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In the observations brought forward in this paper I propose to show how several forms of congenital cataract are the outcome of arrests of development in the lens at different stages in its development.

I am, naturally, aware that to attribute a condition to an arrest of development offers only a partial explanation of its etiology, leaving the cause of the arrest still to be accounted for. I think, however, I shall be able to demonstrate that a recognition of the stage at which the arrest of development has taken place, though we do know the actual cause of the arrest, helps to a clearer insight as to the meaning of the appearances presented by congenital cataracts, and also affords valuable hints as to the most suitable operative measures to employ in their treatment.

For descriptive purposes, I propose to summarize the development of the lens into the three following stages and then to point out how certain arrests of development occur during each stage :

(i) The down growth of a fold of cuticular epiblast, which is separated in the form of a hollow vesicle from the rest of the surface epiblast by intruding mesoblast, and surrounded by a hyaline capsule.

(ii) The lengthening out of the cells composing the posterior layer of the vesicle until they fill its entire cavity. These are the first formed lens fibres and exist as the most central part of the fully developed lens.

(iii) The proliferation of the cells lining the anterior capsule and their transformation at the sides of the lens into lens fibres. A transformation which is effected by their lengthening out anteriorly and posteriorly, so as to encircle the fibres developed from the posterior layer, lines of sutures resulting where their ends come in contact. This laying on of fresh fibres laterally goes on throughout life, but its rapidity is lessened as life advances by the increasing intra-capsular tension tending to check the proliferative activity of the cells.

*An address read before the Oxford Ophthalmological Congress, *July 17th*, 1908.

(1) The arrests of development which I have to describe during the first stages have mainly to do with the capsule of the lens.

The hyaline capsule of the lens was at one time regarded as a structure of mesoblastic origin, and the product of the fibro-vascular sheath, which encircles the lens in fœtal life. There is, however, considerable evidence to show that it is really epiblastic in origin and formed as a kind of secretion from the epithelial cells lining it.

This evidence may be considered as embryological, anatomical, chemical, experimental and pathological.

Kölliker and Kessler pointed out that the capsule is present before the fibro-vascular sheath makes its appearance. We find also that the anterior and lateral parts of the capsule continue to grow and thicken with age, long after the vascular sheath has disappeared.

The anterior and lateral parts, which are lined by epithelial cells throughout life, are markedly thicker than the posterior, which is only lined by cells for a short time during early fœtal life. The anterior capsule in the adult lens has been estimated as 0·016 m.m. in thickness and the posterior as 0·008 m.m.

Though classed as an elastic membrane, it is found not to resemble yellow elastic tissue of undoubted mesoblastic origin, either in its chemical composition or in its staining reactions. In these particulars it has close affinity to the basement membranes of glands, which it is very probable are also the product of epithelial cells.

*Schirmer has shown that after experimental wounds of the anterior capsule, the gap first becomes closed by some rows of cells resulting from proliferation of the capsular epithelium beneath a layer of fibrin. Later a new hyaline layer makes its appearance across the space left by the retraction of the two extremities of the wounded capsule. This new hyaline layer is evidently the product of the epithelial cells on its inner surface, and is formed quite independently of any cells of mesoblastic origin.

In connection with anterior polar cataracts, new formations of a layer of hyaline capsule are sometimes met with beneath the opacity. A study of sections from a series of lenses with this form of cataract, which vary in the length of time which has elapsed since its formation, clearly establishes the epithelial origin of these new hyaline layers.

Anterior polar cataract most frequently occurs as a sequelæ of ulceration of the cornea in infancy; it may, however, be congenital. I have examined anatomically a lens with this form of opacity from an eye with congenital aniridia, in which the cornea was perfectly clear and healthy and had never been inflamed or ulcerated. In this specimen the microsopical

* *Archiv f. Ophthal.*, Bd. XXXV, Ab. 1, S. 220.

appearances of the affected area were precisely similar to those seen in anterior polar cataracts, resulting from corneal ulceration.

*I have published a description of the histological appearances of anterior polar cataract in six eyes, in which the time which had elapsed from the formation of the opacity to that at which the eye was excised ranged from six weeks to 21 years.

In two, in which the opacity was of only six weeks' and eight weeks' formation respectively, the changes found consisted in some disintegration of the lens fibres at the anterior pole and proliferation of the cells lining the anterior capsule in that situation. As to which is the primary of these two processes there has been much discussion.

The one theory holds that the primary factor is the proliferation of capsule cells excited into an excess of activity by the neighbouring inflammatory condition of the cornea. It affords, however, no explanation of the occurrence of congenital anterior polar cataract, which, as I have already pointed out, may be present in an eye where there was no history of any inflammation having occurred, and where, after the most careful microscopical examination, not the least trace of any past inflammatory trouble can be detected.

The other theory, in support of which I have myself published some observations,† assumes the disintegration of the lens fibres to be the first change. This disintegration apparently results from a localized disturbance in nutrition due to contact of the lens at its most prominent part, the anterior pole, with the inflamed and swollen cornea. In an infant's eye, with its exceedingly shallow anterior chamber and very globular lens, such contact is easily affected. The disintegration thus brought about in the lens substance causes a lowering in the intra-capsular pressure in the affected area; so removing the restraining influence which the intra-capsular pressure exerts on the proliferating activity of the capsular epithelium, and allowing of its irregular multiplication.

The fact that similar changes in the lens are excited by contact with it of a sarcomatous growth of the ciliary body favours this theory. The congenital anterior polar cataracts can be accounted for in accordance with it by imagining the apposition of the lens and cornea which normally exists for a part of fœtal life to have been abnormally prolonged, after the disappearance of the fibrovascular sheath which supplies the lens with nutriment in its early stages.

The mass of proliferated capsular epithelium, which is at first met with in an anterior polar cataract, gradually becomes converted into a condensed mass of tissue showing only a slight lamination, and studded here and there with a few flattened elongated epithelial cells which have retained their nuclei. Beneath

*_Trans. Ophth. Soc. of U.K._, Vol. XII, p. 89.

†_Trans. Ophth. Soc. of U. K._, Vol. XVIII, p. 124.

such a mass, epithelial cells, continuous with those lining the capsule in the unaffected parts, gradually insinuate themselves, separating it off from the adjacent lens substance. In an anterior polar cataract of seven months' formation I found a continuous layer of such cells on the posterior surface of the laminated mass, at the margins of which the hyaline capsule and its lining epithelium appeared to part company, the former passing in front and the latter behind.

In the course of time, in front of the epithelial cells which pass behind the laminated mass, a hyaline layer, similar in all respects to the lens capsule, makes its appearance. In an anterior polar cataract of 11 years' formation I found the alminated mass continued between two unbroken layers of hyaline capsule, both of which were continuous with the capsule in the unaffected region at the sides, the posterior of the two being alone lined by epithelium. That this new hyaline layer of capsule is the product of the epithelial cells which have crept in beneath the laminated mass seems to me an irresistible conclusion.

Anterior polar cataract occurs most frequently in infancy long before the lens has attained its maximum dimensions and density. As age advances, new lens fibres are laid on laterally, which, extending backwards and forwards, separate what were for a time the most peripheral fibres away from the capsule. As already stated, in anterior polar cataract, besides the proliferation of the capsule cells, there is some degeneration of the lens fibres. These are at the extreme periphery of the lens at the time the degeneration takes place; as new lens fibres are developed, the disintegrated area and the laminated mass formed from the proliferated cells, gradually become separated by them. So that, in an anterior polar cataract of long duration, two opacities are often seen, the one at the surface of the lens, the outcome of the proliferated epithelium, and the other some little depth down, due to degenerated lens fibres. I have recorded the details of seven cases in which two such opacities were observed clinically, and also the description of one lens in which they were found pathologically. The latter was the case to which I have already referred, where the anterior polar cataract had been of 11 years' duration. At the anterior pole was a laminated mass of tissue enclosed between two hyaline layers of capsule, the posterior of which was lined by cells, then came some layers of healthy-looking lens fibres and beneath them, at some little depth in the lens, was an area where the fibres had broken up into irregular granules and detritus. This latter corresponded fairly accurately in size and extent to the anterior laminated mass.

In some of the cases seen clinically with two opacities the separation has not been complete, a thin band cementing them together, so that they presented much the appearance of a collar

stud. Presumably in such cases the apposition of the ends of the lens fibres which have grown forwards from the sides has not been completely effected, some granular material having remained interposed in the line of suture.

Taking it now that the evidence brought forward suffices to show that the lens capsule is the product of the epithelial cells lining it, it is obvious that the posterior part of the capsule has only a very short time in which it can be formed, for the cells which compose the posterior layer of the lens vesicle soon lengthen out into lens fibres, and then become separated from the lens capsule by the growth around them of the new lens fibres laid on laterally; the posterior capsule for the remainder of life being devoid of any cellular lining. Under these circumstances it is not surprising that defects in the development of the central part of the posterior capsule should occasionally be met with, causing it to be unduly thin or absent altogether.

Several cases have now been recorded in which a congenital gap in the posterior capsule of the lens has been found. The first of the sort, so far as I am aware, was in a specimen which I described and pictured in 1892.* It was the right eye of a child, 9 months old, in which a peculiar appearance had been first noticed when 3 weeks old. This was found on examination to be due to a grey reflex at the posterior part of the lens with a large hœmorrhage on its surface. It suggested the appearance of a gliomatous growth, and the eye was consequently excised. Pathological examination showed that there was no new growth, but an opacity at the posterior part of an otherwise clear lens with a reddish stain on its surface. No remains of any central artery of the vitreous was to be seen. In microscopical sections of the eye passing through the pupil, the anterior capsule of the lens appears normal, it is lined by a single layer of cells which terminate as usual in the nucleated zone. On tracing the capsule backwards from the nucleated zone it is seen as a normal capsule for some distance; then at the posterior pole it is for some distance entirely absent. In the gap where it is absent, and for a short distance in front and behind the capsule on each side of it, there is a mass of tissue composed of elongated nucleated cells and fibres. This tissue forms the anterior part of the vitreous with which it is continuous. It resembles what Hess has described as atypically developed vitreous, *i.e.*, mesoblast which should have developed into vitreous humour, but which has formed a fibro-cellular tissue in place of the usual vitreous structure. When this takes place some of the blood-vessels of the fœtal vitreous usually persist. In this specimen no blood vessels are seen in the mass of tissue at the back of the lens, but there are collections of red blood corpuscles in the posterior part of the lens immediately

* *Royal Lond. Ophthal. Hosp. Reports*, Vol. XIII. p. 363.

in front of it. This hæmorrhage must have come from a branch of the central hyaloid artery before it disappeared. An attempt seems to have been made in this case to compensate for the failure in development of the posterior part of the lens capsule by atypical development of the anterior part of the vitreous.

The eye which contained this lens immediately after its excision was placed in Müller's fluid and hardened in a way which usually allows of the fibres of the lens, even in the fœtal eye, being shown in sections in a good state of preservation. In the sections of this lens, however, the fibres appear to be everywhere much shrunken and to form a sort of network with a mesh between them. The nuclear fibres remain adherent to the posterior pole. The cortical fibres are seen at the sides and have extended forwards round the anterior surface, but have not extended backwards around the fibres developed from the cells of the posterior layer of the vesicle. In the centre of the lens in front of the nuclear fibres is a quantity of coagulated, hyaline, albuminous material. Two cases, in some respects very similar to this, were described by Parsons in 1902,* and he has kindly allowed me to have drawing made from his sections of them. The first specimen was a congenitally small eye, which showed clinically a yellow reflex behind the lens and some opacities in the cortex. It was removed when the child was 10 weeks old and hardened in Mann's solution.

The following is the description of the microscopical appearances of the lens given by Parsons :—

"Lens: anterior capsule normal; the cubicle cells are continued round the equator, and line the peripheral parts of the posterior capsule; the central parts of the posterior capsule are wavy and apparently broken up by the tissue which covers the posterior surface; the peripheral parts of the posterior capsule are thickened and very wavy on one side, as if retracted. The cortex of the lens is maldeveloped, showing swollen, nucleated cells and fibres arranged irregularly. The nucleus is homogeneous, containing some aggregations of small cells with deeply-stained nuclei. Behind the lens, and in apposition to it, is a lens-shaped mass of very dense and cellular connective tissue extremely richly supplied with blood vessels."

Parsons' second case also had atypical development of the anterior part of the vitreous, a congenital gap in the posterior capsule of the lens, and a persistent hyaloid artery. The nucleus of the lens in this case was well developed, but from the way it had stained was sharply differentiated off from the cortex. Parsons' description of the lens in this case is as follows: "Lens fibres and anterior capsule normal. The posterior capsule is replaced by a layer of fibro-cellular tissue, which is continuous with similar tissue forming the suspensory ligament, and with delicate fibres running into the vitreous. The capsule behind

* *Trans. Ophth. Soc. of the U.K.*, Vol. XXII, pp. 253 and 258.

the equator of the lens is much thickened, and is wavy posteriorly, where it dwindles away and disappears in the cellular tissue. In sections near the periphery of the lens it is seen that the lens fibres overlap the irregular margin of the gap in the capsule, and here the fibro-cellular tissue is much thicker."

Having, from these cases, become aware of the not uncommon association of these defects, I was led to examine sections which I had previously prepared with persistent hyaloid artery and atypical development of the vitreous. I found that in those in which I had sections passing across the centre of the posterior pole of the lens a gap in the posterior capsule was present. In those which I had only sections passing through the side of the lens there was some rucking of the posterior capsule, suggestive of the possible presence of a defect in the centre.

So frequent is a congenital formation of fibrous tissue met with in association with a defect in the posterior capsule of the lens that I am inclined to think that the latter is the primary condition, and the former a kind of compensatory change.

In the first case which I described of congenital gap in the posterior capsule, I mentioned that some of the atypically developed vitreous lay just inside the capsule at the margins of the gap. In the next two cases I am going to refer to there was a much more extensive protrusion forwards of this fibrous tissue, of mesoblastic origin, into the cavity of the lens capsule.

The affected eye in one of these two cases was removed from an infant, aged 3 months. The mother stated she had noticed in it two small white specks at birth, which had gradually become larger and covered the sight. There had been no discharge from the eye and no inflammation. On examination, the iris was found to be in contact with the cornea, there being no anterior chamber, and the pupil was blocked by a greyish opacity. The eyeball was removed under the supposition that it contained a new growth, but none was found on pathological examination. A persistent and patent hyaloid artery extended throughout the vitreous ending and breaking up in a mass of fibres and elongated cells at the back of the lens (atypically developed vitreous). In anterio-posterior sections through the centre of the lens there is a break in the continuity of the posterior capsule at the posterior pole. On each side of the break it is seen to taper off and become merged with the mass of fibro-cellular tissue. In sections passing a little to one side of the posterior pole no gap in the posterior capsule is seen, but it is lined like the anterior by a single row of cells on its inner surface.

The fibro-cellular tissue at the back of the lens passes through the gap in the capsule invading the lens substance, and extends almost to the centre of it. Small collections of lens substance are seen in the sections surrounded by fibro-cellular tissue. In part of the fibro-cellular tissue within the lens capsule are some collections of red blood corpuscles.

At the anterior pole of the lens is a mass of laminated tissue presenting the appearance of an anterior polar cataract in which the fibres has become less closely compressed than usual. The lens substance is composed of a network of hyaline material with empty spaces in it.

In this case there is met with within the lens capsule an admixture of lens substance of epiblastic origin and atypically developed vitreous of mesoblastic origin. In the next case I shall refer to, the invasion of the lens substance by the mesoblastic tissue has been even more extensive than in the last.

It was a congenitally malformed cystic eye in which there had apparently been a failure of involution of the primary into the secondary optic vesicle.* The lens showed extensive changes, its anterior capsule was thrown into numerous folds, and lining it were epithelial cells in some places many layers thick. No well developed lens fibres could be detected. There was a considerable gap in the posterior capsule through which atypically developed vitreous, containing blood vessels, extended forwards. The contents of the lens capsule had thus become a vascularized mass of cellular tissue, in which it was difficult to differentiate the mesoblastic and epiblastic elements; granules of calcareous material, staining deeply with logwood, were scattered about in it.

These cases with a congenital defect in the posterior capsule of the lens and with fibro-cellular atypical vitreous filling the gap or extending forwards into the lens substance, are not only of considerable pathological and embryological interest, but also of clinical importance

In the first case I have mentioned, although clinically a grey reflex was seen at the back of the lens, the lens itself appeared clear. Except for some cortical opacities, the same was true of the second case. Microscopical examination of the lenses in both these cases, however, showed extensive changes, which could not be entirely accounted for by the hardening reagents. Both these eyes were removed very early in life, at 9 months and 10 weeks respectively. I think there can be little doubt that if they had been left some months longer the whole lens in each case would have become opaque.

I have recorded a case† in which presumably there was a similar condition behind the lens where such a change was observed to take place.

The case was that of a child, four years old when first seen, in whose right eye the mother first noticed a peculiar appearance three weeks after birth. On examination a greyish-white opacity, with a somewhat metallic lustre, was seen behind the lens; in its centre was a small, dark, round, slightly depressed

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Trans. Ophth. Soc. of the U.K., Vol. XXVI, p. 177.

†*Royal London Ophthal. Hospital Reports*, Vol. XIII, p. 365.

spot with fine blood-vessels passing through it. The lens itself was quite clear, and a red reflex could be obtained round the extreme margin of the opacity in all directions. Four months later the whole lens had become of a milk-white colour, and the changes previously seen at the posterior pole were hidden from view. The eyeball was enucleated and opened in the fresh state. The lens substance, which was milky-white throughout, easily separated from the capsule, at the posterior part of which was a dense white membrane. Passing right through the centre of the vitreous from the optic disc to this white membrane was a thin grey band. Microsopical examination showed the membrane to be composed of a dense mass of fibres and elongated cells, with a few small blood-vessels in it containing red-blood corpuscles. The central parts of the eye with the central hyaloid artery were preserved as a macroscopical specimen, so it cannot be definitely said whether or not there was a central deficiency in the posterior capsule of the lens.

In these cases, if the lens substance in front of the atypically developed vitreous became opaque before birth, or if the patient was not presented for examination until it had become opaque, the condition would be diagnosed as one of complete congenital cataract.

If a discission operation was performed some lens matter might be liberated, exposed to the action of the aqueous, and become absorbed ; but the fibro-cellular tissue at the back would be unaffected by the operation, and would remain as a dense opaque membrane, which it would be found very difficult to tear or to cut with a needle. If its removal was attempted by traction with a pair of forceps, vitreous would be certainly drawn forwards with it, and some would probably escape, seeing that the membrane is attached to, and is really part of, the vitreous. If it were to be incised with a pair of iridotomy scissors the gap formed would remain as a simple linear incision, and show no tendency to gape, the membrane being such a tough and inelastic structure.

All those who have had experience of operative procedures on congenital cataracts will recognise in the above, happenings with which they are familiar.

In two cases, shown to me by my friend, Mr. G. W. Thompson, where a tough membrane which could not be torn with a needle was encountered in connection with congenital cataract, the opaque tissue had ultimately been displaced downwards out of the central part of the pupillary area. A good clear opening in the line of vision had been thus obtained, through which the patient could see, and through which a clear ophthalmoscopic view of the fundus could be obtained. In both these cases a slender fibrous cord was detected passing forwards through the vitreous, from the centre of the optic disc to the back of the displaced opaque membrane, evidently the remains of a central hyaloid artery.

These two cases afford confirmatory evidence that the tough, resistant membranes met with in congenital cataracts are frequently due to a malformation of the anterior part of the vitreous. They also, I think, serve to indicate the way in which such cases had best be dealt with. In them a very satisfactory operative result had been obtained by the somewhat accidental displacement downwards of the opaque tissue. I would advocate that such displacement or couching of the tough membrane should be deliberately carried out I think in most cases of congenital cataract it is first desirable to perform a needling in the usual way : it serves as an exploratory procedure enabling the operator to judge of the consistency of the tissue with which he has to deal ; it exposes to the solvent action of the aqueous humour whatever lens matter there may be present. If, as the result of the preliminary needling operation it is found that there is a tough resistant substance in the pupillary area, which is not an anterior polar cataract, then as a subsequent procedure I would suggest that the operation for its displacement should be performed.

In some of the cases I have mentioned with a congenital gap in the posterior capsule of the lens, there has been an extension forwards of mesoblastic fibro-cellular tissue into the lens.

I now come to a class of cases in which through a gap in the posterior capsule of the lens there was a protrusion backwards of the lens substance into the vitreous ; a class of cases which is known as posterior lenticonus.

For our knowledge of the anatomical changes in posterior lenticonus we are largely indebted to Carl Hess. An excellent little monograph, recording some fresh cases and summarizing the literature of the subject, was published by André Patry.*

There are, he says, 16 published cases of posterior lenticonus, in which an anatomical examination has been made, two in man, and the other 14 in rabbits and pigs. In five of the cases the condition was bilateral.

In 14 specimens a gap in the posterior capsule was met with, in some it was doubtful whether it was intact or not. In three the continuity of the posterior capsule was certain, but in these cases it was abnormally thin.

In those cases with a gap in the capsule, the lens substance was either in direct contact with the vitreous, or separated from it by a connective tissue membrane of variable thickness continuous with the canal of Cloquet.

In some of the cases the only changes in the lens itself were in the fibres projecting backwards into the vitreous, or in those in their immediate neighbourhood. In others the changes were more extensive ; the anterior cortical substance was involved in 10 specimens. In 8 specimens the nucleus was situated near

* *Sur l'Histologie et l'Etiologie du Lenticône Postérieur*, Genève, 1906.

the posterior pole, in some in contact with the posterior capsule.

The views put forward as to the cause of posterior lenticonus are classed by Patry under two headings :—

1. That the rupture of the posterior capsule (as he terms it) is due to an increased size of the lens.

2. That the rupture and deformity of the lens are both due to traction produced by the hyaloid artery.

With regard to the first theory, there is nothing to show why the lens should increase in size or why the posterior pole of the capsule should be the part to give. The increase in size of the lens cannot be attributed to the formation of a cataract, for, as already stated, in some cases the cataractous changes are entirely limited to the parts at the posterior pole protruding into the vitreous. It is difficult to accept Pergens' suggestion of primary new growth of lens substance, or "*phakome*," unaccompanied by any histological changes.

With regard to the second theory, there is the difficulty that in a large proportion of the cases no trace of a hyaloid artery was found. Indeed, in only 6 out of 21 specimens was it present. Moreover, the opacities of the lens met with in connection with lenticonus are some of them different from those met with in traumatic cataract, and are not always entirely localized at the posterior pole.

Through the kindness of my friend, Mr. S. Mayou, I have had the opportunity of examining the sections of two microphthalmic eyes showing posterior lenticonus. Both cases have been recorded by him in the *Transactions of the Ophthalmological Society*.*

In the first case, the eye was an exceedingly malformed one, with a cystic formation apparently due to a distension of the anterior part of the primary vesicle.

The anterior capsule of the lens was lined in the usual way by a single layer of epithelial cells. The posterior capsule was absent, and was replaced by connective tissue, at the junction of which, with the capsule, the latter was thrown into numerous folds. In sections passing through the centre of the lens, I found a knob-like projection of the lens substance backwards through the opening in the capsule. Where the lens substance lay outside the capsule it was in contact with the connective tissue in the anterior part of the vitreous.

This case presented at the back of the lens a gap in the capsule and fibro-cellular tissue, as in the one I have first described, but instead of the fibro-cellular tissue extending into the lens substance, the lens substance extended out into it.

Mayou's other case was a microphthalmic eye with a congenitally detached retina, apparently due to failure in development of the vitreous humour. The lens had a conical projection posteriorly ; its posterior capsule was intact, but

* Vol. XXIV, p. 340 and Vol. XXVIII.

appeared thinner than usual. There was some distortion of the posterior fibres of the lens, but no backward displacement of the nucleus and no posterior vascular capsule.

Neither of the two theories already referred to adequately explains the causation of posterior lenticonus when all the recorded cases are considered together, and I would suggest that a better explanation is to be found in regarding the gap in the posterior capsule as a defect in development rather than a rupture. The fact that Hess† has found and pictured the condition in a chicken embryo 150 hours old is, I think, strongly in support of this view. As I have pointed out, the development of the capsule at the posterior pole has to be completed exceedingly early, and should it fail to form there is no means in later life by which the defect can be made good.

The gradual growth of the lens would cause such a gap to expand, and if not too much blocked up by fibrous tissue formation in the anterior part of the vitreous, the lens fibres at the posterior pole would protrude through it, forming the conical projections which are met with.

In some cases the faulty development appears not to result in a complete absence of the capsule at the posterior pole, only producing an undue thinness and weakness, which on growth of the lens results in a bulging outwards in that region.

Of the backward displacement of the nucleus of the lens which is met with in these cases of posterior lenticonus I shall have more to say later on.

The points which I have endeavoured to bring out in the foregoing observations as to the development of the lens capsule may be summarized as follows:

(i) That the lens capsule is the product of the epithelial cells lining it.

(ii) That the time during which the posterior capsule of the lens has to complete its development is exceedingly short, as the cells forming the posterior layer of the lens vesicle soon lengthen out into lens fibres and become separated from the capsule.

(iii) That congenital gaps in the posterior capsule are met with which are bridged across by fibrous tissue formation in the anterior part of the vitreous.

(iv) That this fibrous tissue formation in the vitreous, which sometimes contains blood-vessels from the central hyaloid artery, may extend through the gap in the posterior capsule, an admixture of mesoblastic and epiblastic tissue within the lens capsule resulting.

(v) That this fibrous tissue formation at the back of, or within, the lens capsule, accounts for those cases of exceedingly tough, shrunken, congenital cataracts which show little tendency to absorb after discission.

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†Pathologie und Therapie des Lensensystems, *Handbuch der gesamten Augenheilkunde*, Bd. VI, S. 201.

(vi) That the protrusion of lens substance through a congenital gap in the posterior capsule, or the bulging of a congenitally thin posterior capsule, offers the best explanation of cases of congenital posterior lenticonus.

2. Passing now to defects occurring during the second of the three stages into which I split up the development of the lens, that in which the nuclear fibres are formed from the cells lining the posterior part of the lens vescicle.

Before the lens vesicle has become completely closed anteriorly the cells which come to form its posterior layer show signs of elongation. After the vesicle is complete this elongation progresses until its whole cavity is filled by these cells transformed into lens fibres. These lens fibres later become surrounded by others laid on at the sides and form the nucleus of the lens.

If the cells forming the posterior layer of the lens vesicle for some reason failed to lengthen out into lens fibres, then no nucleus of the lens would be formed, and the laterally developed lens fibres would have nothing to encircle.

I wish to point out that there is a form of congenital cataract in which the pathological appearances are best explained by considering such an arrest of development in the nucleus to have taken place.

The clinical appearances of these cataracts are, I think, such as allow of them being differentiated from other forms of congenital cataract, and the recognition of their nature is of some practical utility in their treatment by operative measures.

The condition is one which has been spoken of as disc-shaped cataract, and four cases have been published in which anatomical examinations have been made. The first by Becker* in 1882, one by Vossius† in 1893, one by myself‡ in 1898, and the other by Hess§ in 1905.

The appearances of the lenses in anterio-posterior section have been figured in the three last. A comparison of the three figures and the descriptions given show that they presented the same chief characteristics. These are as follows :—flattening of the lens anterio-posteriorly, most in the centre ; so that in a section two rounded masses are seen connected by a band, an appearance comparable to that of a dumb-bell. In the central flattest part of the lens a laminated mass of tissue is situated, similar to that met with in anterior polar cataracts. It extends backwards from the anterior to the posterior capsule, no lens substance intervening. In the rounded parts, at either side of the central laminated mass, there is lens substance,

Zur Anat. d. Gesunden u. Kranken Linse., Wiesbaden, 1883.

†*Beit. z. Augenheil.*, Bd. IX.

‡*Trans. Ophth. Soc. of the U.K.*, XVIII, p. 124.

§*Hand. d. gesamten Augenheilk.*, Bd. VI, p. 145.

showing a variable amount of disturbance in the vicinity of the central part, and becoming more normal and regular in its arrangement towards the periphery.

Evidently, in these lenses there is an entire absence of the nucleus. Its failure in development does not seem, however, to have checked the activity of the cells lining the anterior and lateral parts of the capsule. The laminated mass in the centre is, as I have said, precisely similar in appearance to the laminated masses met with at the anterior pole of the lens in anterior polar cataract. These, as I have shown in the early part of this paper, result from the proliferation of the cells lining the anterior capsule, as the outcome probably of a decrease of the intracapsular pressure in their vicinity. In these lenses, in which the nucleus fails to develop, there would likewise be an abnormally low intracapsular pressure in the neighbourhood of the anterior pole of the lens, and so the formation of a large anterior polar cataract is what might be expected.

The activity of the cells lining the sides of the capsule not being interfered with, a formation of the laterally developed lens fibres has taken place. These having no nucleus around which to group themselves, have formed accumulations at the sides of the anterior polar mass, with irregularities and vacuolations of varying distribution.

The specimen showing this condition, which I examined, was described by me in a paper dealing with the pathogenesis of anterior polar cataract, and quoted as an example of a congenital formation of that form of cataract. I had not at that time appreciated the significance of the flattened condition of the lens as evidence of the non-development of the nucleus.

The lens was met with in a congenitally buphthalmic eye which was removed from a girl aged seven years. The eye had been noted as larger than its fellow from birth, and there had never been any inflammatory symptoms noted in it. There was a congenital adhesion of a portion of the iris to the cornea, a little above the centre of the latter, which showed no scar formation or any sign of part inflammatory infiltration. The condition of the lens could not be made out clinically.

In Vossius' case there was a leucoma of the cornea which also prevented the clinical appearances of cataract being seen.

In Hess' case the patient was first seen when 10 months old and had an iridectomy performed. Both eyes were similarly affected. Clinically there was a dense white central opacity with radiatory streaks from it. Around this central opacity was a typical zonular opacity and outside that a zone of clear substance. The edge of the lens was seen in the coloboma.

I have seen several congenital cataracts clinically which I think there can be little doubt belonged to this class. In some the results of operation tended to confirm the diagnosis.

The appearances which seem to be characteristic of this condition are: the presence of a dense circular central white opacity which, while extending to the anterior capsule, is at a distinctly deeper level than that of the surface of the iris at the pupillary margin: some irregular greyer opacity in the immediate vicinity of the central white patch, in which the latter appears to be set. With an undilated pupil this central opacity may fill the whole of its area, and appear to be a much shrunken opaque lens. If, however, a sufficient dilatation of the pupil can be obtained, a peripheral clear area is discovered. If an iridectomy is performed the edge of the lens will become visible, but it will be seen not to present its normal rotundity, appearing too thin from before backwards.

When a discission operation is performed on these cataracts, with a little manœuvring the white central opaque part can be separated from the rest of the lens, and will then probably drop down in the lower part of the anterior chamber, where it may remain for an indefinite time, showing no tendency to become absorbed or to give rise to irritation. When the dense, white patch has been picked off in this way, a central black opening is left, as can be easily understood from a study of the anatomy of these cataracts. After discission a small amount of lens matter usually becomes liberated and absorbed, but comparatively only a very small amount to what is so affected in other cataracts.

In this class of congenital cataracts, most satisfactory results are to be produced by needling operations, especially if the nature of the condition which is being dealt with is recognised. One or two discissions usually suffice to allow of good vision.

In a child of 18 months, with this form of cataract, whom I operated on in January, 1906, I succeeded in each eye in tearing away the central dense white *plaque*, which appeared to have calcareous matter in it. In each eye it has fallen into the lower part of the anterior chamber and remained there since. After its separation a good central opening, through which a clear view of the fundus could be obtained, was left.

3. My third stage in the development of the lens is concerned with the formation of the laterally developed lens fibres which surround the nucleus.

In the last class of case dealt with, some dystrophic influence, acting on the lens vesicle exceedingly early in fœtal life, resulted in the failure in formation of the lens fibres which develop from the cells of the posterior layer of the vesicle, but had not prevented the next stage in the formation of the lens, *viz.*, the development of lens fibres laterally from the cells at the sides of the capsule.

It is easy to imagine the exact reverse of this taking place, *i.e.*, a normal, or comparatively normal, development of the lens fibres from cells forming the posterior layer of the vesicle, and

the occurrence of some dystrophic influence later, interfering with the development of the laterally-formed lens fibres from cells at the sides of the capsule.

Such an occurrence, I would suggest, offers an adequate explanation for another variety of congenital cataract which is met with, also presenting distinctive clinical characteristics. It is that form of congenital cataract in which a small nucleus, comparatively well-formed, is found in a capsule distended with opaque milky fluid, which latter has resulted from the proliferative activity of the cells at the sides of the capsule instead of lens fibres.

I have examined sections of one such cataract, which occurred in a congenitally microphthalmic eye removed from a child two months old. The lens appeared completely opaque, and showed some dense white calcareous-looking granules anteriorly. There were several other congenital malformations in the eye, a nearly complete absence of the iris, a persistent pupillary membrane, and a persistent hyaloid artery. In sections the lens was seen to be much smaller than normal, and to present a very irregular outline. Its nucleus was the only part in which lens fibres were well formed, they were there concentrically arranged and free from vacuoles; in the cortical portions there was much irregularly hyaline substance. Cells lined the whole of the posterior as well as the anterior capsule. At the anterior pole the capsule was much corrugated, and on its inner surface was some dense laminated tissue, with flattened cells and calcareous deposit between the laminæ, a condition like that of ordinary anterior polar cataract.

In this class also may be included the case with total congenital cataract, the microscopical appearances of which are recorded by Hess.* The condition was bilateral, and the child died when six months old of pneumonia, 28 days after the lens in one eye had been needled. Sections of the lens in the unoperated eye showed the whole of the cortex to be composed of a homogeneous material, except for a few well-formed fibres in contact with the capsule at its equator. The nucleus which was suspended in this homogeneous material was composed of concentrically arranged fibres presenting changes similar to those met with in zonular cataract. In its peripheral parts there were numerous, variously sized, vesicles, and in the centre small drops staining deeply with hæmatoxylin.

In this case, I would suggest that the fibres arising from the cells of the posterior layer of the lens vesicle and forming the nucleus developed in the normal way. Then came some dystrophic influence which, besides affecting the fibres already formed so as to bring about the vacuolation in them, checked the

* *Handbuch d. Gesamten Augenheilkunde.* Bd. VI, S. 151.

development of the fibres which should have been laid on laterally, causing instead the formation of the albuminous fluid.

Clinically this form of congenital cataract presents the appearances of a Morgagnian cataract of late life. It has a characteristic milky-white homogeneous colour, and sometimes shows chalk-white specks on the surface.

I have operated on several congenital cataracts of this description; when the capsule is pricked with a needle there is an escape of the milk-white fluid, often containing some flocculent particles, into the anterior chamber. In two cases in which I left the opaque fluid in the anterior chamber I found the needling operation followed in the course of 24 hours by increase of tension. I now always adopt the precaution of evacuating it after making an incision with a broad needle.

The lens fibres developed laterally are formed by the lengthening out, forwards and backwards, of cells derived from proliferation of those composing the anterior layer of the lens vesicle. They gradually come to encircle the nuclear fibres developed from the cells of the posterior layer of the lens vesicle, and so cut them off from contact with the lens capsule. The separation of the original nuclear lens fibres from the capsule gradually increases with the growth of the lens.

Some laterally formed lens fibres lengthen out most backwards and others most forwards, but in the normally developed lens the nucleus is situated equidistant from the anterior and posterior capsule. Displacement backwards of the nucleus of the lens is sometimes met with as a congenital malformation, there being a partial or complete failure in the backward growth of the laterally developed lens fibres. Complete failure is sometimes due to adherence of the nucleus to the posterior capsule.

Patry, in his monograph on lenticonus posterior, in which he analyses the histological appearances in 16 recorded cases, says:—

"The nucleus is often (8 cases) situated very near the posterior pole, sometimes in immediate contact with the posterior capsule. This position is most often in association with a rupture of the posterior capsule."

As I have already pointed out, where he speaks of rupture of the posterior capsule in connection with this condition, I should say failure of development.

I have had a case under my observation for some years of a congenital cataract in which the nucleus appears abnormally far back. When first seen, the child was four years old; her left eye had been noticed to turn in since the age of eight months. The right eye appeared in every way normal and the lens was clear. In the left eye there was a dense white opacity at the posterior pole of the lens, and in front of it, close to if not in contact with it, a less dense greyish opacity which apparently involved the whole nucleus. It had a well-defined margin, but

was not perfectly circular, being roughly triangular in shape. All the cortical lens substance at the sides and front of the opaque nucleus was clear; the latter was evidently situated much closer to the posterior than the anterior surface of the lens.

Hess* has pictured the microscopical appearances of a lens from a rabbit's eye presenting a condition very similar to what was apparently present in the above case. In it a much altered nucleus is seen situated too far back in the lens and imperfectly separated from the posterior capsule; the cortical lens fibres at the front and sides were healthy.

Knies† has suggested as an explanation of the changes in some cases of congenital axial cataract which he met with, that adhesion of the nucleus to the anterior and posterior capsule had taken place, and that consequently it assumed a spindle-shaped formation on the expansion of the lens by the development of cortical fibres laterally.

Vossius‡ adopts the same explanation for a case of his in which there was in one eye a typical anterio-posterior fusiform opacity, and in the other an anterior polar opacity and a lamellar cataract with a filamentary opacity passing between the two.

There is a variety of congenital axial cataract, termed by Gunn§ "coralliform," for which it seems to me difficult to accept the explanation suggested by Knies for his cases. In this coralliform cataract tube-like opacities are seen to radiate forwards and outwards from the centre of the lens towards the capsule and to end in an ampulliform manner. In the absence of any histological proof of the nature of such opacities, I would suggest that they are probably situated in the lines of suture between the fibres.

The lines of suture between the ends of the lens fibres vary in their arrangement in different animals, and in the human lens the arrangements which are met with in different animals are met with at different levels in its substance.

In the most primitive form of lens, such as the globular lenses of some fishes, all the lens fibres extend from a central point at the anterior pole, to a central point at the posterior pole, and the suture passing antero-posterior is a single straight line.

The simplest divergence from this arrangement is for the ends of the lens fibres, instead of meeting at a point, to meet along a line; the line at the anterior pole being at right angles to that at the posterior. This permits of some shortening of the lens fibres, those which have to extend furthest anteriorly not having to extend so far posteriorly, and *vice versâ*.

A further modification effecting the same purpose is for the

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* *Handbuch d. gesamten Augenheilk.*, Bd. VI, S. 158.

† *Archiv f. Ophth.*, Bd. XXIII.

‡ *Beit. z. Augenheilk*, Bd. IX.

§ *Trans. Ophth. Soc. of U.K.*, Vol. XV, p. 119.

line of junction of the fibres to have a triradiate arrangement, the three arms of the triradiate figure at the anterior pole being situated in the reverse direction to those at the posterior.

A more elaborate arrangement still is the formation of a star-shaped figure, with primary and secondary radiations, along which the ends of the fibres come into contact.

The human lens in fœtal life is at first globular ; the fibres which are developed from the posterior layer of the secondary vesicle, and which form the extreme nucleus in the adult lens, have their extremities all meeting anteriorly and posteriorly.

As new lens fibres are laid on at the sides, the lens alters in shape, the lateral diameter increases more than the antero-posterior. A different mode of meeting of the lens fibres at the surface is then met with. At birth the simple triradiate figure at the anterior and posterior pole is seen.

With the further growth of the lens and the formation of fresh cortical layers the simple triradiate suture becomes in them replaced by the more complicated star, which at the surface of the lens in the adult is seen to present primary and secondary radiations.

In sections of a human fœtal lens, or of the lens at birth, the lines of suture running antero-posteriorly between the laterally developed lens fibres can easily be seen. Where the hardening of the lens has been imperfectly carried out and the lens fibres have shrunk, some expansion of the lines of suture frequently takes place. In sections of the adult lens the lines of suture are not to be detected.

It is well known that both in congenital cataracts and in cataracts occurring in late life disturbance may take place at the surface of the lens in the lines of junction between the lens fibres, resulting in a superficial triradiate or star-shaped opacity. What I now suggest is, that disturbance in the lines of sutures, not only at the surface, but also in their course as they radiate forwards and outwards from the nucleus, accounts for lines of opacity in congenital cataracts which are seen to radiate in that direction.

The defects which I have described as arising during this third stage of development in the lens may be summarized as follows :—

(1) The formation of congenital Morgagnian cataract from failure in development of the lens fibres laid on laterally, and their replacement by opaque albuminous fluid.

(2) Congenital displacement backwards of the nucleus of the lens, associated with a defective growth backwards of laterally developed lens fibres.

(3) Congenital fusiform axial cataract (of Knies and Vossius), due to adhesion of the nucleus of the lens to the anterior and posterior capsule, and defective growth forwards and backwards of the laterally developed lens fibres.

(4) Axial opacities radiating forwards and outwards from the nucleus, and located in the lines of sutures between the lens fibres laid on laterally, which run in that direction.

Conclusion.

In conclusion, I would, as the outcome of my clinical and anatomical studies, make the following suggestions concerning the treatment of congenital cataract:

(1) To wait until a child is 10 months old before operating. At an earlier age the cornea is so small and the anterior chamber so shallow that the necessary instrumentation cannot be so satisfactorily carried out as in a more fully developed eye. Moreover, the amount of aqueous humour is so small that it does not suffice for solution of the liberated lens substance.

(2) In some cases in which the pupil is small and does not dilate well with atropine, it is best to commence with an iridectomy.

(3) In nearly all cases it is well to begin with a needling. For valuable information can be obtained by its means as to the thickness of the capsule and consistency of the lens, even should it fail to liberate much lens matter to the action of the aqueous.

(4) If the cataract is a dense, white, anterior polar one, set in a ring of clear, or partially clear, lens substance and apparently flattened from before backwards, the so-called disc-shaped cataract, then an attempt should be made to separate the central white opacity with a needle and let it fall into the anterior chamber. Two needles are sometimes required to effect this.

(5) If, on pricking the capsule, milky-white fluid escapes into the anterior chamber (congenital Morgagnian cataract), it is well at once to evacuate this fluid, for fear of increased tension ensuing.

(6) In some cases of congenital cataract, the whole lens and capsule can be removed in a most satisfactory way by forceps. Very often, however, such a procedure is followed by escape of vitreous. It is difficult to differentiate which cataracts can be safely dealt with in this manner. They generally seem to be complete cataracts with a tough capsule and lens matter of a gelatinous consistency.

(7) If after a needling and some absorption of liberated lens matter, a dense, tough, white, fibrous-looking membrane remains, there is probably some atypical development of the anterior part of the vitreous. Under such circumstances, an attempt had best be made forcibly to displace the membrane downwards and backwards out of the axis of vision.