What may be the future of these spared eyes?

Without a long period of observation, it is impossible to say. One of our cases has been observed from the age of 45 to 62 years, and the spared eye was quite normal 14 years after the diagnosis of glaucoma on the other side (intraocular pressure : 15 mm.Hg; C = 0.31; visual fields normal).

Treatment

We emphasize only one point: if the diseased eye, with poor vision, is controlled only by the maximum medical therapy, and if sulphonamides by mouth have disagreeable side-effects, the patient prefers to have an operation on the glaucomatous eye and to have no treatment to the good eye.

We have never seen unilateral cortisone-induced glaucoma as observed by Chandler and Grant (1965).

Conclusions

(1) Angle-closure glaucoma may remain unilateral in 50 per cent. of acute cases. It is impossible to predict future structural changes in the anterior segment. Provocative tests are always valuable and prophylactic conventional iridectomy should be carried out on the fellow eye.

(2) Chronic simple glaucoma may be markedly asymmetrical, but the asymmetry is rarely so important that the glaucoma appears to be unilateral. If the spared eyes are studied by tonography, the dexamethasone test, and cyclopentolate tonography, they nearly all react like glaucomatous eyes.

So, if one accepts a hydrodynamic definition of glaucoma, unilateral glaucoma does not exist: there are only asymmetrical glaucomatous eyes. But if one does not accept this hypothetical concept unilateral glaucoma must exist.

(3) To explain why only one eye is diseased is another story.

My thanks are due to Dr. Y. Delmarcelle for his help in the preparation of this paper, and also to Miss M. T. Roque and Mrs. M.-C. Grenier.

Glaucoma associated with spontaneous displacement of the lens

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There are a number of disorders which may be associated with spontaneous displacement of the lens and it is instructive to study the mechanism of the glaucoma which may occur in certain of these conditions (Table I).

Simple ectopia lentis is a genetically determined condition with an autosomal dominant mode of inheritance. Glaucoma eventually develops in about one-quarter of patients

Table I *Spontaneous displacement of the lens*

Simple ectopia lentis
Ectopia lentis et pupillae
Late spontaneous dislocation of the lens

Associated with other ocular anomalies

- (1) Rieger's anomaly
- (2) Aniridia

Associated with spherophakia

- (1) Marfan syndrome
- (2) Marchesani syndrome
- (3) Homocystinuria

with this condition, but only if the lens becomes *dislocated* anteriorly or posteriorly. The glaucoma in these patients is truly lens-induced. The same can be said for the less common ectopia lentis et pupillae, an autosomal recessive condition in which the lens and slit-shaped pupil are displaced in opposite directions. Late spontaneous dislocation of the lens, the most common of these three conditions, is probably a variety of simple ectopia lentis. In this autosomal dominant condition, the patient, usually a young adult, presents with a dislocated lens with no previous history of subluxation.

If we compare the glaucoma in these isolated conditions with that in the next group, a difference emerges. In Rieger's anomaly, or mesodermal dysgenesis of the anterior segment, there is a faulty differentiation of the anterior chamber. In the angle, numerous pectinate-like fibres extend from the iris to a prominent ring of Schwalbe, while in severely affected cases the angle may be occluded by a forest of criss-crossing strands. The anterior stromal layer of the iris is markedly hypoplastic, and there may be pseudo-polycoria and corectopia. Dental anomalies occur frequently in association with the ocular changes: the teeth may be congenitally absent or small and malformed, or there may be defects in the enamel. Various skeletal changes have also been reported in patients with Rieger's anomaly. In these patients, whether or not the lens is subluxated, glaucoma is due to the developmental anomalies in the angle.

In aniridia, glaucoma occurs in about one-third of cases, while subluxated lenses are uncommon. Glaucoma is due either to mesodermal tissue filling the angle or to the development of anterior synechiae between the rudimentary fringe of iris and the cornea.

In Rieger's anomaly and aniridia, therefore, glaucoma in association with a subluxated lens is due to developmental anomalies of the angle and not to the displaced lens.

The third group of conditions, that associated with spherophakia, is the most important and the most interesting. The Marfan syndrome is characterized by skeletal, cardiovascular, and ocular changes. The skeletal changes are well known and include long extremities, arachnodactyly, kyphoscoliosis, and pectus excavatus. A long narrow face and a high arched palate are common. The cardiovascular changes consist of abnormalities in the media of the aorta and pulmonary artery which may lead to the development of diffuse dilatation or dissecting aneurysm. The classical ocular change is ectopia lentis, which can be found in over 95 per cent. of cases. The lens is small and spherical and is most commonly displaced upwards. Defects in the cornea and sclera may result in high myopia, keratoconus, megalocornea, and blue sclerae. The angle of the anterior chamber is frequently abnormal, containing pectinate strands, iris processes, or mesodermal tissue. Glaucoma may occur either as the result of these anomalies or following anterior or posterior dislocation of the lens. Anterior dislocation is the more common lenticular cause of glaucoma.

The much rarer Marchesani syndrome is also characterized by skeletal and ocular changes, the skeletal changes being the antithesis of those found in the Marfan syndrome. The patient has a short stature with short limbs and fingers, well-developed musculature, and an abundance of subcutaneous fat. In the eyes, spherio- and microphakia occur and a subluxated lens is frequently found, although it is said to occur later in life than in the Marfan syndrome. The angle of the anterior chamber is often abnormal with changes similar to those seen in the Marfan syndrome; these changes may produce glaucoma. Glaucoma may also occur if the lens dislocates anteriorly or posteriorly.

In the Marfan and Marchesani syndromes, therefore, glaucoma may be truly lens-induced if the lens becomes dislocated, but may in some cases be due to developmental anomalies of the angle.

The third condition in this group, and the one most recently described, is homocystinuria. This is an inborn error of metabolism due to the absence of an enzyme, cystathionine synthase, and is transmitted as an autosomal recessive trait. Skeletal, cardiovascular, central nervous, and ocular changes may be present, although not every affected individual needs to show all these changes. Kyphoscoliosis, genu valgum, and joint laxity are frequently seen, and excessively long extremities, resembling those of the Marfan syndrome are common. In the cardiovascular system thromboses in arteries of intermediate size, particularly the coronary, cerebral, and renal vessels, may occur and frequently cause death in these patients. Increased platelet stickiness has been reported by some workers but not confirmed by others. Vascular thromboses, however, are particularly liable to follow surgical procedures. Mental retardation occurs in about two-thirds of these patients who are often highly nervous individuals with symptoms resembling schizophrenia. Localized neurological signs may occur, because of intracranial vascular thromboses. Subluxated lenses are present in about 80 per cent. of patients after the first few years of life and downward displacement of the lens is more common than in the Marfan syndrome. The angles of the anterior chamber are normal. Acute glaucoma, due to an anterior dislocation of the lens, is common and is often the presenting symptom.

In spite of its rarity, homocystinuria should always be borne in mind in patients with subluxated lenses, all of whom should be screened for this condition. The cyanide-nitroprusside test is a simple screening procedure which any laboratory can perform in a few minutes. The diagnosis has to be confirmed by urinary and sometimes blood amino-acid analysis, or by enzymatic studies.

In homocystinuria, therefore, glaucoma is lens-induced and occurs when the lens becomes dislocated, usually anteriorly.

Management

The management of glaucoma in the presence of a displaced lens depends upon the cause of the glaucoma. I do not intend to discuss the management of glaucoma due to anomalies of the angle and will restrict my comments to the management of glaucoma due to the displaced lens. This depends upon the position of the lens.

If an anterior dislocation of the lens results in the development of acute glaucoma, two lines of treatment are possible, either extraction of the lens or its reposition behind the iris. Extraction is frequently accompanied by loss of vitreous, while reposition, which I favour, risks a recurrence of the anterior dislocation as well as the possible dangers resulting from a lens dislocating posteriorly into the vitreous.

Acute pupillary-block glaucoma, resulting from the lens becoming incarcerated in the pupil, may be treated either by a lens extraction or by mydriatics which usually relieve

the condition. Recurrences can usually be prevented by performing a peripheral iridectomy. Because of the surgical complications which frequently follow a lens extraction in these patients, I favour the more conservative course of action.

If glaucoma occurs in the presence of a subluxated lens, the glaucoma is not lens induced. It is usually due to the presence of angle anomalies; occasionally it is caused by vitreo-pupillary block.

Glaucoma in the presence of a posteriorly dislocated lens may have one of several causes and at times considerable clinical acumen is required to determine the cause (Table II). Vitreo-pupillary block is characterized by a shallow anterior chamber, iris *bombé*, and vitreous blocking the pupillary aperture. It is treated by intensive mydriatics in the first instance; if these fail to relieve the block, a peripheral iridectomy may be necessary. Rubeosis secondary to a retinal detachment is usually easy to diagnose on careful examination; treatment is of no avail. Phacolytic glaucoma occurs with an anterior chamber of normal depth, which contains a flare and cells, in the presence of a mature or hypermature cataract. If there is doubt about the diagnosis, a paracentesis of the anterior chamber should reveal the characteristic macrophages filled with eosinophilic-staining material. Treatment is removal of the lens, preferably under direct vision. Angle anomalies can only be diagnosed gonioscopically. Coincident primary glaucoma can be diagnosed only by exclusion. Treatment is that of the primary glaucoma.

Table II *Glaucoma with a posteriorly dislocated lens*

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(1)	Vitreo-pupillary block
(2)	Rubeosis secondary to retinal detachment
(3)	Phacolytic glaucoma
(4)	Angle anomalies
(5)	Coincident primary glaucoma
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The treatment of glaucoma resulting from the lens being dislocated into the vitreous depends, therefore, upon the cause of the glaucoma. Only when a phacolytic glaucoma occurs is extraction of the lens mandatory. The eye appears to tolerate a lens dislocated into the vitreous remarkably well, as long as the capsule remains intact. A breach in the capsule is liable to result in the development of phacolytic glaucoma.

The treatment of a dislocated lens in a patient with homocystinuria merits a few comments. There is no doubt that vascular thromboses are liable to occur in these patients, particularly after surgical procedures, and because of this, surgery should be contemplated only as a last resort. If surgery is inevitable in order to save the eye, certain precautions should be taken. The patient should be mobilized immediately postoperatively and steps should be taken during surgery, such as additional corneo-scleral sutures, for this to be safe. Anticoagulants should not be given, as haemorrhage into the eye is liable to follow their use. Pyridoxine should be prescribed and, if there is time in the preoperative period, a low methionine diet should be instituted. It is my experience that, if these precautions are taken, the risks of surgery are less than suggested in the recent literature.

Summary

The aetiology and management of glaucoma in the presence of a spontaneously dislocated lens are discussed.

I wish to thank my colleagues both at Moorfields and elsewhere for referring cases to the Genetic Clinic.

COMMENTARY

TREATMENT OF THE DISLOCATED LENS

If the lens dislocates anteriorly and can be repositioned behind the pupil, a peripheral iridectomy usually prevents further anterior dislocation of the lens. If the lens is dislocated posteriorly, it should be left alone unless there is some very strong indication for its removal; *e.g.* when the lens starts to leak and produce a phacolytic glaucoma.

In anterior dislocations of the lens, aspiration of the lens is a reasonable technique especially if there is a strong possibility of vitreous loss at the time of operation.

CAUSE OF GLAUCOMA IN DISLOCATED LENSES

In the Marchesani syndrome changes in the angle are explicable on the basis of a pupillary block causing angle closure. Posterior dislocations do not seem to occur in the Marchesani syndrome. There is no evidence that a posteriorly dislocated lens causes glaucoma through stimulation of the ciliary body.

GLAUCOMATOUS CHANGES IN HOMOCYSTINURIA

A great many patients with homocystinuria who develop optic atrophy with cupping resembling glaucomatous optic atrophy in the absence of a raised intraocular pressure, also have increased platelet stickiness. Dr. Spaeth felt that the platelet stickiness changes were universal in homocystinuria and that if they are not demonstrated this is probably the fault of the technique. As there is a direct relationship between the amount of stickiness and the level of the homocysteine it seems possible that the increased platelet stickiness produces an occlusive vascular disease and hence atrophy of the optic nerve head. Homocystinuria is an extremely rare disease even amongst the mentally retarded. Dr. Spaeth surveyed 10,000 individuals who were mentally retarded and only found three cases of homocystinuria. However, as it now appears that some of these patients are likely to be treatable with low methionine diets and/or pyridoxine, it seems worth while locating them early. Very little is yet known of the natural history of the condition. One young girl followed by Dr. Spaeth since 1965 and treated with diet and pyridoxine has so far had no ocular complications. The most satisfactory screening test for homocystinuria is the silver modification of the cyanide-nitroprusside test (Winston and Barber), which practically eliminates most of the false positives found with the original cyanide-nitroprusside test.

Treatment of juvenile glaucoma

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COMMENTARY

Patients with juvenile glaucoma respond extremely badly to the standard surgical procedures, but many have anterior insertions of the iris and angles which look rather like those seen in infantile glaucoma. Some of these patients do well with goniotomy using the Lister modification of the Barkan knife made by Grieshaber. This is a sharp instrument making it much easier to enter the anterior chamber and to dissect over the anterior aspect of the trabecular meshwork. Bleeding which cannot be readily washed out as with infants, is particularly severe in young patients, but has never been so disastrous as to require further surgery. If goniotomy has been unsuccessful Professor Richardson has found that goniopuncture is, occasionally, a worthwhile procedure, though it may not succeed in more than one in ten or one in fifteen.

In young children with the Sturge-Weber syndrome it was necessary to use cyclocryo-therapy as a primary procedure. This could be repeated on several occasions if necessary.

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Dr. Hetherington's manuscript was unfortunately lost in the post.

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