Editorials

Cataracts in children[1]

The possibility of improving the vision in children with congenital cataract has been with us for many years. It is over 150 years since John Cunningham Saunders, an ophthalmologist at St Thomas' Hospital, described in great detail his method of treating congenital cataract. Both Saunders (1816) and his Scottish counterpart William MacKenzie (1840) were well aware of many of the problems that still confront us now. In his book on the practical management of diseases of the eye, MacKenzie stated, 'the answer decidedly is, operate in infancy, and if possible before teething commences'; by this statement predating the ophthalmologists who in the last two decades, taught by scientists about amblyopia, started again to recommend early surgery.

Apart from minor differences in surgical technique, varying from one to another like the hemlines of fashion, there was little change until the impact of the work begun in the 1960s by Wiesel & Hubel (1963) led to a profusion of exciting studies into visual development.

Leinfelder (1968) was perhaps the first to appreciate the importance of this new information to patients with congenital cataract. He thought that 'failure to obtain good vision is usually the result of the coincidental association of congenital amblyopia'. Perhaps his interpretation of congenital amblyopia is not the same as that which a modern developmental ophthalmologist might have, but there is no doubt that amblyopia due to the failure of development of the visual pathways at all levels from the retina to the visual cortex, both neurophysiologically and structurally, that is caused by the defocused image during a critical period of development, is the most important single factor in the preventable part of the visual handicap in congenital cataract.

We know that, given appropriately timed surgical and optical treatment, patients with bilateral cataract achieve high levels of vision (Taylor *et al.* 1979, Gelbart *et al.* 1982). From the theoretical standpoint it is of even greater importance that it is possible to achieve good levels of acuity in patients with unilateral congenital cataracts by very early surgery and optical correction (Beller *et al.* 1981, Benezra & Paez 1983). It was previously thought that the treatment of unilateral cataract, providing the visual defect was truly congenital, resulted in no significant improvement in the vision because of irreversible amblyopia. Although these newer visual results are very encouraging, there are substantial disadvantages to operating on the infant's eye. The disadvantages of early surgery, the disruption to family life at a crucial time, the risks of operating on a smaller eye the pupil of which does not fully dilate, the rapid opacification of the posterior capsule which gives rise to difficulty in refraction and the prescription of the appropriate powered optical correction, and the greater difficulty in postoperative examination should make the ophthalmologist faced with an infant with congenital cataract duly cautious. Congenital cataract is still the most frequent cause of preventable childhood visual handicap (Taylor & Rice 1982) but, by early detection, new techniques of surgery and optical correction, the outlook for the future may be improved.

The preoperative assessment of vision has been helped by studies and observations on visual development, and relatively objective techniques of visual assessment have given depth to our understanding of vision in these children. Pattern visually evoked responses (Odom *et al.* 1981) and forced preferential looking (Jacobson *et al.* 1981, Dobson & Teller 1978, Atkinson *et al.* 1977) have enhanced our ability to know whether and when to operate. However, careful clinical assessment of how the infant reacts to his environment is still the standby of virtually every clinician however much he may be helped by further aids.

Preoperative investigation of the child with congenital cataract has undergone refinement in the last few years. There have been several studies into the aetiology of congenital cataract, most of which have indicated that hereditary idiopathic cataracts are common, accounting for about one-third of congenital cataracts. The list of associated metabolic disorders and syndromes which have congenital cataract as one of their manifestations is enormous, but it seems unnecessary to exclude each one of these in turn in every patient. Clinical examination by an ophthalmologist should exclude certain developmental anomalies of the eye associated with cataract, and simple tests for urine reducing substances in the child, and after a milk 'load' in his mother, backed up where necessary by galactose enzyme studies, together with urine, amino-acids and blood electrolyte, haemoglobin and calcium estimations, will reduce the workload of the hospital laboratories and will not miss any otherwise treatable patients. The role of the paediatrician in this regard is most important. He will be able to relate the cataract to any dysmorphism or systemic disease and help the

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ophthalmologist with developmental assessment and examination of the infant, and the management of any other aspects of the systemic disease.

It is fashionable to decry the advances made by the recent methods of surgery for congenital cataract. However, there is little doubt that improvements in techniques of extracapsular cataract extraction practised in adults have made a great difference to the results of the now classical technique of aspiration introduced by Scheie (1960). Although Scheie's technique and its modifications have been very satisfactory in terms of vision, it is only the use of more modern techniques that has made it less likely for the child to have the numerous secondary needling operations that were characteristic of aspiration by the old technique. Opacification of the posterior capsule is not important in older children or adults who can have elective surgical or laser capsulotomy, but it is of vital importance in the management of infants with congenital cataracts since it will cause amblyopia either by direct effect or by preventing accurate refraction and dispensing of the appropriate optical correction.

One answer is the removal of the whole lens, including the capsule, by the technique of lensectomy. A variety of instruments introduced for vitrectomy are now available for use in the anterior segment. The anterior approach through the limbus or through the cornea is the usual one in infancy (Taylor 1981) because of the uncertain anatomy of the pars plana in younger children. Lensectomy not only removes the problem of opacification of the posterior capsule, but the suction-cutting instrument can also remove many of the associated intraocular abnormalities such as persistent hyperplastic primary vitreous, or enlarge tiny fixed pupils which can interfere with postoperative optical correction. The major disadvantage of lensectomy is that we do not know whether the disturbance to the vitreous, which is inevitable with the removal of the posterior capsule and the vitrectomy that is carried out to prevent the vitreous prolapsing into the wound, will influence the risk of retinal detachment seen with earlier techniques of congenital cataract surgery.

Congenital cataract surgery is not without its complications, principally glaucoma (Phelps & Arafat 1977) and retinal detachment (Kanski *et al.* 1974). Characteristically retinal detachment is delayed in onset, occurring two or three decades following cataract surgery. This complication is largely seen in eyes that are predisposed to retinal detachment by axial myopia and anomalous vitreoretinal adhesions, and is a consequence of late posterior vitreous detachment. The prognosis for such detachments has in the past been poor because of impaired visualization from capsule and cortical lens remnants, bound-down pupils and vitreous opacification. However, with modern methods of cataract surgery such as lens aspiration, it is hoped that the prognosis for repair of any subsequent detachment will be improved, but only time will tell whether such methods of cataract extraction will influence the long-term incidence of retinal detachment. Inflammatory complications are less common than in the days of Frederick Cordes (1957).

Whilst surgery is of obvious importance and usually uppermost in the surgeon's mind, from the point of view of vision the provision of an adequate optical correction, which results in the treatment of the associated amblyopia, is almost as important in achieving a high level of visual acuity and it is certainly much more difficult both for the doctor and the parents.

Whilst intraocular lenses have gained widespread acceptance in adults, their use in congenital cataract in childhood has still not gained widespread acceptance for three main reasons. Firstly, the refractive error of aphakic infants often requires a contact lens of over +30.00 dioptres, and an intraocular lens with sufficient power to correct this degree of refractive error would have to be of a power of up to +40.00 dioptres. Such lenses are not yet widely available. Secondly, the eye grows substantially in the first two years of life and any intraocular lens that corrected the initial refractive error would certainly be wrong for the refractive error present at the end of the first year. Thirdly, the complications of lens implantation must be remembered (Hiles 1977). The presence of a lens in the eye excites a response which may result in degradation of parts of the lens and this response may be more vigorous in childhood (Mehta 1979). The lenses will remain in the eye for a lifetime since removal is traumatic, and it is difficult to believe that implantation of lenses in young children, whose life expectancy is virtually all of three score years and ten, is currently justifiable. However, with the advent of better techniques for lens implantation and better design and construction of lenses from one piece of relatively inert material, there may yet come a time when lens implantation in childhood is more widely accepted, especially for traumatic unilateral cases in preverbal and other young children. This should not be taken as an exhortation to use intraocular lenses more widely, especially since virtually every child, given appropriate training of the parents, can be fitted with a contact lens.

The use of contact lenses in infancy is now widespread and their use depends on close cooperation between the parent, child, ophthalmologist and optician; although encouraging results have been reported, the risks and difficulties of contact lens treatment in infancy cannot be discounted.

Keratorefractive surgery, in which the refractive power of the cornea is increased by a variety of means such as the use of on-lay grafts of the cornea (Morgan *et al.* 1981), may provide a further option in the future by providing at least part of the optical correction of aphakia. However, the technique is elaborate, expensive and unproven.

Whilst few would disagree that the treatment of unilateral congenital cataract by surgery alone is not indicated because of the poor visual results, there are a few encouraging results with the achievement of high levels of acuity in unilateral congenital cataracts, where the visual defect has been truly congenital, by neonatal surgery and optical correction, either by contact lenses (Beller *et al.* 1981) or by the use of intraocular lenses. It is extremely difficult to achieve high levels of acuity since the results of Beller *et al.* have not been repeated and they were achieved under exceptional circumstances. It is, however, reasonable to offer parents the possibility of surgical and optical correction in unilateral congenital cataract if they fully understand the risks and difficulties of treatment, and if there is any possibility that the visual defect (not just the cataract) has been acquired after birth, so that the eye has been, albeit for a short time, visually experienced. Another situation in which treatment may be indicated is the congenital rubella syndrome, where there is an incidence in the second decade of life of disciform degeneration at the macula destroying good vision of the eye unaffected by the cataract. Unilateral congenital cataract may also give rise to a cosmetic defect, but this is not severe unless there are associated abnormalities. If the decision is made to remove the cataract on cosmetic grounds, it is probably best to remove it in early childhood – but after the first two years of life – so that it does not become calcareous and difficult to remove by conventional surgery.

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Psychotherapy research

Between the two world wars, psychotherapy – usually meaning psychoanalysis or a derivative – while ignored by organically-orientated psychiatrists, was assumed by others to be an effective, possibly the only effective means of treating neurotic and related disorders. This assumption was based on slight objective evidence and suffered a grave affront from a paper by Eysenck (1952) in which it was argued that improvement following psychotherapy is no greater than that to be expected from rates of 'spontaneous' recovery. This view has been supported in later papers by Eysenck and by others (e.g. Rachman & Wilson 1980), and contradicted (e.g. Bergin & Lambert 1978), and remains a focus of controversy. Mean-

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