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AN ADDITIONAL CASE OF DOUBLE CONGENITAL MICROPHTHALMOS.

BY CASSIUS D. WESCOTT, M.D.

During a trip in Minnesota last September, I was asked by my friend, Dr. E. J. Lewis of Sauk Center, to see Maxwell James, aged 2 years, who was said to have been blind from birth.

Upon examination the child seemed to be in perfect health and his development was good for his age. I have been unable to secure a photograph showing the condition of the eyes, but the one which I present will give a general idea of the little fellow's appearance. The lids and orbits were normal. The palpebral fissure of the left eye was not smaller than usual, but the lids of both eyes appeared sunken. Both eyeballs were smaller than normal, the diameter of the cornea measuring no more than 6 or 7 mm. in left eye and 5 or 6 mm. in the right. The iris was present in the left eye, but the pupil was not well made out, because of a small central opacity of the cornea, and the fact that the opaque sclera seemed to shade off into the cornea, leaving a narrow ring of clear cornea between an outer opaque ring and a central circular opacity. The right eye was decidedly smaller than the left. There was almost no clear cornea, the iris showing faintly through the almost opaque membrane. The movements of the eyes were normal, but limited. No cysts could be demonstrated in either orbit and the conjunctiva was smooth and free and gave no sign of past inflammation. There

was evidently vision in the left eye, for the child could see to get about, and was immediately attracted by bright objects held before it.

The baby had always been perfectly well and presented no other physical anomaly. The parents were Americans of the middle class, and had always enjoyed good health. They had the appearance of health when I saw them, but both were spare and the mother did not seem to be especially vigorous. She had borne but two children, gestation being without incident and delivery natural in both instances, but both children had been born blind. The first, a girl, had died at 18 months, of some acute infantile disease, but up to that time had seemed well and normal, except that the eyes were almost exactly like those of the little boy, whose case I have just described.

A careful inquiry failed to elicit any cause for these two cases of microphthalmos in the family, and the parents had no theory to suggest. So far as the mother knew, or would tell, nothing had happened during the intra-uterine life of the infants which might interfere with their development. Although an itinerant professor had promised to cure the eyes of the little boy, for $50 cash, in advance, I gave it as my opinion that little change was likely to occur, although it might be possible to slightly improve the vision of the left eye by iridectomy, if it should increase in size with the growth of the child.

Treacher Collins has reported ("Transactions of the Ophthalmological Society of the United Kingdom," Vol. xiii) two cases of congenital microphthalmos, in which he secured specimens and made elaborate examinations. He divides these cases into two classes: First, those in which the eyeball is simply abnormally small, *i. e.*, very highly hypermetropic eyes, in which, beyond the defect in size, there has been no arrest in development. Second, those cases in which the eyes, besides being small, have some other congenital abnormality, resulting from imperfect closure of the fetal ocular fissure. This last class may again

be divided into those where there is only a slight defect, and where the eyeball retains nearly its normal shape, and those in which the accompanying abnormality is very gross, the eye being usually exceedingly small, while connected with it are one or more cysts. Thess cysts are often large, sometimes concealing the globe, which is situated at the back of the orbit, so that its presence can not be ascertained by clinical examination alone.

His first case was of the class to which my own probably belongs, and I trust you will pardon me if I quote his report:

The patient was an infant who died when four days old, from congenital heart disease. The aorta and pulmonary arteries formed one common trunk, and there was a defect in the ventricular septum. The right eyeball measured 19 mm., antero-posteriorly and laterally, and 8.5 mm. vertically. There was a small prominence in the sclerotic posteriorly, immediately below the optic nerve. The iris was well developed, but several delicate fibers stretched across the pupil, evidently remnants of the pupillary membrane. The ciliary body presented its usual appearance. The lens was in its normal position and the vitreous appeared healthy. The retina had numerous little rucks in it. At the seat of the prominence below the optic nerve there was a gap in the sclerotic, through which a fold of retinal tissue projected.

Microscopic examination revealed no central blood-vessel in the optic nerve. The nerve fibers, after having passed through the lamina cribrosa, all sloped somewhat downward; the upper ones afterward curved backward to the upper part of the retina. Below the optic nerve there was at first no retina, no uveal pigment layer and no choroid, only a mass of nerve fibers with fibrous tissue external to them. This fibrous tissue, representing the sclerotic had, a short distance from the lower margin of the outer optic nerve, a break in its continuity. Into the gap thus formed the nerve fibers passed, expanding somewhat after they were through it and becoming mixed with bodies like those found in the nuclear layers of the retina; external to this nerve tissue was a thin layer of fibrous tissue. On the side of the gap in the sclerotic, farthest from the nerve, the retina commenced with its usual layers, being continuous with the nerve fibers emerging from the gap; around the edge of which, also, the uveal pigment layer turned. The tunics of the eye were traced forward from the break in the sclerotic. The retina was at first seen to possess its usual layers, the uveal pigment was present, also the choroid, which, however, was more cellular than normal and devoid of pig-

ment. A short distance farther on the choroid ceased and the uveal pigment layer became an unpigmented layer of cells.

The left eyeball measured 18 mm. antero-posteriorly, 18 mm. laterally and 16.5 mm. vertically. The cornea measured 8.25 mm. latterly and 7.5 mm. vertically. There was a prominence in the sclerotic posteriorly, below the optic nerve, more pronounced than in the right eye. There was a small coloboma of the iris downward. The lens was situated nearly in the center of the globe, a considerable space being left between it and the posterior surface of the iris; it was almost globular in shape, measuring 5 mm. antero-posteriorly and 5.5 mm. laterally. The ciliary process, thin and much elongated, passed almost directly backward toward the sides of the lens. In the center of the vitreous was a band of fibrous tissue, which passed forward from the back of the globe below the optic nerve to the posterior pole of the lens; this band was broad posteriorly and gradually tapered off as it passed forward. Around this central band the vitreous appeared normal. The retina had some small rucks in it. Below the optic nerve, in the situation of the prominence seen externally, a fold of retina seemed to be included in a gap in the sclerotic.

Microscopically the structure of the cornea appeared normal. In the position of the coloboma of the iris the uveal pigment ended about on a level with the point of termination of Descement's membrane in the fibers of the ligamentum pectinatum. The fibers of the suspensory ligament, passing from the much elongated ciliary processes, sloped backward and a little inward to the sides of the displaced lens, which was very rudimentary in structure. The ciliary body, choroid, uveal pigment, and retina in the upper part of the eye appeared healthy. In the lower part, the ciliary muscle was much elongated, and the choroid could only be traced backward as far as the equator of the globe, its posterior part being absent. The uveal pigment layer, just previous to the termination of the choroid, was thrown into several folds; from this position it was continued backward, first as a layer of unpigmented cells, and afterward as tissue which looked like degenerate retina. The band of tissue passing through the center of the vitreous was composed of bundles of nucleated fibers, with small blood-vessels coursing among them; it was adherent posteriorly to the sclerotic, and anteriorly, where it was very thin and reduced to but a few fibers, it joined the posterior capsule of the lens. The vitreous was continuous with it at the sides. There were no central blood-vessels in the optic nerve; all its fibers, after passing through the lamina cribrosa, bent directly downward, the upper ones curving round again to the retina above, which began in a fold. The lower ones seemed to end abruptly at the band of fibrous tissue which passed forward through the vitreous. Below this band of fibrous tissue some very degenerate looking retina, consisting mostly of nuclear bodies, was seen

embedded in a gap in the sclerotic. The retina in the lower part of the eye had its inner granular separated into two; some fibers separated by spaces running transversely between them.

Hirschberg reports (*Berliner Klin. Wochenschrift*, 1870) the case of a girl of three months, with double microphthalmos. She was well developed, but her eyeballs, though well formed, were very small. The horizontal corneal diameter on both sides was about 5 or 6 mm. There was coloboma of the right iris, below and inward, and persistent pupillary membrane. In the left eye there was luxation of the opaque lens downward, and a persistent hyaloid artery. The fundus showed an extensive discoloration (coloboma). The child continued to develop and the eyes grew somewhat larger, but no perception of light could be discovered. The parents were perfectly healthy.

Rindfleish described (*Archiv. f. Ophthalmologie*, Leipsic, 1891) a case of bilateral microphthalmos, with cystic posterior ectasia, complicated with hydrocephalus. The ventricles were dilated, the cerebellum was cystic and the hemispheres of the cerebrum were separated from the cranium by a collection of subarachnoid fluid. The irregularities of the base of the skull had been obliterated by internal pressure. The roofs of the orbits were concave upward, and the orbits thus depressed contained microphthalmic eyeballs, with connecting cystic structures.

Williams of Boston has reported (*Boston Medical Journal*, 1850, Vol. xliii, page 421) a case of microphthalmus, complicated with congenital cataract of both eyes, in a boy 13 years of age. The entire globe was extremely small, and much sunken in the orbit. The iris, in both eyes, was hardly more than half the average size in a new-born infant, the pupil scarcely larger than a mustard seed. The entire field of the pupil in the right eye was occupied by a cataract, apparently formed by the capsule only, the lens having been absorbed. The pupil of the left eye presented an opacity covering nearly its entire area, but a small point existed toward the temporal side, which was not occu-

pied by cataract, and the opacity was less white than in the right eye. Dr Williams successfully operated upon the cataracts, but with what result as to vision I am unable to say.

Bernheimer reports (*Archives of Ophtholmology*, 1894) a remarkable case of hydrocephalus internus, in which the eyes of the patient were normal in external appearance, but very small. The muscular and lachrymal apparatuses were normal in both. The left pupil was not circular. The axis of the left eye measured 12 mm., and lying to the lateral side of the optic nerve was a cyst 6 mm. long, filled with clear fluid. The right eye was continued posteriorly in a thick stalk, and was about the size of the left. In place of the nerve was a sheath containing but a single vessel, and to its lateral side was a cyst continuous with the posterior and inferior portion of the ball. The lens was small, egg-shaped and cataractous. The retina was completely detached and much folded. The optic nerve had a peculiar appearance, and nerve tissue could scarcely be found. A third of the nerve tissue was converted into an egg-shaped cyst, communicating by a narrow cleft with the cavity of the eyeball. The pigment epithelium and the choroid were normal and in position. Near the choroidal ring the choroid was somewhat thickened and choroidal pigment was traced back to the wall of the cyst.

FitzGerald reported (*British Medical Journal*, 1886, Vol. ii, page 1385) a case of double congenital microphthalmos to the Royal Academy of Medicine in Ireland, and exhibited the patient. There were two undeveloped globes, with cornea and iris, but little or no anterior chamber; illumination of the fundus was impossible. The father of the child thought it could see when light was brought into the room, but after having several times made trial with both lamp and ophthalmoscope, Dr. FitzGerald was unable to satisfy himself that the child saw anything. The mother in this case gave a history of a fright during gestation, but at what period was not stated.

Brailey reports (Transactions of the Ophthalmological Society of the United Kingdom, Vol. x, page 139) a case of double microphthalmos, with defective development of the iris, in a patient with imperfect teeth and arms, seen at Guy's Hospital. Vision of the right eye equalled fingers at 10 feet, and the field was much narrowed all round. Tension was +1 and the eye was myopic about twelve diopters. Vision was not improved by lenses. The cornea was small, about 9 m.m. in diameter. The iris was absent except a narrow crescentic piece in the inner side, which occupied about two-fifths of the circle. The lens was in situ. Its diameter seemed to equal that of the cornea. The disc was cupped and had a myopic crescent to the outer side, and opaque nerve fibers to the inner side. Vision of the left eye equalled fingers moving. Tension was +2 and the eye was myopic. The cornea was the same as in the right eye. The iris was wanting except for three small isolated bits with rounded edge, situated on the inner side of the vertical line. The disc was cupped.

De Vincentiis of Pavia, reports (*Ann. di Ophth.*, Vol. xiv) a case of congenital bilateral microphthalmos, with defective development of the heart. Mayer described a case of microphthalmos with cysts in the lower eyelids. Ginlini reports (*Ztschr. f. Verleich. Augenh.*, Wiesbaden, 1891) a case of double congenital microphthalmos in a dog four weeks old. Holtzke (*Archives of Ophthalmology*, Vol. xii) made careful studies of the eye of a rabbit with coloboma of the iris, ciliary body and choroid, and general microphthalmos. He closes his report as follows: "The explanation of the above-described changes is very simple. There have evidently been extensive inflammatory processes in the eye, and I see no reason why we should not adopt Deutschmann's view (*Klin. Monatsbl. f. Augenh.*, March, 1881) that the arrest of development is only a changed development of the eye in consequence of an intra-uterine inflammation. This inflammation has probably taken place in the

latter half of the third week; the fetal fissure was nearly closed, or after having been closed, was again opened; the iris, on the other hand, had not begun to develop. Why this inflammation was just at this fissure, which is so important for the development of the eye, can not be decided, unless it was favored by the greater vascularity of this neighborhood."

Hess reports (*Archiv. f. Ophth.*, Bd. xxxiv) and gives details of the microscopic examination of a series of microphthalmic eyes with colobomata; he found in his cases no sign of past or present inflammation and concludes that it has very much less to do with the production of congenital malformation than is generally assumed.

As I have no specimen to present, and have had no opportunity to make pathologic studies of microphthalmic eyes, I have no theory to offer as to the causation of these anomalies. The appearance of the eyes in my own case would cause one to think immediately of an inflammatory process.

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