EXAMINATION OF YOUNG CHILDREN

3

Objectives

In examining the eyes and vision of preschool children, under the age of 4–5 years, our approach and method have to be modified from the routine appropriate to older children and adults. It is likely that there will be much less cooperation on the part of the very young patient so we need to use quick, simple tests that can be applied for the short time we can count on the patient's attention. Precise measurements may not always be possible and we have to look for significant departures from normal.

A young child cannot give subjective symptoms and we have to rely on the observation and impressions of the parent. The family history will be very important. Children whose parents or whose brothers or sisters have strabismus or amblyopia are very much more likely to develop these conditions.

This chapter is mainly concerned with the professional eyecare of young children, rather than with screening methods. No visual acuity test is likely to be adequate for screening by itself: binocular vision tests and ophthalmoscopy are essential for detecting a wide range of potential problems (Rydberg & Ericson 1998). Regular eye examinations or vision screening throughout school life seems important, since 70% of children who have significant ocular conditions go undetected by their parents or teachers (Rose et al 2003). Screening methods for preschool children have been reviewed (Bishop 1991, Simons 1996) and a new computerized system shows promise for screening school children (Thomson & Evans 1999).

Unfortunately, many areas of the UK do not have good preschool or school screening programmes in operation and optometrists should therefore seek to examine all children (from neonates onwards) at routine intervals. This may not be necessary if there is a local screening programme that is both thorough and rigorously audited. However, even the best screening programme may fail to detect some anomalies and children with risk factors (family history, birth factors, symptoms) should always receive professional eyecare.

Active pathology

Anomalies of binocular vision in children, as in adults, may be a sign of active pathology. The first responsibility of the practitioner is to investigate this possibility. It is particularly important to check for incomitancy, note the palpebral openings and carry out careful ophthalmoscopy. Where there is any doubt, the patient will need to be referred for medical investigation before proceeding. It must be remembered that there are methods of investigation that are available in some hospitals but are seldom possible in primary eyecare practices.

Development of vision

In order to be able to assess the eyes and the vision of infants, it is important that we should have some idea of how vision normally develops from birth through infancy and childhood to adult vision. Although a lot more is now known about this, there are still many gaps in our knowledge. Normal vision requires a good optical system with a focused image and good resolution. Optical resolution has also to be matched with a neural receptor system of good resolution and a neural image processing ability leading to psychological perception. The perceptive level itself is very dependent on previous visual and other sensory experience. Obviously this sensory experience cannot be present at birth. As experience grows, reflexes are reinforced and associations between different sensory input and experiences are formed. For example, it is clear that very early in life an infant learns to recognize the mother's face and the meaning of different facial expressions. All visual functions are built as they are reinforced by experience acting on the anatomical and physiological systems, which, although not complete at birth, mature early in life and allow the full potential of the visual system to develop.

The macular region of the retina is poorly developed at birth and both this and the visual cortex continue developing after birth. One would expect, therefore, that the spatial visual functions of neonates are significantly below the accepted norms for adults and this is the case. It should be stressed that there is a wide variability in the development of visual functions and the figures given below are illustrative typical values from the literature. All aspects of visual development, normal and abnormal, have been reviewed in depth by Simons (1993). In contrast to major spatial resolution deficits seen in young infants, temporal resolution (e.g. flicker detection) is remarkably good (Teller 1990). Some theories account for this and other findings by considering the development of various parallel pathways, including cortical and subcortical components (Teller 1990).

Contrast sensitivity at birth is approximately 1/30th of its eventual level and it improves rapidly over the first 6 months, achieving adult levels at the age of 3 months for low spatial frequencies but taking up to 3–4 years to reach adult levels at high spatial frequencies. Faces are attractive to infants, and studies have shown that these are fixated at the age of 2 months but not at 1 month. Infants have some ability to discriminate between expressions at about 3 months and between faces at about 5 months.

Visual acuity

The rate at which visual acuity appears to develop in human infants depends on how it is defined and on how it is investigated. Using the objective assessment of the visually evoked potential (VEP), it appears that the infant's ability to resolve patterns improves from a level equivalent to about 6/38 at the age of 1 month to the equivalent of 6/15 at about the age of 6 months (Teller 1990). Another method, physiological optokinetic nystagmus (OKN), is mediated via a different nervous pathway to normal visual acuity. OKN methods are not in common clinical use and will not be covered here.

The most common approach to assessing visual acuity in infants is based on 'preferential looking', when the practitioner observes whether the infant turns the head or eyes to look at a grating or picture rather than a grey patch of equal size and luminance. If no preference is shown for the grating, it is assumed that it cannot be resolved; although it should be noted that this approach assesses extrafoveal vision. This method indicates that visual acuity is approximately equivalent to 6/180–6/90 at age 1 month, 6/90–6/36 at 3 months and 6/60–6/18 at 6 months (see Appendix 2).

Obviously, a Snellen-type acuity measurement cannot be made until the child is older, even if specially designed tests are used that employ pictures. These tests suggest that 6/6 acuity is not achieved until over the age of 3 years. All these Snellen-type tests, however, involve an element of form perception. At the simplest, the child must be able to recognize the difference between a square, a circle and a triangle. Form perception is developed later than simple resolution, so that Snellen-type measurements assess a more advanced form of vision than preferential looking tests. Visual acuities with Lea symbols are typically about one LogMAR line better than with Snellen (Vision in Preschoolers Study Group 2003). Most children can letter-match by the age of 3 years but monocular acuities are only possible in two-thirds of children aged 3–4 years and nearly all over 4 years (Salt et al 1995).

Some norms for various clinical visual acuity tests can be found in Table 3.1 and a guide for clinical use is given in Appendix 2.

Refractive error

The refractive error during the first year of life is very variable in most infants. At birth it is of the order of +2.00 DS (SD = 2.00 DS). It is approximately +2.50 DS in about 50% of infants. There is hypermetropic astigmatism in 29% and myopia in 23% (Cook & Glasscock 1951). In many infants, high degrees of astigmatism are observed during the first year, but this is variable and usually disappears before the end of the first year. On average, the hypermetropia decreases rapidly during the first year to a mean level of about

Table 3.1 Various estimates of the development of visual acuity with age. Approximate Snellen equivalents are given

Age (months)	*Test*	*VA (Snellen equivalent)*	*Source*
Newborn	Unspecified	6/300	Grounds 1996
	Preferential looking	6/360–6/120	Stidwill 1998
	Unspecified	6/240	Ansons & Davis 2001
1	Preferential looking	6/180	Teller 1990
	Unspecified	6/200–6/90	Grounds 1996
	Preferential looking	6/480–6/120	Stidwill 1998
	Unspecified	6/180–6/90	Ansons & Davis 2001
3	Unspecified	6/90 to 6/60	Grounds 1996
4	Preferential looking	6/50	Teller 1990
	Preferential looking	6/120–6/30	Stidwill 1998
4–6	Unspecified	6/18 to 6/6	Ansons & Davis 2001
6	Preferential looking	6/30	Teller 1990
	Unspecified	6/60–6/36	Grounds 1996
	Preferential looking	6/90–6/24	Stidwill 1998
9	Unspecified	6/46–6/24	Grounds 1996
	Preferential looking	6/90–6/24	Stidwill 1998
12	Preferential looking	6/24	Teller 1990
	Unspecified	6/24	Grounds 1996
	Preferential looking	6/90–6/24	Stidwill 1998
12–17	Cardiff cards preferential looking	6/48–6/12	Adoh & Woodhouse 1994
18	Unspecified	6/18–6/12	Grounds 1996
	Preferential looking	6/45–6/15	Stidwill 1998
18–23	Cardiff cards preferential looking	6/24–6/7.5	Adoh & Woodhouse 1994
24	Preferential looking	6/12–6/9	Teller 1990
	Unspecified	6/12–6/9	Grounds 1996
	Preferential looking	6/30–6/12	Stidwill 1998
24–29	Cardiff cards preferential looking	6/15–6/7.5	Adoh & Woodhouse 1994
30–36	Cardiff cards preferential looking	6/12–6/6	Adoh & Woodhouse 1994
36	Preferential looking	6/6	Teller 1990
	Preferential looking	6/12–6/5	Stidwill 1998
	Single optotypes	6/6	Ansons & Davis 2001

(Continued)

Table 3.1 (Continued)

Age (months)	*Test*	*VA (Snellen equivalent)*	*Source*
36–48	Single optotypes Crowded optotypes (Cambridge cards)	6/6 6/12–6/9	Atkinson et al 1988 Atkinson et al 1988
36–48	Crowded optotypes (Cambridge cards)	6/6	Atkinson et al 1988

+1.50 D at age 1 year and then decreases slowly at the average rate of about 0.1 D per year until the age of 10–12 years, when the typical rate of change slows down even more. High degrees of myopia present at birth can lead to esotropia with onset in early childhood (Ansons & Davis 2001). Myopia over 5 D in children under the age of 10 years can be associated with systemic or ocular pathologies and it has been recommended that these cases are referred (Logan et al 2004), preferably to a paediatric ophthalmologist.

Nearly three-quarters of children with esotropia and/or amblyopia have a 'significant' refractive error (myopia, hypermetropia ≥ +2.00 D, anisometropia ≥ 1.00 D, astigmatism ≥ 1.50 DC) and children with these refractive errors have a one in four chance of developing strabismus and/or amblyopia (Bishop 1991). Hypermetropic children are at higher risk of developing accommodative esotropia if there is a positive family history of esotropia, subnormal random dot stereopsis or hypermetropic anisometropia (Birch et al 2005).

At the age of 6 months hypermetropia over +4.00 D in any meridian is abnormal and 91% of 6-month-old infants have less than +5.50 D hypermetropia by cycloplegic retinoscopy (Ingram et al 2000). At the age of 1 year, hypermetropia over about +3.00 D is abnormal, as is hypermetropia over +2.50 D at 3 years. The management of abnormal degrees of hypermetropia is controversial and will depend on the degree of departure from normality, symptoms (parental reports), family history and other optometric findings. At the least such cases should be examined very regularly (sometimes initially monthly) and parents should be alerted to look for strabismus. Where there is a high risk of developing strabismus, refractive correction should be considered. In the absence of a manifest strabismus the full refractive correction may be undesirable because this might prevent the 'emmetropization' process (Hung et al 1995) and a partial correction will have less effect on emmetropization (Ingram et al 2000). The emmetropization process appears to be deficient in strabismus (Ingram et al 2000), so there is no reason not to prescribe the full hypermetropic correction in these cases.

Anisometropia of more than 1.00 D or astigmatism over 2.00 D at any age over 1 year indicates the need for a correction to be considered. Normal acuity development requires a good and equally sharp image in both eyes.

Low degrees of myopia are less of a concern because young children spend most of their time viewing near objects. Ehrlich (1996) argued that, up to the age of 2 years, myopia should only be corrected if over −3.00 D. Some prescribing guidelines for refractive errors can be found in Appendix 2, although other factors always need to be considered, including visual acuities, binocular vision function, the reliability of the clinical test results and the likelihood of parents returning for scheduled appointments and for unscheduled appointments if any problems are observed.

Uniocular fixation

Normally, the peripheral retina is well developed at birth but the central 5° of the retina is at best partially functional at birth. Hence, in the first few weeks of life precise foveal fixation is unlikely but fixation of suitable targets may take place at non-foveal retinal locations. The tendency to fixate new objects increases during the first 3 months of life. The fixation reflex requires reinforcement by active vision if it is to develop normally. If the system is faulty in some way, this may prevent normal development of central fixation and therefore normal acuity.

The fixation reflex does not become firmly established until later and, if anything impedes it during the first 2–3 months of life, central fixation can easily be lost. This period of 2–3 months of rapid maturation is known as the critical period so far as fixation is concerned. In comparison, the critical period for acuity development is 2–3 years. The critical period is followed by a further interval during which the system can easily break down: the plastic period. Central fixation can still be lost up to the age of 3 years if anything disturbs the system.

Occasionally there are abnormalities in the foveal nervous system that are present from birth but these account for only a very small number of eyes with fixation failure. Most loss of fixation arises from the lack of a central image in a strabismic eye or, occasionally, from a very blurred image. If either of these is present during the critical period there will be a failure of normal fixation and acuity to develop and, unless treatment is given before the end of the plastic period, it is unlikely that central fixation can ever be achieved. The longer the strabismus or the blurred vision is left untreated the less chance there is of ever getting central fixation with full acuity. This obviously emphasizes the need for early detection and treatment. It has also been shown that, if an eye is occluded for a long time for any reason (e.g. ptosis or cataract) during the critical period, this too will impede the acuity development (deprivation amblyopia). If the occlusion occurs before the age of several months, central fixation will also be lost.

Blinking reflex, vestibulo-ocular reflex, saccadic and pursuit eye movements

At birth, a blinking response to bright lights should be present (Mehta 1999). The vestibulo-ocular reflex is present (in full term infants) by the seventh day (Mehta 1999). Saccades are readily apparent in neonates but tend to be small and are relatively unresponsive to novel stimuli in the periphery. By the second week of life small saccadic eye movements can reliably direct the line of sight towards a peripheral target and after the second month large single saccades occur. Although this resembles the situation in adults, adult levels of saccadic accuracy have still not been reached at 7 months of age (Harris et al 1993). Compared with adults, the saccadic latencies are prolonged in infants, preschool children and possibly even older children.

Pursuit eye movements are present in neonates but are brief, intermittent and frequently interspersed with saccades. Parents and clinicians should be able to detect the behavioural sign of infants fixating and following on targets of interest by the age of 2 months. The visual system becomes better at pursuing faster targets over and beyond the first 10–12 weeks. In infantile esotropia there is usually crossed fixation so that fixation occurs with the right eye for objects in the left field and with the left eye for objects in the right of the field. Under these circumstances, the pursuit reflex may develop normally in each eye for half the field. In the other half, the eye may not follow correctly if the other eye is covered. This will give the appearance of a lateral rectus palsy. As the crossed fixation is a form of alternating strabismus, it usually allows the development of good acuity.

Fusion, vergence and stereopsis

Rudimentary binocular alignment without cosmetically noticeable strabismus is often present at birth but true bifoveal fixation probably does not occur until the age of about 2–3 months. Occasional (<15% of the time) *neonatal misalignments* of the visual axes are common and usually innocuous in the first month of life but should become much less common in the second month (Horwood 2003b). These are most often convergent, probably reflect the normal development of vergence control and only require referral if they worsen after 2 months or if there is an intermittent deviation at 4 months (Horwood 2003a).

Conjugate eye movements may or may not occur in neonates, although convergence may not occur for 2 months. One view (Schor 1993) is that tonic, proximal and accommodative vergence are present at birth but fusional vergence develops later (at about 4 months), possibly in line with improved visual acuity. A clinical implication of this is that infants are unlikely to show a vergence response to horizontally orientated prisms until fusional vergence develops. One would expect the development of sensory fusion to be closely interlinked with motor fusion, and this appears to be the case. Several measures of cortical binocularity confirm that sensory fusion is, on average, first found at the age of 3–4 months (Birch et al 1985). The ultimate goal of binocularity is stereoacuity and it is not surprising that research techniques have shown that stereoacuity is initially absent and then develops abruptly and rapidly from the age of about 3–4 months (Birch et al 1985). Clinically, stereopsis is detected at different times using different tests; these are discussed later in this chapter.

Cortical cells require an input from both eyes if they are to become the 'binocularly driven' cells of the normal adult system. This must occur during the critical period. A strabismus or occlusion of one eye will prevent this bifoveal stimulation and, if not checked during the critical period for this function, binocular vision may never be possible as the binocularly driven cells will not develop but will become monocular cells. Nelson's (1988a) review of the literature on the 'risk of binocularity loss' concluded that 'relative plasticity' was at its maximum at age 1–3 years. The plasticity then reduced rapidly at first (to 50% of its maximum at age 4 years) and then more gradually, to 20% of its maximum at age 6 and about 10% at age 8. Obviously the 'infantile esotropia syndrome' (Ch. 15), with its onset under the age of 6 months, needs early referral. If such patients are not seen until they are over a year old, it is very unlikely that anything other than a cosmetic operation will help. Indeed, the prognosis for achieving binocular vision in infantile esotropia is never very good.

Accommodation

Accommodation is probably present from birth but is initially (until the age of about 3 months) inaccurate and principally operative over a short range (about 20–75 cm). It is thought that the main constraints on accommodative function in infants are attention and detection of the blur signal. Under ideal conditions of attention, accommodative function varies from one infant to another (Hainline 2000), but is probably good enough to give them the acuity that their sensory system can resolve (Aslin 1993).

Examination

During the examination young children should usually sit on the carer's knee, where they will feel more secure in otherwise strange surroundings. The mother may be able to help in eliciting the child's attention when required, or in steadying the head. Give time for the patient to get used to the situation while you are taking the history and symptoms from the mother. Try to relate to the child in a friendly way at appropriate moments before carrying out any 'tests'. Where possible, tests are presented as games to be played. A third adult in the room may be of help in holding test cards and fixation targets for distance vision. Do not darken the room unless absolutely necessary, and then it is better to adjust the lighting slowly. It is sometimes advantageous to wear informal clothing, avoiding clinical white coats. Picture books may be useful and small attractive toys to hold attention for fixation are necessary, preferably ones that can be 'squeaked' to give reinforcement. It is also useful to have toys in the reception area and ideally a children's area, with a small table and chairs, as this helps children to feel at home from the beginning of their visit. For toddlers, it is useful to have a car booster seat to place on the consulting room chair.

Methods and equipment

As explained above, the normal routine examination is not appropriate for children under the age of 5 years. We therefore need to decide exactly what we want to know as a minimum, and the quickest way to arrive at an answer. It is important to be quick and to frequently change tests to maintain interest. If a strabismus is suspected, the priorities are to ask the following questions:

(1) Is binocular vision present?
(2) Are there any signs of a strabismus?
(3) Is the unaided vision the same in the two eyes?
(4) Is the refractive error normal for the age?
(5) Is the corrected acuity normal for the age?

It may not be possible to answer all these questions for every child. With others, much more can be done in addition. A lot will depend on the level of cooperation of the young patient. Great patience and more than one visit may be required. The following procedures are typical of those that can be used in preschool children. The order of testing will depend on the child's cooperation and the practitioner's personal preference.

Vision and visual acuity

A variety of clinical tests allow the practitioner to measure uncorrected vision and visual acuity in children of any age, although monocular testing is particularly difficult at about 1–2 years (Shute et al 1990). For infants, preferential looking cards are usually the best method of assessment (Fig. 3.1), and a single presentation is used when the looking behaviour is clear, with a maximum of three presentations when equivocal (McCulloch 1998). Classic grating pattern preferential looking cards, such as the Keeler acuity cards or Teller cards, are required for infants below the age of about 6 months. After this age children become bored with these tests (Teller 1990) and, especially over the age of 1 year, children usually respond to the more interesting vanishing optotype cards (Cardiff Acuity Test; Adoh & Woodhouse 1994). The Cardiff test is not good at detecting amblyopia (Geer & Westall 1996), so crowded optotype tests should be used as soon as the child is capable.

Where preferential looking tests are not available, other acuity tests for infants are to observe behaviour when one eye is covered, optokinetic nystagmus and the 10 Δ base down test. In this test a 10 Δ lens is introduced vertically in front of one eye while the patient fixates an accommodative target. Spontaneous alternation of fixation should occur or, if one eye is preferred, then the non-preferred eye should maintain fixation for at least 5 s if the preferred eye is covered (Mehta 1999).

By about the age of 2 years, many children can do picture matching tests in which they are required to match a distant picture with the correct one from a range of large pictures held close to. The Ffooks tests requires children to discriminate between a circle, a triangle and a square, but even with this it must not be assumed that a child can necessarily tell the

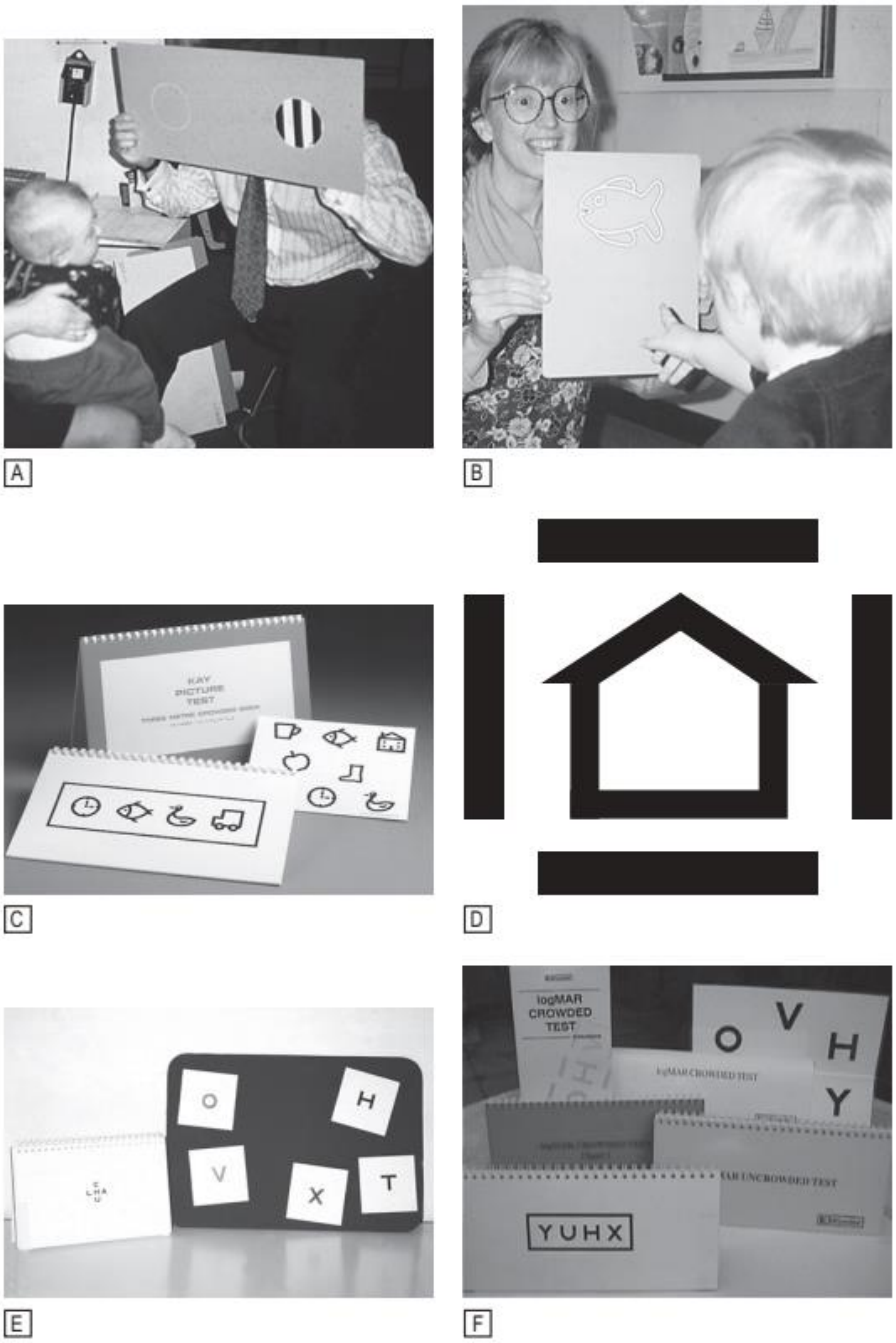

Figure 3.1 Some visual acuity tests used with children. (A) Preferential looking grating test (Courtesy of Dr Simon Barnard). (B) Cardiff acuity cards. (C) Kay 3 m crowded book (Courtesy of Hazel Kay). (D) A Lea symbol presented in a crowding box on Test Chart 2000 (reproduced with permission from Thomson Software Solutions). (E) Cambridge Crowding Test. (F) Glasgow Acuity Card.

difference between these shapes and it may be necessary to teach the patient to do so first. Other tests, for example those designed by Kay and Lea, use pictures constructed on Snellen principles and one version of the Kay test (Fig. 3.1C) uses a crowded design (below). Inevitably, test results are affected by patient cooperation; however, if monocular acuities with Lea single optotypes are possible, then each eye's result should, in 90% of cases, be within one line of the other eye (Becker et al 2002).

By the age of about 2.5–3 years most children can carry out tests where they are required to match letter acuity targets with large reference letters in a chart or book held close to. The traditional test of the type, the Sheridan–Gardiner test, employs single letters. A major disadvantage of this test, and of any test using single optotypes, is reduced sensitivity in detecting amblyopia because of the crowding phenomenon, or contour interaction. These terms describe the effect of adjacent contours in reducing target detection. Contour interaction, which is particularly marked in strabismic amblyopia, occurs when an adjacent contour is placed at a distance equivalent to the width (or diameter) of the target letter and is maximal when adjacent contours are 0.4 letter diameters away. A linear version of the Sheridan–Gardiner test, the Sonksen Silver test, demonstrates some sensitivity to the crowding effect. However, the tests that are most sensitive to this effect are the Cambridge Crowding Cards and the logMAR Crowded Test (Glasgow Acuity Cards). The latter test also benefits from several other design features that make it particularly well suited to the detection of change in visual acuity (McGraw & Winn 1995) and compares well with conventional logMAR charts (McGraw et al 2000). The Kay Crowded Picture Test is of similar design but using pictures instead of letters. These two tests have been shown to produce comparable results (Jones et al 2003) and both can be reproduced on the computerized Test Chart 2000 (Thomson 2000).

Indeed, the greatest flexibility for testing the vision of children from about the age of 2 years is to use the computerized Test Chart 2000 (Fig. 3.1D). This allows the practitioner to choose one of a variety of optotypes (e.g. Lea symbols, Kay pictures, Makaton symbols, lower case letters, numbers) and allows for the optotypes to be presented individually with crowding bars. This latter method of presentation may be ideal for young children since it achieves the simplicity of single optotype presentation without sacrificing crowding. Computerized charts also allow practitioners to randomize the optotypes, which is very helpful to prevent children from memorizing the chart.

Various visual acuity tests are illustrated in Figure 3.1 and norms for visual acuity tests are given in Table 3.1 and summarized in the clinical worksheet in Appendix 2. Visual evoked potential measures of acuity are not included, since they are not generally used in primary eyecare practice, but they tend to give higher estimates than preferential looking, with acuities as high as 6/9 at 6–12 months (Teller 1990).

Binocular vision

Cover test A cover test is usually possible in most infants if a sufficiently 'interesting' target is used (e.g. brightly coloured squeaky toy). As children become older more detailed targets, and more accurate results, can be obtained. The palm of the hand or thumb is used for occlusion rather than the usual 'occluder', which distracts attention. Sometimes it is obvious that the patient objects to one eye being covered but not the other, suggesting a difference in acuity.

The most common deviation in young children, which usually presents in the first 6 months of life, is the 'infantile esotropia syndrome' (Ch. 15). It usually has a large angle, over 40 Δ, which is the same for distance and near fixation. There is often a higher degree of hypermetropia than normal for the age and the deviation may be partially accommodative. There is usually crossed alternating fixation; i.e. the right eye tends to fixate objects in the left part of the visual field and the left eye fixates in the right field. The change of fixation can be seen if the patient can be persuaded to follow a target moved across the horizontal. It seems that about half of patients have amblyopia with eccentric fixation (Dale 1982). There may be latent nystagmus (Ch. 18), and sometimes dissociated vertical deviation (Ch. 9).

Congenital exotropia is rarer than congenital esotropia and is different in that congenital exotropia is typically present from birth. The angle is usually fairly large and constant and may be associated with neurological abnormalities (Ch. 15).

Another infantile anomaly is the 'nystagmus compensation (or blocking) syndrome' (von Noorden 1976). In this condition, the convergent strabismus seems to be adopted in order to lessen nystagmoid movements, which are reduced on convergence of the eyes. The patient's head is usually turned away from the side of the fixating eye so as to produce further convergence of this eye, and there may be the appearance of a lateral rectus palsy (Ch. 17). Neither of these conditions can be treated by refractive or orthoptic means alone and a surgeon's opinion should be taken as soon as possible.

With some young children a reliable cover test result cannot be obtained and other methods of assessing ocular alignment are required. Three such methods are described below.

Hirschberg's method and Krimsky's method The degree of deviation may also be estimated by the Hirschberg's method, using the position of the corneal reflection of a pen torch. Figure 3.2A shows, for two deviations, how the reflection of a light in the cornea appears displaced from the centre of the pupil by 1 mm for each 20 Δ (11°) of deviation of the eye (various estimates range from 7° to 15°; Spector 1993, Pearson 1994). Figure 3.2B shows the appearance for a left divergent strabismus of about 20 Δ. Figure 3.2 assumes that the angle lambda (see Glossary) is zero, and the reflex is central in the right undeviated eye. Angle lambda at birth is typically 13.75 Δ, reducing to 9 Δ by 5 years old (Pearson 1994). The position of the reflex in the non-amblyopic eye (with the other eye covered) should be noted before the angle of strabismus is estimated binocularly by judging the difference in position of the reflex in the strabismic eye. In a right convergent strabismus with a large angle lambda, the reflexes would therefore appear in the same positions as those shown in Figure 3.2B.

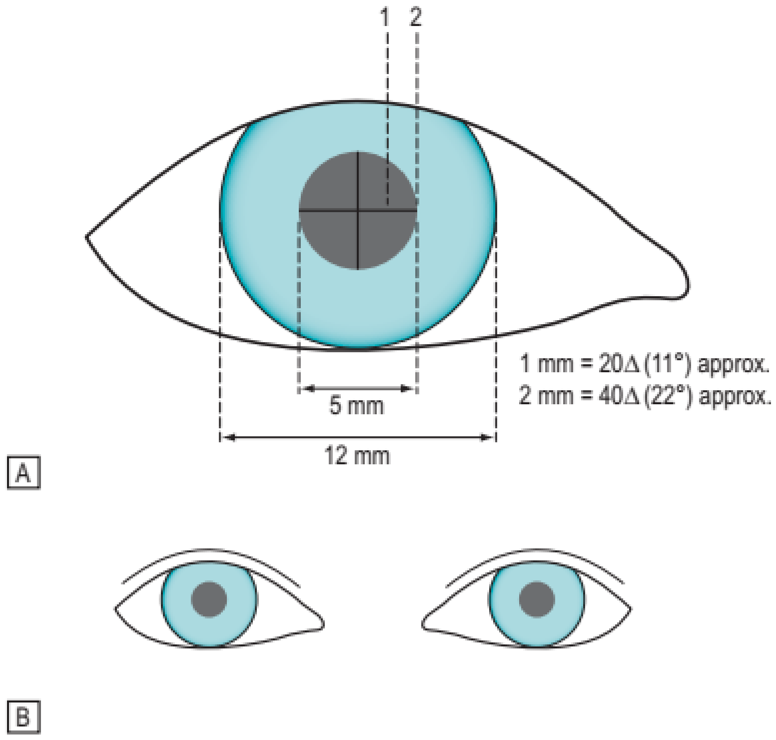

Figure 3.2 Hirschberg's estimation method for the angle of strabismus. See text for details.

Krimsky's method is a modification of Hirschberg's method where prisms are placed in front of the deviating eye until the prism power is found that makes the corneal reflexes appear to occupy the same relative positions (Krimsky 1943). These methods are imprecise: deviations of up to 14 Δ may be overlooked (Spector 1993).

Bruckner's test The practitioner views both eyes through a direct ophthalmoscope at a distance of 75–100 cm. If the fundus reflexes are equally bright then this suggests that there is no strabismus. If they are not equally bright, then there is either a strabismus (in the eye with the brighter reflex), pigmentary difference, unequal pupil size or anisometropia (Griffin & Grisham 1995, p 107). Von Noorden (1996) said that asymmetric fundus reflexes may be normal in infants up to the age of 10 months.

Motility The presence of an incomitancy will increase the risk of pathology being present. It is usually possible to check for incomitancy by holding the child's head still and attracting the attention so that the eyes turn into the tertiary positions of gaze. With infants it is generally better not to hold the head but to move the motility target further than usual and to allow patients to move their heads. Alternatively, the parent can rotate the child, whose attention is held on a stationary fixation target.

Convergence and motor fusion Convergence can be assessed by moving the target towards the nose after near cover testing.

Motor fusion can be assessed by testing for convergence when base-out prisms are introduced monocularly. A convergent movement of the eye shows that binocular single vision, at least in the periphery (Kaban et al 1995), is present; a versional movement suggests that the other eye is not fixating and the prism should be tried before this eye. If no movement is seen, it suggests that binocular vision is doubtful. Infants should be able to overcome 20 Δ base-out by 6 months of age (Riddell et al 1999). By the age of 4–5 years it should be possible to measure full fusional reserves; the eyes should be observed to check the break and recovery points objectively. Some paediatric norms for fusional reserve tests can be found in Appendix 2.

Stereoacuity Sensory fusion can be assessed with tests requiring stereopsis. A range of stereoacuity tests are illustrated in Figure 3.3.

About half of 6-month-old children and 80% at 9–17 months give a positive response to the Lang stereotest. However, only 65% of 9–11-month-old infants give positive responses to the Frisby stereotest, although by the age of 2 years 100% of children with normal vision responded to the Frisby and Lang stereotests (Westall 1993). Successful stereoacuity testing is possible in virtually all children at the age of 3 years (Shute et al 1990). Several studies have suggested that the random dot E test is particularly good for screening (Hammond & Schmidt 1986, Ruttum et al 1986, Pacheco & Peris 1994, Schmidt 1994), although the method of use is important (Fricke & Siderov 1997). Specifically, patients should be asked on which plate the 'E' is present and not just which plate looks different.

Romano et al (1975), using the Titmus test, found that at age 3.5 years the lower limit of normality is 3000″. At 5 years it had improved to 140″, and did not reach a normal adult value of 40″ until the age of 9 years. It appears to be a little better with the TNO test. With conventional clinical tests, the improvement from 18 months to 5.5 years may be the result of improved attention rather than changes in neurophysiology (Ciner et al 1991). Heron et al (1985) found that with the Frisby, Randot, TNO and Titmus tests adult-like levels are reached between the ages of 5 and 7 years.

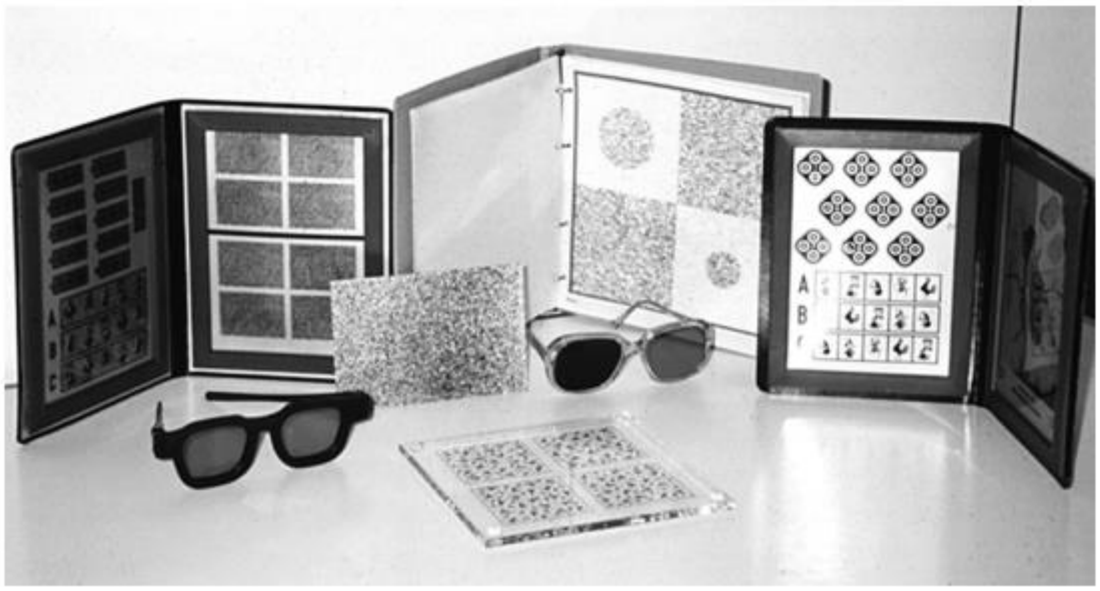

Figure 3.3 Stereoscopic tests. In the foreground is a plate of the Frisby test and in the background, from left to right, the Randot, Lang, TNO and Titmus tests (sometimes called the Wirt test, which was in fact a precursor of the Titmus circles test).

The Lang stereotest detects every case of constant large-angle strabismus, 90% of cases of microtropia and 65% of cases of anisometropic amblyopia (Lang & Lang 1988). The future of stereoacuity testing for young (Ciner et al 1996) or handicapped (Ciner et al 1991) children may lie with preferential looking methods. Using a preferential looking technique, Schmidt (1994) found that the random dot E test was better for screening preschool children than a visual acuity stimulus. Preferential looking stereoacuity cards are not widely available at present but show promise as a test for children in the first year of life (Calloway et al 2001).

The distinction between global (random dot) and contoured stereotests was discussed on page 33. Some norms for stereotests for young children can be found in Appendix 2.

Ocular health

As some strabismus in children under the age of 5 years may be due to a pathological cause, it is very important to do everything possible to examine ocular health. For example, the palpebral openings should be noted: the two lid openings should be equal and symmetrical. Any inequality in a strabismic child may indicate a growth of extra tissue in the orbit. This would push the eye forward and disturb the eye movements. It could be caused, for example, by a dermoid cyst, sarcoma of the muscles or glioma of the optic nerve.

An attempt should be made to look at the media and fundus but, even if a full ophthalmoscopic examination is not possible, look at the colour of the fundus reflex. This may be done with an ophthalmoscope or with a retinoscope moved around so that all areas of the fundus are checked: the retinoscope gives a narrow concentrated beam of light and can be used from a greater distance, which may more readily be tolerated by the patient. White areas of the fundus can be a sign of retinoblastoma, and a strabismus may be the first sign of this condition, which usually begins before the age of 4 years. A white reflex, whatever the cause, requires urgent referral: even congenital cataract should be removed as early as possible, ideally within 6 weeks of birth (Birch & Stager 1995).

Usually, in children, the pupil is large and the media clear, which helps in seeing the fundus. Indirect ophthalmoscopic techniques may be particularly appropriate to get an overall view of the fundus quickly, and mydriasis may be required in some cases. Indirect ophthalmoscopy facilitates comparison of the size of the optic discs, particularly when a graticule is used. Optic nerve hypoplasia is a cause of poor vision that may be difficult to recognize and hence may be incorrectly diagnosed as amblyopia (Ch. 13).

In infants where the eyes appear large or there is the suspicion of asymmetry or of pale discs, it is useful to record the horizontal visible iris diameter. This is likely to be abnormal in infantile glaucoma and norms are given in Appendix 2. Other signs of infantile glaucoma include hazy, opaque corneas and the triad of epiphora, blepharospasm and photophobia.

If an infant is uncooperative on a particular day and a detailed inspection of the fundus is not possible, then a follow-on appointment can be arranged in the near future.

Refraction

Initially, we need to discover if there is any significant refractive error, rather than to measure the exact refraction. Refractive errors outside the normal ranges (see Appendix 2) indicate a need for further investigation.

Lens racks are designed for use with children who are too young to wear a trial frame. The most common method for refracting infants is to hold single trial lenses before the eye and this is reasonably effective. Refractor heads and phoropters are not appropriate to young children but can sometimes be used from the age of about 4 years if introduced in a child-friendly way.

The refraction will need to be carried out objectively. For example, hold a single trial case sphere before one eye and observe the type of movement of the reflex with the retinoscope. The power of the sphere will be the average refraction for the age, plus 1 DS, plus the 'working distance' lens. It is better to work at a distance of 0.5 m for very young patients. An 'against' movement may indicate less hypermetropia or that the patient is accommodating. A cycloplegic refraction can follow at a second visit by the patient. The corrected acuity can also be measured. During the cycloplegic examination, the patient can be encouraged to fixate the retinoscope light.

There is some debate over when to carry out a cycloplegic refraction. On one extreme, some authors recommend carrying out a cycloplegic refraction to determine the full refractive error as a matter of course in the majority of young patients; others argue that a cycloplegic refraction represents an abnormal state and is hardly ever appropriate in optometric practice. A sensible 'middle-ground' approach is not to perform a cycloplegic refraction on every child but rather when a preliminary examination reveals one of the risk factors in Table 2.3. In infants a greater reliance will be placed on the objective signs in Table 2.3. Even when a cycloplegic refraction is performed, it is still important for the practitioner to know the non-cycloplegic refraction, since this can influence the sensory status (Kirschen 1999). Patients under the age of 3 months who need a cycloplegic refraction are best examined in the hospital environment because of the risk of systemic effects of the cycloplegic agent (Edgar & Barnard 1996).

Some practitioners use Mohindra's technique of retinoscopy as an alternative to cycloplegic refraction (Mohindra & Molinari 1979). This is carried out in darkness with one eye occluded and with the child fixating the retinoscope light. To take account of the patient's accommodation, for a working distance of 0.5 m an allowance of 0.75 D is subtracted from the result (Stafford & Morris 1993).

A fairly reliable objective assessment of the refractive state can also be obtained by photorefraction. This term covers three different techniques, which all involve the analysis of a photograph of the image of a flash source that has been refracted on entry and exit from the eye (Thompson 1993). The sizes of the photographed light patches depend both on the defocus of the eye relative to the camera distance and on the pupil diameter, so that a computerized system can calculate the refractive error from the reflex sizes. Photorefraction is claimed to be particularly useful for screening large numbers of infants, although it may lack sensitivity for detecting anisometropia (Fern et al 1998).

Management

First, the question of a referral for medical investigation should be considered, and this is essential if there is any doubt about the presence of pathology. It should also be recommended where it is clear that the circumstances would not allow a reasonable prognosis for refractive and orthoptic treatment by the optometrist. If the patient is going to require medical attention, the sooner the referral the better. Care should be taken not to delay other treatment when it is clear that the condition will not respond to the methods available in a primary care setting.

Preschool children are often too young to cooperate with any form of orthoptic exercises. Some can carry out simple exercises and a range of suitable exercises are described by Wick (1990). Refractive error should be corrected if possible, and amblyopia may need treatment by occlusion (Ch. 13). The importance of correcting significant hypermetropia to prevent amblyopia is discussed in Chapter 13.

Modern methods of acuity assessment in very young children suggest that vision develops at a much earlier age than previously believed. This emphasizes the need for examination as early as possible, so that measures can be taken to prevent the failure of the development of acuity. Children with a parent (or other close relative) with amblyopia or strabismus are particularly at risk of amblyopia. It is therefore important that these children should be seen as early in life as possible and that young adult patients with amblyopia should be advised that their children's eyes need to be examined.

The general principles of strabismus management are described in Chapter 15.

Clinical Key Points

- Precise results are usually not possible with young children and frequent appointments are often appropriate
- Visual acuity is readily assessed in young children by preferential looking and norms vary for different tests
- Refractive errors are very variable in the first year, but at the age of 1 year more than 3 D of hypermetropia is a risk factor for strabismus
- Binocular functions, including stereoacuity, are usually present by the age of about 4 months
- Always attempt a cover test and motility test. Stereotests and the 20 Δ base-out prism tests are also useful