Congenital duplication of the lens

R. H. B. GREY AND N. S. C. RICE

From Moorfields Eye Hospital, City Road, London

Congenital anomalies of the formation of the lens from the lens plate or lens vesicle are rare and it would seem that reduplication is exceptionally uncommon.

Duplication of the nucleus and cortex within one lens capsule has been recorded on several occasions (Von Graefe, 1854; Erwin, 1894; Clegg, 1914; Marquez, 1922). Although Erwin's case also had glaucoma none of these cases had any other abnormality of the eyes and the condition was unsuspected until the patients underwent cataract extraction. The operations were performed when the patients were aged between 57 and 73 years and were described as uneventful but, after removal of the lens nucleus, a second nucleus was subsequently expressed. These cases would seem not to have been true lens duplications. Von Szily (1938) in the Doyne Memorial Lecture demonstrated the mechanism of the origin of these duplicated nuclei and pointed out that in some areas the epithelium extended posteriorly behind the equator of the lens, thus leading to an irregular arrangement of lens fibres and to two nuclei and cortices, one in front of the other.

Logetschnikow (1893) described a case of a 48-year-old woman on whom he performed a cataract extraction. He noted a large temporal lens and a small nasal lens as opposed to two nuclei separated by a coronal split, but this case also appeared to have one capsule.

True duplication of the lens with separate capsules would seem to be uncommon. Richardson (1951) presented a case of a child of 11 months of age in whom there was considerable maldevelopment of the anterior and posterior segments of the eye. A central corneal tumour was present with a coloboma of the iris, ciliary body, and choroid; there were also two lenses. There was mesenchymal tissue extending from the cornea to the iris surface and the retina was attached to the posterior surface of one of the lenses.

Lyford and Roy (1974) published a case report of an eight-year-old girl with absence of one side of the nose, mental retardation, and unilateral

ocular defects. There was a bifid upper lid, coloboma of iris, ciliary body, and choroid and duplication of the lens.

This paper describes a case of true duplication of the lens which has features that distinguish it from previously reported cases.

Case report

A White seven-week-old baby boy was brought to an outpatient clinic by his parents. From the time of his birth they had noticed that his left eye seemed enlarged and had an irregularly-shaped pupil. Although the cornea was clear it appeared larger than the right cornea and there was a typical coloboma of the iris. The mother reported a normal pregnancy followed by an uneventful delivery; the birth weight was 7 lb 6 oz (3.4 kg). There was no family history of ocular abnormality. A diagnosis of congenital glaucoma was suspected and the child was admitted for examination under general anaesthesia.

The left cornea was oval measuring 13 mm in the 45° meridian and 11 mm in the 135° meridian. The structure of the cornea appeared normal, with no splits in Descemet's membrane, and it was transparent (Fig. 1).

FIG. 1 Photograph to show oval cornea and iris coloboma

The anterior chamber was of normal depth but the iris had an infero-nasal coloboma involving the whole width of the iris (Fig. 1). Behind the iris there were two well-formed, clear lenses both lying in the coronal plane of the eye. The supero-temporal lens appeared slightly larger than the infero-nasal lens and neither appeared to be dislocated. Both lenses appeared to be circular apart from the upper margin of the inferior lens which had a slightly concave outline (Fig. 2). There was a fine strand stretching from the superior margin of the inferior lens to the nasal edge of the pupil margin and from this point the strand expanded as a thin triangular fibrous sheet on the iris surface reaching the angle of the anterior chamber covering a short area of the trabecular meshwork. Elsewhere the trabecular meshwork appeared normal.

Examination of the fundus showed a coloboma of the choroid with an appearance similar to a drumstick; a thin choroidal defect extended forwards from the disc and expanded in the periphery. There was pigment hypertrophy around the edge of the expanded area, but the retina appeared to be intact over its surface with normal retinal vessels running across it (Fig. 3). The optic disc appeared normal. The intraocular pressure was 16 mmHg. The right eye was normal with a normal refraction.

General examination of the child revealed no other defects and x-rays of the head revealed no developmental abnormalities of the nose or skull. The mental development appeared normal and has remained so.

Discussion

During normal embryonic development the lens plate develops in the surface ectoderm in the

FIG. 2 *Photograph with retroillumination showing iris coloboma and duplicated lens*

FIG. 3 *Painting of typical partial coloboma of choroid*

second week (4 mm stage). This invaginates and forms a pit (5 mm) which then develops into a vesicle by the fourth week (7 mm). Some time before the fifth week (10 mm) the lens vesicle separates from the surface ectoderm and then becomes engulfed in mesodermal tissue. The determinant of these stages of development is the optic vesicle developing from the forebrain. The outer layer of the optic vesicle also acts as the determinant of the development of the choroid.

The case described in this paper showed a typical, partial coloboma of the uvea—that is, in the line of the embryonic cleft and involving only some of the layers of the developing eye. It would seem that there was a mild abnormality arising in the fusion of the outer layer of the optic cup at the 12–13 mm stage which led to the coloboma. Whatever the defect was that prevented this fusion it would seem reasonable to suggest that it had affected, at an earlier stage (5–7 mm), the optic vesicle determining the developing lens plate. Instead of the plate forming a single lens pit and vesicle the defective optic vesicle led to the formation of two lens pits, thus two separate lenses.

In the embryo before the 13 mm stage there are fibrils extending from the lens vesicle to the surface ectoderm—that is, the anterior vitreous. It has been suggested that these fibrils act as the basis for the mesodermal invasion, which later forms the corneal stroma. It would seem probable therefore that if there was an abnormality of the lens vesicle there would be a corresponding abnormality of anterior vitreous fibrils. This could explain the long axis of the oval cornea being in the same meridian as the duplicated lenses in the present case.

Richardson suggested that an abnormality of the central lens plate had led to the formation of two lens vesicles but this would not explain the development of a coloboma, which seems to be present whenever the lens is reduplicated. Also, unlike Richardson's case the other mesodermal structures of the eye appeared largely unaffected. The zonule of the lenses appeared normal and the iris stroma, corneal stroma, and anterior chamber were normal

apart from one fine strand from the inferior lens to the iris border and drainage angle.

The patient of Lyford and Roy had an atypical complete coloboma on the lateral side of the eye with unequal lenses. Presumably in this case the more fundamental defect of the fronto-nasal process had secondarily affected the normal fusion of the optic vesicle leading to abnormal determination of the lens plate.

Summary

A case of reduplication of the lens with uveal coloboma is described. This is a rare condition and, unlike the two previously reported cases, the other ocular structures and adnexae appeared normal.

The authors wish to express their thanks to Mrs W. Pennel for secretarial help and to Mr K. Sehmi for photographic assistance.

References

- CLEGG, J. G. (1914) *Trans. ophthal. Soc. U.K.*, **34**, 163
- ERWIN, E. J. (1894) *Ophthal. Rec.*, **3**, 433
- GRAEFE, VON A. (1854) *v. Graefe's Arch. Ophthal.*, **1**, 323
- LOGETSCHNIKOW, S. (1893) 'Khirurgitscheskaya Lepotis', vol. 3, pt 4, p. 946
- LYFORD, J. H., and ROY, F. H. (1974) *Amer. J. Ophthal.*, **77**, 315
- MARQUEZ (1922) *Klin. Mbl. Augenheilk.*, **68**, 305
- RICHARDSON, O. (1951) *Trans. Canad. ophthal. Soc.*, **14**, 102
- SZILY, VON A. (1938) *Trans. ophthal. Soc. U.K.*, **58**, 595