A CASE OF ORBITAL ENCEPHALOCELE WITH UNIQUE MALFORMATIONS OF THE BRAIN AND EYE.

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THE following case shows extremely rare congenital abnormalities of the eye, and an apparently unique malformation of the brain.

Alice B. was brought to one of us (J. H. P.) during the first week of life. The parents are both healthy; the mother is 21 years old, and this is the first child; there have been no miscarriages. There is no history of malformations, &c., on either side.

The child was healthy and well nourished, born naturally at term. At birth a large dark swelling was noticed projecting from the right orbit. The child was kept under continual observation as an out-patient at the Children's Hospital, Great Ormond Street, for seven months, when it was operated upon. The condition did not vary materially, though there was some increase in the tumour. From the mother's description it would seem to have been especially prominent and dark soon after birth.

Fig. 1 shows the condition of the child when 7 months old. There was a large cyst-like mass in the right orbit, which pushed forward the lids, so that they were stretched and enlarged. The orbit was enlarged, especially in the outer part, the upper border extending upwards here so that the

vertical diameter measured $1\frac{1}{2}$ inches as opposed to $\frac{3}{4}$ inch on the left side. The eye was displaced down and out; it was usually covered completely by the lids, which were kept closed.

The tumour caused two separate swellings, pushing forward the upper and lower lids; the conjunctiva looked bluish, but not markedly so, and was otherwise normal. Beneath the lower lid the tumour felt nodular, reminding one of the feeling of a plexiform neuroma. Deep palpation failed to reveal any gap in the orbital bones.

The right eye was considerably smaller than the left, though it otherwise showed no obvious external anomaly; the cornea measured 9.5 mm. in diameter. On ophthalmoscopic examination, after dilatation of the pupil, considerable abnormality found in the fundus. The fundus was difficult to examine, and the interpretation of the results was also difficult. There was a coloboma of the optic nerve. Outside the disc there was a very marked parallax. A pale, more or less triangular, patch was seen outside the disc, with retinal vessels running over it. There was no typical coloboma of the choroid.

The left eye was normal. There were no other congenital abnormalities. There was no swelling over the temporal region such as is usually associated with plexiform neuroma. The palate and mouth were normal, and suction was perfect.

The case was shown at the Ophthalmological Society when the child was 13 weeks old. A variety of suggestions were made as to the nature of the tumour, *e.g.*, meningocele or meningo-encephalocele, hæmangioma, lymphangioma, plexiform neuroma, congenital cyst of the orbit associated with microphthalmia. Against meningocele was the fact that at this time there was no definite pulsation. The colour was against hæmangioma, and lymphangioma is so rare in the orbit that such a diagnosis was improbable. The general character of the mass and the absence of swelling over the temporal region made plexiform neuroma unlikely. The absence of a coloboma of the choroid and the relatively large size of the eye made congenital cyst associated with microphthalmia also improbable.

On the possibility of its being angiomatous, and from the fact that Sir Victor Horsley has obtained improvement in meningoceles with this treatment, electrolysis was tried. During the operation, clear fluid escaped and continued to do so for some time. No beneficial results followed. Some fluid was drawn off with a hypodermic needle and tested for reducing agents. It did not reduce Fehling's solution, but the amount obtained was small.

In the course of the next three months the child thrived, but the swelling increased in size and the disfigurement became very great (fig. 1). There could no longer be any doubt about pulsation in the tumour. The fact that meningocele without encephalocele is practically unknown in the orbit (*see* Parsons, "Pathology of the Eye," vol. ii., p. 721) led to the definite diagnosis of a meningo-encephalocele. The correlation of this condition with the ocular abnormalities caused some difficulty, which is only partially elucidated by the anatomical examination.

After consultation with my colleague, Mr. Kellock, I (J. H. P.) determined to exenterate the orbit, hoping that I might be able to close the aperture in the skull by stitching the meninges over it. The whole of the contents of the orbit were removed, including the eye and a mass of brain. The meninges were adherent to the globe and were so much attenuated that it was found impossible to close the aperture in the skull. During the following days there was a large amount of leakage of cerebro-spinal fluid, the dressings and pillow being soaked. The child died on the fourth day.

At the *post-mortem* examination the cranial cavity was found to communicate with the right orbit by a large hole, 10 mm. in diameter, below the lesser wing of the sphenoid; it obviously represented the combined optic foramen and inner part of the sphenoidal fissure.

The mass of brain removed from the orbit, the whole of the inner part of which it occupied, measured 42 mm., by 25 mm., by 18 mm. It had a somewhat convoluted surface, was very congested, and contained a space filled with what looked like broken-down cerebral substance. Micro-

scopic sections show that it is made up of brain substance containing large cells resembling those found in the cortex (fig. 2).

The condition of the brain is shown in the accompanying photographs (figs. 3 to 6). Fig. 3 shows the left side, which is normal. Fig. 4, showing the right side, demonstrates the presence of a large mass lying under the right frontal lobe, which it presses upwards, and in front of the right temporo-sphenoidal lobe, which it presses backwards. Fig. 5 represents the appearance of the brain from below, with the mass *in situ*. The mass, which measures 8.5 cm. by 8.4 cm. by 3.0 cm., is covered by pia mater and is attached to the rest of the brain by a pedicle of pia mater, the third nerve, which passes through it, and by blood vessels. The surface is almost smooth, but here and there, especially on the inner side, there are shallow vertical grooves. The upper surface is quite smooth; on the inferior surface, near the centre, there is a rough patch from which the pedicle of the intra-orbital portion sprang.

The brain proper is apparently quite normal, apart from the results of pressure. It will be noticed (fig. 6) that the orbital convolutions, as well as those of the frontal lobe (fig. 4), are well marked. The optic chiasma is intact. The optic nerves are elongated and flattened, but contain normal nerve fibres. The mass is of course anterior to the chiasma.

The intracranial mass is a solid block of brain substance. On cutting it through in a sagittal direction it is seen to be covered by a thin layer of white matter, under which there is a thin lamina of gray matter, then another thin layer of white matter, and finally a large core of gray matter. Microscopical sections show that it consists of brain substance of the cortical type (fig. 7). We have not succeeded in working out any definite analogy in the form and structure of the cells with any particular part of the cortex or basal nuclei.

We have been unable to discover the record of any case similar to the above in neurological literature. We sent photographs to Professor Cunningham, of Edinburgh, who

was so good as to inform us that he had never seen a similar condition, nor met with any reference to anything like it.

There are two theories which might account for the development of a large isolated mass of brain substance such as that described. It is possible that it might be the teratological development of a second brain. It is more probable, in our opinion, that at an early period in foetal life a small portion of potential cortical brain substance was separated more or less from the main mass and grew and developed independently, finally becoming completely isolated. Professor Cunningham suggests from the absence of any central cavity and the position of the stalk of pia mater—near the anterior perforated space—that there was originally some connection with the caudate nucleus region.

Pathological examination of the eye. — The eye was hardened in formalin, divided horizontally, and cut in serial sections.

Macroscopical examination. — The globe is small, its diameters, not including the cysts, being as follows: antero-posterior 18 mm.; transverse 19 mm.; vertical 16·5 mm. The transverse diameter of the cornea is 9·5 mm. The anterior half of the eye is normally formed. The posterior half is distorted and flattened upon its inner and lower aspects, and shows the following abnormalities (fig. 8): (1) An adhesion of the dura mater of the encephalocele to its inner and lower surfaces, corresponding with the area of flattening; (2) a very thin-walled cyst which springs from the junction of the nerve with the globe on the inner side; (3) a cystic bulging separated from the latter by a constricted neck which seems to involve the nerve itself.

(1) The dura mater, which is adherent to the sclera, is thick and tough. The adhesion is extremely dense so that it is wholly impossible to strip it off. It covers most of the inner and lower aspects of the posterior part of the globe, and becomes lost indefinitely over the cysts behind. Its exposed (cerebral) surface is smooth and glistening, and the same appearance is evident in the external lining of the cysts.

(2) The more anterior of the two cysts forms an almost

perfect hemisphere about 5 mm. in diameter. It is so thin that it appears bluish, and its wall is indented with the slightest touch. It rises up rather sharply from its surroundings, and is seated at the point of junction of the nerve with the globe. In position it is directly on the horizontal nasal meridian.

(3) The more posterior of the two cysts is separated from the other, and from the posterior part of the globe by a constricted neck. Its greatest transverse diameter is 6.5 mm., and its walls are not so thin as those of the more anterior cyst. From a point somewhat below its posterior pole a thin, but apparently normally-formed nerve projects. The cyst appears therefore to be an expansion of the nerve as a whole with spreading out of the nerve fibres over its surface. It will be seen from the microscopical description that this appearance is partly fallacious.

On dividing the globe the most evident abnormality is an exceedingly deep pit or funnel in the position of the optic disc. The retina has become detached in the hardening fluids so that it stretches across the mouth of the more anterior of the two cysts and partially conceals it. There is nowhere any evident defect of the retina. On all sides it can be seen to reach up to and pass over the edge of the pit. This edge is very sharp and prominent on the temporal side, more shelving on the nasal. The retinal vessels emerge chiefly on the nasal side in several isolated trunks, and the papilla seems to be stretched out over the wall on this side. The cyst ends far back in the nerve in a rounded fundus, but, as only a small circular piece was removed from the upper part of the globe, this could not be very closely examined. All the anterior parts of the eye are normal, and the large white patch visible with the ophthalmoscope on the temporal side of the disc is not visible, being concealed by the artificially detached retina.

Microscopical examination.—Three main abnormalities are found which may be conveniently described separately. (1) A cystic bulging on the nasal side involving all the tissues of the globe, and corresponding with the more anterior of the two cysts seen macroscopically (figs. 9 and 10, A).

(2) A large coloboma of the nerve entrance on the temporal side corresponding with the posterior cystic swelling of the nerve (figs. 9 and 10, *B*). (3) A defect of the pigment epithelium in the macular region corresponding with the white patch seen with the ophthalmoscope in this situation (figs. 9 and 10, *C*).

(1) Over this area the sclerotic and choroid are very greatly thinned. Immediately in front of it the sclerotic is

FIG. 10.

thicker than normal, owing to the fusion of the dura mater of the encephalocele with it. At the edge of the cyst it thins off rather suddenly so as to consist of only a few fibrous trabeculæ. It remains of fairly even thickness over the whole area, and posteriorly abuts directly on the nerve. It is lined on its inner surface by a layer of somewhat looser tissue which is continuous with the choroid anteriorly, and is very much atrophied in places, and not to be followed with

certainty as a continuous layer. On its inner surface, again, the pigment epithelium is found. Over the summit of the cyst the cells are partially depigmented, but it can be easily followed as a continuous layer in all the sections; if absent at all it is only in minute spots and patches. The retina proper is also uninterrupted, and is quite normal in structure. It is somewhat folded and has become partially detached in the hardening fluids, but there can be no doubt that during life it lined the excavated area.

(2) The larger cyst is a so-called "coloboma of the nerve entrance," but not an excavation of the nerve proper. On each side the sclera and choroid come to their normal terminations (figs. 10, *t* and *n*). This is especially well seen on the temporal side where the intervaginal space (fig. 10, *v*) gives a good rallying point. Just within it the sclera (*s*) terminates in a sharp promontory; the choroid (*c*) ends in the same position, and the retina (*r*) a little farther in, being dragged round into the excavation for a short distance. On the nasal side the intervaginal space is left open, and therefore does not furnish so good a point for orientation; but the lamina cribrosa (*e*) gives the indication required. On this side also the sclera, choroid and retina end in practically normal relations to one another. There is therefore no coloboma outside the normal limits of the porus opticus. It is in fact that small portion of the scleral promontory which immediately adjoins the lamina cribrosa and lies over the termination of the intervaginal space and to the inner side of that spot which has given way, or perhaps more properly, has never been normally formed.¹

All the abnormalities found in the nerve entrance follow from this giving way of the sclera at its margin (*see* figs. 11 and 12). Since the bulging occurred to the inner side of the termination of the intervaginal space, the latter is not displaced, though spread out over the surface of the cyst. In especial its anterior blind end retains its normal relations. On the other hand, since the bulging is to the temporal side

¹ For convenience of description we shall continue to speak of the area as having "given way," though it is probable that the essential factor is defective development.

of the nerve itself, the latter has become carried over as a whole to the nasal side. In the normal papilla the lamina cribrosa arises from the area in question, but in the present instance it has not shared in the bulging, but has been displaced bodily to the nasal side; and since the relations of the nerve and sclera remain normal on that side, the lamina has become slewed round in the process, so that the surface

FIG. 11.

FIG. 12.

of the papilla looks temporalwards. Theoretically there should be a connection between the papilla and the retina on the temporal side, and in fact some greatly distorted remains of retina are to be found in the excavation (r'). Owing to the great distension, however, they have in places become completely atrophic, so that no continuous nervous layer can be traced. Corresponding with this fact the

nerve-fibre layer of the retina is highly atrophic everywhere on the temporal side, but well preserved on the nasal.

The nerve has not simply been displaced by the excavation, it has also been hollowed out. That is to say, it does not simply run over the wall of the cyst in the form of a cord, but has been spread out over a large area of its wall. Hence, in any given section, in addition to the thin nerve which runs round the nasal side of the cyst (*o*), transversely cut nerve fibres are found for a considerable distance towards the temporal side (*o'*), and in peripheral sections the nerve has the appearance of being split up its centre. These transversely-cut fibres are merely fibres mounting over the wall of the cyst to reach the lamina at some point above or below the plane of section; they do not enter the globe on the temporal side. It will be noted, therefore, that although the nerve is excavated, there is nothing analogous to a glaucomatous cup. The nerve is hollowed out by invasion from the side, not by pushing back of the lamina cribrosa. The lamina, as before stated, is not involved in the bulging, but merely displaced bodily to the nasal side and slewed round so as to face temporally; it has no convexity backwards.

Together with the process of distension of the sclera a process of thickening of its fibrous tissue has gone on. Hence the fibrous wall of the cyst, in spite of its considerable size, is not thinned, but very considerably thickened. Within this thick pad of connective tissue, which lines the excavation, a few distorted remains of retinal tissue are found on the temporal side (*r'*).

The nerve is atrophic, but well formed. The central vessels pass up the middle, and enter the globe as usual in the centre of the lamina cribrosa.

(3) In the macular region there is a defect in the pigment epithelium over an area about 3 mm. in lateral, and probably about the same in vertical diameter (*c*). The cells cease quite abruptly on each side, but there are some small patches of depigmented epithelium within it. The choroid generally on this side is thin and poorly developed, but there

is no defect in it—it can be traced as a continuous layer over the area in question. The retina also shows no gap. It is not quite normally formed, showing some thinning and irregularity of its nuclear layers, but these changes are slight, and a fovea is quite distinctly recognisable (*f*), as shown by thinness of the whole retina, relative thickness of the ganglion cell layer, and the presence of cones alone. It is situated towards the temporal side and somewhat above the centre of the gap. As before mentioned, the nerve-fibre layer is almost absent on the temporal side, but well formed on the nasal. The ganglion cell layer is present, however, and all the other layers are practically normal, except for a few oedematous spaces in the inner nuclear and internuclear layers.

As already stated, the dura is intimately adherent to the sclera on the nasal side. Microscopically it is so intimately fused that only a very indefinite line of demarcation can be made out except where it is reflected from the sclera. It is lined on its outer (cerebral) side by a thin layer of tissue having the structure of arachnoid (*ar*), and tracing this backwards it is found to line the thin cyst on the nasal side externally, and to be continuous with the arachnoid lining of the nerve. There is therefore no intervaginal space on this side, and the nerve has no dural sheath. It is not easy to say how this has come about, but the most probable explanation would seem to be that the dural sheath of the nerve (like the sclera farther forward) has become intimately adherent to the dura mater of the encephalocele, and has subsequently been dragged away from the nerve, leaving the intervaginal space open. Or, possibly, the dural sheath has never been properly formed on this side. Or, again, it may have been dissected off at the time of operation, though the lack of any stump of dural tissue (whereas one is present on the other side) renders this improbable.

All the anterior structures of the eye are normal. The corneo-iridic angle is somewhat of the foetal, or infantile, type, with a few trabeculae of the ligamentum pectinatum crossing it, but probably not more so than corresponds with the age of the child.

This interesting globe shows no fewer than three different congenital abnormalities, each of which is most unusual.

(1) With regard to the staphylomatous bulging on the nasal side of the nerve, its position is its most notable feature. "Colobomata of the choroid" in any position except downwards and in the macular region are exceedingly uncommon, and not a few of the published cases suggest patches of old choroido-retinitis rather than true congenital abnormalities. They have, however, been seen outwards (Adams Frost [1]—a case with a coloboma of the iris also outwards), and upwards and outwards (Rindfleisch [26], Nuel and Leplat [21], Nuel? [20], Fuchs [8]). It is worthy of note that atypical coloboma of the iris may be associated with coloboma of the choroid in the usual position (v. Mittelstädt [22], v. Reuss [25], Hess [14]). We have only been able to find two cases of inward coloboma. In one of these, recorded by Randall and de Schweinitz [24], there was a deeply excavated, bluish-white defect one and a half times the diameter of the papilla, and situated inwards and a little downwards from it. In the other recorded by Steinheim [28], there was a large inward coloboma, engulfing the papilla, with a coloboma of the iris in the same position. We believe the present case to be the first atypical coloboma (apart from macular colobomata) submitted to pathological examination. It was not found to differ in its essential microscopical details from colobomata elsewhere. As has frequently been found in other cases, especially in eyes obtained shortly after birth, there was no actual defect in any layer, but only a considerable stretching and thinning. In particular the retina was perfectly normal except for a degree of atrophy of the nerve fibre layer, which was probably to be attributed to the condition of the nerve entrance.

(2) Several cases of coloboma of the optic nerve entrance have been examined microscopically. They fall usually into one of three classes. The most common are colobomata of the choroid reaching up to the nerve, and are therefore not true defects of the nerve itself. When these become ectatic the lower border of the nerve becomes displaced upwards and backwards, so that the surface of the papilla

comes to face downwards, exactly as in our case it came to face temporally. Naturally in such cases the dural sheath and intervaginal space go with the nerve and are not involved in the coloboma (Manz [18], Bock [3], Görlitz, [11], Devereux Marshall [19], Knapp [15], Tschermossow [29], v. Duyse [5]). The second form includes cases in which a pocket of retina is snared off just at the lower edge of the papilla, and projects into the sclera (Liebreich [16], da Gama Pinto [9], Manz [18], Bach (Case 1) [2], E. v. Hippel [12], v. Duyse [5]). Usually, if large enough to appear externally, it comes out below the dural sheath, so that in this case, as in the last, the vaginal space is not interfered with, but is displaced bodily with the nerve. Insensible gradations are found between this type and the well-known cases of microphthalmia with extraocular cysts containing distorted retina. These two forms of coloboma of the nerve may be combined and both have been found in the condition described by Fuchs, and known as inferior crescent or conus—a condition which is congenital, and usually associated with a considerable degree of amblyopia. A third, and much rarer, form of coloboma is an actual defect in the nerve itself, and lamina cribrosa being imperfectly formed (Ginsberg [10], Görlitz [11], Bach [2]).

The present differs to some extent from all these cases. In the first place, the position of the coloboma is quite peculiar, being on the temporal, not on the inferior side of the papilla. Again, the choroid is not defective. It is the point of junction of the sclera with the lamina cribrosa which has become ectatic. The ectasia has therefore taken place to the *inner* side of the intervaginal space, and the latter has not been displaced with the nerve, but has retained its normal position. The theoretical stages through which this has come about are shown in figs. 11 and 12, while fig. 10 represents the actual state of affairs found. The only reference to such a condition with which we are acquainted is contained in a paper by Elschnig on inferior conus. He found the tissues between the "sclera-dura" and the nerve ectatic and bulging into the intervaginal space, but, as this occurred in a very myopic globe, it must be doubtful

whether the condition was congenital or due to myopic expansion.

(3) The case furnishes a good example of a central or macular coloboma. As in the coloboma on the nasal side of the disc so here, there was no actual gap in any layer though there was defective formation. In the retina there was even an imperfectly-formed but quite recognisable fovea. This has not, we believe, been previously found in a macular coloboma. The most defective layer was the pigment epithelium. It was present only in islands and had entirely lost its pigment. It should be noted that neither here nor in the other colobomata was there the least evidence of old or recent inflammation. Indeed, the absence of all adhesion between the retina and the choroid is direct proof to the contrary. Hence the case adds to the already very large amount of evidence against Deutschmann's theory that all colobomata are due to inflammation in early foetal life. There was also no bulging of the ocular tunics. It seems evident, therefore, that the adhesion between retina and choroid, the extreme atrophy and distortion of these layers, and the deep excavation which have usually been found in pathological examinations of macular and other colobomata, are secondary phenomena, due to subsequent inflammation, or to stretching and giving way of a weak part of the globe. It is highly probable that they were not present in this instance because the eye was obtained from an infant, and therefore before secondary changes had occurred. Confirmation of this view is obtained from a case reported by Pause, in which the retina and choroid were both quite intact over a coloboma of the choroid, while the pigment epithelium was present, but deprived of its pigment. The case, therefore, closely resembles ours, and it occurred in a child which died at birth. Depigmentation of the pigment epithelium has also been noted by Bock and others over typical colobomata of the choroid. The changes more commonly found in macular colobomata are described in the papers of Hess [13], Bock [3], Deyl [4], and v. Duyse [5].

It seems evident that in this case the abnormalities

present in the eye were not independent of the orbital encephalocele. Had the colobomata been of the ordinary type it might have been possible to suppose that they were one expression of a general tendency towards the formation of congenital abnormalities, while the brain tumour was another expression of the same tendency, just as, for instance, supernumerary digits on the hands and feet are frequently associated, yet not dependent the one on the other. In view of the fact, however, that the brain masses in the orbit directly pressed upon and flattened the globe, and since two at least of the colobomata were of quite an atypical variety, it seems much more likely that they stood in the relation of cause and effect. Moreover, microphthalmia with colobomata has been found associated with contraction of the orbit from hydrocephalus (Rindfleisch, Hans Virchow).

A possible explanation would be that the apparently isolated mass of brain substance had been snared off at an early period of foetal life, and had interfered with the proper budding out of the primary optic vesicle, or had even been derived from a portion of brain substance which should have gone to the formation of the vesicle. It seems highly improbable, however, that if this had been so the globe would have been so well formed, especially in its anterior parts. Since there is no defect below either in the nerve or retina, it is evident that a secondary optic vesicle also formed and closed in the usual manner. We believe it to be certain therefore that the defects in the globe were caused by the pressure of the growing brain masses on the growing secondary optic vesicle.

The explanation of colobomata which do not occur in the line of the foetal cleft is always a matter of difficulty, but in the present instance this is not so great as usual. It was evident that the mass entering the orbit from the inner side had chiefly pressed upon the nasal side of the globe. This was shown by the marked flattening of the sclera, and intimate adhesion of the dura to it on that side, as well as by the marked displacement of the eye to the temporal side. The encephalocele had a lobulated surface, so that its

pressure must have been unequal, and it is not improbable therefore that the ectasia on the nasal side of the nerve was formed by the giving way of the ocular tunics at an unsupported spot between two lobules. The junction of the sclera with the lamina cribrosa is in any case a weak spot, and possibly the deep recess at the junction of the nerve with the sclera was also a part poorly supported by the lobulated encephalocele; hence the giving way of the tunics in this situation. The macular coloboma can scarcely have been due to direct pressure of the tumour, and its relation to the encephalocele must, we think, be left an open question.

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FIG. 1.

FIG. 2.

FIG. 3.

FIG. 4.

FIG. 5.

FIG. 6.

FIG. 7.

FIG. 8.

FIG. 13.

FIG. 9.

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DESCRIPTION OF FIGURES.

FIG. 1.

Appearance of patient at the age of seven months.

FIG. 2.

× 230. Section of the orbital encephalocele.

FIG. 3.

Brain viewed from the left side—normal.

FIG. 4.

Brain viewed from the right side, showing mass of brain substance pressing frontal lobe upwards and temporo-sphenoidal lobe backwards.

FIG. 5

Brain viewed from below with mass *in situ*.

FIG. 6.

Brain viewed from below with mass displaced forwards, showing that the orbital convolutions are intact.

FIG. 7.

× 230. Section of intracranial adventitious mass of brain substance.

FIG. 8.

General view of the globe to show the two cysts, one at the junction of the nerve with the globe, the other involving the nerve itself. The figure also shows a piece of the dura mater of the encephalocele inseparably adherent to the globe.

FIG. 9.

Section of the whole eye to show general relations. All anterior parts of the globe are normal. The shape of the lens is due to shrinkage in formalin. The detachment of the retina is also artificial.

A, Thin-walled ectasia on the nasal side of the nerve; B, Coloboma of the nerve entrance; C, Position of the macular coloboma.

FIG. 10.

Diagram of the posterior part of the globe as re-constructed from serial sections.

A, Ectasia on the nasal side of the nerve; *B*, Coloboma of the nerve entrance; *C*, Macular coloboma; *ar*, Arachnoid lining of the encephalocele; *SD*, Sclera and dura of the encephalocele inseparably united; *s*, Sclera; *Ch*, Choroid; *r*, Retina; *f*, Fovea; *n*, Nasal side of porus opticus; *t*, Temporal side of porus opticus; *o*, Optic nerve; *o'*, Tongue of optic nerve consisting of fibres mounting over the ectasia to pass through the lamina at a different level; *l*, Lamina cribrosa; *r'*, Remains of retina within the fibrous tissue lining the coloboma; *v*, Intervaginal space not displaced with the nerve, but retaining its normal relations to the ending of the sclera and choroid.

FIGS. 11 AND 12.

Diagram to show theoretical stages in the formation of the coloboma of the nerve. Lettering the same as in the last figure. Fig. 11 shows the junction of the sclera with the lamina cribrosa commencing to bulge backwards into the intervaginal space. Fig. 12 shows a further stage of the same process. The nerve is being carried over to the nasal side, and the lamina cribrosa and surface of the papilla are being slewed round so as to face temporally. The ending of the intervaginal space remains in its normal position. The connection between the temporal side of the papilla and the retina is becoming greatly thinned.

FIG. 13.

Nasal side of the coloboma of the nerve to show the manner of entrance of the nerve.

o, Nerve curving strongly round the nasal side of the coloboma; *n*, Ending of the sclera and choroid on the nasal side of the nerve; *l*, Lamina cribrosa; *s*, Junction of the sclera with the lamina cribrosa on the temporal side of the nerve; *r*, Retina.