ARTICLE

The Doyne Lecture

Congenital cataract: the history, the nature and the practice

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Introduction

In his 1933 Doyne Lecture A.B. Cridland cited Emmerson: 'An institution is the lengthened shadow of one man.' The Oxford Congress is such an institution, founded on 15 July 1909,[1] with Robert Walter Doyne as the first Master; it holds a special place in the academic calendar of ophthalmologists around the world because of its academic excellence, the beautiful city in which the meeting is held, and the warmth and conviviality of the members. I was particularly pleased and proud to be asked to give the 75th Doyne Lecture in the 80th year of the lectureship because this is the first such lecture to be given by a British paediatric ophthalmologist and it is given at a time when paediatric ophthalmology in general and in the British Isles in particular is flourishing clinically and academically. I chose congenital cataract as my subject not only because it has been a lifelong interest: it is still the major preventable cause of lifelong visual handicap, and there are a number of controversial areas which have not yet been resolved.

History

Although couching for cataract was practised in the Hellenistic period,[2] its use in children was not ideal: an already tricky operation was turned into an exceptionally difficult one by the child's anatomy and the lack of an anaesthetic. Pott,[3] the ophthalmologist to St Bartholomew's hospital, introduced the discission operation which was to remain in use for nearly 200 years. It consisted of making an incision of one or another shape and size in the anterior capsule, leaving the aqueous to absorb the lens material over a few weeks; however, a thick capsular membrane usually resulted, and glaucoma was frequent. Most practitioners of discission used an anterior approach but Saunders[4] advocated a posterior approach, because the dispersion of the lens material into the vitreous was, he considered, more effective. Ziegler[5] in 1921 described a technique of 'through and through' needling, using two bold V-shaped cuts: 'Boldness in incision is a virtue, timidity a vice'. There could not have been enough sufficiently bold surgeons around as the technique never caught on, although Ziegler's knife can still be bought today.

Gibson,[6] from Birmingham, used a technique in which the couching needle was used to rupture the anterior capsule and the eye was left to settle over a period of 2 or 3 weeks. Then an incision, made 'by a corneal knife of the largest size' was used to extract the cataract: 'as the knife is withdrawn, aqueous and some of the cataract evacuate spontaneously or helped by a curette'. Gibson's technique was to become known as linear extraction, and was used world-wide for nearly 150 years. Many authors used a combination of the techniques; linear extraction gave the best vision, but discission, being the simplest, was the safest.

Scheie and Ewing[7] described the history of aspiration of the lens through a small incision. Rhazes, a Persian physician and philosopher, mentioned its use by Antillus, a contemporary of Galen, in the fourth century AD. Aspiration was practised in Japan in the fifteenth century, in Italy in 1829, and by Laugier in 1847, and modifications of the technique came into and just as easily went out of fashion, mostly because of technical difficulties. Teale's[8] cannula was an ingenious device in which a fine cannula that was inserted into the lens via a small incision at the limbus had suction applied to it through a rubber tube running to the surgeon's mouth. Scheie[9] described the technique of lens aspiration that was to be the basis of numerous subsequent methods; the use of the operating microscope in infant cataract surgery,[10] new sutures and instruments took it to new levels.

Although there were some advocates of intracapsular cataract extraction in children,[11,12] complications were too frequent.

As the Industrial Revolution brought about improvements in the quality and availability of surgical instruments, so surgeons became bolder in their techniques. Treacher Collins[13] simply couched a persistent hypoplastic primary vitreous (PHPV) membrane; others, later attempted to remove them. Alexander[14]

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'seized the disc by the iris forceps and extracted it only a drop of vitreous escaped and the result has been entirely satisfactory'. Some of the applications of new technology were most imaginative: Nutt,[15] for instance, preceded radiofrequency capsulorhexis by 35 years, even if his diathermy technique didn't catch on!

Amblyopia in congenital cataract

Juler,[16] in 1921, noted that traumatic cataracts operated before 5 years of age did badly, whereas those operated after 5 years did relatively well. As ophthalmologists became aware of the ground-breaking work of the amblyopia researchers, so they realised how it affected their management of congenital cataract.[17,18] There are a number of clinical observations that reinforce the importance of amblyopia on the visual outcome following cataract surgery:

1. Early treatment brings about better visual results. Once the importance of amblyopia became evident, good visual results were obtained by the early treatment of previously unrewarding cases of unilateral cataract.[19]
2. Early and continued optical correction is essential in infant surgery. It is likely that the poor results of cases operated in infancy in early studies[20,21] were due to the lack of optical correction. Today, cases with early surgery but later optical correction have a worse prognosis than those with early optical correction.[22]
3. Occlusion of the phakic eye in unilateral cataract is as essential as early surgery and optical treatment.[23]
4. Poor compliance with optical treatment and occlusion reduces the quality of the visual results.[24–27]
5. In bilateral cataract, the first operated eye achieves the better vision[28–30] in the absence of pre- and immediately post-operative occlusion, in cases with even only a few days between the operations on the two eyes.
6. Later deprivation does not give rise to additional visual defect.[16,31]
7. Brief early deprivation, before the critical period, does not affect vision.[32,33]
8. In asymmetrical, bilateral cataracts, the more severely affected eye develops the worse vision.[18]

Early referral

As the importance of amblyopia became established,[17,32] so did the necessity for early referral. Many ophthalmologists went through a stage of treating congenital cataracts as a 'semi-emergency'.[34] Although not strictly necessary on the grounds of treating amblyopia, the attitude led to earlier referral through publicity and the teaching of paediatricians about the need to detect cataracts in neonates.[35–37] In most units specialising in congenital cataract the majority of referrals are now infants in the first weeks of life.[38]

Epidemiology

Population-based studies have suggested a prevalence of 1.2 per 10 000 total births,[39] 2.1 per 10 000 live births[40] or 2.3 per 10 000 total births.[41] These figures are almost certainly low as the data are gathered at diagnosis, or from inpatients symptomatic in the first year of life,[40] or from discharge abstracts of live or still-born babies,[39] or from a malformation register.[41]

Screening of 2447 4-year-olds[42] gave 7.7 cataracts per 10 000, and two birth cohort studies suggested a prevalence of 5.3[43] and 4.4 per 10 000.[44,45] Between 40%[43] and 55%[40] of these cases are unilateral. A current British nation-wide active surveillance study with high ascertainment by Dr Jugnoo Rahi at the Institutes of Child Health and Ophthalmology in London suggests that the cumulative incidence in the UK is between 2 and 3 per 10 000 live births per year.

New registrations of patients with congenital cataract, aged 0–15 years, from the National Registers of Partial Sight and Blindness, England and Wales suggest that there may be a decrease in the percentage of registrations due to cataract. This decrease may be due to an improvement in management and to changes in attitudes towards registration.

Natural history

Pathogenesis

The mechanisms of cataractogenesis are not well understood. Insights have been gained from conditions such as galactosaemia where water is osmotically drawn into the lens by the accumulation of galactitol, causing the 'oil-droplet' refractive change in the central part of the lens. Further hydration leads to the swelling and rupture of lens fibres, and the formation of a cataract.[46] Bilateral cataract is almost invariable in untreated 'classical' galactosaemia due to galactose-1-phosphate uridyl transferase deficiency;[47] it occurs in the rare epimerase deficiency,[48] but in galactokinase deficiency the lens is less certainly involved. Galactokinase-deficient persons may be prone to presenile or infantile cataracts[47] but some studies have failed to confirm this.[49,50] Marginal maternal deficiency of galactose enzymes may underlie some congenital cataract.[47,51]

Familial sorbitol dehydrogenase deficiency is also associated with cataracts,[52] which probably have a similar largely osmotic mechanism. Accumulation of mannose-enriched oligosaccharides in the posterior parts of the lens is associated with capsular opacities in mannosidosis.[53]

Hypoglycaemia in infancy, of any cause, may give rise to cataracts,[47,54–56] but the mechanism is not clear. The metabolic defect is usually profound; most affected infants have convulsions and many have permanent brain damage.[57]

Hypocalcaemia also causes infantile cataracts,[58-60] usually of less functional significance than hypoglycaemic cataracts. Calcium plays an important

role in cell signalling[61] related to acetylcholine receptors which liberate calcium from intracellular stores in the lens.

The occurrence of cataracts in the Smith-Lemli-Opitz syndrome,[62,63] in which there are low levels of plasma cholesterol and high 7-dehydrocholesterol, in cerebrotendinous xanthomatosis, which also has high levels of cholestanol in tissues including the lens, and in mevalonic aciduria, together with cataract production by inhibitors of cholesterol in the lenses of experimental animals, suggest an important role for cholesterol in the structure and maintenance of lens cell membrane structure.[64]

Oxidative damage has been implicated in cataractogenesis: oxygen-derived free radicals are highly reactive compounds which can oxidise lipids and proteins can cause cell membrane damage and alterations to cell function[65,66] this damage is accelerated by UV light, ionising radiation, hyperbaric oxygen[67] and high concentrations of transitional metal ions.[68] This damage is normally prevented by cellular antioxidants such as superoxide dismutase, glutathione peroxidase, ascorbate and vitamin E. Thirty-three per cent of young rats brought up on a diet containing low levels of both tryptophan and vitamin E develop cataracts.[69] Patients with Down's syndrome have increased free radical activity,[70] perhaps due to the trisomy giving high levels of CuZn-superoxide dismutase, the gene for which is carried on chromosome 21. Cataracts may be prevented by antioxidants.[61]

Transient cataracts occur in premature infants;[71–74] in most cases the pathogenesis is obscure. Alden *et al.*'s cases[72] had mostly received kanamycin, and had been acidotic, suggesting a metabolic cause. In some, the cataracts have been clearly associated with treatment for retinopathy of prematurity;[75,76] 93% were associated with laser treatment and 3% with cryotherapy.[76]

Ninety per cent of water-soluble lens proteins are crystallins.[77] They are contained in the lens fibres, and have a spatial arrangement and order which is imporant for the clarity of the lens. Three major classes of crystallins – α, β and γ – are distinguished on their chromatographic properties, the α being of the highest molecular weight, the γ the lowest. They are very stable proteins, resisting denaturation by oxidation, UV radiation and racemisation. The γ-crystallins mostly occur in the lens nucleus,[78] the site of the Coppock cataract in which a mutation of the γ-crystallin gene has been found.[79] A cerulean cortical cataract is linked to the β-crystallin region on chromosome 22q; β-crystallins are mostly found in the lens cortex.[78] The gene for the major intrinsic protein (MIP) of the human lens fibre cell membrane has been mapped to chromsome 12q.[80]

Genetic heterogeneity

Genetic heterogeneity has been demonstrated in a number of families using a variety of gene markers.[81–84] There is probably a close association between the morphology and the gene locus;[85] hence a pulverulent cataract involving the cortex as well as the nucleus is not linked to the Duffy blood group[86] as is the Coppock cataract[87] (Table 1).

Morphology

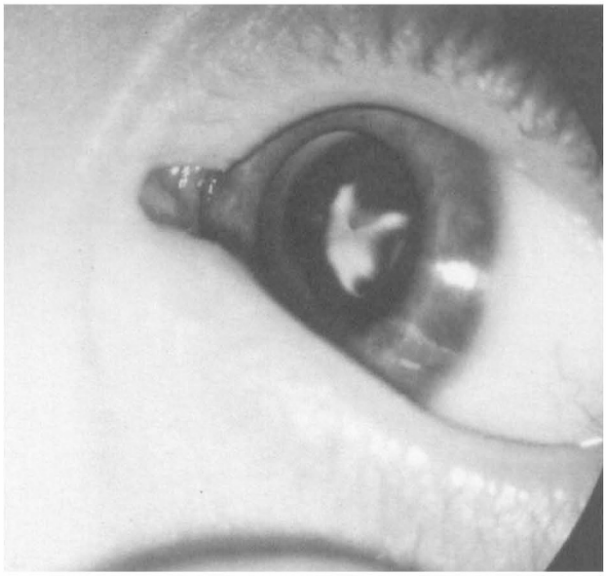
The morphology of cataract is important for several reasons: it can give a clue to the age of onset, to the visual prognosis, it may suggest heritability, and it may give a clue to the aetiology. Some morphological types have a better visual prognosis than others, with lamellar cataracts and posterior lenticonus doing well and dense central cataracts relatively poorly.[103–105] Phenotypic heterogeneity (Fig. 1) is a prominent feature in many families, particularly with autosomal dominant congenital cataract;[82,84,85,106,107] there are inter-ocular (Fig. 2), morphological and intrafamilial differences within a pedigree.

In congenital Morgagnian cataracts, named after Giovanni Morgagni who described them in 1762, the outer zones of the lens become liquefied whilst the nucleus remains intact. This allows the nucleus to fall by gravity in any direction, depending on the position of the head.

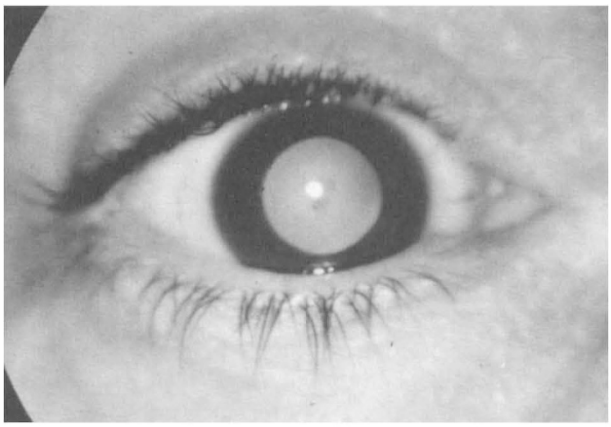
Table 1. *Gene loci linked with cataract types*

Cataract type		Author and reference	Gene locus
Volkman, nuclear	PD	Eiberg *et al.* (1995)[88]	1p36
Posterior polar	AD	Ionides *et al.* (1997)[85]	1p
Zonular pulverulent	AD	Renwick and Lawler (1963)[87]	1q21–25
Coppock-like	AD	Lubsen *et al.* (1987)[89]	2q33–36 (γ-crystallin region)
Polymorphic	AD	Rogaev *et al.* (1996)[90]	13q
Polymorphic	AD	McKay *et al.* (1997)[91]	13q
Nuclear and lamellar	AD	Marner *et al.* (1989)[92]	16q
Anterior polar	AD	Berry *et al.* (1996)[93]	17p
Zonular, sutural	AD	Padma *et al.* (1995)[94]	17q11–12
Cerulean	AD	Armitage *et al.* (1995)[95]	17q24
Cerulean	AD	Kramer *et al.* (1996)[96]	22q (β-crystallin region)
Nance-Horan syndrome	X-LR	Lewis *et al.* (1990)[97]	Xp22.2–22.3
Nance-Horan syndrome	X-LR	Zhu *et al.* (1990)[98]	Xp22.2–22.3

AD, autosomal dominant; XL-R, X-linked recessive.
Chromosomal translocations 2;14,[99] 2;16,[100] 3;4,[101] 3;18[102] have suggested candidate sites.



(a)



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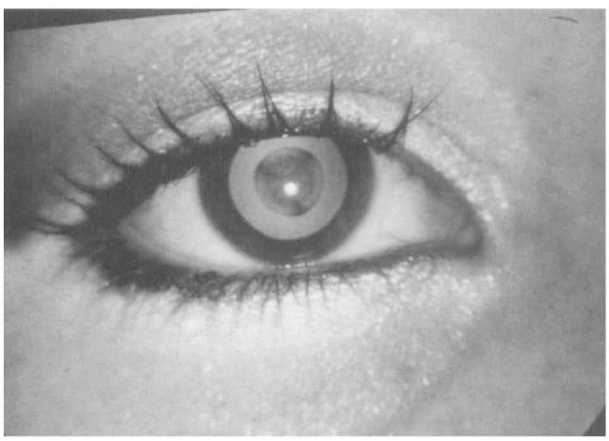


(c)

Fig. 1. *Autosomal dominant congenital cataract presenting in infancy (a). Also shown are the patient's mother (b) and grandmother (c), who are asymptomatic.*

Total cataracts involving the whole or nearly all of the lens occur in Down's syndrome, acute metabolic cataracts and congenital rubella (where 'shaggy' nuclear cataracts are more common), but may be seen in familial or sporadic cases.

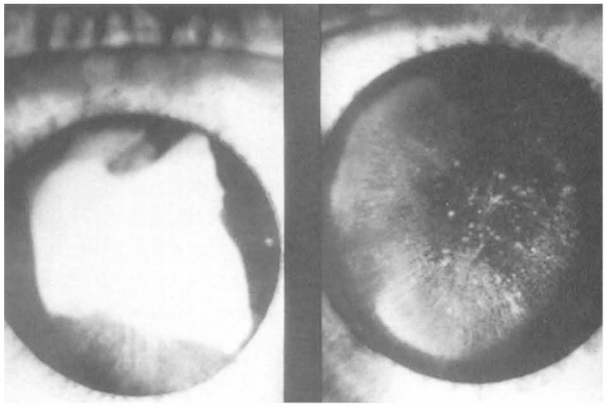


Fig. 2. *Interocular phenotypic variability in a patient with pulverulent cataract.*

Disc-like and membranous cataracts represent varying stages of reabsorption of the lens, leaving either a disc of lens material or a bag of milky or crystalline substance, or the anterior and posterior capsules fused together (membranous cataract). It is common in persistent hypoplastic primary vitreous (PHPV),[108] and has been described in congenital rubella,[109–111] in the Hallermann–Strieff syndrome,[112,113] aniridia,[114] Lowe's syndrome[115,116] and in a patient with the Pierre Robin sequence,[117] and it may occur after rupture of an anterior lenticonus.[118] When the reabsorption occurs in a localised area, a hollow doughnut-shaped[119,120] or a sector-shaped membranous[121] cataract may result.

Dot-like anterior polar cataracts were described early in the twentieth century.[13] These are tiny white dots on the anterior surface of the lens in the axial area that probably represent abnormalities of lens vesicle detachment. They vary in size from one case to another,

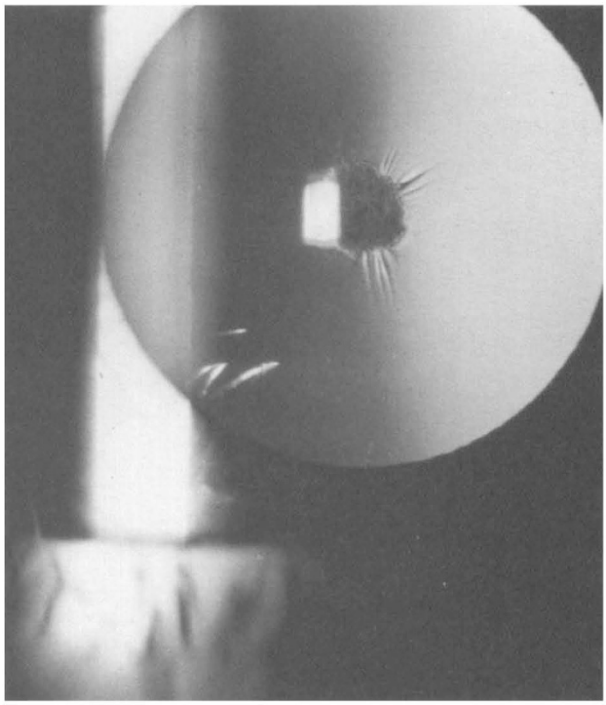


Fig. 3. *Anterior polar cataract showing capsular wrinkles that are probably associated with the astigmatism that occurs in some cases.*

and although they are often not in themselves visually significant, they may be associated with refractive errors[122] (Fig. 3), amblyopia and strabismus. Usually static, they may occasionally progress[123,124] and become visually significant. Some cases are inherited as an autosomal dominant trait, and they may be associated with cornea guttata.[125,126]

Anterior pyramidal cataracts probably also represent anomalies of lens vesicle detachment; they are larger axial opacities than the dot-like anterior polar cataracts, may even extend anteriorly and rarely fuse with the cornea. They are fibrous, and more likely to be visually significant and to progress than anterior polar cataracts. They may become detached[127] and form an anterior chamber foreign body.[128]

Plaque-like anterior polar opacities are axial or eccentric opaque plaques associated with a persistent pupillary membrane (Fig. 4). Presumably they are caused by abnormalities of pupillary membrane regression.

Anterior subcapsular cataracts are usually associated with acquired disease such as uveitis, trauma, irradiation or atopic skin disease, but they may be part of a more widespread cataract. They may, rarely, be associated with anterior lenticonus such as that seen in Alport's syndrome.

Anterior lenticonus is most frequently encountered in association with Alport's syndrome of nephritic haematuria and deafness. The lenticonus may be a manifestation of a basement membrane disorder;[129] it can be congenital and is found in about 10% of affected young children,[130] but may increase in frequency with time up to 30%.[131]

Mittendorf's dot represents the remains of the anterior end of the hyaloid artery. It appears as a small axial or paraxial grey-white dot opacity at the posterior apex of the lens, often associated with the thread-like remains of the hyaloid artery. It is visually insignificant unless it is large, when it may then represent a form of PHPV.

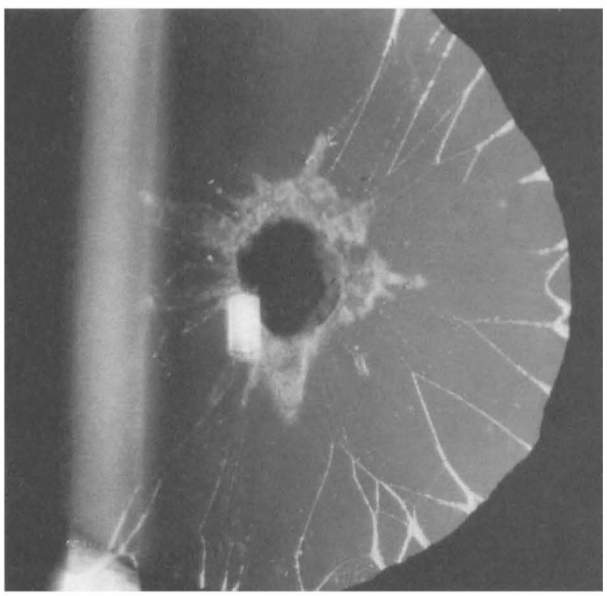


Fig. 4. *Plaque-like anterior polar cataract showing its association with remnants of the pupillary membrane.*

Posterior lenticonus or lentiglobus is a unilateral or bilateral and asymmetrical, thinning and posterior bowing of the posterior lens capsule which has variable effects on the adjacent lens cortex: sometimes it manifests as a high degree of astigmatism, often irregular, without cataract or as a progressive, localised opacity in the abnormal area. It may be present at birth[132] or progress in the first months[133] of life. It is not usually associated with any systemic disease. Amblyopia is frequently present;[134] however, vision may be improved by post-operative occlusion,[135] and because there is a possibility of the visual defect being acquired, surgery may still be indicated even in cases where one might normally expect a poor visual result.[136] The visual results are generally better than with many other causes of infantile or congenital cataract.[104] There is a localised thinning of the posterior capsule.[137–141] Sometimes the junction between the lenticonic and the adjacent capsule is sharply defined and may be seen as a 'ring of fire' on slit-lamp examination.[142] Presumably, the cause of the cataract is largely mechanical; as the capsule bows posteriorly there is progressive distortion of the lens fibres themselves, and cataract formation which can be rapid. Sometimes there may be a hyaloid remnant attached to the lenticonic area:[137,143,144] the pathogenesis of these cases (Fig. 5) may be different from that of the majority without a hyaloid remnant. Although sporadic cases exist, many are inherited as an X-linked[145,146] or autosomal dominant trait.[144–147]

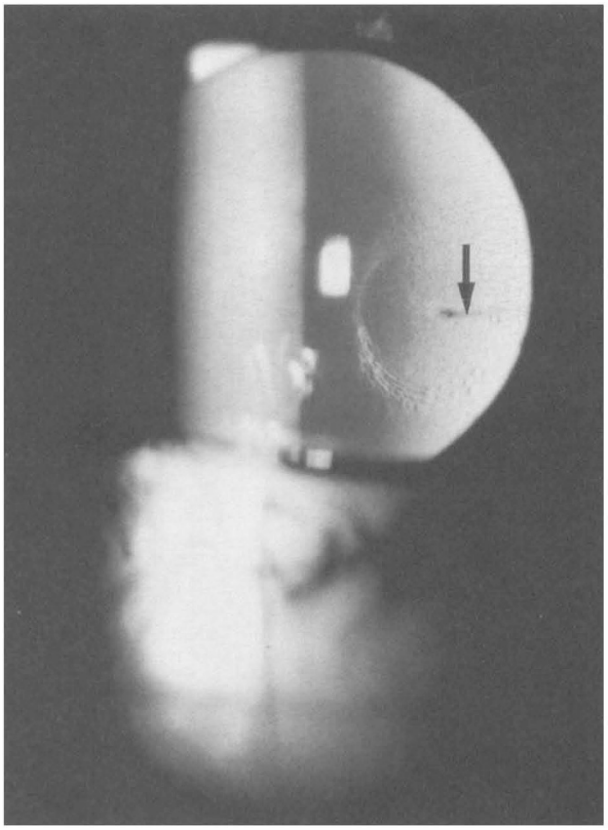


Fig. 5. *Posterior lenticonus with attached hyaloid artery (arrow).*

Posterior lens reduplication is a rare congenital anomaly in which there is a small, distinctly separate, additional lens, usually cataractous, on the posterior pole of the normal lens (Fig. 6); it probably results from an abnormality of lens placode invagination, and needs to be distinguished from posterior lenticonus.

Lamellar or zonular cataracts are common forms[148] involving one or more layers or zones of the lens. They are often inherited as an autosomal dominant trait. Typically, they are bilateral but slightly asymmetrical, and are composed of a layer of minute white dots in a single or more layers of the lens not involving the embryonic nucleus, but sometimes involving the fetal nucleus, and with clear cortex outside them. They are often incomplete, and they may have projections from their outer edges known as 'riders'. The visual prognosis, especially in partial cataract, is better than in many other morphological types;[104,149] many cases can be managed conservatively.[103,150] There is often a marked interocular and intrafamilial variability.[107] They presumably are related to a transient disturbance of lens metabolism; although most usually no underlying cause is found, they may occur in cases of known transient metabolic disturbance, such as galactosaemia.[151]

Nuclear cataracts are opacities of the embryonic or fetal nucleus, similar to lamellar cataract, and often not highly visually significant. Vogt's anterior axial embryonic cataract is a visually insignificant group of opacities lying near, but posterior to, the anterior upright Y. Vogt[152] thought that it may be remnants of primary lens fibres; Mann[137] felt it was more closely related to sutural development. Crystalline nuclear cataracts have been described in association with an abnormality of the hair.[153]

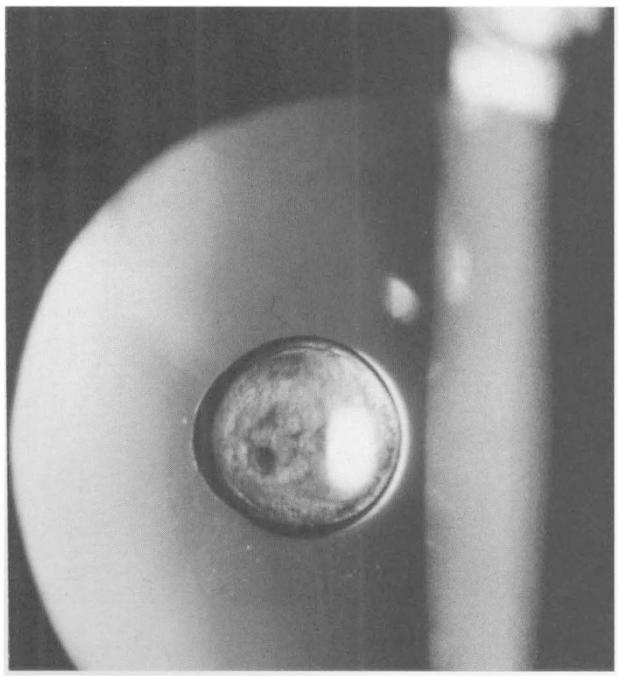


Fig. 6. *Posterior lens reduplication showing a clearly defined cataractous 'blister' on the posterior surface of, but demarcated from, the lens. It remained static for 4 years.*

Central pulverulent cataracts are composed of myriads of ('pulverised') tiny dots. They may be familial as in the autosomal dominant 'Whalsay' cataract,[154] which affected descendants of a Dane, born in 1745, who immigrated to the Shetland Isles.

Nettleship and Ogilvie described the central pulverulent cataracts of the Coppock family in 1906:[155] 'Our attention was originally drawn to the family by Mr Doyne who saw and described the first case, William Coppock, senior in 1888 at the Oxford Eye Hospital . . . The cataract is always very partial and circumscribed, sometimes being so slight as to require careful seeking . . . a sharply defined circular disc placed deep in the lens between the nucleus and the posterior pole'. They distinguished it very carefully from lamellar cataract. Adams,[156] also at the Oxford Eye Hospital, described a patient who was 'one of the original family': the cataract was not discoid, but central; this was confirmed by Rosen's[157] slit-lamp photographs. There is frequently considerable intrafamilial variation[85] (Fig. 7).

Oil-drop cataracts are classically seen in infants with galactosaemia. There is a central area of different refraction to the surrounding lens that looks like an oil droplet floating on water. If treated late or if the diet is not strictly adhered to, a lamellar opacity may develop. Other forms of cataract occur in galactosaemia including posterior subcapsular, small nuclear and cortical.[158] They may regress if dietary control is good.[158]

Cortical cataract is unusual in childhood. A few families have been described with an autosomal dominant inheritance.[85] The onset of the cataract may be post-natal – at least the visual defect may become significant after infancy. The nucleus is not involved.

Blue-dot, cerulean, coronary or punctate cataracts have a beautiful sky-blue or sea-green (cerulean) hue on slit-lamp microscopy;[159] other colours may be seen, including red, brown and opalescent white. They are bilateral, largely stationary, and visually significant. They are often concentrated in the equatorial region of the lens, and they have a variably sized dot-like shape;

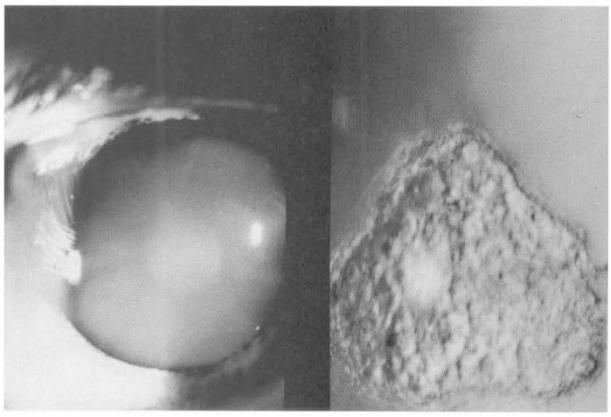


Fig. 7. *Intrafamilial phenotypic variation in central pulverlent cataract. On the left is the cataract of the asymptomatic patient who was detected by screening; on the right is the posterior cortical cataract of her mother.*

sometimes they are elongated. Coronary cataracts are elongated or club-shaped cerulean opacities that form a ring around the equator.

Punctate lens opacities occur in 13–20% of patients with Down's syndrome.[160,161] The opacities occur in the peripheral cortex, and at the anterior and posterior poles,[162] and they increase with age. They were thought to arise from excrescences of the capsule[163,164] but a subsequent study has failed to corroborate this.[165] In carriers of Lowe's syndrome, punctate grey-white opacities occur in all layers of the cortex, but not in the nucleus;[166–169] they increase in number with age, and since these opacities may be found in the normal population, their numbers must be compared with those in age-matched controls.

Sutural opacities around or involving the sutures are very common, and not usually visually significant. They are often familial and range from an increased density of the sutures to a variety of whitish or cerulean dots clustered around either or both sutures. They may progress, and form nuclear or central cataracts.[170] Sometimes, sutural cataracts are the only manifestation of involvement in asymptomatic relatives. They may be inherited as an autosomal dominant or X-linked recessive[171] trait. They are rarely found in association with the sutures more peripheral to the Y sutures.

Coralliform cataracts are usually static, central, complex cataracts that cut across normal anatomical boundaries, and are composed of multiple round or fusiform white or cerulean opacities. They are arranged in a coral-like,[172] fusiform, or spindle-shaped fashion and they may be mainly axial. The anatomical arrangement suggests that they may be due to a primary abnormal arrangement of lens fibres. They are often visually insignificant, and can be inherited as an autosomal dominant trait.[173,174]

Wedge-shaped or sector cataracts occupy a sector of the lens, or if they are larger they are known as semilunar. They have been described in Conradi's syndrome,[175] when they may represent an example of Lyonisation, and Stickler[176–178] syndrome.

PHPV must be considered in any discussion of congenital, especially unilateral, cataract management. In 1840 Mackenzie[179] noted 'In some congenital cases we find the pupil widely dilated and round the cataract the appearances of a black zone, the lens and capsule being so small in consequence of impeded growth as to drag into view the ciliary processes'. The surgery consisted of couching or needling the membrane after lens discission:[13] the visual results were not reported in detail! PHPV was characterised by Reese[180] and more recently by Goldberg[180a] as a variable developmental abnormality of the primary vitreous and hyaloid vascular system. The eye is usually microphthalmic, there is a membrane of very variable extent and thickness behind, and usually inseparable from, the lens that is attached via the apices of its scalloped margins to a variable number of ciliary processes. The membrane itself is relatively avascular, but there are usually vessels that pass to the ciliary processes and the iris, and the hyaloid vascular system is present to a variable degree, but only occasionally is there significant flow: it is probably leakage from this vessel that causes intralenticular haemorrhage.[181–183]

The lens itself may be of a normal size, which gives rise to a shallow anterior chamber as the retrolental membrane shrinks and thrusts it forward; this occurs usually in the first months of life and may be progressive, giving rise to glaucoma. The lens may spontaneously reabsorb,[108] but the eye is still at risk, if the membrane is thick, of dislocation of the ciliary body and hypotony. In one histopathological series[184] 58% were unilateral without associated ocular anomalies, 31% unilateral, with ocular anomalies, and 11% were bilateral with ocular or systemic abnormalities. Systemic disease is so rare in unilateral PHPV as to make routine investigation unrewarding.

The indications for surgery are threefold: first, to prevent the complications of glaucoma and hypotony, second for cosmesis, and third for vision. It is known that, if treated early, the visual prognosis may be sufficiently good[136,185] to warrant the arduous optical correction and occlusion regime that is the same as that for unilateral congenital cataract; the visual results are probably better in the milder cases.

The untreated state

What happens to untreated congenital cataracts? The shape of central cataracts is altered by the growing cortex[186] in that they are compacted at a decreasing rate with age, the eye remaining emmetropic despite changes in the parameters that determine refraction.[187] Many cataracts are sufficiently mild and little changed by time that they can be managed conservatively.[103,105,150,188] Often, in lamellar cataracts, the child is able to go through a normal education, perhaps with a little extra help, and it is the desire to obtain a driving licence that brings about the surgery.

Some cataracts progress rapidly; for instance, untreated galactosaemic oil-drop opacities become lamellar-like cataracts and, if treated, some of these cataracts may disappear,[158,189–192] or at least fail to progress.[193–195] Monitoring of galactosaemia dietary control by observation of the cataracts has been suggested.[158] Many more progress very slowly.

In a small proportion of untreated cataracts spontaneous reabsorption takes place.[4,117,179,196] due to leakage of lens material, degeneration, liquefaction and absorption leaving behind the capsular leaves with a variable amount of residual lens matter.

Bilateral cataract

Aetiology

There have been numerous excellent reviews of aetiology[60,66,148,197–200] and the differential diagnosis,[59,201] but since the incidence varies so much with the location of the investigator there can be few implications that can be gained from any one study, at

any one time. Rubella, for instance, has nearly disappeared in developed countries but is still the main cause of cataract in others. The reader is referred to the papers cited above for a fuller discussion.

Investigation and family history

Ideally, the investigation of the child with bilateral congenital cataracts is undertaken by the ophthalmologist and a paediatrician, and sometimes a geneticist, a dysmorphologist, a counsellor or vision development team organiser and a developmental paediatrician. The education authorities may be involved early. The first role of the ophthalmologist is to see whether the problem is a purely ocular one; posterior lenticonus, unilateral PHPV and most familial cataracts rarely require the involvement of a paediatrician. The parents and any siblings should be examined, preferably dilated and with a slit-lamp, even in the absence of a positive family history.

The paediatrician will need to assess the overall development of the child, and look for the presence of any dysmorphic signs, or signs of metabolic or other disease. There is no need to carry out a battery of biochemical or other tests in every case; further investigations should be directed appropriately by the paediatrician There are, however, some investigations that are useful as a routine such as urine amino acids, reducing substances and organic acids, and blood electrolytes, amino acids and fasting lipids. Some cases may require bone X-rays (Conradi's syndrome), chromosome analysis (dysmorphic children), galactose enzymes, virus antibodies or cultures of the lens aspirate.

Visual assessment and the indications for treatment

When the visual defect in a child with congenital cataract is severe enough to interfere significantly with visual development then surgery and post-operative optical treatment, together with occlusion therapy, if indicated, are necessary. However, if the vision of the infant is good enough, active management is best postponed until the child is older, as it may be possible at the later age to carry out a more satisfactory optical correction as the change in power of an eye becomes less. To be sure of the best course for each individual, careful, and sometimes repeated, visual assessment is mandatory.[202,203]

There are many congenital cases where the state of the vision is obvious; if the cataract is dense and large, or total, then there is no doubt that surgical treatment is necessary. In partial cataract, the assessment is much more difficult, and clinical observation is still the most important and reliable method of determining whether treatment is in the best interests of the child. Although qualitative, it is reliable and can even be used to monitor occlusion post-operatively.[204]

The fixation pattern of each eye – whether it is central, steady and maintained, whether there is nystagmus and whether the child has fixation and refixation reflexes appropriate for his or her age – are all observations that are used as the basis for the decision whether or not to operate. An infant with a total cataract, especially if it is acquired, may fix a light normally, despite being effectively blind to solid objects. It is most important not to be influenced unduly by the density or extent of the cataract on ophthalmoscopy: it is very misleading and can either under- or over-estimate vision.

There can be no hard and fast rules as to whether surgery is indicated, and the principle should be whether or not there is sufficient visual defect to significantly interfere with a reasonably normal visual development. If there is any doubt it is always better to reassess the situation after a short period.

There are two main ways of measuring acuity in preverbal children. These are particularly useful in carrying out serial measurements in the management of the post-operative occlusion, but may also be used in pre-operative assessment. Visually Evoked Cortical Potentials (VEPs) to transient patterned stimuli,[19,205–207] or 'steady-state' or 'swept' VEPs[207,208] have been successfully used to measure pre- and post-operative acuities; although they suffer the disadvantages of cost and requiring very substantial expertise, they are the only way of measuring vision without the need to observe eye movements, and they may be more suitable for less co-operative and less attentive infants. Flash VEPs may be useful in complete cataract to establish the gross integrity of the neural visual pathways. Forced Choice Preferential Looking (FPL) is now commonly used in clinics managing patients with congenital cataract.[209-212] The method requires meticulous technique if it is to be reliable, but with appropriate training, it can be performed by a wide variety of personnel. It is relatively inexpensive.

Conservative management

Because very many individuals with congenital cataract have a good prognosis untreated, it is vital that they should not have surgery without due caution and assessment. The use of mydriatics may be helpful in some cases, especially during re-evaluation, but in the long-term the effects on accommodation and glare are significant limiting factors.[105] Optical iridectomy was popular previously[103,213–215] because it avoided aphakia and its correction, usually by spectacles; it was most useful in partial cataracts.[213] As methods of optical correction became better, the relative disadvantages of iridectomy therefore became greater, and it became less popular also because the visual results were less than satisfactory,[105,216,217] probably because paraxial light rays through the peripheral parts of the lens used after the iridectomy are optically inferior to axial ones.[218]

Surgery

Bilateral synchronous or asynchronous surgery?

In some centres bilateral, synchronous cataract surgery has been routine,[219] and Guo *et al.*[220] felt that bilateral

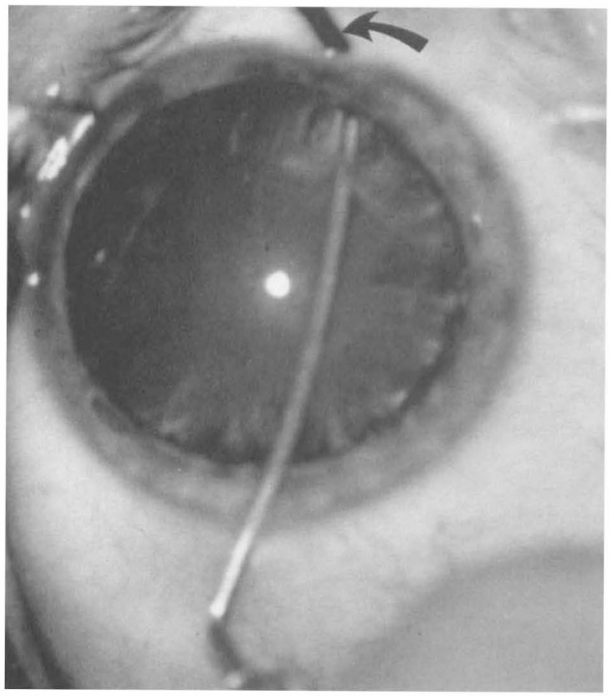
synchronous cataract surgery had the benefits of reducing the anaesthetic risks, reducing hospital in-patient time and costs and allowing earlier optical correction, reducing interocular differences. It is generally felt, however, that the risk of bilateral synchronous endophthalmitis is much greater than the risks of anaesthesia,[221–223] and the cost savings and optical benefits are not important if there are just a few days between operations. Bilateral, synchronous surgery should therefore be reserved for those rare cases that are a significant anaesthetic risk even in a sophisticated paediatric service.[224]

Operation sequence in bilateral cataract

Bouzas[29] noted that the visual results in one eye in bilateral cataract were worse if the other eye was operated much earlier. Even a short interval between the operations on the two eyes[28,225] may result in significant differences in the final acuities. This difference can be mitigated by synchronous surgery[219,220] or, more safely, by occluding both eyes until the second eye is operated.

Lens aspiration

Lens aspiration became used extensively after it was popularised by Scheie in 1960.[9,226] Scheie's technique was to use a 19 gauge thin-walled needle with a rounded point;



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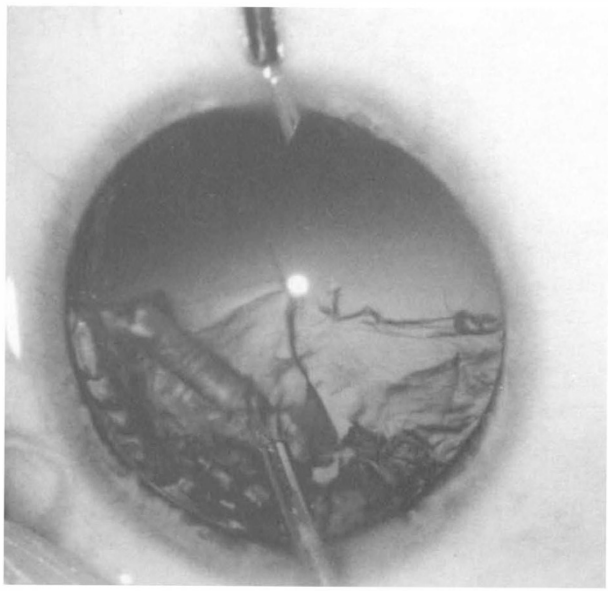

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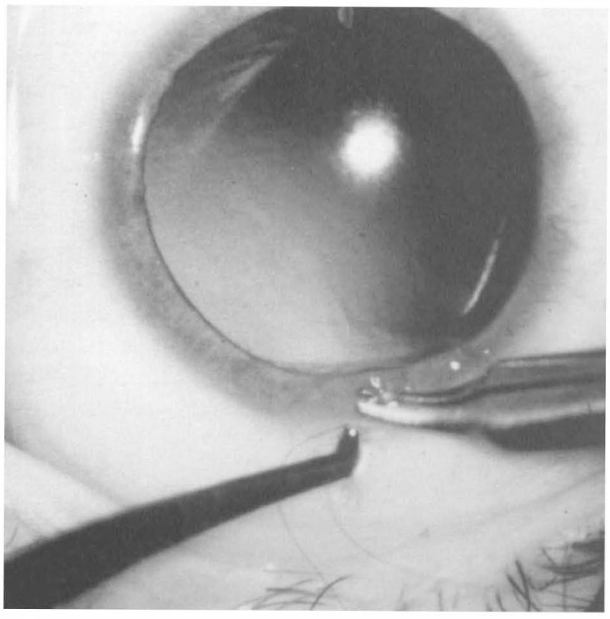


(c)

Fig. 8. *Simples lens aspiration. (a) A wide 'can-opener' anterior capsulotomy is made to remove as much of the anterior capsule as possible. The arrow indicates a small self-retaining infusion cannula. (b) The lens cortex and nucleus are aspirated through a blunt-ended cannula. (c) A clear posterior capsule is left; a primary posterior capsulotomy or capsulorhexis can be performed at this stage. Even if clear, the posterior capsule will opacify in nearly all infants.*

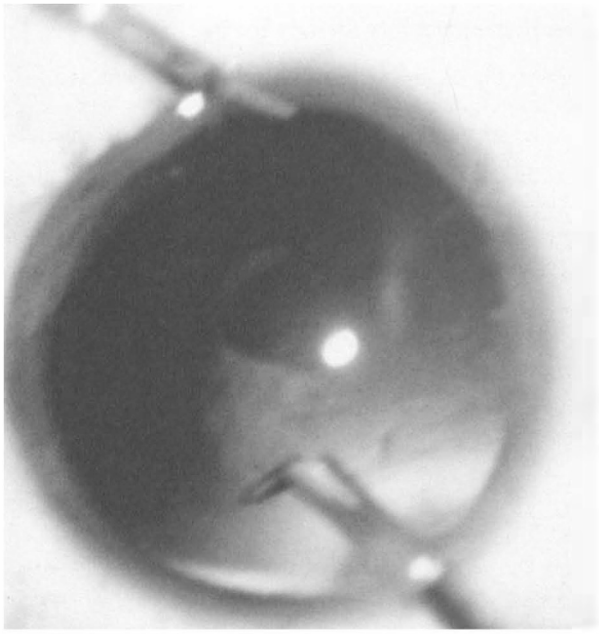


Fig. 9. *A posterior capsule plaque is peeled off the posterior capsule to leave a clear axis.*

he just aspirated, replacing the aqueous if necessary. There have been many improvements in this technique,[10,216,227] particularly the addition of one or other form of irrigating cannula to maintain the anterior chamber (Fig. 8), and the routine use of the operating microscope.

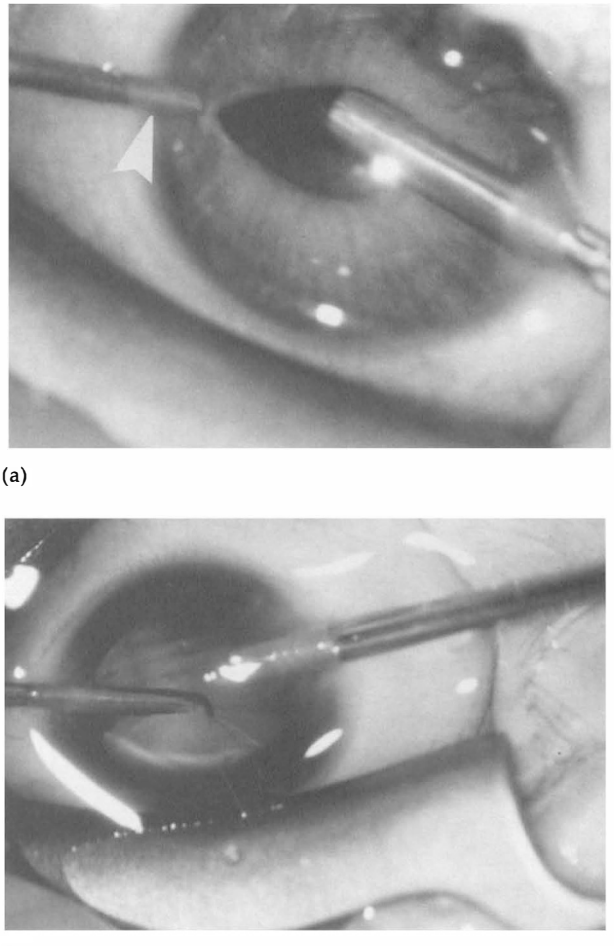
The main problem with aspiration is that the posterior capsule is left behind, and it becomes opaque, even if it is meticulously cleared of any membranes and cleaned (Fig. 9), leading to amblyopia and necessitating repeated surgery.[228] Capsulotomy can be done at the time of the primary surgery, either using a knife-needle[10]or with a vitrectomy machine.[229,230] In small infants there is a risk of posterior synechiae formation and iris bombé. This may cause glaucoma, and a peripheral iridectomy is recommended by some[231] to prevent this; most recommend that in infants of less than 18 months old, a lensectomy, without irridectomy, is performed.[232]

The most frequent indication for lens aspiration today is in an older infant in whom the surgeon may not wish to use an intraocular lens due to the potential growth of the eye, or a likely lack of co-operation with post-operative follow-up. Below the age of 1 year to 18 months, lensectomy is probably the preferred technique.[228,232] Long-term follow-up suggests that the complication rate of simple lens aspiration is low.[10,233]

Lensectomy

As vitrectomy machines came into common use in the late 1970s they were used to get over the main problems of lens aspiration: capsule opacification and vitreous strands through primary capsulotomies. The former was a significant cause of amblyopia in infants, in whom the opacification was most rapid.

An anterior approach (trans-corneal)[228,229,234–237] was used because it was felt that an adequate exposure could be obtained in all cases by modifications of the surgical



(a)

(b)

Fig. 10. *Bi-manual lensectomy. (a) The irrigating cystitome is being used to retract the iris in order to expose the parts of the lens covered by the iris in a case where the pupil has failed to dilate. (b) The irrigating cystitome is being used to deliver lens material to the vitrectomy machine.*

technique (Fig. 10). Earlier posterior approach (trans-scleral) techniques were based on adult anatomy with the entry being made 4 mm behind the limbus.[238] It soon became clear that this gave rise to damage to the retina and vitreous base in infants, so empirically[239–243] or based on anatomical studies[244] a pars plicata approach was adopted, with the entry 1.5–2.5 mm behind the limbus.

The advantage of lensectomy was that the visual axis was cleared permanently and re-operation for an occluded axis was rare,[228] although secondary cataract was recorded even after capsulectomy and anterior vitrectomy.[245]

Complications of surgery

Operative and immediately post-operative complications

Endophthalmitis following intraocular surgery in children is not common, but a survey of over 500 paediatric ophthalmologists[223] suggested an incidence of 7 per 10 000 cases. The infection was diagnosed by the third post-operative day in 82% of cases, and an organism was found in 65% of cases. Nasolacrimal duct

obstruction and upper respiratory tract infections are significant predisposing factors[222] and the visual prognosis is poor, 65% having no light perception.

Taking inadvertent small 'bites' at the pupil margin is quite common but it is probably not very important visually; larger ones may be. Posterior synechiae occur, especially after infant lens aspiration, in PHPV, and in intraocular lens implantation.[246,247] Many pupils are small in congenital cataracts, and the necessary surgical manipulation or stretching by iris hooks may damage the sphincter muscle.

Vitreous strands or 'wicks' are not unusual, even with careful surgical technique, because they may be difficult to detect and if the baby cries in the early post-operative period the chances of vitreous prolapse increase.

A knuckle or iris often becomes attached to the incision site, giving a distortion of the pupil; this is due to wound leakage in the immediate post-operative period, enhanced by crying. Larger iris prolapses can only occur with adult surgical-type incisions.

Transient flame-shaped retinal haemorrhages are not infrequent after lensectomy;[248,249] they are probably due to hypotony and vitreous traction.

Hyphaema is not uncommon in minor degrees but it is sufficiently serious to figure in some series that discuss surgical complications.[20] It may be a prominent part of the process that leads to phthisis bulbi.[250] Intralenticular haemorrhage suggests that PHPV is the underlying diagnosis.

Endothelial cell loss of up to 10%[251–254] may occur, irrespective of the method of surgery.[252] Persistent corneal oedema was much more frequently seen when rubella was more common.[255] Endothelial cell loss may be severe with the use of some types of intraocular lenses.[256]

Vitreous loss of lens material (Fig. 11) needs to be detected by fundoscopy at the end of the procedure, and treated immediately by vitrectomy.

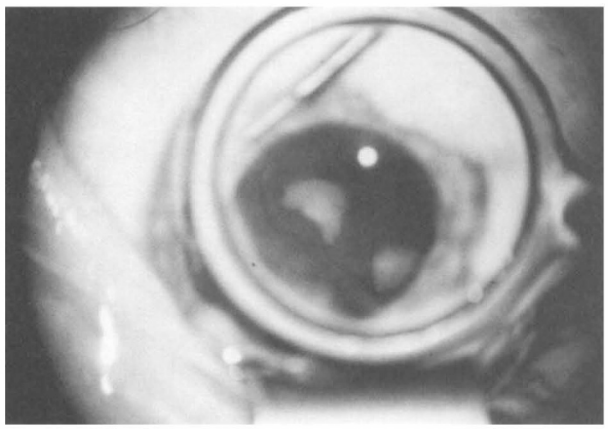


Fig. 11. *Vitreous loss of lens material. Two white, fluffy lumps can be seen adjacent to the inferior temporal vascular arcade. Removal by vitrectomy and aspiration should be carried out to avoid inflammatory consequences.*

Later post-operative complications

Where the posterior capsule is left intact in children, the visual axis will become obscured in the majority,[213,228,257,258] with potentially serious effects on visual development.

There are two main types of posterior capsule opacification (PCO). In the first there is a rapid fibrosis from metaplasia of the epithelial cells that line the lens capsule.[259–262] The second form of opacification[260] is due to more gradual proliferation of equatorial lens epithelial cells that form the commonly found 'pearls' that may completely cover the posterior capsule. A Soemmerring's ring is formed when the proliferating lens epithelial cells are enclosed by both the anterior and posterior capsules.[260] Clinical observation suggests that PCO is more rapid and severe if the eye is inflamed post-operatively, but human lens epithelial cell proliferation occurs in a protein-free medium as well as in one with added protein,[263] suggested that inflammation is not essential for PCO.

PCO represents one of the major problems in congenital cataract management;[264] it can be avoided by surgical technique, or treated. A lensectomy usually, but not always, avoids the need for surgery for secondary cataract, but leaving a substantial proportion of the membrane at the end of the operation improves the possibility of using a secondary intraocular lens at a later date, and disturbs the posterior segment less. Leaving the capsule intact, but performing a capsulectomy at the end of the procedure, with or without a vitrectomy, is one choice.[228–230, 235, 265–267] Posterior capsulorhexis (Fig. 12) has become more widely used, greatly aided by viscoelastic substances. It may be combined with a small vitrectomy and the lens optic may be slipped behind the posterior capsulorhexis[258,268] as a way of reducing opacification.

The alternative to prevention, and the management of those in whom prevention has been unsuccessful, is to perform a secondary capsulotomy by needling, YAG laser[269,270] or by an anterior or posterior[238] approach using a vitrectomy machine.

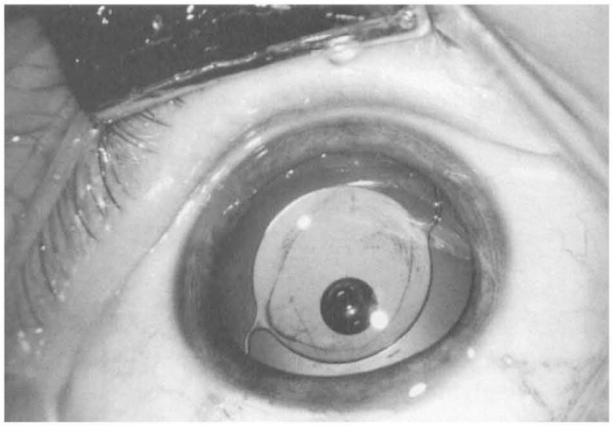


Fig. 12. *An intraocular lens 'in-the-bag' with an anterior and (rather large) posterior capsulorhexis.*

Table 2. *The incidence of glaucoma as a complication of congenital cataract*

Series	Operation	Incidence of glaucoma
Hammami (1972)[215]	Linear extraction, discission, or intracapsular	6.5%
Rice (1977)[10]	Lens aspiration	0.9%
Chrousos (1984)[257]	Lens aspiration	6.1%
Keech (1989)[267]	Lens aspiration or lensectomy	11%
Simon *et al.* (1991)[277]	Lensectomy	24%
Robb and Peterson (1992)[278]	Various	27%+
Schrader *et al.* (1994)[279]	Pars plana lensectomy	4.1%
Biglan *et al.* (1997)[280]	Cataract + secondary IOL	7.1%

Early papers on retinal detachment following childhood aphakia were influenced by the very poor prognosis in nearly every case and it is still difficult to get a reliable insight into the incidence due to the long latent period and the poor long-term follow-up in all series. The incidence is between 1% and 10%.[215,267,271,272] The latent period (the interval between the cataract surgery and the detachment) is long at, around 30 years,[273–275] and although basal vitreous gel incarceration[276] was a concern after lensectomy, it has not proved to be a significant problem yet. Although anatomical success is good, the visual results are worse than in many other types of detachment.

Glaucoma is the most frequent serious complication encountered in the management of congenital cataract, but there is no very accurate estimate of the incidence. The wide variation in the results (Table 2) suggests a low rate of detection in some series, variations in incidence with different surgical techniques and with different timing of surgery.

Children with aphakic glaucoma present, if they have not been detected by screening, because of the appearance of corneal clouding, pain and redness, or because of the appearances associated with shallowing of the anterior chamber in pupil block. The eye may enlarge and become buphthalmic, but often this enlargement is subtle and is only shown as an excessive loss of hypermetropia;[281] this loss of hypermetropia is normal in aphakic children, so it is important to monitor both corneal diameter, the state of the optic disc and intraocular pressure.

There are various ways in which glaucoma can occur: pupil block and iris bombé[231,282,283] occur from pupil occlusion by vitreous, secondary membranes or synechiae. Glaucoma is rare after modern intraocular lens implantation;[265,284–286] it may be, however, that this is due to selection bias: very few young infants and very few abnormal eyes are implanted, and, it is these eyes that are particularly prone to glaucoma. Pseudophakic malignant glaucoma[287] has been described. Rapid lens swelling following discission[288,289] was one of the main reasons why this procedure was abandoned and Treacher Collins[137] knew that needling of Morgagnian cataract led to glaucoma; this complication cannot occur after an adequate removal of lens material by aspiration or lensectomy. There may be a familial predisposition or an anterior segment anomaly.[290,291] Chronic open angle glaucoma is probably the most common form, comprising 79.7% of Asrani and Wilensky's series of 64 affected eyes.[292] The cause is not certain, although it may be more frequent in microcornea,[104,293,294] with small pupils,[293] and with surgery before 1 year of age.[267,278,293] The cause may be an accumulation of lens material in the angle.[295] It is unlikely that the use of steroids is a significant factor in many cases because they are used for only a short time. Although the glaucoma may be associated with pre-operative factors,[267,290] it seems likely that the surgery itself plays a major role; it occurred in only the operated eye of a patient with bilateral cataract.[277] Vitreous may cause glaucoma by pupil block,[282,288] perhaps aided by premature posterior vitreous detachment, explaining some cases that occur months or years after the initial surgery. The presence of large amounts of vitreous in the anterior chamber, spreading over the iris, may cause aqueous outflow defects. Glaucoma may be less common in patients who have had a lensectomy/vitrectomy.[292] Lens-induced uveitis and glaucoma are rare.[296,297]

All aphakic children should be considered as being at risk for life. In the first years examinations under anaesthetic may be indicated; many patients can be followed by non-contact tonometry, observation of the optic disc, corneal diameter measurement, and ultrasound and refraction to monitor eye growth, before full glaucoma monitoring is instituted.

Although one paper[298] tentatively suggested that cystoid macular oedema may be an important cause of visual morbidity after congenital cataract surgery, especially lensectomy, the incidence is now thought to be considerably less than 10%.[243,299–302]

Post-operative inflammation, leading to a high incidence of phthisis bulbi, occurred in the congenital rubella syndrome;[226,303] the high incidence was reduced once it was realised that the virus remained in the lens for many years post-operatively[304–306] and subsequent surgery included more complete aspiration and steroid cover.[307–309] Chronic post-operative inflammation in relatively mild forms is not uncommon, Jameson *et al.*[310] found it was less common in children up to 1 year, but personal experience suggests that it may be more common in younger patients or at least that inflammatory complications are less frequent in older children.

In infants, even with careful technique, non-absorbable sutures become loose very frequently and need to be removed to prevent their becoming a focus for infection. They can be removed routinely from the first

operated eye when the second eye is operated, or if there is another anaesthetic for any reason, but most surgeons now use absorbable sutures routinely.

Optical correction

Introduction: the changing visual environment

It has been known for 20 years that, experimentally, immature eyes can be made myopic by manipulating their visual environment[311–317] and that humans may have the same effects from disease.[318–320] Gordon and Donzis[187] established that there is normally a substantial decrease in the power of the eye in the first years of life, particularly the first 2 or 3 years. It was not surprising, therefore, that Morris *et al.*[321] (Fig. 13) found that there was a steady and significant decrease in the power of the contact lenses used to fully optically correct their infant patients' aphakic eyes as they grew. Other studies confirmed this myopic shift.[316,322,323] It is almost universal and usually greater in unilateral cases;[316,324] this effect also occurs in pseudophakic eyes[325–327] although in humans[328,329] and in monkeys[246] the effect may be less marked than in aphakic eyes. This refractive change has to be taken into account if the amblyopia is to be managed optimally.

Spectacles

'The child is very awkward in using convex glasses, and extremely unwilling to wear them. The early use of them is inexpedient'.[4] Saunders was, unusually, wrong: spectacles can be worn at any age, they are not unduly expensive, they can be changed readily as the child grows, they are safe, their magnifying effect may improve the child's acuity and make microphthalmic eyes appear a more normal size. Spectacles can be the only form of optical correction that is available in a community,[330,331] and most children using contact lenses should have a pair of aphakic glasses as a 'spare', for when they are not able to use contact lenses. A child wearing contact lenses, or with an intraocular lens, will need bifocals if his or her acuity is good enough.

Spectacles, however, have important disadvantages: an infant's ears and nose are often too insubstantial to support aphakic glasses, and they also have optical and cosmetic disadvantages.

Contact lenses

Contact lenses were infrequently used to correct aphakic children before the 1960s. In England, the problem was addressed by Mr Montague Ruben, at Moorfields Eye Hospital, who had already shown an interest in contact lenses for infants.[332] He collaborated with the manufacturers of the lathes used to make hydrophilic lenses so that they were able to cut at previously unavailable radii, and a technician was employed to make the lenses specially for infants and young children. These lenses were used extensively, often on a continuous-wear basis, and the early results were encouraging.[28,333] In North America, hydrophilic lenses were not so readily available and most of the early lenses were rigid forms: many were successful.[17,334–336]

The effectiveness of contact lenses in the management of amblyopia is emphasised by the fact that in many units specialising in the correction of infant aphakia rates of successful wear for the critical years of visual development approach 90%.[337–342] This still makes contact lenses the treatment of choice for infant (defined as a child in the phase of ocular and visual development) aphakia,[342,343] and the standard against which other methods of optical correction should be compared before they are used.

Contact lenses have advantages in the correction of infant aphakia;[344] the parameters can readily be changed. Morris *et al.*[321] found that the contact lenses, used to fully correct aphakic infants, reduced in power very substantially in the first years of life; this myopic shift has been confirmed in other series of patients treated with contact lenses.[322,323,345] They are safe,[339–341,346–348] probably more safe than intraocular lenses.[349] They can be used in combination with spectacles for a near addition and to correct aniseikonia.[350]

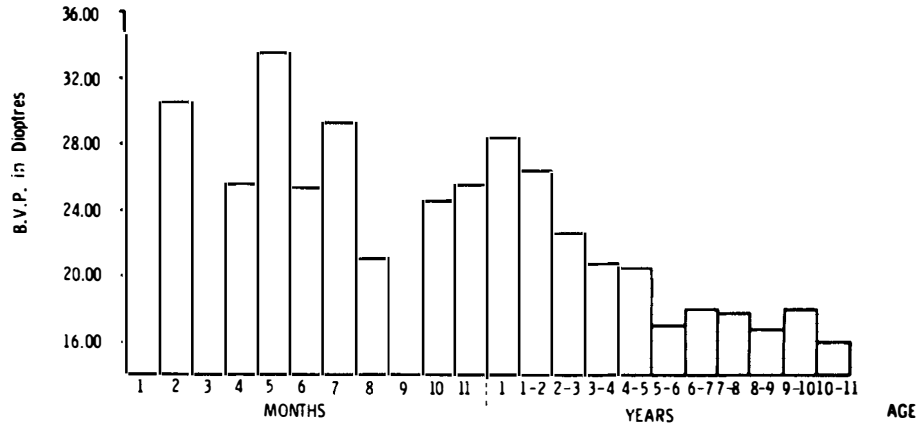


Fig. 13. *The changes in back vertex power (B.V.P.) of aphakic contact lenses in relation to age. The changes are most marked in the first 2 years of life, underlining the need for any form of optical correction to take a continually changing refraction into account. From Morris* et al.[321]

There are, however, a number of risks and disadvantages: hypoxic keratitis,[351,352] infective keratitis,[339] and red eyes without ulceration[339] which are usually caused by chronic hypoxia or loose sutures. Corneal pannus of a millimetre or more[339,346] suggests hypoxia; changing to a rigid gas-permeable lens from an extended-wear lens is effective.[353] Increased corneal thickness occurs but human endothelium studies have not shown significant changes associated with contact lens wear,[347] despite suggestive changes in infant primates.[354] Contact lens fitting and tolerance problems, difficulties with occlusion, and high turnover of lenses may be a source of failure.[27,339,340] The costs can be substantial, especially if the rate of lens turnover is high, if there are complications requiring frequent outpatient visits or admission to hospital, or if the parents live far from the contact lens practitioner, and have to miss work to come for consultations. In 1990 the cost of lenses and solutions in the UK was £187 per year.[339] Contact lenses are difficult to wear if there is external eye disease (as is frequent in Down's syndrome), if the eye is dry,[355] or in microphthalmic eyes or eyes with corneal anomalies.

Epikeratophakia

Early attempts[356,357] were accompanied by failure rates of up to 30% but success rates quickly improved.[358-361] The Committee on Ophthalmic Procedure Assessment[362] found that the refractive prediction was poor overall, there may be a significant time before the graft clears, making the treatment of amblyopia more difficult, and visual acuity after grafting may not improve adequately.[361] Technical success rates in young children, with all that that implies for amblyopia, are lower – 63% in one series[359] – and there is a significant myopic shift in the first years of life.[359,363]

Epikeratophakia, therefore, has rather special indications in patients with congenital cataract. It may be indicated when there is contact lens failure in the older monocular aphake who is less susceptible to amblyopia, or as an alternative to primary or secondary intraocular lenses in older bilateral aphakes who are contact lens and spectacle intolerant. Elsas[23] made the very relevant point: 'The critical factor in getting good vision following monocular cataract extraction was the patient's willingness to patch the better eye, not the method of correction of the refractive error'.

Primary intraocular lenses

As the early published reports of intraocular lens (IOL) surgery indicated,[364–369] there is not any technical problem with the implantation process, but with the management of amblyopia[370] and the short- and long-term complications. Many of these early reports paid not even lip-service to the amblyopia question, and there is still a tendency to regard IOL implantation as a technical problem and an end in itself, not a part of the management of amblyopia.[371]

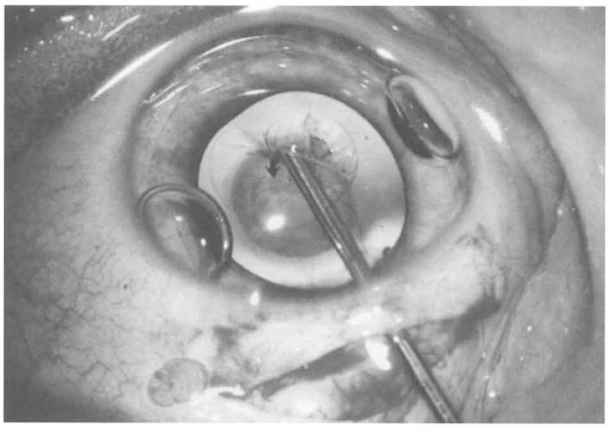


Fig. 14. *Anterior capsulorhexis. The capsulorhexis is proceeding from left to right and is extending too far towards the equator of the lens; the forceps are applying force in the direction of the arrow. The infant lens capsule is very much more elastic than the adult's and the forces needed to produce a small, round, capsulorhexis must be vectored differently, often at more than a right angle to the intended direction.*

For safe IOL implantation in children, a number of factors need to be taken into account: the lens needs to last the lifetime of the child, say 80 years for an infant.[201] Although polymethylmethacrylate (PMMA) lenses in one form or another have been in use for decades and are known to be safe, their use combined with nylon loops has led to problems.[372] Similarly, some flexible lenses were widely used for children in the 1980s[349,373,374] only to be withdrawn in the early 1990s. New materials may make implantation easier, or enable a small incision to be used,[328] but one must be sure of its likely lifespan as removal of a lens is a major intraocular procedure. One-piece PMMA lenses are currently the most used lenses.

The implant needs to be of an appropriate size for the age and eye-size of the patient.[375] Many would be cautious about using an IOL in a microphthalmic eye,[376] whatever the size of the optic and haptics.

Anterior chamber lenses are easily implanted but have an unacceptably high complication rate.[377–380] Today, an 'in-the-bag' technique, through a capsulorhexis that is smaller than the IOL optic diameter (Figs. 14, 15), is the favoured option by nearly all surgeons;[201,285,329,381] a few believe that a sulcus-fixated lens is adequate.

Fig. 15. *A too-small capsulorhexis may contract to form a tiny round aperture.*

There is general consensus that IOLs are appropriate for most children with congenital cataract over 2 years of age.[201,382–384] Younger patients can ethically be implanted if adequate attention can be paid to their optical correction and post-operative management.

To take account of the future growth of the eye the use of a lens of less than the power to give emmetropia may be used.[385] A rule of thumb is to use a lens of 3 dioptres under-corrected for a 2-year-old rising proportionally to a lens to achieve emmetropia at 8 years, though there is no study yet to substantiate this or any other 'formula'. An over-correction with contact lenses or spectacles may be indicated in younger patients. The idea of implanting two lenses, one to be permanently in place in the capsular bag and one in the sulcus to be removed after the growth of the eye has taken place, is probably too invasive. Side effects and complications were frequent in earlier series,[369,380] and are still important. Prolonged, meticulous and honest[377] follow-up is the only way that we are going to find out how much early enthusiasm[386] needs to be tempered with caution.

Posterior capsule opacification is common.[264] There is little doubt that posterior capsule opacification is reduced by posterior capsulotomy or capsulorhexis with or without an anterior vitrectomy.[258,265,268,329,377,387,388] It may occur even after a posterior capsulotomy,[370] probably by invasion of the anterior vitreous or hyaloid face by proliferating lens fibres or fibroblasts, and Metge[389] found the incidence of opacification was 73% in those with an intact posterior capsule and 63% in those who had had a posterior capsulorhexis or anterior vitrectomy.

Uveitis shows itself as a red and photophobic eye, as an eye whose vision is unexpectedly poor due to cystoid macular oedema, and on slit-lamp examination there may be flare, cells in the anterior chamber and keratic precipitates (KP) on the posterior surface of the cornea and both surfaces of the lens. Apple *et al.*[390] noted several of the ways in which post-operative uveitis can occur: mechanical, immunological and complement activation, or the 'toxic lens syndrome', due to the leaching out of toxic chemicals from the lens. Phacotoxic and phacoanaphylactic reactions are more likely to occur as a reaction to the crystalline lens itself[296,297] rather than the implant. Fibrinous uveitis is frequent in the early post-operative period[391] and may result in posterior synechiae or secondary membranes;[241,247,267,374] occasionally it may be severe and the result disastrous (Fig. 16). The membrane may require the use of tissue plasminogen activator,[392] hirudin (experimentally)[393] or YAG laser treatment.[394] Most frequently it is prevented by the use of heparin[395] at a concentration of 5 IU/ml of infusion fluid, and careful handling of the tissues, especially the iris. Unilateral[250] and bilateral[396] blinding uveitis have been described in patients who have had iris-fixated lenses.

Posterior synechiae are common,[370,397] usually related to the post-operative fibrinous uveitis, but reduce with time.

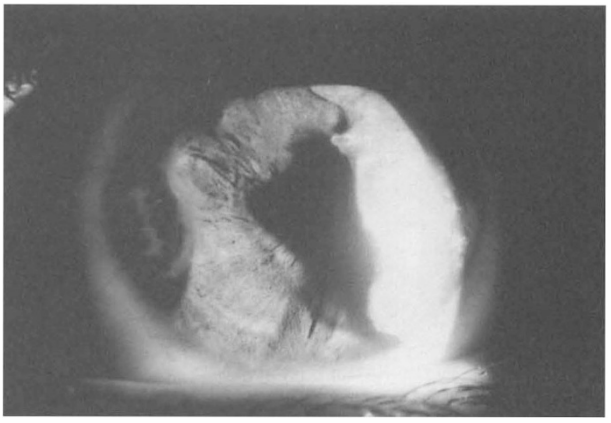


Fig. 16. *Following a severe post-operative fibrinous uveitis, secondary membrane formation and glaucoma this pseudophakic eye became phthisical.*

Lens displacement, dislocation and iris capture are not infrequent in sulcus-fixated lenses[398] but are less frequent with newer posterior chamber lens techniques.

Haptic breakage has been reported in monkeys[399,400] but is probably not significant in man, at least for the current follow-up periods.

In the experimental situation glaucoma can be frequent after neonatal implantation.[401] Malignant glaucoma has been described,[287] and glaucoma occurs after anterior chamber lens implantation,[377] but is not likely to be more frequent after uncomplicated 'in-the-bag' lens placement.

Corneal endothelial counts are reduced after the implantation of an IOL. Lenses implanted outside the posterior chamber are probably worst,[253] whilst posterior chamber lenses may have little effect on the endothelium.[402]

Iris damage, sphincter damage or rupture, irregular pupils and iris atrophy were all common in early series.[369,377,380,390,403] They are largely avoided or reduced by: good pre-operative pupil dilatation, careful technique, avoiding iris touch, pre-operative non-steroidal anti-inflammatory agents (orally or topically), maintaining pupil dilatation by 1:10 000 adrenaline (2.5 ml of a preservative-free solution in 250 ml of infusion fluid) and the use of iris hooks if the pupil is undilatable. Iris prolapse[264] is uncommon now that a scleral tunnel is the most commonly used technique.

Secondary intraocular lenses

Secondary IOLs can be successfully implanted in children who have an adequate posterior capsule to support the lens.[280,398,404] The site of the implant is less satisfactory than in primary implants because secondary IOLs are usually in the ciliary sulcus. It is possible to reconstruct the capsule to, in effect, place the lens 'in-the-bag'.[405] Sulcus ring or suture fixation[280] may well not last the child's lifetime, and in adults secondary IOLs may relatively predispose the eye to later endophthalmitis.[406]

Table 3. *Factors influencing visual prognosis in bilateral cataract*

Factors giving a good prognosis	Factors giving a poor prognosis
Late onset of the visual defect	Early onset of the visual defect
Early surgery, if surgery indicated	Late surgery, if the visual defect is of early onset
Early post-operative optical correction	Late post-operative optical correction
Non-dense opacity	Dense opacity
Lamellar or nuclear opacity	Large opacity or total cataract
Otherwise healthy eye	Other eye abnormalities, i.e. microphthalmos, PHPV, corneal anomalies
Small opacity	Operative or post-operative complications
Good compliance with amblyopia management	Pre-operative nystagmus
	Un-occluded squint
	Poor compliance with amblyopia management

Amblyopia management in bilateral cataract

The most important part of the management of the amblyopia in bilateral congenital cataract involves prompt surgery and accurate optical correction, but if these two factors only are relied on there will almost invariably be one eye that will be amblyopic. The acuity can be measured by Forced Choice Preferential Looking techniques (FPL), and the occlusion can be modulated according to the measured acuity.[210]

The Great Ormond Street Occlusion Protocol for Bilateral Cataracts is as follows:

1. 0–1.2 octaves inter-ocular difference in visual acuity ⟶ no occlusion
2. 1–2 octaves difference ⟶ 1–2 h of occlusion of the preferred eye
3. More than 2 octaves difference ⟶ 2–4 h of occlusion of the preferred eye
4. If no improvement after stage 3 ⟶ Full-time occlusion, review every 1–2 weeks.

Older children who can cooperate with letter matching or other acuity measurements can be occluded in a similar manner; it must be remembered that single-letter acuity measurements may over-estimate the acuity. If the acuity cannot be measured, but the child has a squint, or there is objection to the covering of one eye more than the other, then occlusion of the preferred eye can be carried out empirically. If there is no squint there is a case for occluding alternate eyes for less than 1 h per day.

Visual results in bilateral cataract

Many of the earlier series[20,21,30,272,407–409] that analysed the factors that led to good visual results noted that early surgery was associated with poor visual results – quite the opposite to the situation today. It seems likely that these results were caused by inadequate optical correction and occlusion.

As the experimental work on amblyopia was applied to the clinical management of congenital cataract.[17,333,410] so the beneficial effects of early surgery on visual development became apparent.[25,219,225,336,411,412] Age at surgery is not the only important factor (Table 3), and in one series[26] was not a significant factor. Most series contain a number of biases, including the inclusion of various ages of onset of the visual defect, varying cataract morphology, different ages at operation and differing surgical techniques. It is not yet certain whether visual results differ with different types of optical correction.[381]

It is in infants that the achievement of good visual results is most difficult. Lorenz in 1994[22] summarised her own work and that of others;[25,38,149,333] 50% of her own cases operated in the first year of life with total or axial cataracts achieved 20/50 or better, and 70% achieved better than 20/100. Fifty per cent of the children with early surgery and optical correction had some binocular function. Hing *et al.*[38] retrospectively looked at the medical records of 212 patients with all types of crystalline lens problems requiring surgery. They included 59 patients with simple congenital cataracts, of whom 26 had linear acuity measurements. Their results for those patients who had severe visual deprivation from birth are tabulated in Table 4. It is therefore important to realise, and to communicate to the parents, that in the early onset cases there is likely to be a visual defect even after optimal management.

Hing's[38] infantile group, as expected, did better, with a mean visual acuity of 6/13, and the juvenile group, with a mean age at surgery of 82 months, achieved 6/9. Most of the recent series[329,387,398] that give visual results after IOL implantation show the very good visual results that are expected with most of the patients having later-onset visual deprivation.

Table 4. *Bilateral cataracts with congenital-onset visual deprivation*

Age at surgery (months)	1–2	3–4	5–6	7–12
Mean binocular VA	0.32 (6/18)	0.18 (6/33)	0.23 (6/26)	0.25 (6/24)
No. of patients	6	7	7	6
No. with VA of 6/24+	5	2	4	3
Mean follow-up (months)	48	67	63	57

Unilateral cataract and related disorders

Aetiology

Unlike bacterial congenital cataract, unilateral cataracts are not usually associated with systemic disease. Many apparently unilateral cataracts may be bilateral but asymmetrical and slit-lamp examination is mandatory. Many cases are idiopathic but PHPV and posterior lenticonus are frequent. They may occur with congenital infections; rubella cataracts may be unilateral, without exceptional intraocular damage, but in varicella, cytomegalovirus or toxoplasmosis a cataract is usually secondary to extensive retinal disease. Microphthalmos is a frequent association.

Amblyopia, and the visual results

Published results of unilateral cataract treatment from the early part of this century were universally poor;[17,18,20,21,29,103,215,272,408,413] 'surgery for unilateral congenital cataract is strongly advised against',[103] 'surgery for unilaterals is not justified',[20] 'everyone knows the uselessness of operating on unilateral congenital cataract'.[272]

As amblyopia research reached the ears of clinicians, the first attempts were made at management by early surgery, optical correction and occlusion, some with vision monitored by VEPs or FPL.[17,34,217,414–416] Mostly these were not consistently successful, although promising. 'The dictum of extreme conservatism in the management of monocular cataracts in children needs to be re-evaluated'.[417]

Beller *et al.*, in 1981, occluded both eyes of their early operated patients until the contact lens was fitted, then patched the fellow eye for 96 h, then for a variable amount, depending on the results of VEP monitoring; 5 of their 8 patients achieved 6/9 or better! It is most likely that it is the occlusion more than any other factor that determines the visual outcome.[23,27,418]

A number of studies[19,22,27,204,205,209–212,412,418–423] strongly suggest that surgery before 2 months of post-term age, early and consistent optical correction, and early and full compliance with occlusion treatment are the keys to the achievement of good vision (say 6/9 acuity in the majority) in the monocularly aphakic eye.

There is some evidence that eyes which may appear to have PL visual acuities within the normal range in infancy and early childhood may fall below normal as time goes by.[212,420,423] Not all recent visual results are in accord with the importance of early treatment in the attainment of good acuities;[381] the poor results with early treatment may be due to compliance or optical problems.

The patients who have achieved high acuity levels had a wide variety of occlusion regimes, many monitored by FPL. At Great Ormond Street Hospital, since there is some evidence that excessive early occlusion may be associated with an increase in nystagmus and effects on the phakic eye, we now follow the regime of phakic eye occlusion: 1 h per day for each month of age until the baby is 6 months old; from 6 months old the vision continues to be monitored by FPL, and the amount of occlusion of the phakic eye is modulated, depending on the inter-ocular difference:

1. 0–1/2 octave difference ⟶ 50% of waking hours
2. 1–2 octaves difference ⟶ 75% of waking hours
3. >2 octaves difference ⟶ 100% of waking hours

(reviewed every 2 weeks)

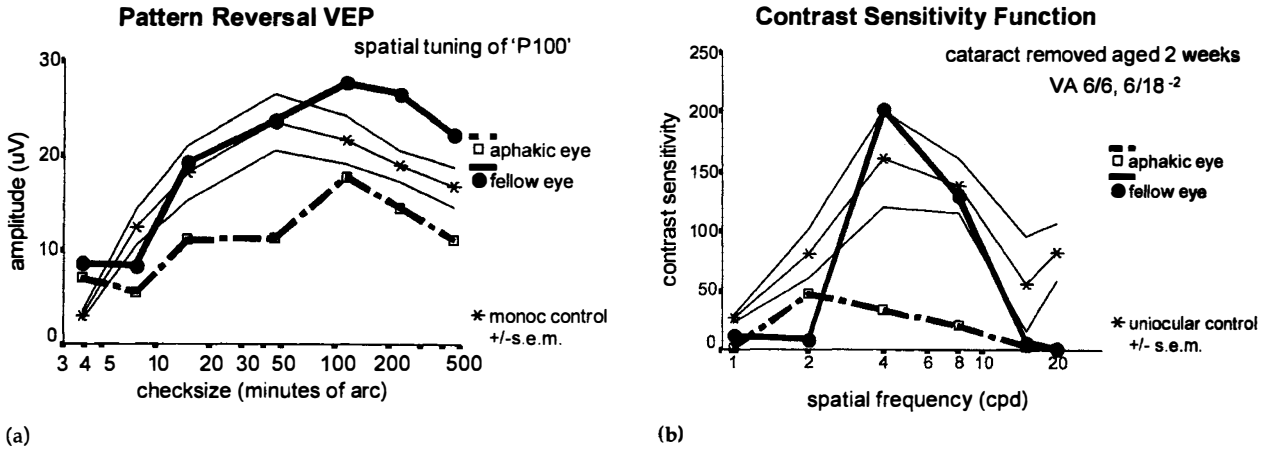


Fig. 17. *Uniocular aphakia. (a) The amplitude of pattern reveral VEPs from aphakic and fellow eyes of a 6-year-old patient are plotted as a function of check-size. These data illustrate the best patient outcome of the ongoing study of Thompson* et al.;[208] *the patient's unilateral congenital cataract was removed at 2 weeks old, and the fellow eye was patched for 50% of the waking day for the first 9 months of life. There is no detectable strabismus, monocular OKN is symmetrical, there is no nystagmus, but no stereoacuity was demonstrable. Pattern reversal VEPs were elicited to 80% contrast black and white checkerboard phase-reversing at 1.5 Hz, 3 reversals per second. (b) Contrast sensitivity functions from the same patient as in (a) also demonstrate that the fellow eye approximates to the mean contrast sensitivity values of the uniocular control group data. The aphakic eye, however, demonstrate marked loss of sensitiviity at all but the lowest spatial frequencies in spite of a linear visual acuity of 6/18 part in this eye. Contrast sensitivity functions were measured to horizontal sine wave gratings presented in a Two Alternative Forced Choice (2AFC) paradigm.*

Binocular vision after treatment for unilateral congenital cataract

Untreated later-onset monocular traumatic cataracts may be associated with horror fusionis without amblyopia, but congenital, unilateral cataracts have traditionally had little[424] or no binocular vision, even when the acuity in the affected eye is good.[22,425] Some cases were demonstrated to have rudimentary binocular vision[423,426,427] and a few had some stereopsis.[428–430] One patient who had had a progressive occlusion regime achieved 20/25 acuity and 50 seconds of arc stereoacuity confirmed by an independent ophthalmologist.[431]

Practical management

Fully informed consent by the parents is a vital prerequisite to proceeding with treatment. It is an essential *fact* that treatment is not mandatory. The parents must be given a clear account of all the alternatives, compatible with their understanding of the matter, and when possible they should be the ones to make the decision. It is vital that the parents are appraised of the fact that the surgery is a small, but important, part of the treatment of amblyopia; good visual results depend on the prolonged use of one or other form of optical correction, but mostly on the maintenance of a regime of occlusion,[23,27] for many years.

Unilateral congenital cataract is not a socially significant disease: it must be a coincidence of considerable rarity if a person who has had such a cataract successfully treated (creating a 'spare eye') blinds the phakic eye. By reducing things to the absurd, there are many circumstances where treatment is actually contraindicated. More obvious ones include first the child living in circumstances where treatment is economically unjustifiable: who would treat a unilateral congenital cataract in many a developing country when there may be many times more who are blind from bilateral cataract? Second, there are the cases where the parents are unable to carry out the all-important post-operative care (one must remember that in many 'advanced' countries there are areas where the adult illiteracy rate is substantial: these disadvantaged people are probably the least likely to go through with the rigours of treatment). Failure to carry out either occlusion or optical correction will inevitably give poor visual results.[23,27] Third, treatment is inappropriate where life expectancy is reduced. Last, there are the cases where the prognosis is very poor; in only exceptional circumstances is a good visual result obtained with treatment that starts after the first months of life.

The parents need to be appraised of the fact that there is a significant chance that the eye may be damaged by the surgery itself, by glaucoma, retinal detachment, or by the wearing of a contact-lens (see above). Even the phakic eye may be blinded.[396]

If taking days off work, long distance travel, repeated clinic visits,[420] fees and all other disbursements are taken into account, the costs and the disturbance to the daily lives of patients and their parents are very substantial.

Although there are now many studies that demonstrate that good visual results can be obtained in a substantial proportion of cases, these results have mostly been obtained by patients at renowned referral centres, with the excellent treatment and parental motivation that that implies. There are a few dozen of these cases in the world literature: the numbers that have not had this happy outcome is not known, but it must be many times the number of good ones. Even in major referral centres, the ability to achieve a good functional result (i.e. driving vision) is unlikely to be more than 50% of patients presenting with unilateral congenital cataract, before exclusions are made.[225] The achievement of good binocular vision is so rare as to be noteworthy, and strabismus is the rule in congenital cataract.[432]

There is a possibility that the vision of the phakic eye may be altered by the treatment of the cataractous eye. This may include defects of acuity[208,433] (Fig. 17) or contrast sensitivity;[433] rarely patients may fix with the phakic eye. These complications are subtle and not universal.[319,423]

Manifest latent nystagmus (MLN) is frequent after unilateral cataract treatment, and it may also occur in untreated cases. Because there is often a nystagmus null position in the adducted position of the phakic eye, there is often an abnormal head posture with the face turned towards the phakic eye. The nystagmus may be less frequent when the monocular deprivation is profound enough to give marked abnormalities in the visually evoked cortical potentials. In these profoundly deprived cases, the persistent optokinetic nystagmus (OKN) asymmetry that is seen in cases where the deprivation is less profound (such as treated unilateral cataract) is not observed, suggesting that it is the lack of unequal input and interocular competition that allows normal OKN development.[434] It has been suggested, to the contrary, that early surgery for monocular congenital cataract (which reduces the deprivation and increases interocular competition) may lessen the nystagmus by allowing a more equal sensory input.[435]

Although the psychosocial consequences may be a significant problem,[337] children who do have behavioural abnormalities that interfere with or even stop treatment may also have other family, medical or environmental disturbances, and these problems are not inevitable[436] or even frequent. An unpublished study from Great Ormond Street Hospital failed to demonstrate any significant problems with bonding and attachment.

There are, of course, important reasons why treatment may be in the best interests of the child; a poor cosmetic appearance because of a white pupil, especially in a microphthalmic eye, is a cosmetic blemish that is unsightly and difficult to treat with a cosmetic shell or contact lens in a young child. The achievement of good vision (say, that compatible with passing the driving test) is regularly achieved. This has the benefit that a 'spare'

eye is created, in case of accident or disease; an amblyopic's fellow eye may be more at risk than the fellow eye of a normal.[437] In a small, but increasing number of cases some degree of binocular vision is achieved,[423,431] which has functional benefits of its own but also reduces the normally very high incidence of strabismus and may reduce the incidence and severity of nystagmus.[435] There is also a case to be made that the 'sound' eye may not always be as sound as it appears at presentation,[438] which may increase the need to treat the cataractous eye.

Most parents are usually keen for their child to have the cataract removed and to undergo the optical treatment and amblyopia management.

Optical correction in uniocular cataract

Aniseikonia makes the use of spectacles the last resort as a primary correction, but they can be useful as a way of fine adjustment to an IOL or for the near/distance correction of bifocals or as a part of a contact lens–spectacle combination to counter aniseikonia.[350] In the latter technique a combination of a high-plus contact lens is used with a minus spectacle lens; the method has not been widely used because the amount of aniseikonia with contact lens correction is not enough to interfere with the development of binocular vision, where this is possible. Contact lenses are the standard at the moment, because of their safety and ability to be changed. IOLs have many advantages, and are the treatment of choice in children over 2 years old;[384] they are being considered for infants.

Postscript

'In cases of congenital cataract ought the operation to be delayed till the patient has attained an age sufficient to enable him to understand the importance of an attempt to restore sight or ought it to be practised during infancy? The answer decidedly is, operate in infancy, and if possible before teething commences' (William Mackenzie, 1840[179]).

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