de Schweinitz (G. E.)

Coloboma of the Iris, Polycoria,

AND

Primary Glaucoma.

By G. E. de SCHWEINITZ, M.D.,

Of Philadelphia.

[*Reprinted from American Ophthalmological Society Transactions*, 1891.]

[Reprinted from American Ophthalmological Society Transactions, 1891.

COLOBOMA OF THE IRIS, POLYCORIA,

AND

PRIMARY GLAUCOMA.

By G. E. de SCHWEINITZ, M.D.,

OF PHILADELPHIA.

The occasional association of primary glaucoma with complete or partial congenital absence of the iris, has been noted in several instances. The most recent paper on the subject is one by E. Treacher Collins (*Ophthalmic Review*, April, 1891), in which he reports a case of primary glaucoma with apparently complete aniridia, and refers to the case of congenital coloboma of the iris and primary glaucoma recorded by Mr. Lang (Trans. Ophth. Soc., U. K., Vol. X, page 106), the instance of double microphthalmos and glaucoma, with partial absence of the iris, reported by Brailey (*Ibid.*, page 139), and an example of glaucoma secondary to dislocation of the lens in a patient with nearly complete aniridia, described by Armaignac (Mémoires et Observations d' Ophtalmologie pratique, page 239). To

this list I will add a case of coloboma of the iris, polycoria, and primary glaucoma. The history is as follows:

Mrs. M. M., aged 52, an American by birth, presented herself for treatment January 26, 1891, with the following statement: For several years she had noted with her left eye iridescent circles surrounding lights, most marked, according to her statement, when looking at the moon, but had experienced no pain. In the spring of 1890, the vision in the left eye became very poor, with intermitting pain in the peri-orbital and temporal regions. Two weeks before her visit for treatment, she suffered violent pain in the left eye and almost complete suppression of sight, the attack lasting about forty-eight hours. Since then she has had a number of similar attacks of less severity, and characterized chiefly by temporary obscurations in vision. The patient is a well-preserved woman, in good general health, and with a clean family history. Her previous illnesses have been rheumatism and malaria, neither of violent type. Her children are healthy, and have normal eyes and natural vision. There is no instance of blindness or congenital ocular defect in her ancestors. The following is the record of the examination of the eyes:

O. D. $\frac{20}{xl\frac{1}{2}}$ with $-$ 1.25^c axis 15 $\frac{20}{xx}$; $+$ 2.50s. D = 0.50 p. p. 22 cm.

O. S. $\frac{20}{c}$ unimproved by glasses; with $+$ 2.50 spells D = 1 with difficulty.

O. D. Oval disc, gray red; small central excavation; several spots of disseminated choroiditis.

O. S. Oval disc, excavated to the scleral border, the center of the excavation measuring -2 D; strong arterial pulse; below the disc a large irregular patch of retino-choroiditis, at first yellowish-white in appearance and farther towards the periphery presenting pigment dots. A smaller similar patch in the macular region. Coarse injection of the episcleral vessels; steaminess of cornea; shallow anterior chamber; T $+$ 3. The iris of this eye presented a congenital coloboma down and in its axis at 75 degrees. Up and out from the coloboma were

three supernumerary pupils, the upper being crossed by two fibres. In addition to these defects of the iris there were considerable patches of atrophy, the largest being adjacent to the uppermost supernumerary pupil, lying to its inner side. (See colored drawing.) In the center of the pupil lying on the capsule there was a small, irregular, brownish, faintly granular tag. Figure 1 represents the field of vision. The outer boundary marks the limits of the normal field ; the transverse shading where vision was lost. It will be noticed that the greater portion of the nasal field is wanting, together with a small strip on the temporal side. Iridectomy was declined ; eserine was

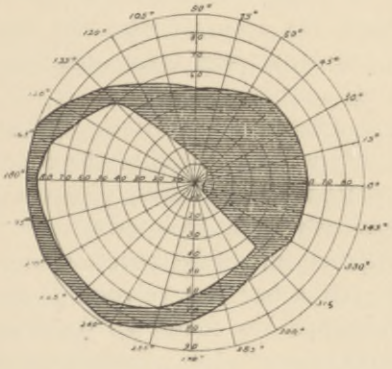

FIG. 1.

ordered. The next day the vision of the affected eye had risen to $\frac{20}{XL}$. In the fields of vision for white, blue, and red, in addition to the loss of the greater portion of the nasal field, there was slight increase in the shrinking of the field below. Both the blue and red fields were contracted, the greater proportional contraction being present in the red field. Two days later, in spite of eserine, the vision was $\frac{20}{C}$, and the day succeeding this, $\frac{20}{O}$, fingers being poorly counted at one foot ; T + 2. The patient was not seen after this until the 13th of February, having used the eserine continuously, and also having taken iodide of potash. She then reported one attack of general head pain, with some redness of the eye. The vision was barely $\frac{20}{CC}$, the

cornea very steamy, and the tension + 2. The lens was slightly opaque, and no view could be obtained of the eyeground. Iridectomy was urged, and the following day when the tension was unchanged, but the steaminess of the cornea and the pain had materially increased, and the central vision had sunken to counting fingers; this was permitted. This was performed by inserting the keratome at the position of the congenital coloboma, seizing the rim of iris and a portion of the adjacent tissue and excising it. A sudden movement of the patient caused the point of the keratome to come in contact with the capsule of the lens. Reduction of tension, and freedom from pain were the immediate results of the operation, but the accident just noted caused an opacity of the lens. No glaucomatous attack has occurred since the operation, and the patient has been free from pain. An extraction of the opaque lens may restore vision to the eye.

The first point of interest in this case and in all cases analogous to it, evidently is the relation of the congenital defect of the iris to the development of primary glaucoma. This vice of conformation apparently ought to interfere with the substantiation of the idea that closure of the filtration angle and increased tension bear to each other the relation of cause and effect Fortunately, Mr. Collins has found an opportunity of making pathological and microscopical examinations in two cases which throw light upon this subject. The first case was one of congenital aniridia in which the cornea ulcerated, the eye became glaucomatous and a staphyloma formed in the ciliary region. After the removal of this eye it was found that the ciliary body, with its processes, was present, and ended anteriorly in a small projection which proved to be the representative of the iris. The microscope revealed that the filtration area of the cornea was occluded by a close adhesion to it of the stump in which the ciliary body terminated. Mr. Collins's second case was one of traumatic aniridia, the eye having become blind and glaucomatous, the nerve being deeply cupped. On section the following lesions were found: round celled infiltration of the anterior part of the cornea; avulsion of the iris at its extreme root; atrophy of the ciliary body; and intimate adhesion of the most anterior ciliary processes to the posterior surface of the

cornea at its periphery, in the region of the ligamentum pectinatum. Illustrating the same point from the clinical standpoint the case of A. W. Natanson (Abstract in *Centralblatt f. prak. Augenheilkunde*, Supplementheft zum Yahrgang, 1890, page 489) may be quoted. This observer studied an example of aniridia and traumatic aphakia, with elevation of the intra-ocular tension (T+1). As a result of a blow with a piece of wood, the left eye of the patient was deprived of the iris and lens, and none the less secondary glaucoma with increased tension, developed twenty-seven days after the accident. From the cases examined by Mr. Collins, it fairly may be concluded, as he points out, that examples of partial and even complete aniridia form no exceptions to the rule that development of glaucoma is associated with blocking of the filtration area of the cornea.

The second point of interest in this case is the character of the congenital defect itself. The coloboma does not extend entirely across the iris, but stops a short distance from the ciliary margin and in this sense is incomplete. This fact, added to the others quoted from Collins's researches, serves to explain the existence of the glaucomatous tension. As is usual with unilateral defects of this character, the left eye is affected, and the fissure is directed downward and inward, inclination from the perpendicular being about 15°. In addition, three distinct apertures or supernumerary pupils are present, occupying the upper and outer part of the iris, the uppermost of the three in a line with the coloboma, being crossed by two thin bands, probably remains of the pupillary membrane. The remainder of the iris is considerably atrophied, especially in three patches, the largest one of which occupies the upper and inner quadrant, and the smallest, oval in shape, is adjacent to the lowest aperture from which it is separated by a band of nearly normal iris tissue. This patch of atrophy and the aperture near which it is situated, have almost exactly the same shape and size. Had the iris tissue in the three atrophic areas completely absorbed, six apertures, in addition to the coloboma, would have existed. As is well known, the number of so-called supernumerary pupils may be considerable. Mittendorf (Trans. Amer. Oph. Soc., 1884) has reported one case in which five distinct openings were

present. It seems probable, at least in human eyes, that no true instance of doubling of a normal pupil has been observed, and that the term of polycoria describes the existence of several openings in one iris.* In the majority of cases, in addition to the central pupil, one or more apertures are placed eccentrically which do not represent malformations due to arrested development, but either are defects in the iris tissue arising from an incomplete evolution of the choroid, or are dialyses, *i. e.*, separations of the attachment of the iris. The patches of atrophy in the present case appear to be imperfect apertures, the defects not having gone on to complete absorption of the structure. The small granular tag lying in the pupillary space (not shown in the colored drawing), undoubtedly represented the remains of the capsulo-pupillary membrane. The association of capsulo-pupillary membrane and coloboma of the iris, and the etiological questions which this raises are interesting points which have received considerable attention in the papers of O. Plauge (*Archiv. f. Augenheilk*, Bd. xxi, page 194) and Seggel (*Klin. Monatsbl. f. Augenheilk*, August, 1890, page 299), the former observer concluding that in laterally placed iris colobomas, in contradistinction from those which are situated below, anomalies of the pupillary membrane of the embryonal eye should be regarded as the most important etiological factor. The patches of retino-choroiditis present in both eyes deserve notice. In the left, or glaucomatous eye, the large area below the disc at the first glance suggested the possibility of coloboma of the choroid, an anomaly reasonably to be expected owing to its frequent association with a similar defect in the iris. The lesion, however, was an ordinary area of exudative retino-choroiditis, and not a developmental deficiency. In this connection it is interesting to recall the theory that many cases of arrested development are due to intra-uterine sclero-choroido-retinitis (Deutschmann), and the fact that cases of polycoria have been placed upon record (Seeley), the subjects of which have become blind from choroiditis. Finally, the choroiditis may have been the primary cause of the glaucoma. In one of Mr. Collins's cases, this was attributed to a hemorrhage into the lymph space between the choroid and sclerotic.

*Franke E. Ueber Angeborene Polykorie Klin, *Monatsbl. f. Augenheilk*, August, 1889, s. 386.