A CLINICAL ACCOUNT OF A SERIES OF CASES OF CAPSULAR (? CAPSULO—SUBCAPSULAR) CATARACT ASSOCIATED WITH THE DEPOSIT OF PIGMENT AT OR AROUND THE CENTRE OF THE CAPSULE, WITH CONSIDERATIONS AS TO THE AETIOLOGY

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PART I.

The following series is brought forward with the object of calling to such cases the attention of clinicians, and possibly by some piece of good luck, of pathologists, to their characteristics, and in order that similar types may be more adequately described than is here possible.

There are seven cases in all. They occurred in the routine examination of school children who had been presented on account of defective vision, and who numbered approximately 7,500. The incidence is, therefore, something under 1 in 1,000. In spite of this considerable frequency a reference to a certain number of standard text-books, to the Ophthalmoscope and the British Journal of Ophthalmology, to the Transactions of the Ophthalmological Society, U.K., and to the American Encyclopedia of Ophthalmology, has produced but little to the point. At the same time some of my friends, including Mr. Sydney Stephenson, and
I myself, am under the distinct impression that we have from time to time observed similar cases in the course of clinical work. The probability is that the condition is not very rare but, that, being of academic rather than practical interest, it has not stimulated the clinician to the exercise of his descriptive powers, nor come under the, much less probable, observation of the pathologist.

Case I.

It has been hinted above that the cases here under notice have not been very perfectly recorded. This is due to the fact that very few of them have been seen more than once, so that it has been necessary to rely on the original hasty notes in the case book which were accompanied, fortunately, in most instances, by a rough sketch of the conditions present. The crude diagrams published herewith, and for which I offer my apologies, constitute merely an attempt to indicate the general arrangement, the similarities, and the differences of the abnormal appearances. Emphatically they must be regarded as mere diagrams in which the relative size of the interesting parts is perhaps exaggerated, and their positions not necessarily exact.

Case II.
The seven cases, which occurred in the County of Lanark (but no two of them in the same district of the county, to say nothing of the same family), all show a deposit of pigment at or around, or partially around, the centre of the anterior lens capsule in one or both eyes. In four, the fellow eye was normal, in three, the condition was similar in the two eyes. In addition to the pigment deposit, which varied considerably in amount, six out of the seven presented small opacities which were white or bluish-white like boiled white of duck’s egg. Of these six cases with white opacities, in four the latter were rounded, and in two they were radially placed. In two cases out of the seven there were adhesions of the iris to some part or parts of the central pigmented region of the capsule, while in one of these two there was a tag of persistent pupillary membrane as well as an iris adhesion at another portion of the iris border. In none of the cases was there any reason to think that there had been a post-natal iritis. The white, or bluish-white, spots occurred in every case outside the central pigmented area though apparently in relation
to it. In one, the white spots were so close outside the ring of pigment that, with transmitted light, in which they appeared black, the whole appearance on casual examination might have suggested a lamellar cataract with riders. It is not possible from the available notes to state with certainty the exact depth at which all of these rounded or radiate spots occurred, but in one case (No. 6),

which has been more fully examined than the others and was submitted for observation at a recent meeting of the Scottish Ophthalmological Club, the round white spots seemed definitely to be an integral part of the capsule. The radial white flecks in two of the cases perhaps suggest a sub-capsular position. The most that can be said from memory is that all the opacities seemed to be very superficial and were classed in my mind as capsular or sub-capsular opacities. It is unfortunate that of the seven cases five, including the two with radial opacities, have gone quite out of reach and cannot now be followed up. It was, indeed, only on the conclusion of my period of service with the Lanarkshire Education Authority that I had leisure to collate the cases, and by that
time only two remained at school. It so happens that these two are the two showing iris adhesions (Nos. 6 and 7).

Notes of the individual cases follow:


Case III. Francis B., 11 years. Small ring of pigment with spots around it, the colour of boiled white of duck’s egg. One of these spots is considerably larger than the others, and has been visible through the undilated pupil since birth (mother’s statement). Fellow eye normal. V.R. with correction 5/9 partly, V.L. 5/6.

Case IV. James M., 7 years. Both eyes affected. There is a somewhat irregularly shaped pigment area in the centre of each capsule. The original note says, “There is an irregular brown opacity which, on magnification, is seen to be made up of minute dots. In addition, there are in each eye one or two bluish-white opacities which lie in a radial position near the brown opacity.” The position of the radial opacities differed in the two eyes. (This case was seen six years ago. It is evident from the V.A. that the pigment shown in the diagram is much too dense, especially in the R.E.) V.R. 6/12, 6/9? V.L. 6/36, 6/24? Left convergent squint.

Case V. Esther McG., 13 years. Both eyes affected. The original note says, “In both eyes at the centre of the anterior part of the lens capsule there are opacities. Parts of this are pigmented in the form shown in the diagram. The other opacities
are the colour of boiled egg white, small, few in number, and arranged radially.” The radial opacities in the diagram are probably slightly too large relatively to the diagram, while the pigmented area is almost certainly exaggerated. V.R. 5/12 partly, with letters down to 5/6. V.L. 5/6.

Case VI. James D., 6½ years. This case has been more fully studied than any of the others, and was shown to the Scottish Ophthalmological Club at Glasgow on November 5, 1921. Both eyes affected. In the centre of each anterior capsule is a circular area of fine pigmentation. In the R.E. there are no white opacities. Upwards and inwards there is an adhesion of the iris which can be traced right into the central pigment. It has the appearance of an iritic adhesion. Extending from below upwards to the pigment area there is, as generally agreed by the members of the Club, a thread of persistent pupillary membrane. This, when the pupil is contracted doubles up into an S-shaped loop. In the L.E. there are three round white spots, very small, close to the edge of the pigment area. There is also a very small iris adhesion above, which had escaped notice and is not shown in the diagram.


Case VII. Catherine C., 9 years. The right eye only affected. This case is different from all the others in that instead of a ring, broken ring, or plaque of pigment, this is arranged in the form of five little masses which, if joined by a line would complete a somewhat irregular circle. At the outer margin of most, not all, of these pigment masses, and in the position which, in the other cases, would correspond to the outer border of the pigment ring, are small white dots of the colour of boiled egg white. Three of the pigmented masses have comparatively broad adhesions to the iris which are of the iritic type. V.R. 5/60 not improved. V.L. 5/60 corrected for mixed astigmatism 5/9.

From the clinical appearances alone one can merely speculate upon the aetiology. At first I was inclined to consider the condition as a purely developmental anomaly, but, when it became obvious in the two cases 6 and 7 which occurred in 1920 and 1921, that there were in them, though otherwise so similar to the previous cases, adhesions of the iris indistinguishable from those formed as the result of iritis, I became uncertain as to the parts played respectively by faulty development and ante-natal inflammation. Further, Dr. Leslie Buchanan, to whom I had talked about these capsular cataracts, happened just at this time to see an infant eight months old who was brought to him merely because the mother had noticed white spots in the child’s pupils. When atropin was used there were iritic adhesions and white spots in both eyes, which latter in appearance corresponded to the
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spots in my cases. Owing to extreme restlessness a complete examination of this child was practically impossible.

Unfortunately Case VII could not be shown to the Scottish Ophthalmological Club, owing to an attack of mumps.

Some may be inclined to suggest that these are really cases of post-natal iritis with pigment left on the lens capsule. There are various reasons for not accepting this view. In the first place there is, so far as I know, no history in any one of them pointing to inflammation of the eyes, although such inflammation cannot be excluded positively. These children were all brought before me as the result of school medical inspection, in the course of which defective vision had been recorded. Secondly, the relatively high frequency of incidence. Thirdly, it is not usual in post-natal iritis to have such a marked deposit of pigment followed by complete or nearly complete withdrawal of the pupil margin, even when the iritis has been recognized and treated with atropin, and one may suppose that, had these been untreated cases of post-natal iritis, in no single instance would the pupil have remained free. Fourthly, post-natal iritis does not result in the formation of egg-white spots on, in, or under the anterior lens capsule. Post-natal iritis may presumably be put out of the reckoning. But it seems to be a fact that in two of the cases there has been iritis which might be regarded as ante-natal. The remaining five cases are so similar in all respects, except the presence of posterior synechiae, that one feels bound to admit that there might have been iritis in them too. But if one base one’s ideas on what occurs in post-natal iritis it is somewhat difficult to understand why, in view of the marked pigment deposit, there were not decided posterior synechiae in all of them. In putting the cases, or rather one case and the accounts of the other six, before the members of the Scottish Ophthalmological Club, I asked the meeting to discuss what connection there might be between a developmental error as represented by the egg-white opacities and the persistent pupillary membrane on the one hand, and the evidence of iritis on the other. Did a developmental fault lead to the iritis? The only positive suggestion was that of Dr. A. J. Ballantyne, who thought it more likely that pre-natal iritis might be the cause of faulty development. I cannot do better than transcribe what he has written down for me as the substance of his remarks.

Dr. Thomson asks for opinions as to the nature of the changes seen in his cases. He has referred to the fact that two of them show, in addition to the lens opacities, true posterior synechiae, while one of these two shows remains of the pupillary membrane as well as synechiae; and he suggests that there may be some connection between the incomplete disappearance of foetal structures and the occurrence of inflammation of the iris. I was interested
in a somewhat similar problem in a case of anterior synechia of pupillary membrane remains which I reported in the Transactions of the Ophthal. Soc., Vol. XXV, and it has been discussed by Treacher Collins in these Transactions, Vol. XXVII and elsewhere. I would hazard the suggestion that in these cases of Dr. Thomson's the deposits, pigmented and unpigmented, on the lens are remains of the tunica vasculosa lentis, and that the occurrence of pre-natal inflammation of the uvea caused the persistence of fragments of this foetal structure."

I am not by any means sure that Dr. Ballantyne's ingenious explanation is the correct one. The most common cause of iritis is syphilis, yet Hutchinson says that iritis in infants due to hereditary syphilis is rare. He himself, with all his experience saw only twenty-three cases ('Syphilis,' 1899, p. 239). Therefore it does not seem probable that ante-natal syphilis will be any more frequent. My cases showed a frequency of about 1 in 1,000 children examined for defective vision, and, further, presented no obvious signs of this disease. Syphilis, therefore, seems an unlikely cause of the iritis. What other causes of ante-natal iritis are there? In a report on 500 cases of iritis, Jennings and Hill (Ophthalmology, April, 1909, quoted by American Encyc. of Ophthalm.) give intrauterine inflammation as the cause in two cases. No hint is here given as to the cause of the intrauterine inflammation, but if syphilis be excluded one would be inclined to think that some form of toxaemia is the next most probable explanation of it.'

PART II.

Since the foregoing was written, one of the Members of the Scottish Ophthalmological Club has made a suggestion as to aetiology. This suggestion demands a further argument.

(I.) It has been suggested that "most, if not all" of these seven cases may be cases of "quiet" iritis of infants. For confirmation of this I was referred to the work of the two Hutchinsons. Consequently I carefully re-read what J. Hutchinson, Senior, said in "Syphilis" (v. supra) and studied J. Hutchinson, Junior's article on "Quiet Iritis" (Trans. Ophthal. Soc., of U.K., Vol. VIII). To my mind neither of these articles lends any particular support to the view that such cases as these are post-natal and syphilitic. There are certain facts pro and others con. The principal fact pro is that quiet iritis in infants may be overlooked owing to absence of obvious symptoms, and that the results of such iritis are not discovered till later on, as in such cases as these. Con we have (a) the great rarity of infantile iritis. Hutchinson, Sen., while holding the opinion that the cases
of infantile iritis may be overlooked, also says, "Yet in proof
that, however carefully looked for, it is really very rare, I may
mention that during seven years' practice at the Metropolitan Free
Hospital I never treated a single case in connection with that
Institution, although numbers of congenito-syphilitic patients
presented themselves and I scrupulously looked at the eyes of
all" (loc. cit.); (b) the relative freedom of the pupil in my cases.
Compare what Hutchinson says: "Notwithstanding the ill-
characterised phenomena of acute inflammation, the effusion of
lymph is usually very free and the danger of occlusion of the pupil
great" (Ibidem); (c) neither of the Hutchinsons, with their oppor-
tunities of observation and skill in the doing of it, have described,
in the works referred to, the other characteristics of the type of case
at present in question, and it is fair to assume that such character-
istics were absent in their cases. In only one is there mention
made by Hutchinson, Jun. (loc. cit.) of "white and brown opacities
on the surface of the anterior capsule."

(2.) Cases to all intents and purposes identical with mine have
been reported by Jessop (Trans. Ophthal. Soc., U.K., Vol. VIII)
and by Stephenson (Ibidem, Vol. XXX). In both of these the
cause was stated as ante-natal. Jessop refers to "foetal iritis,"
and describes in the same case an iritic adhesion and a thread of
persistent pupillary membrane, with "milk white" spots in the
lens (cf. my case VI). Stephenson regards his case as "a rare
type of persistent capsulo-pupillary membrane," and the descrip-
tion is almost identically that of several of my cases.

(3.) Lastly, and most importantly, there is the article by
Cosmettatos, of Athens (Ann. d'oculistique, 1912, p. 241), entitled
"Des Restes congénitaux du segment antérieur de la tunique
vasculaire du crystallin." The author goes into the whole question
of the varieties of persistent remains of the capsulo-pupillary and
pupillary membranes, and explains them developmentally. The
capsulo-pupillary membrane extends, during intra-uterine life,
from the equator of the lens as far as that part which corresponds to
the anterior extremity of the secondary optic vesicle. The
capsulo-pupillary membrane covers the part of the lens not covered by the
secondary optic vesicle, and of which the central part later on
forms the pupil. The author refers to the rarity of literary
references to vestigia of the capsulo-pupillary membrane. Only
eight cases were collected by Brückner, and Cosmettatos himself
adds a few more, including one by S. Stephenson (Ophthalmoscope,
May, 1908). This is what he says of their appearance:—
"Vestigia of the capsulo-pupillary membrane are represented by
little opacities in the form of dots, striae or spots of different sizes.
Their colour is white, brown, or clear. They vary in number, and
are situated at the periphery of the anterior capsule. But sometimes
the opacities occupy the pupillary region also. In this case we have
a combined form of vestigia of the capsulo-pupillary and pupillary
membranes.” No doubt some of the appearances correspond to
those described by myself, but a more important statement from
the point of view of the question of the aetiology of my cases is the
following. The author is speaking of vestigia of the pupillary
membrane as distinct from the capsulo-pupillary membrane.
“Membranous vestigia of the pupillary membrane which start
from the pupil border seem to be very rare, according to
Brückner, since this author has never been able to find a typical case. Never-
theless, there have been seen vestigia of the central part of the
pupillary membrane which leave the posterior border of the pupil
in the form of thin veil-like membranes or of membranous filaments,
and portions of this membrane have also been seen occupying the
pupil area partly or completely and attached to the capsule. According to
Brückner these vestigia occur in connection with the border of the embryonic pupil before the formation of the pupil border proper (bord pupillaire definitif) which takes place after the
eversion of the pigment layer of the iris on to the anterior layer
(ectropion of the pigment layer of the iris). . . . The filaments
which start from the pupil border are more common. They
present the appearance of posterior synechiae of the iris, and are
either inserted into the anterior capsule or float free in the pupil
area. In the former case atropin does not dilate the pupil.”

One could go on giving extracts from Cosmettatos on this
interesting subject. I have, however, made sufficient reference
to his work to emphasise the point that, after all, these seven cases
may be purely developmental, every one of them. Until the advent
of the two last cases (Nos. 6 and 7) in which the appearances
suggested past iritis, I had no doubt about it. I am inclined to
go farther, and, as it were, to carry the war into the enemy’s camp
by suggesting that some of the cases which, faute de mieux, have
been described as “quiet iritis” are in reality cases of develop-
mental vestigia.

As a last word I may make the remark that the occurrence of
seven similar cases—or six if one excludes Case I—of
this kind in 7,500 children is of sufficient significance to suggest
that such cases should be carefully looked for and further studied,
not only because of their inherent interest, but because it is
important to distinguish cases of iritis or sequelae of iritis from
congenital anomalies in the development of the iris and the anterior
lens capsule.
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