

Update on squint and amblyopia

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At birth a baby's vision is very immature, with an acuity of around 3/60^{1,2}. Visual experience in infancy and childhood has a major influence on the development of the visual pathways, particularly the visual cortex^{3,4}. During the critical periods of visual development visual acuity develops as do other modalities such as contrast sensitivity, colour, pattern and motion perception, and binocular function with stereopsis^{1,2}. Disturbance to normal visual development will affect the maturation of these different visual functions, with associated structural changes in the visual pathways and cortex, resulting in amblyopia—i.e. reduced acuity in one or sometimes both eyes without any ocular lesion. Either these children lose binocular function as a result of a squint or they acquire a squint because of poor development of binocular function. The commonest causes for amblyopia are a squint (strabismic amblyopia) or blurring of the image in one eye due to unequal refractive power in the two eyes (anisometropic amblyopia). Psychophysical tests show differences between strabismic and anisometropic amblyopia, but for the patient the effects are similar.

Squint and amblyopia are common conditions: about one in fifty children have a squint⁵, and up to 5% of the population have an amblyopic or lazy eye^{6,7}. In view of their lifelong impact on visual function and physical appearance, with consequences for education, jobs and psychological wellbeing⁸, good management offers substantial long-term benefits. Recent work favours early diagnosis and treatment, and there has been increasing effort to treat children as soon as possible. There is also renewed interest in the treatment of adults. This article reviews these areas.

STRABISMUS

Genetics

Siblings of children with a squint are themselves often checked for a squint. The value of this has been shown by the twin eye study based at St Thomas's Hospital, which indicates that genetic influences are important in convergent squints (the commonest childhood squint) although not in divergent squints. Siblings of a child with a convergent squint have a doubled risk of developing a similar condition⁹.

Neonatal deviations and infantile esotropias

Many small babies are thought by their parents to have crossed eyes and the traditional advice has been to wait and see if this settles and to advise ophthalmological referral at about three to four months of age if it does not. In North America some surgeons have been recommending very early squint surgery, sometimes within a few months of birth, to obtain the best chance of stereopsis^{10,11}. Support for an initial wait-and-watch policy has come from Horwood, who has shown in a prospective longitudinal study of 1150 normal children that neonatal misalignments are extremely common¹². However, the 89 children (7.7%) with very frequent neonatal misalignment (more than 15% of the day) were significantly more likely to develop a convergent squint and to have associated refractive errors. Interestingly, children without any evidence of neonatal deviations are at greater risk of astigmatism. In later life the deviations are commonest at about seven weeks of age, with a decline in prevalence by fifteen weeks. They tend to be associated with near fixation, and are thought to be due to immaturity of vergence mechanisms¹³. Two American studies, analysing the histories of children seen with early-onset squints, have confirmed the variability of early eye alignment and have helped to define the risk of development of a true squint^{14,15}. Whereas a small-angle intermittent squint will frequently resolve, a child presenting after ten weeks of age with a constant deviation of more than 40 dioptres on two examinations is very unlikely to recover spontaneously. These data will allow early decisions as to which children will need squint surgery.

Recent studies have suggested that both the duration of ocular misalignment and the age at which the eyes are straightened are of prognostic importance: they correlate not only with improved stereoacuity outcome but also with improved stability of the long-term eye position and a lesser need for corrective surgery at an older age^{10,16}. Stereoacuity develops between three and five months of age and matures to a near adult level during the first two years of life². Surgery before the age of two years—possibly within the first six to twelve months of life—is now believed to maximize the chance of a good outcome¹⁶. Obviously, other points need to be considered such as the technical difficulty of operating on a small eye. If it becomes established that very early surgery yields the best outcomes, this will have important practical implications for early referral, rapid assessment

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and availability of surgeons and anaesthetists experienced in operating on and anaesthetizing small children.

Muscle pulleys

Studies of the functional anatomy of the extraocular muscles have demonstrated the importance of connective-tissue sleeves or pulleys surrounding the rectus muscles just posterior to the equator of the eye¹⁷. These act as the functional origins of the muscles, rather than the origin at the orbital apex, and prevent sideways slipping of the muscles during eye movements. MRI work has shown that mal-position or heterotopia of the pulleys can be a major factor in incomitant squint and can mimic, for example, inferior oblique overaction or Brown's syndrome^{18,19}. Damage to these pulleys during surgery may be a further cause of incomitance in a squint.

Squint surgery in older patients

Strabismus surgery can be valuable not only in children but also in older patients. A US analysis of Medicare claims showed that more than 6500 strabismus procedures were performed on patients over 65 years of age²⁰, whilst one UK surgeon reported that 7% of his surgical strabismus workload was in the over-60s²¹. The range of diagnoses was varied but 48% of elderly patients had diplopia, which is a significant functional disorder that requires correction. No serious surgical or anaesthetic complications were reported. With the increase in the aged population the strabismus surgeon will be dealing increasingly with adult strabismus. If there are no anaesthetic contraindications, age should not be a barrier to ocular realignment.

AMBLYOPIA

In 1997 a National Health Service systematic review reported on preschool vision screening²². It stated that there was a lack of good-quality research into the natural history of amblyopia and the disabilities it caused. It questioned the effectiveness and value of screening of preschool children for amblyopia and the effectiveness of available treatment. It also suggested that treatment for amblyopia might be psychologically damaging and advised further clinical studies into the extent of disability attributable to the amblyopia, the incidence of blindness or partial sight due to amblyopia with subsequent loss of the fellow eye, and the prognosis for vision in the amblyopic eye under these circumstances. The conclusion of the British report was the reverse of similar reviews undertaken in North America^{23,24}. Much evidence has since accumulated to show that screening and treatment for amblyopia are important, with lifelong benefits to patients. Some of the studies are discussed below.

Disability due to amblyopia

Patients with amblyopia may be debarred from undertaking certain jobs because they fail the required visual standards (in particular they may not be able to hold a class II professional driving licence) and additionally they may be at risk of visual handicap if they should damage or lose the vision of the fellow eye⁸.

A national population-based epidemiological study undertaken through the British Ophthalmological Surveillance Unit examined the risk, causes and outcomes of visual impairment after loss of vision in the non-amblyopic eye, and the likelihood and factors predictive of improvement in the amblyopic eye^{25,26}. The results indicate that both the risk of permanent visual impairment and its consequences, for the individual and for society in general, are greater than previously assumed, with limited scope for secondary and tertiary prevention of visual impairment in this context. Clinically useful improvement in vision in the amblyopic eye is uncommon, but is associated with greater severity of visual loss in the non-amblyopic eye (i.e. the worse the sight loss in the 'good' eye, the better the chance of improvement in the amblyopic eye), age, previous acuity in the amblyopic eye and new optical treatment²⁶. These studies have emphasized the importance of providing children with good vision in each eye, and also undermined the long-held belief that the amblyopic eye will improve if the non-amblyopic eye is damaged.

Screening and treatment of amblyopia

In Sweden and Israel^{27,28} the prevalence and severity of amblyopia have declined substantially where screening programmes are in place. Moreover, vision does not improve in patients not complying with treatment, and can subsequently be improved with supervised compliance²⁹.

The mainstay of amblyopic treatment remains refractive correction with spectacles, and occlusion therapy³⁰. Treatment with spectacles alone may be enough to improve vision in some patients with late-onset amblyopia. Compliance with treatment is the major factor in response, and work with occlusion dose monitors has shown that in many cases compliance is not what is reported. In these circumstances admission to hospital may be effective in securing the necessary compliance²⁹. Most of the improvement in vision is obtained in the first four hundred hours of occlusion therapy³¹.

Drug treatment for amblyopia has been tried, with levodopa and carbidopa. In the small number of trials reported, usually including patching as well, vision has improved modestly³². This treatment is as yet unproven, and many clinicians are wary of using pharmacological treatments. In adults the side-effects of these drugs are well

documented³³; long-term side effects in children, particularly on behaviour, may be difficult to detect.

Assessment of vision defects in children with amblyopia

The accurate assessment of vision in children is particularly important in the detection of amblyopia and in the assessment of response to treatment. The standard method of assessing vision in older children and adults has been the Snellen-based letter chart, but this is not appropriate for very young children. For these, picture-based tests or single letter tests are used. However, it is important to realize that these can give an apparently better acuity than a test with a line of letters, because of the effect of crowding. This means that a child who is apparently doing well with a single letter test can appear to deteriorate when he or she starts with a linear test—which can be very disappointing to parents unless the reason is carefully explained. Because the differences between lines in the Snellen chart are unequal, the research standard for testing vision in adults is a logMAR based chart. Similar logMAR based test cards have now been developed for children³⁴. They seem to be much more sensitive for detecting interocular acuity differences^{35,36}, and are gaining popularity for detecting and managing children with amblyopia.

Treatment of adults with amblyopia

The success of amblyopia treatment declines steadily with age. For most amblyopes it becomes ineffective around the age of eight, although some straight-eyed anisometropes respond well to patching at much later ages. El Mallah *et al.*³⁷ reported that adult amblyopic patients had an increase in acuity in their amblyopic eye after losing central vision from macular disease in their good eye. This observation aroused considerable interest because it raised the possibility of adult cortical plasticity. However, epidemiological evidence indicates that such improvement is uncommon²⁶. Mosley and Fielder³⁸ suggest that a clinical trial is needed³⁸. In any such trial it will be important to determine whether patients have anatomical loss of retinal ganglion cells in their affected eye since such loss will cause transneuronal degeneration in the central visual pathway related to the non-amblyopic eye³⁹. This is likely to increase the scope for recovery of vision in the amblyopic eye, as indicated by the improvement seen in the amblyopic eye of one patient when he had a retinal detachment in the fellow eye and then showed further improvement when that eye had to be enucleated⁴⁰.

CONCLUSION

Treatment of strabismus and amblyopia in childhood reduces the risk of later visual and employment

disadvantage and maintains a more stable ocular alignment. Treatment of amblyopia in adulthood is not yet a possibility. There will be an increasing trend to early squint surgery in young children and an increase in demand for strabismus surgery in elderly patients.

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