A NOTE ON TWO CASES OF MEGALOCORNEA

by

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Although over 80 cases of megalocornea are on record (Duke-Elder, 1938), cataract extractions in the condition are sufficiently rare to rank as unusual surgical procedures. It would seem worth while reporting other cases.

On September 16, 1941, I was asked by Dr. Bourne of Dulwich Hospital to see a man with very large eyes. He was aged 58 years, and was recovering from haemorrhage from a gastric ulcer. He had also had severe bleeding from piles. He complained that since the illness his vision had failed considerably. Previously, vision had been good, in fact, he had been an army driver in the 1914-18 Great War. On examination I found he had bilateral megalocornea. The corneae measured 14 mms. in the horizontal meridian, and were large and lustrous. Slit-lamp examination showed no folds or splits in Descemet’s membrane. Each iris presented a somewhat atrophic appearance with a vertically oval pupil rather like that of a cat. There were very deep anterior chambers. The pupils reacted well to light and dilated widely with homatropine. The tension of the eyes was normal. Bilateral arcus senilis was present. (Figs. 1 and 2).

Both lenses showed cuneiform cortical lens opacities which effectually masked the fundus appearances. He could see hand movements, and projection of light was good in the left eye. In the right eye only the temporal half of the visual field was present. At this period the patient was still weak from haemorrhage from an ulcer.
demonstrated by X-ray on the lesser curvature of the stomach. His haemoglobin was only 48 per cent. of the normal.

By September 30, 1941, his general condition was much better, though the lenses were more opaque. Completely incapacitated by his poor sight he was transferred to Horton Hospital, Epsom, for cataract extraction.

Fig. 1.
The grandfather's right eye.

Fig. 2.
The grandfather's eyes.
Two cases of megalocornea

Enquiry showed that his grandson suffered from the same condition; and I was afforded an opportunity of examining the boy. He was a cheerful lad, his large and lustrous eyes giving him something of the appearance of a spectral Tarsier (Figs. 3 and 4). He too had an arcus senilis; but his pupils, unlike those of his grandfather, were circular. Fundi and lenses were normal and vision was 6/9 in each eye. There was no associated defect in intelligence in either case; the boy had won a scholarship, and the grandfather was a quick-witted cockney workman. I examined the boy's mother, whose eyes were in no way abnormal. No other members of the family were available for inspection; but the testimony of the three seen was so definite that I think the pedigree (Fig. 5) is reliable. It will be seen that my elder patient had an uncle with megalocornea. His mother was "partially paralysed from the knees downward." One of his own daughters had the same defect, which I had no opportunity of seeing. There was no consanguinity in the family.

Fig. 3.
The grandson's left eye.
Fig. 4.

Showing size of grandson's left eye compared to face.

Fig. 5.

(Paralysed feet)
TWO CASES OF MEGALOCORNEA

It will be noted that all three cases of megalocornea were in males.

By November 29, 1941, the patient had recovered sufficiently from his gastric haemorrhage to justify cataract extractions. Owing to the rather intimidating accounts of such extractions in the literature, it was thought worth while to perform a trial extraction on the right eye, in spite of the poor projection. Many of the operative difficulties recorded seem to be due to pressure on the globe during extraction, so that it was thought best to essay an intracapsular extraction with forceps on the eye. The facial nerve was blocked with 5 per cent. novocaine and adrenalin by the O'Brien method; and absolute akinesia of the orbicularis was obtained. A novocaine-adrenalin retrobulbar injection was given and the patient left 15 minutes to allow his intra-ocular pressure to become as much lowered as possible by the adrenalin. I thought a large conjunctival flap would be useful; and never having performed a cataract section on such an eye before, I raised the conjunctiva above the cornea by a subconjunctival injection of saline to help in cutting the flap. It was interesting to see that the injection fluid, creeping forward subconjunctivally, raised the upper portion of the "arcus senilis" with the conjunctiva. A superior rectus stitch was inserted.

My cataract section turned out like any section in a less abnormal eye; and I cut a large conjunctival flap. It was thought that the oval pupil might present difficulties, so a complete iridectomy was performed, followed by an intracapsular extraction in the ordinary manner with Arruga forceps, no complication occurring. The conjunctival flap was sutured into place with 6/0 plain catgut. The lens, examined afterwards, seemed normal in size, a rather soft cataract with unbroken capsule and the usual distortion from forceps traction.

The eye recovered rapidly; and soon the causes of the eccentric projection could be seen. The disc was atrophic with blurring of the temporal margin. About 6 o'clock there was a patch of choroidal atrophy in the far periphery. The ciliary body could easily be seen through the iridectomy. There were some vitreous opacities present; but the patient was delighted with the increase of vision in what he had been assured was likely to be a useless eye (Fig. 6). 

Exactly the same technique was employed when the second cataract was extracted on December 29, 1941. This time the patient was a little nervous, rolling his eye while the iridectomy was being performed, so that a narrow bridge of iris was unintentionally left at the pupillary margin. The bridge was divided with de Wecker scissors before completing an intracapsular forceps extraction as in the other eye.
There was no complication during recovery save a few lines of striate keratitis during healing, presumably from the very large section. There were vitreous opacities in the left eye also; otherwise the fundus was normal. The patient has been frequently seen since. The left vision is 6/9 with a +9.0 D.Sph. and +2.0 D.Cyl. axis 180° correction.

No definite cause was found for the partial atrophy of the right optic nerve. The patient was certain the eye had been good before the gastric bleeding, which may well have been the cause.

Remarks

Cataract is a common complication of megalocornea, Veil and Sarrazin (1939) even stating that it is invariable by the time the patients reach the age of 40-50 years. Vail (1931) summarised 69 reported cases of which 27 had cataract. Of these six are described as having anterior capsular, four posterior subcapsular, and two nuclear opacities. There were three, Morgagnian and two black cataracts. No details are given of the remaining 10 cases. Vail (1931) reported 18 extractions by eight surgeons, and three cases
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have been reported without detail since. The favourite technique seems to have been iridectomy followed by a spoon extraction, with or without counter-pressure (10 cases). Iridectomy, extracapsular extraction, and later needling, have been done in two cases. One case each are on record by simple extraction, delivery by needle, delivery by forceps, and the Madras method. Vail noted that vitreous was certainly lost in seven cases, probably he thinks, in three other vaguely reported cases. Three extractions were complete failures, three were followed by retinal detachment later, and in three cases iritis is noted. Probably in 12 cases useful vision was obtained.

So many cases are reported with a tremulous lens that intracapsular extraction seemed worth trial in the case reported above.

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REFERENCES


INCLUSION CONJUNCTIVITIS

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INCLUSION conjunctivitis is a term which includes three forms of follicular inflammation of the conjunctiva; these are swimming-bath conjunctivitis, conjunctivitis of swimming-bath type but contracted elsewhere than in swimming-baths, and inclusion,blennorhoea or non-bacterial ophthalmia neonatorum.

Each of them is caused by an identical virus infection which consists of bodies included within the cytoplasm of the epithelial cells of the conjunctiva. The inclusion bodies exhibit two phases of development known as elementary bodies and initial bodies, collectively they are called Halberstaedter-Prowaczek Körperchen (or for short H.P.K.) after their original discoverers.

Swimming-bath conjunctivitis is rarely seen in England but is not uncommon among young men on the continent of Europe. It is propagated by persons infected with inclusion urethritis or