BILATERAL CONGENITAL ANOPHTHALMOS*

BY

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BUDAPEST

ANOPHTHALMOS is one of the rarest of the developmental anomalies of the eye and it is usually bilateral. In the case described below an orbital cyst and other developmental anomalies were present.

Congenital anophthalmos was first described by Lyscostenes and later by Schenk (1609) and Barthelin (1657), but Briggs was the first to mention the hereditary character of this disease (Sorsby, 1934). At the beginning of the nineteenth century Briggs described a family in which, out of seven children of healthy parents, three were born with bilateral, and one with unilateral anophthalmos. Later Monteath (1821) and Walker (1831) emphasized the hereditary nature of the disease.

Manz (1876) says that in most cases of congenital anophthalmos rudiments of the eye can be found in the orbit. When this is not the case, however (Röder, Gradenig), the optic nerve is imperfect; it may be represented by strands of connective tissue, or it may be absent (Rudolphi). The foramen opticum may be constricted. Rudolphi claims that it is impossible to distinguish exactly between this picture and microphthalmos, and that anophthalmos is not a developmental defect of the eye, but atrophy due to some unknown injury in foetal life. He believes the pathogenesis of microphthalmos and anophthalmos to be the same.

Landesberg (1877) pointed out the importance of blood relationship between the parents. Hippel (1899) showed that cases of microphthalmos and of bilateral anophthalmos can occur in the same family.

Recent reports tend to confirm these earlier views. Cecchetto (1920), Ougaud (1922), Langon (1926) and Wirth (1938) all mention the parents as being related by blood. Hanke (1904), in a case of congenital bilateral anophthalmos could find rudiments of the eye, consisting of very small choroidal remnants embedded in fibrous connective tissue only by histologic examination after death. The infant had neither optic nerve nor tract on either side. Triepel (1920) reports that in his case there was atrophy of the optic nerve and chiasm. Gallemaerts (1924), in a case of bilateral anophthalmos found, after histologic examination, rudiments of the eye on the left side only; on the right side there were no traces of the eye.

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Neither side had optic nerve, or chiasm; the corpora quadrigemina were defectively developed. Ougaud (1922) studied a family in which anophthalmos, choroidal coloboma and interstitial keratitis occurred. Canneyt and Vandemeulebroecke (1937) found unilateral and bilateral anophthalmos, microphthalmos and iris coloboma in four generations of one family. Wirth (1938) gave the history of three generations of a family with unilateral and bilateral anophthalmos, coloboma of the iris, the choroid and the crystalline lens, and congenital cataract.

In 1948, in this Journal, I presented the case of a ten-day-old boy with right-sided anophthalmos and orbital cyst, together with other developmental anomalies (proboscis on the right side, hare-lip and cleft palate). The rudiments of the eye were apparent only by histological examination of the contents of the orbit. Soon afterwards, the children's ward of our hospital, from which this patient had come, sent a six-months-old boy with congenital bilateral anophthalmos to the eye-department for examination.

FIG. 1.

FIG. 2.

He was the second child of a healthy mother of 22 years of age. The parents were not related. Wassermann reaction was negative. No developmental anomaly had ever occurred in the family.

Ophthalmological report:—Lids sunken on both sides, palpebral fissure shorter than normal (9 mm.), no eye on either side conjunctival sac very narrow. Rudiments of the eye neither visible, nor palpable. No other developmental anomalies. (Figs. 1 and 2).

X-ray examination:—The bony wall of the orbit, as well as the optic canals much narrower than usual on both sides. In profile the forehead appeared flattened, the cranial basis shortened, the three cranial *scalae* situated on one level. The sella smaller than normal, but clearly defined. Encephalography revealed no essential anomaly.

The child being alive and well, there was neither *post-mortem*, nor histological examination.

According to the literature, in cases of congenital anophthalmos there is generally a record of blood relationship between the parents or a family history of anophthalmos or other developmental anomalies. It may happen, however, as in the cases of Hasner (1876), Wicherkiewicz (1899), Hanke (1904), Triepel (1920), Ventola and

Zembrano (1946) that neither were the parents related, nor—which is more important—did the condition appear to be hereditary or associated with other developmental anomalies. This is not inconsistent with the hereditary character of the disease. It may be that the parents cannot give exact information about their family history; moreover, the child afflicted with anophthalmos may become the founder of a family suffering from this or some other developmental anomaly.

By clinical examination I could not detect rudiments of the eye on either side. But it has been shown in the literature that when the child dies and *post-mortem* or histological examination can take place, rudiments of the eye are usually found in the soft parts of the orbit. From the X-ray examination we may deduce that the optic nerves or perhaps the chiasm and tracts were defectively developed but positive radiological proof of this is lacking.

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ACUTE A THERAPEUTIC STEP IN CHRONIC GLAUCOMA*

BY

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A MEASURE that is sometimes very useful in treating an attack of acute glaucoma is the retrobulbar injection of novocain (procain), administered not pre-operatively, but as an isolated therapeutic incident (S. Gifford, M. J. Icaza). It does not of course have a permanent effect, so that an operation becomes necessary sooner or later; but the object of the injection is to avoid the performance of

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