Congenital Aphakia with Hyaloid Artery and Retinal Fold

H.

Weve

From the University Eye Clinic Utrecht [Director: Prof. Weve]

Our observations lead to the opinion that every transition is to be found between simple arteria hyaloidea persistens and so-called pseudoglioma caused by congenital detachment.

Congenital Aphakia with Hyaloid Artery and Retinal Fold.

A beautiful example of so-called hyaloid artery combined with partial “retinal fold” recently came under my observation.

The 16-year-old Jan v. O. from De Bilt was sent by his employer because his right eye showed a convergent squint. This has existed from earliest youth. He showed the interesting picture of congenital aphakia with hyaloid artery and partial retinal fold.

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By slitlamp examination and dilated pupil one saw a completely flat and transparent membrane. In the periphery, it contained some white cataractous lens residues with cristals and some semi-transparent masses resembling spawn of frogs. Much deeper in the vitreous and completely free from the residues of the lens one saw at the nasal side, the head of a massive arteria hyaloidea. From this head a star of fine lines (threads or folds in a flat membrane of hyaloid) emerged in the direction of the ora serrata.

The hyaloid arteria forms a thick white skin of about the diameter of the optic disc and runs straight in the direction of this disc which is partly covered by it. From here it runs vertically down in the shape of a retinal fold adhering to the choroid in a cicatricial and pigmented area. This fold is about 5 papil-diameters long.

To imitate the stereoscopic effect of the ophthalmoscopic picture a shadow is painted on the fundus oculi.

Another little fold runs temporally upwards.

The eye has never been wounded and no trace of perforation of the cornea could be found. We must, therefore, suppose that the condition of the lens is not of traumatic origin.

The eye is amblyopic.

The left eye is myopic (9 dptr) and shows changes of the optic disc that are probably also congenital. The optic disc is oval shaped and shows at its temporal side an atrophic area that is not sharply contoured as myopical degenerations generally are.

The mother is 5 dptr hypermetropic.

A 9-year-old sister is strongly myopic with myopic astigmatism.

A 13-year-old brother is emmetropic in both eyes.

A 10-year-old cousin is hypermetropic 3 dptr. One eye is amblyopic and squints.

There is no consanguinity of the parents.