COMMUNICATIONS

UNI-OCULAR ZONULAR CATARACT*

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In a paper entitled "The Aetiology of Lamellar Cataract," read before the North of England Ophthalmological Society at Liverpool in April, 1920, Treacher Collins makes the stimulating statement: "It is very desirable that a larger number of cases of Unilateral Lamellar Cataract should be collected and analysed..... it would seem that when only one eye is affected some local cause can be discovered to which the condition can be attributed, such as an injury or an attack of inflammation." It is in an attempt to prove my contention that the tentative suggestion made in the latter part of his statement is true that I have investigated the condition; it is because the term lamellar cataract has come to convey to the minds of most ophthalmologists a definite phenomenon with an accepted aetiology that I have adopted the alternative adjective zonular to express what I believe to be a separate clinical entity.

Literature

An important attempt to collect and review the existing knowledge on this subject was made by Walter in 1917; he described

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the condition as extremely rare, and the opportunity to follow the development of zonular cataract as even rarer. He refers to cases described by von Graefe, Leber, Vossius, Weiss, Stein, and others. I have consulted those original papers which are truly relevant to the subject, and will discuss them in detail later.

Walter objects to Ruhwandl's query as to the accuracy of the histories in such cases—the latter tends to disregard the importance of the factor of injury. While agreeing with Ruhwandl as to cases of bilateral cataract, he disagrees that in every case of uni-ocular zonular cataract there was present a congenital cataract before the injury, and suggests that a one-sided zonular cataract may follow trauma, and that new transparent lens fibres are laid down subsequently over the opacity.

This idea is supported by many described cases. von Graefe in 1857, in considering the formation of zonular cataract in traumatic dislocation of the lens, describes three cases. The first refers to a boy of 19 who, 8 years previously, had sustained an injury to his right eye from a blunt cross-bow. The lens was dislocated upwards and inwards, and in this region projected forwards so as to bulge the iris locally. On maximal dilatation of the pupil the lens was found to possess an opacity similar in appearance to a lamellar cataract; moreover, the condition remained stationary through the several months that the patient was under observation.

The second case provided the opportunity of following the development of the opacity, as the patient was seen a few days after a contusion to the eye, with traumatic iridodialysis and hyphaema. As soon as the lens was visible it was noticed to be dislocated and a cataract was in process of formation. In a few weeks this attained the appearance of a typical lamellar cataract, after which the condition remained quite stationary.

The third case was seen 10 years after the injury; again the lens was dislocated, and showed a peripheral, perfectly transparent zone, within which was an opaque region through which a clear central interval could be made out. von Graefe notes here that the equator of the lens was angular — "polygonal" — not showing the natural curve, and explains the phenomenon by the observation that the zonula was not torn in spite of the subluxation, and that it exerted an irregular traction upon the periphery of the lens.

A similar case was described by Merz-Weigandt in 1900; here a man, aged 31 years, had sustained a blow on the eye from the knot of a leather whip, and a hyphaema followed. Gradual failure of vision ensued, and 4 years later the appearance of a "typical lamellar cataract" was presented. The importance of the case from a medico-legal point of view is stressed, and the fact
is noted that before the accident the patient was a first-class marksman in the army.

Weiss in 1902, in a paper on the subject, referred to von Graefe's cases, and also to a case of Vossius which the latter had needled for myopia; there followed the formation of a nuclear or lamellar cataract. He remarks upon the similarity of a case of his own, with the difference that in his case there was a nuclear opacity, and two lamellae of opacity, formed in the lens of a boy, aged 18 years, who had sustained an injury to the eye which dislocated the lens. Weiss considered that the injury to the capsule from the dislocation caused the nuclear opacity, and that transparent lens fibres were then laid down over this; he is at a loss to explain the later formation of two concentric lamellae of opacity.

Five cases of unilateral perinuclear cataract were described by Stein in 1904. The first differs from cases already described in that the injury, in a boy, aged 11 years, was a perforating one, through the sclera. A copper foreign body was removed from the vitreous, and the lens was subsequently noted to be clear. Twelve days after the injury a diffuse central clouding was seen in the lens, bounded by a well-marked curved line laterally, while medially the lens was obscured by many small wart-like excrescences, giving it a toothed appearance, especially distinct in the region of the wound. These are of especial interest in that they resemble the appearance presented in my Case I, and may be susceptible of a similar explanation to that put forward in my description. Clear lens matter was present outside the zone of opacity; from the description it would appear that the central interval presented an appearance very similar to that described in my Case II.

Stein's second case presented a perinuclear cataract estimated at 8-9 mm. in diameter, which was revealed after optical iridectomy in a girl, aged 14 years; the operation was undertaken for a vascularised opacity of the lower half of the cornea, with anterior synechia, which followed an attack of inflammation at the age of 4 years. A case with a similar history, seen at the age of 16 years, showed a very imperfect lamellar opacity, which would appear to have some connection with an anterior polar cataract which was present in the eye; this condition was also present in the previous case, but no connection was noted between the two opaque zones. It is possible that in each case the anterior opacity represents the direct, and the lamellar opacity the indirect, result of an affliction in early life of the cornea which was perforated at one stage.

The fourth case described by this writer sustained a blow to the left eye at the age of 12 years, before which the eye was reputedly normal; the injury was followed by a white spot, the size of a pin's head, in the pupil, with no impairment of vision. After
the receipt of a second injury 10 months before examination at the age of 20 years, the white spot grew in size and vision deteriorated. Stein described what can only be assumed to be persistent pupillary membrane, between the insertion of two threads of which was a capsular opacity; this was connected to a disc-like perinuclear cataract, surrounded by an irregular annular opacity. Peripherally, the lens was clear. The rather obscure history complicates the elucidation of the findings; it may be that the capsular opacity was an ordinary congenital polar cataract and the perinuclear opacity a traumatic zonular cataract, or alternatively that the anterior cataract was a capsular "imprint" of a cataract for which the trauma was entirely responsible. The existence of persistent pupillary membrane makes the first explanation more probable; it is quite possible that the first injury was trivial and actually caused no opacity of the subcapsular region of the lens, but that the second was entirely responsible for this.

His fifth case is unimportant in that, although an interesting condition of perinuclear cataract surrounded by a second layer of clouding in one eye only is described, there is no previous history given, and no explanation of the condition is volunteered.

Three other cases which appear to be similar have come to light. One, described by Marcus Gunn in 1895, presented himself at the age of 44 years having sustained a blow to the eye 20 years or more before. There was a rupture of the iris, and the lens was dislocated up and in; in the lens was to be seen a zonular opacity. Gunn states that the opaque zone when he examined the case was smaller than it was soon after its causation on account of compression by the lens matter laid on externally. This may be a statement made from previous experience, or merely a surmise, since there was obviously no chance of observing it as a fact in this particular case.

Mayou gives a very short description of the case of a man, aged 32 years, who sustained a "black eye" at the age of 9 years, there being no other relevant history. As far as one can gather from the description, the opacity found was zonular, though it would appear to have been more marked in, if not actually limited to, the inner quadrant. The margin was well defined and the opacity was covered by clear lens matter.

Finally, Collins in 1916 described the case of a girl, aged 9 years, whose right eye had been injured at birth; the injury had also caused some cervical deformity which had demanded operative remedy. There was, again, no history of fits or rickets, and the teeth were sound. The lens showed a grey semi-opaque zone, well defined laterally, though the vision was not much affected. Collins notes eccentric dilatation of the pupil, and also that the zone appeared to be eccentric in the lens; both of these observations suggest very strongly that it was rather the lens itself which was
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the eccentric factor, which would bring the case into line with many others here described. On account, amongst other things, of the persistence of pupillary membrane in the eye, Collins considers the possibility of the opacity being due to a developmental defect; he finally, however, rejects that idea and offers an explanation compatible with that which I shall bring forward to cover all these cases.

Clinical

My personal observations of this extremely rare condition are limited to six cases. I have had access to notes of two further cases, patients at the Royal London Ophthalmic Hospital, which describe a condition so apparently similar that the inclusion of these cases is justified.

Case I.—M. W., a woman, aged 33 years, gave a history of having fallen heavily at the age of 5 years, striking the left side of her forehead; the superficial injury necessitated skin suture. There was no history of convulsions, fits, or rickets. Not until the age of 10 years was any visual affection noticed by the patient, when it was remarked that the left vision was worse than the right. She attended a General Hospital where glasses were prescribed; feeling that these were of no benefit to her, she came to the Royal London Ophthalmic Hospital in 1928, complaining of defective vision in the left eye. Vision was found to be R. 6/6 partly; L. 6/24. In spite of her statement to the contrary, the use of her glasses (R. + 1'0 D.S. c + 0'25 D.C. → 180°. L. + 4'5 D.S. c + 0'5 D.C. → 150°) enabled her to obtain 6/5 with the right eye and 6/12 partly with the left.

The pupils were active and tension normal. Homatropine and cocaine produced normal dilatation of the right pupil, and full examination showed this eye to be normal; poor dilatation of the left pupil resulted. There was no sign of a perforating injury; some congenital pigment was found on the anterior lens capsule, and a slight degree of atrophy of the iris margin was noted at “2 o’clock”; it is possible that this represented the site of the main point of injury (cf. Case II).

The lens was subluxated to a slight degree, upwards and inwards. A zone of opacity occupied the lens, placed concentrically in it and covered by clear cortex except at an area corresponding to the site of iris atrophy, where for a small part of the circumference the cortex was stippled with opaque dots. The opacity was seen only in the lower, outer, and part of the upper quadrants; it was difficult to decide whether or not the circle was completed in the rest of the circumference by an extremely faint line of
opacity. Again corresponding to the point of iris atrophy was a large dense opacity in the periphery of the affected zone; elsewhere it was composed of striae and stippled areas, while peripherally below was a very regular line of cuneiform opacities, between which opaque lens fibres were very plainly seen. It is suggested, by the regularity of these peripheral opacities, that they represent the result of the tearing off of bundles of fibres of the suspensory ligament. The zone of opacity was occupied by a central clear interval, in which the anterior and posterior sutures were clearly seen.

Case II.—I. G.—A very instructive case, which I showed before the Ophthalmological Section of the Royal Society of Medicine in October, 1930, having first seen her in March of that year, and again at intervals until May, 1931, during which time I have observed no change in the lens. There was no history of rickets, nor of the occurrence of any fits or convulsions in infancy; the patient, a girl, aged 14 years, is one of a family of 9, all healthy, and is described as a very healthy child; she sustained a non-perforating injury of the left eye 5 years ago from a twig. The vision of the left eye was noted to be somewhat defective 3 years ago; as far as the patient can judge, there has been no change in either direction since then.

The right eye had 6/5; vision, was emmetropic, and appeared normal in all respects; there were many strands and fibrillae visible in the vitreous on microscopical examination, which were almost certainly normal. The left vision was less than 6/60, the pupil circular and active, and the tension normal. The pupil dilated well with a mydriatic, and revealed a shell of opacity in the lens; at about "6.30 o'clock," on the peripheral part of the anterior capsule, was some iris pigment which at one point was continuous with the iris, forming a fine pigment synechia.

Clear peripheral cortex was seen in every position except down and out, where the shell of opacity extended past the fully dilated pupillary border; the impression given was that the opacity was situated concentrically in the lens, and that the whole lens was dislocated, to a very slight degree downwards and outwards. The opacity consisted of anterior and posterior lamellae; the anterior lamella was densest down and in, whereas the posterior varied in density irregularly over its area. The edge where visible, was quite clean-cut, and no riders were present; the cortex of the lens outside this edge was everywhere clear except in a position corresponding to the point where iris pigment was found on the lens capsule. It is suggested that the pigment disturbance was due to the original injury and represented the site of reception of greatest injury; at this point, and at this point only, was the
cortex of the lens outside the main opacity affected—a precisely
similar state of affairs to that described in Case I.

The cataract, in its anterior lamella, consisted of dense cumuli
of opacity with striae below, with outlying opaque areas, less
dense, extending round the periphery, and centrally, from this
area. Upwards and outwards there was practically no opacity to
be seen, except where the two lamellae joined to form the well-
defined boundary; in the dense area opposed to this was an
optical appearance practically identical with lamellar separation,
which Koby describes as indicating hydration of the lens, but is
usually regarded as a senile phenomenon. The posterior lamella
consisted again of cumuli, here most marked up and out, and
tending to fade away down and in. The central interval was not
optically empty but was occupied by fine short spicules of opacity
set irregularly throughout the space; the anterior Y suture was
identified with ease, but the posterior could not be traced.

Examination of the parts of the eye posterior to the lens, so far
as this was possible, revealed no abnormality.

*Case III.*—J. L. a child, aged 4 years, with no history of birth
injury or other trauma, nor of any relevant disease, was brought
to the Royal London Ophthalmic Hospital because her parents
noticed that the left eye tended to turn up and out. The child was
one of twins, the brother being apparently quite normal.

The media of the right eye were perfectly clear and the eye
appeared normal in all respects. The left lens showed a centrally
placed opacity covered by clear cortex; this opacity, as far as
could be ascertained, certainly gave the impression of being
composed of a "shell" like a lamellar cataract; but owing to the
age and disposition of the patient, detailed examination was
difficult, and microscopy impossible. The outline of the opacity,
while clear cut, was irregular; the surface was also irregular, and
the opacity had a "cracked ice" appearance, of varying density
in different areas. There was also a posterior cortical opacity of
a vague stellate shape, which appeared to be not entirely separated
from the posterior lamella of opacity. The lamellar opacity (if
such it was) in this case was of a much smaller size than in the two
preceding cases; this is consistent with the disparity in the ages of
the patients.

*Case IV.*—L. S. was first brought to the Royal London Ophthal-
mic Hospital at the age of 6 years, in November, 1929. The boy
is in the charge of an Orphans' Institution and it is impossible to
obtain a detailed history; nevertheless, it is clear that "something
happened" to the child at the age of 2 years, and that at some
subsequent stage an abnormality of the right eye was noticed, for
which he was brought to hospital. At that time the right eye was noted to be convergent and cataractous.

I saw the patient myself for the first time in April, 1931. The left eye, apart from a low degree of hypermetropia, was normal. The right eye was externally normal, no scar or other evidence of injury being visible, the pupil active, and the tension normal. The pupil dilated circularly and well with homatropine and cocaine, and revealed a zonular opacity of the lens. The size of the opacity was such that there was a narrow ring of clear red reflex between it and the fully dilated pupillary margin. There was no sign of dislocation of the lens.

The edge of the opacity was very irregular, and not so sharply defined as in some of the other cases; at irregular intervals round it were projections into the overlying lens substance, which projections bore a vague resemblance to the riders of a lamellar cataract. The opacity itself was mainly striate, with woolly patches interspersed between the radiate striae; apart from a few very fine spicules, the thin layer of cortex overlying the cataract was clear. The opacity was dense enough to make it impossible to see the anterior Y or to determine whether there was a central clear interval; and to this extent it may be objected that the term zonular is probably a misnomer; be that as it may, the important fact of a superjacent clear cortex is established. No further abnormality of this eye was noted.

In spite of repeated enquiries, no relevant medical history was forthcoming; the boy's teeth were normal in appearance. The first impression gained from a macroscopic view of this opacity was that it belonged to that class associated with hyaloid remains, and this must be borne in mind as a possible alternative diagnosis; nevertheless, after microscopy and full consideration, I am fairly confident that the case resembles in its mode of origin the others in this series.

Case V.—C. D., was brought to hospital in June, 1930, being then a boy aged 1½ years. The history was that the parents had noticed a mist over the left eye for eight weeks; "the pupil had begun to go white," and the first sign of this had occurred six weeks after a serious fall which, according to the story, had severely damaged the patient's nose. The mother—an intelligent person—was quite sure that no sign of this whiteness was present before the accident, and was equally sure that both eyes had good vision; since the fall, she had on occasion tied up the right eye and satisfied herself that the vision of the left was defective.

On examination after mydriasis, which was full and circular in each eye, the right eye was found to be normal. The left showed an opacity of the lens, situated in the central part, concentrically;
there was apparently no dislocation of the lens. Slit-lamp microscopy was impossible. Examination with the loupe showed the opacity to be zonular in type. The posterior lamella was by far the denser of the two; nasally it was composed of a layer of cloudy opacity of regular density, while temporally it was more irregular, resembling rather, secondary cataract in appearance. The anterior lamella was very thin, and, indeed, appeared to be incomplete temporally; I have no doubt, however, that the microscope would have revealed a very faint layer of opacity, completing the zone. At three points round the periphery there were striate opacities resembling the riders of a lamellar cataract, though much more irregular in disposition and arrangement; at one point there was one rider, at the others two. They were of particular interest because of the ease with which two of them could be seen actually "riding" the lamellae, being staple-shaped opacities, lying in clear cortex in the antero-posterior plane; it is rarely one has the opportunity of so readily verifying the aptness of the term applied to these opacities. A similar appearance was noted by Stein in 1904, in examining a case of complicated cataract in a girl, aged 16 years.

The central interval of this cataract appeared clear, and there was a layer of clear cortex covering the whole; a good peripheral reflex was obtained in which the riders showed clearly. The fundus was apparently normal, though minute examination was difficult. I have examined this boy a year after his first visit and can see no change in the appearance of the opacity; it is of course impossible to measure the depth of clear cortex covering the cataract, though I have no doubt but that it is greater than it was a year ago.

Case VI.—F. B., a male nurse, aged 35 years, attended hospital on June 10, 1931, bearing a letter from his temporary employers requesting an opinion as to whether the cataract in his right eye was likely to be followed by a similar occurrence on the left, in view of his impending appointment to a permanent post. The fact that he was a regular soldier until 12 years ago proves that the cataract had formed since that period; he had passed all his medical examinations, and became a first-class marksman, shooting from his right shoulder. Eight years ago he was thrown on to the point of his chin in a road accident and was unconscious for a short time. Three or four years ago a foreign body in his left eye drew his attention to the fact, for the first time, that his right vision was defective; he has carefully noted that since that time there has been no increase in the density of the "film" over his right eye.

The left eye showed, on examination, no abnormality of any kind, and vision was 6/5. Right vision was down to counting
fingers at half a metre; the pupil was active, and tension normal. Full circular dilatation of the pupil occurred under the influence of a mydriatic, and revealed a diffuse clouding of the central part of the lens; this was poorly demarcated from the superficial lens matter, and, although not so dense in its centre as more externally, could not in any sense be described as possessing a clear central interval. In this respect the opacity was certainly not truly zonular in type, there being but a faint indication of this kind of structure by the slightly less dense central interval; it is highly probable that the comparatively advanced age of the patient at the time of the receipt of injury was responsible for this. The opacity was covered by a much more nearly clear cortex, consisting only of a very thin shell; again his age may be invoked to explain this. There were small radiate peripheral opacities in this cortex, somewhat suggestive of riders. The anterior Y was well seen, enabling measurements to be made; a good annular reflex was obtained, and a very limited view of an apparently normal fundus.

I have had access to the notes of two other cases of uni-ocular zonular cataract. The first describe the case of a male infant, aged 7 months, in August, 1925, whose right eye had been noticed to be cataractous for two weeks. There was no history of birth trauma or other injury, but the child had had a very bad attack of whooping cough 3 months previously. The right lens showed an irregular opacity, dense in parts, less dense in others; below these were opacities in the “cortex” suggestive of atypical riders. The appearance was then described as that of an “Atypical unilateral lamellar cataract”; the left eye was quite clear. On examination in February, 1926, the opacity was noted to be denser in the centre, the “cortex” being still clear; the left eye was normal.

The other notes referred to a boy who was aged 16 years when first examined in June, 1928. Again the patient was one of twins; no forceps had been used at birth, but there was a vague history of convulsions at the age of 1 day; the baby had been very small and backward and had been thought to be moribund on occasions. When examined in hospital, the left media were clear, and vision was 6/6; the right eye was described as possessing a “lamellar cataract denser on the temporal side;” vision in the eye, with a – 9.0 D. sphere, was 3/60.

As indicated in the opening paragraph, it is my endeavour in this paper to establish the fact that a uni-ocular zonular cataract, has an origin entirely distinct from that of the common lamellar cataract, and that the factor most probably responsible for it is injury. In this respect it will be apposite to consider a few details of the growth and nutrition of the lens and also, so far as they are known, the aetiology of lamellar cataract and the mode of formation of traumatic cataract.
The Growth and Nutrition of the Lens

Up to the ninth week of intra-uterine life, all the substance of the lens is formed from the cells of the posterior wall of the lens vesicle. These primary fibres, as they are termed, constitute what can be still recognised in the adult lens as the central dark interval. The anterior wall of the lens vesicle persists as the lenticular epithelium; over this, by the 13 mm. stage, is laid down the hyaline capsule, the origin of which is at present not definitely known. By an elongation of the bodies of the cells of the definitive postero-lateral wall, the nuclei of these cells come to occupy an equatorial position; whence are laid down the secondary fibres, overlapping the primary and forming, from the third month until birth, the foetal nucleus. Sutures are found in this and in all subsequent zones of the lens; in their simplest form, that of the anterior and posterior Y, they mark the limit of the foetal nucleus, and at birth are situated immediately subjacent to the capsule. The Y sutures of an adult lens thus indicate the extent of the lens at birth, though probably not the size, for it appears likely that compression of the older parts of the lens occurs as the newer fibres are laid down over them, causing an apparent shrinkage of the foetal nucleus. The fibres laid down from birth to puberty form another zone, referred to as the infantile nucleus; after this time, the adult nucleus is laid down over the preformed layers, and later the superjacent cortex; these are to be considered respectively adult and senile in time of formation.

Growth of the lens, therefore, continues throughout life; knowledge of its pre-natal rate of growth is fairly complete, whereas we are practically uninformed as to the post-natal rate. An important point of difference in the development of the lens from that of other epiblastic structures should be noted. In a structure such as the skin, the oldest part is external, and continuous surface death and loss is replaced by growth of the deeper layers. Since the lens is formed, however, by an involution of surface epiblast—a characteristic which it shares with the dental enamel—its oldest part is internal, and growth occurs by the laying on of fresh lens fibres externally. In the consideration of the aetiology of lamellar cataract this is a point of moment, since it is reasonable to suppose that a morbid condition of the whole organism might be reflected in a similar manner in two separate parts of that organism of similar embryological origin.

The nutrition of the mammalian lens is carried out in its early stages through the medium of its vascular tunic; a rich network of tissues derived from the hyaloid and long ciliary arteries. This fibro-vascular sheath normally disappears before birth, after which disappearance the lens depends for its nutrition primarily upon
the uvea, through the medium of the aqueous and vitreous humours. The metabolism must be carried on by a process of diffusion through the lens capsule, since all investigations have failed to find any support for the theory of the existence of circulatory channels in the lens substance.

It is reasonable to assume that the oxygen content of the aqueous and vitreous is insufficient in itself to carry out the nutrition of the lens; and the lens has been proved to possess certain oxidative enzymes, together with an auto-oxidisable substance glutathione, which make up an internal oxidation system responsible for its nutrition. A derangement of this internal oxidation system is productive of opacities in the lens, the result of denaturation and coagulation of the lens proteins. In this manner the importance of radiant energy in the production of lens opacities can be appreciated, either direct or through an affection of the ciliary body; for in the latter case the aqueous, a product of the ciliary body, will in its turn be affected, causing an upset in the oxidation system—an occurrence seen in its crudest form in the production of complicated cataract. Concomitantly with this, an alteration of capsular permeability will increase the likelihood of opacity formation; and such alteration can be even more readily attributed to trauma than to radiant energy.

**Lamellar Cataract**

In considering the much discussed question of the aetiology of lamellar cataract—using the term in its conventional sense—one remembers that it was Davidson and Horner who in 1865 first propagated the idea that such a cataract was the result of rickets. Many and varied have been the theories since adduced to account for this comparatively common condition, and it is even now impossible to hold the belief that, except in most general terms, one single cause is responsible for all lamellar cataracts.

Lamellar cataract is a disease of early infancy, and by most writers described as "probably congenital." It consists in the presence in the lens of a layer of opacity situated around a central core, which core often shows fine punctate opacities. There may on rare occasions be two such layers arranged concentrically; the cortex outside is clear, with the exception of the usual presence of the radial "riders," and remains so except in a very small minority of cases in which the opacity has been seen to become complete. The opacity is found in both eyes, to quote the majority of writers, "almost always"; in view of my thesis I would delete the former adverb.

Histologically, the opacity is seen to be caused by interfibrillary globules; fissures are also seen, some of which are manifested
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clinically as riders; and irregular circular spaces are found in some cataracts which account for the occasional presence of white dots of greater density than the general opacity. As to size, the average of 10 measurements made by four different observers and quoted by Collins was 4.775 mm.; the average of 10 measurements made ophthalmoscopically by Dub was 4.92 mm. Now the diameter of the lens at birth is about 5-75 mm.—a figure arrived at by many observers and agreed to by most ophthalmologists. It is also agreed that the size of a lamellar cataract is never greater than that of the lens at birth—a point stressed by, amongst others, Parsons, Collins and Dub, and verified by the above figures. Hence it must of necessity follow either that the formation of the lamellar cataract is prenatal in time, or that the zone of opacity at the time of formation is not the most peripheral part of the lens.

In an important paper dealing with the incidence of lamellar cataract and rickets in many parts of the world, Collins in 1892 found good reasons for believing in the inter-relationship of these two diseases; while he disagreed on the whole with the conclusion of Davidson and Horner, he held the opinion that some predisposing condition was to be held responsible for both. The histology of the affected lens suggests that some nutritional or metabolic disturbance would appear to be a likely cause, since the globules found must represent either non-developed lens cells, or lens fibres which, having commenced to develop, have through defective nutrition degenerated; and there is not the slightest evidence of this partial failure of nutrition being due to a local cause. Arlt, on the other hand, noting the frequent occurrence of convulsions in infancy in the subjects of lamellar cataract, pronounced a local mechanical explanation of the disease, suggesting that the convulsions caused a loosening of the “nucleus” of the lens from the “cortex,” the layer of fibres affected becoming clouded as a result. The presence of such a local cause would seem to me to exclude the opacity from the category altogether, as in a case quoted by Collins of a child, aged 8 months, who had an opaque staphylomatous cornea, the result of ophthalmia neonatorum. He found here, in addition to the much more usual anterior polar cataract, a zonular opacity of the lens, evidently due to mechanical disturbance of nutrition at an early age. Similar cases have been recorded by Becker and Schirmer.

Horner objects to Arlt’s theory on the ground that it is proved that the cataract is originally an affection of the whole lens; he quotes an experiment of von Hippel, who subjected rabbits in utero to the influence of X-rays, and subsequently watched transparent lens fibres being laid down over a total cataract. Peters, on the other hand, claimed to have watched two lamellar cataracts in the process of formation, and hence believed that an affection of the
nucleus could take place in a sound cortex; he postulated a transfer of fluid from nucleus to cortex due to shrinkage of the former. Hess stated, in opposition to this, that the fluid content of nucleus and cortex was the same, and gave, as a possible explanation of the occurrence of lamellar cataract, late closure of the lens vesicle; this is at best, however, a surmise and inapplicable to all cases of cataract formed post-natally. Peters accepts Horner's theory of the one-time totality of a zonular cataract in the case of bilateral congenital cataract, but inclines to the opinion that other types are caused by an alteration of the composition of the aqueous due to an affection of the ciliary body, which prejudices the nourishment of the lens; presumably he infers that the part of the lens farthest from the nutrient medium is affected to a greater extent than, or in contradistinction to, that situated nearer, an idea also subscribed to by Collins. In considering the possibility of this influence, as opposed to that of a general intra-uterine toxic nature, while one is assisted in the explanation of the cause of a traumatic zonular cataract, the question as to the nature and cause of the disturbance of the ciliary body remains unanswered; the fact that lamellar cataracts are bilateral increases the difficulty. On the other hand, the well-known occurrence of the clearing of a posterior cortical traumatic opacity lends support to the argument.

Parsons gives the cause as a period of malnutrition in late intra-uterine or early infantile life. He disagrees on the whole that rickets is the cause; in experiments on rats, nutritional deficiency caused lamellar cataracts but no bony changes, and in older animals rickets was often observed, whereas no lens opacity followed. The diet was deficient in vitamin A, fat and phosphorus, and the parts most obviously affected were those epithelial structures where desquamation does not occur, i.e., the lens and the teeth. Bennett calls lamellar cataract an early infantile disease, due to a derangement of general health; he makes a great point of the simultaneous affection of the lens and teeth, and again indicates their similar embryology. Nettleship, in his masterly study of heredity in connection with eye disease, differentiates between hereditary and non-hereditary cases, and states that in the former class of case, the cataract is just pre-natal in time of formation.

An interesting side light on our knowledge of the aetiology was thrown by Jeremy in 1919, in connection with parathyroid deficiency. He noted the development of a cataract of lamellar type in a patient who had undergone thyroidectomy 3 to 4 years before; cases were quoted in which cataract had followed 6 months (Schiller) and 5½ years (Westphal) after similar operations. Confirmation of the presence of this interconnection was provided by Edmunds (1916), who described the occurrence of cataract after experimental thyroidectomy in a 'dog.'
Uni-Ocular Zonular Cataract

It will thus be seen that many clinical phenomena—rickets, tetany, hypoplasia of teeth, parathyroid deficiency—have been proved to be associated with the occurrence of lamellar cataract, and that over all the cases hangs the vague shadow referred to as "disturbance of nutrition." What can provide a link to join up all these apparently isolated factors? I suggest that a consideration of calcium metabolism will do this. The influence of nutrition in general and vitamin supply in particular, upon the metabolism of calcium in the young growing organism is accepted; calcium plays an obvious part in the formation of teeth; rickets and tetany are known to be diseases intimately connected with calcium metabolism; and the influence of the parathyroid glands in this direction is also proved. Lamellar cataract is therefore to be looked upon as a manifestation of a disturbance of the metabolism of calcium in the growing organism, be it before or after birth; absence of substances exerting an influence on this metabolism—vitamin A, parathyroid secretion—may cause its appearance; diseases known to be intimately associated with this metabolism—rickets, tetany—are frequently complicated by its appearance.

Traumatic Cataract

The exact mode of formation of opacities in the lens, whether due to traumatic or constitutional influence, is still the subject of research and discussion; we are here concerned only with that group of opacities caused by concussion of the globe. In 1887 Schlösser gave it as his opinion that the opacity caused by non-penetrating injury of the globe was due to a distension of pre-existing lymphatic spaces in the lens, a theory which he claimed to support by experiments upon rabbits. A year later Fuchs described eleven cases of posterior traumatic cataract, and agreed with Schlösser as to the mode of their production. zur Nedden in 1904 produced a contrary theory, to the effect that the lens suffered a movement within its capsule, their connection being thereby severed; by this means fluid gained access to the cement substance and separated the lens fibres. The idea behind this has appealed to many writers; for instance, Friedenwald has lately suggested that the cause is a subcapsular epithelial injury, or an alteration in capsular permeability. One cannot but feel that such a change plays a large part in the pathological process; but it must be added that the first goes no way towards explaining the posterior limited opacities, nor does the second do more than suggest at most the first stage in the process.

Hudson, in a study of posterior traumatic cataract, gave yet another explanation, suggesting that the opacity was due to a
swelling of the lens fibres themselves. He quotes Leber as saying that the latter's researches almost definitely disprove the existence of lymph channels in the lens—a proposition now generally accepted; and rightly adds that Schlösser's hypothesis explains neither the appearance nor the progress of the opacities. As an explanation of the fact that such opacities are usually post-equatorial, he mentions that the posterior capsule is thinner and less resistant than the anterior, providing greater facility for swelling of the lens in that region; moreover, the equatorial fibres are younger and contain more fluid protoplasm, hence their reaction would be expected to be more severe. Rather than blame access of aqueous for the formation of the opacity, Hudson considers that direct concussion of the lens itself is the more probable cause. In an analysis of his cases, I could find only six that suffered non-penetrating injuries, and of these but three were sustained in early life; none of the three cataracts was zonular in type, though it is possible that one, which was removed when the opacity was total, would have gone on to become a cataract of this kind. In 1916 Whiting, studying anterior and posterior cortical traumatic opacities, found nothing to contradict Hudson's hypothesis.

To refer again to Leber's work, an experiment of his may be aptly quoted, in which he removed a small piece of the lens of a rabbit, and examined the lens on the rabbit's death 321 days after. A somewhat shrunken lens showed a scar in the capsule at the site of injury, from which there extended an opaque track to join a central deep opacity; all the periphery of the lens was clear with the exception of the capsular scar and the opaque track. Leber's explanation was that the injury was followed by a total cataract; proliferation of the capsular epithelium made a scar which closed the opening, and clear lens fibres were subsequently laid down over the opaque lens. Another case of this sort was reported by Vossius in 1898, who noted the formation of a nuclear or lamellar cataract after discission for myopia. The opacity of the ring type associated with this writer's name should be mentioned in considering concussion opacities, though it is probable that this is due either to a deposit of iris pigment upon the lens capsule (and this is not a cataract at all) or to transient degenerative changes in the capsular epithelium caused by pressure of the iris; I have never seen a case of this type in which the opacity did not clear completely and permanently.

Bearing in mind the changes in the lens caused by a penetrating injury of the capsule, it does not seem possible to neglect the influence of free access of fluid to the lens in the formation of traumatic cataract in general. At the present stage of knowledge of concussion opacities and their pathology, the most probable
Uni-Ocular Zonular Cataract

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explanation is that excess of fluid gains access to the lens fibres by reason of an alteration in the capsular permeability, such alteration resulting either directly from the injury, or from the severance of the physiological connection between the capsule and the subjacent epithelium and lens fibres. The presence of this fluid, with its salt content, then causes either swelling of the lens fibres, or of the inter-fibrillary cement substance, or both; the spaces between the fibres become filled with coagulum, the fibres degenerate, and, according to the extent of these changes, a partial or total opacity results.

The Aetiology of Uni-ocular Zonular Cataract

In reviewing the foregoing description of some 22 cases of uni-ocular zonular cataract, together with that of the nutrition of the lens, and the suggestions made as to the causative factors in lamellar cataract and traumatic cataract, several points come to notice. The first is the very high percentage of cases in which there is a definite history of non-penetrating injury, sustained at an early age; only two of the injuries were sustained after the age of 20, and the great majority happened before 12 years of age. Only two reports describe the occurrence of a penetrating injury; the one accidental, the other operative. Two further cases were the subjects of severe inflammatory disorder at an early age; one was certainly a perforated corneal ulcer, the other probably.

Of the cases due to non-penetrating injury, as many as half of them are described as having a dislocated lens on the affected side. It is at least possible that some injury occurred to the two solitary cases of which the description is lacking in this respect; it is about equally possible that a minor degree of dislocation of the lens was present in some of those cases in which this phenomenon is not described. But even apart from these surmises, it is justifiable to draw some conclusions as to the aetiology of the condition under review, from a consideration of the reports of the cases as they stand.

In considering lamellar cataract, it is difficult to see why the constitutional derangement responsible for the condition should bring about a unilaterial opacity, unless one recalls Arlt's hypothesis; with this mechanical explanation in mind, two facts become clear: first, that a cataract so caused may be justifiably included in the category of traumatic cataracts, and secondly, that it would appear to be an even chance whether the mechanical disturbance to the body as a whole should cause a cataract in one eye or both. I have accepted as evidence of injury, in two quoted cases, the occurrence of very severe whooping cough in a 4 months' infant, and that of convulsions in an infant at the age of one day.
It has been proved by the examination of many cases of concussion cataract, that the injury was sufficient to rupture the lens capsule, enabling fluid to gain free access to the lens; any subsequently formed lens fibres were also subjected to the action of the fluid, and the ordinary type of complete concussion opacity resulted. Bearing in mind the pathology of traumatic cataract previously outlined, one can deduce what would occur if fluid gained access to the lens for a limited period only, subsequent new lens fibres being again laid down under approximately normal conditions. Such an event could result from a temporary alteration in capsular permeability caused by a severance of the physiological connection between the capsule and the lens, with a consequent alteration in its osmotic properties, or—alternatively and rarely—by a perforating capsular injury which subsequently healed. In each of these cases, degenerative changes would occur in the periphery of the lens as then constituted; if an approximation to natural conditions was restored without delay, the nutrition of the central lens fibres would not be disturbed and the fibres laid down subsequently would again be transparent. According to the severity or duration of the alteration in physical properties of the capsule, so would the transparency of the central lens or the younger peripheral fibres be affected to a greater or lesser degree. I have endeavoured in my case descriptions to show how this degree of transparency does vary.

It is in view of the reasonableness of this theory that I have emphasised the high percentage of cases in this series showing dislocation of the lens. Such an eventuality would certainly alter the relations between capsule and lens; a minor degree of dislocation, perhaps clinically undetectable, would also cause an alteration in their relationship; and, by deduction, a disturbance of the lens insufficient to dislocate it may cause, again, a disturbance of an exactly similar nature. It would also seem, from a consideration of my cases I and II, that in addition to a general disturbance of the capsule, there may be a more severe local injury affecting the properties of the capsule permanently, the affection being manifested by a local opacity in the cortex outside the affected zone.

It is necessary to remark briefly upon those cases in the series which do not quite fall into line. The two who suffered a penetrating injury could be included, in view of what has just been said, on general grounds had they both behaved typically; but it will be remembered that the case of intra-ocular foreign body is described as showing the central opacity in twelve days—that is, a zonular opacity appeared as such. Again, von Graefe’s second case is similarly atypical since he describes the opacity as assuming in a few weeks the appearance of a typical lamellar cataract. It is difficult at present to see why either a penetrating or a non-
penetrating injury should cause the appearance of an opacity within transparent lens fibres; it is on the other hand easy to see, in the other case, how the periphery of the lens should be rendered opaque, with the subsequent laying down over the opacity of transparent lens fibres. In the two inflammatory cases, it must be assumed that the nutrition of the lens was disturbed either by an alteration in the chemical properties of the aqueous, or that the capsular permeability was altered by a toxic mechanism, or both. It is not safe to draw any conclusions from either of these cases, reported as they are at second hand; after careful consideration of Stein's description I was inclined to doubt whether the use of the term zonular was really justified in either of the cases, though it was as such—"perinuclear"—that he described the opacities.

Conclusions

1. The cause of a uni-ocular zonular cataract is trauma, almost invariably of the non-penetrating variety.

2. The injury is of such kind as to cause a tendency towards dislocation of the lens; in quite half the cases, dislocation actually occurs.

3. The cause of the zonules or lamellae of opacity is a temporary disturbance of the nutrition of the lens, brought about most probably by a severance of physiological connection between lens and capsule, with a consequent alteration in the capsular permeability.

4. The recorded cases support the view that such a type of opacity is more likely to occur in a young, actively growing lens than in an adult lens.

5. While it might be suggested that inflammation is an alternative cause of a zonular opacity, the immediate cause presumably being a chemical or toxic action by the nutrient fluids, such a suggestion finds no support in this investigation.

Postscript.—In examining cases of traumatic zonular cataract one is provided with an important and hitherto unattainable piece of information. It is assumed from what has been said that the zone of opacity occupies, in its inception, the most superficial part of the lens, and that the subsequent laying down of clear lens fibres causes it to occupy an intermediate position; knowing, then, the age at which the injury was received, and measuring the relative position of the opacity, one should be able to deduce the relative
Epith. to Opac. = \(a\)

Opac. to Y. = \(c\)

Epith. to Y. = \(b\)

<table>
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<tr>
<th>Case</th>
<th>Age when measured</th>
<th>Age at injury</th>
<th>(a)</th>
<th>(b)</th>
<th>(c)</th>
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<th>Period (c)</th>
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<td>15</td>
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<td>5</td>
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<td>3. Case 6</td>
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**GRAPH I.**

- Growth of Lens mms./10
- \(b\)

**GRAPH II.**

- Periods, Birth to Injury and Injury to Examination, Years.

- Age in Years.
or actual post-natal rate of growth of the lens. It was my intention to carry out this investigation with the material at my disposal.

The measurements were made by means of the micrometer drum (Ulbrich) on the binocular microscope of the Gullstrand slit-lamp. The lamp was arranged so that the emerging beam was as nearly axial as possible; the microscope therefore had only to be racked straight forward in order to take the two readings. The beam was first focused (by racking away from the eye) on the epithelium of the lens, the drum thereby remaining at zero; the microscope was then racked forwards till the anterior lamella of the opacity was focused, and a reading was taken; further racking forward focused the anterior Y, and another reading was taken. Three readings were made at each position and the mean taken. These readings gave the actual depths of the zone of opacity, and of the anterior Y, in the lens, in tenths of a millimetre; the error introduced by the refractive media is so small as to fall well within the limits of experimental error and may be disregarded.

My attempt to produce some data in this investigation from which one could generalize as to the post-natal growth of the lens was frustrated. In the first place it was essential that the zonule of opacity should not be so dense as to render impossible the accurate focusing of the anterior Y; in only three of my cases was this condition fulfilled. Of the three remaining cases thus measured, two gave consistent figures, whereas the third was quite incompatible. The possible reasons for this are many; an inaccurate history, a mistake in technique, or an abnormal shape of the lens consequent upon dislocation (a very probable factor) would each render the figures valueless. It is unfortunate that this was the case; the figures obtained are, however, reproduced below for analysis, where the discrepancies shown by Case I will be noticed; these are even more strikingly seen in the graph (I). Owing to the extremely small number of lenses measured, it is not even justifiable to conclude here that the rate of growth of the lens is an individual factor, varying in different cases; a useful conclusion can only be arrived at by measuring many more cases of what is, unfortunately for this investigation, an extremely rare condition.

Graph II is appended to show the result of measuring the distance between the epithelium and the anterior Y in a number of normal lenses.

For access to cases, and to notes of cases, relevant to this investigation, I am indebted to Sir John Herbert Parsons, Mr. F. Juler, Miss Mann, and Mr. R. C. Davenport, of the Royal London Ophthalmic Hospital.
GLIOMA OF THE OPTIC NERVE

BY

BERNARD GLUCK

CARDIFF

GLIOMATA of the optic nerve are rare, but not excessively so. Surveying the literature up to that date, Hudson¹ in 1912 was able to collect 118 cases which, after reviewing the published reports and where necessary modifying the original diagnosis, he was able to classify as gliomata or probable gliomata. In 1930, Mathewson² was able to trace 52 additional cases, so that up to the present less than 200 cases have probably been reported. But in the lifetime of the individual surgeon it is unlikely that more than one or two cases will come under his care; thus, according to

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