. In 64 of the author's cases where the interstitial

keratitis had just manifested itself 100 per cent. showed a positive Wassermann reaction, but in 8 old cases where the disease had run its course more than ten years previously, the Wassermann reaction was negative, although in 5 of these a luetic curve was obtained by Lange's Colloid Gold test. A description of this test was published by Bigland in the *Lancet* of Oct. 2nd, 1920, and by Lange in the *Berlin Klin. Woch.*, May 6th, 1912.

Navarro prefers a flocculation test such as the Kahn to the Wassermann reaction in congenital syphilis.

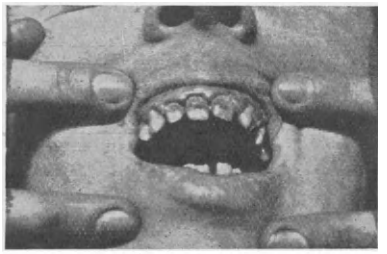
Involvement of the central nervous system in hereditary syphilis is relatively uncommon according to Jeans, and by statistical study there is no difference in the incidence of central nervous system involvement in hereditary and in acquired syphilis. The finding of an increased number of cells, globulin, albumin and a positive colloid gold reaction though important has not the same significance as the Wassermann reaction. Of 78 infants (under two years) examined, the serum of all but one was strongly positive, the cerebrospinal fluid in 31 was positive ; all of the latter had clinical signs of hereditary syphilis. Of 66 latent infections 20 per cent. had a positive cerebrospinal fluid Wassermann reaction. Of 70 actively syphilitic patients (from two to fourteen years of age) 22 (or 31 per cent.) had a positive cerebrospinal fluid Wassermann reaction. Some of the children showed meningitis with or without convulsions, also convulsions classified as epilepsy, hydrocephalus, hemiplegia, spastic quadriplegia, spastic paraplegia, optic atrophy five times (twice with associated lesions) ; fixed pupils without optic atrophy were seen four times. The nervous system was involved in 40 per cent. of the syphilitic infants and 31 per cent. of the older children.

In passing it may be mentioned that not only in acquired syphilis but in the congenital form may septicæmia occur. I have had a case of bilateral interstitial keratitis (in a boy aged fourteen years) who showed a positive Wassermann reaction and suffered so severely that he had to enter a nursing home. His temperature rose to 105°, his throat was foul and his knees became enormously swollen. Full specific treatment was steadily given resulting in restored health and eyesight.

Whether hereditary or acquired, Jeans says the earlier the treatment the better, and in the early stages intravenous or intramuscular medication should be given.

Hassmann and Meier have drawn attention to the fact that

formerly cutaneous manifestations of congenital syphilis were more common than evidences of nervous system involvement, but latterly there has been a decided increase in the neurological manifestations of congenital syphilis and so not only the public, but the medical profession are inclined to overlook such cases until in later years the disease has broken out in some complaint of the nervous system. They point out that while neuro-syphilis was not demonstrable at birth and was comparatively rare between one and three years of age, it reached the appalling frequency of 85·7 per cent. between the ages of three and six years.



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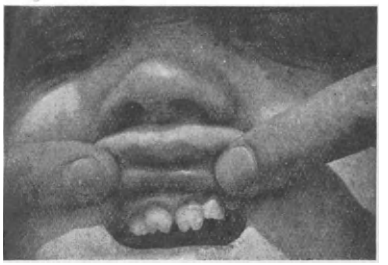


(*b*)

FIG. 112.—Two types of teeth seen in congenitally syphilitic children :—
(*a*) Peg teeth widely separated from each other.
(*b*) Notched teeth (Hutchinsonian).
(Rea's Interstitial Keratitis.)

It is usually stated that infection of a fœtus takes place about the fourth or fifth month of pregnancy, but the author believes infection takes place much sooner ; the anlage of the permanent teeth is laid down in the second month and it is in these teeth that the typical notching and peg-formation are found. Binary syphilis has been described by Harrison, quoting Hoffman and Schilling, who observed an infant which died seven weeks after birth, whose bones and organs showed typical syphilitic manifestations and yet had a primary sore rich in Sp. pallida on the scalp.

The common stigmata of a congenital syphilitic child should be known ; these are—the depressed flat bridge of the nose, scarring or ulceration at the angles of the mouth (rhagades), snuffles in the young child, notched (Hutchinsonian) teeth or peg-shaped teeth (Fig. 112) ; probably the corneal scars of an old interstitial keratitis are present, and Clutton's joints. Graves thought a scaphoid shaped scapula was pathognomonic, but much more so is the exostosis of the external end of the clavicle diagnostic of congenital syphilis

(Higoumenakis). When examining a case of interstitial keratitis the author always looks for evidence of bilateral sero-synovitis of the knee-joints (Clutton's joints) which is extremely common in congenital syphilis. It should be remembered also that bilateral syphilitic affection of the eye, such as interstitial keratitis, disseminated choroiditis and retinitis, is indicative of congenital syphilis, whereas the unilateral affections are due to the acquired disease. A case, for example, was that of a young woman who came complaining of " congenital syphilis " and who had received treatment for same at a large general hospital. She showed, however, the presence of a widely disseminated choroido-retinitis *in one eye* and also had been treated with " black ointment " for swollen inguinal glands, neither of which conditions in the author's experience is commonly found in congenital syphilis. Close questioning revealed the fact that the girl at the age of eight years had been raped and so the blame could not be laid at the door of her parents any longer. I have never seen a case of squint (apart from tabes) due to paralysis in a congenital syphilitic. If the squint is present it is due to a refractive error. A unilateral, highly myopic eye is often seen in congenital syphilis as a sequel to stretching when the eye was attacked by keratitis, choroiditis or scleritis, or a combination of all three. Lacrimation is common and is due to destruction of the lacrimal and ethmoidal bones before birth.

The one outstanding fact about the treatment of congenital syphilis is the tremendous improvement in the general constitution after thorough specific treatment ; the toxic appearance is lost, growth is stimulated and healthy babies are born to young mothers who in earlier life have had such thorough treatment. But in comparison to this, how discouraging the prospect of cure or even benefit in children showing parenchymatous or late meningovascular involvement of the central nervous system. Treatment may be given until the blood and the cerebrospinal fluid are negative in every respect, yet by such, sight has never been brought back to a case showing the presence of complete optic atrophy.

The ophthalmologist is not likely to see as many cases of the meningovascular form of congenital syphilis as the neurologist, although the number of the meningovascular exceeds the parenchymatous form. Hydrocephalus, convulsions, mental deficiency and plegias are seen. In connection with these there may be optic atrophy due to involvement of the tissues of the eyeball, or, as in

many cases which have been observed where the retina is quite sound and normal in appearance, yet complete atrophy apparently has followed a syphilitic meningitis. In such cases treatment is unavailing as regards restoration of sight; the same also has been found in regard to deafness in similar cases. There may be pupillary changes such as loss of reaction to light or both to light and accommodation.

Juvenile Tabes. Juvenile tabes is more commonly found than general paresis, the proportion being about 10 to 1. It occurs in congenital syphilitics or in those who have acquired syphilis in infancy or early childhood. According to Jeans males and females are equally affected. It begins somewhat later in life than general paresis, usually about the end of the second decade or may be as late as the thirtieth year with rarely any signs of congenital syphilis in preceding childhood. Although the parents of juvenile tabetics or general paretics are generally tabetics or general paretics, yet this distinction does not follow any known hereditary law. There is no sign or symptom in adult tabes that has not also been described in juvenile tabes; there are certain peculiarities which distinguish the one from the other—first, the essential chronicity of juvenile tabes, secondly, the frequency of vesical symptoms, headache, optic atrophy, Argyll Robertson pupils, oculo-motor paralysis, lightning pain and sensory alterations, thirdly, the frequency with which the affection is associated with general paralysis (Mott's tabo-paresis).

Not only does one observe the presence of Argyll Robertson pupils, but the entire absence of pupillary reaction is sometimes seen—the " frozen pupil " of Grinker. The pupils may be unequal and irregular also. Ataxy is rare and when present is slight and seldom extends to the upper arm. Crises and trophic changes appear to be uncommon in juvenile tabes.

The association in a syphilitic child of bilateral, simple optic atrophy with loss of knee jerks, would, by itself, point to the provisional diagnosis of juvenile tabes.

Juvenile Paresis. This also occurs in congenital syphilitics and develops, as a rule, between the ages of from seven to fifteen or it may be delayed until after thirty years of age. Owing to the patient's undeveloped state the mental symptoms usually resemble those of imbecility and are rarely hallucinatory or maniacal, rather they are taciturn and apathetic (Grinker). The physical signs are almost identical with those of acquired general paralysis and there

are in addition the stigmata of congenital syphilis. I have seen a case of a young man aged nineteen years who had interstitial keratitis when ten years of age, but for whom specific treatment had not been prescribed and who was now showing the early symptoms of juvenile paresis.

Optic atrophy is common while the pupils are usually dilated and sluggish. Neither tabes nor paresis of the juvenile type is very amenable to treatment. Worster-Drought says the cerebrospinal fluid is usually positive and remains so in spite of energetic treatment.

General Paralysis

For many years general paralysis had been recognised as a clinical entity, although for a much shorter period it has come to be regarded as a syphilitic manifestation. It is an organic disease of the cerebral cortex, usually occurring in the fourth decade of life. It displays a large array of clinical symptoms leading to progressive motor paralysis and profound mental deterioration, terminating fatally usually in two or three years. It does not appear to be so prevalent as it used to be. Professor George M. Robertson, in the Seventh Maudsley Lecture, called attention to the sudden fall in 1919 in the number of deaths from general paralysis of the insane, a decrease which he said was maintained. He stated that the symptoms of general paralysis do not occur with any frequency until seven years after infection, and the patient usually lives two years after the disease has declared itself. Therefore, nine years before 1919 something had happened to account for the observed fall in the accurately recorded death rate of this disease. In 1910, the year in question, Ehrlich introduced " 606."

Noguchi demonstrated the presence of spirochætes in the cerebral cortex of patients who died from the disease. The Wassermann reaction of the cerebrospinal fluid is always positive, and also, but not so commonly, is that of the blood. Juvenile cases are seen in congenital syphilis. A young man was found in a private asylum apparently suffering from general paralysis, but in reality he was there due to a remark made by a celebrated doctor with whom he was dining, who during the course of the meal suddenly exclaimed " Why you have Argyll Robertson pupils ! "—but Wisdom is justified of her children. The disease has been seen in a child seven years of age.

General paralysis occurs more commonly in males than females. The proportion is 4 to 1. The juvenile variety, however, is found

equally among males and females as both sexes are equally exposed to the inheritance of syphilis. General paresis may resemble diffuse meningo-vascular neuro-syphilis, not alone clinically, but in its histo-pathology, yet ordinary syphilitic manifestations such as iritis, ocular paresis, gummata, etc., are extraordinarily rare in patients suffering from general paralysis. However, just as in tabes, there may be atrophy of the optic nerves. About 8 per cent. show primary optic atrophy. It is towards the end of the disease that complete optic atrophy is liable to appear, yet it has been known to occur at a very early period, and has even preceded every other symptom by several years. The Argyll Robertson pupil appears in half the number of patients. The pupils also may be minute in size, that is miotic, or dilated, mydriatic or irregular, as in tabes. Consensual pupillary reaction, that is, contraction when light is thrown into the opposite eye, may be abolished before, simultaneously with, or subsequently to, loss of the direct reflex. Dr. Bevan Lewis considered loss of the sympathetic reflex (dilatation on pinching the skin of the neck) to be the earliest pupillary sign of general paralysis. The so-called paradoxical pupil symptom may be an early symptom of the onset of general paralysis. This condition is shown by throwing a strong beam of light into the eye by means of focal illumination. The pupil at first contracts, then dilates, then contracts again, and after a few such oscillations finally remains widely dilated.

Ocular palsies are not commonly found.

Hallucinations of visions occur in about 25 per cent. of cases. Transient blindness has been known to occur, and mind blindness is seen in the advanced stages.

Retrobulbar Neuritis

Retrobulbar Neuritis.—When inflammation affects that portion of the optic nerve which lies between the entrance of the retinal artery into the nerve (about 12 millimetres behind the globe) and the eyeball, there is a definite ophthalmoscopic change observed in the disc, the œdema present producing the change known as neuritis. But if the inflammation of the optic nerve takes place behind the entrance of the retinal artery then the condition known as *retrobulbar neuritis* does not produce a change in the appearance of the disc. The symptoms are subjective, although a descending atrophy—partial or complete—may follow. The inflammation in the nerve affects the connective tissue between the nerve bundles or the nerve

sheath, but does not affect the nerve fibrils, medullary substance or neuroglial sheaths, although later secondary degenerative changes do occur in these.

Traquair quotes Roenne's definition of retrobulbar neuritis as " a disease in the optic nerve characterised by a selective affinity for the papillo-macular fibres." The disease, characterised by the presence of a scotoma in the field of vision, is really not confined to any particular bundle of the nerve, evidence of which is often shown by the presence of a scotoma which extends far beyond the limits of the centro-cæcal area, and may even break through to the periphery, or the disc may show a definite neuritis (papillitis). However, much more commonly the scotoma is centrally situated, and there is no change found in the nerve head.

The average age of onset in Traquair's 139 cases of acute unilateral type of retrobulbar neuritis was thirty-two years.

When first seen a case of *retrobulbar neuritis* presents the following clinical picture. The patient complains of pain in the eyes and around the orbits ; there is pain on pressure being applied to the eyeballs, and pain produced by moving the eyes from side to side. Seldom does the patient complain of headache or vomiting. There is diminished vision, or the patient may say he has noticed a letter missing in a word. One patient stated she could not see the " o " in the word " Bovril " on an advertisement across the street ; while others complain of almost complete loss of vision, this loss occurring rapidly in a day or two. Some patients state they seem to be looking at objects through smoke.

In what is usually recognised as *acute retrobulbar neuritis* only one eye is affected ; there is no change found in the optic disc, the pupil when examined by strong light contracts but does not maintain its contraction, and the scotoma in the field of vision is central. If such conditions remained *in statu quo* then we would be justified in defining acute retrobulbar neuritis as a clinical entity, but we have already found that the disease may spread to both eyes, and neuritis or atrophy may appear and the central scotoma may so spread that vision becomes completely lost. However, we must bear in mind that cases characterised by (1) acute onset, (2) unilateral incidence, (3) central scotoma, and (4) tendency to recovery, are usually referred to as instances of *acute retrobulbar neuritis*. They show rapid loss of vision, and also rapid return. The vision may be lost in a few hours or days, and may return just as rapidly ;

the central vision being restored so that visual acuity again becomes 6/6, but searching the field with small coloured objects may still show a residual defect. Vision for white always returns more rapidly than for colour. The scotoma is usually central, but may extend towards and break through to the periphery. Sometimes an annular scotoma appears around the fixation point, and a small area of intense loss of vision may be found within the existing scotoma. It has been pointed out that if the lesion is situated at the junction of the nerve with the chiasma a unilateral scotoma with a straight vertical edge is produced, and is situated at the apex of the upper and outer quadrants of the field of vision.

Ophthalmoscopically the disc is usually observed to be normal, but a faint indistinct margin may be seen, or occasionally a raised disc surface discerned.

It is difficult to determine by means of the ophthalmoscope whether a swollen disc when present is due to raised intracranial pressure—plerocephalic œdema—or due to œdema of the nerve—neural œdema. The writer has observed on more than one occasion when there was an inflammation of the bulbar end of the optic nerve present that a small hæmorrhage has been found floating in the vitreous, and the patient will state that he has already seen the floating spots with the affected eye. But this is not observed in plerocephalic œdema. A comparison of the fields of vision show that in raised intracranial pressure there may be peripheral contraction of the field ; whereas in inflammation of the optic nerve, especially when the papillo-macular bundle is affected (axial neuritis), the defect is a central scotoma. In plerocephalic œdema, or as it is commonly called, papillœdema, central vision may remain normal for months, but it is rapidly lost in neural œdema.

Many cases are observed where a permanent scotoma remains, central vision becoming worse and then improving with long intermissions, and a slight temporal pallor of the disc may supervene. One such case the author has observed for ten years, but this young woman has shown no further development, such as disseminated sclerosis or other form of disease. Both eyes may be implicated ; the etiology of these cases presents decided difficulties.

As an example of the bilateral form of retrobulbar neuritis observed during pregnancy the following may be quoted. A young woman twenty years of age about one month before her confinement noticed her vision had suddenly diminished. On examination I

found the central vision of each eye such that fingers could only be counted at 20 inches. Ophthalmoscopic examination showed each fundus to be normal. After recovery from confinement vision gradually improved until 6/60 was reached in each eye, and it has remained so for two years. Now it can be seen that the temporal portion of each disc (papillo-macular bundle) has become paler in colour than the nasal side.

Causes.—To the ophthalmologist acute retrobulbar neuritis is usually considered to be idiopathic in origin, but to the neurologist it appears to be due mainly to disseminated sclerosis. Adie compared the age of onset and sex incidence in acute uncomplicated cases of retrobulbar neuritis and in cases of established disseminated sclerosis in whom a history of an acute attack of visual failure in one eye was given as their first symptom. In 85·8 per cent. of the former and in 86·9 per cent. of the latter the acute attack occurred between the ages of twenty-one and forty, in the majority between the ages of twenty-one and thirty. Adie further stated that acute unilateral retrobulbar neuritis, uncomplicated, was found in 64·7 per cent. females; history of unilateral retrobulbar neuritis as first symptom in patients with disseminated sclerosis, 63·7 per cent. females. These agreements are remarkable, and clearly suggest a common cause; also that females are more liable to both diseases than are males. We are therefore justified in stating that the common cause of retrobulbar neuritis is disseminated sclerosis.

Of the more chronic forms of retrobulbar neuritis where the symptoms are unilateral we must mention among the causes that of raised blood pressure. A hæmorrhage into the optic nerve is not at all uncommon. From a small scotoma occurring in any part of the field to complete blindness, such is the disturbance produced by a local hæmorrhage into the nerve. Of three cases which have been observed recently, one, a lady in her forties suffering from hyperpiesia, while playing tennis noticed the onset of a sudden diminution of vision in one eye, the investigation of which led to the treatment for raised blood pressure, and while treatment was being administered month by month the large scotoma became smaller and smaller, until at last only a vertical streak was found above the central area, and finally this disappeared. The second case was one of complete sudden unilateral blindness in a case of extremely high blood pressure, and in a short period of time after this occurrence the patient died. The third case is illustrated by Fig. 49, in

which blindness at the macula gradually recovered in a period of a few months.

Inflammation of the frontal, ethmoidal and sphenoidal sinuses may be responsible for retrobulbar neuritis, chiefly of the unilateral and more chronic type. The toxins escaping from suppurating sinuses easily invade the orbit. I have seen a suppurating frontal sinus cause blindness together with complete paralysis of the extra-ocular muscles. Watkyn Thomas operated twice on the frontal sinus, and in three months there was complete restoration of function of the orbital muscles and nerves. Van der Hoeve described cases of disease of the accessory sinuses and its effect on the optic nerve. He described the typical scotoma defect and enlarged blind spot (van der Hoeve's sign) in these cases, also atypical forms showing peripheral contraction of the field of vision and annular scotomata. He further stated, "The ophthalmologist has in the eye no sign to distinguish the origin of a retrobulbar neuritis," and also admits that X-ray examination may fail, but that an exploration made surgically with evacuation of pus often leads to rapid restoration of vision. It stands to reason that if the optic nerve is exposed to toxic influences for a lengthy period degeneration of some fibres will eventually take place and some portion of the field of vision will be permanently lost. Before one would explore the nasal region, it must not be forgotten that, co-existing with the nasal or other diseases, there may be a diabetic or syphilitic condition present also. Professor de Grosz of Budapest found in 18,587 patients 58 cases of optic nerve disease of nasal origin.

Many conditions have been quoted as being causal factors in the production of retrobulbar neuritis, such as chill and infectious diseases, poisons such as nicotine, wood alcohol, iodoform, etc., are responsible at times, while Leber's atrophy must not be forgotten. From the perusal of the various causes of retrobulbar neuritis one deduces the fact that the optic nerve in this disease may be attacked by both local and constitutional conditions. The disease may be in the optic nerve alone, or may be far removed from it. Loewenstein has reported that he found tubercle bacilli in the blood of cases of disseminated sclerosis with retrobulbar neuritis, but it must be remembered that Kolle and Küester have not been able to substantiate Loewenstein's findings.

C. B. Walker, Traquair, Peters and many others, emphasise the value of quantitative perimetry in the investigation of retrobulbar

neuritis before any surgical procedure is undertaken. This method requires more time than ordinary perimetry with the 5 or 10 millimetre discs, but the results are exceedingly finer. Walker quotes a case of post-ethmoidal disease where ordinary perimetry failed to record any change, yet quantitatively there was found not only an enlarged blind spot but an upper temporal field defect also. The posterior ethmoidal cells and sphenoidal sinus were found filled with pus and polypi, and when these were curetted the field of vision thirty days later returned to normal, leaving only a slightly enlarged blind spot.

Differential Diagnosis.—There is a rapid onset of the symptoms of retrobulbar neuritis in cases of neuromyelitis optica of Devic, but in this latter disease there is spinal involvement (see p. 324), together with bilateral neuritis.

I have seen syphilis causing a form of retrobulbar neuritis with the production of a large horseshoe-shaped scotoma, the macular area escaping.

Although disseminated sclerosis is accountable for many cases of acute retrobulbar neuritis yet, as van der Hoeve says, we must not say it is disseminated sclerosis until definite symptoms of this disease appear.

Although Leber's disease is a form of retrobulbar neuritis, yet it is distinguished from the acute form by both eyes being always affected, bearing in mind that the affection in one eye may precede the other by a few days up to a year or so. In Leber's disease, too, there is a central scotoma for both white and colours, and the fundus may appear to be perfectly normal. Later, however, a partial optic atrophy ensues with the pallor on the temporal side of the disc, or the whole disc may be completely involved. Leber's disease also attacks members of the same family.

The scotoma formed in tobacco amblyopia is never so dense as that found in retrobulbar neuritis. There is no pain produced by moving or pressing upon the eye, also tobacco amblyopia is bilateral, although one eye may be more affected than the other. The recovery of vision in tobacco amblyopia is not so rapid as is usually found in acute retrobulbar neuritis.

The defect caused by vascular disease is usually sudden, unilateral, and recovery is slow—if at all. The field defects are more or less extensively sectorial in shape rather than a central scotoma.

Hysterical blindness is never accompanied by a demonstrable central scotoma.

In those cases of retrobulbar neuritis accompanied by swelling of the optic disc an important point of distinction between this condition and a similar one caused by intracranial pressure is that the loss of vision in the former is much more marked than in the latter ; also the loss in the field of vision which gradually supervenes in persistent raised intracranial pressure is that of peripheral concentric contraction.

Finally, a tumour pressing on the nerve will produce at first a central scotoma, but the failure of vision is steadily progressive, this failure being unilateral. (See p. 248.)

In all cases of suspected retrobulbar neuritis it is of the utmost importance to examine the field of vision, not only of the affected eye but of the apparently normal one.

Prognosis.—As regards the recovery of sight in the acute forms the prognosis is good. In the more chronic and bilateral forms the recovery is not so good, and a slow recovery of vision does not augur well for a complete recovery. Cases which result in complete blindness may recover, but may leave severe and extensive field defects, while if the duration of the blindness or partial blindness is lengthy, recovery may be far from complete.

Treatment.—Should the cause be difficult to determine, it is always well to remember that in inflammation of any part of the body absolute rest is essential, so therefore, should a case suffering from retrobulbar neuritis appear to be in normal health, yet this matter of rest must not be forgotten. There is surely no need to emphasise the necessity of preserving at rest such a delicate structure as the optic nerve. Rest in bed, avoidance of reading, etc., should be followed. Diathermy may be of the utmost helpfulness. Such an apparatus as Siemens' Ultratherm, 6 metre wave-length, is spoken of most highly by Dr. Sainsbury of the West End Hospital for Nervous Diseases in connection with ocular therapeutics.

The iodides in 10 grain doses, t.i.d., are useful, also inunctions of mercury especially around the orbit, salicylates and tonics may be prescribed ; but if a constitutional cause is found treatment must be specifically given in that direction.

Disseminated Sclerosis

(*Synonyms :* Insular sclerosis ; Multiple sclerosis).—This disease is commonly found in European countries, particularly in Norway ; also in Switzerland, where the incidence is twice that of England and

Wales. There is a widespread divergence of opinion as to the etiology of this disease. Spirochætes have been described (Kalberlah, Pette), also a filtrable virus (Bulloch), while minute spherical bodies—*Spherula insularis* of Chevassut—have been stated to be the cause of multiple sclerosis. Brain, in his critical review of disseminated sclerosis with 218 references, says that the evidence appears to be more in favour of the view that disseminated sclerosis is due to neurotropic infection, and is an encephalo-myelitis characterised pathologically by perivascular demyelinisation with a definite relapsing tendency. Patches of demyelination followed by gliosis are found in the optic nerves, the neighbourhood of the cerebral ventricles, and many other situations throughout the brain. Also the posterior columns, and the margins of the spinal cord. The cerebellar lobes are comparatively rarely attacked (Buzzard and Greenfield). After a time secondary degeneration occurs, due to destruction of the axis cylinders. When the axis cylinder and myelin sheath are destroyed, then the signs and symptoms of the disease are permanent. It is said that males are more affected than females, yet judging from the number of cases of idiopathic retrobulbar neuritis seen at hospital, one would infer that females appear to be more commonly attacked by this disease than males. The disease attacks young adults, chiefly between the ages of twenty and forty. Children and young adults have been brought to ophthalmic hospitals suffering from a certain degree of blindness, or even complete amaurosis, followed by recovery. These cases a few years later have shown signs and symptoms of disseminated sclerosis.

The onset of disseminated sclerosis is usually gradual, but an acute or apoplectic form of multiple sclerosis does occur (Weschler). Many writers (Guillain, André Thomas) have stated that the onset is insidious, others that it is sudden. If one is to judge retrobulbar neuritis to be an early symptom of disseminated sclerosis, then both from the ophthalmological and neurological points of view the onset of disseminated sclerosis is gradual. Adie states that retrobulbar neuritis as an early symptom may precede the general appearance of signs and symptoms by many years. Many patients when closely questioned will state " they had a dragging limb or weakness long before they came up for examination," or " they saw double some years ago, but it passed away." It is commonly accepted, however, that in this disease there is a history of frequent relapses or remissions.

Optic Nerve.—The disease may attack the optic nerve at many points. Patches have been observed close to the central vessels, while in the chiasma and optic tracts these may be either subpial or subependymal in relation to the chiasmatic recess of the third ventricle. Rönne says that involvement of the chiasm and optic tract is frequent both in disseminated sclerosis and acute myelitis and gives an interesting diagram of the field of vision in one case which includes both a scotoma and a hemianopia. There is marked

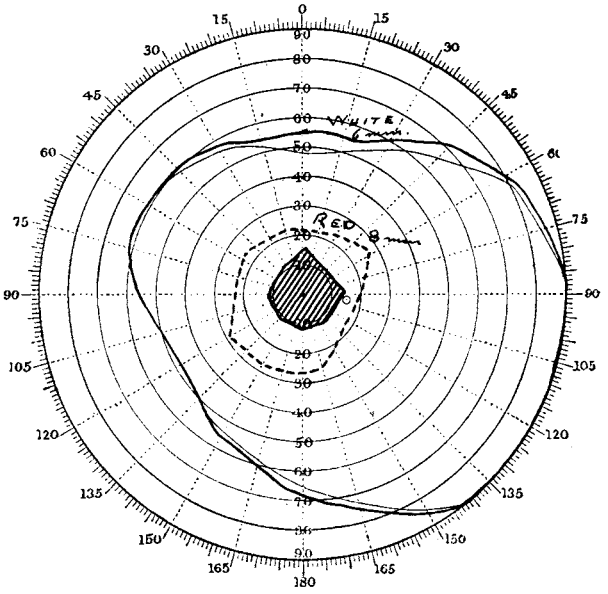


FIG. 113.—Field of vision of right eye in a case of disseminated sclerosis mapped out when the disc was a bright red colour and its surface elevated one diopter. There was a central scotoma, vision fell in four days from 6/12 to counting fingers at 8 inches. Finally central vision remained at 6/36.

reaction of the connective tissue as well as of the glia and the optic nerves (Dawson). The whole intracranial course of both optic nerves may be devoid of myelin, and the optic radiations may be involved also by extensions from the periventricular sclerosis around the posterior horn (Brain). In the earliest ocular manifestations of the disease, when most probably a retrobulbar neuritis is present, the disc, as a rule, does not show any appreciable difference from normal. Yet in a few cases there are slight changes, such as a deepening or flush of the pink colour of the disc surface, and large veins with

slightly blurred edges of the disc. In the more advanced stages of the disease there is a partial optic atrophy (see Plate VIII), due to atrophy of the papillo-macular bundle. This pallor extends to the edge of the disc, but when a deep shelving physiological pit is present it is difficult to define, but it is a very definite whitening of the temporal edge of the disc, the whiteness extending to the extreme edge of the disc on the temporal side. Uhthoff states that 3 per cent. of the cases of disseminated sclerosis show complete optic atrophy, 37 per cent. partial optic atrophy. This latter figure, however, may be easily increased to 70 per cent. according to the state of the disease as seen among the patients at the West End Hospital for Nervous Diseases. Nettleship has observed some cases of mild degree of papillœdema, while Leslie Paton records three cases showing active changes of the disc. (There was recovery of vision synchronous with the subsidence of the optic neuritis.) Paton suggests that disseminated sclerosis should be thought of when disc changes are found to be of very short duration. The main change observed in the optic disc is, however, one of atrophy, partial or complete. It may be assumed that if active changes are taking place close to the nerve head the atrophic appearance may be preceded by that of a mild neuritis, viz., redness of the surface, blurred edge or slightly swollen disc.

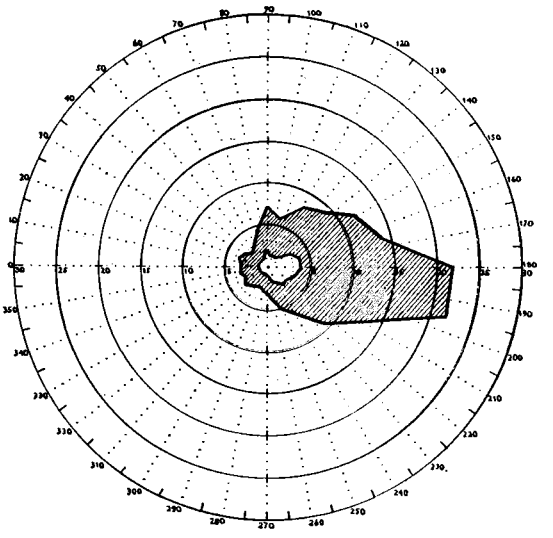


FIG. 114.—Unilateral central scotoma found in conjunction with the presence of a partial primary optic atrophy (see Plate VIII). Central vision reduced to 6/60.

Fields of Vision.—The changes in the field of vision are toxi-inflammatory in type (see Figs. 113 and 114). Single or multiple scotomata which may or may not involve the central vision are to be found. Central scotomata may be bilateral and often relative,

with no change in the peripheral field. However, irregular peripheral contraction is found without loss of central vision (Fig. 115). If the chiasm or optic tracts are affected, quadrantic defects in the field of vision occur, or homonymous hemianopic defects are found involving central vision. If a quadrantic or hemianopic defect suddenly develops, particularly in a young woman, and as quickly passes away, the possibility of the presence of disseminated sclerosis should be borne in mind. The ophthalmic surgeon more commonly meets with changes symptomatic of acute retrobulbar neuritis. Defects in the field of vision may be seen before any change is observed to have taken place in the optic nerve head. It is characteristic of the disease that defects in the field of vision may alter in shape, position or intensity. Those cases of diminished vision resembling acute retrobulbar neuritis may recover quickly. The frequent fluctuations in the field of vision are due to resolution of the lesion in one part of the nerve followed by development of fresh foci elsewhere.

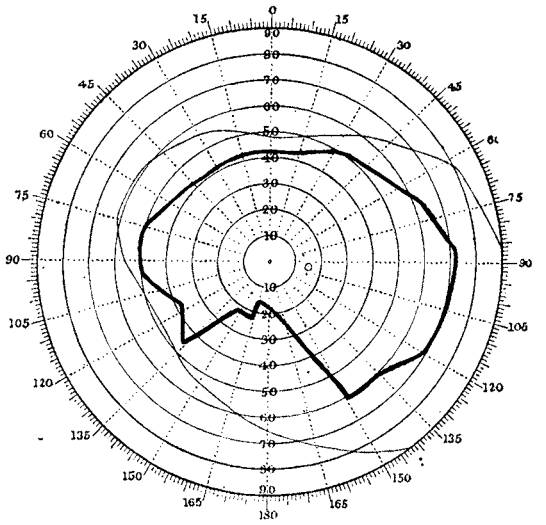


FIG. 115.—Contraction of peripheral field due to disseminated sclerosis without loss of central vision for form.

Nystagmus has always been mentioned as one of the classical signs of disseminated sclerosis, but it is frequently absent in this disease. It is sometimes present when the condition is not that of disseminated sclerosis, but is due to a lesion of the cervical portion of the spinal cord (Brissaud). Nystagmus is frequently seen during some stages of the disease (70 per cent.). The nystagmoid movements elicited in healthy persons on looking strongly to the right or left should not be confused with the nystagmus found in disseminated sclerosis or other disease of the nervous system. The nystagmus is frequently so fine that it is not seen

by the naked eye, but when the disc is magnified seventeen times by direct ophthalmoscopic observation the fine movements can be perceived. Observation by Frenkel's spectacles is useful.

The relationship of retrobulbar neuritis to disseminated sclerosis is a very much debated one. Many ophthalmic surgeons have seen cases of retrobulbar neuritis for which no adequate cause could possibly be found. These cases also were unaccompanied by symptoms of a nervous nature, and so were not then seen by a neurologist. But many neurologists on the other hand, have also seen cases in a state of early disseminated sclerosis who stated that years previously they had suffered from an attack of unilateral blindness which had passed off. Traquair states that " multiple sclerosis is accepted by all ophthalmic surgeons as a common cause of retrobulbar neuritis, but its incidence is estimated as widely as from about 20 per cent. to about 80 per cent. Adie examined 70 cases of acute retrobulbar neuritis within a week or so of onset. In 31·3 per cent. the patients were already suffering from disseminated sclerosis, and in 41·8 per cent. the diagnosis " probably disseminated sclerosis " was made. Both Traquair and Adie found that just over 60 per cent. of cases of acute unilateral type of retrobulbar neuritis were females ; this may suggest a common cause, viz., disseminated sclerosis. (See Retrobulbar Neuritis.) Marcus Gunn found in 233 cases of retrobulbar neuritis 51 showing symptoms of disseminated sclerosis.

Ocular Palsies.—Instead of retrobulbar neuritis being the initial symptom of disseminated sclerosis, ocular palsies of a fleeting nature have been observed. If a plaque is present in the mid-brain or pons, paralysis of conjugate movements may be present. I have seen a young woman with each eye deviated outwards, which was due to a bilateral nuclear paralysis of both oculo-motor nerves. Sometimes there is only a paresis found which upsets the muscle balance, the paresis being measureable by means of the Maddox rod. The sixth nerve is most commonly affected. Partial dissociation of conjugate movements may occur, resulting in divergent squint and diplopia. The lesion in these cases may be in the posterior longitudinal bundle.

Pupillary Changes.—There may be miosis of the pupils or they may be unequal ; sometimes they simulate a unilateral Argyll Robertson pupil, but as a rule the pupillary reactions are normal. Total ophthalmoplegia interna has been described.

Brief Contrast of Ocular Signs and Symptoms in some of the Commoner Diseases of the Nervous System

Tabes Dorsalis.	Disseminated Sclerosis.	Hysteria.	General Paralysis.
Optic nerve. 20 per cent. show optic atrophy.	3 per cent. show complete optic atrophy. 60 per cent. show incomplete optic atrophy.	Normal discs.	8 per cent. show primary optic atrophy.
Paralyses. Third cranial nerve most commonly affected.	Associated movements may be affected, also nystagmus may be present.	Normal muscles.	Paralyses occur similar to tabes but the percentage is much smaller.
Pupillary changes. Chiefly Argyll Robertson pupils (70 per cent.), also miosis, irregularity of pupillary outline and unequal pupils.	Miosis of the pupils, unequal pupils. As a rule pupils are normal.	No change.	Argyll Robertson pupils (about 50 per cent.), also paradoxical pupil.
Fields of vision. Symmetrically contracted.	Scotomata may be present. Central colour scotoma common.	Fields vary from day to day.	Loss of field depends on extent of optic atrophy.
Vision. Central vision usually remains until complete atrophy is present.	Central vision usually good. Sudden loss of sight may be due to retrobulbar neuritis.	Normal.	Mind blindness.

Neuro-myelitis Optica

Synonyms : Diffuse myelitis with optic neuritis : la neuro-myélite optique aiguë, or Devic's disease.

This disease was first described by Allbutt in 1870, and later by Achard and Guinon in 1889. Devic in 1894 gave it the name of neuro-myélite optique aiguë. Both sexes from the age of twelve to sixty are liable to this disease. There is massive demyelination of both optic nerves and spinal cord, generally in the lower cervical and upper dorsal regions. The lesion in the optic nerve produces a neuritis which may be severe and without pain, or symptoms of a

retrobulbar neuritis with little change at the nerve head, but with severe pain on movement of the eyes or by pressing on them with the fingers. The spinal cord lesion may occur before, at the same time, or after the appearance of the ocular symptoms. Goulden states that the optic neuritis appears first in four-fifths of the cases. Beck showed that in one case examined the spinal cord exhibited massive demyelination from the seventh cervical to the twelfth thoracic segments, with small areas of rarefaction in which the axis cylinders were also destroyed, although Gordon Holmes states that a considerable proportion of axis cylinders persist. In the demyelinated areas there is marked perivascular infiltration with cells of the mononuclear variety. The lesions in the optic nerves are most marked anterior to the chiasma, but may extend throughout the optic tracts. These lesions are produced by a common virus acting on both spinal cord and optic nerves.

The pathology of the optic nerves in Beck's case showed a great deal of demyelination of both nerves. Each was extremely cellular, most of the cells being neurological astrocytes. Many microglial cells were found particularly near blood vessels. Near the eyeball behind the lamina cribosa there was a certain amount of round-cell infiltration, while in the most posterior part of the optic nerve were found a number of cavities which extended into the chiasm and optic tracts. (See also Rosenbaum's case.)

A case of the author's presented herself in the out-patient department of the Western Ophthalmic Hospital. A young mother about twenty-eight years of age complained of loss of vision during the past week or so. Vision was 6/36 and 6/24 in each eye respectively. On examination of the fundus a 3 dioptre papillœdema was found to be present. There were no other signs or symptoms apparent. A week after admission to hospital the patient became completely blind. The only neurological symptom to be discovered was slightly increased deep reflexes pointing probably to a pyramidal tract lesion. Three weeks later vision returned, and soon central vision became the normal 6/6, with subsidence of the swelling of the nerve head, leaving behind slight pallor of the surface of both discs, but with no obvious change in the fields of vision. This case might be classified with a number of others which showed the presence of neuritis with very slight or absent signs of spinal cord involvement. The case just mentioned was given injections of acetylarsen with rest in bed.

Differential Diagnosis.—In neuro-myelitis optica the characteristic defect in the field of vision is a bilateral central scotomata, which may rapidly proceed to complete blindness and may just as rapidly recover. This is in direct contrast to what is found in papillœdema, the result of raised intracranial tension produced by a brain tumour. Here the vision may not be affected even for months, although a papillœdema of four to six dioptres may be present. Again, contrasting neuro-myelitis optica with disseminated sclerosis, it must be remembered that a bilateral neuritis is seldom seen in the latter. In both of these diseases a central scotoma is extremely common, relapses occur in disseminated sclerosis, but not so common in neuro-myelitis optica.

Prognosis.—As in the case already quoted, there may be complete recovery, although the accepted mortality is about 50 per cent.

Encephalitis Periaxialis Diffusa (*Synonym:* Schilder's disease)

The disease first described by Schilder in 1912 has fortunately become less common in recent years. Collier, Greenfield, Stewart Symonds and others have made contributions to the subject. The disease generally occurs in children and young people. Most probably it is inflammatory in origin, affecting the white matter of the cerebral hemispheres, resulting in widespread demyelination followed by destruction of the axis cylinders, the greatest change taking place usually in the white matter of the occipital lobes. The disease is bilateral, spreading in a forward direction, the temporal lobes becoming involved together with the optic radiations, so that an early symptom may be a hemianopia or complete blindness. The pupillary reflexes, however, are preserved as the primary centres are not involved, nor the pupillary pathways unless atrophy of the optic nerves supervenes.

The onset is rapid, usually without fever. Headache and giddiness may be present. Mental changes occur leading to progressive dementia. Epileptiform attacks may precede or follow the visual disturbances; these may occur at any stage of the disease; the fit may take the form of conjugate deviation of the eyes followed by tonic and then clonic spasm of the limbs. Occasionally the disease begins first in one occipital lobe, so that an homonymous hemianopia on the opposite side results; but this is soon followed by the opposite occipital lobe becoming involved, and this leads ultimately to complete blindness.

Papillœdema occurs in 25 per cent. of cases, but the degree of swelling of the optic disc is always of small amount. Primary optic atrophy is rarely seen, but if present the pupillary reflexes are affected. Occasionally the sixth cranial nerves are involved, causing diplopia, while nystagmus is sometimes seen. It has been stated that the disease is not familial, yet the author has observed three cases occurring in one family—all were under twelve years of

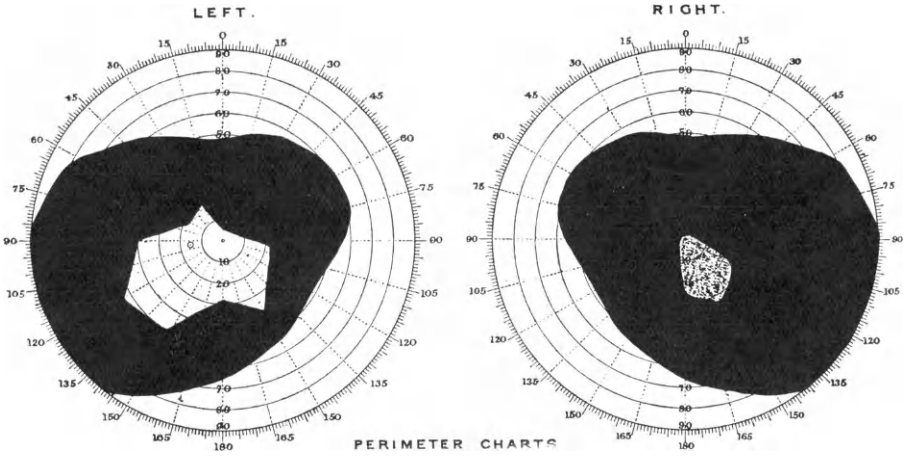


FIG. 116.—The left visual field was taken with a white disc 1 cm. in diameter. the right with a piece of white paper 10 cm. square. The woman aged forty-three had suffered from migraine for years. While at work, sight of the right eye became dim. Two days later she became blind in both eyes. She had not even perception of light. Three days later it was possible to take the above fields. Two days later she was completely blind again. Death ensued eight days after the onset of the illness. (From Stewart, Greenfield and Blandy.)

age. Two of these cases were under the care of Dr. Worster-Drought, and the third was under the care of the late Dr. E. D. Macnamara. Blindness occurred early in each case, and was rapidly followed by increasing dementia, together with a marked spastic paralysis of the limbs, with all the signs and symptoms of pyramidal tract disease. The fundus of each case remained normal throughout the duration of the disease. There were no changes to be seen at the optic disc, and although a finely scattered pigmentation was observed over the fundus the same appearance has been seen in normal children. Stewart, Greenfield and Blandy report three cases, aged three years, eight years, and forty-three years respectively. Although the average duration of the disease is fourteen months, yet the

woman aged forty-three died in nine days. These writers also observed that deafness may be an earlier symptom than blindness. Their pathological investigation showed the disease may have a much wider involvement in the centrum ovale, that it may affect the pons, optic nerves and cervical cord. The visual fields, which are illustrated (see Fig. 116), are those of the woman aged forty-three. The optic discs were swollen, the condition being a "true optic neuritis with papillitis" (Paton), not a papillœdema.

The disease invariably terminates fatally.

Differential Diagnosis.—Cerebral neoplasms produce papillœdema, headache and vomiting, but the vision is not diminished for several months, although the papillœdema may be of marked amount; whereas in Schilder's disease the papillœdema seldom measures more than 1 dioptre and blindness rapidly ensues.

Brain and Strauss state that there is really no other disease affecting children or young subjects which presents the following triad of symptoms: (1) Progressive loss of vision of the cerebral type; (2) progressive spastic paralysis; and (3) progressive mental dissolution.

Diffuse Cerebral Sclerosis.—Closely allied to Schilder's disease is that known as diffuse cerebral sclerosis (Krabbe), which is somewhat more familial than Schilder's disease, but various authors consider these diseases identical.

There may be optic atrophy, but blindness is probably due to involvement of the optic radiations, while squint and nystagmus may be observed.

Migraine

Synonyms: Hemicrania; Bilious attack; Periodical sick headaches.

Migraine is characterised by paroxysmal headache which is usually accompanied by nausea and visual phenomena. The headache, although beginning at a certain spot, the eye or the temple, and remaining unilateral for a time, subsequently spreads. In only a small proportion of Campbell's cases, did the headache remain unilateral. Most of his cases showed symmetrically distributed headaches over the anterior part of the head, or over the forehead alone. Sometimes the pain reached the bridge of the nose.

In the large majority of cases the headaches begin at puberty, but cases as early as eight years of age have been recorded, while

some in middle age have suffered from their first attack. It is more commonly seen in females than in males. In females menstruation is a decided factor in bringing on an attack of migraine. In other cases diet seems to influence the disease. Campbell suggested that in some cases where the patient's eyes were subjected to severe strain he could often cure himself by wearing suitable glasses and diminishing the supply of nitrogen (fats) in his food. The author knows of a doctor who dare not take milk lest it precipitate a most violent attack of migraine. As the patient grows older the attacks change in character, or may disappear altogether, or the migraine of early life may be replaced by epilepsy in later life (Ely).

It is a well-known fact that during pregnancy migraine is modified. Fisher believed this was due to the profound change which takes place in the pituitary gland at this period.

There are cases where the attack consists of nothing else than simple bilious headache, while others, for a day or two previously, experience premonitory symptoms, which consist of a low-spirited, languid or drowsy feeling, constipation, followed by visual hallucinations, consisting of scintillating scotomas, fortification spectra, (known as teichopsia), or the visual disturbance may show itself as a hemianopia. The scintillating scotoma may begin as a spot close to the central point of fixation which spreads peripherally in a scintillating manner, objects within this area becoming invisible. In fifteen to thirty minutes this amaurosis is followed by severe paroxysmal headache. The teichopsia or fortification spectra may take the form of battlement of zigzag lines with coloured edges, sometimes a spot of light appears in a scotomatous area, rapidly extends, developing into a zigzag pattern. In fifteen to thirty minutes these visual disturbances are followed by severe paroxysmal headache and nausea, after which the patient gradually feels better. If hemianopia occurs instead of the scintillating scotoma, it is found to be homonymous, each half of the visual field on the same side being affected. (See also Fig. 117.)

Migraine in which hemianopia is present may in a few cases resemble ophthalmoplegic migraine, inasmuch as in the latter paralysis of the ocular muscles becomes permanent, so in a few cases of migraine the hemianopia remains established. Such cases have been recorded by Thomas, Uhthoff, Ormond and Adie. They have described cases where permanent hemianopia was observed after repeated attacks of migraine. Gowers drew attention to those

cases of migraine which suffered loss of sight lasting for a few hours or a day or two, sometimes apart from attacks of headache, at other times in association with the pain. The failure of sight, usually transient, sometimes becomes permanent. Infrequently temporary blindness, or complete peripheral loss of field, leaving only central vision, is described. Double vision has rarely been complained of.

It must be remembered that visual hallucinations may be associated with intracranial tumour. These, together with headache and vomiting, may cause confusion where the diagnosis lies between an intracranial neoplasm and migraine beginning in a middle-aged person. Brain has seen migraine simulated by an aneurysm pressing

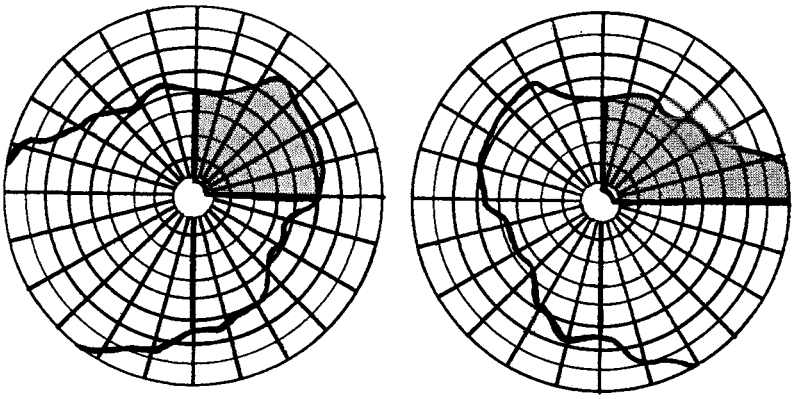


FIG. 117.—Homonymous quadrantic loss of vision in a case of migraine. (Peter.)

upon the optic nerve which caused flashes of light in the affected eye with headache; papillœdema was also present. Scintillating scotomata, if seen repeatedly, may be associated with cerebral hæmorrhage, cerebral anæmia, tumour of the brain and ocular lesions. In the true migraine attacks there is no change to be seen in the fundus. In this connection recently an interesting case was brought to the author's notice. It was that of a man, sixty-five years of age, who since boyhood had suffered from mental storms. On rising from bed and while dressing he would rave over some small detail of his past life, or storm about some simple domestic item, such as his grandchild being taken to see an oculist—he had little faith in these new-fangled medical ideas. These storms would pass away in an hour or so, and therefore did not interfere with his heavy daily duties. But periodically an attack of migraine came

on, during which the patient remained in bed in complete darkness, often for two days at a time, all work being suspended. Lately, he suffered an attack of headache of great severity, but this time on recovery speech was found to be impossible and remained confused for several weeks. Examination of the fundi showed a papillœdema of 1 dioptre in each eye, the diagnosis being a meningeal hæmorrhage in the region of Broca's area.

Campbell, in his study of 80 cases of this disease, has noticed that nervous subjects of an intellectual or studious type are much more likely to suffer from the classical symptoms, while those who are not of this type, but who do suffer from the classical form of this disease, have a strong neurotic taint; he said, "I would be greatly surprised if I did not find among his relations a history of epilepsy or insanity, or both." He believes that the disease assumes its most specialised form in highly neurotic individuals, and such individuals owe their neurotic diathesis essentially to heredity. Liveing looks upon migraine as the sensory equivalent of epilepsy. The disease is probably of an angio-neurotic nature, and as such affects the structures concerned in sight, producing such disturbances as flashes of light and temporary loss of field of vision. Spasm of the cerebral vessels has been held accountable for such a condition as migraine. Recent experiments by Florey and Forbes and others have demonstrated nervous control over the cerebral circulation. Some have apparently observed a constriction of the central artery of the fundus of the eye during the preliminary stage of the aura, followed by throbbing and dilatation of the vessels. This suggests a vasomotor disturbance, probably spasm. Riley says, "The rôle of the vasomotor apparatus in the production of migraine remains as the most plausible explanation for the varying manifestations of the disorder." Whatever is the cause of migraine, in several respects, however, it resembles epilepsy; there are prodromal symptoms, the attacks are explosive in character, and visual symptoms occur (Ely). It is thought that the correction of errors of refraction does not influence the onset or course of either migraine or epilepsy; but the author, like the late Ernest Clark, is strongly of the opinion that a fine estimation of the refraction, together with a careful measurement of the muscle balance, may help very considerably in giving relief to sufferers from these diseases. An example of a migrainous patient obtaining such relief is as follows: A young lady bank-clerk, who could perform her duties for only half a

week at a time, and who had to remain indoors and often in bed the remainder of the week with severe headache, migrainous in type, did not absent herself a single day for six months upon wearing glasses which merely corrected a moderate degree of hypermetropic astigmatism. Another patient was seen no less than eleven times during a period of three months. She had come from abroad and, indeed, had failed to obtain relief in any direction. She wore her glasses for six years with great amelioration of the headaches. The minutest amount of astigmatism should be corrected, the muscle balance fully determined, prismatic correction given if necessary, especially for hyperphoria, and finally the glasses, while being worn, should be examined by the oculist or a thoroughly reliable optician as to their correct interpupillary fitting and angular alignment.

A short time ago Adson made a communication to the Proceedings of the Staff Meetings of the Mayo Clinic on the surgical treatment of migraine. His patient, a man of fifty-three years of age, complained of a boring or pressure type of pain behind and over the right eye, which extended over the parieto-temporal region. The first attack of this pain had occurred nineteen years before, had been periodic, and now had become a daily affair for the past four years. The pain had become so severe that the man had to give up his work as a salesman. The etiology of the symptoms was unexplainable. Adson first performed a peri-arterial sympathectomy of the common carotid artery for a distance of 2 centimetres from the bifurcation downwards to obliterate any afferent or efferent impulses travelling along the internal carotid artery. Next he ligated and divided the external carotid artery just above the bifurcation of the common carotid artery to obliterate the throbbing sensation of the pulse beats and also to interrupt impulses travelling along the middle meningeal artery and its branches. Further, to assure complete interruption of sympathetic pathways and communications with the vagus and glosso-pharyngeal nerves, he removed the superior cervical sympathetic ganglion and upper sympathetic trunk for a distance of 6 centimetres. The post-operative results were most gratifying. There was instant relief from the boring, pressure type of pain. Adson suggests that this case and its results confirm his suspicion that headache, whether it be migraine or the result of brain tumour, is caused by irritation of the meningeal arteries rather than by an intra-cerebral lesion,

with the exception of those lesions which involve the thalamus and sensory fibres. He draws attention to the fact that when a craniotomy is performed under local anæsthesia it is obvious that little if any pain is experienced during manipulation or resection of cortex, cerebral vessels or dura, but if the meningeal vessels are touched the patient will cry out with pain.

The many and various forms of treatment of migraine have been summarised by Alvarez, Riley and others. It has been noticed that an attack of typhoid fever can give relief for a year or two and therefore injections of peptone, or foreign proteins are sometimes helpful. Ergotamine tartrate as suggested by Tzanck and reported on by Lennox, von Storch and O'Sullivan has proved of great value. Although this drug was used at first with the idea that it quietened the sympathetic nervous system, Soloman concludes his investigation with the statement that migraine is not a disease caused by a generalised sympathetic dysfunction. The point is emphasised by Alvarez that if any medicinal preparation is to be taken internally it should be given promptly before the onset of nausea. In extreme cases when everything fails, Critchley and Ferguson suggest decompression. Holmes said he had never known migraine to persist in a patient who had had a surgical decompression performed.

Ophthalmoplegic Migraine

The two diseases, *migraine and ophthalmoplegic migraine*, should be considered as being two separate entities. Severe headache is common to both, but there the similarity ends. In 1890 Charcot gave his opinion that the term *ophthalmoplegic migraine* described cases in which there is a palsy of an ocular nerve, generally the third, at first temporary but later becoming permanent and associated with migraine. He described the condition as recurrent oculomotor paralysis. This affection is not common. In the *Lancet*, 1882, Saundby described a case of megrim with paralysis of the third nerve. G. H. Oliver remarks that patients suffering from ophthalmoplegic migraine are usually of the poorer classes, and are seen among hospital patients, whereas migraine is by no means confined to such. One of Oliver's cases was a woman who had passed the menopause and who presented herself as an out-patient suffering from a complete oculo-motor paralysis, internal and external. She stated that from early life she had suffered from pain in the head, vomiting and paralysis of eye movements; the latter in-

cluded ptosis. She saw double until the pain passed off. Gradually in recurring attacks the pain lasted longer, and the paralysis was of a more lengthened period until finally complete paralysis supervened. Moebius, who early described this condition in its entirety, thinks of relapsing oculo-motor paralysis as a condition in which from childhood or youth paralysis is limited to the third cranial nerve, commencing with headache and vomiting, returns at intervals, and affects nearly always one and the same eye. Oliver describes the three periods as the period of pain, the period of paralysis, and the period between the attacks. I have freely abstracted from his paper.

Period of Pain.—The pain is not a true neuralgia (Moebius). It is a diffuse pain such as migraine, beginning in the temporal region and spreading to the occiput, accompanied by nausea. The pain and vomiting always precedes the paralysis and then ceases as soon as the paralysis is fully developed. Moebius thinks the pain has its origin in the region of the nuclei of the third cranial nerve as the cell groups of the origin of the fifth cranial nerve lie to the outer side and above the third nerve group. As the fifth cranial nerve supplies the eye and dura mater as regards sensibility, so an œdema produced by a toxin affecting the third nerve origin could easily affect the fifth nerve cells, thus producing headache and vomiting ; these passing away leave behind a condition of paralysis. The pain may last from days to weeks.

Period of Paralysis.—It is usual in the earlier attacks for the paralysis to pass off, but gradually this condition becomes permanent. Ormerod and Spicer record cases where the paralysis was at once complete. All the muscles may not be equally paralysed. The iris may be paralysed, the accommodation escaping. The paralysis of accommodation, if present, and the mydriasis are the last to clear up. Both pain and paralysis may co-exist.

Period Between the Attacks.—Cases have been recorded in which patients have suffered at one time from true migraine and at another from ophthalmoplegic migraine, sometimes alternately, but always conscious of which condition was present. The paralysis always distinguished the ophthalmoplegic migraine from true migraine. Migraine attacks may be weekly or monthly, but attacks of ophthalmoplegic migraine may occur only a few times during the patient's life, and each attack may continue for days, weeks or months. In ophthalmoplegic migraine the paralysis may entirely disappear

between the attacks, or some muscle or muscles may remain paralysed.

There are many conflicting opinions regarding the cause of ophthalmoplegic migraine, but many believe it is a disease *sui generis*, the cause of which is unknown and not to be considered in the same category as those described by Simmonds as being due to subarachnoid hæmorrhages, or by Adie and Bramwell as a leaking aneurysm, or by growths, such as Karplus's case where a tumour and neurofibroma the size of a split pea was found on the trunk of the third nerve. In addition to the foregoing, one must diagnose the condition from cerebral syphilis, tabes, chronic meningitis, multiple neuritis, syringomyelia, hysteria and multiple sclerosis.

Moebius considered a toxin to be the probable cause of ophthalmoplegic migraine. Townsend relates a case in which septic teeth were removed after the first attack, but this procedure did not prevent the onset of a second and third attack. It is interesting to note that in this case the third attack differed from the previous two in that the sixth nerve became paralysed instead of the third. This patient was one of a large family, all of whom were subject to bilious headaches, while one sister suffered from epilepsy.

The involvement of the oculo-motor nerve in all of the published cases is constant, but the pathological pictures show no uniformity in the morbid processes (Riley).

Epilepsy

Attention has already been drawn to the relationship between migraine and epilepsy. Ely, writing of the migraine-epilepsy syndrome, says that he found 60·8 per cent. of epileptic patients giving ancestral histories of migraine, but only 14·03 per cent. giving ancestral histories of epilepsy, and 5·7 per cent. of patients with migraine showing epilepsy in their ancestral histories. Russell Brain found in his series of 200 epileptics a family history in 28 per cent.

So frequently has it been stated that errors of refraction are a probable cause of epilepsy or epileptiform attacks that it would be well to examine briefly the etiology of this disease. At once we are faced with a difficulty, for Cobb enumerates fifty-six direct causes of fits and postulates thirteen physiologic mechanisms to explain them. He does not include ophthalmic conditions in his list. Gowers described a case of a young man who suffered from

severe convulsions of the hysteroid type with paroxysms of struggling which only ceased when ice was applied to the spine. After a few days Gowers noticed blurring of the optic discs with some swelling which subsided after the cessation of the fits. The patient died in three months, and at post-mortem no trace of disease visible to the naked eye was found. Such cases as these are termed idiopathic epilepsy. Yet in a great many cases of epilepsy post-mortem evidence of meningeal inflammation, atrophy of the brain and dilatation of the ventricles is found. While studying for a final examination one of the author's patients found great amelioration from headaches by the use of proper glasses. Previously to this the patient had met with a serious motor accident in which a large piece of the parietal bone was torn away, and on healing scar tissue was formed between the scalp and the dura mater. But finally convulsive seizures came on and only by the separation of the scalp from the dura by interposing a bone graft did the fits cease. Foerster and Penfield have described contracting scars as a cause of traumatic epilepsy. Of intracranial neoplasms only 25 per cent. appear to produce convulsive seizures, and these are chiefly tumours in the anterior fossa, while the high pressure produced by tumours in the posterior fossa is seldom the cause of convulsions (Parker). In Penfield's study of 30 cases examined on the table during the convulsive period he found 26 showed vascular changes. A constant visible phenomenon was cessation of arterial pulsation. Cerebral pallor was sometimes present during the seizure, but more commonly followed it. He found the epileptic brain subject to vasomotor reflexes never described in the normal brain. These reflexes were probably not sustained by autonomic centres outside the cranial cavity, but by centres within the brain itself. "Physiological instability of the blood vessels seems to be the abnormal state common to epileptics of all varieties." The term idiopathic as applied to epilepsy really means that the cause is unknown, but gradually there is more light being thrown on this most distressing disease. During the last ten years it has been proved that in many cases there is an underlying metabolic cause. Epileptic children in whom a condition of ketonæmia has been produced by giving a ketogenic diet (a diet rich in fats and poor in carbohydrates) have been freed from fits in 31 per cent. of cases, and in a further 23 per cent. the number of attacks was reduced. Adults, however, have not responded so well (Bridge).

In Gower's series of 3,000 cases the proportion of males to females was 12 to 13. In 75 per cent. the attacks began before twenty years of age. There was an aura or warning of the attack in three-fifths of all his cases. During the pre-convulsive symptoms special sense auræ took place. Gowers says a visual warning of epilepsy is twice as common as all the other special sense auræ together. The visual auræ consist in seeing flashes of light or balls of fire, sudden loss or dimness of vision, complex scenes, scintillating scotoma, macropsia or micropsia (objects appearing large or small), animals, etc. During the convulsive state, which may last half a minute, the pupils are widely dilated and fixed, and the corneal reflexes are lost. Light thrown on the pupils does not produce a reflex. Gowers, speaking of his series of cases, says that the only change in the vessels of the fundus seen was a distension of the veins during the period of lividity. The author has frequently noticed in some cases of petit mal and mild cases of epilepsy that the veins of the fundus appear somewhat varicose in form. This was not due to arterio-sclerosis. The patients were chiefly young women about thirty years of age. In one case the epileptic attacks were so mild that the patient considered herself fit to drive a motor car. (The writer's opinion is that anyone liable to fits, no matter how mild in character, should not be allowed a driver's licence.) During the attacks minute hæmorrhages may form on the retina such as are seen in agonal states.

In epilepsy (as in migraine) every peripheral source of irritation should be excluded. Errors of refraction should be carefully corrected and the muscle balance measured, and if necessary prismatic correction should be prescribed. In one year the author saw 5 cases, a boy three years of age, another boy fourteen years of age, a girl of eighteen, and two older patients, whose attacks were practically abolished by the wearing of glasses for the first time to correct errors of refraction. These were usually of the hypermetropic astigmatic variety with obliquely placed axes. In all the large nerve hospitals the physicians have found it of the utmost advantage in helping the progress of a case to refer such patients suspected of suffering from errors of refraction to the ophthalmic clinic where special facilities are available for prescribing the necessary glasses.

It is remarkable that Mayer obtained so many varying conditions of the eye in the examination of ninety-six patients. He found

defective vision in 41 cases, very few with muscle imbalance or pupillary abnormalities. The majority of the epileptic patients examined in the ophthalmic clinics of hospitals for nervous diseases do not show any marked error of refraction or departure from the normal fundus. Spiral and bizarre fields of vision have often been described, but Mayer, while examining the fields of vision by means of flashes of light, does not find any constriction of the visual field. In organic cases of epilepsy, as would be expected, he did find optic atrophy, arterio-sclerosis and in one case choked disc which later proved to be due to a tumour of the brain. Although he concludes his paper by saying, "To-day it is known that there is no direct relation between errors of refraction and epilepsy," yet I maintain when such is found it is the duty of the physician or ophthalmologist to insist upon a correct estimation of the refraction and that the correction shall be worn constantly.

Primary Thyrotoxicosis

Synonyms : Exophthalmic Goitre ; Graves', Parry's or Basedow's disease.

This disease named after Robert James Graves, son of a Dublin doctor, was described by him in 1835 ; but a much more complete account of the disease was given by Caleb Hillier Parry of Bath (1755–1822). It was left to John Basedow in 1840 to make a full and exact account of the disease. This disease characterised by exophthalmos, enlarged thyroid, tachycardia, tremor and nervousness is still undefined as regards its etiology. It affects women much more commonly than men, about six to one (Joll). The symptoms usually begin about thirty years of age, but it is seen both in the young and old. Joll says that it is probable, but difficult to prove, that all forms of thyrotoxicosis have an extra-thyroid origin. If the sympathetic system is at fault why are the pupils not dilated ? If the thyroid is not at fault, why does partial thyroidectomy do so much to restore the patient to health again ? (although, as one has often observed, such an operation does not cure the exophthalmos. Joll states 50 per cent.).

There is an undoubted relationship between the other ductless glands and the thyroid. A considerable amount of work has recently been done which seems to co-relate the functions of the anterior lobe of the pituitary, thyroid, ovaries and adrenals. Anderson and

Collop have reported the production of hyperplasia of the thyroid in the rat with a marked hyperthyroidism by means of a crude alkaline extract of anterior pituitary and a killed staphylococcic culture. A purified thyrotropic extract administered to eight hypophysectomised rats prevented the atrophy of the thyroid which invariably occurred in the untreated hypophysectomised animal. Riddle says it would seem that the thyrotropic response is produced by the gonad-stimulating hormone of the anterior lobe of the pituitary gland. Schochaert and Foster found the total iodine content of the thyroid gland in the young duck was rapidly reduced by injection of a saline emulsion of the anterior pituitary lobe. They also found that this lobe not only possesses a growth hormone but possesses a thyrotropic action also. It is well known that acromegaly may exist at the same time as Graves' disease, or may follow it in later years. The author has just seen a case of pituitary disease with typical bilateral hemianopia and early optic atrophy, but this young woman of twenty-six years of age has never had a menstrual period. Such cases are common, and such are also found in Graves' disease. Langdon Brown says that sympathetic irritation is well known to be a factor in producing thyroid enlargement and Graves' disease. Intensive degenerative changes in the sympathetic terminal reticulum have been found by Sunder-Plassmann, which might offer a possible explanation for the often-described psychic trauma co-existing with severe Graves' disease. (See also paper by Harrington.)

In Gardner Hill's etiological study of primary Graves' disease he found that the extrinsic exciting factors such as shock, mental strain and infections have played a prominent part in the onset of this disease in a large percentage of his cases. Psychic trauma alone was responsible for 50 per cent., while infections such as tonsillitis and influenza accounted for 20 per cent. The disease was precipitated in 80 per cent. of his women cases by extrinsic factors such as shock, mental stress and sepsis. He found that sex events in the female—puberty, pregnancy, the puerperium and the climacteric—appeared to be predisposing factors.

A six-year study of thyroid disease in Dunedin Hospital has been reported by Fitchett, Hercus and Bell. They state that the pathological changes which induce toxicity are the same whether they occur in a previously healthy or an already diseased gland. They suggest four groups : (1) simple goitre (the colloid or nodular

type); (2) the thyrotoxic class, including primary, secondary and Graves' disease; (3) hypothyroid states—myxœdema, cretinism; and (4) malignant thyroid disease. Their findings also showed that the iodine content of the blood was higher in primary Graves' disease than in simple goitre, and that the iodine content of blood, fæces, and gland were all lower than normal in simple goitre.

Ophthalmic Signs and Symptoms.—Exophthalmos or prominence of the eyeballs is present in roughly 75 per cent. This proptosis is sometimes unilateral at first; one case that came before the author was unilateral from the beginning until the disease was cured. The cause of exophthalmos is definitely not known. In emaciated subjects excess of fat in the orbits was found on autopsy. If this proptosis suddenly subsides at death then one must admit that the orbital tissues are in an œdematous state. Cushing believes the exophthalmos seen in pituitary tumours is due to blocking of the cavernous sinus and its tributaries from the orbit. Dudgeon and Urquhart have described lymphorrhages (lymphocytic infiltration) in the extra-ocular muscles in exophthalmic goitre. There is indeed a definite change in the function of these muscles. If one has kept a record of the muscle balance of each patient, the discovery of an indefinite muscle imbalance in the same patient later on who had showed a normal balance will help to diagnose this disease in its early stages. For example, a young woman aged twenty years, in 1932 had an esophoria equivalent to 3 prism dioptres, while the vertical error was nil, yet three months later the esophoria had increased to 6 prism dioptres, and now there was a right hyperphoria of 2 prism dioptres. Such a change in the muscle balance could not be produced by a definite paralysis, so further examination of the patient by a physician revealed the fact that this patient was in the early stages of a thyrotoxicosis. I would stress the importance of measuring the muscle balance in every suspected case of thyrotoxicosis. The probable explanation of a muscle imbalance would be the amount of fat infiltrating the extra-ocular muscles, as has been observed by Silcock. Swollen fusiform bellied muscles were found by Foster Moore and Naffziger instead of the normal flat bands. This, too, would tend towards the production not only of an apparent paresis but to exophthalmos as well.

Sometimes the exophthalmos becomes extreme. I remember a case under Treacher Collins' care in which the eyelids would not meet over the eyes. Stitching the lids had proved useless. Sight

was lost due to ulceration of the cornea from exposure. Cecil A. Joll has just performed Naffziger's operation in such a case. The aim of the operation was to enlarge the capacity of the orbits. In this operation a frontal osteo-plastic flap is raised, the orbit approached from above, and the latter decompressed by removing its superior wall. The operation, however, is a severe one, and would not be well borne by all patients suffering from thyrotoxic symptoms.

Patients have complained that they thought one eye had lately become larger, but examination may show in these cases that the apparent enlargement is due to slight protrusion forwards of the eyeball, combined with Dalrymple's sign, which consists of retraction of the upper lid. The larger palpebral aperture gives a false impression to the lay mind that one eye is larger than the other.

In 1859, Mueller described a layer of unstriped muscle bridging over the sphenomaxillary fissure, corresponding to a more largely developed layer found in the extensive aponeurotic part of the orbital wall of various mammalia. These involuntary muscles are supplied by the cervical sympathetic, and in certain animals may have the power of causing some protrusion of the eye ; but Symington states that this can only happen to a very limited extent in man. I confess that while demonstrating a very large number of subjects in the dissecting room I was never able to define such a muscle. There are, however, numerous smooth muscle fibres in Tenon's capsule which are innervated by sympathetic nerves from the cavernous plexus *viâ* the ciliary ganglion and the long ciliary nerves. Starling says that stimulation of the nerves described causes contraction of these muscle fibres, protrusion of the eyes, and rise of intra-ocular pressure. One of the explanations of the protrusion of the eyes in this disease is that it is due to stimulation of the sympathetic nerves in the neck by the local pressure of the thyroid tumour. Removal of the superior cervical ganglion was thought to bring relief in this condition, but the modern surgical treatment of exophthalmic goitre does not tend towards this view. The pathology of the orbit shows that the disease is of a more profound origin. John Griffith found after removal of both eyes in a severe case of exophthalmos that the lids could not be closed over the orbital contents. The marked pulsation of the arteries of the head and neck may be seen in the arteries of the fundus oculi (Gowers), and the latter vessels may show slight enlargement of

their calibre. Venous pulsation at both optic discs was noted by Marcus Gunn.

Choroiditis, cataract and optic atrophy have been seen in cases of exophthalmic goitre, but one cannot say for certain if their presence was a coincidence or an association. The size of the pupils is normal, the pupillary reflexes are normal.

Most of the patients who suffer from progressive exophthalmos have had a normal or low basal metabolic rate. This is significant, for Cole says that in moderately severe cases +40 is a frequent figure.

The amount of proptosis of the eyeballs may vary from time to time. If the extra-ocular muscles have become œdematous (as, indeed, has been observed by Foster Moore), then we can understand the apparent variations that take place in the paresis of these muscles as shown by measuring the muscle balance. Now and then one sees limitation of movements of the eyeballs in all directions. (Apparent external ophthalmoplegia.) Exophthalmos may continue for years after all the other thyrotoxic symptoms have disappeared. (Forty-nine years—Goulden.) Even should the lids meet easily over the eyeball, the protrusion of each eyeball leads to an exposure conjunctivitis and epiphora. Patients often complain of the staring appearance of their eyes, together with this redness, a condition which can be greatly ameliorated by a partial tarsorrhaphy or uniting of the lid edges at the outer quarter. In this way the operation of external cantho-tarsorrhaphy at once hides the deformity and clears up the hyperæmia of the conjunctiva.

Dalrymple's sign consists of retraction of the upper lid, which produces a peculiar staring appearance that is characteristic of this disease. There are other conditions which will produce this staring appearance such as hysteria in women, tentanus, stimulation of cervical sympathetic, local inflammatory and neoplastic conditions of the orbit, tower skull, and high degrees of myopia.

Von Graefe's sign indicates the failure of the upper lid to follow closely the downward movement of the eye. This sign may be present in unilateral proptosis or even in its absence; it is also seen in Thomsen's disease and other forms of goitre. Von Graefe himself believed this sign is due to stimulation of the fibres of Muller's palpebral muscle in the lid from the sympathetic nerves. (Two examples of the pseudo-Graefe phenomenon were described by Gowers in 1879.)

Stellwag's sign indicates the absence or incompleteness of the act of winking. Instead of the normal act of winking taking place about five times a minute, the intervals may be prolonged to minutes or even half an hour. Swanzy believes this imperfect winking is due to insufficiency of the orbicularis rather than overaction of the levator. This insufficiency may be due in part to lessened reflex irritation of the cornea and conjunctiva, and may also be partly related to the diminished flow of tears sometimes found in Graves' disease. It was believed by Sattler that there is a reflex centre in the brain governing the retina, cornea and conjunctiva, and it is therefore due to a disturbance of this centre that Stellwag's sign is brought about. Occasionally the opposite condition prevails and the eyes wink too frequently.

There is also *Moebius's* sign, which consists of a weakness of binocular convergence, and is not the result of exophthalmos. The reader will remember that in encephalitis lethargica there is often a failure of convergence also ; the former, however, is due to the local orbital condition, while the latter is the result of a lesion in the neighbourhood of the third nerve nuclei in the midbrain.

Kocher's sign relates to the immobility of the eyeball when the lid moves upwards and the eye is attempting to follow an object being moved in a vertical direction. This is really the reverse of Von Graefe's sign. Kocher believed that tremor and retraction of the upper eyelid as an early sign of Graves' disease precedes the exophthalmos.

Joffroy's sign is observed when on the patient looking upwards there is absence of the usual wrinkling of the forehead.

Jellineck's sign. This is a very inconstant sign, and consists of pigmentation of the eyelids seen on the outer surface of the closed lids. Joll says it is most conspicuous in juvenile forms of primary toxic goitre.

Rosenbach's sign is that of trembling of the lids while attempting to close them gently.

Many other names have been mentioned associated with such conditions as epiphora, loss of eyebrows and eyelashes, œdema of eyelids, difficulty in everting the upper lids, pulsation of retinal arteries, and limited movements, but these are merely associated conditions and should not be classified as definite signs.

Nervous System.—By extending the hands with the fingers separated a distinct tremor of the fingers can be seen. The patient

often appears to be nervous, while in severe cases mental symptoms such as acute mania may be present. Although worry is considered one of the probable causes of Graves' disease yet the author has recently had a case under observation, that of a middle-aged woman, where a history of worry of any kind could not be elicited. In fact, she was calm and placid, and only on the operating table did she show any signs of nervousness. Fright from air raids, earthquakes, etc., have been responsible for an increase in the normal number of cases presenting themselves. The disease may be complicated by encephalitis lethargica, epilepsy or any form of neuro-syphilis, especially tabes. Headache is common, and visual hallucinations sometimes occur. When mental disorders supervene the patient was probably predisposed to insanity (Joll).

Secondary thyrotoxicosis or toxic non-exophthalmic goitre is associated with the presence of an adenomatosis of the thyroid gland, hence the term nodular goitre with hyperthyroidism. In this form of thyrotoxicosis exophthalmos is rare, or if present is slight, so also are nervous symptoms.

Treatment of Exophthalmos.—The treatment of exophthalmos due to Graves' disease presents a constant problem which seems to remain far from solution. The removal of the orbital fat has been done by Moore. In 1899 Abadie and Jonnesco described the operation for the removal of the cervical sympathetic, this operation being primarily used by Jonnesco in Basedow's disease and for the reduction of tension in chronic glaucoma. Since then the same procedure has been carried out in the attempt to reduce the exophthalmos by lessening the action of the nerves on the thyroid. In some cases the operation proved of some assistance (Juler). Sewing the eyelids together with the idea of holding back the eyeballs in place is hopeless.

Probably the greatest advance in the treatment of thyrotoxic exophthalmos has been recorded by Naffziger. In a severe case where sight was rapidly diminishing, he removed the roof of the orbit (by means of a transfrontal operation) as far as the ethmoid and sphenoidal cells, anteriorly as far as the frontal sinus. Laterally the entire plate was removed and posteriorly as far back as the lesser wing of the sphenoid.

Although the orbital contents bulged markedly through this opening, he did not find any excess fat but discovered that the cone of muscles was greatly swollen, each individual muscle being much

larger than normal and was fibrous in texture. It was due to this condition that each eyeball was proptosed. Microscopic examination of portions of the muscles showed round cell infiltration, marked œdema, destruction of the muscle fibres, complete loss of muscle architecture with increase in fibroblasts and generalised fibrosis. The temporal and thigh muscles however were normal. Apparently the fibrosis followed on the œdema of the muscles. By splitting the muscle cone of origin around the optic nerve the choked disc which had been present was relieved and sight began to improve rapidly.

If such a condition of fibrosis is common in chronic cases of exophthalmos due to Graves' disease, injections of such substances as prostigmine, thyrotropic hormone, however useful they may be in the early stages, can be of little avail. At the commencement of evidence of muscle imbalance daily inunction by iodine ointment of the lids appears to be of some value.

Myasthenia Gravis

Synonym : Asthenic bulbar paralysis

This is a disease chronic in nature, in which the patient suffers from abnormal muscular fatigue, which at first is confined to a small group of muscles, later spreading and even involving the muscles of the limbs. The condition was first described by Wilkes in 1877. The disease does not involve the nervous system, so that there is no interference found in the reflexes, no sensory disturbance, and no fibrillation. It is a disease of the muscles exhibiting great fluctuations in the intensity of the symptoms. Exacerbations and omissions occurring alternately are very characteristic of this condition. The disease shows a predilection for the bulbar muscles, such as the eyelids, face, tongue, throat and neck. It begins in adult life between the ages of twenty and fifty ; females are more affected than males. Ptosis of one or both eyelids is often the first symptom of this disease (40 per cent.) (Oppenheim).

The ocular symptoms are as follows : The patient complains of finding difficulty in keeping the eyelids open for any length of time, especially in the evenings ; reading can be continued for only a few minutes owing to the failure of the action of the muscles of accommodation and convergence ; the ptosis is nearly always bilateral, although the first symptom of which the patient may complain may be a unilateral ptosis. The wrinkling of the brows, which

is a usual feature of ptosis from other causes, is not observed, as the compensatory action of the occipito-frontalis is absent. Ophthalmoplegia externa, partial or complete, follows the ptosis in half the cases. Collier asserted that it is well known that the paralysis of myasthenia gravis may become complete and permanent. The affected muscles are generally much stronger early in the day than towards the evening, but the muscle imbalance is not constant.

As slight exophthalmos is often observed. it is necessary therefore not to confuse myasthenia gravis with exophthalmic goitre, in which there is also a certain amount of fatigue shown by the muscles.

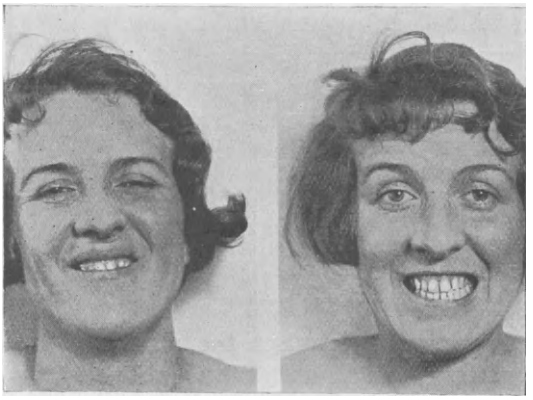


FIG. 118.—In these photos the patient is making a maximum effort to show her teeth with the jaws shut. The weakness of the retractors and the substitution of the elevators of the upper lip is seen in the left-hand photograph, taken before the injection of prostigmine. The increased power of the retractors after injection is seen on the right. (Blake Pritchard.)

Jolly, in 1895, described the *myasthenic reaction* of the nerves and muscles in this disease. He found the faradic current rapidly exhausted the nerve and muscle reactions, but these again responded quickly after rest. He did not observe these phenomena after galvanism.

Due to fatigue of the tensor tympani the range of hearing of high-pitched notes may be diminished.

Boldt and Mendel have observed the *myasthenic reaction* of the pupils to light ; at first the pupil would react promptly, but after a few more attempts were made to elicit the reaction it would become slow and feeble, but was never completely abolished.

In 1934, Dr. M. B. Walker published a description of the unmistakable improvement she had noticed in a case of myasthenia gravis as a result of giving physostigmine. Later Blake Pritchard reported on the results of injecting several patients with prostigmine and atropine. The effect lasted eight hours. Strangely enough the only untoward symptom following the injection was the feeling

of hardness or tightness around the orbits, increased flow of tears and twitching of eyelids. Blake Pritchard says the probable effect of the prostigmine is to delay the action of acetylcholine at the motor endings by the choline esterace normally present in the blood.

Patients may live for years, but in a few cases the disease has terminated life in a few weeks. The outlook is best when the ocular muscles are first affected.

Ocular Lesions associated with the Trigeminal Nerve

The fifth or trigeminal nerve is the nerve of sensation to the ocular apparatus. The course of this nerve from the Gasserian ganglion peripheralwards has been described in Chapter III. The motor root arises as fibres from the motor nucleus of the fifth nerve beneath the floor of the upper part of the fourth ventricle, and joins the bundle known as the descending motor root of the fifth nerve which springs from a long column of large nerve cells close to the cerebral aqueduct and which extends from the superior corpus quadrigeminum to the level of the main motor nucleus. These fibres leave the pons above the sensory root and passing forward come to lie beneath the ganglion, where they join the mandibular division of the fifth nerve. The sensory fibres pass backwards from the Gasserian ganglion situated in the cavum Meckelii on the apex of the petrous portion of the temporal bone, from whence the fibres cross the subarachnoid space to reach the side of the pons at the union of the latter with the middle cerebellar peduncle. Within the pons, medulla and spinal cord a large part of these fibres called the descending sensory or bulbo-spinal root pass as low as the level of the second cervical root, giving off collaterals which arborise round cells of the substantia gelatinosa of Rolando. The three main groups of cells constituting the bulbo-spinal root are the nucleus sensibilis *a* of Winkler, the nucleus sensibilis *b* of Winkler, and the nucleus gelatinosus. Those fibres, called the ascending or mesencephalic root, arborise round cells constituting the sensory nucleus of the fifth nerve placed to the outer side of, and somewhat deeper than, the main motor nucleus. Hewer and Sands state that the secondary neurone is constituted by fibres from both these nuclei crossing and running up as the trigeminal fillet (found lateral to the median fillet) to the optic thalamus. The tertiary neurone, being formed by fibres from the

optic thalamus, passes through the anterior part of the anterior limb of the internal capsule to the post-Rolandic cortex. (See Fig. 119.)

Not only to the eye and its adnexa is the trigeminal nerve the nerve of sensibility, but also to the face, part of the external ear, nose, the mouth and dura mater. In addition it supplies secretory fibres to the lacrimal gland, also dilator pupillæ fibres which leave the naso-ciliary nerve by the long ciliary nerves, while fibres pass to the unstriated muscle of the levator palpebræ superioris. It joins up with the cervical sympathetic conveying vaso-constrictor impulses, and finally it is probably trophic in function also.

Neuroparalytic Keratitis

When for the relief of trigeminal neuralgia the Gasserian ganglion is extirpated or its function abolished by injection of alcohol, one of the first results is a flushing of the vessels of the conjunctiva covering the eyeball—lasting for a week or so. If the eye is protected by a pad and bandage with a very soft lubricating ointment containing 5 per cent. boric acid the eye will in all probability remain sound. Sometimes, however, in spite of every care the cornea becomes denuded of its epithelium, first in the centre and then gradually spreading to the periphery. The cornea becomes slightly cloudy, and if at this stage due care is not taken the cornea becomes more opaque in appearance, turning a yellow colour, and then breaks down in a state of ulceration with pus in the anterior chamber (hypopyon). There is marked ciliary injection around the corneal margin of the sclera as in iritis, but owing to the anæsthesia pain is not present and lacrimation is absent, or nearly so. Fortunately many cases recover from the desquamation of the epithelium, and not even showing a trace of the disturbance, but often a scar of more or less density is left behind, even although ulceration has not taken place. Of 60 cases of alcoholic injection for destruction of the Gasserian ganglion Hinds Howell found 36 had complete anæsthesia over the area supplied by the first division of the trigeminal nerve ; 16 of these developed corneal ulcer. He said the period of greatest danger of corneal ulcer supervening was in the first fortnight following the injection. This condition is a peculiar one, in that, although it is associated with a lesion of the trigeminal nerve, yet in herpes zoster ophthalmicus, where often there is anæsthesia over the whole distribution of the ophthalmic division

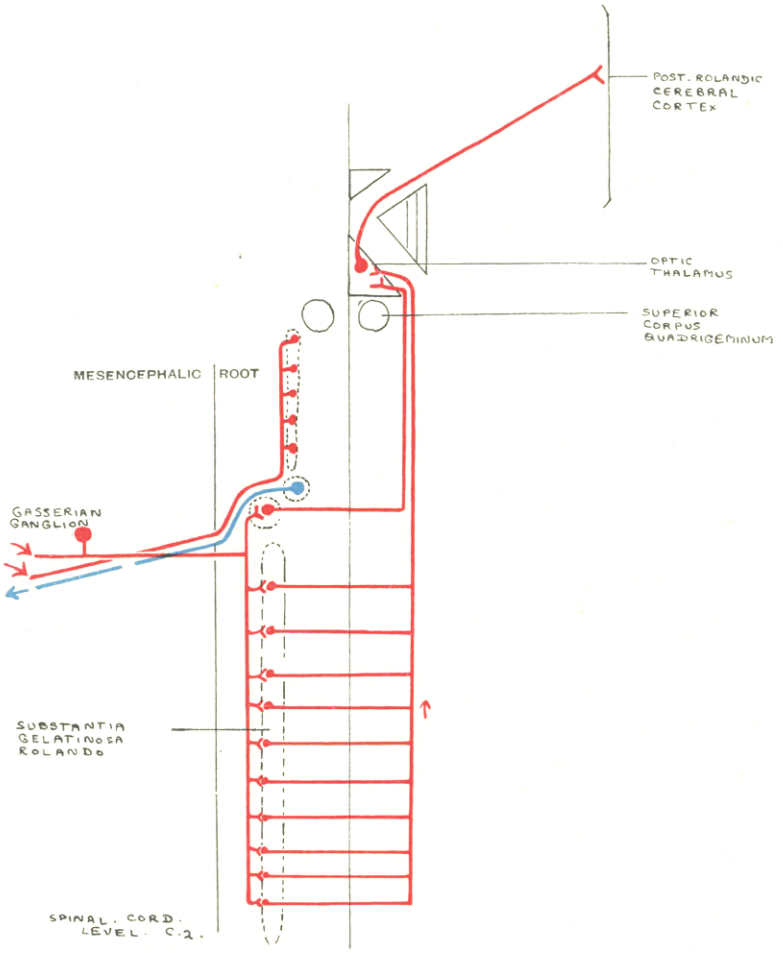


FIG. 119.—Diagram of the path of the fifth nerve fibres in the brain. Red: sensory. Blue: motor. (From Hewer and Sandes' "Study of the Nervous System.") (Wm. Heinemann Medical Books Ltd.)

[*To face p.* 348.

of the trigeminal, desquamation does not take place. The epithelium of the normal cornea if not moistened will rapidly show desiccation and become exfoliated. During ophthalmic operations when the lids are retracted by means of a speculum it is necessary, from time to time, to moisten the cornea with normal sterile saline, or place a simple contact glass filled with normal saline on the cornea in order that the epithelium shall be preserved intact.

Several things occur which are responsible for the condition of neuroparalytic or neurotrophic keratitis. The trophic influence is removed from the delicate epithelium ; there is diminished secretion of the lacrimal gland, and so the effect of the latter is enhanced by the former in a much more marked degree than if normal corneal resistance and sensation were present. An analysis of the recorded cases shows that the keratitis occurs only in paralytic lesions of the fifth nerve, and that this disease is due to irritative changes in connection with the degenerating nerve, and that the development of the affection is assisted by the diminished lid-reflex and lacrimal secretion ; the desiccation and exfoliation of the corneal epithelium make it possible for the entry of septic infection to the cornea. The disease therefore cannot be regarded as of neurotropic origin in the strict sense of the word. (Swanzy.) The trophic disturbances which are to be found associated with nerve lesions according to Charcot are not the result of the absence of the normal trophic impulses, but of inflammatory irritation. The most varied trophic disturbances are to be found when there is disordered activity or irritation of nerve fibres or their nuclei. The trophic theory was first put forward by Majendie in 1824.

Duke-Elder has recently suggested that the formation of histamine in the tissues is under the control of impulses passing antidromically down the sensory nerves, presumably because these nerves control the actual metabolism of the cells. He states that the loss of the normal activity of these nerves will rob the tissue of this means of control, and it would seem likely that this loss and the subsequent antidromic release of chemical materials of this nature which follow a lesion or irritation of the fifth nerve are responsible for the type of disturbance which has been termed trophic.

Neuroparalytic keratitis occurs in all cases of bilateral paralysis of the fifth nerve, but the consensual reflex apparently affords protection when only one nerve is affected. The condition is much

more likely to take place when the facial nerve is also involved, with resulting imperfect closure of the eyelids—lagophthalmos. (Paton.) Denny and Neve have noticed this corneal condition in advanced leprosy and state that it can be brought about by the lagophthalmos of leprosy, one ulcer following another until the cornea is transformed into a dense scar which completely destroys vision.

The lesions producing neuroparalytic keratitis are not confined to the Gasserian ganglion, for further causes of this condition are to be found in fracture of the skull, intracranial neoplasms, and gummatous basal meningitis. Paton reported a case of Foville's syndrome due to a tuberculoma in the anterior part of the medulla in which a severe neuroparalytic keratitis subsequently developed.

The treatment of this condition consists in closure of the lids by a dermal suture following the injection of the ganglion ; this is done by making raw the juxtaposed edges of the lids for about 5 or 6 millimetres a little more on the outer side of the middle line, avoiding the lash-bearing margin, and bringing the lids together by means of two or three fine sutures. The lids may have to remain closed from three to six months. Leslie Paton emphasises the fact that even a small band holding the lids together is sufficient for the protection of the cornea. Within three days from sewing the lids together the corneal epithelium may have regenerated and the cornea become clear. The utmost care has to be taken in finally breaking down the lid adhesions. Others, such as Swanzy and Vaughan Gudden, have relied on simple bandaging of the eyelids together. Cases of the author's have remained perfectly clear and transparent for several years, although corneal anæsthesia has persisted, when the treatment at first was that of bandaging the lids, daily cleansing by lukewarm boric lotion and the application of a soft boric ointment. Before removing the Gasserian ganglion Horsley was accustomed to stitch the eyelids together as a protection for the cornea, and sometimes twisted the central root out of the pons. Hinds Howell, however, points out if the ulceration is due to peripheral nerve irritation it should occur whether the lids are stitched or not.

Herpes Zoster Ophthalmicus

In the year 1867 Bowman described several cases of herpes frontalis seu ophthalmicus, one of which he illustrated, showing the

permanent scars left on the face after the attack had passed off; another case he mentioned was one in which the attack was followed by optic atrophy, while yet a third was accompanied by diplopia. A year previously Jonathan Hutchinson had given his classical account of the disease. Since then a great deal of work has been done in many countries in the investigation of the pathology of the disease and its relationship to varicella. A portrait of the disease was referred to by Hutchinson as being in the atlas published by Danielsen and Boeck, and a wax cast having reference to it is in the Guy's Hospital collection—the case of a boy aged five years.

Clinical Aspects.—Herpes zoster ophthalmicus is a disease of middle life. It is most commonly seen after forty years of age, the average age being fifty-five (Lodge), although Hutchinson reported a case as early as four years of age, and Fisher related a case of a child aged nine months. The author has just seen an extremely severe case in a man seventy-five years of age; the disease, indeed, seems to be much more severe in elderly people than in the young. This disease attacks males and females almost equally in numbers. Two distinct types manifest themselves, namely idiopathic zoster and symptomatic zoster; the first being an acute epidemic disease caused by a filtrable virus running a definite course with constitutional disturbances such as sickness and temperature. The first local symptom of herpes ophthalmicus is neuralgic pains over the distribution of the ophthalmic division of the fifth nerve, one to four days preceding an eruption of the skin which is strictly limited to the midline of the forehead. Some patients have stated that both pain and eruption began simultaneously. There is a distinct erythema as well as vesiculation, the latter rapidly breaking down, together with a certain amount of œdema which involves the upper eyelid; the lid, indeed, being in a state of ptosis, especially if it is the seat of vesicles; the œdema, moreover, may spread to the cheek below and to the outer side of the eye. At first the vesicles are filled with clear fluid which is sterile (Paton), later becoming turbid and yellow. I have seen a case where the pain was so severe that the patient had not slept for nine nights, and when seen only one large blister was found extending from the eyebrow to the scalp, probably due to coalescence of several smaller vesicles. This patient, though it is now six years since the attack, still suffers severe pain, the relief of which has defeated all known therapeutic means. I have not attempted to do what Bowman did

in a similar case ; that was to sever the supra-orbital nerve subcutaneously. The results do not encourage me to copy his example. Many cases presenting themselves for the first time show a number of ruptured vesicles drying up and leaving deep and permanent scars, indicating that both the dermis and epidermis of the skin have been involved, the period of vesiculation, rupture, and drying up lasting about three weeks. At this stage the pain is found to be passing away, leaving the skin in a more or less anæsthetic state, although many complain of the persistence of pain. Of the three branches of the ophthalmic division, namely the supra-orbital, the supra-trochlear and the naso-ciliary, the first two are generally affected or all three may participate, but the naso-ciliary may escape. Head and Campbell describing the pathology of herpes zoster report a case where the eruption followed the distribution of the third or maxillary division of the trigeminal, and Paton has seen a case where the first and second divisions were completely involved. Symmetrical herpes on both sides of the face has been described. Head and Campbell's case belonged to the idiopathic type, but in sympathetic zoster the distribution is not so commonly confined to the ophthalmic division. In 50 to 60 per cent. of the cases the eye is involved showing both extra- and intra-ocular complications. When the eye is involved the conjunctiva becomes chemosed and injected, but vesicles on the conjunctiva are rare. Penman describes 4 cases which exhibited a form of scleritis occurring two or three months after an attack of herpes ophthalmicus. These showed one, two or more nodules of a dark red colour, and about the size of a lentil beneath the conjunctiva. The nodules were slow in subsiding, gradually getting smaller and darker till after some months they finally disappeared leaving sharply defined, slatey grey areas in the sclera where they were situated. Wilbrand and Saenger mention sclero-keratitis and scleritis as a complication of herpes ophthalmicus. Many cases are seen where apparently the conjunctiva alone is affected, yet if a fine wisp of cotton-wool is applied to the cornea there is an absence of reflex

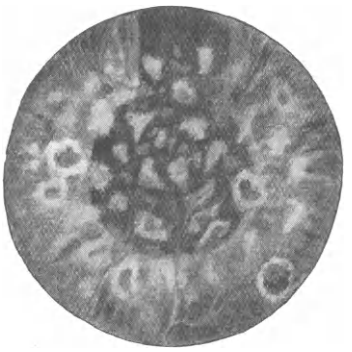


FIG. 120.—Calcification of the cornea sequel to an attack of herpes ophthalmicus. (Harrison Butler.)

winking, showing the cornea has become anæsthetic. These are the cases in which the eye is, in the author's experience, most liable to damage through not impressing upon the patient the necessity of keeping the eye closed with a bandage or straw-plate shield. Cases present themselves at ophthalmic hospitals showing an eye the cornea of which is partly calcified (see Fig. 120). Examination of the skin of the forehead above the eye reveals old herpetic scars. Questioning the patient reveals that little or no care was taken of the eye during and after the active part of the disease. Other conditions show in addition to the chemosed conjunctiva involvement of the cornea, and many forms of keratitis may occur. There may be superficial opacities just beneath the epithelium or deep to Bowman's membrane, which may be scarcely visible, or may be clearly seen as white flakes. Fig. 121 represents the appearance of the cornea in this condition as seen through the slit-lamp. The area on the left of the illuminated portion shows the surface of the cornea, the hollow-shaped area on the right being the depth of the cornea. The opaque dots therefore are found in the more superficial layers of the cornea to about one-quarter of its depth. In addition, herpetic vesicles, phlyctenulæ and bullæ may be seen, and any of these may break down into ulceration. Without ulceration, deep keratitis (keratitis profunda) may follow, and remain so that vision may be greatly diminished due to scarring or opacities. It is the practice of many ophthalmic surgeons to sew the lids together when signs of corneal involvement present themselves; but I have to confess that my practice of bandaging the eye from the onset of the disease with its daily toilet of weak boric lotion and ointment has given me such results that not in one

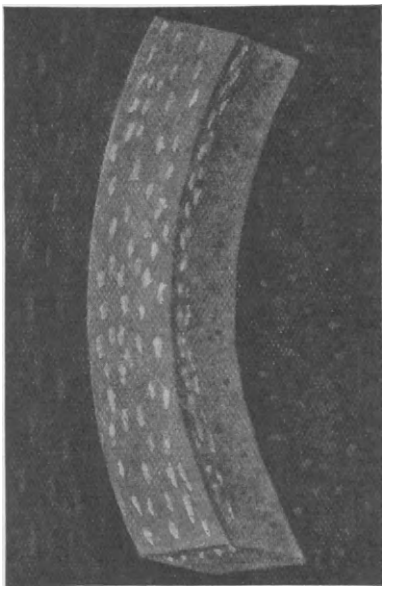


FIG. 121.—Slit-lamp appearance of cornea in which keratitis had quickly supervened on an attack of herpes ophthalmicus. The white opaque foci are situated in the anterior quarter of the cornea under Bowman's membrane. (Harrison Butler.)

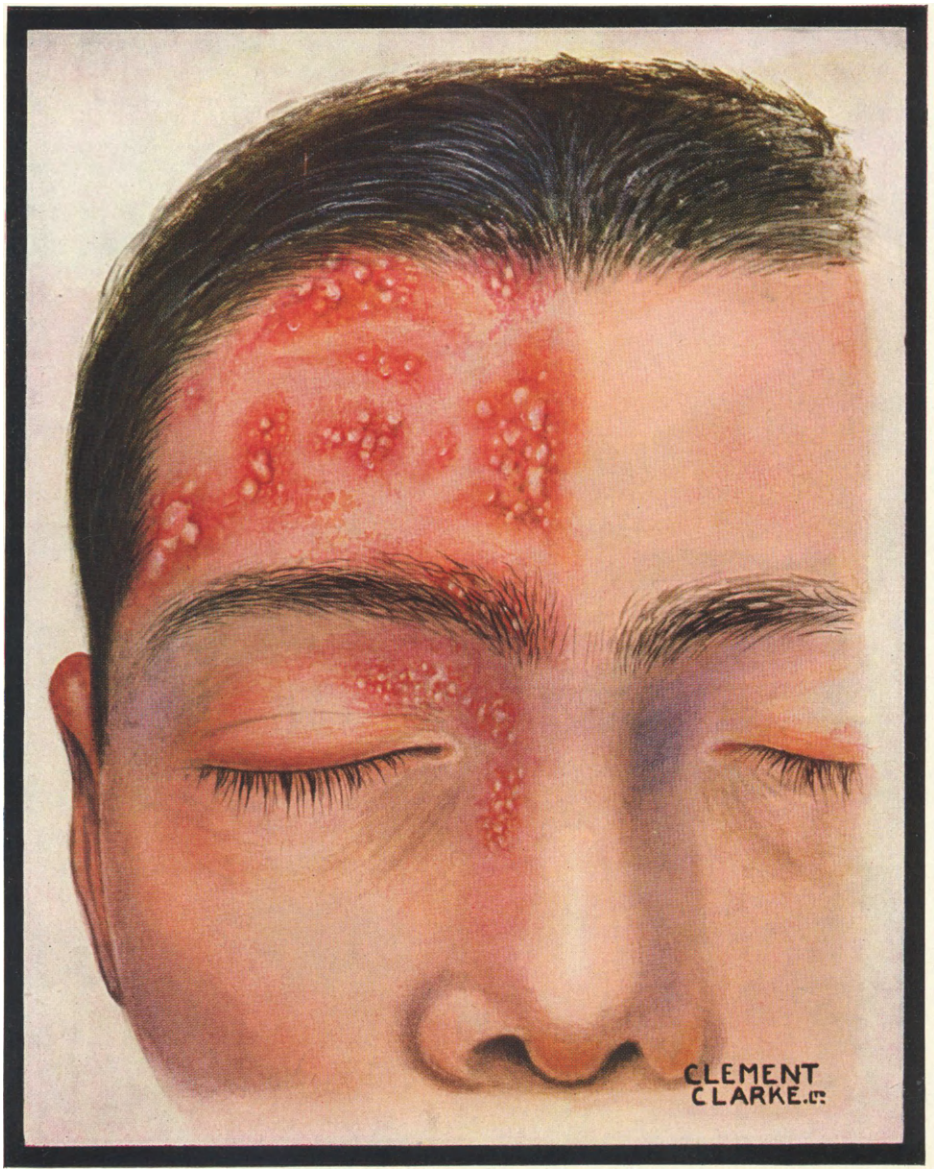
single case have I been forced to unite the lids by suturing. Anæsthesia of the cornea, partial or complete, may remain; recovery is slow and sectors of the cornea may remain permanently insensitive. Iritis and irido-cyclitis are sometimes present, the latter throwing down keratitic precipitates (k.p.) on the posterior surface of the cornea. I have never seen the formation of cyclitic membrane in the vitreous even where keratitic precipitates were present, nor has the experience vouchsafed to others of seeing an associated optic atrophy been mine yet. When iritis is present atropine 1 per cent. can be added to the boric ointment. Paton says iritis has been seen immediately before the skin eruption. Where iritis is not present the pupil may be reflexly miotic, or it may be dilated due to paralysis. Choroiditis and retinitis have been seen, but these appearances are exceedingly rare. Either raised or lowered tension of the globe may occur.

Ocular palsies associated with herpes ophthalmicus have been described. Metz has enumerated fifty cases of ocular paralysis associated with this disease of which involvement of the oculomotor nerve was 74 per cent. Collier in Price's "Practice of Medicine" says that syndromes have been observed in the eye where a prodromal attack of pain has been followed, not only by corneal anæsthesia, but also by a motor nerve paralysis (internal ophthalmoplegia)—a "motor herpes." Wyss found numerous hæmorrhages in the muscle substance of the ocular muscles, hence ascribing the paralysis to a thrombo-phlebitis (see Fig. 122).

In a paper written by Chance he quotes Hunt, who found that of 158 cases of ocular paralyses of various kinds associated with herpes ophthalmicus 18 cases involved the third cranial nerve, 1 the fourth, and 5 the sixth nerve. Immobile pupils may be the result of ophthalmoplegia interna, but may also be caused by posterior synechiæ from the presence of iritis. Possibly encephalitis may be at the root of some of these associated palsies. Pointing out the rarity of the facial nerve involvement Chance also states that these palsies have shown a slow but complete recovery as a rule (see Fig. 122).

Herpes ophthalmicus has been, and is, so frequently mistaken for erysipelas that one cannot do better than quote Hutchinson's own words regarding its diagnosis. He said: "Herpes frontalis is always limited to one side. It never transgresses the median line of the forehead and nose. It never affects the cheek, although there may be some sympathetic œdema (œdema of contiguity).

PLATE XVI



HERPES ZOSTER OPHTHALMICUS

The vesicles are unilateral and do not transgress the middle line. The naso-ciliary nerve is involved. (Drawn from a case under the author's care.)

[*To face p.* 354.

There is less general swelling of the skin than in erysipelas, and in some cases very little. The vesicles of herpes are smaller, more defined, more numerous, and altogether much more conspicuous and prominent than are the bullæ of erysipelas (see Plate XVI). There is much more pain and much less constitutional disturbance in herpes than in erysipelas. The strictly unilateral character of the one contrasted with the irregular location of the other is, however, a most reliable feature for the purpose of ready diagnosis." A patient suffering from herpes may be confidently assured there is no danger except to the eye, and, most probably, he will have neither a relapse nor a recurrence. For the purpose of prognosis it might be as well to remember Hutchinson's dictum that if the nasociliary nerve is affected and vesicles form on the side of the nose, the eye is most likely to be involved. But this is not invariably so.

Pathology of Herpes Zoster Ophthalmicus.—The changes that take place in the Gasserian ganglion during an attack of idiopathic or epidemic herpes ophthalmicus are similar to the changes which take place in a dorsal root ganglion in herpes zoster. Sattler, in 1875, examined the Gasserian ganglion of a case in which death supervened fourteen days after the attack. He found the ganglion infiltrated with small round cells with marked destruction of the ganglion cells. The ophthalmic division was also degenerated. Head and Campbell also found small celled infiltration, hæmorrhagic extravasation, destruction of ganglion cells, together with inflammation of the nerve sheath closely resembling the changes seen in acute anterior poliomyelitis. If the attack has not been severe there will be little or no trace left behind in the ganglion.

Etiology.—The disease is caused by an ultra-microscopic virus. Just as symptomatic zoster may occur when destructive lesions caused by tubercle or cancer affect the cord and its envelopes with the dorsal root ganglion, so may symptomatic herpes frontalis be caused by arsenic, mercury, bismuth or carbon monoxide poisoning, also in the course of infections such as pneumonia, tuberculosis, meningitis, cerebral tumour and sub-arachnoid hæmorrhage. In these cases the neuralgia may precede the eruption by many days in contrast to what is found in the idiopathic form, where the pain and constitutional disturbances precede the eruption by one to four days. However, as Head and Campbell have shown, the histological changes of both are indistinguishable.

Relationship of Herpes Zoster Ophthalmicus to Varicella.—Since

1888, when Bokay observed a case of varicella which had occurred ten days after being in contact with herpes zoster, many writers have drawn attention to the relationship between herpes ophthalmicus and varicella. Netter alone recorded 174 cases. Both of these diseases occur together in epidemics. Brain points out that

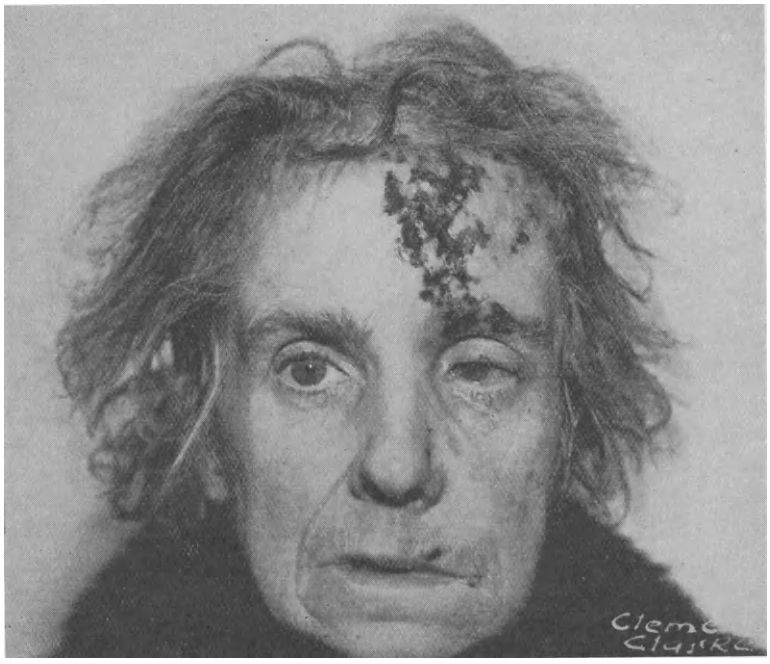


FIG. 122.—A severe case of herpes ophthalmicus accompanied by paralysis of the sixth and seventh cranial nerve. The corneal epithelium was rough and bullous in appearance and the pupil was partially dilated. Ichthyol ointment was responsible for delineating the vesicles on the skin which extended not only to the nose but as far as the upper lip. (Case under care of Dr. Keppel Barrett.)

in Bokay's data concerning the epidemics of zoster and varicella the peak of the zoster curve usually occurs one month before that of varicella. Netter and Urbain investigated the fact that there were antibodies to varicella in the blood of the patient suffering from zoster. They found deviation of complement occurred when the crust of vesicles either from varicella or from zoster were used as antigens against the blood serum of the zoster patients, and the

same results were obtained with the same antigens and the blood serum of patients suffering from chicken-pox. It is possible that herpes zoster and chicken-pox are essentially the same. Examining the vesicle fluid in thirty-two typical cases of herpes zoster, Aimes found in twenty cases elementary bodies morphologically similar to those found in varicella and vaccinia. Pure suspensions of these bodies prepared by high-speed centrifugalisation of zoster vesicle fluid were specifically agglutinated by zoster convalescent serum. Ransom Pickard's view is that " the virus causing varicella and herpes zoster has two phases, easily transmutable. The herpes zoster phase has its primary seat in a posterior root ganglion. It may spread locally, involving adjoining nerves in the case of the Gasserian ganglion ; but its main and normal extension is along the sensory nerves to fibrous tissue covered by epithelium, where it reverts to the varicella phase. It there produces nodules, usually with vesication, sometimes accompanied by ulceration and even sloughing. The affections of the eye in herpes ophthalmicus are explained by this theory, as are those epidemics of herpes zoster unaccompanied by varicella."

It is interesting to note that a patient under arsenical treatment developed herpes zoster, and this was apparently followed by varicella in a child contact (Parkes Weber). Savin has recently suggested the hypothesis that zoster and varicella are caused by modified strains of the same parent virus, which would explain the interchangeable complement-fixation reactions in the two diseases, and the high protection each disease gives against its own recurrence ; also the simultaneous appearance of herpes zoster in patients presumably exposed to a common source of infection. Zoster, varicella and vaccinia have each in common the power to produce a form of encephalitis (Horder, Brain, Perdrau and Winnicott).

Treatment. Immediately the case is seen for the first time the eye should be bandaged. Each day the eye is gently bathed with weak lukewarm boric lotion, and a soft boric ointment (5 per cent.) composed of paraffin and lanoline inserted between the eyelids. Should signs of iritis appear 1 per cent. atropine can be added to the ointment. It is not necessary to stitch the lids together, for the conjunctival sac is kept in a cleaner condition when the lids can be freely opened. The patient should be warned of the danger to the sight, and impressed with the fact that the lids must be closed for three months. In all the cases one sees when the sight

has been lost, it has been found that the patient has not been instructed to protect the eye in the way I have just described. Fig. 120 is an illustration of one such case. It should be remembered that the tension of the eye may increase so that eserine must be used, and also that an attack of glaucoma may supervene months after the disease has passed. In one of the author's cases, a man aged fifty years, an attack of sub-acute glaucoma came on sixteen months after the onset of the zoster. The cornea had remained partly anæsthetic, and minute opacities could still be seen scattered throughout the cornea which had not affected normal vision. This patient years previously had an attack of varicella which had left a large pock-mark above the left eyebrow, and now when attacked by herpes ophthalmicus the first vesicle that appeared was on the site of the old pock-mark, and it was the last to heal. Patients should be kept in bed and warmth applied to the affected parts. Although the skin lesions are unaffected by treatment, yet some relief can be given by applying cocaine ointment 1 per cent. and dusting with starch powder, or calamine-zinc-oxide lotion may be applied. Tincture of benzoin (3 drams to 1 ounce of rose ointment) can be applied to the skin several times daily, while opium and quinine can be given internally. Salicylates may be used to lessen the pain, but Sidlick found the pain relieved for some unexplained reason by intramuscular injection of pituitary (0·5 to 1 cubic centimetre of the ordinary obstetrical preparation given once or twice at forty-eight-hourly intervals). Sanford Gifford has seen several cases in which three such injections were followed by complete relief of pain, lasting two to three days, and not returning after the third injection.

It may be of interest to know that marked alleviation of pain in cases of herpes zoster has been brought about by the injection of thiosinamin ethyl iodide in doses of 1 cubic centimetre daily. It has rarely been necessary to give more than six injections. In post-herpetic neuralgia the same treatment has been found to be effective provided the condition has not existed too long before beginning treatment.

The use of percaine benzyl alcohol in olive oil has been suggested to me by F. Silcock. He injects 2 c.c. through a No. 14 Record needle above the level of the eyebrow.

As already mentioned, there are some cases where apparently nothing can relieve the post-herpetic neuralgia. This pain is probably

of central origin in the pons, and cutting of the sensory root does not always relieve it. Grinker says the only available treatment then is Roentgen radiation to the Gasserian ganglion and pons.

Herpes Febrilis

Herpes corneæ is a condition in which small vesicles form upon the cornea in febrile diseases, especially those of the respiratory tract, such as bronchitis, whooping-cough and pneumonia ; and is perhaps most commonly seen in epidemic influenza. Those who, like the author, saw this epidemic among soldiers in France, will remember that in addition to tender and inflamed eyes small herpetic vesicles were observed on the lips, the alæ of the nose and ears, and even on the genitalia. Herpes corneæ is usually unilateral, and is on the same side as the vesicles upon the face. The vesicles on the cornea are scarcely the size of a pin's head, and are arranged in groups, breaking down rapidly and healing without scar tissue being formed. They are accompanied by pain, lacrimation and photophobia. The condition may prove very obstinate, and instead of healing a dendritic ulcer may follow. Ophthalmic surgeons are familiar with the various forms of corneal ulcer, but one is always struck with the intractability of this particular type. The ulcer heals so slowly, and there is something much more than local involvement. It is indeed a neurotropic condition.

Gruter discovered that herpes febrilis was transmutable, and that the various forms of simple herpes were related to each other.

The virus found in the vesicles of herpes febrilis is an ultramicroscopic filter-passing organism. When the contents of a herpes vesicle is injected into the cornea of a rabbit a condition of herpes corneæ is produced ; but in addition the infection, by passing up the nerves to the central nervous system, there produces encephalitis —*encephalitis herpetica*. While it was contended by Levaditi and others to be the same as encephalitis lethargica, Goodpasteur does not agree with this. Levaditi believes the herpes virus has an affinity for those tissues derived from ectoderm, the cornea, skin and central nervous system. Normally the virus is dermotropic, and so produces lesions in the cornea and skin, but when its toxicity is increased it becomes neurotropic, and thus causes lesions in the central nervous system.

For many years physicians have observed that the occurrence of herpes febrilis is of good prognostic significance in acute in-

fections. In the nineteenth century statistics showed that patients with herpes had a 2–9 times lower mortality than those without, especially in meningococcal meningitis, pneumonia and malignant diphtheria. Naegeli has observed this also in the fever therapy of neurosyphilis. According to him the good prognostic significance of herpes febrilis depends on the neurotropic toxins of the virus working antagonistically on the other organisms.

The treatment of *dendritic ulcer*, which is the result of herpes febrilis, should be by touching the ulcer with absolute alcohol or carbolic acid, atropine and boric ointment inserted between the lids and the eye kept closed. Ultra-violet irradiations could be profitably employed.

Probably the best treatment of herpes corneæ is that described by Gundersen. The cornea after cocainisation is painted with a strong solution of iodine containing 7 per cent. iodine and 5 per cent. potassium iodide in alcoholic solution. With the lids held widely apart the cornea is rubbed over by an orange stick swab dipped in the above solution until a deep chestnut brown colour is obtained. Further penetration of the iodine is stopped by another application of cocaine which forms an insoluble cocaine iodide. The cornea changes from a lighter colour until it becomes white. The epithelium now becomes necrotic so that in twenty-four hours the cornea is entirely denuded of epithelium and a smooth glistening Bowman's membrane can be seen. Complete re-epitheliation quickly follows. The patient may require sedatives to allay the pain during the succeeding four hours after treatment. Gundersen's paper is well worthy of careful study.

While recounting the ocular lesions associated with the trigeminal nerves, Foster Moore would have us include a special type of ulcer which appears years after the patient has been exposed to mustard gas. Just as occurred in the beginning of the trauma, the conjunctiva and the cornea again become milky-white in appearance with vessels irregular in calibre passing over the affected conjunctiva. No further change may take place ; but on the other hand, a deep ulceration may set in with very little degree of discomfort—pain, lacrimation and photophobia being almost completely absent. Closure of the eyes by tarsorrhaphy brings about a cure.

Other conditions such as filamentary keratitis, on account of their trophic nature, might be included in this list of lesions associated

with the trigeminal nerve. Although it may seem unnecessary to have described the motor root of the trigeminal nerve, yet recent work (Grout) has shown that the Marcus Gunn phenomenon can be abolished by section of the nerve to the external pterygoid on the side of the lid-jaw movement. Therefore it will not be out of place at this point to mention briefly Sinclair's classification of abnormal associated movements of the eyelids and jaw.

SERIES I. *Group* 1.—Cases of one-sided congenital ptosis in which the drooping eyelid is raised both when the mouth is opened (digastric ?) and also when the jaw is directed to the opposite side (external pterygoid).

Group 2.—Cases of one-sided congenital ptosis in which the drooping eyelid is raised when the jaw is depressed, but is not raised with lateral movement of the jaw.

Group 3.—Cases of one-sided congenital ptosis in which the drooping eyelid is raised with lateral movement of the jaw (external pterygoid action), but not with simple opening of the mouth (the Marcus Gunn phenomenon).

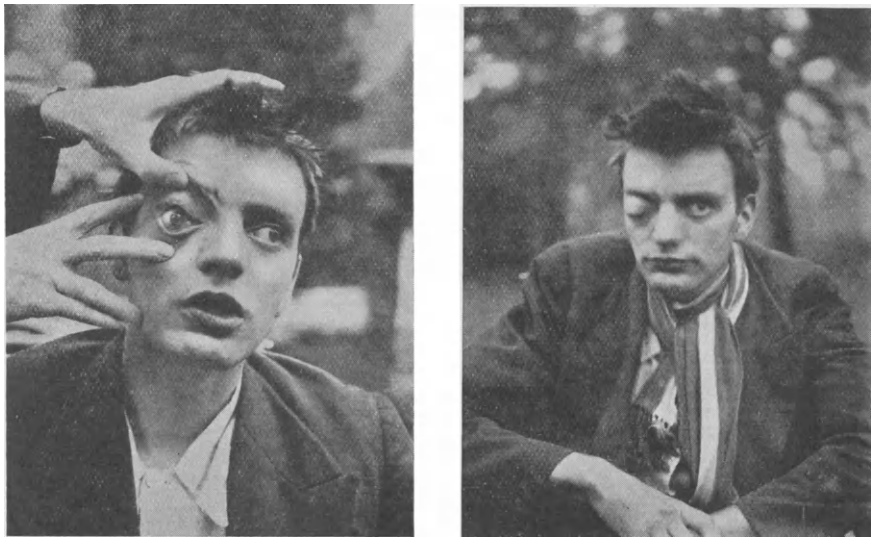
Group 4.—Cases in which similar associated movements of one upper eyelid with movements of the lower jaw occur, but in which there is no ptosis.

The Phakomatoses

Under the heading of "*phakomatoses*," van der Hoeve has grouped the three diseases—*tuberose sclerosis* (which he calls the phakomatosis of Bourneville), *von Hippel-Lindau's disease*, and *von Recklinghausen's disease*. A phakos is a nævus without nævus cells, and so van der Hoeve calls these diseases the phakomatoses (from phakos—the mother spot).

Carnegie Dickson would increase the number of diseases which form tumour syndromes. He points out that not only in a single tumour can there be "polyvalency" in its cells, but in a single instance of a disease (see Fig. 123) there can be an extraordinary variety of tumours present. The case illustrated by Fig. 123 was under the care of Dr. C. Worster-Drought, who in conjunction with Dr. McMenemy, will shortly publish the case in full. There were present not only two large bilateral acoustic tumours—so-called cerebello-pontine tumour—showing a structure somewhat intermediate between a typical neurofibroma and a typical meningioma, but there were also small tumours on all the cranial nerves. The

proptosis of the right eye was due to a meningioma spreading along the ethmoid wall, with invasion of the ethmoid sinuses and orbit. There were also numerous growths—meningiomata and meningiomatous plaques of various sizes—projecting inwards from the inner surface of the dura, also small tumours on almost all the nerve roots of the spinal cord. At many levels of the cord there were central ependymal proliferations and tumours as well as



A. B.

FIG. 123.—A case of multiple tumours. A shows the right eyeball displaced forwards and outwards by a meningioma spreading along the ethmoidal wall. B illustrates the presence of paralytic ptosis due to the presence of intracranial tumours on the cranial nerves including the oculo-motor. Bilateral papilloedema was present while vision was still nearly normal.

gliosis and the formation of syringomyelia-like cavities. Carnegie-Dickson described this case and others at the Section of Pathology and Bacteriology, B.M.A., Dublin, 1933. He put forward the suggestion that there is a distinct relation between the various groups of cases or types characterised by the presence of multiple tumours involving especially the central nervous system, thus enlarging the group suggested by van der Hoeve.

The following table after van der Hoeve shows the phakomatoses so named by him, and gives an idea of the many signs and symptoms produced by these three diseases. The list does not include glaucoma,

buphthalmos, epilepsy, mental defects, acromegaly, dwarfism or plexiform neuromata (see page 371).

All the phakomatoses may be accompanied by congenital abnormalities, such as horseshoe or single kidney, syndactyly, etc., which facts should be borne in mind when patients suffering from adenoma sebaceum may present themselves for other apparently unrelated complaints.

Tuberose Sclerosis. *Synonym :* Epiloia (Sherlock)

This rare congenital disorder, described by Hartdegn and Bourneville in 1881, chiefly affects the cerebrum. It is associated with adenoma sebaceum and tumours elsewhere in the body, together with epilepsy and mental deficiency. It begins in early life ; the adenoma sebaceum appearing on the face in the naso-labial folds as early as the fifth year, later spreading on the face in a butterfly fashion. Apparently the disease is not found in the coloured races. It is a congenital abnormality occurring early in embryonic life. Fabing reports a case of tuberose sclerosis with epilepsy in identical twins. He suggests that the lesion which was destined to evince itself as tuberose sclerosis was present shortly after insemination of the ovum. In this disease the naked-eye appearance of the cerebrum presents numerous whitish, slightly prominent areas, which on examination are more palpable than visible. Tumours may occur in the retina without accompanying mental or physical changes. Van der Hoeve discovered such a retinal tumour, but its real nature was revealed when investigation into the family history laid bare the fact that five brothers and sisters were suffering from advanced tuberose sclerosis. He has also observed white spots in the retina, either combined with, or without tumour formation ; these retinal tumours, he states, are found in the fibre layer of the retina, and consist of neural fibres or neurocytes, or both. They nearly always contain cysts, and these cells may readily become malignant.

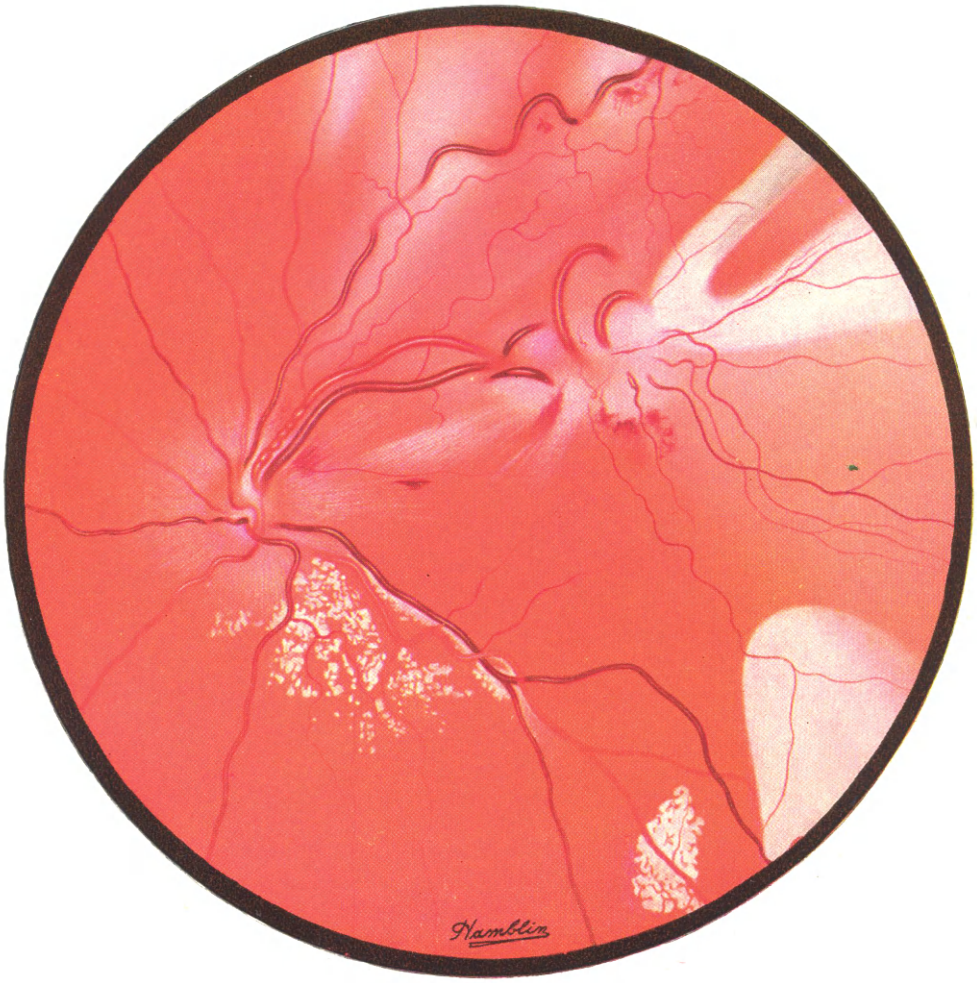
Lindau's Disease or von Hippel-Lindau's Disease.—Angioma of the brain occurring in connection with angiomatosis of the retina may be found associated with such congenital abnormalities as angiomata of the spinal cord, cysts of pancreas and kidneys, hypernephroma, and cavernosis of the liver. Treacher Collins was one of the first to describe, in this disease, a capillary nævus of the retina, and to demonstrate that tortuosity of the blood-vessels of

the eye may be associated with hæmangiomatous formations in the skin, and at times occur in connection with symptoms indicating pressure on the cerebrum probably due to some tumour formation. Angiomatous cysts of the cerebellum frequently co-exist with retinal angiomas, hypernephromas or cystic disease of the kidneys or pancreas. (Sargent and Greenfield.) Lindau, in 1926, showed that retinal angioblastomata may be associated with similar formations of a cystic nature in the cerebellum, spinal cord, pancreas and kidney. Treacher Collins emphasised the importance of recognising that these angioblastomata are neoplasms of mesoblastic tissue, growing in a neuro-epiblastic structure, and that in this respect they differ from the so-called gliomatous growths of the retina, in which also there is a new formation of blood-vessels, but which are neoplasms of neuro-epiblastic tissue with mesoblastic vascular accessories. In eyes affected by such angioblastomatous growths hæmorrhages occur, sometimes associated with nose bleeding, detachment of the retina, iritis and secondary cataract. The detachment of the retina is due to cystic formation in the angiomatous growth, and is similar to the cystic formation in angiomatous growths in the cerebellum, Cushing and Bailey favour the view that the cysts are due to transudation of lymph from the vessels of the growths, rather than mere replacement of fluid from an area of degeneration. The white plaques observed on the retina by the ophthalmoscope are due to fibrous tissue formation in sub-retinal blood clots, and are met not only in connection with retinal angioblastomas but also in connection with vascular disease, associated with hæmorrhages from the vessels. (Treacher Collins.) When the retina is the seat of an angiomatous growth, blindness always ensues, the eye becoming glaucomatous irido-cyclitic or staphylomatous.

Both van der Hoeve and Leslie Paton have observed by means of the ophthalmoscope the changes taking place from time to time in the retina ; the former in tuberose sclerosis, and the latter in an angiomatous condition associated with von Hippel-Lindau's disease (see Plate XVII).

In the differential diagnosis between retinal angiomatosis and congenital arteriovenous communication Kravitz and Lloyd point out that in the latter no " bee-nests " (telangiectases) could be found on the retina, also it is most uncommon for vision to be preserved in the former. They note that implication of the brain is indicated

PLATE XVII



THE RETINA IN V. HIPPEL-LINDAU'S DISEASE

The fundus of the left eye of a girl aged sixteen years who had been under the care of the late Dr. W. J. Adie. There was advanced retinal angiomatosis in the right eye, while now for fourteen months Paton and Williamson-Noble had observed the progress of the disease in the left eye. This girl suffered from severe headaches, vomiting and visual phenomena like teichopsia. Greenfield's pathological examination of the tumour in the right eye led him to the conclusion that although it was at first difficult to be sure whether the tumour was essentially neuroglial or angiomatous and as no neuroglial tissue was found in the central angioma it might be safely assumed that this was a true capillary or plexiform angioma. The central tumour resembled very closely the tumours in the cerebellum which have been described by Lindau, Cushing and Bailey. (By kind permission of L. Paton, F. A. Williamson-Noble and J. G. Greenfield.)

[*To face p.* 364.

by field changes such as homonymous hemianopia, also an arteriogram might show the twisted and tortuous vessels running far back into the occipital region on the involved side.

Von Recklinghausen's Disease

Synonym : Neurofibromatosis, fibroma molluscum

This is yet another type of the multiple tumour syndrome, and is a disease of congenital origin, characterised by cutaneous pigmentation and multiple tumours of the cranial and peripheral nerves, including the sympathetic system. We have seen that in tuberose sclerosis it is the cerebrum which is principally affected, and in Hippel-Lindau's disease the cerebellum. In all three diseases nævoid growths are present, although true nævus cells are not found in the growths.

Penfield and Young, in describing the involvement of the nerve trunks, meninges and central nervous system in von Recklinghausen's disease, state that throughout the three tissues in which neoplasms appear there is to be found definite hyperplastic reaction of the cells peculiar to those tissues, indicating that in this disease an irritant or stimulating influence is exerted on the tissues, causing hyperplasia in them which may be followed by true neoplastic growth of these cells.

The growths in von Recklinghausen's disease are not true neuromas, which must consist of both nerve tissue and nerve cells but are false neuromas, termed " neurofibromatosis " by von Recklinghausen, who believed that the tumours were chiefly fibrous and arose from the connective tissue sheath of the nerves.

Tumours on the cranial nerves in this disease are found chiefly on the auditory nerves, but are also found on the optic and trigeminal nerves. Bilateral acoustic nerve tumours are usually associated with von Recklinghausen's disease. In a case reported by van der Hoeve he found in one eye swellings at the optic disc and a detached retina, in the other eye choked disc and two small retinal tumours. Several years later this man died, and an autopsy was performed, when, on microscopic examination these retinal tumours were found to be composed of the same type of fibrous cells as are found in tumours similarly situated in the disease tuberose sclerosis and von Hippel-Lindau's disease. They are not exactly the same, but are closely allied. It is agreed that in all the phako-

matoses tumours both of mesenchymal and ectodermal origin may be found in the same case. Tumours of the optic nerve are found in association with von Recklinghausen's disease ; for example, fibromata, sarcomata, myxomata and gliomata. Sometimes the tumour spreads along the optic nerve and becomes intracranial (Hudson), making the diagnosis from pituitary tumour very difficult. But the rapid loss of vision, even before there are fundus changes, and later exophthalmos, help to differentiate one from the other. In one case removal of the bone forming the roof of the optic canal has stayed the loss of vision from pressure for over a period of nine years (van der Hoeve). Just as the tumours of von Recklinghausen's disease become more marked at puberty (although they may be present in childhood), so blindness as evidence of pressure on the optic nerve by a tumour in this disease may occur before twenty years of age. Cutaneous pigmentation may be present at birth.

A new growth of the optic nerve is situated within the cone of the extraocular muscles, so that although there may be proptosis, yet the movement of the eye is not impeded until quite late. The tumour does not invade the eyeball to any extent, but by pressing on the globe behind causes a slowly increasing amount of hypermetropia (see Figs. 85 and 86). Radiography of the optic canal may reveal great enlargement, and so help in the diagnosis.

Sometimes an entire nerve trunk and all its branches may become invaded in von Recklinghausen's disease and so produce a condition called *plexiform neuroma*, seen most commonly on the temporal side of the head, the upper eyelid (see Fig. 124 and 126), and back of neck. In some cases when the lids have been affected an enlarged eyeball has been found—buphthalmos.

Treacher Collins and Rayner Batten have drawn attention to the intraocular neurofibromatosis which was the cause of such buphthalmos. Hypertrophied nerve-endings in the choroid, thickening of the peri- and endoneurium of the ciliary nerves where they lie in the uveal tract, sclera and cornea were responsible for the condition of buphthalmos. When, therefore, we see a case of buphthalmos, the patient should be examined for cutaneous pigmentation, moles and nodules (mollusca fibrosa), or other manifestations of von Recklinghausen's disease.

Parkes Weber, having collected eight cases in which buphthalmos was associated with cutaneous and cerebral (not cerebellar) hæman-

giomatosis, suggested that while retinal hæmangiomatosis was most likely to be associated with disease below the tentorium cerebelli (Lindau's disease) buphthalmos was generally associated with supra-tentorial hæmangiomatosis.

When in von Recklinghausen's disease optic atrophy is found, it may be the direct result of pressure on the optic nerve by a tumour,

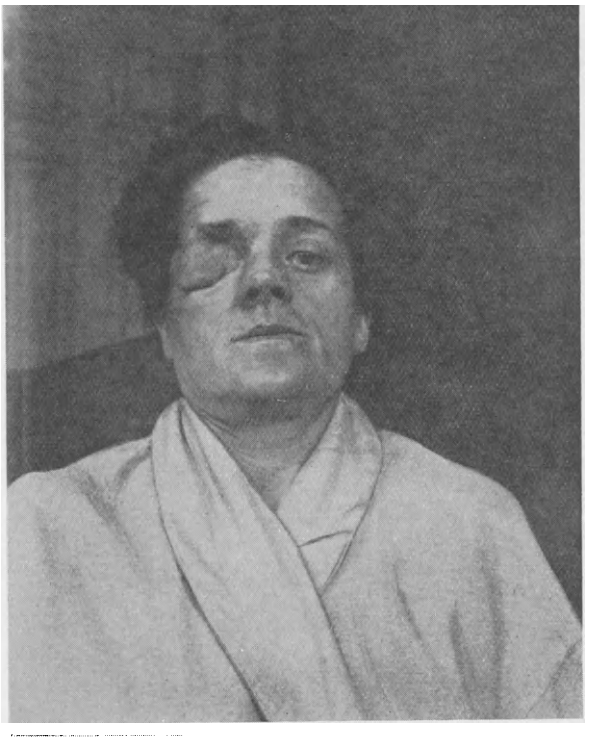


FIG. 124.—Plexiform neuroma of the eyelid in a case of von Recklinghausen's disease. (Case under the care of R. Batten).

or may be consecutive to a papillœdema resulting from raised intracranial pressure produced by an intracranial growth.

The association of tumours of the von Recklinghausen type with intracranial tumours should not be forgotten. Fig. 92, kindly supplied by Jefferson, illustrates in a case of von Recklinghausen's disease, the growth of a fibroma enlarging the right optic canal. The tumour was removed by opening up the roof of the orbit and canal. The patient, a boy, is alive and well now three years after the

operation. One of Martin and Cushing's cases of tumour of the chiasm was associated with von Recklinghausen's disease. Again, Shapland and Greenfield describe a case of neurofibromatosis with meningeal tumours. There were tumours (neurofibromas) in the skin of the lumbar region, there were tumours (psammomata) attached to the cranial dura mater as well as to the meninges of the

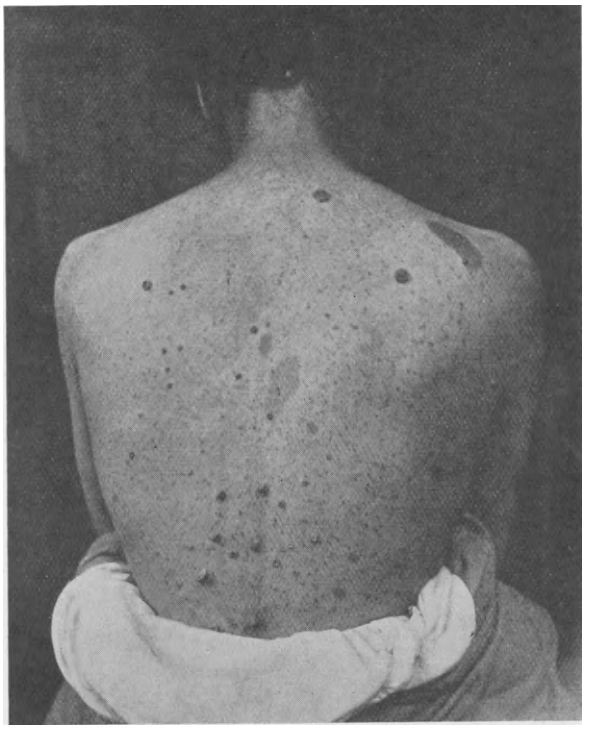


FIG. 125.—Moles and nodules on the back of the patient shown in Fig. 124.

cord. Tumours were found on the right oculo-motor nerve, on the left trigeminal, on the right auditory and on the spinal part of the spinal accessory.

Rayner Batten has provided me with the illustrations of a case which came under his care. The patient, a middle-aged woman (Fig. 124), began to suffer from proptosis of the right eye while the upper eyelid became greatly thickened. Finally, the eye had to be removed, revealing within the orbit as well as in the eyelid masses

of thickened tissue. Fig. 125 is the reproduction of a photograph of the patient's back. It shows numerous moles and nodules. These nodules are small tumours on the superficial nerves and are rarely painful. The bluish spots seen on the skin prove at times to be neurofibromas. The lesions in this disease are cutaneous,

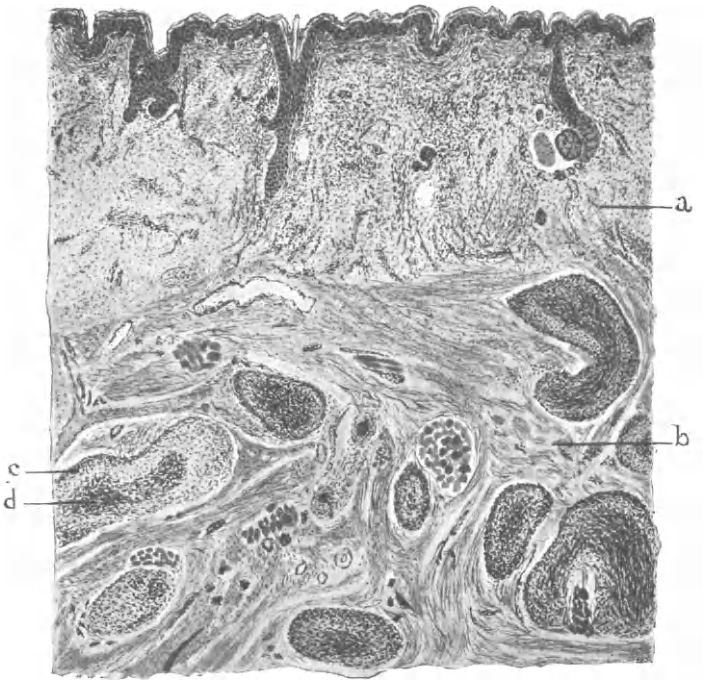


FIG. 126.—The microscopical appearances of a section through the skin of the eyelid in a case of neurofibromatosis. (a) Thickened chorium; (b) subcutaneous tissue with thickened nerves in it cut in various directions; (c) the thickened perineurium of one of the nerves; (*d*) nerve-fibres in the centre of the mass of thickened fibrous tissue. (From Snell's paper on Plexiform Neuroma.)

nervous and bony. (Weber.) The bony changes are similar to those of osteomalacia. The condition of plexiform neuroma has been described. It is from Snell's paper that Fig. 126 has been taken. The condition is similar to that in Batten's case (Fig. 124). It is worth remembering Weber's dictum that "a chronic non-inflammatory swelling on one half of the tongue, or a loose fold of superabundant skin and subcutaneous tissue over one eye or at the side of the head, have a significance in favour of neuro-fibromatosis."

Let it be understood that every case of buphthalmos is not associated with von Recklinghausen's disease, but in every case of this form of enlarged eye in a child search should be made over the body for the presence of moles. The case of proptosis of the eye described by MacMillan and Cone was associated with fifteen pigment marks on the body.

The optic nerve is infrequently attacked in this disease, but such has been described by van der Hœve, while a psammoma in the sheath of the left optic nerve was present in the case of Greenfield's cited above.

In classifying the encapsuled tumours of the peripheral nerves Penfield recognises two types : the perineural fibroblastoma and neurofibroma. The latter, rather than being true neoplasms, are but one manifestation of a systemic disease, yet while in MacMillan and Cone's case the neurofibroma of the orbit was associated with moles that of Motto's case was not. The derivation of neurofibromata is still in doubt ; both von Recklinghausen and Penfield consider their origin to be the endo- and peri-neurium, a mesoblastic connective tissue, while Masson holds that they spring from the cells of the sheath of Schwann—a derivation of the neural crest.

A summary of Farberow's article on Neurofibromatosis Recklinghausen includes :—

(1) X-ray examination of the skull is indicated in cases where there are swellings on the head.

(2) The most common deformations of the skull are in the region of sight.

(3) An enlargement of the sella turcica is most commonly met with in cases of neurofibromatosis Recklinghausen when tumours are present in the orbit.

(4) The enlargement of the optic canals and formation of defects in the orbital walls are uncommon in cases of tumours in the neighbourhood of the orbit.

(5) The roentgenological findings offer no grounds for the assumption that the hypophysis plays a part in the development of neurofibromatosis Recklinghausen.

A syndrome has come to be recognised and known as Sturge-Weber's disease. In this condition we may find a nævoid or angiomatous condition of the cheek, eye, and part of the brain. There may be present buphthalmos, epilepsy and hemiplegia.

Phakomatoses (after van der Hoeve)

	Bourneville (Tuberose sclerosis).	Von Hippel-Lindau.	Recklinghausen.
Nervous system	*Brain.*	*Cerebellum, medulla, spinal cord.*	*Brain.*
	Tumefaction in the cortex.		Heterotopical spots.
	Proliferation of the glia.	Angiomata.	*Cranial nerves, peripheral nerves, sympathetic nerve.*
		Cysts.	
	Tumours in the ventricles.	Proliferation of the glia.	
	Heterotopical spots in the white substance.	*Brain.*	Tumours.
			Ganglioneuromata.
	Cysts.	Sometimes the same.	Neurofibromata.
		Syringomyelia.	
	Medulla, cerebellum, spinal cord.		
	Sometimes tumefactions.		
Eyes	*Optic disc and retina.*	*Optic disc.*	*Exophthalmos pulsans :*
		Angiomata.	*Optic disc.*
	Cysts.		Degeneration and glious proliferation.
	Tumours.	*Retina.*	
	Neurofibrillomata.		Cysts.
	Neurocytomata.	Angiomata.	
		Gliosis.	*Retina.*
		Cysts.	
		Degeneration.	Cysts.
			Tumours.
			Neurofibrillomata.
			Neurocytomata.
			Degeneration with angiomatous parts.
			Optic nerve :
			Tumours.
			Atrophy.
			Iris :
			Tumours.
Other organs	*Heart :*		*Thorax :*
	Rhabdomyomata.		Intrathoracic tumours.
	Kidneys :	*Kidneys :*	
	Cysts.	Cysts.	
	Tumours.	Hypernephromata.	
	Leiomyomata.		
	Angiomata.	*Pancreas :*	
	Hypernephromata.		
	Fibromata.	Cysts.	
	Lipomata.	Tumefactions.	
	Uterus :	*Ovary :*	*Abdomen :*
	Fibromata.	Cystadenomata.	Intra-abdominal tumours.
	Leiomyomata.		
	Intestinal :		
	Lipomata.		
	Thyroid gland, Mammæ :	*Suprarenal glands :*	*Endocrine glands :*
	Adenomata.	Tumours.	Various affections.
	Skin :	*Skin :*	*Skin :*
	Nævi, etc.	Angiomata.	Nævi.
	Moles.	Moles.	Moles.
	Tumours.		Tumours.
	Adenomata.		
	Fibromata.		
	Angiomata.		
	Lipomata.		
	Bones :	*Bones :*	*Bones :*
	Defects in the skull : anomalies in the vessels of the diploe of the skull.	Anomalies in the vessels of the diploe of the skull.	Defects in the skull.
			Affections and deviations of the spinal column.
			Cysts.
			Tumours.
Congenital anomalies	Spina bifida, ectopia testis, horseshoe-shaped kidney, etc.	Anastomosis arterio-venosum.	Spina bifida, syndactyly, one kidney, etc.

If the skin supplied by the first division of the trigeminal is affected, then we may find an angiomatous condition of the occipital lobes. If the second or third division is also involved, then an angiomatous growth of the temporal lobes also may be found.

Spasmus Nutans. *Synonym :* Head-nodding of Infants

This is a disease of infancy occurring entirely in the first two years of life. It is characterised by rhythmic nodding, lateral or rotary movements of the head, or any combination of these. The head movements are usually preceded by nystagmus. The eyes rhythmically converge and diverge alternately, or the nystagmus may be dissociated ; that is, the movements in one eye may be in the horizontal plane, but up and down, or rotary, in the other. The nystagmus may be limited to one eye, which may be the only manifestation of the disease (Still). There is no other disease of the eyes or nervous system present, and the prognosis is always favourable.

Donald Paterson and Ellis, writing in *The Lancet* of October 3rd, 1931, state that such a condition as spasmus nutans is often associated with rickets ; it is rare in children of the well-housed classes, and the onset of the disease frequently occurs in the darkest months of the year. They associate such a condition with defective lighting, and with the nursing of these children in basements or top rooms of houses for a considerable period before the onset of this condition. Removal from present surroundings, together with anti-rachitic treatment, seems to bring about a cure.

Gerlier's Disease. *Synonyms :* Paralysing vertigo ; Kubisagari

This disease was first described by Gerlier in 1887. It was seen in Switzerland among farm hands who worked in stables, chiefly at harvest time. It has also been found in Japan. It manifests itself by the onset of vertigo, general paresis, pain in the neck, diplopia, ptosis, dimness of vision and a narrowing of the visual field. The ptosis, which is unequal, is usually the first to appear in the disease and the last to disappear. The dimness of vision may be due to fine retinal hæmorrhages (Rehsteiner).

A somewhat similar disease in fowls has been described by Galloway. It is a paralysis of fowls (neuro-lymphomatosis), a specific condition recorded in Germany, America and Holland. It has been found also in England, and is of particular interest owing to the intense infiltrative lesions in the peripheral nerves. It is

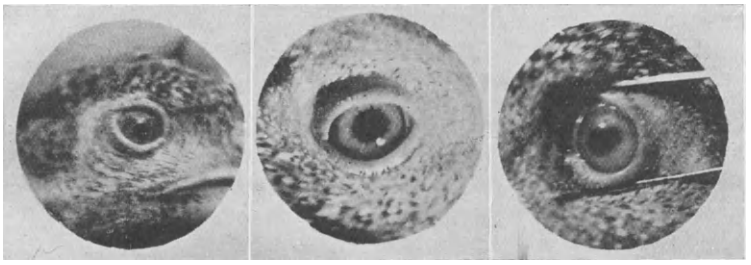


FIG. 127.—Pupillary changes in fowl suffering from neuro-lymphomatosis.

most probably an infectious disease. Galloway succeeded in transmitting the disease to fresh susceptible fowls by intra-cerebral inoculation of the emulsions of the nervous system of diseased birds. Fig. 127 illustrates the change in the pupil found in birds : (*a*) is the pupil of a normal bird, whereas (*b*) and (*c*) show the dilated and uneven pupillary aperture of these paralysed birds. I have to thank Mr. Galloway for the photographs from which this plate was made.

Chronic Progressive Ophthalmoplegia

Synonym : Graefe's disease.

In this rare congenital disease there is chronic progressive nuclear degeneration in the floor of the third ventricle and aqueduct of Sylvius, resulting in diplopia following ocular paralysis. There is ptosis, and later the muscles of both eyes gradually become paralysed. In the end there may be both internal and external ophthalmoplegia. After some years the disease may bring a fatal termination.

In 1868 v. Graefe demonstrated a patient showing a clinical picture not previously described in literature, *i.e.*, all the muscles moving the eye gradually become paralysed with narrowed field of vision and occasional strabismus. The diplopia is slight and sometimes hardly noticed by the patient in spite of the paralysis of the extraocular muscles.

Hereditary Ophthalmoplegia Externa (Treacher Collins)

This is also a chronic affection with gradual onset, occurring in infancy or adult life, and in which one after another of the external muscles of the eyes, including the levator palpebræ superioris, become paralysed. The internal muscles always escape. This may

be nuclear in origin or due to an actual abiotrophy of the muscles themselves. Wilbrand and Saenger collected thirty-two cases of this form of ophthalmoplegia. They described the disease as a bilateral, slowly progressive paralysis of the external ocular muscles without any other signs of disease. Langdon and Cadwalader have described a case including the necropsy report. They found decided changes in the character of the cells constituting the oculo-motor nucleus, together with those of the fourth and sixth nerve nuclei.

Irido-Cyclitis-Parotitis-Polyneuritis

Synonym : Uveo-parotid tuberculosis

As early as 1909 Heerfordt described a clinical syndrome—febris uveo-parotidea subchronica—which was characterised by inflammation of the uveal tract, enlargement of the parotid gland, and in some cases paralysis of cranial nerves, usually the facial. The case described by Feiling and Viner showed severe double irido-cyclitis with cycloplegia (paralysis of accommodation), bilateral parotitis, bilateral facial palsy, absence of knee and ankle jerks, paræsthesia, and an erythematous rash resembling small patches of erythema nodosum on the anterior aspects of the legs and lower parts of the thighs. Garland and Thomson collected and published a review of forty-seven cases. In eighteen of their cases the ocular manifestations showed the development of posterior synechiæ with permanent pupillary changes, such as irregularity of outline and inequality. Iris nodules were noted in one-third of their cases, and in some of these a definite diagnosis of tuberculosis was made. Vitreous opacities also occurred. They put forward strong evidence that the etiological factor in the syndrome of uveitis and parotitis was tuberculosis.

In one of three cases described by Tanner and McCurry a biopsy of the parotid gland gave evidence of tuberculous change, and tubercle bacilli were recovered from the patient's sputum.

The parotitis differed from mumps in that the swollen glands were harder and less obvious ; it was only the pre-auricular portion of the gland which was affected. In Mikulicz's disease, in addition to enlarged parotid glands the sub-maxillary and lacrimal gland may also show swelling, the latter being usually first affected, but neither the eye nor cranial nerves are involved.

The neurological aspect of uveo-parotid fever includes, according to Levin, transient seventh-nerve paralysis, bilateral nerve deafness,

numbness of the face, polydipsia and polyuria. Lesions occur both in the brain stem and cerebrospinal nerves.

Uveitis with associated Alopecia, Poliosis, Vitiligo and Deafness

A uveal syndrome has been described in recent literature, the bibliography of which is given by W. S. Davies.

The uveitis, which is always bilateral, is so severe as to resemble sympathetic ophthalmia. It occurs at the average age of twenty-one years. The prodromal symptoms usually consist of headaches, drowsiness, nausea and a feeling of heaviness about the head. There is circumcorneal injection with cloudiness of the cornea, œdema of iris with keratitic precipitates, together with the formation of posterior synechiæ.

About one or two months after the onset of the uveitis the hair of the scalp begins to fall out, while the eyelashes, eyebrows and hair of the head begin to whiten. This alopecia and poliosis occur in all cases.

There is difficulty in hearing (dysacousia) associated with tinnitus. The auditory affection is generally due to a transient middle-ear catarrh. Vitiligo was noted in 54 per cent. of the cases, occurring soon after the onset of the alopecia and poliosis. These whitish areas of skin denuded of pigment occur on the eyelids, shoulders and backs of hands. Both Wasserman and tuberculous reactions are usually negative. The theory has been put forward by Elschnig that the alopecia and poliosis in association with sympathetic ophthalmia result from the antigenic effect of the uveal pigment, the hair being involved by the marked amount of pigment present. The etiology of this disease is really unknown and treatment can only be directed to relieving the uveitis.

SPINAL CORD

That portion of the spinal cord which has a common interest both for the ophthalmologist and neurologist is the lower cervical and upper dorsal segments. Here each segment of the spinal cord measures 12 or 13 millimetres in depth, and the root bundles of successive nerves are only 1 to 2 millimetres apart. The first thoracic nerve is opposite the sixth or seventh cervical spine. The cervical enlargement of the spinal cord related to the large nerves given off to the upper limb has its greatest width opposite the fifth or sixth cervical vertebra, where it measures 14 millimetres

from side to side (Symington). The arterial supply of this portion of the cord is similar to any other part of it in so far that the anterior spinal artery which descends in front of the medulla spinalis, and lying in the antero-median fissure, is reinforced by branches from the vertebrals, and at this particular level by branches from the ascending cervical artery which is a branch of the inferior thyroid artery. Also spinal branches are given off from the cervicalis profunda, which is a branch of the superior intercostal artery. These pass through the intervertebral foramina. The posterior spinal artery, derived from the vertebral artery, gives branches which anastomose around the posterior roots of the spinal nerve and communicate with the vessels of the opposite side. It has already been mentioned that the dilator nerve fibres of the pupil pass downwards in the lateral part of the pons, medulla and cervical cord, as far as the first and second dorsal segments. The preganglionic fibres take origin from the cells in the lateral horns and leave the cord by the anterior nerve roots, passing by the white rami communicantes of the sympathetic to the cervical sympathetic nerve trunk to end in the superior cervical ganglion. The further course of these nerves is described in Chapter II.

Injuries to the Spinal Cord

During the war the author saw many cases of spinal injury due chiefly to penetrating foreign bodies such as pieces of shell, bullets, etc., also pieces of smashed bone. Many of these were in the thoracic region. The immediate effects were extreme shock and pain; indeed, of all the war wounds one saw these appeared to cause the most severe pain and suffering. There was instantaneous paralysis of all that portion of the body below the wound, and many were rapidly fatal. In civil life, however, fractures and fracture dislocations are the commonest causes of spinal cord injury. These cases are seen where men are employed in shipbuilding yards, also in motor-cycle accidents, diving accidents and falls from horses. They are also caused by motor car and train accidents. In any of these conditions the cervical cord may be affected.

Thrombosis

Thrombosis occurring in arterio-sclerosis may affect a portion of the spinal cord, leading to softening of a definite portion.

Hæmatomyelia

The condition known as *hæmatomyelia* is a sudden hæmorrhage into the grey matter of the cord. It occurs most commonly in the cervical region, causing damage to cells and fibres especially in the region of the central canal, simulating an acute syringomyelia or hemisection of the cord. It occurs at any period of adult life, and its cause is generally trauma. There is a sudden onset with pain in the neck and upper limbs, with paralysis of the upper limbs. Involvement of the phrenic nerve or the presence of Horner's syndrome will indicate that the cervical part of the cord is involved.

The sequel to railway and motor accidents found in many people is a condition known as *spinal traumatic neurasthenia*. These patients often find difficulty in reading, due to a weakness of the accommodation. If an hysterical element is added the fields of vision will become contracted also.

In all of these conditions the question is how much involvement of the spinal cord has taken place ? An injury which affects the lower cervical and upper dorsal segments may show Horner's syndrome, that is, miosis, ptosis and enophthalmos, due to the involvement of the ocular sympathetic. A fine penetrating body such as a rifle-bullet or stab from a knife may also cause a Brown-Séquard syndrome—loss of pain and temperature sensation on the opposite side to and below the level of the wound. Touch is scarcely affected. On the same side there is loss of appreciation of posture and movement of joints, vibration and tactile discrimination. There are also paresis and pyramidal tract signs below the lesion on the same side.

Lesions high up in the cervical region may show papillœdema, nystagmus, pupillary changes, together with dyspnœa and rise of temperature.

Compression of the spinal cord by disease of the vertebral column, such as tuberculosis, syphilitic and malignant disease, may affect the conduction of the cord. This may be caused also by intramedullary and extramedullary spinal tumours, abscess, pachymeningitis, cysts and Hodgkin's disease.

Nystagmus may occasionally be seen in tumours of the spinal cord.

The Klumpke Syndrome

The Klumpke syndrome originally described by Mme. Déjerine-

Klumpke is the result of traumatic avulsion (at birth or under other circumstances) of the last cervical or first dorsal roots. There is a motor paralysis of the hand and part of the forearm. On the corresponding side is found Horner's syndrome, pallor of face and neck, unusual dryness of the nostril and mouth and diminished sweating over the face, neck, arm, and chest.

Facial hemiatrophy has been observed occasionally with Klumpke's syndrome.

Acute Anterior Poliomyelitis

This is an acute infectious disease of the central nervous system. The lesions are mainly found in the anterior horns of the grey matter of the spinal cord, but sometimes extend upwards as high as the basal ganglion and grey matter of the cerebral cortex (polio-encephalitis). The organism believed to be responsible for this infection is a minute " globoid body " recoverable from the brain and spinal cord tissues, also from the naso-pharynx (Flexner and Noguchi). The cervical and thoracico-lumbar regions are the most affected.

When the cervical portion of the cord is implicated there may be paralysis of the ocular sympathetic. If the lesion is higher, say in the brain stem, then ocular paralyses may occur, but these are rare. The paralysis of the sixth cranial nerve seen so commonly in young children is often attributed to a previous attack of polio-encephalitis, but the author's experience does not agree with this. The great majority of these cases have no history whatever of any infection since birth, nor have suffered any form of illness. A slight intracranial injury at birth is the most probable explanation. (See p. 50.)

Nystagmus may result from an attack of acute anterior poliomyelitis.

Acute Ascending (Landry's) Paralysis

This is a rapidly ascending motor paralysis beginning as a flaccid paralysis of the legs followed by paralysis of the trunk and upper limbs. Involvement of the medulla and pons leads to death. The duration of the disease is from a few days to several weeks

It is stated that there is no one pathogenic factor, either bacterial or toxic, that can be regarded as a specific cause of Landry's paralysis.

Ocular symptoms are rare in this disease, but Swanzy says that there may be paralysis of some of the orbital muscles, paralysis of accommodation, mydriasis or loss of light reflex.

Subacute Combined Degeneration of the Spinal Cord

Combined degeneration of the spinal cord is a subacute progressive disease in which the white matter of the spinal cord shows degeneration. The posterior and lateral columns are chiefly affected. This disease is often associated with the severe anæmias, and is observed in individuals of middle age. Retinal hæmorrhages due to the anæmia may be present, also bilateral primary optic atrophy may occur (5 %). As optic atrophy may occur in this disease, it is important to distinguish the condition from tabes dorsalis. In the former, pupillary changes and oculo-motor paralyses are not found, but nystagmus is commonly present ; while in the latter, pupillary changes and ocular paralyses are pathognomonic of the disease, and nystagmus is usually absent. McAlpine has pointed out the relationship of combined sclerosis with pernicious anæmia, together with the appearance of mental changes, neither of which is usually associated with tabes.

Hypertrophic Cervical Meningitis

This rare disease occurs most frequently in the cervical region, and is caused chiefly by syphilis and tuberculosis. Pain is a prominent symptom, radiating upwards and downwards from the neck even to the hands. Horner's syndrome may be present.

Friedreich's Ataxia

Among the hereditary ataxias may be mentioned Friedreich's ataxia, a familial and probably heredo-degenerative disease in which degeneration takes place in the ganglion cells of Clarke's column, the lateral and posterior columns, and spinal and cerebellar tracts.

Nystagmus is common in Friedreich's ataxia ; the movements are more pronounced upon directing the gaze laterally, while optic atrophy is only occasionally seen.

In some of the hereditary ataxias the affection of the nerves going to Deiters' nucleus is indicated by the frequent presence of nystagmus. (Wilbrand and Saenger.)

The eye symptoms associated with Friedreich's ataxia have been

shown in table form by Barrett, the perusal of which reveals that of 38 cases 24 had nystagmus, 2 optic atrophy, 4 ptosis, 3 what were described as Argyll Robertson pupils and one case of ophthalmoplegia. Nystagmus, optic atrophy and ophthalmoplegia were present in one of Barrett's cases. He concludes by stating that the most constant and frequent symptoms of Friedreich's ataxia are ataxia of both lower and upper extremities, speech defect, scoliosis, foot deformities, absent or weak knee and ankle reflexes and nystagmus.

The resemblance between Friedreich's ataxia and tabes dorsalis is suggested, not merely by the pathological changes in the spinal cord in both diseases but also by the presence of clinical symptoms that are common to both.

Marie's Ataxia

In hereditary cerebellar ataxia optic atrophy is more common than in Friedreich's ataxia. There may be loss of pupillary reflexes simulating Argyll Robertson pupils, also there may be changes in the visual fields and diminution of central vision.

Sanger-Brown's Ataxia

This form of ataxia, also hereditary in character and affecting the cells of Clarke's column, the posterior columns and direct spinocerebellar tracts, more commonly exhibits ocular changes than does either of the two hereditary diseases just mentioned. Optic atrophy, ptosis, strabismus producing diplopia, or complete internal and external ophthalmoplegia are found. In these hereditary diseases retinitis pigmentosa is sometimes observed. In hereditary spastic paraplegia optic atrophy and retinitis pigmentosa have been described.

Myelitis

Inflammation of the spinal cord may result in a transverse or ascending myelitis at any level of the cord. Syphilis, tuberculosis and pyogenic organisms from a wound are among the common causes of myelitis. A previous history of retrobulbar neuritis or nystagmus would probably point to disseminated sclerosis as a cause of myelitis. If the cervical portion of the cord is affected there will be paralysis and anæsthesia below the level of the lesion, with the possible addition of inequality of the pupils due to interference with the cervical sympathetic nerve, or Horner's syndrome

may be present. It must be remembered that a virus may affect both the optic nerves and the spinal cord simultaneously ; but in neuro-myelitis optica the optic nerves may be affected before signs or symptoms of the myelitis appear.

Amyotrophic Lateral Sclerosis

Amyotrophic lateral sclerosis or progressive muscular atrophy is a disease affecting the anterior horns of the cord, pyramidal tracts and motor nuclei of the medulla. There is wasting of the muscles of the upper limbs, first seen in the muscles of the hands, especially those of the thenar eminence. Muscles supplied by nerves taking origin in the medulla may be affected at the same time, so that there may be wasting of the orbicularis palpebrarum and other facial muscles. Rarely are the ocular nuclei in the bulb affected, and Horner's syndrome is seldom seen.

Syringomyelia

Syringomyelia is a chronic and progressive disease of the spinal cord. Long cavities form in the substance of the cord, which may or may not communicate with the central canal ; the latter itself is rarely affected. The syrinx usually involves first the cervical enlargement and then the upper dorsal regions of the cord, so that ocular symptoms are common. These may be inequality of the pupils, ptosis and enophthalmos. Optic atrophy and papillœdema have been described as rare signs. Schlesinger suggests that the papillœdema when present is due to internal hydrocephalus. He also pointed out that concentric contraction in the fields of vision, especially for colours, was found in 38 out of 130 cases. The fields of vision were taken by Jameson Evans of a case of syringomyelia under the care of Sydney Short, which was described in the *B.M.J.* some time ago. The chief feature was the great contraction in the field of vision to the green rays. Due to involvement of the medulla and pons (syringobulbia), there may be paralysis of the fourth, fifth and seventh cranial nerves, the fifth causing pain and later anæsthesia, including that of the cornea. Paralysis of the seventh cranial nerve will cause loss of power of the orbicularis resulting in incomplete closure of the eyelids (lagophthalmus). Rotary nystagmus is common in this disease, due probably to vestibular disturbance.

Five cases of syringomyelia complicated by the presence of

choked disc (papillœdema) have been described. The result of an autopsy in such a case has been recorded by Alpers and Comroe, who state the choked disc was due to internal hydrocephalus secondary to a closure of the foramina in the roof of the fourth ventricle.

At the Medical Society of London, in March, 1935, Professor Kelly of Liverpool spoke of ten cases which had undergone operation by him. Laminectomy with drainage of the fluid cavity had been done. He could not speak optimistically of his results. It is well to remember that wounds in cases of syringomyelia heal badly.

The Amyotrophies

Among the disorders of muscles in which ophthalmic symptoms are found may be mentioned myotonia congenita, or Thomsen's disease, myotonia atrophica, myasthenia gravis, facio-scapulo-humeral dystrophy of Landouzy-Déjerine and neuritic muscular atrophy (Charcot-Marie-Tooth).

Myotonia Congenita

Myotonia congenita, a hereditary disease appearing in childhood, is characterised by the occurrence of spasm in voluntary movements so that the primary contraction of a group of muscles is prolonged followed by gradual relaxation. This can be demonstrated by asking the patient to close his eyelids and open them; jerky movements followed by slow relaxation of the orbicularis oculi follow. Swanzy states that amblyopia (failure of sight) of a temporary character has been observed.

Myotonia Atrophica *Synonym:* Dystrophia myotonica

This disease, like the one just described, is familial and sometimes hereditary. Since 1909, Batten and Gibb, Curschman and others, have grouped a number of supposed atypical cases of myotonia congenita and classified them as a separate disease.

Myotonia Atrophica.—Patients affected by this disease usually show fully developed symptoms between the ages of twenty-five and thirty. Greenfield was the first to describe the presence of cataract in association with this disease. In 1911 he described a family of thirteen children, of which five members had dystrophia myotonica; two of these had associated cataract; two other children had cataract alone. A paternal aunt and grandmother had developed cataract at sixty-nine and sixty years of age respectively. The typical signs and symptoms of dystrophia myotonica are: altered

facial expression (resembling the Landouzy-Déjerine myopathy) due to wasting of the facial muscles, including the masseter, temporal and sterno-cleido-mastoid muscles, the muscles of the upper arm and thigh. The myotonia of the facial muscles is responsible for

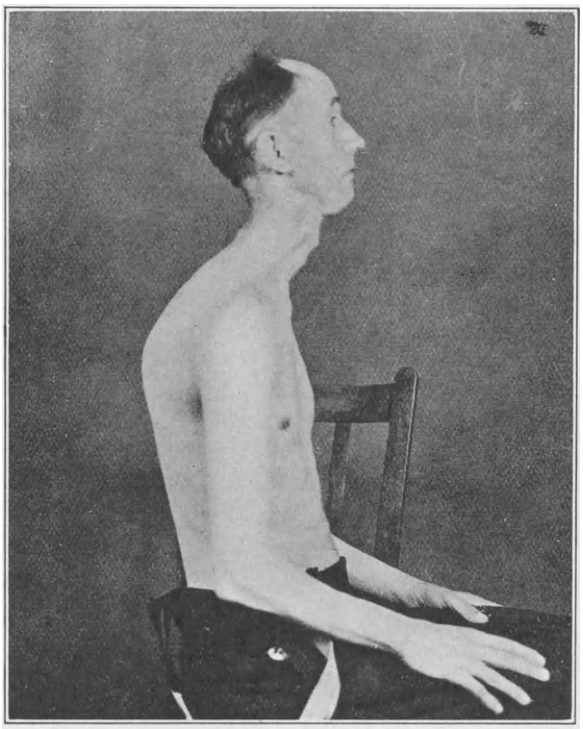


FIG. 128.—Case of dystrophia myotonica fifty years of age. (After Caughey.)

the long-continued smile, difficulty in mastication, etc., due to long relaxation period.

Dystrophic symptoms include atrophy of the sex glands, which in turn produce signs of premature senility, such as frontal baldness and loss of sexual desire. Cataract, which is associated with the disease, or sexual aplasia, may be the only signs of the disease in one generation, while in the succeeding generation the entire family usually show both myotonia and muscular dystrophy (Fig. 128).

Adie states that in a dystrophic generation some members remain healthy, some may have premature cataract alone, some

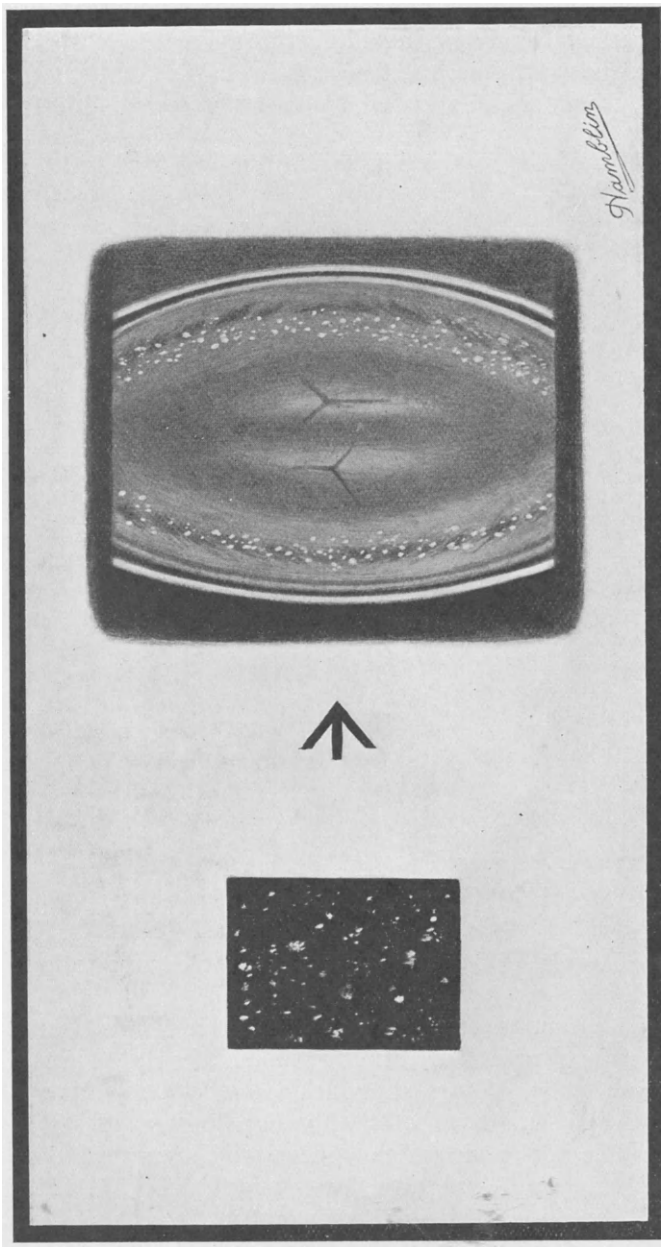


FIG. 129.—Slit-lamp appearance of the lens of the left eye of case shown in Fig. 128. The small illustration on the left is the surface view of the lens. (After Goulden.)

The left illustration shows, according to Goulden, tiny angular opacities crowded together in the superficial parts of both the anterior and posterior zone of disjunction. The space between the capsule and the zone is perfectly clear as is also the deeper part of the cortex and the nucleus of the lens. Scattered amongst these whitish opacities are many iridescent crystal-like bodies, red and green, similar to the ophthalmoscopic view of the bodies in synchysis scintillans. The cataract may set in between twenty and thirty years of age and matures early.

Goulden states that the distribution of lenticular opacities is similar in cases of dystrophia myotonica, meningitis, traumatic myxœdema and post-operative tetany.

may suffer from a muscular disease in an incomplete form, one of the cardinal symptoms such as muscular atrophy being absent; while others present the combination of muscular atrophy with myotonia and extra-muscular symptoms, such as cataract, atrophy of the testicles with impotence, baldness, vasomotor disturbances and general atrophy with loss of weight. Caughey and others have mentioned the influence of decaying social status in relationship to this disease.

The cataract appears much earlier than ordinary senile cataract, and tends to appear earlier in each generation. At first the lens changes may be observed in one eye only, but as a rule both eyes show rapidly ripening cataracts. Fleischer, in 1918, gave an account of thirty-five cases of cataract in families with dystrophic symptoms seen at the Ophthalmic Department of the Tuebingen Hospital. He described the cataract as beginning at the posterior pole, and from the centre lines radiate out to the periphery. Koby, Goulden and Caughey have described the slit-lamp appearance of this particular type of cataract. The anterior and posterior zones of the lens substance just beneath the capsule are clear, then follows a band deep to these areas in which minute whitish angular opacities are present. Among these are iridescent crystal-like bodies, red and green, resembling the ophthalmoscopic appearance of synchysis scintillans (cholesterin crystals floating in the vitreous). (See Fig. 129.) Later posterior and anterior stellate opacities appear as the cataract ripens.

Operation for removal of these cataracts is usually uneventful.

Juler operated on a woman aged twenty-six, whose father, aged fifty, also had an operation for cataract the same year, but only the daughter showed signs of dystrophia myotonica.

The Facio-Scapulo-Humeral Type of Myopathic Atrophy (Landouzy-Déjerine).

This disease, which is hereditary, usually begins in childhood and affects the facial muscles first. The wasting of the orbicularis palpebrarum and the orbicularis oris makes the complete closure of the eyelids impossible, and renders whistling difficult.

Neuritic Muscular Atrophy

Synonym: Charcot-Marie-Tooth's Disease

Although neuritic muscular atrophy is characterised chiefly by wasting of the feet and legs, due to involvement of the peroneal

nerves, yet some years later the forearms may be affected. This is due to involvement of the spinal cord. If the disease affects the lower cervical region there may be inequality of the pupils, due to interference with the cervical sympathetic. Optic atrophy has been described.

Brain states that involvement of the ocular muscles in dystrophy is extremely rare. He has seen one case in a woman aged twenty in whom there was complete bilateral external ophthalmoplegia with normal pupillary actions associated with enlarged and somewhat weak calf muscles and sluggish ankle jerks. Gowers, Willbrand and Saenger have also referred to the extreme rarity of involvement of the extra-ocular muscles in progressive muscular dystrophy (primary myopathy).

Functional Nervous Disease or the Neuroses

A neurosis is an abnormal mental reaction to a situation causing mental stress (Brain). The underlying etiological factor is usually some difficulty in the path of life encountered by the patient. Patients react variously to this difficulty, and therefore produce varying symptoms of nervous disturbance which manifest themselves as (1) neurasthenia, (2) anxiety neurosis, (3) hysteria, (4) obsessional neurosis, (5) occupational, and (6) traumatic neurosis. Although Stoddart and others classify the first three as the psychoneuroses, it is simpler to think of all six simply as the neuroses. Pavlov, who has bestowed upon the world such a wonderful array of physiological experiments, has attempted to reach a physiological interpretation of obsessional neurosis and paranoia. His experiments are, as usual, on dogs. He found it possible to produce a pathological disturbance of the activity of the nerve cells; an alteration of the normal balance between two sides of their activity (the excitatory and inhibitive processes) with an abnormal predominance of the excitatory process which was reducible with bromides. Obsessional neurosis and paranoia, he says, might prove to be pathological states of the corresponding cells of the cerebral cortex—a state of pathological inertness. Pavlov argues that human neuroses might be traceable to irregular development and occasional accentuation of one or other of the emotions, and disease of some organs or of a whole system, which causes the corresponding cortical cells to be temporarily, excessively, or unlimitingly excited. This brings about their "pathological inertness"—an irresistible con-

ception and sensation which continues to last long after its real cause has been withdrawn.

The doctor in his consulting-room is at times forced to make a patient disclose facts which apparently have no bearing on his particular specialty. The ophthalmologist or neurologist does not, or should not, attempt to become a psycho-analyst or psycho-therapeutist. If he does so, he soon loses real interest in his own specific job. I do not mean to say that a doctor should be ignorant of the principles of psychology, quite the opposite. Does it give a doctor mental pleasure to ask a patient sexual questions ? If so, the doctor must utterly and entirely suppress any thought of asking a question the irrelevancy of which to the disease under discussion he is fully aware. But will the unconscious mind some time at a future period bring forth these suppressed thoughts ? Has the doctor acquired a complex which he cannot get rid of ? If he has, its origin goes back a long way, not, I think, to the period of infancy or early childhood, but to those years when the use of his will-power was most in need. Quoting W. R. Bousfield : " Freud has taught us to look at the unconscious mind as a power-house which gives out the strongest determinants of conduct. He has failed to point out that the contents of this power-house, when a person has reached the age at which serious conflicts begin, are largely under his own control." When one has tried to wade through the writings of various thinkers in the realm of psychology, it is refreshing to find men like Bousfield who realise that there has been a " neglected complex " in the generally accepted psychology of the day, which shows, for instance, that love in its highest form—the love " which seeketh not its own," and whose end is giving rather than getting—is an attribute of the psyche and not of the soma. One reads of Jung, when addressing the students of Zurich University, saying : " Love has more than one element in common with religious conviction ; it demands an unconditioned attitude, and it expects complete surrender. Only that believer who yields himself wholly to his god partakes in the manifestation of divine grace. Similarly, love reveals its highest mysteries and wonders only to him who is capable of unconditioned surrender and loyalty of feeling. Because this is so hard, few indeed of mortal men can boast of achieving it. But just because the most devoted and truest love is also the most beautiful, let no man seek that which could make love easy. He is a sorry knight of his lady who recoils from the difficulty of love.

Love is like God—both give themselves only to their bravest knights."

Let us now return to a brief consideration of the neuroses in so far as they concern us.

(1) **Neurasthenia.**—In the condition of neurasthenia there is marked mental and physical fatigability. Both neurasthenia and anxiety neurosis are expressions of excessive reaction to the stresses and strains of life, and of failure to adapt oneself to them. Nowhere can the subject of neurasthenia be more definitely observed than in the ophthalmic department of a hospital for diseases of the nervous system. The statements made by neurasthenic patients are totally unlike those heard in an ordinary ophthalmic hospital. Exaggeration of slight neuritic pains—" not feeling very grand "—they cannot tolerate light, cannot read or, if able to read, do so but for a short time, they quickly become tired when examined by the perimeter, with the result that often a contracted visual field is found. Fatigue is indeed a cardinal symptom of neurasthenia. One must listen sympathetically to such patients, realising that they are suffering from a very definite disorder. Even although the examination of their refraction is carried out most carefully, they will return from time to time complaining of a pain in one eye, or inability to continue using their eyes. But, in contrast to the patient at an eye hospital who returns with his spectacles in his pocket saying he cannot wear them, the neurasthenic will wear them and will not do without them. Nevertheless, any patient complaining of headache should have the fundus of the eye and his refraction examined. The early stages of general paralysis, tabes dorsalis and exophthalmic goitre exhibit a superficial resemblance to neurasthenia. The pupillary light reflex and the fundus should always be examined.

(2) **Anxiety Neurosis.**—No hard and fast line can be drawn between neurasthenia and anxiety neurosis. The symptoms of the latter are a combination of nervous irritability with morbid anxiety, fear and dread. It is the fear associated with the uncertainty that produces the anxiety. Patients suffering from anxiety neurosis are usually hypersensitive, thus they cannot bear bright light or noise. They have constant fear—do not like to open a letter lest it contain bad news. Such patients suffer from dyspnœa, palpitation, vaso-motor disturbances, digestive trouble and phobias. Names have been given to many of these phobias, such as agoraphobia (fear of

open spaces), claustrophobia (fear of being in an enclosed space), coprophobia (fear of evacuating bowels at an awkward moment), acrophobia (an abnormal fear of heights), photophobia (dread of light) and nyctophobia (fear of the dark).

The physical examination of these patients often shows dilated pupils, with slightly diminished central vision.

(3) **Hysteria.**—Undoubtedly the neurosis which gives rise to the gravest symptoms is hysteria. This condition is characterised by abnormal reactions of the personality to mental stress. The functions become dissociated and, in extreme cases, sight, hearing, feeling or speech may be dissociated from the personality. Somnambulism (or sleepwalking) is an almost complete form of dissociation.

In the text-books of neurology and psychiatry the condition of hysteria is there found fully treated. Here it is not necessary to say more than that hysterical patients exhibit multitudinous signs and symptoms suggestive of organic nerve disease, yet on examination decided differences are found ; for example, where the paralytic tries to support himself, the hysterical patient tries to fall ; the epileptic is unconscious during his fits, not so the hysteric, nor is his tongue ever bitten through, as in epilepsy. The author once tried to help a hysterical patient who was suffering from a fit by the time-honoured method of applying cold water, but the patient forestalled him by emptying the jug of water over the author. In the hysterical fugue the patient wanders from home, losing his identity. Such hysterical amnesias and fugues are reactions to difficulties which are making life unbearable. A patient under the author's care had suddenly wandered from home in London, and was found on the outskirts of Liverpool, having walked there in complete oblivion of time or circumstance. Examination showed the presence of cataract in each eye, one being completely ripe. His fear was blindness and the consequent difficulty of supporting wife and family. Removal of one cataract with restoration of vision brought about a cure.

The hystero-epileptic fit seen in Eastern races almost invariably begins with an attack of an epileptic nature ; the terminal stage, following violent movements of the body, is one in which visual hallucinations are experienced.

Women suffer from hysteria more commonly than men, and many cases are seen among the uneducated masses. The mode of production of hysteria is exemplified in a case cited by Stoddart of

the woman who did not wish to go abroad with her husband. Slowly but surely she developed hysterical paraplegia. The paraplegia was a suggestion and at the same time an expression of an idea in the woman's mind, and also it became the means to achieve her purpose. Again, a young girl who detested the manager of her office developed hysterical blindness ; the fact that her love affairs were in order did not prevent the onset of hysteria, and so her blindness prevented her working at the office and probably secured her removal from it. These two cases illustrate what is known as *convertion hysteria*, because the mental compromise is effected through physical manifestations. In *anxiety hysteria* the mental symptoms predominate, and are mainly those of anxiety or fear.

The physical manifestations of *convertion hysteria* are manifold. We are here concerned with the ocular symptoms. Of these, blindness is common. The ophthalmologist notices an extremely lowered visual acuity as revealed by the test types, but normal vision when the patient wishes to see a particular person or thing. The bilateral blindness resulting from a head injury may be perpetuated for a long time, and such patients may have their sight suddenly restored. Bilateral blindness may be present, but if unilateral it may be associated with a hemi-anæsthesia. If fields of vision are recorded they may be found to be reduced, being smaller on the anæsthetic side. Recently the newspapers described the marvellous restoration of vision under the care of my friend, Mr. Arthur McCurry, but, as he pointed out to me, mention was not made of how the patient found his way alone eight miles to hospital. But " X-rays " perform miracles at times.

In all these cases of hysterical blindness the fundus oculi is normal in appearance. If squint is present, it is associated with contractions of the orbicularis palpebrarum, without any counteraction of the occipito-frontalis. During a hysterical fit, if the eyelids are parted the eyeballs are found rolled up. Often a ptosis is seen, and by its means of partially or wholly covering the pupils sight is diminished. The patient cannot read the test types, and if an attempt is made by the oculist to raise the lids above the pupils they are found to be firmly held down—a true hysterical reflex unconscious action. But take her spectacles and ask which she uses for reading and which for distance, and while her mind is so diverted the lids are seen to move upwards, and the patient uses her eyes to a definite purpose. Ptosis may be accompanied by painful vision—

dysopsia algera—or by photophobia and blepharospasm or contraction of the upper and lower eyelids. Macropsia, in which objects appear large, and micropsia, in which they seem small, have already been mentioned.

The pupillary reflexes in these hysterical patients are normal. If diplopia is complained of it may be found to be present when only one eye is in use. Spasm of the extra-ocular muscles associated with spasm of accommodation is sometimes found. In spasm of accommodation objects are out of focus on the retina, and the patient, already in an excited condition, thinks she is going blind. When perimetric examination of such patients is repeated, the field with each examination becomes smaller; a spiral or helicoid field may be thus obtained at one sitting. Dyscromatopsia and acromatopsia, perverse or loss of colour perception, are rarely found. In organic disease the red field is found to be first to show contraction; but in hysteria the opposite may be seen, the red may be the last to disappear.

When there is a sense of strain, or weariness in the eyes and head, brought about by the use of the eyes, the condition is known as asthenopia. Now in the ophthalmological department of a hospital for nervous diseases cases are found where the physician has referred the patient to the oculist with the query: "Does the patient need glasses, or a change of glasses?" The physician has found no evidence of organic disease, and the oculist does not find any error of refraction, or the glasses being worn are correct. This form of asthenopia is not accompanied by red conjunctivæ, nor cephalalgia. The patient just states: "I cannot use my eyes for any length of time." One of the author's patients did all the sewing herself for her friend's trousseau, but for months afterwards she could not keep up the continuous use of her eyes. Another, a lady musician in an orchestra, suddenly saw a flash of light, whether real or imaginary she could not state, but for years she could not earn her living reading music. These are examples of what is known as *hysterical asthenopia*.

Stoddart has observed spasmodic convergent strabismus in quite a number of hysterical patients. Hallucinatory delirium is seen in cases of hysterical insanity or at the conclusion of hystero-epileptic fits. The patient sees animals, has visions of God, or emotional incidents of his past life.

Perhaps one of the most rational statements made regarding hysteria is that by Stoddart, when he says: "The only radical cure

of hysteria is to discuss the unconscious strivings which have given rise to the disease and in so doing to reveal them to the patient."

(6) **Traumatic Neurosis and Shell-Shock.**—Spinal traumatism, as is so commonly seen after railway accidents, is peculiarly liable to produce an attack of hysteria. Another form of hysteria is found when patients have suffered an injury to one eye. Although perfectly able to work again they are afraid to do so, and cannot bring themselves to start work afresh until compensation is paid. At once we are faced with a great difficulty—is the patient malingering or is he genuine ? The system which pays such a person his half-pay and ends by paying compensation, generally after a legal fight, is to blame for producing a real neurosis. The longer compensation is delayed, the worse his vision becomes, and his eyes apparently refuse to become normal.

During the Great War many of the soldiers among the evacuated wounded were found to be suffering from a nervous phenomenon which was attributed to the explosion of shells in their immediate neighbourhood, and other terrifying experiences. The condition soon came to be known as *shell-shock*. It was a true neurosis in which reduction of vision was a common manifestation ; in some cases there was complete blindness for weeks, months or even years. It has been stated that through the process of auto-suggestion the loss of vision was sustained. Sometimes this was accompanied by blinking. The fundi and fields of vision were found to be normal. Ormond, in a paper on " Concussion Injuries of the Visual Apparatus," has stated that " the defects of accommodation have been brought to light as a result of shell-shock, and this has led to marked improvement in the vision of many cases by the use of suitable lenses."

From many years' experience the writer has come to the conclusion that patients suffering from functional or organic disease of the nervous system should have a thorough ophthalmic examination. The correction of errors of refraction is most important. Indeed, a pair of spectacles may have been found to do infinitely more for such a patient than all the bottles of sedative mixtures previously prescribed. Most ophthalmic surgeons are aware that in many cases severe epileptiform fits and melancholic attacks of the mind are banished by the entire relief of ciliary strain and the careful correction of a muscle imbalance which is beyond the normal amount of error.

The Psychoses

The psychoses are the true insanities, and, unlike the neuroses, the patients do not as a rule recognise their suffering and infirmity. Dr. H. C. Beccle reported to me that as the result of examining over 1,000 psychotics at the Springfield Mental Hospital he found remarkably few suffering from ocular symptoms. Other reports, however, show a greater number of ocular manifestations among such patients.

Melancholia.—This is a phase of periodic insanity in which the mental attitude of the patient is one of deepest misery. In keeping with the diminished amount of secretion everywhere, the lacrimal gland also suffers, so that the flow of tears is diminished. The palpebral aperture is widened, due to excessive wrinkling of the occipital frontalis. Stoddart has observed nystagmus on extreme lateral deviation together with diminished power of accommodation. Glasses stronger than the age of the patient warrants have to be prescribed. This loss of accommodation may be regained on the recovery of the patient. Hallucinations do not present themselves to the melancholics.

Paranoia.—A mental disorder in which systematised delusions develop as a result of the patient's projecting his unconscious wishes on to his environment. Subjects passing through the slow insidious development of this disease may show on examination of the eyes nothing abnormal except a certain unsteadiness of gaze, but vision is not affected, and hallucinations do not occur.

Dementia Præcox.—This psychosis, which appears in adolescent and early adult life, is characterised by a variety of mental symptoms, physical signs, and disorders of conduct which appear in predisposed persons, tending towards a characteristic and profound dementia.

During the stages of this disease tremor of the closed eyelids may be seen, and although the pupils are usually dilated they react well to light. Also there are faint mental pictures not so vivid as true hallucinations, and therefore called pseudo-hallucinations. In *dementia paranoides* visual hallucinations and illusions occasionally occur.

Epileptic Insanity.—Most epileptics in asylums are regarded as potential dangers. The conduct of many of these patients is cruel and brutal; they are mischief-makers and tale-bearers, yet at times exhibit periods of religiosity. The repeated occurrence of

fits helps to produce dementia. In what is known as larval epilepsy automatisms occur in which the patient's mind is usually a blank as regards what he has done. During the epileptic delirium there is the presence of terrifying hallucinations, which may even drive the patient to homicide. Stoddart thinks in these cases the peripheral fields of vision are contracted. The pupillary reflexes respond but feebly to light. In the latter stages of dementia hallucinations are not at all frequent. Nystagmus may be observed.

As mentioned in a previous chapter, all peripheral sources of irritation in these patients should be removed, and perhaps not the least important is the presence of ciliary strain in ametropic eyes, which is relieved by the wearing of proper glasses.

Delirium Tremens.—During an attack of this acute disorder the patient's face is flushed and the conjunctiva of the eyes red and suffused, the pupils are at first contracted, but later they dilate. There is also contraction of the visual field. As is well known, visual hallucinations dominate the clinical picture. Rats, devils, enormous spiders and all kinds of strange animals are seen ringed, most probably with some colour, such as blue. This colour is not changed by looking through glass or any other colour. Mental pictures are produced by pressing the closed eyes. One can remember during routine hospital work that, although in the midst of an attack of delirium, a sharp word will bring the patient to his senses and he will talk quite rationally for a short time. Colours are confused, but this is psychical.

CHAPTER XIV

AFFECTIONS OF THE VEGETATIVE NERVOUS SYSTEM

In contradistinction to the central and peripheral nervous systems which serve the senses and activate the muscles controlled by will there is what in the past has been termed the sympathetic or autonomic system which is purely involuntary, the fibres of which pass to organs having smooth muscles, such as the intestines, blood vessels, glands, ducts and skin. Also to such organs as the heart, the muscles of the reproductive organs and glands.

There is a close relationship between the cerebrospinal nervous system and the autonomic; there are autonomic centres in the former while the latter is not altogether involuntary. The functions of the former have been obvious, but those of the autonomic have been slowly coming to light, revealing a widespread vegetative nature.

Physiological and anatomical evidence for the existence of nerve tracts connecting the hypothalamus with the spinal sympathetic centres has been put forward by Beattie, Brown and Long. Stimulation of the hypothalamus is followed by marked increase in blood pressure, as was shown by Karplus and Kreidl. These investigators found that on stimulation of the hypothalamus centres were found to be present which controlled the smooth musculature of the eye, regulated the lacrimal and sweat glands and the vaso-constrictor and temperature-regulating mechanisms. The probable position of these centres was in the corpus subthalamicum.

Clinical evidence of vegetative centres in the region of the hypothalamus has been supplied by Penfield, who described under the title *Diencephalic autonomic epilepsy*, the symptoms produced by an encapsulated tumour which periodically pressed on the thalamus of both sides. The tumour contact and pressure were clearly above and anterior to the nuclei of the hypothalamus. Autonomic convulsions were discharged by the irritating pressure of the movable tumour and by the interruption of the pressure of the cerebrospinal fluid when the tumour became wedged in the

foramen of Monro, thus shutting off the flow of fluid from the lateral ventricles to the third ventricle. The tumour, in fact, was acting as a ball valve. The patient showed the following sequence of phenomena : there was a prodromal phase of restlessness, followed by a marked rise of blood pressure with flushing of the face, pilomotor disturbance causing the appearance of " goose flesh," lacrimation and protrusion of the eyes (not always constant). There were no convulsive seizures. To Penfield it seemed obvious that the recurring attacks from which the patient suffered were made up of discharges from the centres which controlled or partly controlled the vegetative nervous system.

Another aspect of the function of the vegetative nervous system has been demonstrated by Müller and Chura. Certain forms of skin irritation were brought about by intradermal injections of inert and physiological solutions, even of distilled water. Müller noticed that following the injections there was a more or less rapid decrease in the number of leucocytes. Later he discovered that simultaneously with the peripheral leucopenia a dilatation of vessels and a leucocytosis occurred in the liver and mesentery. From these results and examination of the literature on the subject Chura is of the opinion that such a reaction is a manifestation of the state of the vegetative nervous system and proposes this procedure as a method of examination of a certain part of it.

The vegetative nervous system is divided into two fairly distinct parts (Langley). That which arises from the brain and from the lower sacral region is sometimes called the mesencephalic-bulbo-sacral outflow, autonomic or parasympathetic ; while that portion arising from the thoracic and lumbar regions is called the thoracic-lumbar outflow or sympathetic. (For a full description of the vegetative nervous system and its affections I would refer the reader to the work by Kuntz, also White and to that by Grinker.) The importance of these two divisions in the general make-up of mankind has been demonstrated by Eppinger and Hess. In the translated work of these men by Kraus and Jelliffe two distinct clinical syndromes known respectively as *vagotonia* and *sympathicotonia* have been described. By vagotonia is meant excessive irritability of the parasympathetic system, while sympathicotonia may be looked upon as the antagonistic condition brought about by over-stimulation of the sympathetic. Vagotonics show increased sensitiveness to atropine and pilocarpine. There is contraction of

the pupils, accommodative spasm which in young people is relieved by atropine, the palpebral apertures are narrowed and Mœbius's sign (insufficiency of convergence and accommodation) may be present. The vagotonic has a slow heart, cold extremities, asthmatic spasm and low blood pressure. One may easily think of the opposite condition—sympathicotonia—where the pupils are dilated, there is flushed skin and excessive sweating. These conditions described by Eppinger and Hess are not seen frequently enough, and indeed do not give the pharmacological response that would be expected from them, but such theories impress upon one the various functions of the widespread vegetative system (Sheehan).

Two phenomena have been described, one by Löwi. In the normal eye adrenaline cannot overcome the permanent tonus of the sphincter pupillæ or of the muscle of accommodation. Only when the tonus of the sympathetic is increased generally, for example, in de-pancreatised dogs, can adrenaline acting as a stimulant of the sympathetic nervous system have its full mydriatic action. Löwi has described this phenomenon in several cases of Graves' disease and in diabetes.

The other phenomenon illustrates the reflex stimulus which passes *viâ* the vagus upon stimulation of the trigeminal nerve. Aschner found that pressure upon the eyballs will cause a slowing of the heart beat ; this test of Aschner's is supposed to be particularly effective in vagotonics. This reminds one that a blow from an object, say a golf ball, on the eye will cause the heart to stop, hence the sudden collapse of such patients at the moment of impact.

It is not necessary for our purpose to describe fully the anatomy of the vegetative nervous system, but we must give some attention to the cervical sympathetic (Fig. 130). There are three ganglia, the superior, middle and inferior connected by intervening cords. The superior, the largest of the three, is placed opposite the second and third cervical vertebræ. Reddish in colour and fusiform in shape, it lies in relation in front with the internal carotid artery and internal jugular vein ; behind, the Longus capitis muscle. Of its branches the inferior communicate with the middle cervical ganglion ; its lateral branches, consisting of grey rami, communicate with the upper four cervical nerves and to certain of the cerebral nerves, to the ganglion nodosum of the vagus, to the hypoglossal nerve, while a filament, the jugular nerve, passes upwards to the base of the skull

and divides to join the petrous ganglion of the glossopharyngeal and the jugular ganglion of the vagus. Its anterior branches give rise to the carotid nerves which form the important sympathetic plexus

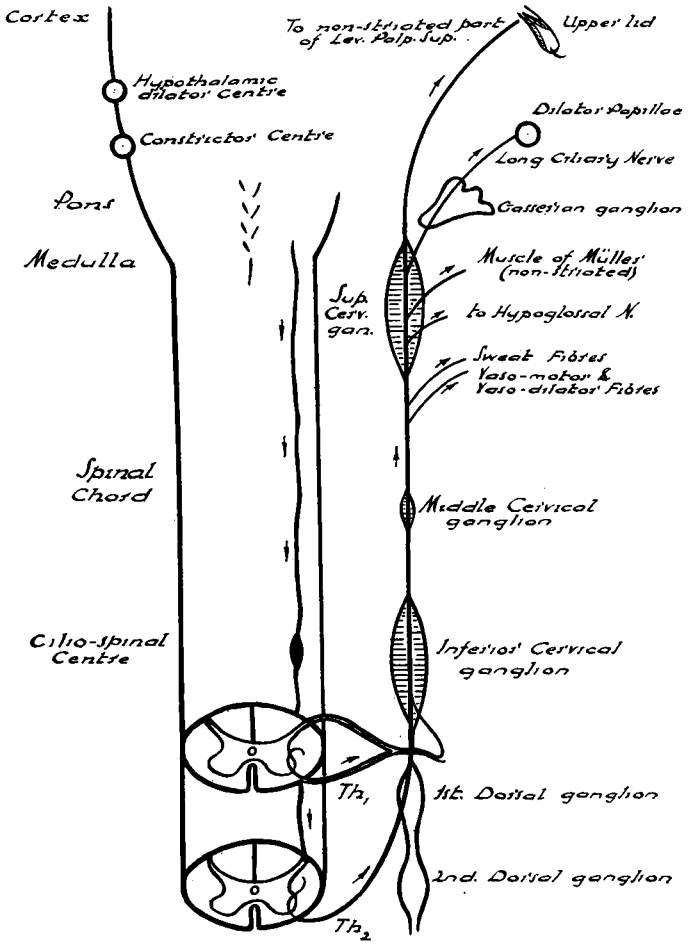


FIG. 130.—Diagram illustrating the path of impulses from the cortex cerebri through the hypothalamus, pons, medulla and spinal cord as far as the ciliospinal centre : their continuation upwards through the cervical sympathetic ganglia and cord.

along the internal carotid artery and its branches. From the plexus on the middle meningeal artery a fine branch passes to the otic ganglion, while another, the external superficial petrosal nerve,

passes to the genicular ganglion of the facial nerve (see Fig. 27). The superior ganglion gives rise to the superior cardiac nerve.

The middle cervical ganglion, the smallest of the three, is placed opposite the sixth cervical vertebra close to the inferior thyroid artery and gives off grey rami to the fifth and sixth cervical nerves. It also gives off the middle cardiac nerve.

The inferior cervical ganglion is situated between the base of the transverse process of the last cervical vertebra and the neck of the first rib on the medial side of the costocervical trunk. It is connected with the middle cervical ganglion by two or more cords, one of which forms a loop around the subclavian artery and supplies branches to it. The last is called the ansa subclavia of Vieussens. The inferior ganglion gives off the inferior cardiac nerve and also the grey rami to the seventh and eighth cervical nerves. But infrequently the inferior cervical ganglion is found fused with the first thoracic ganglion—rarely the second—to form the *stellate* ganglion. When this occurs an additional branch passes to the first dorsal nerve, while occasionally a branch from the second thoracic ganglion (nerve of Kuntz) passes also to the first dorsal nerve. On this account the second thoracic ganglion must also be removed when operating for the relief of vasomotor abnormalities of the arm (see p. 32).

From the carotid plexus branches pass to the submaxillary, otic, sphenopalatine and ciliary ganglia. Conveying impulses from the brain stem fibres pass to innervate the smooth muscle of the glands, the lacrimal, carotid and salivary, and to the dilator muscle of the iris.

Horner's syndrome, which has been described in Chapter IX and is illustrated by Fig. 131, is due to a lesion of the cervical sympathetic trunk, the grey rami entering into it or the cells in the spinal cord which give off these rami. Hence lesions of the lower cervical segment of the spinal cord, the ciliospinal centre at the level of the seventh and eighth cervical and first dorsal nerves are responsible for this syndrome. Tumours or syringomyelia of the cord at this level may be the cause ; enlarged glands pressing on the cervical sympathetic, an enlarged thyroid, neuroma of the sympathetic cord, aneurysm, cervical rib, tuberculosis of the apex of the lung or war injuries may bring about this condition. It is included in the Klumpke syndrome. One may include higher up within the cranium thrombosis of the posterior inferior cerebral artery and

Raeder's "paratrigeminal paralysis of the oculo-pupillary sympathetic."

Enophthalmos is not usually evident after cervico-thoracic sympathectomy, but, lest it should appear and to avoid the contracted pupil, Cardozo and Telford have evolved a certain technique in their operations which avoids such a result. After cervico-thoracic ganglionectomy Mutch found that homatropin produced full dilatation, but cocaine not only failed to dilate the pupil after sympathectomy but also prevented it from dilating fully in the dark. Adrenaline does not affect the pupil after cervico-thoracic ganglionectomy, but when combined with cocaine in daylight the

FIG. 131.—Bilateral partial ptosis due to bilateral cervico-thoracic ganglionectomy. The pupils do not dilate with cocaine. The patient's only complaint is that her friends tell her she has acquired a "dreamy look."

pupil on the denervated side dilates more readily than the normal pupil, a reaction termed by Anderson "the paradoxical pupillary reaction."

After removal of the cervical ganglia the tension of the eye falls immediately; this led to the belief that such an operation could influence the course of glaucoma, but Axenfeld, Lagrange and many others found that the removal of the superior cervical sympathetic has little effect on any form of glaucoma except the very chronic. After cervico-thoracic ganglionectomy the tension of the eye falls but rapidly rises again.

That the sympathetic has the function of a truly trophic nerve has been shown by Asher, who, having cut the cervical sympathetic on one side, exposed both corneæ to the rays of a quartz lamp of the same intensity and duration. Either only the corneæ on the side

without the sympathetic showed lesions (observed by the slit-lamp) or if both corneæ have been affected, on the side without the sympathetic the lesion is the greater and takes longer time to heal. The older contention that permeability is under the regulation of the sympathetic thus receives new support.

Angioneurotic œdema or Quincke's disease is an affection associated with the vegetative nervous system. The disease occurs in people who are of a neurotic disposition and is usually hereditary in character. (Osler.)

By nature it is possibly allergic, many kinds of foods and drugs being responsible for the condition. The symptoms occur suddenly and are confined to areas from one to several inches in diameter. The eyelids, lips and cheeks may be the site of an attack. They become swollen in a sudden manner without pain but sometimes preceded by itching, heat and redness. I have seen it occurring in the lids, in one case without symptoms or redness, in another with intense itching so severe that the skin was torn by the nails. It is not painful but the intense swelling may close the eyelids, the condition lasting for a day or may remain for a week. It is usually discovered that some indiscretion in eating is responsible. One agrees with Atkinson that shell-fish taken with alcoholic drinks is frequently responsible for the onset of this condition, also in some cases the partaking of strawberries.

The type which attacks the larynx may prove fatal, also circumscribed œdema of the brain (Stone) has been known to bring about raised intracranial pressure and paralysis.

The condition of angioneurotic œdema is sometimes seen in tabetic subjects. It is often associated with urticaria, a condition which sometimes yields to calcium medication. The main thing is to eliminate from the food such articles which we suspect are the cause of the disease. Adrenaline or ephedrine used locally or injected may be of benefit and ice or lead lotion applied locally helps to diminish the swelling.

Progressive Facial Hemiatrophy

Synonym : Romberg's Disease

Although described in most text-books as a rare disease, it is really not so. Over 400 cases have been reported in the literature and referred to by Archambault and Fromm. The disease attacks both sexes equally, and either side of the face may be affected. It

may occur in infancy or even in advanced life but it is more commonly met with in the second decade of life. Its hereditary character has not been proved nor has it been found to show a familial incidence.

Progressive facial hemiatrophy is an atrophic process which involves the skin, subcutaneous fat and connective tissues, muscle and bone. The muscular atrophy, however, is not due to loss of muscle tissue but to loss of its fat and connective tissue, the muscular power remaining almost unimpaired, as is easily demonstrated in the case of the extraocular muscles when the orbit is involved. The atrophy may begin at any point, from the forehead or orbit to the mandible or upper part of the neck. In rare cases the facial hemiatrophy has been associated with atrophy of the opposite-sided extremities, a condition in the vegetative nervous system comparable to the crossed hemiplegia in the cerebrospinal system.

Traumatism has been assigned as a cause for facial hemiatrophy. History of a trauma long past or recent has been frequently given. This, however, may only be an exciting cause. Local infective conditions have been mentioned; tonsillitis, angina, alveolar abscess or periostitis of the bones of the jaw. Infective diseases as an ætiological factor have been recorded. Such diseases are diphtheria, erysipelas, scarlet fever, etc. Tuberculosis has been mentioned so frequently that attention has been drawn by Archambault and Fromm to the possibility of apical infection producing a lesion of the cervical sympathetic. These authors, adducing many facts, including the deductions of André Thomas regarding the function of the sympathetic system, have come to the conclusion that their cases and probably most of those described in literature exhibit inescapable evidence of disturbances in the domain of the cervical sympathetic system.

Facial hemiatrophy has thus been held as symptomatic evidence of a lesion of the sympathetic fibres originating in the cilio-spinal centre of the lower cervical cord or of the descending bulbo-spinal sympathetic pathway.

The symptoms begin insidiously, there is wasting at one part of the face; gradually spreading, it involves the fat, then the bones, last and least the muscles. There may be complete facial asymmetry or the area affected may be localised. It is not uncommon to see the orbit singled out. The appearance is characteristic, and on examination one is struck by the dissimilarity of the two

orbits and their contents. The eyelids are thin, the eyelashes may be white in colour or almost absent, the eyeball is retracted into the orbit, the pupil contracted, but the movements of the eyeball are unimpaired. Sometimes the pupil is dilated. Fig. 132 illustrates a case under the author's care—a woman of middle age who has

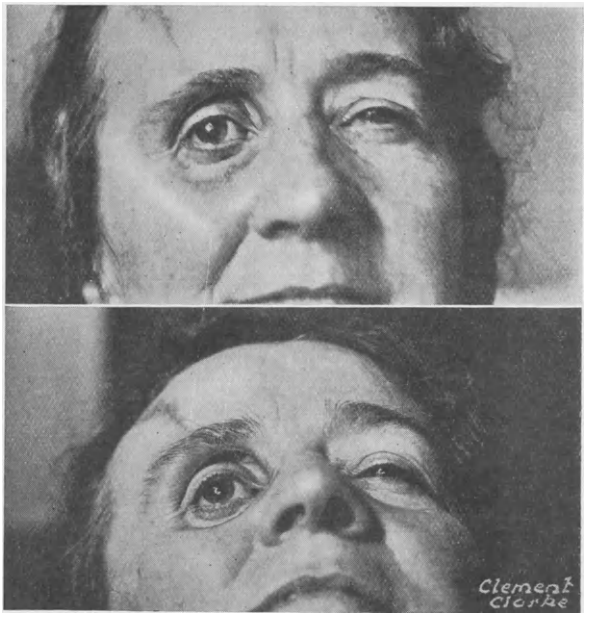


FIG. 132.—Partial facial hemiatrophy confined to right orbit. Note the loss of fat in the orbicularis palpebrarum muscle resulting in extremely thin eyelids; the loss of fat in the orbit causing enophthalmos. The lower photograph shows the deep hollow beneath the superior orbital margin due to the great loss of orbital fat.

The pigment on the forehead marks the place of an old injury.

The right pupil is dilated owing to its being fixed in this position by atropine in the treatment for tuberculous iridocyclitis. The left eye is still sensitive to light therefore the partially closed eyelids.

suffered from tuberculous iridocyclitis and has responded well to tuberculin treatment. Note the thin eyelids, the hollow appearance above the eyeball beneath the sharp edge of the roof of the orbit. The pupil is permanently dilated, due partly to the pupil being attached by adhesions in a dilated position. This patient received a severe blow on the forehead above the right orbit twenty years ago, followed not long afterwards by apparent shrinking of the orbital contents.

In orbital cases Horner's syndrome is present—ptosis, enophthalmos and miosis, but sometimes the pupil is dilated. Also pigmentation of the skin may be present as on the forehead in Fig. 132. Many and various clinical associations, such as sclerodermia and syringomyelia, have been described with facial hemiatrophy. Trotter showed a case of recurrent herpes zoster of the face with localised facial hemiatrophy. Flint demonstrated a case where a previous history of facial paralysis was given and now a persistent recurrent ulceration of the cornea was present. He drew attention to the enophthalmos and the enlarged pupil on the same side as the facial hemiatrophy. On this side there was no reaction to light directly or consensually but there was sluggish reaction to accommodation.

Trigeminal neuralgia and injuries to the trigeminal are common but facial hemiatrophy is not common. It is therefore unlikely that the cause of a facial hemiatrophy can be imputed to the trigeminal nerve.

Both epilepsy and migraine have been mentioned in association with cases of facial hemiatrophy or in their relatives, while psychoses have been observed in cases where the disease has developed later in life.

Surgery of the Sympathetic Nervous System

The volume of literature on experience gained by surgery of the nervous system is rapidly increasing. Recent text-books of surgery, such as " Post Graduate Surgery," have devoted considerable space to the subject. As Abel says, " The proper functioning of the autonomic system depends on perfect adjustment of its components, and overaction of its sympathetic portion results in spasm of blood vessels or of the intestinal musculature." By interrupting the sympathetic part the passage of abnormal impulses is arrested. When the vasoconstrictor fibres to the blood vessels are divided there is an increased flow of blood to the part concerned. If, however, the tissues and vessels show pathological change as in retinitis pigmentosa little result may be expected from any form of operation. Good results have been reported commonly in connection with Raynaud's disease, thrombo-angeitis obliterans (Buerger's disease), retinal arterial spasm, essential hypertension, causalgia and cardiospasm. White fully discusses the effect of cervicothoracic ganglionectomy in the treatment of epilepsy. Penfield (in a letter to Dr.

Stanley Cobb) reported that in a case of bilateral epilepsy the convulsions became localised to one side of the body after a unilateral sympathetic denervation. Crile has performed the operation of bilateral adrenal denervation on the theory that epilepsy results from rhythmic hyperactivity of the entire energy system of the body. Further investigation is required to establish definite conclusions regarding the effect on epilepsy by ganglionectomy.

There is no definite evidence that sympathectomy can influence generally the course of migraine, although Dandy reported two cases of intractible migraine successfully relieved by cervicothoracic ganglionectomy.

Pain of sympathetic origin which has been relieved by operation on the sympathetic nervous system has been described in the report of the clinic of Swift and Flothow. The case quoted was that of a young woman who had sustained a laceration over the first metacarpal bone. In spite of all suggested treatment the pain remained. Two types of pain were present, one a constant ache, the other an acute shooting type. Injection of novocaine of the right dorsal sympathetic trunk was done. This produced various changes. The right hand became distinctly warm and there was an absence of perspiration. Within twenty minutes there was a complete Horner's syndrome. The pain in the hand entirely disappeared. This led to the operation of dorsal sympathetic ganglionectomy being done. Since then there has been no recurrence of pain.

Reports from this clinic state that Horner's syndrome has been avoided by removing the second and third ganglia rather than the first and second, as it is found that vasomotor impulses to the upper extremity arise not higher than the third dorsal segment (see p. 32).

A case is also reported by Harris of a young woman who had suffered from shooting pain in the right cheek up to the temple. Previous injections of alcohol were given without benefit, the infra-orbital nerve was avulsed, the Gasserian ganglion was twice injected with only slight improvement. Procaine injection of the right stellate ganglion led to the disappearance of pain for several hours. The resection of the right stellate ganglion was followed by permanent relief from pain.

Two cases of recurrent hemicrania relieved by stellate ganglionectomy have been reported by Craig. One of these cases suffered from severe right temporal neuralgia with lacrimation from the right eye and nostril. The attacks began after exposure to cold.

This was a case of paroxysmal sympathetic neurosis and not hereditary migraine. (Harris.)

The recital of these cases seems to lead one to the conclusion that both afferent and efferent impulses pass through the sympathetic nervous system, a statement supported by that of Helson, Frazier and many others; also by studying the reactions in Raynaud's disease and comparing the effects of ulnar anæsthesia evidence has been obtained that the human sympathetic nerves contain vasodilator as well as vasoconstrictor fibres. (Lewis and Pickering.)

In a few days after cervico-thoracic ganglionectomy ptosis is very marked, the conjunctival vessels are engorged, the tension of the eye falls to such an extent that accommodation is impaired. Glasses have to be altered. After a fortnight the ptosis is lessened and the tension rises. Enophthalmos is not usually marked. (Gask and Ross.) It is noteworthy that in many cases one has seen where the cervico-thoracic ganglion has been removed enophthalmos is very slightly present, if present at all.

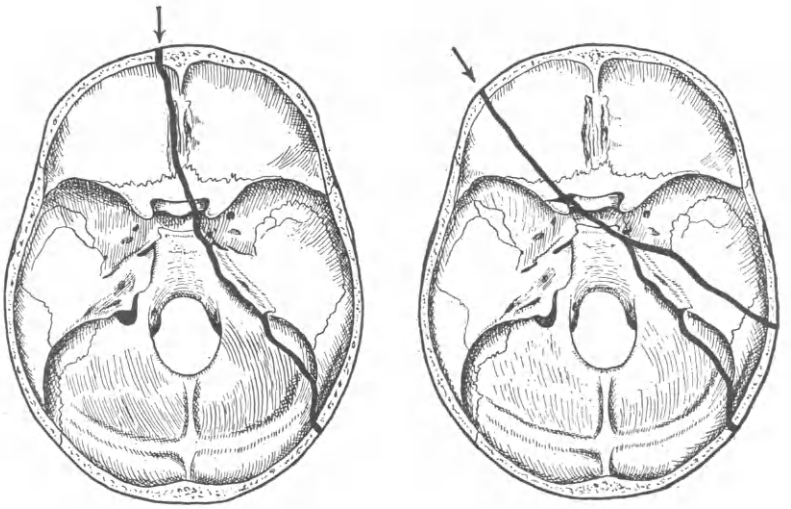
In cases of severe facial paralysis Leriche has advocated the removal of the superior cervical ganglion in order that the upper lid should fall down somewhat for the protection of the eye from undue exposure.

Adson and Brown, as well as Lewis, have shown that pilocarpine injections can induce sweating in an area deprived of its sympathetic supply presumably because the drug acts directly on the sweat glands. It is therefore useless to use this method in a test for sympathetic denervation.

CHAPTER XV

OCULAR MANIFESTATIONS OF HEAD INJURIES

OWING to the enormous expansion of motor traffic the number of accidents occurring has rapidly increased during the past few years. Almost daily in private practice or in hospital one has to



(*a*) (*b*)

FIG. 133.—(*a*) Force applied to the median frontal region: The fracture passes backwards from the perpendicular plate of the frontal bone to the cribriform plate of the ethmoid, thence between the optic foramina to the body of the sphenoid: from there the fracture diverges to the opposite side, and tearing off the posterior clinoid process passes along the petro-occipital suture to the jugular foramen. (Rawling.)

(*b*) Force applied to the lateral frontal region in the situation of the external frontal process. The fracture tears away the anterior clinoid process, comminuting the roof of the sphenoidal sinus. (Rawling.)

deal with these cases. In the past, when horse traffic was the rule, people were "run over" and injured generally by pressure, but with motor traffic the commonest occurrence is that of a pedestrian being struck by a car and pitched violently to the ground or the passenger in the car being thrown forwards from his seat; in either

case violent force is applied, often to the skull, the result of which varies greatly from simple concussion to extensive fracture. As Bacon and Le Count have said, "there is no evidence that their cranial injuries are received while the victim is still erect and they have failed to find much evidence of crushing of the head beneath wheels of vehicles." From a neuro-ophthalmological point of view these cases are of extreme interest. An intimate knowledge of the ophthalmic symptoms following cranial fractures lends considerable aid to the accurate diagnosis of the position and extent of injury in such cases, and although the problem of surgical intervention is one which presents itself to the neuro-surgeon, yet the solution is greatly aided by the knowledge and recognition of these ocular manifestations.

A study of the records of cases of fractures of the skull treated in the various hospitals brings out a surprising variety of causes, some of which are tabulated below :—

Causative Agents.	McCreery & Berry.	Vance.
Automobile	110	189
Falls	179	156
Blows	62	33
Bullet	3	
Unknown	140	100
Street car	14	19
Train	3	
Crush	5	
Horse and Wagon	4	

In many cases there is partial or complete blindness immediately. This is explained, not by the number of fractures which pass through the optic foramina, but most probably due to extravasation of hæmorrhage into the sheath of the optic nerve. In Hoelder's cases 53 out of 86 showed injury to the bones forming the optic canal, but Rawlings says, "The vast majority of anterior fossa fractures avoids the immediate vicinity of the optic foramen, passes by preference between the two foramina or diverges towards the sphenoidal fissures." In a report on 512 necropsies, Vance showed that in only 10 per cent. did fissured fractures involve the optic foramen. It is probable that fracture and displacement of the anterior clinoid processes pressing upon the nerve may be account-

able for a large proportion of cases affecting the optic nerves. Hogarth Pringle examined 174 cases post mortem which died from fracture of the skull. Of these, only 13 showed a fracture involving the optic foramen. In 2 of these the foramen was fractured through its roof, yet the optic nerves were not injured nor was hæmorrhage

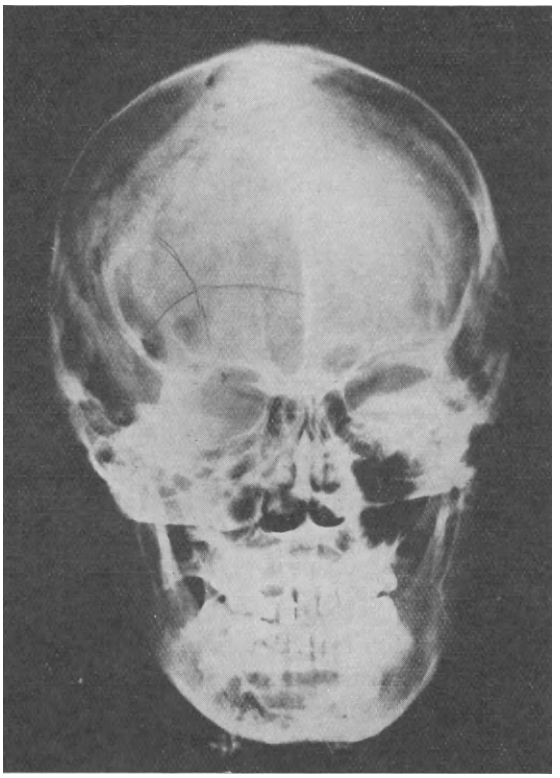


FIG. 134.—Skiagram of skull taken antero-posteriorly. A stellate fracture of the frontal bone is seen extending downwards through that portion of the right wing of the sphenoid bone which forms part of the lateral wall of the orbit. (Taken by H. M. Worth.)

found in the sheath. In 16 cases hæmorrhage into the nerve sheath was found. This leaves a large proportion of cases of blindness which are unexplained by fracture through the optic foramen or by hæmorrhage into the sheath of the optic nerve. Laceration of the nerve has been found at autopsy in some cases, while in others there is a state of physiologic block due to concussion or from stretching.

According to Traquair, Dott and Russell, damage to the optic canal as evidenced by the occurrence of temporal defects in both visual fields is an uncommon complication of head injury. They made an examination of 27 cases. They point out that chiasmal damage may easily escape notice, especially when visual acuity is little impaired. It is suggested that in both traumatic and tumour cases the primary damage is to the vessels supplying the chiasma. There is much resemblance between the field changes which occur in injury and in tumour cases.

Rawling's investigations show the tendency of most fractures of the skull to converge towards the pituitary region (see Fig. 133). The optic canal too may be involved in any fracture of the skull, but it is not commonly involved when there is violence applied in the neighbourhood of the external angular process of the frontal bone (Fig. 134). Many cases of fractured base involving the bones of the pituitary region in the neighbourhood of the floor of the third ventricle show the presence of sugar in the urine, a symptom which might be more commonly sought for (Bacon and Le Count).

The earlier classification of acute cerebral traumas was based chiefly on the type of bone lesion, but lately the tendency is to consider these cases from the viewpoint of increased intracranial pressure and the accompanying neurological symptoms. It has been pointed out by Bagley that considerable aid is forthcoming from a classification based on post-mortem findings. His classification is as follows :—

Group (1) Simple depressed fracture.
,, (2) Compound fracture with or without depression.
,, (3) Extradural hæmorrhage.
,, (4) Blood overlying the cortex.
,, (5) Cortical injuries.
  (*a*) cortical lacerations ;
  (*b*) diffuse superficial extravasations ;
  (*c*) single or multiple cortical clots.
,, (6) Extravasations in the vein of Galen systems.
,, (7) Hæmorrhage in the brain stem.

Nevertheless, there are certain symptoms which draw our attention to the location of the fracture, *e.g.*, in fracture of the anterior fossa there may be palpebral, peripalpebral and subconjunctival hæmorrhage. Blood is found in the tissue of the eyelids

usually making its way from the inner canthus, while subconjunctival hæmorrhage is discovered by raising the upper lid, thus exposing a hæmorrhage descending from above and from the outer side. Hæmorrhage into the orbit associated with injury to the cavernous sinus or internal carotid artery results in severe and immediate proptosis. Hæmorrhage from the lesser vessels such as the ethmoid will not produce such marked proptosis. As so many fractures, involving both the anterior and middle fossa, traverse the body of the sphenoid bone it is not difficult to appreciate the fact that the cavernous sinus can thus be readily injured and that portion of the internal carotid artery which lies embedded in the outer wall of the cavernous sinus may also be involved; this results in the formation of an arterio-venous aneurysm, which later leads to the condition known as traumatic orbital aneurysm, in which the proptosis takes place possibly one month or so after injury (see p. 217). (Fig. 135.)

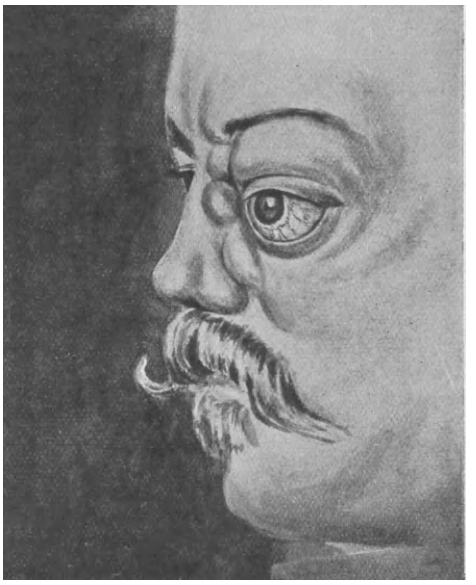


FIG. 135.—A case of traumatic orbital aneurysm. (Rawling.)

An unusual case which terminated fatally has been related to the author by his friend W. Rutledge. A young man was flung off his motor cycle and sustained severe concussion. While the patient was still alive it was noticed that one eye was proptosed and was pulsating. This condition followed the accident immediately. At autopsy the roof of the orbit was found to be severely fractured and brain matter was protruding into the orbit causing proptosis: the pulsation of the brain was thus communicated to the globe.

The following points are of importance in diagnosing a case of *traumatic orbital aneurysm* or *pulsating exophthalmos*:

Proptosis.—The first symptom is proptosis with dilated con-

junctival and eyelid blood vessels; this takes place after the lapse of one or two months from date of injury. The proptosis can be diminished by steady pressure backwards.

Aneurysmal Symptoms.—The eyeball and conjunctival vessels pulsate synchronously with the heart's beat; this, however, is modified or abolished by pressure on the ipso-lateral common carotid artery.

By palpation a definite thrill can be felt and by auscultation a loud murmur, as of a waterfall, can be heard. This is audible over a wide area, including the bony orbit and bones of the vault. The patient experiences noises in the head, a whirring noise, which is intensified by bending down.

Trophic Symptoms.—Owing to pressure there is lymphatic obstruction, so that the cornea may become cloudy and chemosis of the conjunctiva takes place. If the lids cannot be closed there is great danger to the eye from keratitis or sloughing of the cornea. Pressure may bring about diplopia from involvement of muscles or nerves. The pupil may be dilated due to paralysis of the third nerve. Through involvement of the fifth nerve trophic changes are brought about and considerable pain may be experienced from stretching of its branches.

Fundus.—The ophthalmoscope shows great engorgement of the retinal veins and a swollen optic disc. Vision rapidly diminishes, even to complete blindness.

Rawling says repeated hæmorrhage and secondary infection may place the patient in imminent danger of his life.

Locke says 588 cases of pulsating exophthalmos have been described since 1809. 23 per cent. were spontaneous, while 77 per cent. were traumatic in origin, and of the latter 94 per cent. had an arterio-venous communication between the internal carotid and the cavernous sinus. A pulsating swelling above the inner canthus is usually seen with the arterio-venous type and the exopthalmos is greater than in other purely aneurysmal forms. He recommends prolonged carotid compression before tying the carotid artery. The exophthalmos in rare cases subsides spontaneously.

Fracture of the ethmoidal bones is sometimes associated with the escape of air which may pass into the orbit, so that proptosis is produced by the patient blowing his nose. These cases often recover in two or three weeks. In such cases the patient is well advised to refrain from blowing his nose.

Among the cardinal symptoms of fracture of the cranium Jentzer describes the importance of the " intervalle libre "—the period of consciousness which takes place immediately on recovery from the initial concussion and which presumes an anatomical lesion, a short interval indicating severe hæmorrhage.

Pupillary Changes.—At this stage one might be tempted to dilate the pupils to observe if there is any sign of raised intracranial pressure taking place, but, mydriatics must not be used lest the important symptom of the dilated and fixed pupil be overlooked. Morphine also should be avoided if possible as it causes contracted pupils and at the same time conceals signs of oncoming stupor.

As long ago as 1887 Macewen drew attention to the difference in size of the pupils in certain head injuries. These were cases of traumatic intracranial hæmorrhage. He noticed that one pupil became dilated and fixed while the other remained acting normally, and that during the lucid interval it was sometimes discovered the patient could not see with the eye in which the pupil was dilated—indicating severe pressure on the entire cerebral hemisphere. Some observers have noticed the dilated pupil becoming normal during the release of pressure as the operation proceeded (Macewen and de Quervain-Hoessli). The importance of the unilateral dilated pupil has been stressed by Cushing, Holman, Scott, Rand and Jentzer. All of these agree that the dilation and fixation of the pupil is a valuable aid in determining the location of the intercranial injury and hæmorrhage following head injuries. Indeed, it has proved to be of greater localising significance than bleeding from the opposite ear or even a hemiparesis of the same side (Rand). There is one precautionary point, however, to which Goulden has drawn attention and which has been observed several times by the author, namely, that direct injury to the optic nerve will result in a dilated pupil which, however, will contract on illuminating the opposite eye but remains inactive to direct light stimulation. Holman and Scott say " that operative intervention should be directed towards the side on which this dilation and fixation first appear," also " its transitory character makes accurate and oft repeated observation necessary from the moment of injury," while Rand states that in cases in which hemiplegia and the dilated pupil were homolateral intracranial hæmorrhage was found on the same side.

Rand admits it is often difficult to explain the occurrence of homolateral hemiplegia, but, as Kernohan and Woltman have shown,

it is possibly due to displacement of the mesencephalon and pressure or contusion of the opposite cerebral peduncle against the sharp free edge of the tentorium cerebelli.

Papillœdema.—The difficulty of examining the fundus of an uncontrollable patient without even the influence of a mydriatic has been acknowledged by most neuro-surgeons. Choked disc was not observed in any of Cohen's 75 cases of fractured skull. McCreery and Berry say that examination of the fundi would probably show a larger percentage of changes if carried out as a routine and at frequent intervals, but they acknowledge that in the hands of the average general surgeon with an uncontrollable patient and without artificial dilation of the pupils the findings may be questioned. They urge, however, repeated funduscopic examination as a routine part of the treatment of all cases of intracranial injury. Cairns in his series of 80 cases of head injury in civil life observed definite papillœdema in only 7 cases, the earliest onset of which occurred five days after injury, but Holman and Scott observed its onset eight hours after injury. In cases of blurring of the disc, Cairns has found a large blood clot, severe cerebral œdema and also a collection of cerebrospinal fluid in the subdural space. As Cohen pointed out, the examination of the fundus immediately following skull injuries may reveal pre-existing contributory factors and so be helpful from the medico-legal point of view. A large number of funduscopic examinations have been made by Kearney immediately after accidents causing fracture of the skull. The invariable finding was an œdematous retina in which even the small veins were enlarged out of all proportion to the accompanying arteries. In some cases the entire eye contents seemed œdematous and all fundus details were obscured. Cases of marked papillœdema were not observed at first.

Dr. Liebrecht agrees with Cairns as regards the time of onset of papillœdema after the accident, namely, three to eight days, and says the type of papillœdema is somewhat different from the papillœdema of raised intracranial tension caused by tumour of the brain. In the case of injury to the head the blood in the neighbourhood of the disc is more apparent and the exudates larger and more marked than in papillœdema caused by the presence of an intracranial neoplasm.

Blindness.—Hæmorrhage into the sheath of the optic nerve has been commonly reported and commented upon. The source of the

hæmorrhage may be from the subdural cranial space, from rupture of vessels passing between the nerve and its covering or from the central vessels of the retina which run a short distance inside the dural sheath before they enter the optic nerve. In 8 of Pringle's 16 cases with intravaginal hæmorrhage, the hæmorrhage was seen in the sheath or cut surface of the nerve as it presents after the brain is removed from the cranium, and in all there was blood in the subdural space in the middle fossa of the skull, but in 7 cases no blood showed on the cut surface of the nerve. The blood in these places was only present in the orbital portion of the nerve and it seems certain that the blood was effused into the nerve sheath

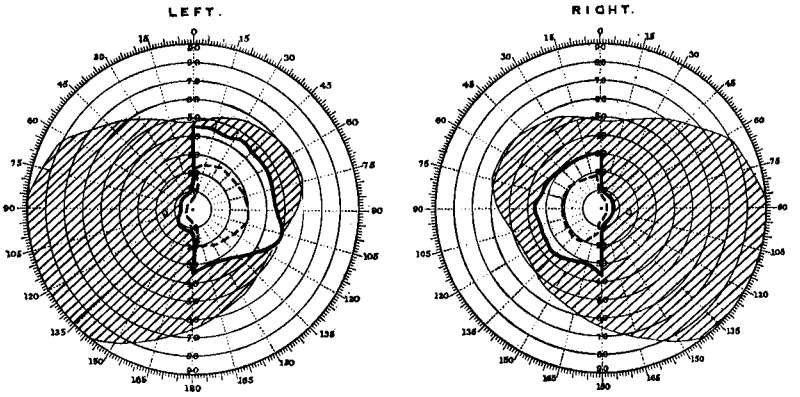


FIG. 136.—Bitemporal hemianopia due to partial optic atrophy following fracture of the skull. (Peter.)

in the orbital or distal side of the foramen, and Pringle was convinced the hæmorrhage began at the distal extremity of the nerve close to its entrance into the eye.

From the number of cases of monocular blindness following slight injury we must lean to Pringle's theory, that the intravaginal hæmorrhage is brought about as the result of the varying tensions produced in the tissues in the orbit by the violence. Where fracture of the foramen was found only 3 of the 13 cases showed hæmorrhage into the sheath.

Damage to the optic nerve may be complete or incomplete. The history usually given is that after an injury such as a blow on the head there was sudden blindness on one side. Ophthalmoscopically nothing abnormal is seen, the pupil on the affected side is dilated and inactive to direct light stimulation, but in the course

of two or three weeks the beginning of optic atrophy is observed. Cases have been reported where the injury was not so severe and only partial or quadrantic loss of vision was found (Fig. 136).

Two cases have been reported by Grey Clegg—one a boy in whom the upper half of the field of vision was lost due to a fall; the other a woman who had been struck by a banged door on the nose and as a result the lower half of the field was now absent. Bachauer illustrates the fields of vision of such a case where, not only symmetrical quadrantic defects were present, but absolute scotomata were found in the remaining portion of the fields, indicating that damage to crossed and uncrossed fibres had taken place at the chiasm. The Barkans also have reported on 5 cases in which quadrantic defects were found, some in association with a central scotoma, and in all some pallor of the disc was observed. Traquair points out that defects occurring as the result of injury are usually absolute and have steep edges. If the chiasm is injured there may be complete blindness on one side and a quadrantic defect in the field of vision of the other eye.

Delayed Blindness.—There is a condition which is becoming more evident from day to day and which up to the present has not had sufficient attention drawn to it, namely, delayed cases of blindness in which optic atrophy slowly supervenes on an accident in which injury has been applied to the skull, generally in the frontal region. The author has under his care the case of a young woman who five years ago was knocked down by a motor cycle. She recovered without obvious injury and soon seemed to be in much her usual state of health; then, four years after the accident she noticed her vision was diminishing and on examination the ophthalmoscope revealed the presence of a partial primary optic atrophy which one year later had become almost complete, leaving only perception of light. The fields of vision had shown steadily concentric contraction. Reference has already been made to similar cases reported by Margoline. Cushing has operated for the removal of a pituitary tumour and found on exploration nothing but a cystic condition of the arachnoid membrane present. Thirty-three cases of cysternal arachnoiditis have been described by Horrax. All were operated on with the presumptive evidence of tumour, and in none was a tumour found. In the 5 cases that died all showed histological thickening of the arachnoid membrane forming the cerebellar and basal cisternæ. Of the etiology of this condition Horrax mentions

among other causes trauma, infected contusion of the scalp, ocular and naso-pharyngeal infection, and extension inwards of the infection in otitis media. The process has been attributed to a serous meningitis, inflammatory gliomatosis or œdema of the brain. Such a condition may involve any of the cranial nerves, and if the chiasmal region is affected a bilateral affection of the optic nerves may take place. Papillœdema has been observed in the localising form of chronic arachnoiditis, while anæsthesia of the corneæ has been described by Monier-Vinard in a case of cystic arachnoiditis involving the cerebello-pontine angle (see p. 275).

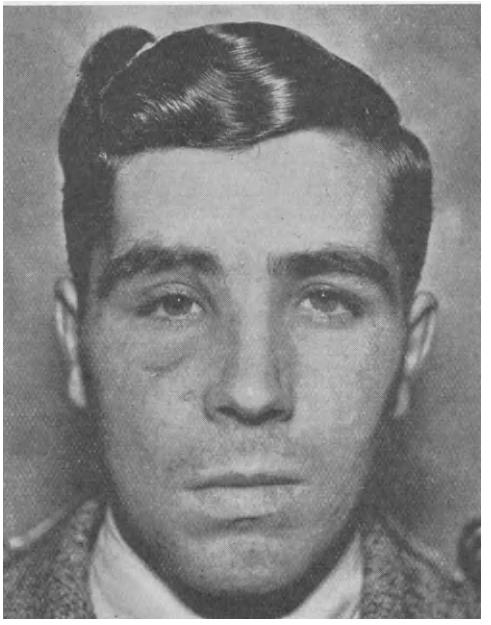


FIG. 137.—Intracranial injury resulting in right-sided ptosis and enophthalmos. There is vertical and lateral diplopia which increases on looking downwards and to the right due to paralysis of the inferior rectus, inferior oblique and external rectus muscles.

Owing to injury of the branch of the third nerve to the inferior oblique (which branch conveys the fibres to the ciliary ganglion for the innervation of the sphincter pupillæ) the pupil is dilated, although the remainder of Horner's syndrome is present due to intracranial injury of sympathetic fibres.

Lateral Deviation of the Head and Eyes.—As mentioned in Chapter III, when a paralytic lesion is present on one side of the cerebral hemisphere there is lateral deviation of the eyes to that side and away from the paralysed extremities. So in cases of head injuries the deviation of the head and eyes is found to be towards the side of the major lesion.

Cranial Nerve Palsies.—Lesions producing disturbances in the ocular movements by interfering with the cortical centres or the connecting fibres between these centres and the brain stem nuclei always affect lateral or vertical deviation of the eyes, but do not so commonly affect single nerve nuclei.

A lesion of the third or oculo-motor nerve is uncommon, although

Grinker has seen some instances due to petechiæ in the distribution of the oculo-motor nerve nuclei. Paresis in the vertical plane due to contusion of the midbrain may be present and may affect one or both eyes, while nystagmus may be present. It is more common to find isolated branches of the oculomotor nerve affected although complete external and internal ophthalmoplegia have been seen.

In Chapter III Wolff's theory is described regarding the cause of the sixth or abducent nerve palsy which is so common in brain lesions. The suggestion is that owing to the relationship of the sixth nerve to the back of the apex of the petrous portion of the temporal bone (see Fig. 24), pressure or traction at this point can easily result in injury to the nerve (Fig. 137).

The fourth or trochlear nerve is the least commonly affected cranial nerve according to Sherren. In one case of the author's the only after-effect of an injury sustained while driving a car which came into collision with another was a fourth nerve palsy; the patient had permanent double vision on looking downwards and to the side.

The fifth nerve is not often affected. The second and third divisions pass respectively through the foramen rotundum and the foramen ovale two foramina which lie in front of the spheno-petrosal suture, a suture traversed by the majority of middle fossa fractures. Therefore these two nerves are seldom involved.

The first division, however, as it passes through the superior orbital fissure (sphenoidal fissure) may be injured so that complete anæsthesia of both cornea and conjunctiva may follow, and if the eyeball in such cases is not protected corneal ulceration or sloughing may ensue. Fortunately, such complete injury to the ophthalmic division of the trigeminal is uncommon.

The seventh or facial nerve is the most commonly affected of the cranial nerves and is often associated with deafness due to a concomitant injury to the eighth nerve. Complete facial paralysis may be associated with ectropion or eversion from paralysis of the orbicularis palpebrarum which, due to the displacement of the punctum lacrimale, allows tears to fall over the cheek; the eyelids do not close properly so that the conjunctiva becomes infected and chemosed, and even the cornea through exposure may ulcerate. Delayed facial nerve paralysis may be the result of involvement of the genicular ganglion or it may indicate meningeal infection.

SUMMARY OF SYMPTOMS OF FRACTURE OF THE BASE OF THE SKULL.
(After Rawling)

Anterior Fossa.	Middle Fossa.	Posterior Fossa.
Hæmorrhages.	Hæmorrhages.	Hæmorrhages.
Subconjunctival.	Into the temporal region.	Into the nuchal region.
Palpebral	From the mouth.	Into the occipital region.
Peripalpebral.	From the nose.	Into the post-auricular region.
Orbital.	From the ear.	
Retinal.		
From the nose.		
From the mouth.		
Cerebrospinal fluid.	Cerebrospinal fluid.	Cerebrospinal fluid.
From the nose.	From the nose.	None.
From the mouth.	From the mouth.	
	From the ear.	
Brain-matter.	Brain-matter.	Brain-matter.
From the nose.	From the ear.	None.
Air-escape.	Air-escape.	Air-escape.
From the frontal sinus.	From the mastoid antrum.	From the mastoid antrum.
From the ethmoidal cells.		
Nerve-involvement.	Nerve-involvement.	Nerve-involvement.
Olfactory.	Fifth (second and third divisions).	Seventh.
Optic.	Sixth.	Eighth.
Third.	Seventh.	Ninth.
Fourth.	Eighth.	Tenth.
Fifth (first division).		Eleventh.
Sixth.		Twelfth (?).

The following instances of sequelæ to cranial injury may be quoted. In 2 cases described by Lillie and Adsom, unilateral central and annular scotomata were produced by callus from a fracture extending into the optic canal (see Fig. 47). At the time of injury both patients had ecchymosis of the eyelids and the pupils were unequal, but vision did not become affected until six weeks later. Radiograms were made according to Camp and Gianturco's technique and these gave evidence of abnormality of the contour of the optic canal involved. An exploratory decompression in one case led to the removal of detached pieces of bone, including anterior clinoid processes. The fracture had involved the optic foramen on its lateral and inferior surface, the resulting callus had compressed the optic nerve to two-thirds its normal size. The optic nerve was decompressed thoroughly, callus and bony fragments being removed. There was no hæmorrhage in the orbit nor in the optic nerve sheath. The earlier radiograms did not reveal any fracture of the foramen, and as operation was done too long after injury the defects in the field of vision were permanent.

Another interesting case was that of a boy aged five years who was knocked down by a motor car. Extensive fracture of the skull

led to partial primary optic atrophy of the left eye, paralysis of the left sixth cranial nerve allowing the left eye to converge and paralysis of the facial nerve which recovered. A mastoid operation was performed by Deacon on the left side six weeks after the accident. Now exactly one year after the accident during an examination, when a mydriatic oculet was placed in the conjunctival sac, the little patient began to cry but no tears came from the left eye. His mother then said she had noticed this absence of tears since the accident. Apparently the nerve of secretion to the lacrimal gland was injured and, as the eye had its normal amount of moisture, only that portion of the secretory nerve which is concerned with emotional weeping, namely, the seventh cranial nerve, had been involved (see Chapter III, p. 59).

Cases of gross dislocation of the eye have been reported, one by Goulden in which fracture of the skull by compression led to complete removal of the skull cap, leaving the dura intact, the patient being conscious throughout. The left eye was dislocated forwards, protruding beyond the margin of the lids. It was easily restored under an anæsthetic and next day the patient could read small print with this eye.

Kearney, quoting Dr. Sharpe, advises that all patients with fracture of the skull should be placed in bed, kept absolutely quiet, free catharsis maintained, given a liquid diet and an ice helmet applied to the head. If no signs of intracranial pressure develop the foregoing treatment is all that is given, but if the intracranial pressure is slightly increased or up to nearly double the normal, then in selected cases repeated lumbar puncture may be performed and the condition relieved. If, however, the cerebrospinal fluid pressure is found, both with the ophthalmoscope and at lumbar puncture, to be double the normal or even more than double, and the pulse rate may or may not be greatly lowered from medullary compression, then he advised a simple cranial decompression to be performed for the relief of pressure and for drainage before a possible collapse of the medulla occurs. An examination of the fundus of the eye twenty-four hours after a lumbar puncture for cranial decompression has been done usually reveals a reduction in the amount of the œdema that had existed previous to the operation.

Extradural Hœmorrhage.— In all fractures of the skull there is some blood between the inner surface of the skull and the dura mater ; in many cases the amount is small and is of little or no con-

sequence, but if such a vessel as the middle meningeal artery is torn the amount of blood effused into the epidural space becomes a serious menace to life through the resulting cerebral compression.

The meningeal vessels lie in grooves on the inner surface of the skull. One has often seen these vessels practically enclosed in a bony canal so that when a fracture takes place the artery is readily torn across and bleeding occurs at the line of fracture.

Vance found the greatest number of fatalities occurred between thirty and forty years of age, the age when the dura mater is not so adherent as in later life. The escaping blood forces the dura away from the skull and the underlying brain is compressed.

The history of the fracture involving the middle meningeal artery is briefly as follows: After momentary concussion or a brief period of unconsciousness the patient recovers and seems to be in much his normal state. This lucid interval may last for hours. After this interval the patient complains of headache, there may be vomiting and slowing of the pulse-rate with gradually increasing stupor. Respiration becomes stertorous or Cheyne-Stokes. The pupil on the side of the injury becomes dilated and occasionally papillœdema quickly appears, the corneal reflex on the opposite side may be lost and a progressive hemiplegia (accompanied by aphasia if the clot overlies the speech centre) appears. These symptoms may be called the characteristic syndrome of *hæmorrhage* of the *middle meningeal artery.*

Over one-half of Vance's cases died within twenty-four hours.

Especially fatal are ruptures of the lateral venous sinus in posterior fractures, the brain in this region being extremely vulnerable to pressure.

Subdural Hæmorrhage.—Subdural hæmorrhage may be unilateral or bilateral and results from the soft cerebral tissues coming in violent contact with the bony skull. This type of injury is found in the greatest number of cases and therefore accurate diagnosis is of the utmost importance. The ocular symptoms must be carefully observed; for instance, Holman and Scott describe one of their cases where a left decompression was done because of a right hemiparesis, disregarding the dilated right pupil. Twenty-four hours later the patient died and autopsy revealed an external, right-sided subdural hæmorrhage over the frontal and temporal regions with a long laceration of the cortex of the temporal lobe.

It has frequently been mentioned that later dilatation of the

opposite pupil occurs and remains so, there being no response to light stimulation.

If subarachnoid bleeding takes place, blood is found in the cerebrospinal fluid and hæmorrhage may appear on the retina or even in the vitreous body. Hæmorrhage in the base of the brain is characterised by cranial nerve palsy, rigidity of the neck and symptoms of pressure on the medulla.

Chronic Subdural Hæmatoma.—This condition, also known as pachymeningitis hæmorrhagica interna, results from a trauma which may have been so slight as to be overlooked or the injury may have been severe. Trotter believes the bleeding is of venous origin from the cerebral veins passing from the brain to the tributaries of the superior longitudinal sinus.

Its symptomatology is extremely varied, but one important feature is *headache*, which is the most common symptom (with raised cerebrospinal fluid pressure) accompanied by sleepiness, forgetfulness, vertigo, thick speech and irritability. The headache, usually severe and persistent, becomes worse as the drowsiness passes into stupor. The periods of stupor gradually increase in length and finally merge into a profound coma. At this stage the size of the pupils may change. Although it has been said that papillœdema is not common and hemiplegia is rare, in all of Rand's cases papillœdema was observed, showing from 1 to 6 diopters of swelling, the greater swelling usually occurring on the side of the hæmatoma. In doubtful cases Holmes urges prompt surgical treatment.

Head Injuries in Children.—In children the dura mater has not a firm attachment to the inner surface of the vault of the skull, which explains the possibility of having an extensive fracture of the skull without producing cerebral damage. The dura, however, is firmly attached to the base, hence any fracture in this region lacerates the membrane. Another factor which explains the fewer number of cases of extradural hæmorrhage in children is the shallowness of the grooves containing the meningeal vessels. In Beekman's series of 331 cases of injury to the head in children under thirteen years of age there was not a single instance of extradural hæmorrhage from the meningeal arteries. He noticed also that children stand severe cranial injuries better than adults ; that frequently fractures of the base of the skull in children may be overlooked for lack of clinical symptoms.

The symptoms of fracture of the skull and brain injury are similar to those seen in the adult. *Headache* was the commonest symptom. In many cases the child's behaviour after recovery from the accident changed. In 10 per cent. of cases originally diagnosed as psychopathic personalities, Kasanin found a history of brain injury during childhood or adolescence.

The corneal reflex has already been briefly mentioned. As the trigeminal innervation from the cortex is contralateral a cerebral hæmorrhage on one side may cause loss of corneal sensitivity on the opposite side, shown by touching the cornea with a wisp of cotton wool, avoiding the eyelashes in so doing. Monrad-Krohn has pointed out that loss of corneal reflex is often an early sign of a trigeminal affection, and indeed sometimes of the facial nerve in cerebello-pontine tumours, and is then ipsolateral.

If any injury has involved the trigeminal or facial nerve the corneal reflex can be made use of for diagnosis. If the trigeminal nerve is injured, touching the cornea is not followed by blinking of the lids on either side, but injury to the facial will abolish winking on the same side only, the contralateral escaping.

As a point of general interest and probably guidance to the reader, radiography has been called to our aid not only for the demonstration of fractures, but also in the form of arteriography for the revelation of the state of the blood vessels within the cranium following on head injuries. (Löhr.)

CHAPTER XVI

POISONS WHICH AFFECT VISION

MANY classifications have been made of those substances which affect vision, particularly those producing central scotomas. De Schweinitz has grouped those drugs which have a deleterious influence on vision and has made a comprehensive list in his "Toxic Amblyopias," while Casey Wood more briefly classifies poisons according to their direct action on the optic nerves, the production of a chronic retrobulbar neuritis and retinal disease. Uhthoff also provides us with a classification comprising those poisons, such as alcohol and tobacco, the consumption of which produces amblyopia of more or less severity, auto-intoxications from such conditions as diabetes mellitus, puerperal fever and pregnancy; and finally, chemicals, such as quinine, salicylic acid and lead. He also includes pellagra and snake poison.

Acetylcholine

This is a white hygroscopic crystalline powder with a saline, bitter taste and a characteristic odour. Its formula is

$$(CH_3)_3N(OH)CH_2.CH_2.O.CO.CH_3.$$ (B.P.C.)

This substance has already been mentioned in connection with the *humeral transmission of the nerve impulse* (see p. 37). Loewi and Navratil showed that vagus stimulation liberated a chemical agent in the frog's ventricle which can produce a vagus effect on another heart. The substance liberated is probably acetylcholine. Stimulation of the third cranial nerve leads to liberation of acetylcholine in the aqueous humour. Probably all parasympathetic nerves act similarly although Loewi found that physostigmine and ergotamine do not sensitize for the action of all vagus-like acting drugs, for example, not for that of choline and muscarine, but they do for that of the vagus substance and acetylcholine.

It is stated that acetylcholine is 100,000 times more active than choline on arterial pressure but only three times as toxic. Choline in small doses lowers the intraocular pressure, but in larger doses

raises both the intraocular pressure and the general blood pressure, but the former is out of all proportion to the latter. The extrinsic muscles of the eye contract under the influence of choline. (Duke-Elder.)

Dale suggests that nerves which liberate bodies resembling adrenaline and acetylcholine should be called respectively "adrenergic" and "cholinergic."

Acetylcholine is used in such diseases as arterial hypertension, Raynaud's disease, paralytic ileus, tobacco amblyopia and embolism of the central artery of the retina.

The effect of acetylcholine on the blood pressure resembles the effect of antidromic stimulation on the posterior nerve roots, *i.e.*, it dilates the limb blood vessels.

The dose of acetylcholine is 0·02 to 0·2 gram ($\frac{1}{3}$ to 3 gr.), dissolved in 5 ml. ($\frac{1}{6}$ oz.) of sterilised water. It is given subcutaneously or intramuscularly, but intravenous administration is dangerous.

Acetylcholine injected subcutaneously and intramuscularly in doses up to 0·5 gm. has no appreciable effect (Carmichael and Fraser), but intravenous injection of 0·03 gm. produces cardio-inhibitory effects with flushing of face, neck and upper thorax, intensified by eserine and abolished by previous injection of atropine. But Orr and Young have used acetylcholine in embolism of the retinal artery by subconjunctival injection and found atropine did not alter its action and no symptoms of collapse followed using 8 mins. (B.D.H.).

Alcohol

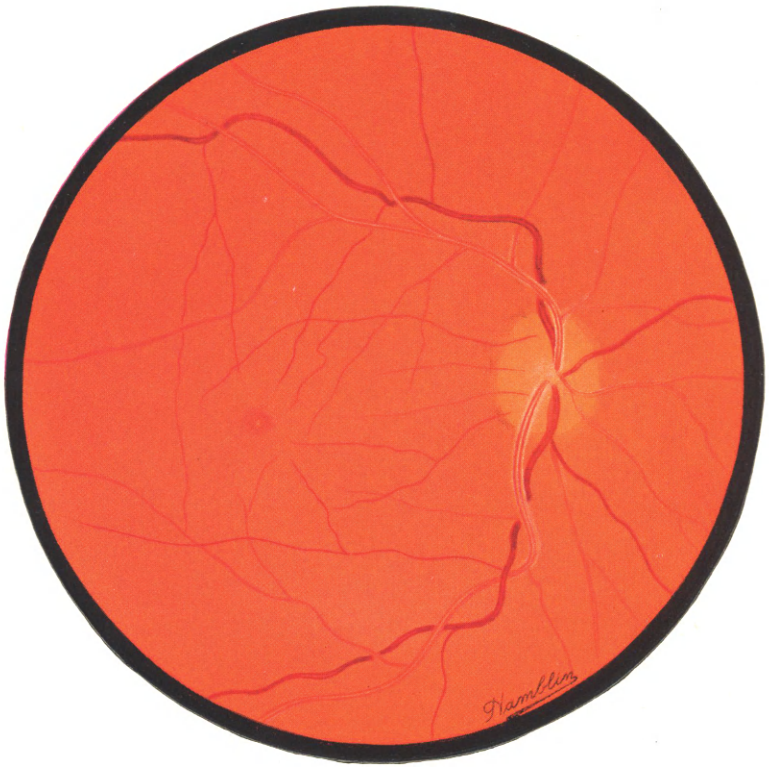
Acute Alcoholism.—That it is the nervous system which is chiefly affected as the result of consuming quantities of alcohol is clearly shown by the muscular inco-ordination which follows, together with mental disturbance and, finally, narcosis. A doctor may be called upon at any time to examine a patient suspected of having consumed more liquor than is deemed wise. Where exactly one should draw the line has never been determined ; what in one country is considered an amount which a gentleman should take would be looked upon with horror by members of a neighbouring state. The problem, "Why do certain men take alcohol in excess ?" is such a many-sided one that it is impossible to discuss the matter to any extent in such a book as this. I would refer my reader to Bernard Hart's contribution to the Mott Memorial—"The Ætiology

of Alcoholism "—in which he discusses this question. He freely quotes from Stockert, Trotter and others who have contributed to this subject. The author's simple advice to young men is avoid the habit of taking any beverage containing alcohol. From the writer's observance in the Great War he would emphatically state that the surest way of losing one's nerve is to pursue the constant habit of whisky drinking. In times of severe stress and strain such as war, mountain climbing, severe athletic contests, alcohol will always defeat the best endeavours. Nothing can so alter judgment and produce faulty ideas in action as the imbibing of alcohol in any quantity—large or small. At one time the Berlin police recommended chauffeurs of all classes of motor vehicles to become teetotallers. A report on the " Relation of Alcohol to Road Accidents " has recently been given by a special committee approved by the British Medical Association. It supports what is already well known, that alcohol depresses or even suspends the function of the brain, that in the performance of mental tasks the taking of whisky reduces the brain's efficiency, although, curiously enough while proving this, the subjects submitted to the tests by the various examiners were convinced that they had been doing these tasks better after taking whisky, and were surprised to find their records proved the contrary.

The French Academy of Medicine has passed a resolution urging the new French Government to resume the campaign against alcoholism, to limit the number of licences for the sale of alcoholic beverages, and to promote garden cities.

When driving a car the neuro-muscular co-ordination is in play all the time. Nowhere can the effects of drinking alcohol be more clearly seen than on the driver of a car who has indulged too freely—for example, when a car is driven round a gyration the opposite way to the normal, fouling several cars in its passage, the taking of blind crossings at 40 m.p.h., etc. I have already mentioned the method of measuring the muscle balance of the eyes with the Maddox rod, yet every oculist knows how impossible it is to get a constant result when examining a patient who is the worse for liquor. A slight muscle imbalance is exceedingly common, but this condition, in order to secure single vision, is overcome by many persons without conscious effort. Give alcohol, however, to such people and soon the power of accurate fusion is lost. Think of what this may mean to a driver, an aeroplane pilot and others whose lives and those

PLATE XVIII



ARTERIOSCLEROTIC RETINOPATHY

The arteries are brighter in appearance than normal, the branches of the artery leaving the disc have their walls considerably thickened and where an artery crosses a vein a kink in the latter is observed. Normally the vein when lying behind the artery is visible through its walls, but when the artery becomes thickened the vein is not visible through the vessel. This is the stage preceding what is commonly termed arteriosclerotic retinitis in which minute hæmorrhages and white spots of exudate make their appearance. (From E. Clarke's book, "The Fundus of the Human Eye.")

[*To face p.* 420.

of their fellow-men depend on their visual judgments. We often judge speed by the movements of the eyes and the amount of accommodation exerted. If a muscle imbalance is produced or aggravated by alcohol, how is a driver to acquire a quick mental appreciation of another car's movements ? Those who are in such a position that the lives of their fellow-men depend on their actions should avoid alcohol in any form.

There is one aspect of the effect of alcohol on the human body which apparently has received little attention. A young man, twenty years of age, may play the fool with alcohol, the effects of which are to set going arterio-sclerotic changes in his arteries. If at forty years of age he relinquishes the alcoholic habit, his arterio-sclerosis goes on just the same as if he had not stopped the habit, with the usual dire results. A man fifty years of age beginning to form the habit of spirit drinking also soon acquires arterio-sclerosis and it little matters whether he gives it up or not, the vascular disease has begun. The fundus of the eye is the best place in the human body where these changes can not only be seen, but their progress can also be followed (see Plate XVIII).

As a restorative, alcohol has not its equal. Exposure to cold and wet may not produce any harm if such a stimulant is taken immediately afterwards. However, in our present state of civilisation even the most gentlemanly of men may be found from time to time to have overstepped the mark of discretion in this respect. It is difficult when called, say, to a police station to diagnose drunkenness, when the prisoner is surrounded by earnest members of the Law endeavouring to find out his actual state of inebriation. Examination of the pupillary reactions will help. The pupils should be examined in a dark room. If the nervous system has been affected by an undue amount of alcohol the pupils are found to be dilated and do not react smartly to stimulation by light, also in some cases acute nystagmus is observed and the temperature of the patient is frequently below normal. A patient might be brought to a hospital by the police, suspected of being drunk, when in reality he may be dying from apoplexy. It is not an unusual occurrence for a person who has been drinking heavily to suffer from a stroke during his state of drunkenness. In this condition the coma is usually deeper, breathing is stertorous and there may be signs of hemiplegia, as evidenced by a greater degree of flaccidity of the limbs on one side. If the pupils in the precomatose state do not

react to light it is an indication for the Wassermann reaction to be done rather than an estimation of the extent of drunkenness. In the comatose state the size of the pupils may correspond to that of deep chloroform anæsthesia. It should be mentioned while referring to alcohol that patients who suffer from raised blood pressure should avoid this drug. Alcohol raises the blood pressure, and may do so to a dangerous degree, hence the reason for making the above statement that it is not uncommon for a drunken person to suffer from apoplexy.

Chronic Alcoholism.—The poisonous effects of alcohol are manifested first as a functional poison, as a poison of the tissues constituting the parenchymatous elements, particularly epithelium and nervous tissue, producing a slow degeneration, while the changes in the blood vessels are those of thickening due to fibrosis.

The state of the pupils in chronic alcoholism is varied ; they may be normal, sluggish, or simulating Argyll Robertson or even reversed Argyll Robertson reactions, but true Argyll Robertson pupils are never produced by alcohol alone. Vision may or may not be affected. Some writers have stated that they have never seen amblyopia caused by alcohol itself but always in association with the use of tobacco. However, amblyopia may result from chronic alcoholism alone, some of the worst cases being found in those who habitually make a practice of drinking methylated spirit, which is a mixture of alcohol 19 parts, and wood naphtha 1 part.

Pure alcohol amblyopia is rare and, indeed, its occurrence is denied, by some authorities. Neither Nettleship nor Marcus Gunn has ever seen a case of pure alcohol amblyopia. Those cases which have been recorded are not free from the suggestion that tobacco was the main factor in its production. Uhthoff quotes 6 cases, which were supposed to be free from the influence of tobacco. Each one of these suffered from delirium tremens, having been heavy drinkers and practically free from tobacco. The pathological investigation showed that the papillo-macular bundle was chiefly affected, but even when gray atrophy of the optic nerve was present, some normal fibres were found in the diseased area, whereas in tabetic atrophy normal fibres are not found in this situation.

From the researches of Lundsgaard and others we know that alcoholics acquire temporary or permanent night blindness. Those who are permanently night blind have had no preceding stage of

temporary night blindness. The field of vision for white or red is contracted also. Fig. 138 was kindly lent by Dr. Roenne, which illustrates the degeneration in the external corpus geniculatum following the degeneration of the papillomacular bundle in a typical case of amblyopia.

In the case of toxic (alcohol) amblyopia Uhthoff demonstrated cross-sections of the optic nerve which showed changes of an interstitial inflammatory nature. The process of proliferation in the nerve septa was shown by increasing density, cellular infiltration, proliferation of blood vessels and changes in their walls with proliferation of the glia.

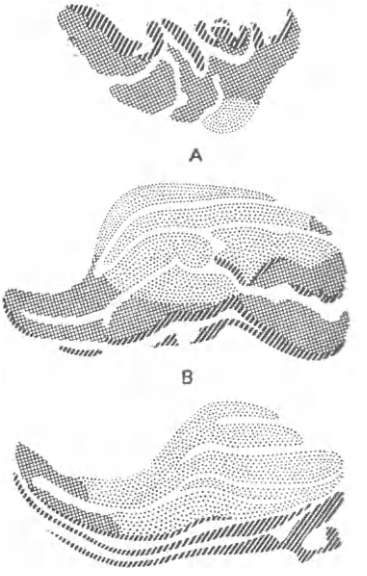


FIG. 138.—Frontal section of the geniculate body 1·8 mm., 3·1 mm. and 4·5 mm. respectively behind the beginning of the ganglion, the light dotted area in each section representing the degeneration of the ganglion cells following upon macular degeneration. (Roenne.)

The actual visible result in the retina of the eye produced by chronic alcoholism is a state of arterio-sclerosis. The arteries present a burnished copper-wire appearance; wherever an artery crosses a vein a kink in the latter is produced and in a more advanced degree fine hæmorrhages, flame shaped, are seen where the various vessels cross. A more advanced stage, known as arterio-sclerotic retinitis, shows hæmorrhages and fine white exudates scattered everywhere. The edge of the disc is blurred and its surface is raised, and finally, if the patient is still living, the optic disc becomes pale. A patient may live ten years after the onset of arterio-sclerotic retinitis, but swelling of the disc head with loss of distinctness of the disc edge is indeed an ominous sign. The worst example of this I have ever seen was in a middle-aged man who had become quite temperate during his recent years, but who as a young man placed no restraint on his consumption of alcohol. Within six months from examination he was dead.

The arterio-sclerosis seen in hard-working people who have been

life-long teetotallers presents a somewhat different appearance in the retina from the alcoholic variety. Examining the fundus of a young man in his thirties who was in the habit of consuming a bottle of whisky or more in a day, I could discern a slightly œdematous appearance over the retina particularly adjacent to the blood vessels, an appearance which is absent in the former variety or what might be termed the "dry" form of arterio-sclerosis.

Three kinds of pathological changes of the retinal vessels are known (Friedenwald). In the first group the primary lesion appears to be a proliferation of the intima with the development of atheromatous plaques. Fibrosis of the media and adventitia follow on this change. The second group comprise the condition of arteriosclerosis characterised by hyaline lipoid infiltration of the intima and media of the vessels. Thirdly, there is obliterative endarteritis.

Among the various forms of hypertensive retinopathies Fischberg includes arterio-sclerotic retinopathy and describes it fully. It was Foster Moore who first drew the attention of the medical world to the retinal appearances in renal disease and differentiated them from those which occurred as the result of arteriosclerosis without obvious kidney disease or from diabetes.

The *mental condition* of chronic alcoholics has been described by Jackson and others. Korsakoff mentions one type as a psychosis polyneuritica and this symptom complex is known by his name. McAlpine has described a case of Korsakoff's syndrome with double sixth nerve palsy due to alcohol. It should never be forgotten that chronic alcoholism is one of the important elements in the strain which leads to mental breakdown. Both insanity and epilepsy may result directly from chronic drinking. The pathologies due to chronic excess in adults may be detected in children to whom alcohol in the shape of wine and beer has constantly been given. Obarrio quotes a girl of five with hepatic enlargement and impaired tendon reflexes, a boy of five suffering from alcoholic polyneuritis and cirrhosis, while in all his cases he has found dreamlike hallucinations.

Alcoholic polyneuritis has been regarded by Strauss as similar to the polyneuritis of beri-beri, and he has treated the condition by giving yeast extract which contains the vitamin B complex. He used Marmite.

Stockert's experimental work shows clearly that alcoholic parents produce far fewer offspring than the normal as these for the most part are weak and may die before or shortly after birth, but he

found that those which survived were stronger and healthier than the average. The suggestion is that alcohol may be one method of eliminating the weak, allowing only the strong to survive.

Methyl-Alcohol Poisoning (Wood-Alcohol Blindness).—This is a form of poisoning which has commonly been found both in America and Germany. Many reports have been made of cases of severe poisoning (often leading to death) as the result of convivial gatherings where the liquid taken has been strongly fortified by methyl-alcohol.

In cases where wood-alcohol has been taken, either by accident or design, the severest toxic symptoms follow, such as intense headache, vomiting, abdominal pain, slow and weak pulse, delirium, convulsions, stupor and death. In cases that have recovered, loss of sight may supervene. Ziegler states that sudden blindness with vomiting and abdominal pain should always arouse the suspicion of methyl-alcohol poisoning, especially if diplopia or ptosis is associated. He draws attention to the fact that during the year 1914, in Great Britain, over 1,000,000 gallons of methyl-alcohol were used in varnishes. It can be easily understood that the presence of such a liquid in large amounts and of easy access may become a danger. It has been found that the commonest way of producing poisonous symptoms has been the taking of the alcohol by the mouth, or from fumes or by cutaneous absorption from toilet preparations or liniment.

The various conditions of the optic nerve head in cases of poisoning are described by Ziegler as (*a*) neuritis or papillitis, when the swelling of the disc may even measure 2 diopters ; (*b*) retrobulbar neuritis, with shrinking of the nerve head, exposing the lamina cribrosa at the bottom of the pit ; and (*c*) sudden sclerosis of the nerve head producing a chalky white appearance of the disc. The fields of vision show both peripheral contraction and scotomata in varying degree. It should be remembered when diagnosing the condition of wood-alcohol poisoning that part of the poison is excreted by the kidneys as formic acid, which reduces Fehling's solution and may therefore lead to the suspicion that the case is one of diabetes. Acidosis appears early.

The *prognosis* in cases of wood-alcohol poisoning is very grave, death often supervening in a few hours or days. Ball states that it is safe to say that those who recover remain blind for life with few exceptions.

Regarding the *treatment* of methyl-alcohol poisoning Ziegler says that as the greatest amount of the poison is found in the stomach

on the second or third day, therefore constant daily lavage of the stomach with alkaline solution can eliminate a great deal of it. The intestinal tract should be cleared by purgatives and profuse sweating be obtained. Negative galvanism should be employed to stimulate the optic nerve and restore its function before actual atrophy sets in.

Arsenic

Arsenic is such a widely distributed element that very few substances in their composition are without traces of it. In the past, various things, as wall-paper, etc., have been blamed for cases of poisoning, but Turner-Wilson showed that in such a case which had been reported in the newspapers, the wall-paper contained 0·00278 gr. of arsenic per square yard of wall-paper, whereas in Massachusetts, U.S.A., 1/10 gr. of arsenic per square yard is permitted, and in Holland 5 mg. per square metre is actually allowed, which is equivalent to 0·0645 gr. per square yard. He points out that in another case of arsenic poisoning attributable to wall-paper, the analysis proved that this poor lady might have eaten 343 yards of the paper, 21 inches wide, with impunity so far as its arsenical content was concerned.

Apparently an immunity can be developed against arsenic, such as is seen among the arsenic workers in the mines of Styria. In chorea and pernicious anæmia steadily increasing doses are often given until the patient takes from 15 to 20 drops of Fowler's solution thrice daily. Whitla quotes the work done by Besredka, who found that an antitoxin had formed in the blood-serum of animals fed on arsenic, which so stimulated phagocytosis that the drug is absorbed by the leucocytes and liver cells in which the arsenic may long remain stored up in a harmless state. When large doses of arsenious acid are taken in half an hour there is burning in the throat and stomach, rapidly followed by excessive vomiting, colic, diarrhœa, thirst and collapse. Death results from cardiac and respiratory failure. The more chronic form of arsenic poisoning produces œdema of the eyelids, especially the lower; there is severe irritation of the mucous membrane of the throat, nose and conjunctiva, also there is vomiting, diarrhœa and peripheral neuritis, the legs being attacked first, with deep pigmentation of the skin on the trunk and lower limbs and keratosis which is rarely followed by epithelioma. The arsenic may be largely recovered from the hair and nails. The introduction of arsenical preparations for the

cure of syphilis has drawn attention to the effect of the drug on the organ of vision. Some of these are apparently safe, yet in one preparation to which neurologists are turning their attention, namely, tryparsamide, which is found to influence both the cerebral and spinal syphilis more than novarsenobillon, there lies the possibility of damage to the optic nerve. It is a remarkable fact that a previous course of treatment with the novarsenobillon seems to enhance the effect produced by the subsequent use of tryparsamide in the cure of syphilis. In connection with this latter drug, it is stated that visual impairment is seen in 3 to 10 per cent. of cases (Lees), but Johnson-Abrahams tells me he has repeatedly seen cases of lowered vision caused by various arsenical substances, but the recovery of sight is usual. Probably 1 per cent. show primary optic atrophy. Tryparsamide is therefore safer to use in the case of general paralysis where primary optic atrophy is less common than in tabes. The author, when asked his opinion as to whether tryparsamide should be used in a case of syphilis, examines the optic nerve carefully for any sign of atrophy, including the visual fields. If the slightest evidence exists of the presence of degeneration of the nerve, the advice given is to use another form such as N.A.B. or the original " 606."

It is interesting to know that an attack of herpes ophthalmicus has followed the use of arsenic. Nystagmus has been observed in chronic arsenic poisoning, also optic neuritis and optic atrophy have been seen. There is a singular difference in the visual symptoms produced by the various forms of arsenic compounds ; the inorganic compounds may have little or no effect on the field of vision (Schirmer). When they do, it is a central scotoma that is found, while the organic forms such as tryparsamide and atoxyl produce a marked change in the nerve elements, resulting in a peripheral contraction of the field of vision, beginning on the nasal side. The former tend to recover, but in the case of the latter, once the destructive processes in the nerve have begun, there is no recovery and complete blindness finally ensues.

The *treatment of chronic arsenical poisoning* consists in giving daily intravenous injections of 0·6 grams of sodium thiosulphate dissolved in 10 c.c. of sterile distilled water in order that a harmless compound containing the arsenic should be formed in the body. In acute arsenical poisoning antidotum arsenum should be administered. This is a mixture containing a strong solution of ferric

chloride 30 ml., light magnesium oxide 10 grams in distilled water. The dose is 4 fluid ounces. After giving this mixture the stomach is subsequently emptied, stimulants given, together with the application of warmth (Whitla).

Atropine

Atropine is *dl*-hyoscyamine, an alkaloid obtained by the racemisation of *l*-hyoscyamine extracted from *Atropa Belladonna* Linn., *Hyoscyanus muticus* Linn., and other solanaceous plants (B.P.C.). When the crystals are dissolved in water it gives an alkaline solution. Atropine sulphate is the sulphate of this alkaloid.

Atropine is extensively used in medicine. It paralyses the terminations of the parasympathetic nerves which supply plain muscle, the heart and glands. It thus diminishes the secretion of gastric juice, tears, saliva and sweat. Atropine raises the lysozymic action of the tears (Ridley) and thus it can be of great help in treating some of the severer acute and chronic forms of conjunctivitis. Atropine acts as a powerful stimulant to the central nervous system, especially the motor area, thereby affecting co-ordinated movements. Small doses cause dryness and redness of the mouth, dilated pupils and disturbance of vision. Larger doses are followed by active brain excitement, restlessness, delirium and talkativeness. Atropine raises the arterial tension at first, but later the tension falls : by its depressant action on the vagus the heartbeat is quickened, and it is thus of use in cases of poisoning by pilocarpine and muscarin where the heart is slowed down. Atropine relieves bowel colic and is therefore of the greatest value in lead colic and colon spasm.

But it is in ophthalmology that this drug is of primary importance. Instilled into the conjunctival sac a drop of 1 per cent. atropine sulphate produces dilatation of the pupil which dilatation begins to show itself in twenty minutes or so. This mydriatic action lasts from seven to ten days, sometimes much longer. For the purposes of refraction examination in children of twelve years of age and under, a drop of $\frac{1}{2}$ per cent. atropine sulphate should be applied three times over the previous eighteen hours. The accommodation is also paralysed, but this action is not so lasting as the effect on the pupil. Dilatation of the pupil is brought about by the paralysing action of atropine on the myo-neural junction between the nerve endings of the oculo-motor nerve and the sphincter muscle. However, in 1926, Loewi and Navratil showed that the action of

atropine on the pupil is brought about by inhibiting the vagus substance (which resembles acetylcholine) liberated by the parasympathetic nerve endings. These experimenters found by the use of physostigmine and ergotamine pre-treatment that the heart is sensitised not only for the effect of vagus stimulation, but also for the vagus substance. They conclude by saying that the sensitisation by the above-mentioned alkaloids may be regarded as a consequence of the diminishing of the esterase effect caused by them.

Acetylcholine stimulates the parasympathetic nerve endings in the iris, an effect that is abolished by atropine (see p. 37), so that the constrictor effect produced by the parasympathetic nerves is abolished by atropine, and, therefore, dilatation of the pupil takes place. Atropine raises the tension of an eye when the corneo-iridic angle is smaller than normal which prevents the normal rate of the exit of the intraocular fluid. (Colle and Duke-Elder found in all their experiments that atropine produced a fall in the intraocular pressure in the normal eye.) Hence the great danger of instilling atropine when the intraocular tension is already raised by bringing on an attack of glaucoma. Instead of using atropine a solution of homatropine hydrobromide can be used which dilates the pupil in ten to twenty minutes, the effect both on the pupil and accommodation lasting about twenty-four hours. Intraocular tension is also raised, but not so markedly as with atropine and the effect of the homatropine is quickly neutralised by instilling eserine salicylate. After using homatropine, and at the completion of a fundus or refraction examination, $\frac{1}{4}$ or $\frac{1}{2}$ per cent. eserine will usually reduce the size of the pupillary aperture in half an hour. If raised intraocular tension is suspected then 1 per cent. eserine should be used. Eserine has little effect on a pupil dilated by atropine.

Atropine poisoning is sometimes seen when children mistakenly have eaten belladonna fruits. The poison being excreted by the kidneys, the bladder should be emptied by the catheter to prevent reabsorption. According to the B.P.C., in cases of poisoning 1·2 grams (20 gr.) of tannic acid should be given in 120 ml. (4 fluid ounces) of water and the stomach then evacuated by the syphon tube or by emetics. Caffein should be used to combat the depression which follows the preceding excitement.

That atropine disturbs the heat-regulating centre in the hypothalamus has been shown by Finkelman and Stephens, who measured the increase in oxygen consumption and the ability to

maintain the normal temperature on exposure to cold in patients with chronic epidemic encephalitis as compared with normal subjects. The greatest loss in temperature on exposure to cold was found in cases of encephalitis which had been treated with atropine.

Carbon Monoxide Poisoning

Carbon monoxide, McNally says, is one of the most important poisons associated with human life and industry. Since the advent of the motor car, poisoning by this gas in garages has been of frequent

Percentage Saturation of the Blood with Carbon Monoxide and Corresponding Physiological Effects

Per cent. of Hæmoglobin in Combination with Carbon Monoxide.	Physiological Effect.
10	No appreciable effect except shortness of breath on vigorous muscular exertion.
20	No appreciable effect in most cases except short wind even on moderate exertion; slight headache in most cases.
30	Decided headache; irritable; easily fatigued; judgment disturbed.
40–50	Headache, confusion, collapse and fainting on exertion.
60–70	Unconsciousness; respiratory failure and death if exposure is long continued.
80	Rapidly fatal.
Over 80	Immediately fatal.

occurrence. Furnace workers and miners due to their calling are particularly liable to the effects of this gas poisoning. The most frequent cause of death by carbon monoxide has been the fumes from the internal combustion engine and the escape of gas, normally used for illuminating purposes, to bring about suicide or death by accident.

Carbon monoxide is odourless, colourless and tasteless. The victims of fire are often unconscious when the fire actually reaches them. I have seen eight bodies removed from a burning hotel. The faces showed no trace of terror or struggle, but the bodies were intensely red, due to the formation of carbon monoxide hæmo-

globin in the blood. Milroy illustrates the spectroscopic appearance of carboxyhæmoglobin, a method which is used as a test for carbon monoxide poisoning. The smaller the animal the quicker is it poisoned by carbon monoxide—hence the use of mice or small birds for the detection of the gas of a dangerous amount when present in fumes. On post-mortem examination all the tissues are found to be reddened, hæmorrhages are present in the pleura and intestinal mucosa, the blood is fluid and coagulates slowly. The colour of the blood produced by cyanide poisoning is also red, but not so lasting as that produced by carbon monoxide.

McNally's table of percentages shows the physiological effect of the various amounts on the system (see p. 436).

Symptoms.—Headache and vertigo are warnings that carbon monoxide is being inhaled, and usually fresh air is at once sought for. Mild poisoning will produce dilated pupils and central loss of vision, further poisoning will cause loss of vision and acute ophthalmoplegia, with rapid respirations leading to unconsciousness, convulsions and death. Haldane believes that it is due to lack of oxygen that such effects are brought about by carbon monoxide poisoning.

Those who continue to live may show various sequelæ. Grinker has seen a temporary acute chorea which completely disappeared in a few days, also he has described a Parkinsonian syndrome after poisoning which, however, gradually ended in death. Some of the sequelæ are herpes ophthalmicus, pemphigus, nystagmus, congested fundi and immobile pupils, coarse lateral movements of the eyes, severe psychotic symptoms, ataxia and polyneuritis. The late effects of carbon monoxide poisoning have rarely been noticed. Petersen says out of 1,400 cases only five or six have had serious late effects. In one case, a year after poisoning in a garage the patient was still tired, apathetic, very forgetful, hardly able to do a day's work although previously a very energetic man. Another case developed mental symptoms, which promised to become permanent.

The changes in the brain have been described by Strecker, Pollak, Rezek and many others. The outstanding constant pallidal necrosis is described by each. The anterior end of the globus pallidus which is supplied by the recurrent artery of Huebner is typically attacked. Symmetrical softening of the globus pallidus with degeneration in the arterial walls leading to the deposit of

lime salts even by the third day is found. The caudate nucleus and putamen in children are affected. Diffuse hæmorrhages in the pia and the cerebral white matter are found together with demyelination of the nerve fibres and axonal destruction.

The toxin acts primarily on the vascular system, causing vasomotor paralysis and vascular stasis (Grinker).

Treatment.—McNally believes the most effective method of treatment is to take the patient out into the open air, giving oxygen combined with 5 per cent. carbon dioxide under pressure until the cheeks are distended, at the same time inducing artificial respiration. The body meanwhile must be kept warm.

Cocaine

Cocainomania is less common still than morphine insanity. Patients exhibiting the former are emaciated, have a quick pulse, they suffer from a sensation as if foreign bodies were under the skin, sleeplessness, loss of memory and delirium. The hallucinations produced by this drug are of the Lilliputian order, being that of small objects. The habit produces rapid mental, moral and physical degeneration.

Cocainism is sometimes begun by the taking of cocaine snuff. I have seen a patient who came in almost every day asking for cocaine drops to ease his eyes from pain, but it was soon discovered that he was a cocaine addict and wished to get it in any form.

Of all local anæsthetics cocaine is easily the best. Since its introduction by Koeller in 1884 it has been of the highest value in ophthalmology. Cocaine dilates the pupil, although in a dark brown eye or that of the negro it is practically useless for this purpose. It slightly paralyses the accommodation and tends to lower the tension of the eye. But its instillation into the conjunctival sac lessens the reflex of winking and so exposes the corneal epithelium unduly. In cataract operations, when cocaine is used too freely the corneal epithelium is seen to desquamate ; indeed, to avoid such desquamation the cornea should be constantly moistened with saline during the various operations on the eye. Some patients have a marked idiosyncrasy towards cocaine. One patient of the author's has fainted after the instillation of 5-per cent. drops into one eye, and the same occurred a few days later upon the instillation of cocaine during the author's absence. If a patient suffers from any form of heart disease it is wise not to use more than 2-per cent. cocaine,

and that sparingly. This drug should never be injected beneath the conjunctiva, mucous membranes or skin, especially when such an effective and non-toxic anæsthetic as novocaine is so readily procurable.

Filix Mas (Male Fern).

Synonym : Aspidium.

Male fern consists of the rhizome and frond-bases of Dryopteris Filix Mas, a fern indigenous to Great Britain. It contains a yellow amorphous substance of an acid nature, termed filmarone, which decomposes into filicic acid and aspidinol.

Male fern is a tænicide, being a most reliable remedy against t. solium, t. mediocanellata and bothriocephalus. Its use, however, has often produced amblyopia, especially when given with castor oil, in which the active principles are readily soluble. Japanese observers have found on ophthalmoscopical examination contracted vessels of the fundus as in quinine poisoning. Optic atrophy has resulted from poisoning by filix mas. Parsons relates a case where a drachm of extract of male fern was ordered three times a day and was taken for ten days, followed by total optic atrophy in one eye with a much contracted field of vision in the other eye. Sidler-Huguenin in 1898 collected 78 cases of poisoning by male fern, 12 died, 18 became blind in both eyes, 13 in one eye, and 5 showed greatly reduced vision.

Iodoform

Iodoform has been used in the past for surgical dressings. Cases have been reported where its use was followed by central blindness which, however, is of short duration. As a dressing it acts by the liberation of iodine, which quickly forms protein compounds. Absorption with symptoms of iodism sometimes occurred, accompanied by excitement passing on to narcosis. The antidotes to iodine poisoning are large draughts of milk, starch or wheaten flour mucilage, followed by stimulants and warmth.

Lead

Lead poisoning is much less common now than in former times. Thirty to forty years ago admissions to hospital were not at all uncommon. These patients were found suffering from colic, wrist and shoulder drop, also with varying degrees of lowered vision.

Men occupied in the lead industries (such as plumbers, painters and glaziers) were chiefly affected. The lead was probably ingested by the mouth by men who had to mix the paint—a process which to-day is done by machinery. Legge has shown that in recent years, by improved methods of working, there has been a reduction of more than 50 per cent. of these cases. Drinking water which has been carried for a considerable distance in lead pipes was accountable for a number of cases, and there are cases even to-day resulting from this method of lead contamination. Two of these have lately been described by Wilfred Harris ; one was that of a man whose condition was diagnosed as duodenal ulcer, but wrist drop suddenly occurring in his butler led to the correct diagnosis. Findlay points out that in the case of lead poisoning in infancy the intake of lead may be from the lead preparations applied as soothing lotions to the mother's nipples, also certain face powders used in Japan contain lead, and this lead, absorbed by the skin, is excreted in the mother's milk. In older children symptoms may follow from handling painted toys, or playing about railings, such as the cases quoted by Gibson of Queensland, where 200 cases of lead poisoning were admitted to hospital, the source of the poison in these cases being traced to the very soluble lead carbonate in the white paint with which the railings around the houses were painted. The heat of the summer had converted the paintwork of the railing into fine powder, which easily became attached to the moist hands of the children playing around. The total number of cases of plumbism (Gibson states) admitted to the Hospital for Sick Children, Brisbane, between 1891 and 1905, was 200.

Lead poisoning, with predominantly gastro-intestinal symptoms, has been found among agricultural workers in Austria who partake freely of grape-juice from the press. Fermenting must dissolves lead easily so that the U-tubes connecting the press and vat and lead-glazed tankards are responsible for the formation of lead sulphide (Duy).

Another source of lead poisoning is to be found in tobacco factories where zinc plates are used on which the cigar makers cut the tobacco (Jordans).

Roentgenology comes to our assistance in the diagnosis of lead poisoning in children, for Park has shown that in these cases the deposition of lead at the end of the diaphysis of the long bones casts a shadow as of a dense band. Rogers recently published

illustrations of such cases. The outline of the bone is unchanged, the epiphysis is normal but the diaphyseal end shows a band of increased density which varies with the amount of lead absorbed.

Lead encephalopathy occurred in 45 of the 89 cases of children recorded by McKhann and Vogt. This is the most serious of the acute manifestations of poisoning in children. Papillœdema, indicating raised intracranial pressure due to acute cerebral œdema, vomiting, headache with a bulging fontanelle, followed by delirium, stupor or coma, may be seen. There are paralyses of the ocular or limb muscles with head retraction. The prognosis of these cases is serious.

A late sequel of lead poisoning in children may be chronic interstitial nephritis during adolescence. This has been described by Nye following on Gibson and Turner's researches into lead poisoning in children.

Calcium salts have proved valuable as a means of bringing about rapid deposition of lead in the bones, but in children an increased calcium intake should be accompanied by vitamin D in order that the calcium-phosphorus balance is not upset, causing an increased loss of both (Shipley). Recently, Irving Gray has published his method of dealing with plumbism in children. He noticed that a diet low in calcium with the addition of phosphoric acid definitely causes an increased excretion of lead. He gave a high phosphoric, high calorie diet, with sufficient vitamin content with which he improved the general condition of the patient while the rate of excretion of lead was maintained. This treatment must be repeated (he says) if continued excretion of abnormal amounts of lead is found in the urine and fæces. During this increased lead excretion there are no acute toxic symptoms, so that there is no parallel between the absorption and excretion of lead and the toxic manifestations.

It is probable, as Rogers has pointed out, that lead is conveyed in the blood as colloidal lead phosphate which at the normal pH of the blood is very stable, but when slight acidic changes take place it is easily converted into the more soluble di-lead salt. Acids and acid-forming salts will cause an excretion of lead in these cases, which is accentuated if the diet is poor in calcium.

Ninety-five per cent. of the lead is stored in the bones as insoluble phosphate (Aub), which may be removed and excreted by the use

of parathyroid extract (Hunter and Aub). They have advocated the use of Collip's parathyroid extract.

The possibility of diachylon poisoning should be borne in mind when a woman of child-bearing age, who is anæmic and in whom there is a history of amenorrhœa, complains, during a consultation, of severe headache and vomiting.

Reznikoff and Aub, in their "Experimental Studies of Lead Palsy," came to the conclusion that

(1) Experiments with isolated nerve-muscle preparations from frogs have shown that the onset of fatigue in muscles which have been exposed to lead is much more rapid and complete than normal.

(2) A change in the permeability of the surface of muscle cell after exposure to lead is evidenced by an increased diffusion of inorganic phosphate from the muscle into the surrounding Ringer's solution.

(3) As far as can be seen from the response of muscle to nerve stimulation, lead salts seem to have no deleterious action on the conductivity of the nerve of an isolated nerve-muscle preparation from a frog.

(4) Such experiments indicate that the physiological lesion of lead palsy is in the muscle itself and that the muscles which are fatigued are most susceptible to lead paralysis.

The palsy produced by lead poisoning begins in the upper extremities and picks out certain muscles in such a manner that it seems most probable that it is the ganglia and the anterior horns of the cord which are affected, and not the peripheral nerves. Also the ganglion cells of the cortex are involved, producing chronic cerebral symptoms and general progressive muscular atrophy. In acute encephalopathy the lead poison has produced primary endarteritis with hæmorrhages and softening. In chronic encephalopathy there may be constant headaches with gradual mental changes. In both conditions papillœdema may be present and may have even proceeded to primary or consecutive optic atrophy producing blindness, together with certain cranial nerve palsies.

When diagnosing diachylon poisoning the blue line on the gums should be looked for, while examination of the blood shows marked basophilia with stippling of the red cells. There is severe anæmia present, also uræmia and nephritis. Porritt draws attention to a little-known, slow, subtle, insidious saturation of the system, producing symptoms of lethargy and weariness of mind and body with constipation. A change in air, or rather a change in water supply, has beneficial results. It has been pointed out that the so-called brass and bronze poisoning is really lead poisoning.

The author can remember a man being examined by Treacher Collins who showed a distinct papillœdema in each eye without any other sign of lead poisoning, yet he admitted that he was a lead worker. Such cases, however, are uncommon. The reverse holds, for often cases present themselves showing wrist drop, etc., but it may be a year later before optic neuritis is discerned. Gibson found in his cases that there was severe swelling of the disc, often to the extent of 5 or 6 diopters. He never found optic neuritis present without an external rectus muscle paralysis or paresis being present also, rarely the internal rectus, but in his most severe cases all the external muscles were paralysed, the balls were stationary and the ciliary muscles were paralysed also. He says, however, he has not seen ptosis among these cases.

If a child is brought to the consulting room suffering from a recently acquired internal squint, due to paralysis of one or both external recti, and if the presence of a swollen disc is discovered, the diagnosis may provisionally be taken as lead poisoning (Gibson).

The effect on sight is that a transient amblyopia may be produced or a permanent amblyopia may follow. In either case the fundus may show little or no change. Optic neuritis may be present, and is due either to the action of the lead on the nerve tissue itself or to cerebral or nephritic changes. In addition to an optic neuritis there may be a retinitis present, especially in chronic cases, this retinitis being similar to albuminuric retinitis. The saturnine variety, however, may be recognised by the history of the case, the blue line of gums, wrist drop, etc., and the presence of lead in the urine.

Hypertensive neuro-retinopathy, the name given by Fischberg for the condition formerly known and termed albuminuric retinitis by Liebreich, may occur in acute lead poisoning without any evidence of renal disease as well as in chronic renal disease of plumbic etiology.

As in other forms of toxic amblyopia, lead may produce a bilateral central scotoma which may be relative or absolute. The amblyopia may be transient or permanent and in neither case may exhibit any change of the optic discs. The permanent amblyopia finally ends in optic atrophy with complete blindness. The peripheral vision in these cases is usually unaffected until a change in the optic nerve takes place. Consecutive and secondary optic atrophy as well as primary optic atrophy are found in lead poisoning, the first following papillœdema due to increased intra-

cranial pressure caused by lead, the second due to the local effect of lead on the optic nerve itself or the effect of lead on the vascular and renal systems, while the third may be caused by the local effect of the lead without any previous sign of inflammation. Retinitis occurs simulating the retinitis of Bright's disease, but most probably this state is really associated with the renal condition. Colour blindness associated with the amblyopia may be observed, also paralysis of the external muscles, chiefly the external rectus, which is supplied by the sixth cranial nerve. In Gibson's cases of complete paralysis of the extra- and intra-ocular muscles, all these ended in complete blindness, the optic discs showing post-neuritic atrophy. All his cases of extraocular paralysis recovered. Some of his chronic cases showed a consequent partial optic atrophy, giving the disc a pale appearance resulting in concentrically contracted fields of vision and lowered central vision.

Prognosis.—The prognosis as regards the eyes must depend on the general health of the patient. If cerebral or renal symptoms are present the prognosis is grave. Those cases which show symptoms of a central toxæmia of the optic nerve and not of a general atrophy of the nerve are the most hopeful.

It is useful to observe in the bicarbonate of lead poisoning that drinks containing sulphuric acid form insoluble lead sulphate, and also, potassium iodide aids in the excretion of lead from the system. When lead salts are accidentally swallowed, milk and white of egg should be given immediately to render the lead insoluble. The formation of lead albuminate forms a protective coating to the mucous membrane of the alimentary canal, preventing penetration of the metal. (See British Pharmaceutical Codex, p. 818.)

Morphine and Opium

Morphine and opium are practically synonymous as regards their pharmacological and therapeutic action (Whitla). These drugs check every secretion in the body except those of the skin and mammary glands. Moderate doses produce constriction of the pupils but large doses are followed by extreme miosis. It is admitted by physicians generally that when sleeplessness is caused by pain, opium is one of the most reliable hypnotics known : it may be used as an hypnotic in the delirium of fevers, acute inflammations, mania, melancholia and delirium tremens. Whitla says " morphine may be given for organic disease and its administration should tide

over the period during which attempts are made to cure the disease." It is well to remember that morphine, opium and heroin are poisons to the liver and kidneys, so that if there is evidence of suppression of the function of these organs then these drugs should be withheld.

Morphine is extremely dangerous when the intracranial pressure is high owing to its depressant action on the respiratory centre already labouring under difficulties.

It is remarkable that morphine is taken not for its pleasant sensations but rather to avoid the tortures of it being withheld. Comparatively speaking, insanity due to morphinism is rare as compared with that produced by alcohol, but then the cost in the former case is much higher than in the latter.

The ambulant treatment of morphinism has been tried lately by Koenig with considerable success. Twilight sleep is induced by the use of Pernocton (Riedel de Haen). Three injections at 8 a.m., 4 p.m., and midnight are given. Pernocton 2 c.c. intravenously and 2·2 c.c. intramuscularly are injected thrice daily. To avoid sweats, colic, diarrhœa, headache and vomiting, atropine ·002 gr. can be added to the intramuscular injection. Fluid food is given before each injection, and fruit juices and glucose in between. The patient must learn to enjoy nocturnal sleep which can be done by the use of simple sedatives.

Physostigmine

The alkaloid physostigmine obtained from the Calabar bean in the form of physostigmine salicylate or eserine salicylate is used extensively in ophthalmology. Its action is similar to eserine sulphate, but this form of the drug is more stable. It is twice as active as pilocarpine nitrate. These drugs by stimulation of the nerve endings in the gland cells increase secretions, namely, sweat, saliva, tears, gastric juice and pancreatic fluid. Instilled into the conjunctival sac eserine causes the pupil to contract quickly, reaching its maximum in thirty to fifty minutes. This is due to increase in parasympathetic activity. The 1 per cent. solution so commonly used in hospitals is too strong for ordinary purposes, it causes spasm of the accommodation which brings with it intense pain. According to Anderson pilocarpine stimulates directly the parasympathetic endings in the sphincter, but spasm of the accommodation does not follow. In glaucoma intraocular pressure is diminished by eserine and pilocarpine in spite of the fact that both dilate the minute

intraocular vessels and increase the permeability of their walls. By acting on the ciliary muscle the trabeculæ at the angle of the anterior chamber and the canal of Schlemm are pulled open by the scleral spur and so drainage of the intraocular fluid is promoted. But in the normal eye eserine raises the intraocular pressure (Colle and Duke-Elder).

Van Heuven mentions that occasionally the instillation of pilocarpine in chronic glaucoma is followed by lowered vision for one to one and a half hours afterwards. It occurs in people over fifty years of age, but not in young people. The same effect may occur with eserine. Although Hess considered it to be due to spasm of the capillaries or an opacity of the retina, Van Heuven was unable to confirm this on animal experiments. Using a Scheerer entoptoscope, an initial slowing down of the blood stream near the macula was followed by an enlargement of the area where no vessels are to be seen. (Maxwell's spot.) This is coincident with the onset and duration of reduced vision. The effect may be overcome by small doses of nitro-glycerine by mouth or prevented by the addition of cocaine to the pilocarpine. Poisoning by physostigmine is combated by the use of 0·2 per cent. potassium permanganate as a stomach wash together with the use of atropine and strychnine administered hypodermically.

Although physostigmine is not the slightest use in counteracting poisoning by atropine yet atropine is specifically used as an antidote in eserine poisoning.

Quinine

Just as certain individuals show an idiosyncrasy towards nicotine and other drugs so do many exhibit the same to quinine. The treatment of malaria by quinine has been followed in some instances by " cinchonism " and, indeed, among malarial subjects, quinine amblyopia is mostly found. It has been recorded that 2 grams given over thirty-eight hours has caused diminished vision. In South Africa quinine is injected intramuscularly as therapy and prophylaxis against malaria. Grinker relates a case where the drug was injected about the sciatic nerve causing severe sciatic degeneration. There are people who upon taking a moderate dose of the ammoniated tincture will suffer temporarily from diminished vision. Quinine to such will cause headache, tinnitus aurium with dimness of vision. The loss of sight, indeed, may be absolute and

may be accompanied by deafness, the latter usually preceding the former. The blindness affects both eyes and may last for days, some entirely recovering, while in others a permanent defect remains. Very rarely complete blindness has followed for two years (Claiborne). The pupils become widely dilated and inactive to light, the fundus of the eye seen by the ophthalmoscope at first resembles that of embolism of the central artery. There is marked ischæmia of the optic discs and retina with a white haze around the macula. In severe cases tubular vision (seeing as if through a tube) remains. It should be remembered therefore that quinine cannot be taken without due care being given as to the correct dose. Many people have recorded that after taking an indefinite amount in a single dose they have discovered a few hours later their vision had greatly diminished. They have also suffered such unpleasant symptoms known as " cinchonism," namely, ringing noises in the ears, deafness, diminished vision, sickness, headache, delirium, convulsions, paralysis, stupor and collapse.

The dose of quinine causing ocular symptoms shows startling variations. In this connection I would like to quote the following from " Ball's Modern Ophthalmology " :—

" (1) *Solubility.*—Whilst the acid hydrochloride dissolves in an equal quantity of water, the sulphate requires 800 parts to bring it into solution.

" (2) *The Vehicle.*—Solutions, especially spirituous ones, are absorbed quickly ; pills, tabloids and powders are slowly absorbed, or may be expelled speedily by vomiting or otherwise.

" (3) *The Method of Administration.*—Intravenous injection gives a quicker and greater concentration of the remedy than other methods.

" (4) *Idiosyncrasy of the Patient.*—This constitutes the greatest menace. In some cases a dose of 2 grains, given by the mouth, has caused the physician grave anxiety (Elliot).

" (5) *Age and Sex.*—Women and children are especially liable to quinine poisoning."

The symptoms of quinine-amaurosis are first, dilation of the pupils with loss of light reflex ; secondly, marked contraction of the retinal blood vessels, together with pallor of the fundus and discs ; thirdly, diminished vision, the visual fields showing contraction or rarely total blindness ; there may also be delirium and visual hallucinations.

In experimental work on dogs Holden and Birch-Hirschfeld have shown that the toxic effect is exerted chiefly upon the ganglion

cells of the retina, due probably to the contraction of the vessels, followed by an ascending atrophy of the optic nerve.

In most cases the blindness brought on by quinine disappears but not completely, permanent amblyopia and contracted visual fields remaining. With the return of vision the pupils recover their mobility. The ophthalmic picture in the late stage of quinine poisoning resembles optic atrophy, the disc remaining white and the retinal vessels narrow, accompanied in some cases by white lines resembling Plate IV, Fig. 2.

It is interesting to contrast the way in which vision is affected with what occurs in tobacco amblyopia ; in the latter there is loss of central vision with normal peripheral fields, but in quinine amblyopia, peripheral vision may be largely lost, while central vision is retained.

The treatment of quinine poisoning is to withhold any further preparation of cinchona. The poison can be eliminated by the bowels, kidney and skin. The following have been used to aid recovery—potassium iodide, amyl nitrate, hydrobromic acid, and strychnine. When vomiting follows an intramuscular injection of quinine, or when vomiting occurs for the first time Smith says this is an important sign of danger from quinine poisoning, while Elliot states that once quinine blindess has manifested itself there is always danger of a relapse after the use of doses of the drug, which formerly would have been harmless.

It will be of interest to know if poisonous symptoms follow the use of atebrin and plasmoquin (Bayer), which have been used recently instead of quinine in the treatment of malaria by the Indian Medical Service.

In September, 1935, a patient of the author's who is a missionary in Nigeria, West Coast of Africa, returned suffering from a marked muscle imbalance. In order to obtain binocular vision prisms had to be placed before him. He was suffering from an exophoria of 4 prism diopters and a right hypophoria of 9 prism diopters. He had suffered from malaria and been heavily dosed with quinine. For the three months he was at home in England the quinine was stopped. In February, 1936, his muscle balance was practically normal. The only conclusion to which one could come was that the muscle imbalance was due to excessive doses of quinine. The advice was given that on his return to Africa he should use atebrin. I received the following letter :—

"With reference to the use of atebrin, I discussed this on my return with Dr. Fraser of the Government hospital here. He has had some twelve years' experience in the Gold Coast. He said that they had not found atebrin to be an efficient prophylactic against malaria, therefore could only recommend quinine for regular prophylactic use, though he pointed out, the use of atebrin during a malarial attack had been proved of great value, and meant that quinine need not be used in more than 5-gr. doses, against the 15-gr. doses hitherto used during an attack. I found also that one of our own men had tried atebrin as a prophylactic, and after a few weeks had come down with a bad attack of malaria.

"Seeing that Messrs. Bayer & Co., the makers of atebrin, claim that it is an efficient prophylactic against malaria, I took the trouble to write them on this point. Their reply was that as all who had been in West Africa for some time had malarial parasites in their blood, a special initial treatment was necessary. This was: A five days' course of atebrin, followed by a pause of two to three days, then two plasmoquin tablets a day for four days. This, they claim, would rid the system of malarial parasites already present and following this course atebrin could safely be used as a prophylactic.

"I received this reply last month. Until then I had been taking the 5-gr. quinine hydrochloride daily. Then I took up the preliminary course as Messrs. Bayer advised and have now been on the atebrin for about one month. So far it has kept me free from malaria, though naturally a longer trial is necessary before I could say that it does prove an efficient prophylactic, and also before my eyes would benefit much from the dropping of quinine. A six months' period will be a better test as I continue with the atebrin."

Quinidine

Quinidine is an alkaloid isomeric with quinine and is derived from the bark of various species of cinchona. Its sulphate is used in the treatment of auricular fibrillation. Occasionally quinidine produces unpleasant symptoms, such as palpitation, headache, nausea, dizziness and lowered vision. It should be withheld when there is a history of embolism or where there is cardiac hypertrophy, especially enlargement of the left ventricle or heart block (see Osler and McCrae).

Optochin

Optochin or ethyl-hydro-cuprein has been used in the belief that it has a specific action on the pneumococcus. It is considerably more toxic to the visual neurons than quinine. Traquair says the field changes are similar to those of quinine poisoning but are distinguished by the relative frequency of a central scotoma without peripheral contraction of the field of vision.

Santonin, etc.

Yellow vision—Xanthopsia—is seen in *picric acid* and *amyl nitrite* poisoning, also on taking *santonin*. Occasionally santonin poisoning produces violent disturbance of the nervous system. Both dilation and inequality of the pupils may be observed.

Violet vision—ianthopia—has been seen in acute poisoning by *cannabis indica* or *hasheesh* (often sold in cigarette form), while blue—cyanopia—and green vision—chloropia—has been observed in *digitalis* poisoning.

The rapid spread of *marihuana* smoking, especially in America, calls attention to the use of cannabis indica in this form. Cannabis consists of the dried flowering and fruiting tips of the Cannabis sativa Linn. Marihuana is the Mexican name for the extract from the hemp plant and it has been known for centuries throughout the world by its Persian name of hasheesh.

Bromberg has written upon the menace of marihuana. The habit is rapidly spreading, beginning at seaports and also in the States adjacent to Mexico. In the last eight years it has increased greatly in the large cities all over the United States.

The intoxication caused by the smoking of marihuana is initiated by a period of anxiety and the fear of death which quickly gives way to a feeling of calm and ease. Soon the subject becomes talkative, elated, happy and laughs uncontrollably. He becomes brilliant but cannot impart his ideas. Addiction among marihuana smokers is unlike addiction among users of morphine or heroin. In the latter two drugs the tolerance developed in the body is a potent fact in addiction—the victim must have the drug to feel normal. With marihuana the addict wants to recapture the euphoric state into which the drug lifts him. It calls for sexual intercourse where heroin kills the sex desires. If the smoker leaves off, he has no craving for the drug. In addition to being sexually excited, he

may have visual hallucinations—he sees beautiful colours (*vide supra*). Excitement of great intensity with malicious and destructive tendencies appears during the acute intoxication. Possibly 40 per cent. of marihuana users are homosexuals. Bromberg concludes by saying, " the fundamental point is that the anti-social, sadistic and psychopathic elements in the make-up of the users of the drug are responsible for crime rather than any specific iniquitous property of marihuana."

The fumes of *bisulphide of carbon* and *nitrobenzol* have been responsible for many cases of lowered vision, either central scotoma or the reduction of the peripheral field. Stephenson has stated that complete or partial blindness has resulted from inhaling fumes of exploding dynamite or of nitrobenzol.

Poisoning by *metadinitrobenzine* has been recorded by Erskine of four sisters who suffered from methæmoglobinæmia due to absorption through the skin of this compound contained in furniture polish. The poisoning produced marked cyanosis of the skin and mucous membranes and in two of the cases methæmoglobin appeared in the blood. Apical systolic murmurs persisted for several days after the disappearance of the cyanosis. The respiratory depression was combated by the administration of carbon dioxide and oxygen.

Sodium Salicylate

Sodium salicylate is another drug towards which certain individuals show a marked idiosyncrasy, although everyone is liable to certain symptoms of an objectionable nature upon taking large doses of this drug. Thus resembling quinine, large doses will produce fullness of the head, buzzing in the ears and disturbance of vision. Upon further increasing the dose, deafness, squinting, sighing respiration, delirium, with dark albuminous urine, involuntary evacuations and convulsions. The changes in the field of vision are similar to those produced by quinine (Traquair), but the duration of the visual disturbance is shorter and the prognosis quite good. The drug probably acts on the cerebral centres for vision. There are no ophthalmic signs.

As sodium salicylate is used extensively in the treatment of acute rheumatism it would be well to guard against overdosage. A case has been reported by Snell in which a child was given 150 grains of the drug in sixty hours. The child became completely

blind and remained so until her death. Neither pupillary action nor fundus showed any change.

The drug acetylsalicylic acid is in common everyday use under the synonym "aspirin." It can easily be obtained and may therefore be used in the attempt at suicide. From what Neale and Dyke relate of their cases of aspirin poisoning we learn that the dangerous dose of aspirin varies considerably around 400 to 500 gr., the noxious action taking place not at once but after a few hours. Neale says, "that although gastro-intestinal irritation with associated vomiting may occur the leading clinical disturbance is connected with the nervous system, and in the untreated cases, reaches its crescendo in the course of twelve hours or more, when dissolution may take place as the result of profound cerebral depression."

The visceral action of the drug is considerably intensified by the associated dehydration, and it is therefore imperative to combat the depletion of tissue fluids and especially to retain adequate renal secretion. Excretion of the drug occurs in the sweat and the urine. The cerebro-spinal fluid contains the drug in considerable amounts and, therefore, lumbar puncture played a leading part in the recovery of two of Neale's cases. In 4 fatal cases lumbar puncture was not carried out. It is thus suggested that the combined therapeutic effect of the introduction of fluid to the body and the simultaneous aspiration of the cerebro-spinal fluid will be the means of saving an otherwise hopeless situation.

Strychnine

Strychnine is a drug which is used extensively both by ophthalmologists and neurologists. In the mouth it has the action of a bitter, increasing the appetite, but on absorption it exerts its characteristic effects upon the central nervous system—the reflexes are exaggerated and the normal tonus of striped muscle increased. The medulla and sense organs are stimulated, increasing the sense of sight, smell and hearing. It also raises the blood pressure. Upon a poisonous dose being given there is first a sensation of stiffness of the muscles of the body beginning in the neck and thorax and soon spreading to the back and limbs. Severe spasmodic seizures take place; these at first are brief, but later as the effect of the drug deepens they may last several minutes. The facial muscles, by their powerful contraction, produce the "sardonic grin." The cerebrum remains unaffected (Whitla), except by the result of

impeded respiration. The special senses are stimulated so that the field of vision is enlarged and the perception of shades of colour increased. The medulla is partly stimulated so that respiratory and vasomotor actions are considerably increased, but finally, due to failure of the respiratory centres, death ensues.

Strychnine is antagonistic to chloral and alcohol. In strychnine poisoning, urethane is used or intravenous injections of soluble barbitone.

Barlow suggests that the safe dose of pentobarbital for an adult man suffering from strychnine poisoning would be 1/10 grain per pound for the first dose and, should convulsions recur, half this amount for each succeeding dose. Great care should be taken if more than four injections are given during the first two or three hours.

Sulphonal and Barbitone

There are many drugs which are of the utmost importance in the treatment of disease, but the continuous or repeated use of some may produce harmful effects or an overdose may be followed by death. For instance, the drug, dinitro-o-cresol or dinitrophenol, which is used extensively in America for slimming purposes and increasing metabolism without the " side effects " of thyroid, has produced fatal results from apparent idiosyncrasy in certain people. Dermatoses of an unpleasant nature have occurred and in those taking the drug over a long period a very severe form of cataract has supervened.

The dangers associated with various drugs have been described by Sir William Wilcox and others. Take for example some of the sulphone group—sulphonal, trional and tetronal (much used in asylum practice)—the danger lies in their cumulative effect. Coma, mental depression, hallucinations, diplopia, squint, ataxia, drowsiness and stupor have followed their use.

We may also mention that after the prolonged use of the barbituric acid compounds there are actual degenerative changes in the nerve cells. The experimental work done by Dr. Pickworth establishes the pathological basis of the toxic symptoms shown by the nervous system. A patient aged thirty-five suffering from acute rheumatic fibrositis had been given 10 grains of medinal (sodium salt of barbitone) for six nights. The effect was a confused mental condition, bilateral ptosis, indistinct speech and diplopia on looking to the left (Wilcox).

The effect of most drugs taken on an empty stomach is more rapid and more intensive than taken after food. Idiosyncrasy may bring about abnormal results from normal doses, while allergic patients are in general abnormally susceptible. In the case of the barbiturates only a slight degree of tolerance to this group is established over prolonged use. Further, the physical condition of the patient should always be considered before these drugs are prescribed, the state of the renal system, the condition of the liver, the vascular system and the state of hyperthyroidism ; for example, nembutal should not be used as a basal anæsthetic in the last-named condition.

The toxic effects of the barbitone group of drugs on the central nervous system, due either to idiosyncrasy or prolonged use, have been as follows : drowsiness, mental depression, vertigo, ataxy, visual hallucinations, thick speech, diplopia, squint, nystagmus, paralysis of the face, tongue and limbs with tremor of the hands. In prescribing hypnotics, Wilcox advises the use of well-known drugs less toxic than the sulphonal or barbitone groups, such as the inorganic bromides, phenazone and its derivatives, aceto-salicylic acid, phenacetin, etc. These combined with opium preparations or codeine are generally effective. So also are urethane and somnosol mild and safe hypnotics, the former being suitable for children.

Barbitone poisoning which is generally the cumulative result of taking these drugs should be combated by washing out the stomach with warm water, strychnine in full doses, adrenaline or pituitary (posterior lobe) extract injected hypodermically. Food should be given by stomach tube if the coma is prolonged, dextrose-saline by the rectum and, if necessary, to remove the poison from the central nervous system, lumbar puncture should be done.

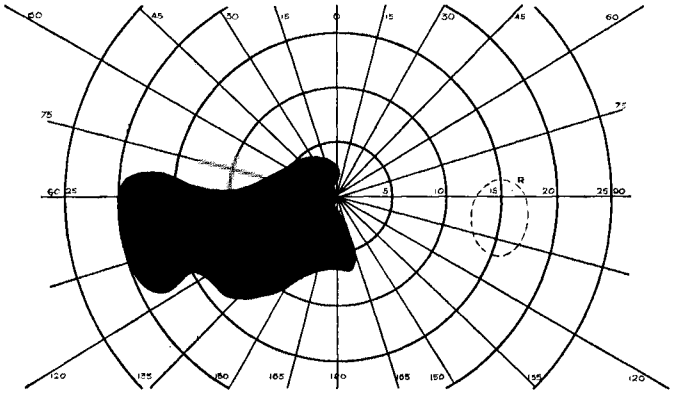
Tobacco

The commonest and most universal form of toxic amblyopia is that caused by smoking. As far back as 1832 McKenzie wrote in his text-book on " Diseases of the Eye," " A majority of the amaurotic patients by whom I have been consulted have been in the habit of chewing and still oftener of smoking tobacco in large quantities." But there are other means whereby nicotine enters the system. It is said to occur in tobacco factories among workers who neither smoke nor chew, no doubt caused by the inhalation of tobacco-laden dust by the workers. It has also been caused by snuff taking.

Many cases occur where both tobacco and alcohol are used, so that in spite of Jonathan Hutchinson's theory that alcohol counteracts tobacco, the worst forms of amblyopia are found in these cases. There is not the slightest doubt but that tobacco used either in pipe or cigarette smoking without the use of alcohol or any other drug can produce greatly lowered vision due to the presence of a marked central scotoma. Why certain people out of all the myriads of smokers should suffer from amblyopia it is impossible to tell, although we know that a lowered state of health may partly account for an attack. It is often seen in men who are greatly worried about business affairs or in those who are confined for most part of the day lacking fresh air and exercise. Men, such as navvies, who have to proceed to their work early in the morning and who smoke heavy shag tobacco on an empty stomach commonly suffer from this disease. It has always appeared to the writer that the amblyopia depends upon the quantity of tobacco smoked and not on whether a pipe or cigarette is used. In Usher and Elderton's cases (1,100 patients) the average amount of tobacco smoked was $2\frac{4}{5}$ oz. per week, but 21 of their patients smoked less than 1 oz. per week. Unfortunately, many women now smoke, and among such I have seen cases of tobacco amblyopia. In women it occurs at a much earlier age than in men. Those I have seen were in their twenties or early thirties, whereas in men it is not common under forty-five years of age—a fact which seems to indicate that a woman's nervous system cannot withstand the effects of smoking as well as that of the male. The habit now pursued by so many women is most unfortunate for one reason alone, they find the habit much more difficult to forgo than do men. When it is so commonly known that nicotine is a definite nerve poison, would it not be well for women who are pregnant to reflect on this so that they might give their offspring the best possible chance of enjoying a full state of health by the omission of such a habit as smoking during this period ? I do not hesitate to say that a pregnant woman should neither take alcohol nor smoke (see paper by A. M. Campbell).

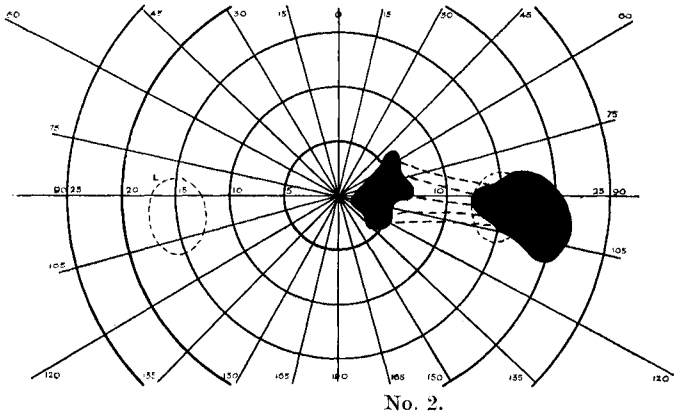
It has been stated that neither a pure alcohol amblyopia nor a nicotine amblyopia has yet been seen, but that always both are combined in greater or lesser degree. Connor, however, has described 27 cases of pure tobacco amblyopia, also 2 cases of pure alcohol amblyopia.

Tobacco blindness is seen in young men as early as twenty years



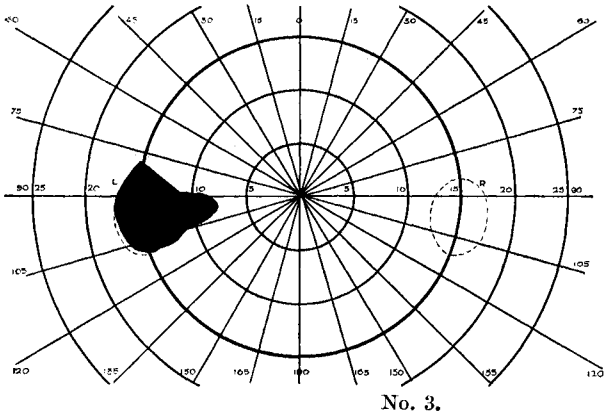
No. 1. — A large scotoma involving the blind spot and the area between the blind spot and fixation, but which stops just short of the actual fixation point.

No. 1.



No. 2.—A scotoma lying close to fixation within the 5° circle, connected by a relative area to the blind spot, which may or may not be prolonged towards fixation.

No. 2.



No. 3. — A scotomatous finger pointing from the blind spot towards fixation.

No. 3.

FIG. 139.—Charts showing three types of scotomata due to tobacco amblyopia. (Doyne.)

of age who smoke while at work in confined rooms. Some of the worst cases the writer has treated were produced by cigar smoking in tropical regions. The scotoma at first may be for form only; perhaps the patient cannot see the lowest two or three lines of the test types while his colour sense is scarcely affected. Later there is a definite scotoma for red and green, while finally, the scotoma may be absolute. De Schweinitz has described blindness occurring in horses in New South Wales, due most probably to the ingestion of the leaves of the Australian tobacco plant, although he mentions that the Virginian deer eat the leaves of the tobacco plant without harm. If smoking is persisted in, in spite of amblyopia being present, primary optic atrophy may ensue. I have seen one definite case of this kind.

Undoubtedly nicotine alone is not the only cause of the central blindness. It is the accumulation of the various poisons which in the end affects the finest divisions of the optic nerve, namely, the papillo-macular bundle. It is not due to their selective action on the ganglion cells of the retina, otherwise tobacco amblyopia would not so continually manifest itself as a central scotoma. Nicotine has a specially selective action as a nerve poison. Cardiologists are aware that smoking can affect the efficiency of the heart producing in the end an extremely lowered systolic pressure, while other vague nervous manifestations seen, especially in women, can be attributed to the habit of smoking—the proof of which is the complete recovery from such conditions on the cessation from smoking. In summing up the evidence of many authors de Schweinitz states that in cases of toxic amblyopia there is an interstitial sclerosing inflammation in the papillo-macular bundle, yet Duggan, who gives a most complete list of all the latest works on the subject of tobacco amblyopia, says that he is convinced that this condition is brought about by vascular spasm in the visual pathway and that the condition should be treated with nitroscleran, which is a vasodilator. Imre told me how he was using amyl nitrite for certain retinal conditions in which he believed there was an element of vascular spasm. As more rapid recovery takes place in those patients who also consume alcohol than in teetotallers it is thought this is due to the dilator action of alcohol. Wright and Moffat, as the result of their experiments, came to the conclusion that smoking produced certain definite pharmacologic effects, such as a marked drop in surface temperature—especially of the extremi-

ties, slowing and stoppage of the blood flow in the capillaries of the nail folds, together with other toxic effects such as anginoid attacks.

Cases suffering from tobacco amblyopia often complain of severe cough, dyspepsia and loss of appetite. This, however, may be due to substances contained in the tobacco other than nicotine. Tobacco tar is deposited from the smoke on the mucous membranes and is liable to set up some form of irritation. Pyridine and ammonia are also liberated during smoking and these also are irritants. Smokers' cough is a common complaint, 100 per cent. cures being effected by breaking off the habit of smoking. The pyridine in large doses is a respiratory paralysant.

Tobacco contains from 1 to 8 per cent. nicotine, and when smoked the nicotine and other substances such as pyrrolidine and resin are partly converted into pyridine, collidine, furfurol, hydrocyanic acid and carbon monoxide. In cigarette smoking carbon monoxide is freely formed, and if inhaled is absorbed by the hæmoglobin, so that a smoker of twenty-five cigarettes per diem may have 5 per cent. of his hæmoglobin temporarily thrown out of use (B.P.C.). Cigar smoking to those unaccustomed to it will raise the blood pressure as much as 40 mm. of mercury, which is rapidly followed by collapse, the pressure falling as much as 50 mm. of mercury. Neuman-Wender, writing in the *Muenchener medizinische Wochenschrift*, recently stated that in an extensive study of the effects of tobacco they found that in addition to nicotine many other harmful products are produced, such as tar, methyl-alcohol, etc. They think the methyl-alcohol may be largely responsible for the depression in vision suffered by smokers. They say a cigarette smoker of twenty per diem inhales about 40 mg. methyl-alcohol.

I had just read " The Nicotine Fallacy " by Green, when accidentally I came across the report by Guenkin and Brown on their examination of 41 male and 14 female factory hands engaged in the extraction of nicotine from tobacco leaves and in the preparation of nicotine sulphate. Nicotine poisoning exhibited itself among those workers and was mainly evidenced as disorders of the autonomous nervous system. Some were traceable to stimulation of the vagus : to these belonged the slow and often irregular pulse, gastric hyperacidity, spasms of the non-striated muscle, salivation and sweating. Cerebral symptoms were caused by the direct action of nicotine on the

central nervous system which resulted in disturbed sleep, weakening of the memory and neurotic symptoms (see Corti).

The scotoma which appears in the field of vision is always bilateral, not quite central, being typically in the centro-cæcal position, *i.e.*, involving both the point of fixation and the blind spot. Doyne has described and illustrated this type of field. The defect for colour exceeds that for white. One rapidly tests for tobacco amblyopia by holding small red or green objects before the tip of the nose, asking the patient not to take his eye from this point. The amblyopic patient cannot recognise the colour, but by moving the coloured object slightly above or below the nose he immediately recognises the colour. Care must be taken not to confuse the centro-cæcal scotoma due to pituitary tumour with that due to tobacco amblyopia. In the former the scotoma is more sharply defined and there is usually an early hemianopic defect becoming evident, while central vision is not so depressed as in tobacco amblyopia (Traquair). Typical tobacco scotomata are shown on Fig. 139.

Ophthalmoscopically, there is no departure from the normal fundus seen except in the most advanced cases, when slight pallor of the temporal side of the disc may be observed. Rarely complete optic atrophy is seen.

The investigations of Rönne in a case of diabetes associated with tobacco amblyopia support the work of Widmark and Dalen ; the former had examined histologically the degenerated macular bundle through the optic tract into the external geniculate body, while the latter in a case of alcohol-tobacco amblyopia made a similar investigation. Fig. 138, which was kindly lent by Professor Rönne, shows the degeneration found in the external geniculate body in the case which he investigated. The three illustrations represent sections at different depths, the light dotted area in each section representing the degeneration of the ganglion cells following upon a macular degeneration. This throws considerable light on the question of the pathology of alcohol-tobacco amblyopia. During life the case described by Rönne showed visual fields with red and green blindness and central scotoma for form.

Winckler has described a non-syphilitic condition closely resembling tabes dorsalis which he calls "pseudo-tabes nicotiniana" ; it is always associated with visual symptoms. His patients were over fifty years of age ; they were undernourished individuals

with a history of excessive smoking for some twenty years. He found absent knee jerks, pin-point pupils irresponsive to light, attacks of giddiness with loss of memory and visual disturbances. He lays emphasis on the fact that the prognosis is very good if the patient gives up tobacco entirely. He considers that the tobacco poison disturbs the fibres in the posterior columns of the spinal cord, while syphilis destroys them. He prescribes the use of sulphur and mud baths with complete abstention from tobacco.

Treatment.—Experience has shown that it is not the slightest use merely cutting down the amount of tobacco consumed but there must be entire cessation of the habit. Treacher Collins was most insistent upon this point. He used to say a single pipeful a day will still maintain the effects of the poison. It is remarkable to see with what regularity a patient suffering from tobacco amblyopia recovers his vision, usually one line of the test type per month, until full vision has been restored. Yet it must be remembered that there are cases where the habit has not been checked in time and permanent damage has been done to the optic nerves, especially the papillo-macular fibres, so that it is impossible in these cases to obtain a complete recovery. The author makes it a habit to ask patients suffering from tobacco amblyopia not on any account to smoke; at the same time he prescribes strychnine, laxatives and fresh air. Percival, reading in *La Semaine Medicale* of the use of expressed juice of watercress as an antidote to lethal doses of nicotine in guinea-pigs, freely prescribes the consumption of watercress in addition to the above treatment. Duggan used from six to ten intravenous injections of from 40 to 100 mg. of sodium nitrite (as nitroscleran), which, he says, considerably shortens the period of recovery.

The composition of nitroscleran (Tosse) is: sodium chloride 0·6 per cent., di-sodium phosphate 0·36 per cent., di-potassium phosphate 0·2 per cent. and sodium nitrite 10 per cent.

In July, 1936, Campbell Orr reported his results of treating several cases of tobacco amblyopia with acetylcholine. The first case reacted in a phenomenal manner. The vision of each eye rose from 1/60 to 4/60 after an intramuscular injection of 0·1 gram of acetylcholine the previous day. Following this 0·1 gram was given daily for six weeks, when the vision was found to be 6/9 with restoration of both colour and form fields. Hearing that J. B. Fenton was

trying out this treatment at the Royal Westminster Ophthalmic Hospital, I asked him for his results. He replied that he was following Campbell Orr's method of giving 0·1 gram daily, subcutaneously however, for one month. Fenton found that the treatment was successful, indeed a definite improvement seems to take place in the first week. In 2 of his 8 cases he was unable to detect any beneficial effect. In 3 there was a very rapid and marked improvement, in the remaining 3 he found varying degrees of recovery. Orr, Fenton and Verriest believe the treatment of tobacco amblyopia by acetylcholine or acecholin (Anglo-French Drug Co.) in a large number of cases gives better and speedier results than any other form of known treatment. Doryl (Merck), which is carbaminoylcholine chloride, has the advantage over acetylcholine and other choline compounds by being stable both in the dry state and in solution, also it may be administered orally or in the form of suppositories.

It seems therefore that up to date acetylcholine is the best therapeutic agent we have for the treatment of tobacco amblyopia.

Vitamins in Ophthalmology and Neurology

Although this chapter deals with poisons which affect vision we must consider such substances as vitamins which by their absence as well as by their excess can have deleterious influences on vision and the nervous system generally.

More than thirty years ago Eijkmann was investigating the factor which caused the disease beri-beri ; his researches on the relation between experimental polyneuritis in fowls and beri-beri in man became classical. At this time little heed was paid to such substances as vitamins in nutrition, but gradually work on this subject accumulated until at the present moment its bibliography has assumed enormous proportions.

Speaking generally this subject is of vital importance to the health of the nation, as is shown by Orr's Report on the Adequacy of Diet in Relation to Income. It is astounding that such a large proportion of our population is underfed and that the food which is consumed by the poorer classes is so deficient both in quantity and quality. Orr says, " The average diet of the poorest group comprising $4\frac{1}{2}$ million people is, by the standard adopted, deficient in every respect." How are we to seek for A1 men among these millions ? As our hospitals draw so largely from this class those of us who are on the staffs of such institutions should continually

bear in mind the possible results not only of food deficiency but also the lack of an adequate supply of vitamins.

The following table with slight change is taken from Foster's paper on Vitamins in Ophthalmology.

VITAMINS IN OPHTHALMOLOGY

Vitamin.	A.	B_1.	B_2.	B_6.	C.	D.	E.
General effect of deficiency.	Keratinized epithelium.	Beri-beri.	Pellagra.	Dermatitis. and Pellagra.	Scurvy.	Rickets.	Sterility.
Source of vitamin.	Carotene. Liver oil. Visual purple.	Wholemeals. Egg yolk. Liver. Yeast.	Fresh meat. Liver. Yeast.	Fish muscle. Yeast.	Ascorbic acid. Fresh fruit. Vegetables not overcooked.	Synthetic. ergosterol. Liver oil.	Wheat germ oil. Greens.
Eye disease due to deficiency.	Hemeralopia. Xerophthalmia. Keratomalacia. ? Retrobulbar neuritis.	Retrobulbar Neuritis. Partial optic atrophy.	Cataract in rats and ? man.	—	Proptosis. ? Hæmorrhagic retinitis.	Lamellar cataract. Myopia.	—
Eye disease from excess.	—	—	—	—	—	Follicular conjunctivitis.	—

The fat-soluble vitamins are A, D and E.
The water-soluble vitamins are B_1, B_2, B_6 and C.

During his investigations into the ætiology of rickets in young dogs, Mellanby noticed an inco-ordination of movement which appeared in these animals. The two factors which determined the onset of this condition were : (1) the absence from the diet of fat-soluble vitamins and (2) the presence of a high cereal intake and especially a large amount of wheat embryo.

Vitamin A or carotene (found in all green plant tissues, carrots, palm fruits, milk, butter and eggs) was found to be the ultimate determining factor in Mellanby's experiments. Degeneration of the spinal cord and peripheral nerves was also found. The chiasma, optic nerves and trigeminal nerves were affected. In 1933 Nicholls inspected a large number of prisoners in Ceylon for signs of nutritional deficiency. Conditions due to vitamin A deficiency were found such as night blindness, diminution of vision, xerophthalmia, keratomalacia, a papular dry skin and neuritis. His post-mortem material substantiated his clinical findings ; he found degeneration of the cord and of the posterior nerve roots.

Visual dysaptation (night blindness) associated with vitamin A

deficiency has been measured and recorded by Edmund and Clemmesen, also by Jeans and Zentmire. Vitamin A plays a direct *rôle* in the bleaching and regeneration of visual purple in the retina. Wood refers to vitamin A deficiency producing a widespread epidemic of night blindness.

Vitamin A is stored in the animal body, chiefly in the liver.

Professor Möllgaard in Denmark, recognising that the vitamin A content of winter butter is comparatively low, has advised the addition of carotin oil which would raise its vitamin A content by about 10,000 units per kilo.

An interesting point has been brought forward by Roller, who says there is an antagonism between vitamin A and insulin. A greater consumption of vitamin A increases glycosuria in diabetes, also he believes that in hypothyroidism increased sugar tolerance is due to insufficient conversion of carotin into vitamin A.

In experimental animals the keratinisation of the cornea takes place due to vitamin A deficiency. Solares has described Bitôt's syndrome where night blindness with xerosis occurred in children four to eight years of age. Light perception measured by Wecker's scale was greatly reduced. These children recovered in one month by giving food rich in vitamins A and D. A young Indian came to London to study but attempted to adhere strictly to the diet prescribed by his caste for him in India. In a few months he suffered greatly in general health. When examined by the author the corneæ were becoming dim, wrinkled and vascularised. Not until he partook of milk, butter, eggs and vegetables did he recover, and that in a few weeks.

This recalls how the importance of vitamin A in vision was first recognised in 1917. At that time the children in Denmark were fed on margarine because the butter was being exported to Germany. These children developed xerophthalmia in which an alteration in the conjunctivæ takes place. occurring in patches and consisting in a dryness of the membrane. At these places (Bitôt's spots) the conjunctiva appears glistening, and is of a whitish colour and looks as if it were covered with foam. Tears do not moisten these spots. At the same time changes take place in the cornea. It becomes dull, lustreless, dry and opaque. In adults, as in the case cited above, the condition may clear up in a few weeks, but in children it may assume a most malignant form. Not only is the conjunctiva affected but the cornea becomes involved (keratomalacia). The

cornea becomes dim, a cloudy infiltrate appears at the centre and then suppurates. These little patients may then die from a general disease condition due to malnutrition. When older children survive their sight is gravely affected, due to severe scarring of the cornea. Keratomalacia generally begins with night blindness.

In conditions of the cornea which suggest a nutritional disturbance the author uses an ointment—vitamin compound (Allen and Hanbury), which helps to restore the cornea and conjunctiva to their original healthy state. The ointment contains vitamins A, B, D and E derived from halibut liver oil and wheat germ oil. The latter contains other substances which make it useful not only in eye conditions but in diseases of the skin, is a useful lactogogue and, indeed, forms a good general tonic.

Vitamin A by maintaining the integrity of the epithelial linings of the body is a powerful factor in resisting the invasion of infection. It is also essential for the proper structure and function of the nervous system.

The daily requirements of an adult are about 2,000 international units, but children require as much as 3,000. The vitamin A and D values of some of the commoner foods are given by Coward and Morgan.

Vitamin B or the anti-neuritic or anti-beri-beri vitamin is a complex, the whole of which is found in milk, liver, fruit and fresh yeast. There are two vitamins, B_1 which is aneurin and B_2 called lactoflavin. These are water-soluble. (Theorell.) The vitamin B_1 value of various foods is given by Baker and Wright. Mellanby believes that both vitamin A and B_1 deficiency is concerned in the production of polyneuritis of beri-beri, and also in the polyneuritis involving demyelination of the nerve fibres. In the polyneuritis of pregnancy where in young women pernicious vomiting has taken place, Wechsler says that since we are dealing with an avitaminosis polyneuritis the vomiting must be stopped even by emptying the uterus.

Probably 500–750 international units of B_1 are the daily requirements of an average person, but in pregnancy it is thought that even as high as 3,000 are necessary.

Vitamin B_2 (lactoflavin) is richly found in liver extract and is also found in the watery fraction of milk. (Euler.) As a yellow pigment it is largely responsible for the peculiar greenish-yellow colour of whey—hence its name. Goldberger has put forward the

theory that vitamin B_2 deficiency is responsible for the disease known as pellagra.

In the experiments on rats conducted by Day, Derby and Langston, it was found that a very large percentage of the rats given a flavin-deficient diet developed cataract, also that cataract may be effectively prevented with pure flavine under rigidly controlled conditions. From this they say that it is apparent that flavine is a specific cataract-preventive vitamin for the rat. In man, however, proof is not forthcoming that cataract follows vitamin B deficiency.

Vitamin B_6. When lactoflavin was removed from B_2 concentrates the anti-dermatitis or anti-pellagra factor remained (Lepkovsky). To this substance B_6 has been applied. Various products differ in their absolute and relative contents of lactoflavin and vitamin B_6. Fish muscle (herring, salmon and haddock) is a rich source of the anti-pellagra source but is practically devoid of lactoflavin, while egg-white, despite its high content of lactoflavin, has no anti-pellagra activity (György). Yeast is the most potent known natural source of the whole vitamin B complex. Also the product Marmite is used extensively on account of its rich vitamin B content.

In Nigeria a widespread occurrence of a disease took place characterised by retrobulbar neuritis and partial optic atrophy which was ascribed to avitaminosis by Moore, the treatment of which was the administration of foodstuffs rich in vitamin B.

Vitamin C (*ascorbic acid*), known as the anti-scorbutic vitamin; it occurs in lemons, oranges, tomatoes and lettuce, indeed in most fresh foods, including milk and meat. It is destroyed by pasteurisation or boiling. Both infantile and adult scurvy is fast disappearing owing to the general recognition of the value of fruit juice in supplementing the vitamin C content of a milk diet. Disease in the tooth pulp, together with generalised hæmorrhages, follow on a diet deficient in vitamin C. Osler states that in addition to mental depression and in some cases headache and delirium, remarkable ocular symptoms are occasionally met with, such as night blindness or day blindness associated with anæmia of the retina. It is stated that in some epidemics of scurvy night blindness is not present but that lack of vitamin C brings about proptosis of the eyes. According to Krause, vitamin C is present in the normal aqueous, lens and vitreous humour, but in the aphakic eye the aqueous humour contains little or no cevitamic acid. The suggestion has been put

forward that in hæmorrhagic retinitis of unknown origin, administration of vitamin C might possibly be of value. From Josephson's work we gather that cataract in man may be influenced by the administration of this vitamin. A certain amount was given to patients suffering from dinitrophenol cataracts and cataracts of other types. The response, he says, especially in the former, was rapidly progressive. Foster says that possibly congenitally dislocated lenses dissolve more rapidly after discission when fruit juice is administered. The potency of ascorbic acid has been standardised by Harris and Ray. Also these experimenters have shown that a low urinary output and a low response to test doses go parallel with a history of vitamin C underfeeding.

Children's requirements are 800–1,000 international units (40–50 mg.) daily, while the adult's requirement is about half that amount. Lemon juice has present 0·5 mgm. per gm. of this vitamin. (Bacharach.)

Vitamin D, the anti-rachitic vitamin, is found in oils such as cod-liver oil, in a much lesser quantity in milk, butter and egg-yolk. In Crooke's halibut-liver oil is found 2,500 international units per gramme, whereas in milk 0·1 is found, or in egg yolk 1·5 international units per gramme. (Coward and Morgan.)

Vitamin D is not identical with calciterol which results from ergosterol by irradiation with ultra-violet light. The natural vitamin D has been isolated, and it has been shown by Eliot, Nelson and others from careful investigation that the effects of vitamin in irradiated ergosterol-calciferol and the vitamin D in cod-liver oil are identically equal in effect when administered to comparable groups of human subjects. The Committee on Dental Diseases appointed by the Medical Research Council, with E. Mellanby as secretary, has published its final report on the Influence of Diet in Caries in Children's Teeth. It is stated therein that a relatively high vitamin D content of the food can do much to diminish the incidence of caries if the vitamin is given during the development of the teeth ; even after eruption the spread of caries is delayed. Foster states that the definite effect on the eyes is indirect. Excessive administration to children who are simultaneously having sun baths has caused follicular conjunctivitis which disappears on withdrawal of the drug.

The association of cataract with rickets, tetanic convulsions, malformation of the teeth, thyroidism, spontaneous hypoparathyroidism or ideopathic steatorrhœa is well known. Langdon-

Brown draws attention to the fact that the one feature common to all these diseases is hypocalcæmia. When visiting Uppsala the author had the pleasure of listening to von Bahr stating his thesis on the subject of zonular cataract in relation to tetanus. In his experimental rats von Bahr found that cataract did not accompany rickets, but only if tetany supervened. Improper feeding can be the cause of rickets, tetany and zonular cataract, and as the fault may be repeated in the children of the same family this probably explains the familial incidence which has been observed. Langdon-Brown concludes that " hypocalcæmia is an essential factor in the formation of this type of cataract. Probably lack of the parathyroid hormone which is certainly less common than lack of vitamin D is of significance only as a cause of hypocalcæmia."

In some advanced cases of scurvy cataractous opacities were found by Moniukova and Fradkin, who also state that vitamin C is not found in the aqueous of a cataractous eye. Their researches were, however, conducted on animals, so that the above cannot be said to be definitely true for man.

All authorities agree that it is advisable to supplement the administration of vitamin D by increasing the intake of calcium and phosphorus. Shipley has shown the value of taking vitamin D in order to maintain the calcium-phosphorus balance of the body. Rea is convinced that the progress of myopia is stayed by giving milk, calcium and vitamin D.

In the rachitic child both calcium and phosphates are absorbed but they are also re-excreted, thus a negative balance is brought about. The bone-forming salts, however, are retained by the administration of either the natural vitamin D or calciferol. The loss by excretion into the gut is prevented and by retention of the bone-forming minerals the calcification of growing bone is aided.

Dosage. Infants up to six months of age require 1,000 to 2,000 international units, while during the rapid growth of older children 2,000 to 5,000 can be given. Adults require 200–500.

Vitamin E is found in greenstuffs and in higher concentration in wheat-germ oil. It is known as the anti-sterility or the abortion-preventing vitamin. Evans and Scott found that rats reared on a diet containing all the known vitamins were rendered partially sterile, but this abnormality could be cured by adding to the diet such natural foodstuffs as green leaves and whole cereals. Work

done by Shute, Vogt-Möller and others showed the value of giving vitamin E in cases of human sterility and habitual abortion.

Just as hypovitaminosis is manifested in many ways such as a sub-scorbutic state (Lund), pellagra, etc., so Schübel draws attention to the possibility of the opposite condition—hypervitaminosis. Although not definitely reported in the human being this condition has been produced in animals. Vitamin A in excess produces hæmorrhagic rhinitis, conjunctivitis and diarrhœa. Vitamin B is rapidly excreted by the kidneys and therefore hypervitaminosis B does not exist, but Schübel states that large doses of vitamin D are definitely toxic. In experimental animals anorexia, loss of weight, diarrhœa, hypercalcæmia with co-existing demineralisation and finally death occurred after the administration of toxic doses. Vitamin D is eliminated through the bowel. Its excess is negatived by vitamin A.

Pellagra

Pellagra is a disease about which a great deal has been written and yet so little has been elucidated regarding its cause, and in spite of our present knowledge the disease is on the increase. It is of interest to readers of this volume, because apart from ulceration and atrophy of the alimentary canal it presents both ophthalmic and neurological symptoms.

The disease is found chiefly in the Southern States of America ; it is also found in southern parts of Europe, while several cases have been reported in England (Hutchinson, Paterson and Bigland). Most of the cases affected are between twenty and forty years of age, the negro being more susceptible than the white. The disease has been found to be endemic in asylums and orphanages ; it is in such institutions that experiments have been carried out resulting in the evidence put forth by Goldberger and Tanner that the disease is one of deficiency and can be cured by diet alone. They changed the diet in several institutions, reducing the maize element in the food and at the same time increasing the amount of protein foods—meat, milk, eggs and legumes, with the result that out of 414 " pellagrins " only one recurrence took place, but upon resumption of the former diet a recurrence of 40 per cent. took place immediately. A suggestion has been put forward by Chick that pellagra may be caused by a toxin derived from the diet which can be corrected by the presence of certain foodstuffs such as meat, milk,

eggs and green vegetables, if given in sufficient quantities. Some hold to a toxin theory (Sabry), while yet again it has been pointed out that a great many cases have been definitely proved to be associated with tuberculosis (Langworthy). It has been suggested by Carmichael and Stern that a common factor may operate in Korsakoff's syndrome and pellagra. The similarity in the histopathological appearance between the two diseases is most striking.

A comprehensive review of the pellagra literature has been given by Douglas Bigland in the *Tropical Diseases Bulletin*. He says: "It is remarkable that those who lean to the microbic theory of pellagra do not pay more attention to the achloryhydria present in a large number of cases when it is remembered that achloryhydria may be present for a considerable time prior to the onset of such conditions as pernicious anæmia and subacute combined sclerosis of the cord, when the sore tongue in the former condition and the combined cord lesions in the latter are taken into account, the possible causal connection is, at least, doubtful."

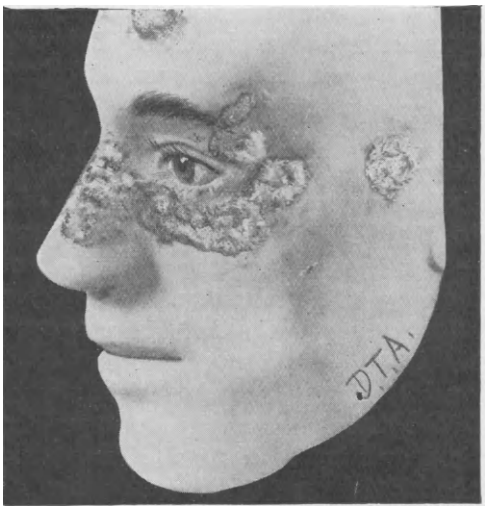


FIG. 140.—The so-called "pellagra mask" due to erythema of the eyelids in a case of pellagra. (Case of Dr. B. H. Vaughan and illustrated by D. T. Atkinson in his book "External Diseases of the Eye.")

Clinically, pellagra is recognised by four abnormalities: symmetrical skin lesions, sore tongue, diarrhœa and nervous or psychiatric manifestations. The symptoms at first are slowly developing weakness, headache and nervousness, while gastro-intestinal disturbances and diarrhœa follow. The tongue becomes red and painful, the epithelium at the tip and sides becomes desquamated and the mucous membrane of the tongue and mouth bleeds. The characteristic skin eruption of pellagra appears on the extensor surface of the extremities and trunk, on the face, vulva

and inner surface of the thighs. If the erythema involves the eyelids it usually extends peripheralwards from the nose and is known as " the pellagra mask " (see Fig. 140). Swelling of the skin takes place followed by chronic thickening of the skin layers, finally leading to atrophy of the skin, so that the skin of the eyelids becomes fissured, especially at the inner and outer canthi. The manifestations on the skin are thought to be due to certain photo-sensitive contents of the blood, possibly dioxyphenylamine (Sabry). Diminution of vision may take place due to formation of cataract or due to the effect of the toxin on the optic nerves, also diplopia occasionally occurs. Hallucinations of sight and hearing, too, are sometimes present.

The disease affects the brain, cord and peripheral ganglia everywhere. In the cases examined by Watson he found in the brain central neuritis, a condition of chromatolysis, first described by Adolf Mayer in 1901 ; the degeneration is of the axonal type —that is, one resembling the effect on a nerve cell when its axon is damaged peripherally. Susman examined the morbid anatomy and histology of a case of pronounced pellagra. He found the meningeal vessels of the cord were congested and tortuous. The cord itself was asymmetrical in many areas. The oldest lesion was in the thyroid where there was proliferation of the vesicular epithelium and increased intracellular granularity and vascularity. In the kidney were found peritubular degeneration and fibrosis, also fibrosis of the liver. The metabolic disturbances which produced the changes in the liver were, he said, due to the thyroid lesion.

The *nervous symptoms* exhibited in pellagra are spasticity, disturbance of sensation, paralysis of the sphincters and loss of the reflexes of the legs. The headache is of a very severe type, while mental changes are often most marked ; these may appear as confusion, dullness, irritability, mania and melancholic states.

Formerly the *prognosis* was bad. In the Southern States of America the outlook has always been serious. The mental deterioration is such that the majority of the patients end their days in a mental hospital. The prognosis is best in chronic cases without mental complications.

The treatment of the severely diseased pellagrins has been described fully by Spies.

Secondary pellagra has been described by Levy Simpson as a condition following gastro-intestinal disease where a limited amount

of food has been taken. Cure is apparently brought about by giving food containing vitamin B_2.

Beri-beri

Synonyms : Kakke ; Endemic multiple neuritis

Beri-beri is a form of peripheral multiple neuritis due to deficiency of the antineuritic vitamin B_1 in the diet. The disease is found in tropical and sub-tropical countries, and occasionally in more temperate ones. It is endemic and sometimes epidemic. Manson says it was formerly the scourge of the mines and plantations of the Malay and Eastern Archipelago. It is found occurring in gangs of coolies and in armies. It is prevalent in China and Japan, in South America and among the fishermen of Norway. Both sexes are affected, and also the infants of mothers attacked by this disease suffer from infantile beri-beri.

Etiology.—Although beri-beri was at first considered to be of an infective or toxic nature it has been conclusively proved by experiment that it is due to a deficiency in the diet of the water-soluble vitamin B_1, which is contained in the pericarp of the rice grain, in the aleurone layer and in the germ of the grain. A similar antineuritic body is found in other cereals, peas, beans, carrots and potatoes, also in yeast, egg-yolk and meat. Polishing the rice and the heat produced in canning food will destroy this accessory factor, and so whenever decorticated rice or canned food is used for any length of time, especially where there are food restrictions, the disease is liable to appear.

Mellanby believes that both vitamin A and B_1 deficiency is concerned in the production of the polyneuritis of beri-beri.

Pathology.—The lesions found in the nerves are those of a degenerative or parenchymatous neuritis. The nerve trunks may show slight medullary changes or even complete Wallerian degeneration. With regard to the ocular paralyses, Wright says there is a mild chromatolysis in the trophic cells of the nerve fibres and the muscles connected with these nerves show signs of a simple atrophy. The chromatolysis is also found in the ganglion cells of the anterior horns and posterior root ganglia of the cord. There is involvement of the phrenic and vagus fibres with degenerative changes in the root of the latter in the floor of the fourth ventricle.

In the dropsical form there is myocardial degeneration, serous

effusion into the pericardium and pleuric cavities, peritoneum and into the cellular tissues of the body.

Symptoms.—Although insidious in onset, beri-beri may also exhibit acute symptoms from the beginning. As in some other deficiency diseases, there are both ophthalmic and neurological symptoms present. Polyneuritis occurs early in the disease (Austregesilo).

There are three forms of beri-beri—the paralytic, the dropsical, and acute, pernicious or cardiac form.

In the paralytic form there is paralysis of a more or less degree, paræsthesiæ and tenderness of the limbs, especially when the muscles are pressed against the bone, and also loss of deep reflexes. The muscles of the forearm and hand are wasted, and may exhibit fibrillary twitching, as may those of the leg and foot. The mode of progression of these patients is that of one suffering from a peripheral neuritis. The muscles of the eyes, face and of mastication are rarely affected, although Elliot, quoting Miyashita, says that the external rectus muscle is affected in many cases, hence the convergent squint that has been seen in cases not only in Japan but in Brazil. Paralysis of accommodation has been described. The cornea and conjunctiva show a distinct decrease in sensibility which disappears on the recovery of the patient. Retrobulbar neuritis is found in some cases of beri-beri, which is evidenced by the presence of a central scotoma for white and colours, but sometimes for the latter only. The peripheral fields of vision may show contraction. The symptom of hemeralopia—seeing better in a subdued light—is also seen, thus resembling what is found occasionally in diabetic and tobacco amblyopia. From the occurrence of central scotoma it is obvious that atrophy of the papillo-macular bundle is sometimes found, but practically never a complete optic atrophy (Elliot).

The wet or dropsical form reminds one of Bright's disease. The tissues are œdematous but more solid than in Bright's disease ; myocardial changes set in with palpitation and rapid action of the heart. There is considerable muscular wasting that may not be apparent until the œdema subsides. The urine in these cases does not show loss of albumen.

The acute or cardiac form is characterised by extremely rapid failure of the myocardium, especially of the right heart. Death may follow in twenty-four hours. In all three forms various paralyses occur.

In infantile beri-beri the death rate, according to Bray, in infants eight to ten weeks after birth is 30 per cent. Many of the mothers do not show the disease although their diet has been proved to be insufficient. The child may be restless and cry continuously; it may show symptoms of sub-acute meningitis, drowsiness, head retraction and lateral rotation of the eyes.

Diagnosis.—In cases of peripheral neuritis with œdematous symptoms coming from a tropical port the existence of this disease should be remembered. Superficially, it may resemble nephritis, tabes, lead poisoning, alcoholic neuritis and heart disease.

Prognosis.—An improvement in the patient's condition may be brought about by administration of a diet rich in Vitamin B, rest in bed and cardiac stimulants. Vomiting is of serious import, while rapid dilation of the heart may suddenly terminate the disease. The treatment of beri-beri is fully dealt with in such a text-book as Manson's " Tropical Diseases."

Central Neuritis of Jamaica

(Scott's Palsy)

In 1918 H. H. Scott described a nervous disease occurring in epidemic form on sugar estates in Jamaica, the cause of which is unknown. As the diet at this particular time (May and June) consists principally of sugar and as the symptoms resemble those occurring in pellagra and beri-beri, one may assume from inference that the disease is caused by some form of food poison.

The pathological changes in the central nervous system are widespread, degenerative changes are found in the posterior root ganglia and spinal cord. The medulla, cerebellum, basal ganglia, optic and peripheral nerves are affected, Wallerian degeneration occurring markedly in the latter.

The onset is sudden, the patient first complaining of an " itching in the eyes "; one eye or both may be affected. The conjunctiva may become red and photophobia may be present, together with a burning taste in the mouth and salivation. Diarrhœa may be so severe that the patient dies in a few days from exhaustion. But it is in the constipated cases that the neuritic symptoms manifest themselves. There is a sensation of numbness and burning commencing in the feet, gradually extending up the leg. Walking is soon affected. The general resemblance to tabes is close (Manson),

although Argyll Robertson pupils do not occur. The sensations of pain, heat and cold are usually retained in the limbs and girdle pains manifest themselves.

Most cases recover, although in a certain percentage dimness of vision, deafness and a peculiar steppage gait with foot drop remain.

There is no particular treatment apart from diet. The occurrence of these cases ceases after the sugar crop has been cut.

Diabetes

Of all the diseases at the moment under the most active consideration and investigation diabetes is one which claims a large

Ocular Abnormalities in Diabetes (Waite and Beetham)

	4,001 Eyes of Diabetics.		914 Eyes of Non-diabetics.	
	No.	Per cent.	No.	Per cent.
Wrinkles of posterior cornea	1,040	26·0	96	10·5
Weakness of accommodation	165	21·0	—	—
Deep retinal hæmorrhages	730	18·0	34	3·7
Waxy exudates in retina	420	10·0	7	0·8
Depigmentation of iris epithelium	258	6·0	21	2·0
Transitory refractive changes	246	6·0	—	—
Cataracta complicata	246	6·0	75	8·0
Iritis	52	1·3	12	1·3
Atrophy of optic nerve	27	0·6	4	0·4
Homonymous hemianopsia	22	0·5	—	—
Flocculi cataract, juvenile diabetics	22	0·5	—	—
Glaucoma	21	0·5	—	—
Argyll Robertson pupils	20	0·5	2	0·2
Paralysis of extrinsic muscles	16	0·4	1	0·1
Tobacco amblyopia	14	0·3	—	—

share of thought. It is tempting, therefore, to discuss questions relating to this disease which are intimately associated with general medicine, but that is not the purpose of this book. To those who wish to know and learn much more about diabetes than can be stated here, the author recommends the perusal of such works as that of Joslin or papers written by Lawrence or Lawrence and Buckley.

As diabetes is greatly on the increase it behoves the ophthalmologist and neurologist to be thoroughly conversant with its various symptoms, especially when such a disease makes contact

with their everyday practice. We can observe symptoms of diabetes in the various systems and special senses of the body, but we have also to observe the effects of other diseases in those already afflicted with diabetes. Lawrence and Buckley's experiments prove that the diphtheric toxin will practically annihilate the action of insulin. That toxæmias and infections acting on thyroid and adrenal (the thyroid-adrenal apparatus) produce over-stimulation of these glands and this over-activity of these two glands is the cause of failure of insulin action in certain diabetics.

We will examine the ocular symptoms associated with diabetes. The table on page 474 is quoted from Waite and Beetham and illustrates the varied character of these.

The *wrinkles in the cornea* occur both in diabetics and non-diabetics—in the former they were seen in 25 per cent. but only in 10·5 per cent. in the latter. The wrinkles are not visible to the naked eye but can be seen by the corneal microscope with slit-lamp illumination ; they are situated at the depth of Descemet's membrane.

Accommodation.—In 165 out of 759 eyes of diabetics there was a paresis of accommodation, due perhaps to excessive glycogen deposits in the pigment epithelium of the ciliary bodies interfering with the action of accommodation, or it may be due to an altered condition of the lens substance.

Pupillary Reaction.—Apparent Argyll Robertson pupils were observed in 0·5 per cent. of cases and these were not associated in any way with evidence of syphilis.

Cataract.—The data given by various workers on diabetes regarding the onset of cataract varies according to what they consider constitutes diabetes. The blood-sugar of normal individuals after an overnight fast is remarkably constant, averaging 0·1 per cent. or slightly less. It may rise to 0·12 per cent. or even 0·13 per cent., which, however, is to be regarded with suspicion. All workers agree that 0·14 per cent. should be regarded as sufficiently abnormal to warrant the diagnosis of diabetes. John Rollo in 1798 (cited by Waite and Beetham), the author of the first detailed monograph on diabetic eye complications, noted the association of diabetes and cataract. Several types of diabetic cataract have been described, but most observers, having the assistance of modern methods of examination, now believe that there is only one typical diabetic cataract ; this type, which is bilateral and showing fine subcapsular dot-like opacities, is seen more commonly in juvenile

diabetics. These were first described by Schnyder in 1923, and again demonstrated and described by Goulden in 1928 (see Fig. 141).

Figures given leave it a moot point as to whether cataract is commoner in diabetics than in non-diabetics, yet one's clinical experience is that the presence of cataract in patients under fifty-five years of age is commoner among the former than in those who are not diabetic.

Proof has been given over and over again that refractive changes in diabetics are due to altered refractive index of the lens. In the aphakic eye, that is an eye from which the lens has been removed, no change of refraction takes place in diabetics. The alteration in the refraction is due not to any change in the corneal radius or alteration in the length of the axis of the eye but, as Granström has pointed out, to the salt retention and osmotic interplay following sudden changes in blood-sugar content. Duke-Elder had previously pointed out that refractive changes were due to osmotic processes involving the lens caused by a variation of the molecular concentration of the blood and tissue fluids with the sugar contents. He described several cases of change of refraction and mentioned many cases already quoted in the literature on the subject. An investigation into the chemical differences between diabetic cataractous lenses and the non-diabetic cataract has been undertaken by Carey and Hunt. The following table is quoted from their paper :—

Chemical Composition of Cataractous and Non-Cataractous Human Lenses

	No.	Cholesterol mg. per cent.	Calcium mg. per cent.	Phosphorus (non-lipoid) mg. per cent.	Ca/P Ratio.
Normal lenses . .	10	406	9·5	16·5	0·6
Cataractous lenses					
A. Non-diabetic .	30	525	30·0	19·7	1·8
B. Diabetic . .	13	511	35·0	6·2	15·5

The point brought out is that although the cataract found in the eye of the diabetic may appear similar to the ordinary senile form, yet the phosphorus metabolism was markedly different.

The change in the refraction of the eye is generally towards myopia, as the blood-sugar content rises and hypermetropic changes are usually seen as the sugar decreases. That this is not invariably so the following case will show. A man aged fifty-nine years was seen by the author in April, 1928. His refraction under a mydriatic was R. −3·25 D.S./−1·25 D.C. 80° and L. −3·00 D.S./−0·75 D.C. 180°, with which he had 6/6 visual acuity in each eye. For nineteen years previously the refraction was practically the same. During October, 1928, this patient returned, saying his glasses were uncomfortable ; examination now showed the refraction of the right eye −1·50 D.S./−2·00 D.C. 80° and the left −2·50 D.S./−2·00 D.C. 180°. From this it will be seen that the change in the right eye was more marked than that of the left. By December the cylinder in the left eye was reduced by one diopter. Examination by the corneal microscope showed the presence of several minute droplets beneath the anterior capsule of each lens but no other opacities. Dr. Calvert examined this patient's blood and found he had a blood-sugar content of 0·15 per cent. but neither sugar nor albumen was found in the urine. The diet was restricted, especially for carbohydrates, of which the patient was too fond. By January, 1936, the refraction was the same in the right eye and the cylinder in the left was then only half a diopter. The blood-sugar is 0·115 per cent. with only a trace of ketone in the urine, but no sugar. After nearly eight years there are in addition to the droplets, which changed but little, minute crystalline bodies scattered throughout the lens substance and a few faint radii are becoming evident in the right lens, while the vision of the right eye is 6/6 partly. One should look with suspicion on a case where in middle-age the refraction changes, especially towards myopia. Often one has discovered long before any lens change was visible that the changing refraction alone indicated a raised blood-sugar content. Several cases of sudden change of refraction towards hypermetropia after an injection of insulin have been recorded by Maxwell. Westcott and Ellis could find no definite relation between the amount of sugar in the urine and the refractive changes—in fact, they say that the most startling changes occur when the patient's condition is improving and the urine free from sugar.

A very thorough study of the refractive changes in diabetes has been done by Granström. One surprising thing he mentions is the large number of eyes which become hypermetropic during the course of the disease.

The one particular type of cataract peculiar to diabetics has been described by Schnyder, Koby and Goulden. The cataract was formed of a cloud of small opacities situated immediately beneath the epithelium resembling flocculi, but many of the opacities had the appearance of minute droplets. In the posterior part of the lens was another layer of opacities (see Fig. 141). Waite and Beetham found 11 cases of this kind out of 297 juvenile cataracts. The great majority of cataracts seen in diabetics are, however, of the

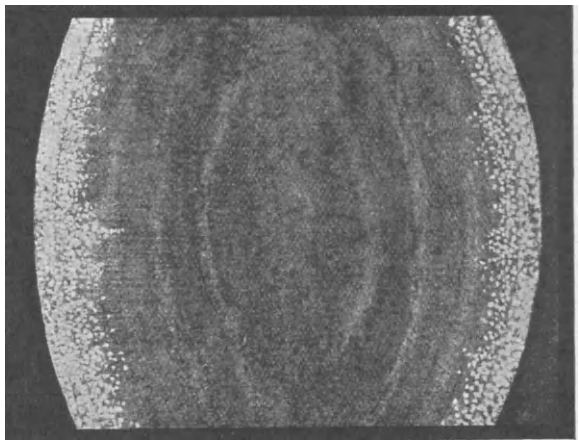


FIG. 141. Optical section of the lens of a case of diabetes, with cataract. Note the arrangement of the opacities which is so characteristic—a great number of tiny white opacities immediately beneath the epithelium of the lens and so invading the space between the band of the lens and the band of disjunction. (Kindly lent by Goulden.)

ordinary senile type, but these lens changes occur somewhat earlier in life than is normal.

It may not be out of place to mention a practical point in connection with cataract operations in diabetics to which little attention has been drawn, namely, the possible occurrence of diabetic scleritis, the onset of which may take place about one week after extraction. My last case of this kind suffered severe and prolonged pain with extreme redness of the eye for a month. A diabetic node developed at 6 o'clock where the eye had been held by the forceps during the cataract operation. The end result, however, showed neither posterior synechiæ nor vitreous opacities and former vision was regained with the proper correction of refraction. The use of the

electric eye warmer was of the utmost benefit throughout, combined with the use of analgesics, especially at night.

From experience the author regards the presence of sugar in the blood as not a positive contra-indication to the operation for cataract removal, but it is certainly a great danger. By dieting and insulin treatment the patient should be rendered as fit as possible before the operation. The presence of gangrene anywhere in the body is an absolute contra-indication to operative interference.

Retina.—In his Atlas of Ophthalmology Jaeger illustrated for the first time the condition known as diabetic retinitis. He believed it was due entirely to diabetes. Now we know that other factors are at work; Waite and Beetham say, "proof of causation of deep retinal hæmorrhages requires more than observed association with hypertension or arteriosclerosis." Also, "evidence points to the capillaries as the source of these hæmorrhages. We must know more of the physiology and pathology of the capillaries of the retina, their normal and abnormal hydrodynamics, about glycogen deposits which are known to affect other ocular tissues, about the normal and abnormal permeability of capillary walls, about thromboses which may form within them and toxins which may affect them adversely." It has been pointed out by Cammidge, Lawrence and Madders that reduced blood-calcium is a possible contributory factor in the production of retinal hæmorrhages but Waite and Beetham cannot accept this as they found only one diabetic out of nine cases of diabetic retinitis having a sub-normal blood-calcium.

Retinal Abnormalities (Waite and Beetham)

	Diabetics in 3.915 visible Fundi.		Non-diabetics in 101 visible Fundi.	
Deep retinal hæmorrhages	730	18·6 per cent.	34	3 per cent.
Waxy exudates	420	10·7 ,,	7	0·7 ,,
Nerve fibre layer hæmorrhages	196	5·0 ,,	33	3 ,,
Cotton-wool exudates	168	4·3 ,,	35	3 ,,
Iridescent crystals	28	0·7 ,,	4	
Proliferation of capillaries in retina	26	0·7 ,,		

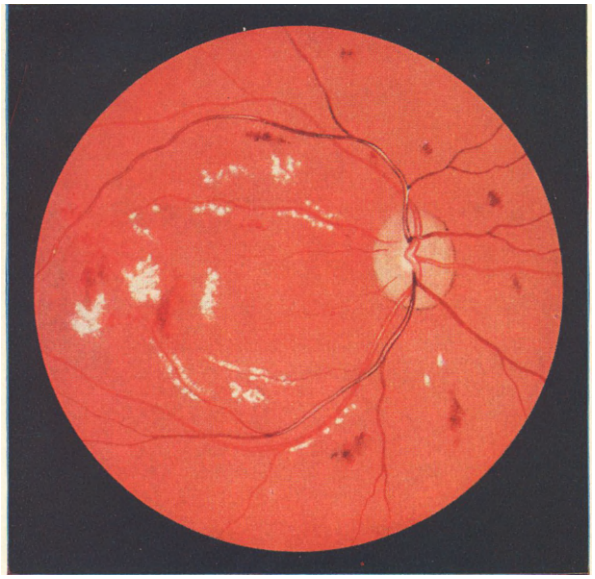
The age at which diabetic retinitis is observed is about fifty-eight years, but it has been seen in juvenile diabetics between

the tenth and twentieth years. This indeed is rare even in severe juvenile cases (Root, cited by Joslin). A certain degree of arteriosclerosis is seen associated with diabetic retinitis in individuals over fifty years of age, but it must be remembered that such arteriosclerosis is not accountable for diabetic retinitis. Waite and Beetham found that deep retinal hæmorrhages were present in 54 per cent. of diabetics that had a systolic pressure under 160 millimetres of mercury. Fischberg encountered three instances of typical arteriosclerotic retinopathy in diabetes in the absence of hypertension or evidence that the blood pressure had ever been elevated (Plate XIX).

The appearance of diabetic retinitis is that of hæmorrhages and exudates. The hæmorrhages are scattered over a wide area in the deeper layers of the retina and are somewhat circular in shape, but large subhyaloid hæmorrhages are often observed. Diabetes of long standing shows the greatest increase in retinal hæmorrhages. The exudates may be waxy with sharp-cut edges or of the cotton-wool variety. They too are scattered over the fundus and do not form a star-shaped figure at the macula as is observed in renal diabetes. In arteriosclerotic retinitis the hæmorrhages are minute, often flame-shaped and are seen particularly at the junctions of arteries and veins, while exudates, consisting of minute white spots, are scattered over the retina or gathered around the macular area, often, indeed, forming a retinitis circinata, the enclosed area of retina usually having lost most of its sensibility to light.

Lipæmia Retinalis.—There are now 48 cases of this condition on record. Of these 17 were under twenty years of age, 16 between twenty and thirty, 9 between thirty and forty, while only 3 instances are recorded over forty years of age. One case had a blood-fat of 21·7 per cent. On a high carbohydrate and low fat diet with insulin the lipæmia disappeared. A boy aged fourteen years was reported by McKee and Rabinowitch in whom the blood-sugar was 0·37 per cent., the plasma cholesterol was 1·40 milligrams and the total blood-fat 9·2 per cent. The description given of the ophthalmoscopic appearance of this boy's fundus was as follows : both fundi were alike, the discs of dirty-white colour, the arteries and veins on the discs were salmon pink in colour and indistinguishable from each other. Gradually the condition cleared on treatment. Formerly the condition of lipæmia retinalis was looked upon as indicating a fatal termination, now we know it is not so.

PLATE XIX



RETINITIS IN DIABETES, OR HYPERTENSIVE NEURO-RETINOPATHY.
(Fischberg.)

In this condition hæmorrhages and exudates are scattered over a wide area of the retina. The hæmorrhages may be found in the deeper layers of the retina and are then somewhat circular in shape, but large subhyaloid hæmorrhages are commonly found; these have the shape of a segment of a circle the upper edge of which is horizontal. The exudates may be either of the cotton-wool variety or waxy with sharp-cut edges; they, too, are scattered over the fundus and do not form a star-shaped figure at the macula.

[*To face p.* 480.

Ocular Tension.—In diabetic coma the ocular tension may fall exceedingly low, but this lowered tension is not invariable. This hypotony was first noticed by Krause in 1904. To avoid error, readings by the tonometer must be taken before intravenous therapy is begun.

Retrobulbar Neuritis.—Every degree of lowered vision may be found in diabetes, from slight mistiness of vision to complete amaurosis similar to that seen in uræmia. Shannon and McAndrews point out the lowered immunity in diabetics to alcohol and tobacco and note the remarkably rapid recovery of vision in such cases when these drugs are withheld. If the scotoma resembles that caused by tobacco it cannot be said that the loss of vision is due to diabetes, but in those cases where sector defects or hemianopias are found, or where the onset of retrobulbar neuritis is rapid and the return to normal takes place in a few weeks without the cessation of smoking, then the loss of vision may be definitely stated to be due to diabetes (O'Donoghue).

Nervous and Mental Complications in Diabetes

Ocular Muscles.—Paralysis or paresis of ocular muscles is found in diabetes. They are not common ; in fact, out of 4,000 eyes of diabetics Waite and Beetham found only 16. The external rectus was the most commonly affected muscle. These paralyses have been thought to be the result of polyneuritis, but it is more likely that they are due to vascular lesions involving the nerves. The paralysis has no bearing on the duration of the diabetes and recovery of function is usually rapid.

It is doubtful whether diabetic polyneuritis is a true neuritis. Due to age there is usually some degree of arteriosclerosis present, and, like diabetic retinitis, is so commonly associated that the inference can only be that often there is a definite relationship.

What is known as diabetic tabes has been described where pain occurring at night, deep, lightning pain, paræsthesia, anæsthesia, nerve tenderness, reflex changes, paralyses, vasomotor and trophic disturbances indicate phases of diabetic neuritis. Jordan grouped 226 cases showing symptoms of neurological disturbance as follows : (1) hyperglycæmic, (2) circulatory, (3) degenerative, (4) neuritic. In the first group the pain involved the legs only. Unlike those cases where the neuritis is of spinal origin where rest is sought, the patients with diabetic neuritis walk about at night for relief of

pain. In the second group, arteriosclerosis is not entirely responsible for recovery from severe pain and paralysis can take place in a few weeks although these may persist for years. The degenerative (3) type showed little pain or paræsthesia, the legs alone in some cases were affected. Pupillary changes were common; some of them have been described as Argyll Robertson pupils. But it is in Jordan's fourth group that those cases belong which ordinarily are termed diabetic tabes. Many had absent knee or ankle jerks. Cranial nerves such as the optic, oculo-motor, facial and auditory were involved in one-fourth of this group.

The *chemistry* of diabetic nerves has been worked out by Jordan, Randall and Bloor. Their results show that the phospholipoids, cholesterol and cerebroside content of diabetic nerves is much lower than in the normal nerve tissue.

In the following paragraph I freely quote from Joslin. In his book Root and Jordan state that insanity rarely develops in diabetes, that senile dementia is common and senility is generally premature. They also quote observations of Kuchens who recorded paralysis agitans in five generations and in the same family diabetes was present four times, being grave in two uniovular twin boys aged fifteen years. Cases of meningitis and encephalitis as complications of diabetes are also quoted.

Epilepsy and cerebral vascular accidents do not occur with any greater frequency in diabetics than in non-diabetic persons, although the incidence of vascular lesions leading to gangrene of the leg and coronary occlusion should not be forgotten.

Overdosage of insulin will produce those symptoms seen in functional hyperinsulism or loss of stored glycogen : such are faintness, weakness, emotional outbursts, diplopia, speech defects, paralysis of arm or leg, delusions and hallucinations, blindness and hysterical weeping.

Diphtheria

Synonym : Membranous Croup

Apart from the faucial, laryngeal and nasal varieties of diphtheria there are other localised forms of this disease. The infection may be found in the conjunctiva of the eye. Just as a membrane forms in the main varieties, so on the conjunctiva the same may appear. On everting the lids the palpebral conjunctiva may be seen to be covered with a whitish membrane which can be peeled off

with little bleeding. The infection of the conjunctiva may be mild (croupous) or marked (deep or brawny), the latter showing thickened lids. The membrane does not occur on the ocular conjunctiva. It must be remembered that other forms of conjunctivitis, for example, the pneumococcus and the staphylococcus, may show the presence of the membrane, yet it is most important that a bacteriological examination may be made to establish the presence or absence of the Klebs-Loeffler bacillus. Confusion might arise owing to the xerosis bacillus, which is a normal habitant of the conjunctival sac, being present. Inoculation into guinea-pigs with cultures of the former is followed by death, but the bacillus xerosis does not kill the animal.

Treatment by antitoxin should be used in all cases whether doubtful or no, and even local instillation of the antitoxin into the conjunctival sac benefits (Parsons).

Uhthoff found by experiment that severe purulent corneal infiltration can be produced by the diphtheria bacillus alone, so in man corneal involvement is sometimes found in this disease both in the croupous and deep forms. The ulceration is severe, often leading to total loss of the eye. After removal of the eye a thick wash-leather membrane may reform in the orbit. Foster Moore has seen it remain for six years.

A metastatic form of purulent choroiditis may occur in diphtheria as well as in meningitis, cerebrospinal fever and other acute infectious diseases.

Paralysis.—Following the course of the main types of diphtheric infection various paralyses may occur; the paralysis might rather be described as a paresis, for in many instances there is scarcely any impairment of function noticed. It was not until the middle of the nineteenth century that the association between the actual faucial attack and the subsequent paralysis became known. Trousseau in 1885 showed that paralysis could follow even cutaneous diphtheria. Rolleston in a series of 2,300 completed cases of diphtheria observed some form of paralysis in 20·7 per cent. He pointed out that the frequency and severity of diphtheric paralysis bear a direct relation to the character of the angina, as shown by the extent and duration of the faucial exudation, nasal involvement, œdema and fœtor and the concomitant adenopathy. Diphtheric paralysis was commoner in children than in adults, the great majority occurring between two and ten years of age. The only paralyses

which occur during the first fourteen days of the disease are cardiac and palatal paralysis. Ocular paralysis takes place three to four weeks after the commencement of the disease and shows chiefly in the form of paralysis of the accommodation. The paralysis is usually bilateral although unilateral cases have been recorded. The patient notices his reading vision is not good and there is difficulty in seeing small objects. Glasses may be required but only for a short time. Among 210 cases of paralysis of accommodation Rolleston observed the average period to be 21·1 days before recovery took place, the longest being 41 days. The external muscles may be affected, the external rectus being the commonest, also the oculo-motor nerve may show paralysis or paresis. Squint and diplopia may therefore follow, but rarely has complete ophthalmoplegia been seen. In 62 cases of squint the average duration was 18·4 days, the longest 47 days. The pupillary reflexes are not affected. Between the fifth and seventh weeks after infection symptoms of polyneuritis occur. At the same time pharyngeal, laryngeal and diaphragmatic paralysis may become evident and is associated frequently with facial paralysis producing a vacant expression. There may be loss or diminution of the knee and ankle jerks with some sensory impairment such as anæsthesia, hyperæsthesia, numbness, formication, loss or diminution of vibratory sensation and astereognosis. Examination of the pupils in adults so affected will help to avoid making a diagnosis of tabes. It may take months before there is complete recovery of the limbs.

In cutaneous diphtheria the toxin spreads upwards by the nerves to the spinal segment which innervates the muscles of the infected region. But in ocular paralysis and polyneuritis the toxin spreads to these parts by means of the blood stream.

Six cases of diphtheric hemiplegia have been recorded by Rolleston. The condition is not due to a primary attack on the nervous system but is secondary to interference in the blood supply by embolism, thrombosis, or hæmorrhagic encephalitis, the first being the commonest. It occurs during the first six weeks. The prognosis is unfavourable as regards complete recovery; epilepsy and mental defects may follow.

In the treatment of diphtheria Rolleston deprecates the doctrine that the antitoxin is of no value after the fifth day; he urges that "the presence of membrane in the throat, nose or elsewhere, however late in the disease, is an indication for serotherapy."

Regarding the Schick test, its principal value lies in the selection of individuals for immunisation. Chesney says : " Alum-precipitated toxoid used in two doses of 0·2 c.c. and 0·4 c.c. at an interval of four weeks gives a high level of Schick immunity. The initial small dose, acting as a detector of hypersensitive persons, eliminates reaction difficulties. The high Schick-negative rate obtained obviates the necessity for routine Schick-testing after immunisation."

Cerebrospinal Fever

Synonyms : Epidemic Cerebrospinal Meningitis ; Spotted Fever

For over 100 years this disease has been known, but not until 1907 and again during the Great War did cerebrospinal fever become prevalent in Great Britain. The disease occurs most commonly in the first five years of life and in adolescence. According to Rolleston, males are attacked in much greater numbers than females. The presence of chronic naso-pharyngeal catarrh seems to be a predisposing factor. Although in the most malignant form death may take place within twenty-four hours, yet in the ordinary form the stage of incubation is four or five days. The disease begins suddenly, meningeal symptoms becoming manifest by intensification of headache and pain along the vertebral column with rigidity.

The meningococcus is a small Gram-negative organism closely resembling the gonococcus morphologically. It is found in pairs or tetrads enclosed within pus cells or may be found free in the blood or cerebrospinal fluid. It is aerobic, non-motile and does not form spores. During the incubation period the patient may show some degree of naso-pharyngitis. This is followed by sudden onset with headache, giddiness and malaise, the temperature rising to 103° or more.

The *ocular symptoms* are found in 50 per cent. of cases (Jochmann). At first the pupils may be unequal and respond sluggishly to light and accommodation. During the acute stage transient paralysis of the cranial nerves is commonly observed, recovery usually taking place as the meningitis subsides ; facial herpes febrilis is frequently present at this stage and facial paralysis may occasionally be noticed. Nystagmus is uncommon. Conjunctival hæmorrhage may be seen and *conjunctivitis* is not infrequent. The appearance of a chemosed conjunctiva may herald the onset of panophthalmitis. Just as there may be a metastatic conjunctivitis in

gonorrhœa, so in cerebrospinal fever a metastatic form may be found from which the meningococcus may be isolated. Axenfeld and McNab give a full account of the finding of the meningococcus in the conjunctiva of cases suffering from meningococcal meningitis, also cases where the disease was not present nor had yet developed, and in the eyes of those who had nursed cases of cerebrospinal fever. They describe the differentiation of the meningococcus, the gonococcus and micrococcus catarrhalis, all of which may be found in the conjunctival sac during an attack of conjunctivitis.

Keratitis followed by corneal ulceration may take place, due particularly to want of proper closing of the lids, and, as from the conjunctiva, so the organism may be obtained from the corneal ulcer. Serious scarring of the cornea may follow, the ulceration resulting in greatly diminished vision.

Metastatic iridocyclitis passing into *choroiditis* takes place in 4 to 6 per cent. (Gaudin). It is most commonly unilateral; beginning as a purulent iridocyclitis, it spreads backwards, filling the globe with pus—panophthalmitis, or the process may be subacute and plastic, ending in shrinking and atrophy of the globe. A suppurative choroiditis may occur, giving an appearance of the presence of glioma of the retina—pseudoglioma. *Hyalitis*, described by Metz-Klockhas, a form of endophthalmitis, resulting in a shrinking of the vitreous, detachment of retina and finally phthisis bulbi, occurs in 3 to 6 per cent. and is similar to what may be found after any general infection, the infection being most probably by the choroidal vessels. These have all ended in blindness until lately, when Netter records he injected anti-meningococcic serum into the vitreous and thus produced cures. Also McLean records a case where the serum was given both intravenously and intrathecally and sight was saved.

In the malignant form of cerebrospinal fever the only neuro-ophthalmological symptoms may be papillœdema and strabismus.

In the first and second year of childhood the fulminating type is rather uncommon, so also is septicæmia.

The complications of the nervous system are hemiplegia and hydrocephalus, the latter being external or internal.

The *hemiplegia* may be transient or permanent, the former when due to œdema, the latter to encephalitis or cerebral hæmorrhage. If there is effusion into the subarachnoid space then the *hydrocephalus* is described as external, but if into one or both

ventricles then the hydrocephalus is internal. The latter variety is the most important complication in cerebrospinal meningitis. Its onset, usually late in the course of the fever, begins with intensification of headache and vomiting. Papillœdema is usually absent, but the vision diminishes and sometimes atrophy becomes evident.

Posterior basic meningitis is a sporadic form of cerebrospinal fever occurring in infants between the ages of four months and two and a half years ; it is bacteriologically identical with meningococcal meningitis. The meningococcus was identified by Still in 1898. It differs clinically from the ordinary type in adults (Rolleston) in its chronic course, the scarcity of eruptions, the frequency of blindness and rarity of deafness ; indeed, hyperacusis is commoner. Opisthotonous is more marked. The onset may be acute with convulsions, vomiting, diarrhœa, or it may be gradual with increasing drowsiness and occasional vomiting. The majority die after a few weeks' illness, the greatest fatality being between the first and second year, that between the third and fifteenth year being the most favourable (Neal). Those who survive may show such sequelæ as blindness, hydrocephalus, and mental deficiency. Nystagmus and strabismus are sometimes observed, and although blindness may be present the pupils react to light. Optic neuritis or papillœdema is rare, the fundus being generally normal in appearance. Sight often returns in a few weeks ; the longest period recorded by Thursfield and Paterson before the sight returned without loss of vision was four months. Brain states the loss of vision is probably due to downward pressure of the floor of the third ventricle upon the optic chiasm and also in part to pressure on the optic radiations and visual cortex by distention of the posterior horns of the lateral ventricles. There is dilatation of the pupils with weak light reflex. Paralyses of the external ocular muscles are common, especially the external rectus. Nystagmus is common and there may be some degree of exophthalmos. Macewan's sign, that is, the presence of a dull tympanitic note on percussion of the fronto-parietal region, is found in many cases. The raised intracranial pressure in infants produces bulging of the anterior fontanelle and even separation of the cranial sutures. Therefore the enlargement of the head in infants is more uniform than in older children. Convulsions may take place, while the limbs become progressively weaker with exaggerated tendon reflexes.

Herpes, which occurs between the third and sixth day of the

disease, may cover the eyelids, ears, cheeks and neck. Mucous membranes may be involved such as the conjunctiva, hard palate and nasal mucosæ.

Leprosy

This disease, although rare in Europe, is found in many parts of the globe. Sir Leonard Rogers estimates the probable number of lepers to be roughly 3,000,000. During recent years many workers have contributed to the subject of leprosy : Danielsen and Hansen of Norway, E. Muir and Sir Leonard Rogers at the Calcutta School of Tropical Medicine, Perkins and Rodriguez of the Philippine Islands, also many others, such as J. Lowe, who saw 5,000 cases of leprosy during eight years at Dichpali. It is owing to the work of these investigators that great advances have been made in the early diagnosis and the treatment of leprosy.

The disease shows a generally infective condition due to the bacillus Lepræ of Hensen, producing granulation-like neoplasms which arise primarily in connection with the skin and nerves.

Leprosy is not hereditary ; the age incidence reveals that the second decade of life is the one most liable to infection, while the commencement of sexual life appears to lower the resistance to the disease. Proof has frequently been given of the wisdom of removing children after birth from their infected parents ; also it is becoming more clearly understood that it is the nodular form of the disease which shows the greater infectivity, due to the copious discharge of the lepra bacilli from the ulcerated mucous membrane of the nasal region and skin lesions. In the nerve form there are comparatively few bacilli present in the nerves.

There are three forms of leprosy : (1) the nodular or tuberculated form affecting the cutaneous tissues chiefly ; (2) the maculo-anæsthetic form affecting the nerves, resulting in a peripheral polyneuritis ; (3) there are also mixed forms.

The incubation period of leprosy may be as short as 100 days, but in 80 per cent. the true incubation period does not exceed five years (Rogers).

The nodular form or cutaneous leprosy begins with a period of malaise associated with dyspepsia and diarrhœa, followed by the appearance of shiny, red, hyperæmic spots situated on cheeks. external aspect of thighs and those portions of the body on which the patient lies. Gradually the patches on the skin coalesce and,

finally, nodules are formed the size of a hen's egg, consisting of granulation tissue containing multitudes of bacilli.

The *maculo-anæsthetic* form begins with a certain amount of malaise followed by sharp and tingling pains along the course of certain peripheral nerves. The ulnar, median, saphenous and peroneal are those most affected. The symptoms of muscular weakness going on to paralysis follow in a symmetrical manner. Paralysis of the facial muscles, including the orbicularis palpebrarum, with anæsthesia, also paralyses of the muscles of the limbs with loss of sensation take place. The skin is not only involved but ulceration, bullæ, necrosis of the distal bones of the hand take place, the latter finally producing deformities of the hands, giving them a claw-like effect. The affected areas of the skin tend to coalesce, forming large raised irregular ovals with a depressed white anæsthetic centre. Thickening of the peripheral nerves and loss of fingers and toes are seen, together with ankylosed joints. In the nerve type the bacilli are very few in number and are found in the fibrous tissue of the nerve trunk and in that of the nerve endings of the skin. The incidence of nerve abscesses has been described by Lowe. Many of these, he observes, have followed on the treatment of potassium iodide. The ulnar nerve above the elbow, the common perineal, superficial perineal, the cutaneous nerves of the forearm and leg, also the great auricular are, he observes, those generally affected.

Rogers draws attention to the self-healing tendency of these nerve cases and thinks this aspect of leprosy is not sufficiently recognised.

The clinical picture of leprosy in *neurological* aspects is entirely due to polyneuritis—of a most peripheral kind (Monrad-Krohn).

The *ophthalmic* lesions in leprosy are numerous. We are indebted to Neve for a full account of those which he observed in the Kashmir State Leper Asylum. He states the proportion is greater in the tubercular form than in the anæsthetic. In the former the spread is by infiltration, while in the latter the results are from exposure.

The early signs of ocular involvement are (1) failure of the pupil to dilate fully and symmetrically on the instillation of homatropine or atropine ; (2) thickening of the bulbar conjunctiva ; (3) infiltration of the cornea in the form of a pterygium-like growth, nodules or interstitial keratitis ; (4) redness or photophobia. Leprosy attacks the eyebrows and eyelids with great frequency. In the nodular form the surrounding skin of the orbits is quickly invaded by the tubercles

of the disease. The eyebrows and eyelashes fall out at a very early stage, sometimes the hairs before falling out become white and splintered. In the nerve form of leprosy the cornea and conjunctiva lose their natural sensitiveness and become anæsthetic, and this condition, complicated by paralysis of the orbicularis allowing the lower lid to fall away from the eye, permits the entry of foreign bodies, dust, etc. At first there is excessive lacrimation just as there is excessive sweating but later the secretion of the tears dries up, consequently the epithelium of the cornea and conjunctiva becomes hypertrophied and chronically inflamed. An atrophic shrinking of both the conjunctiva and cornea may take place, ulceration of the cornea leading to perforation with the loss of the globe.

The paralysis of the eyelids together with the associated hypertrophy of the conjunctiva results in lagophthalmos (inability to close eyelids). The anæsthetic eyelids are liable to irritation and injury from the bites and stings of insects. Nodules appear in the conjunctiva as elsewhere, but these extend from the deeper tissues. Neve has seen keratitis and ulceration of the cornea, but he considers that the most characteristic lesion of leprosy is an infiltration of the corneo-scleral margin in the sclera extending circumferentially. At first there may be only a little dotted deposit of a yellowish colour, possibly vascular, more often it appears as a nodule involving the sclera, cornea and iris simultaneously ; in the cornea it is superficial to Descemet's membrane, from which in some cases it may be dissected. This is a symmetrical lesion. Iritis follows this condition while nodules form in the corneo-iridic angle. Rarely do these nodules form on the edge of the iris. The iris becomes thickened and the deposit of lymph tends to occlude the pupil, producing blindness. If the infiltration begins in the lower angle it resembles an hypopyon ulcer.

Inflammatory lesions of the uveal tract are very painful, and indeed infection of the eyeball is possibly the most painful and distressing condition caused by leprosy (Muir). The posterior part of the globe is but rarely affected although the whole globe may be destroyed by suppuration or general infiltration. The optic nerve is seldom or never the seat of leprous deposits although the ciliary nerves may be infiltrated by spread from the corneo-scleral margin. The treatment of the condition of ectropion which has been followed by McNair, is by the insertion of strips of fascia lata around the

eyelids, a procedure which might probably be taken advantage of more commonly for the condition of ectropion. The electrocautery has been used for the corneo-scleral margin infiltration, not going too deeply to avoid injuring the underlying structures.

For corneal ulceration, especially associated with cyclitis or iridocyclitis, atropine is used, salicylates for pain, while protein shock has been found useful.

Intraocular operations such as iridectomy, cataract, extraction, etc., are usually unsuccessful and are followed by phthisis bulbæ (shrinkage of the globe), although several successful operations have been recorded. In chronic iridocyclitis McNair gave subconjunctival injections of epinephrine hydrochloride followed by powdered atropine. For keratitis he uses gold sodium thiosulphate intravenously and protein shock, while he instills 20 per cent. chaulmoogra oil and dionin into the conjunctival sac. He uses dionin quite freely.

The operation of complete peritomy, that is, resecting a piece of the conjunctiva down to the sclera around the corneal margin, is recommended by Kirwin for pericorneal infiltration. Muir and Chatterji recommend the use of trypan-blue injections. They give an intravenous injection of 1 per cent. solution, using Grübler's in normal saline, giving 3 to 20 c.c. once or twice a week. This can be done for only a limited time as the drug has a selective action on the leprous granulomata. Ulceration with gastro-intestinal disturbances appear, and as the kidneys do not excrete the drug it remains in the body for a long time, staining the skin; but this finally disappears. Perhaps the better way, they say, is to give subconjunctival injections of trypan blue 1 per cent. in normal saline sufficient to inflate the conjunctiva. This method is simpler and probably just as effective.

For the general treatment of leprosy I would refer the reader to many works published on tropical diseases.

Botulism

History.—In 1894–6 van Ermengen defined the symptom-complex of botulism intoxication. He found the bacillus in a ham, the eating of which had caused 50 cases of illness. Botulism is a form of food poisoning due to the toxin of the *B. botulinus* which is formed in the food and absorbed in the gastro-intestinal tract. The bacillus and its spores are widely distributed in nature. It is

anaerobic and produces a powerful exotoxin which is destroyed by boiling. It is an inhabitant of the soil and by soil contamination food is infected. Both animal and vegetable matter may be infected, the toxin thus formed being of deadly virulence. Tinned food when kept at certain temperatures, say, above 68° F., forms a suitable medium for the cultivation of the bacillus and its toxins.

Clostridium botulinum has been separated into several "types" and varieties, so that Types A, B, C, D and Cl parabotulinum are now recognised forms. Man is affected chiefly by Types A and B, which are distinct immunologically, so that the treatment of human botulism with specific antitoxin should be by the use of a serum containing antitoxins for both Type A and Type B. The disease seen in wild birds such as geese and ducks in marshy districts in California is due to Type C, Type B has been isolated in forage poisoning in horses, while Type D and parabotulinum have been isolated from cattle.

Extremely uncommon in Great Britain, yet one remembers the outbreak that occurred in Scotland in 1922 when eight people were poisoned and all succumbed, the first after one day's illness, the last after six days' intense suffering. The first symptoms occurred in fourteen hours after the ingestion of sandwiches made from potted wild duck. A recent epidemic of food poisoning in Stockholm has been described by Bergman. The botulinus toxin was demonstrated by injecting the patients' sera into guinea-pigs and mice.

Osler and McCrae quote Edmunds as stating that the peripheral changes are important, that it is a "curare-like" reaction, producing a paralysis of the motor nerve-endings and that the nuclei most affected do not give the most marked clinical findings. This agrees with the results obtained by Dickson and Shevky, whose researches show that the paralysis is due to muscular weakness owing to the peripheral action of the toxin on the nerve endings. Ophüls after a careful study of a brain taken from a human victim found the nuclei of the third, fourth, fifth, sixth, seventh, eighth, ninth, tenth and twelfth cranial nerves showing the Nissl granules staining well in all his sections; the nuclei of the ganglion cells were perfectly normal.

The pathological lesions show marked thrombus formation in the arteries and veins with hæmorrhages in the meninges and central nervous system. The meningeal reaction is slight, there is

an absence of fever and the sphincters are not affected. The tendon reflexes are preserved. The results are motor, not sensory.

Symptoms.—In the cases reported by Monro and Knox the earliest incubation period was fourteen hours. Symptoms generally showed themselves after eighteen hours. The earliest symptoms are usually visual: ptosis, diplopia, dilated pupils and sometimes complete external and internal ophthalmoplegia. Vision becomes dim owing to the failure of the ciliary muscles which is much more marked when hypermetropia is present, and conversely in myopes, as they have not to use the muscle of accommodation, the loss of vision may not be noticed. In addition there is giddiness, inability to protrude tongue, thick speech, or complete loss of speech, palsy of the diaphragm, paresis of upper limbs, and severe vomiting. Intelligence is unimpaired. There is absence of headache, while the heart may remain unaffected although the most rapidly fatal cases die from heart failure. The cases which live for several days suffer very severely, struggling for breath, the struggle resulting from paresis of the muscles of respiration. However, the picture is one of extreme fatigability and not that of an actual paralysis.

When confusion might arise in the diagnosis between botulism and encephalitis lethargica it is well to remember that the onset of symptoms in the former is much more rapid than in the latter. Also as has been pointed out by Monro and Knox the facial muscles in botulism escape, especially that part below the eyes, and therefore the face does not resemble in any respect the typical Parkinsonian mask.

Treatment.—As food infected with *B. botulinus* gives off a rancid odour such should be carefully avoided and certainly not tasted. It is recorded that half an infected olive was the means of causing a death. To alleviate the intense suffering morphia may be given. The alimentary canal should be washed out proximally and distally as thoroughly as possible. The only remedy known in the treatment of botulism is Botulinus Antitoxic Serum, which must be given early. Suitable supplies of serum are kept at the Ministry of Health and in some of the public offices in the more important centres.

In the Scandinavian outbreak reliance was placed on the botulinus antitoxin. The dysphagia and eye symptoms cleared up slowly over a period of three months.

CHAPTER XVII

HEADACHE AND AMAUROSIS

FOLLOWING the removal from the frontal region of a tumour which has eroded part of the orbital roof or wall, the neuro-surgeon discovers that, owing to the accumulation of hæmorrhage, the eye is proptosed, the conjunctiva is œdematous and the vitality of the cornea threatened. He must turn to the ophthalmologist for help, seeking his aid in the care and protection of the cornea. For constant lavage of the swollen conjunctiva with mild luke-warm boracic lotion and smearing with boracic cream is necessary to maintain an aseptic condition of the eye. So also in the matter of headache one must turn to the other for help and suggestion, for headache is a symptom of a condition, the elucidation of which may need the diagnostic skill of both ophthalmologist and neurologist.

It is remarkable that at the present time one writer after another turns back for reference to Harry Campbell's work on "Headache and Other Morbid Cephalic Sensations," published forty-two years ago. A great deal has been written since and new methods have been utilised for the investigation of disease, but one has to confess that not a great deal has been added to our knowledge of the actual condition known as headache.

To begin with, let us try to differentiate headache from neuralgia. Fully 60 per cent. of the patients visiting the ophthalmologist complain of headache of one sort or another, but when carefully questioned as to where the pain is felt they give vastly different replies. For instance, one patient may say he has a constant pain around the eyes, particularly above the level of the eyebrows, indicating the site of the pain by placing his hand on the forehead, while others say that they feel the pain at the back of the eyes, and again the statement is made that on using the eyes pain is felt in the occipital region. Gordon, from an examination of 1,339 patients, says he is led to the conclusion that the site of ocular pain is extracranial; it is in the superficial muscles of the head. But what would he think if, while attending a clinic in a hospital for nervous diseases,

he were told by many patients that their pain is within the skull; that it is bursting, boring and seems to have no relationship with the exterior? Here is the real difficulty regarding cephalalgia. Let us then accept for the present the word "headache" and try like Galen to differentiate its character according to its causation. In the first place we remember that in blockage of the foramen of Monro by a tumour of the choroid plexus there is distension of both lateral ventricles. If the tumour is mobile, such as described by Dandy, Paterson and Leslie, paroxysmal intermittent headache is of mechanical origin due to obstruction of the circulation of the cerebrospinal fluid. Again, in lumbar and cisternal puncture headache is sometimes a prostrating sequel. The lowered pressure of the cerebrospinal fluid reducing the entire intracranial pressure is the most probable cause of the headache. Some observers have noticed that in those cases of cranial injury where cerebrospinal fluid and blood have escaped by the ear or where lumbar puncture has been done soon after the accident the patient has not complained of the usual intense headache, in some cases of none at all. We see then that an alteration in the volume of the cerebrospinal fluid can cause headache and that this headache is not related to any external structures although it may be localised, *e.g.*, it may be unilateral, frontal, vertical, temporal or occipital.

Secondly, we must consider the intracranial pressure brought about by a tumour. The location of the tumour means not only pressure on surrounding structures such as the small veins which produce local venous congestion leading to œdema of the immediate tissues surrounding the tumour, but also on such structures as the great vein of Galen leading to a condition of internal hydrocephalus. Dandy's explanation of this condition is, that as this vein drains the choroid plexus and if, therefore, a ligature is applied near its origin, it leads to increased formation of cerebrospinal fluid following on the venous congestion of the choroid plexus. In addition then we have increased venous pressure as well as increased cerebrospinal fluid pressure. The skull is a rigid cavity and is incapable of expansion. It contains the brain and its membranes, the blood vessels and the cerebrospinal fluid. The pressure within the skull is stated to be somewhere near the mean of the systolic and diastolic blood pressures. How the pressure is regulated is still a matter for conjecture. Experiments and work done by Stohr, Forbes and Wolff indicate that there is a vasomotor control of the cerebral blood

vessels. The nerves of the pia and choroid plexus are derived at the base of the brain from the third, sixth, ninth, tenth, eleventh and twelfth nerves. The fine meshwork of nerves to the fourth ventricle also is to be found in the pia of the optic nerve ending in club-like swellings. In spite of Florey's work in which he states "the cerebral arteries react to mechanical, thermal, electrical and chemical stimuli by contraction and dilatation," also "no evidence is forthcoming for the presence of any nervous control over the calibre of the cerebral vessels," yet he admits that inflammatory changes observed in regard to the cortical vessels are essentially the same as those occurring in a region supplied with vasomotor nerves. The consensus of opinion seems to be that there is a definite intracranial regulation of pressure and, as Robey points out, the mass of nerves overlying the vegetative centres of the third and fourth ventricles are probably for the protection of the underlying centres against the change in pressure, mass and composition of the cerebrospinal fluid. The conclusions reached by Pickering, Clarke, Hough and Wolff are that in experimentally produced headache by the intravenous injection of histamine, pain is associated with the dilatation and distortion of the intracranial vessels and also that the sites of origin of the nervous impulses experienced as pain are probably the walls of the intracranial blood vessels and the perivascular tissues. This, therefore, is a further evidence that deformation of the intracranial blood vessels is an important factor in the production of headache as manifested clinically. A case of the author's, mentioned in his "Affections of the Eye in General Practice," suffered from constant headache from the month of December till May of the following year, but it was not until then that the first sign of raised intracranial pressure showed itself by the onset of papillœdema in one eye. At the autopsy the tumour was found situated at the base of the brain pressing on the blood vessels, pituitary body and optic tracts. Headache therefore can be produced by a tumour pressing upon the structures responsible for maintaining the intracranial pressure, upon localised tissues such as the dura, nerves, Gasserian ganglion and its branches. Yet one has seen the orbital contents on one side proptosed $\frac{1}{2}$ inch by a slowly growing intracranial cholesteatoma without either headache or papillœdema.

From what has been said it is important to remember that constant headache is of grave prognostic significance. The type of headache which is paroxysmal and wakens the patient at night is

not the type which is produced by disturbances of the ocular apparatus. Combined with papillœdema and vomiting, headache is diagnostic of an intracranial neoplasm; its location, however, seldom helps in pointing out the position of the tumour.

As mentioned in Chapter XII, *pituitary* tumours cause a headache which is not as a rule intense, but is boring in type and may be referred to any part of the head. Suprasellar meningiomas do not usually produce severe headache, but acoustic neuromas may be accountable for paroxysmal attacks of headache radiating over the head and down the spine. Irradiation of certain pituitary tumours causes shrinking and therefore diminution of headache and amelioration of ocular symptoms, including restoration of central and peripheral vision.

Robey, quoting Hodges, contrasts migrainous and pituitary headaches. *Migrainous* headaches occur paroxysmally in neurotic individuals, often with a history of direct heredity, and at almost any time of life. They are characterised by periodic attacks of pain, continuing for varying periods, in the course of the trigeminal and are usually associated with nausea or vomiting and various vasomotor disturbances which are not amenable to treatment. Pituitary headaches are localised and persistent in type and occur in patients often showing clinical dyspituitary disorders; they are usually relieved by continuous and proper glandular feeding.

The use of ergotamine tartrate in the treatment of migraine has revived controversy regarding the causation of the headache in this disorder. By measuring the electrical variation of the skin Solomon has shown that there is no difference in the activity of the sympathetic system between those patients in whom relief from headache has been obtained by the use of ergotamine tartrate and in those where the headache is allowed to run its course. On the other hand Love and Adson found that following the operation of cervicothoracic ganglionectomy for Raynaud's disease those patients who were subject to migrainous headaches obtained relief, the headaches being either abolished or relieved.

The headache following spinal anæsthesia resembles migraine in origin and, as Bacher-Gröndahl points out, is a reflex effect arising in an unstable nervous system and based on anæmia of the brain. Hence his suggestion that 4-6 drops of ½ per cent. alcohol solution of nitroglycerin will bring about relief.

Headache is a marked feature also in various vascular conditions

of the central nervous system, *e.g.*, in *chronic subdural hæmorrhage* it is persistent and severe, becoming worse as drowsiness passes into stupor.

In the author's experience one of the most violent types of headache observed was seen in the final stages of the young woman of whom the ophthalmoscopic appearance of her eye is depicted on Plate XIII., and who died from rupture of a basal aneurysm (see Fig. 78). The pain in a case of this kind radiates all over the head and down the back of the neck.

Severe sudden headache may be due to an *intracerebral hæmorrhage*, while another cause may be *thrombosis* of a cerebral vessel. The latter occurs more commonly than embolism of the cranial arteries. In thrombosis of the cavernous sinus pain is severe and is localised in the orbital or frontal region. Before unconsciousness sets in it is found that hyperalgesia of the ophthalmic division of the trigeminal nerve is present. Headache occurs also in thrombosis of the superior longitudinal sinus and the lateral sinus; in both papillœdema may be present but is usually unilateral, being on the same side in thrombosis of the lateral sinus.

In ophthalmic practice there are three kinds of patients commonly met with who complain of headache. The first consists of those who devote a lot of their time to shooting; they develop "gun headache," which is a most annoying complaint. Such patients are anxious to know if their eyes are at fault, but the only cure known to the author is to stop shooting for a season, for it is really a concussion headache.

The second class consists of those who have met with an accident some time previously. The injury may have been slight and unaccompanied by shock or it may have been more severe; in either case, no obvious cranial injury was noticed at the time of the accident. There are cases belonging to the class suffering from what Trotter has named "persistent cerebral contusion." As mentioned above, the volume of the intracranial content is increased as the result of the accident. A limited swelling of the brain can be accommodated at the expense of the cerebrospinal fluid, but further increase in size leads to compression of neighbouring veins and capillaries. A localised cerebral contusion by a blow, etc., is characterised by small hæmorrhages and extravasations of blood plasma from damaged neighbouring vessels. The rigid skull prevents absorption, contusion therefore may persist for months or

even years. The patient complains of constant headache sometimes paroxysmal in character ; there may be mental disturbances such as irritability, nervousness, inability to concentrate, and usually increased fatigue. One of the author's patients could not continue to manage the affairs of his shop, although he appeared to be fit and well. This inability persisted for over six months. Such patients require rest, perhaps intravenous medication, sometimes decompression. Bearing in mind what has been said regarding intracranial pressure it is most interesting to find that insufflation of air into the cranial cavity introduced by the spinal route, as practised by Penfield, Boyd, Renfield and Norcross, has given great relief to sufferers from post-traumatic headache. These surgeons believe that such a condition is the result of disturbed circulation of the cerebrospinal fluid.

The third type are those who in middle life or later are suffering from raised blood pressure. It is not difficult to interpret the retinal picture shown by many of these patients. The kinking of the veins where the hardened arteries cross them, the bright burnished copper-wire appearance of the arteries and the slightly enlarged veins. Perhaps where the vessels cross minute flame-shaped hæmorrhages are seen or may be scattered some distance away. There may be small white spots of exudate, chiefly found in the perimacular region or in the centro-cæcal area (see Plate XVIII). These patients frequently complain of headache which becomes more severe as the arteriosclerosis advances. Although the headache in *arteriosclerosis* may last for days or weeks it may clear up at times. The headache is of a bursting type and is usually worse at night. The optic disc in arteriosclerosis is usually normal, but if it becomes raised it is an indication that the arteriosclerotic condition is far advanced and that cerebral changes have become marked. I have never seen a case of arteriosclerosis live longer than six months where the optic disc showed a decided papillœdema. Severe headache accompanied by vomiting may be caused by cerebral angiospasm (Oppenheimer and Fischberg). When the three cardinal symptoms of brain tumour are present, namely, papillœdema, headache and vomiting, it is well to remember that there are other alternative conditions which can produce these, one of which is hypertension. McAlpine mentions a case of a young woman aged twenty-two suffering from chronic nephritis on whom a decompression operation was done by Cushing, who found the brain "soggy

and wet." Fischberg, too, warns us that the retinal pictures in hypertensive neuro-retinopathy may closely resemble what is observed in cases of raised intracranial pressure from other causes.

If papillœdema is absent and the headache is occipital then disseminated sclerosis may be borne in mind.

For our purpose it is sufficient to mention further in connection with intracranial causes of headache that *meningitis* in its various varieties, especially the epidemic cerebrospinal and posterior basic forms, is accompanied by intense headache, the posterior basic form especially causing occipital pain with retraction of the head.

Another cause of headache has been brought to our attention in comparatively recent times. It is the sequel of acute or *subacute serous meningitis* in the posterior cranial fossa. After a feverish onset due to sore throat or influenza sudden attacks of pain are complained of and which are felt in the back of the head and nape of the neck. Voss says there are few positive direct findings; slight stiffness of the neck and tenderness over the occipital region and of the muscles of the neck are apt to give the impression of muscular rheumatism. The pupils may be moderately contracted and nystagmus may be present. Under treatment the symptoms begin to clear up in eight to ten days.

Encephalitis lethargica is usually accompanied by mild headache, but sudden visual disturbances such as diplopia, accompanied by a low-grade irregular fever and headache and a gradually increasing lethargy from which the patient cannot be aroused, form an unmistakable picture (Amoss).

Headache and choked disc may occur in *congenital and acquired syphilitic infection*. Locke has quoted cases, some acquired and one congenital, in which the earliest to show signs of increasing intracranial pressure was six months, the longest after infection was eight years. The condition of syphilitic leptomeningitis is beautifully illustrated in his paper. Fuchs says the condition is most apt to occur within three years of infection. In the secondary stage headache may occur at the same time as the skin eruption. Syphilitic headache increases in severity at night time. In all cases of persistent or intermittent headaches the Wassermann reaction of the blood and cerebrospinal fluid should be examined. The pupillary changes in the eye indicate brain syphilis more than anything else. It should not be forgotten, however, that there is such a thing as

congenitally unequal pupils, or the irregularity may result from enlarged mediastinal glands, encephalitis and brain tumours.

In the nervous system *allergic* reactions take the form of localised œdema and arteriospasm (Kennedy). Migraine is one of the commonest results of food allergy. Recurring headaches, too, more or less severe, may be the result of the same condition ; accompanied by sickness and vomiting the condition may simulate an intracranial tumour but papillœdema is never present.

Among the toxic causes of headache stands out *urœmia*. Vision is affected, often very seriously, this being due sometimes to retinal changes, while at others there may be no such change and yet vision may be so severely affected that complete amaurosis is present. In such cases the pupillary reactions are unaffected. Foster Moore examined 20 cases which later died of uræmia. Of these, 19 had retinitis ; therefore he says, " I am convinced that very few patients die in uræmia without changes in the eye grounds." Although it is the rule for " cotton-wool patches " to appear on the retina in renal disease, yet a pure papillœdema may occur without any exudates being present, thus simulating what is seen in the case of an intracranial tumour. The morning headache in these cases of uræmia may be mild and frontal in location, or severe and vertically placed, or it may be generalised.

Many other cases of endogenous origin of headache occur but we are not concerned with them here. Such exogenous causes as carbon monoxide poisoning, lead, tobacco (especially in women), alcohol and drugs such as salicylates are commonly met with, the proof of which is the cure of the headache on the removal of the cause.

Ocular Causes of Headache

In the attempt to differentiate headache from neuralgia we find the greatest difficulty when dealing with ocular causes of cephalalgia. Some patients can describe accurately the kind of headache produced by the state of their refraction. One young lady in her twenties puts aside her concave glasses in order to attend a dance. Next day she suffers from frontal headache, which is ameliorated by aspirin and cold compresses but which is finally abolished by resuming her glasses. The author prescribed glasses for one of his house surgeons. The amount of astigmatism was not great. One day in hospital the young man was observed to be without his glasses. He had forgotten them, with the result that he was suffering from

a headache. He said the moment he would get home he would put the glasses on and that when he would do so it always took fifteen minutes for the headache to disappear. Where then do patients who are ametropes (those who suffer from an error of refraction in one or both eyes) feel the headache ? Is ocular cephalalgia a reflex pain in the extracranial tissues from the nerves to the orbital contents ? Parsons believes that ocular headache is not reflex in character but is protopathic and is influenced directly by the optic thalamus. Gordon, in a critical analysis of the existing theories and in his own studies, came to the conclusion that the site of ocular pain is extracranial, it is in the muscles of the head. He says the dull bilateral diffuse pain suggests a form of myalgia rather than neuralgia ; the latter is sharp and follows the course of the nerve and its branches rather than its peripheral distribution. He also says that the pain is reflex in origin, analogous to the manner in which reflex visceral pain is caused elsewhere.

Just as idiopathic headache is often hereditary by nature, so also is the cephalalgia produced by ocular causes. Astigmatic children have astigmatic parents, uncles or aunts. Such hereditary characteristics regarding ocular anomalies have been fully described by Nettleship.

While organic headache is not influenced by age, purely functional headache does show definite times of life when it is more likely to happen. There is little tendency for it to occur at either extreme of life. Yet children with errors of refraction do suffer from headache and on this account they are brought to the oculist as early as four and five years of age. Children younger than this seldom complain of headache. Headaches become more frequent at the school age and adolescence, also at middle age when presbyopia begins, but in old age there is comparative freedom from headache. Campbell quotes John Wesley as saying, " I am as strong at eighty-one as I was at twenty-one, but abundantly more healthy, being a stranger to the headache and other bodily disorders which attended me in my youth."

The Semitic stock and the Celtic and Italic groups, according to Gordon, appear to possess on an average greater sensibility to headache than the Slavonic and Teutonic groups.

Following Campbell, we may divide ocular causes of headache into three divisions : (1) retinal irritation, (2) organic disease, (3) eye strain.

To leave London on a dull day in January and find oneself the next in brilliant sunshine at St. Moritz is indeed a wonderful and rapid change, but the enjoyment is often marred by the headache which follows from looking all around at the dazzling snow. Sun glasses are immediately sought for, but as this adds to the difficulties of ski-ing one is often forced to leave the glasses aside. However, tolerance of light quickly sets in but not in every case. I have had to bathe a friend's eye every day for a fortnight with zinc lotion to prevent severe swelling of the eyelids, which was merely a reaction to the suffering produced by the dazzling light on the snow. I have seen a young doctor unable to face ordinary light or even look into a microscope for months. He was suffering from *retinal irritation* due to bright light combined with the fatigue produced by severe studying. The only obvious change found in his retinæ was a slight increase in the calibre of the retinal vessels. There are those who in early childhood are fair in colour ; they suffer from a congenital and familial deficiency of pigment in the hair of the scalp and in the retinal and choroidal layers of the eye. As these children grow older the hair turns darker in colour ; there is also an increase of pigment in the eye which is mainly evidenced by the increase of visual acuity. One observant mother noticed that each of her children became darker in colour before ten years of age and that the eyesight of each improved at the same time. In some cases the pigment fails to be laid down in the eye *pari passu* with that in the hair of the head. In these cases throughout life the choroidal system is easily seen through the retina except in the neighbourhood of the macula and disc. These patients are far removed from the albinotic state but they always remain sensitive to strong light. They have a decided intolerance for bright or reflected light of any kind. A tinted glass gives infinite relief, especially when motoring over chalky roads or while cruising at sea. The patient who is suddenly attacked and is suffering from retinal irritation for the first time shows congestive symptoms ; he appears to have conjunctivitis with accompanying swelling of the lids. There is blepharospasm present and photophobia. The " fair " retinal type does not show congestive symptoms, but at all times does not care for bright illumination. Theatres or cinemas which are in complete darkness are particularly trying to such patients, hence architects should remember that in the construction of such buildings, halls or lounges placed between the outside entrance

and the theatre itself should show a gradual diminution in intensity in the lighting system.

Retinal irritation also is found in nervous subjects or in those suffering from a nervous breakdown or in migrainous patients.

Organic diseases of the eye such as conjunctivitis, iritis, glaucoma, retrobulbar neuritis and proptosis are often accompanied by headache. Conjunctivitis, especially when complicated by the presence of minute corneal ulcerations, is productive of severe headache and intense photophobia. A man over thirty years of age who had suffered for years from a hyperæmic condition of the palpebral conjunctiva was given an eye lotion consisting of boric acid and zinc sulphate. He was to use this as a routine measure. To his intense surprise he discovered that he did not suffer from headache when he used the lotion daily but directly he left off using the lotion his headache returned. He vouchsafed this information of his own accord when he next had his glasses altered.

In iritis severe neuralgic pain is felt not only in the eye and orbit but radiates to the temporal region. Salicylates are often employed in heavy doses in the treatment of iritis, but to some such medication has a most depressing effect; this, coupled with the intense pain from the iritis, has sometimes produced a profound depression the cure of which is to stop the use of salicylates immediately.

In acute glaucoma the pain is felt not only in the eye itself but over all the distribution of the trigeminal nerve. Headache is often extreme and is frequently accompanied by vomiting. Does chronic glaucoma produce headache? The answer is that if the intraocular pressure rises with extreme slowness the probability is that headache will not be felt, nor will coloured rings of light be complained of, but in an intermittent or sharp rise of intraocular pressure headache is likely to manifest itself. To this class belong the cases cited by Wilfred Harris, and such cases have been observed by the author also. As French has pointed out, the occurrence of referred pain in chronic glaucoma without pain in the eyeball is important, in that it may be the first thing to draw attention to the unsuspected ocular disease, and the fact that this pain may be severe is borne out by Harris and others who have found that violent neuralgic pain in the face, cheek and temporal regions was mainly due or aggravated by the presence of chronic glaucoma. Relief by trephining the eyeball is extremely obvious to such patients.

The pain from *retrobulbar neuritis* is felt not only in the eyeball and orbit but sometimes on the forehead and vertex as well. Pushing the eye backwards or rotating the eye from side to side will often increase the pain, but on the other hand, many cases of retrobulbar neuritis do not complain of pain at all.

There are many causes of proptosis, the commonest being due to *thyrotoxicosis*. Headache is frequently complained of by these patients. The prominent eyes are not sufficiently protected from the light so that headache accompanied by lacrimation is often present. Tinted glasses give relief, also the eyebath should be constantly used.

A rapidly growing *orbital tumour* or a *cellulitis* may be the cause of severe headache. Cairns has removed a cholesteatoma which had eroded the roof of the orbit and pushed the orbital contents forward, yet this proptosis of the orbital contents had been going on for a space of eight years without pain of any kind.

Finally we come to *eye-strain*. In the first division of Campbell's classification the second or optic nerve was the seat of origin of the headache or referred pain, but now it is the fifth nerve which is affected. Cephalalgia from eye-strain comes chiefly through the ophthalmic division of the trigeminal nerve. The reflex distribution of pain from ocular anomalies is over the distribution of the fifth nerve. As Leslie Paton has pointed out, the central connections of the first division of the fifth nerve passing down so far into the cervical region may explain the headache felt in the region of the distribution of the great occipital and other upper spinal nerves. Patients continually complain to the oculist of occipital pain. A proper correction of errors of refraction may give relief, but in other cases diathermy to the great occipital muscles alone will banish pain, for such a mass of muscle can easily be the seat of a very troublesome *fibrositis*. The deep connections of the trigeminal with the vagus are most likely accountable for the vomiting which often accompanies acute glaucoma, or the relief of dyspepsia which is obtained by the proper correction of errors of refraction or muscle balance. Again, it is due to the deep connections of the trigeminal, which explain sneezing on suddenly entering a brightly lit room or the weeping which results from extreme laughter.

Because idiopathic neuralgia of the first branch of the trigeminal nerve is so rare Voss emphasises the fact that it should never be diagnosed until all other possibilities have been eliminated.

Whether idiopathic or not the supra-orbital nerves are tender on pressure, but this tenderness is limited to the nerves themselves in neuralgia, whereas it often extends to the neighbourhood when the symptoms are due to sinus disease. Harris speaks of migraine and migrainous neuralgia, the first gradually passing into the second over a space of years. He says if the eyeball and back of the eye are especially affected in migrainous neuralgia it will be found that the attacks are often of briefer duration than ordinary migraine, the pain not lasting more than an hour or so or even as brief as ten minutes. This condition he treats by injection of alcohol either into the supra-orbital or infra-orbital nerve, also deep into the temporal region about $\frac{3}{4}$ inch behind the external angle of the orbit. Quoting Harris : " Whereas ordinary migraine is probably dependent on the vasomotor spasm of the cortical branches of the posterior cerebral artery, migrainous neuralgia is probably associated with vasomotor constriction of the middle meningeal artery and other dural branches, and the pain sensations are therefore carried by the recurrent trigeminal branches which are the sensory supply of the dura. Hence Gasserian block or alcohol block of the supra-orbital or infra-orbital nerves, or even a temporal injection may be sufficient to check afferent pain impulses resulting from the sympathetic vasomotor constriction in the dura. Migrainous neuralgia may therefore be compared to the headache following the intravenous injection of histamine, which has been shown to affect only the one side of the head after trigeminal anæsthesia on the opposite side had been produced either by Gasserian ganglion injection or root resection. On the other hand, stellate ganglionectomy does not affect the production of histamine headache."

The pain of tic doloureux spreads from the face and extends to the dura so that what is usually known as neuralgia of the trigeminal nerve comprises also severe headache. During the paroxysms there is excessive lacrimation with dilatation of the conjunctival vessels. There is pain in the eye and supra-orbital region if the ophthalmic division is involved, also photophobia, lacrimation and salivation. One should be alive to the fact that an organic lesion of the trigeminal nerve or its roots may also cause similar paroxysms of pain. Syphilitic infection of the periosteum or the meninges may be responsible for trigeminal neuralgia or may result from tumours situated in the middle fossa of the skull. Such probabilities as the causes of trigeminal neuralgia should be borne in mind and a diagnosis made

by ophthalmoscopic examination of the fundus for papillœdema ; headache and vomiting should also be inquired after. The possibility of a positive Wassermann reaction should never be forgotten.

Once again we turn to food allergy as a cause of trigeminal neuralgia. Such a case was quoted by Kennedy who discovered that a marine fireman suffered for over seven years. It was found to be due to milk allergy, for the attacks were less often at sea where condensed milk only was to be obtained.

A hundred years ago Jungken describing the symptoms of *eye-strain* termed the condition *habitudo visus*. Blurred vision, discomfort, fatigue, tenderness in touching the eyeball, headache and even extreme nervousness all made up this condition. At first it was thought that there was a defect in the visual apparatus of the eyeball, particularly the retina or optic nerve, then the external muscles of the eye were suspected, but it was not until Donders in 1850 made his discovery of hypermetropia that the muscle of accommodation was discovered to be the principal cause of eye-strain. Donders pointed out that not only local diseases of the eye may be produced by faulty optical adjustment, but also all the symptoms that are recognised as eye-strain, including headache and even indigestion. About ten years after Donders published his famous work on " Anomalies of Refraction and Accommodation," Weir-Mitchell wrote on " Headaches resulting from Overuse of the Brain and also from Eye-Strain." Elliot-Smith has shown what an enormous increase of the cerebral cortex has taken place with the development of exact binocular vision due to the presence of the macular region in mammals, especially man ; the great proportion of nerve fibres in the optic nerve conveying macular impulses and the delicate mechanism evolved for convergence and other ocular movements making man's " visual perception as distinctive as the use of articulate speech to give expression to what he sees and thinks."

Although Derby suggests the abolition of the term eye-strain on account of the widespread involvement of the central nervous system when any portion of the delicate mechanism involved in the visual apparatus is at fault, the term however must be preserved, not only for its historical value, but because eye-strain by no means is not always associated with a neurosis. Ocular neuroses should be kept in a class by themselves. As stated in an earlier chapter, it is in the ophthalmic clinic of a hospital for nervous diseases that these

cases are particularly met with, and much less frequently in ophthalmic hospitals.

An interesting proof of the relationship of headache to sight is given by Walton, who found that of those who were blind from birth 66 per cent. were free from headache, while in marked contrast only 31 per cent. of individuals with sight did not suffer from headaches. Among the blind he found migrainous headaches were only one half as frequent as among seeing persons of like age and condition. Nothing allied to scintillating scotoma could be found among the blind he examined.

The infrequency of headaches in persons with monocular vision as compared with those having binocular vision is due, according to Snell, to the absence of effort the former make compared with the latter who have to strain for fusion through a more complex act.

Eye-strain is found in 7 per cent. of ophthalmic patients, according to Snell, while Gordon says 60 per cent. Of the latter, 86 per cent. were due to asthenopia (refractive error) and the rest to other conditions such as iritis, glaucoma, retinal irritation, uveitis, choroiditis, and congestive disturbances of the lids, conjunctiva or lacrimal apparatus. These are pure ophthalmic cases and are given the desired relief by ophthalmic treatment alone. The situation of the pain caused by ocular conditions is most commonly frontal or orbital ; in the latter case the patient says the pain is in or behind the eyes, while much less commonly the pain is described as occipital or temporal. As Campbell says, " I am inclined to think that when ocular headache attacks either the vertex or occiput it is because there is a special predisposition to headache in these parts, a predisposition which would display itself under other exciting causes." In a small book on " Eye-Strain," written by Ernest Clarke, an ophthalmic surgeon of very wide experience, he said the position of the headache varies with the individual, and also notes that the headache is very often periodic.

The pain may begin as a dull ache, gradually increasing to sharp pain, or may be throbbing or stabbing. The time of onset is usually the late afternoon or evening, that is, at the end of the working day. Sometimes headache is complained of in the morning, and by those who we are sure have not indulged in alcohol the night before. The delayed headache is well exemplified by those who suffer severely at the week-ends, although scarcely reading at all at such times. As Griffith says, " although the pain of eye headache may

radiate through any of the trigeminal branches the initial cause is painful contraction of the muscles." Those muscles concerned may be the ciliary muscles, the extrinsic eye muscles and the occipito-frontalis. The reader may remember cases in which he discovered that too strong concave glasses were being worn, causing intolerable pain, or again it was discovered by experience that using 1 per cent. eserine after examining the refraction of a patient under a mydriatic gave certain patients such pain that they were temporarily disabled. In both cases the pain results from over-action of the ciliary muscle.

In an analysis of 300 cases Snell gave the following proportion of those suffering from eye-strain causing headache to be as follows : simple and compound hypermetropic astigmatism 49·3 per cent., hypermetropia 34·3 per cent., simple and compound myopic astigmatism 11·3 per cent., mixed astigmatism 3·6 per cent., and myopia 1·3 per cent. To these Gordon would add 20 per cent. due to muscular insufficiency. The statement that muscular anomalies are the chief causes of ocular headache is quite untrue. Each case of refraction done by the author is accompanied by a muscle balance test, and he would say that 20 per cent. is quite a generous figure. The remarkable thing in connection with the amount of ametropia present is that the smaller amounts are productive of the greatest discomfort. I have seen half to three-quarters of a diopter of hypermetropia corrected by glasses with the utmost relief to those whose calling confined them to an office desk all day. In the case of larger errors of refraction the patients do not struggle to overcome their disabilities and so ciliary strain is not produced, whereas in the smaller errors the eyes are attempting all day long to compensate for them by abnormal ciliary exertion. In a large school where over 400 children are cared for and where headaches used to be a common complaint, now, since the eyesight of each child is examined every eighteen months, headache is rarely mentioned. So evident is this that directly a teacher learns of a scholar suffering from headache he is immediately referred to the oculist. In many of these cases the correcting lens is not more than one to two diopters. The school child suffering the greatest discomfort is the one who has never worn glasses and is suffering from hypermetropic astigmatism with obliquely placed axes. I have even observed epileptiform attacks occurring in such children ; these attacks were abolished by the wearing of proper glasses. Two cases have been seen by the author where the complaint was made that reading or sewing without pain was impossible.

In each case homatropine was used and in neither was any error of refraction found, yet both patients returned to express their grateful thanks for the relief afforded by the use of the " drops." One of these cases, a doctor's wife, has had no pain now for over two years. These cases were not neurotics, their relief came undoubtedly from the abolition of ciliary spasm.

Referring to those cases of cephalalgia caused by muscle imbalance, we must bear in mind that the elevation and depression of the eyes are performed by a mechanism more complex than that used for the lateral movements, and as everyday movements are chiefly lateral in character when vertical movements are called upon in excess of what is ordinarily required pain is the result. The " Academy headache," referred to by Campbell, is a common experience and is caused by the usual elevation of the head together with an unusual amount of ciliary effort in perhaps a somewhat trying illumination. A considerable number of patients exhibit an error of half a prism diopter of hyperphoria ; usually this does not trouble them, but when the elevator and depressor muscles of the eyeballs are called into play continuously for several hours then the error makes itself evident. Excessive lateral movements can produce pain as is commonly experienced by those suffering from " Wimbledon nystagmus." Peter refers to the " panorama headache " and says it is caused chiefly by muscular asthenia, particularly of the hyperphoric variety and not due to ametropia.

The whole world is indebted to a very modest man who lived and worked in Bournemouth for over fifty years. I refer to Maddox, whose writings on the extraocular muscles have stimulated one and all to a better understanding of the principles of refraction in everyday practice.

There is a type of headache which follows operations on the frontal sinuses, in the healing of which some of the muscles situated superior to the eyeball have become involved. Diplopia follows and the images are sometimes set obliquely to each other ; the simultaneous use of both eyes has to be abandoned or if persisted in neurotic symptoms develop. Such cases bring before the oculist a problem which is truly difficult but which should not be despaired of. Repeated operations, elevation, depression or even torsion may be tried and when success follows such efforts the reward is great—particularly when the sufferer is one on whose sight his living depends.

The final muscle condition to which I will refer is that of the occipito-frontalis. The frontales muscles are inserted into the upper lids and each participates in the contraction of the orbicularis palpebrarum. The frontales on contraction pull on the epicranial fascia and this in turn pulls upon the occipital muscles and causes occipital pain. Patients suffering from this type of headache must be given correcting glasses and taught not to screw their lids together. In addition to ocular causes of occipital pain many other conditions might be mentioned but one quoted by Griffith is worth recording. In a case of Lindau's disease he had been observing the angiomatosis of the retina, when the first and only indication that the patient had developed a hæmangioma of the cerebellum was the sudden appearance of severe occipital neuralgia.

Amaurosis

The word amaurosis means total blindness when no change can be seen in the eye sufficient to account for it. Another term—amblyopia—is partial loss of sight often without any departure from the normal appearance of the retina also.

We have already seen that certain poisons such as tobacco, alcohol, etc., can cause a central diminution of vision, but amblyopia from non-use occurs when there has been an obstacle to vision, present since childhood, in the cornea, pupil or lens of the eye. A similar amblyopia occurs in the squinting eye of a child, and such reduced vision begins to take place very early ; indeed, amblyopia is frequently established at four years of age, making it impossible to produce any improvement by orthoptic training during early school days. Indeed, the nearer the age of seven years the less likely is there to be any change in the amblyopic state of the eye. A squinting eye therefore cannot have its refraction corrected too soon—even five or six months old is not too young for glasses to be fitted and worn. If at the age of three years the squint is not corrected it should be operated upon. Children of four years of age operated upon for squint do extremely well. It might be said that this condition of amblyopia need not interest the neurologist but it does. If, as is so frequently done, a cover is placed all day over the good eye of a child of six or seven years of age in order that he should be compelled to use the amblyopic eye, such a procedure often makes the child quite nervous for he is partially blinded, and

if improvement does not take place the child becomes far from well. Leaving the good eye uncovered for part of the day in the attempt to restore vision to an amblyopic eye is utterly useless. A compromise is impossible.

When the sense of strain and weariness in the eyes and head is set up by the use of the eyes the condition is known as asthenopia. It is not, however, a purely physical state. Most observers agree with Best that such a condition is a manifestation of a psychogenic state and that it is indeed only a halfway house to partial or complete amaurosis. Friedenwald suggests special correction for anisophoria (a form of heterophoria in which the degree of muscle imbalance varies with the direction of the gaze) to relieve persistent discomfort in one eye only; while Stutterheim states "asthenovergence" is a principal cause of asthenopia and describes fully a method of kinetic treatment for the removal of such symptoms of eye-strain.

From time to time throughout this book we have read of blindness resulting from an organic lesion in some parts of the central or peripheral nervous system. We have also mentioned such a condition as uræmic amaurosis, but at the moment the subject which claims our attention is the amaurosis which is purely functional in character. Both the oculist and neurologist meet with these cases and pass them one to the other or send them on to the psychologist, for sometimes it is difficult to determine whether these patients are suffering from physical or mental ill-health. Farquhar Buzzard, recognising that the ophthalmic surgeon is brought into contact with a considerable number of neurotic and psychoneurotic patients, says that fear and anxiety are responsible for a great number of such patients seeking advice.

Amaurosis is commonly met with in hysterical conditions; the word hysterical, however, does not cover all those cases of amaurosis, so-called, which present themselves for treatment. Three types of cases are exemplified by the following. The first case, that of a young lady whose life in a large continental city was filled with amusement and gaiety of all kinds; her days were spent in riding, fencing, tennis and so on, but suddenly a crash occurred in the family fortunes, one result of which was that this young woman found herself in London where her daily exercise and diversion was an hour's walk in a park with an elderly female relative. In the girl's demeanour nothing amiss was apparent until one night she

switched on the light in her bedroom and found all was dark. She groped her way into another room and was then gently led back to bed. In the early hours of the morning vision had partly returned and by 10 a.m. full vision was restored. A few hours later the patient was seen and examined, but nothing abnormal was found in the state of each fundus and vision was of normal standard. The second case was that of a girl aged fifteen years, healthy and active, performing her full amount of school study and play as well. Suddenly she too complained of loss of vision. Her father brought the patient immediately to the oculist who had examined her eyes previously since she was eleven years of age. Her vision normally was 6/5, now it was 6/60 in each eye. A mydriatic was instilled but nothing abnormal was found in the fundus of each eye. Perhaps somewhat crudely the condition was put down as brain fag; rest in bed was ordered and in a few days full vision had returned. That was seven years ago and since then there has been no recurrence of the lowered state of vision, the condition of the girl's health being as robust as ever. The third case which one would like to quote was that of an officer who was hard at work in a casualty clearing station X-raying cases. It was during the attack and breaking through of the Hindenburg line. One thousand wounded a day poured into this particular casualty clearing station. After a few days' heavy work this officer on entering the mess and sitting down to lunch noticed that from his usual seat he could not see on his right hand a door opening and shutting which was normally within his peripheral field of vision. To his astonishment he could not see anyone entering or leaving unless he turned his head to look. With a slight feeling of nausea he suddenly realised he was suffering from a right-sided hemianopia. He left the table and proceeding to his tent lay down for a few hours. Upon rising he found his vision was again normal. That was eighteen years ago and no abnormal manifestation of anything physical or mental has taken place since. Neither the young girl nor the officer had experienced the "baulking of fundamental instincts such as self-assertion, self-preservation or sex." They were just mentally tired. These are the cases which must not be classified as hysterical, although one might include as such that of the young woman mentioned in the first instance. Space does not permit one to enlarge much further on the subject of amaurosis. Best has summarised very thoroughly the subject of hysterical amaurosis, including its treatment. An example of this

condition was described by Horgan. He said a young girl aged sixteen years was led into his consulting-room walking with tentative steps and with both arms outstretched. The eyes were wide open and staring vacantly straight forward. The history was that on fleeing from an angry relative she was knocked down by a motor car and immediately after was found to be blind. The pupillary reactions were normal and ophthalmoscopic examination failed to reveal any evidence of organic disease. The result of a sympathetic talk and application of the faradic current was a rapid restoration of vision. Horgan, however, wisely advised a change in environment which entirely prevented a recurrence of trouble. It is interesting to note that in a case of hysterical colour-blindness recorded by Williamson-Noble colour vision was restored by faradism accompanied by a suitable suggestion as to its therapeutic effect. Although it is common to find no change in the reactions of the pupils in hysterical amaurosis, yet in some cases it has been recorded that the pupils were contracted and practically immobile (Ellis), which was possibly due to a state of spasm of accommodation being present.

On going to print, a further case of diminished vision in a little girl of seven years of age has been brought to the author's notice. The mother observed this child holding her book close to her as she read. Being concerned about the probability of the onset of short sight she brought the case for investigation immediately. Under a mydriatic the normal amount of hypermetropia for her age was found, but no correction could raise the vision above 6/60. By the confrontation method both fields of vision were found to be greatly reduced, so perimetric estimation was proceeded with. The result showed both fields reduced to between 10 and 15 degrees from the point of fixation. Each fundus appeared normal. She was then examined by a neurologist and an X-ray of the skull was taken. All proved negative. The child was taken from school, where she had been exceedingly keen about her work, and some tonic treatment was given. In six weeks the vision of each eye was 6/5 and full normal fields of vision had returned.

This is the youngest case of its kind that has come to the author's notice.

To those who may be interested in malingering, Jones and Llewellyn have written fully on the subject of simulation of disease. With reference to sight, however, they have included a special

chapter by W. M. Beaumont, who classifies amaurosis and amblyopia together as follows—

- Amaurosis and Amblyopia
  - True
    - Physical
    - Psychical
      - Hysterical,
      - Psychogenic, but not hysterical.
  - False
    - Entirely false
    - Partly false

and says, "The variety of amblyopia which is sometimes called 'functional' is seen, with certain points of difference, in hysteria, neurasthenia, and in the traumatic neuroses, and in all so uncertain, so unstable are the symptoms that they counterfeit inveracity and simulate simulation."

Neuro-Ophthalmological Examination of the Eyes

(1) Appearance:
- Symmetry of orbits, eyelids and eyes.
- Movements of eyelids, including frontales, ptosis (apparent or real), proptosis.
- Muscular movements, squint (concomitant or paralytic), nystagmus.
- Associated movements.
- Colour and texture of the irides.
- Form, size and position of pupils.
- Pupillary reflexes, direct and consensual.
- Effect of mydriatics, cocaine.

(2) Visual acuity and fields of vision:
- Scotomata, peripheral contraction, including that for form and colours.
- Evidence of night blindness.

(3) State of Muscles:
- Evidence of diplopia from muscular paralysis.
- Muscle imbalance due to paresis, using Maddox rod and Maddox wing test.
- Convergence and accommodation.[1]

[1] The power of accommodation in a child of ten years of age is such that it can see objects clearly at 2½ inches (7 cm.) from the eye, at twenty years of age the near point has receded to 4 inches (10 cm.), at thirty years of age to 5½ inches (14 cm.), at forty-five it has receded to 11 inches (28 cm.), while at sixty it is at 40 inches, or 1 metre (Donders).

(4) Sensation :
Skin sensibility.
Corneal reflex.
Ocular pain.

(5) Examination of the eye :
State of conjunctiva.
Transparency of media.
Appearance of optic nerve, macula, retina and blood vessels.

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