

The assessment and management of strabismus and amblyopia: a national audit

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Abstract

Aims To determine what systems are in place within ophthalmic services for the assessment and management of children suspected of having amblyopia and strabismus. To find out what methods are used for the assessment of these children.
Methods A questionnaire-based study auditing 288 orthoptic departments in the UK.

Results Responses were received from 75% orthoptic departments. Most hospitals employ more than one system for the assessment of strabismus and amblyopia, which is generally dependent on route of referral. These include 'orthoptic assessment without refraction' (66%), 'combined orthoptist and ophthalmologist assessment' (66%), while 22% have an entirely orthoptist/optometric system.

Ophthalmologists are involved in the initial assessment in 145 units (67%), whereas some units involve an ophthalmologist only if response to treatment is poor (15%), or if surgery is required (6%).

Fourteen per cent of units reviewed all children, with discharge criteria being based on normal visual acuity (52%), accurate visual acuity (39%) and a normal orthoptic assessment (42%). Seventy-six per cent of units review some children, commonly as a result of family history (55%), parental concern (43%), poor co-operation (30%) and young age (72%). In the absence of squint or amblyopia children are discharged at the first visit, in only 8% of units.

There is considerable variation in the tests used to assess visual acuity. LogMAR-based tests (eg EDTRS) are not routinely used in 75% of units.

Conclusion Different systems exist for the

assessment and management of squint and amblyopia across the country. While much of this variation is to be expected given their possible aetiologies, some could be reduced to produce a more cohesive service. There is also considerable scope for rationalising the tests used to screen infants and children for amblyopia and strabismus.

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Introduction

Strabismus and amblyopia are the commonest ophthalmic problems affecting young children with prevalences of $\approx 5\%$ of 5 year olds for strabismus,^{1–6} and 1.2–4.4% for amblyopia.^{1,7} It is a strongly held tenet, based on the concept of the sensitive period for amblyopia which ends at around 7 years, that the early treatment of amblyopia is critical and is less likely to be effective if delayed until after school entry.^{8–11} However, while the presence of a cosmetically obvious strabismus generates referral at any age, in amblyopia not associated with an obvious deviation the diagnosis can be delayed.^{12,13}

UK-wide preschool vision screening was advocated in 1976¹⁴ and national preschool vision screening for amblyopia and strabismus identification was set up for children at around 3.5 years of age in 1978. By 1988, 99% of National Health authorities had a screening programme in place.¹⁵ However, in some areas screening has never been established or has recently been disbanded—perhaps in response to the Systematic Review of 1997.¹⁶ Controversy still exists over the need for

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screening,¹⁵⁻¹⁷ the age at which it should be carried out^{15,18,19} and the use of orthoptists¹⁹⁻²² within this programme. However, the Child Health Screening Sub-group of the UK National Screening Committee recommends vision screening by an orthoptist of all UK children at age 4.5-5 years of age.¹⁹ This group also recommends that all children with major disabilities require a full ophthalmic assessment. Currently therefore in the UK amblyopia and strabismus are identified by: population screening by health visitors or orthoptists, referrals via child health surveillance programmes, or referrals by GPs and clinical medical officers (consisting of cover tests with or without visual acuity). Orthoptists may undertake primary screening or assess children in secondary referral community clinics. Some areas have no screening for strabismus and amblyopia.

Children identified by primary population screening are then referred either to an orthoptic or ophthalmic clinic. Apart from population screening, children suspected of having amblyopia or strabismus may be referred to the hospital eye service at any age, by general practitioners, community or hospital paediatricians, health visitors, school nurses or optometrists. Thus there are a number of referral routes by which children with suspected amblyopia or strabismus reach the ophthalmic service, and once there, we have anecdotal evidence that the methods of assessment and systems for management differ.

We have undertaken a survey to determine what systems are in place within ophthalmic services for the assessment and management of children suspected of having amblyopia and strabismus. We also enquired about methods of assessment of these children.

Methods

A questionnaire was sent to every UK orthoptic department on a list compiled by the British Orthoptic Society (BOS) in 1994. The questionnaire was divided into five sections:

Part 1 Requested information about systems currently in place for the assessment of strabismus and amblyopia, and the reasons for choosing them.

Part 2 Enquired about the tests used routinely in the initial assessment of children, eg methods of measuring visual acuity.

Part 3 Enquired about the management of children who had no evidence of strabismus or amblyopia at the first visit.

Part 4 Enquired about whether an entirely orthoptist/optometric service had been considered for the management of children suspected of having strabismus or amblyopia. Reasons for not wanting such a system were then asked for.

Part 5 Enquired about the involvement of an ophthalmologist in the management of children with amblyopia or strabismus.

Results

The British Orthoptic Society list contained 330 orthoptic departments. Since 1994 when the list was compiled, 42 units had either closed down or merged with larger units, leaving a total of 288 orthoptic units in the UK. Questionnaire responses were received from 217 departments, a response rate of 75%.

Part 1 requested information about systems currently in place for the assessment of strabismus and amblyopia, and the reasons for choosing them. Most hospitals have more than one system for the assessment of strabismus and amblyopia, which is generally dependent on the route of referral. The most common are 'orthoptic assessment without refraction' (152 (70%)) and 'orthoptist and ophthalmologist assessment' (149 (69%)) (Table 1). Each unit had considered a number of factors when choosing a system. One hundred and forty-two units had chosen a system because they thought it 'worked well'. Thirty units felt that the system in place had resulted from a shortage of ophthalmologists. Forty-four units had considered 'value for money' when devising their system (Table 2).

Part 2 enquired about the tests used routinely in the

Table 1 Systems in place for the assessment of children with suspected strabismus or amblyopia. Some departments have more than one system in place, thus the total exceeds the 217 respondents

	<i>Number</i>
Orthoptist assessment without refraction	152
Orthoptist assessment with autorefraction	45
Orthoptist assessment with retinoscopy by orthoptist	16
Orthoptist and ophthalmologist assessment	149
Orthoptist assessment and refraction by optometrist in same hospital clinic	81
Orthoptist assessment and refraction by optometrist in same community clinic	33
Orthoptist assessment and subsequent referral to optometrist practice for refraction	43
Orthoptist assessment and referral to ophthalmic medical practitioner in community	23

Table 2 Reasons cited for choosing the systems currently in place for the assessment and management of strabismus and amblyopia. Respondents were allowed to choose more than one reason

	Number
Tradition	62
Continuity of care	63
Works well	142
Shortage of ophthalmologists	30
To improve on previous system	79
Value for money	44
No other option	32

initial assessment of children, eg methods of measuring visual acuity. The initial orthoptic assessment of a suspected case of amblyopia/strabismus was fairly uniform and included cover test, ocular movement, motor fusion and stereopsis.

There was considerable variation across the country in the assessment of visual acuity (Table 3). Twenty-one units use logMAR (EDTRS) to assess visual acuity in children >2.5 years. A large number of units ($n = 171$, 79%) attempt to measure the visual acuity of children under 1 year old as part of their routine assessment for amblyopia. Most units ($n = 186$, 84%) regularly measure the acuity of children in the 1–2.5 year old age group. Kay’s pictures are the most common method in use in this age group ($n = 182$), but other methods commonly in use include Cardiff cards ($n = 49$), and the Sheridan Gardiner test ($n = 48$) and 100s and 1000s ($n = 20$).

Ophthalmologists and/or optometrists are involved in refraction and fundus checks in 182 and 178 units respectively (Table 4). Some units do not undertake refraction or fundoscopy ($n = 32$ and $n = 36$, respectively), but these are community orthoptic clinics, which refer children with any problems to a hospital-based clinic for further evaluation.

Table 3 Tests of visual acuity used in the assessment of children and the age at which they are routinely carried out

Test of visual acuity	Age test performed			
	<1 year	1–2.5 years	>2.5 years	Not done
Acuity card procedure	84	1	0	129
Cardiff cards	138	49	0	25
Kays pictures	1	182	5	4
Keeler	5	4	6	174
LogMAR (EDTRS)	0	2	19	163
Sheridan Gardiner	0	48	125	14
Snellen	0	3	151	27

Table 4 Professionals responsible for carrying out refraction and fundoscopy within a department

	Refraction	Fundoscopy
Ophthalmologist	96	127
Optometrist	40	10
Either ophthalmologist or optometrist	46	39
Not done	32	36

Part 3 enquired about the management of children who had no evidence of strabismus or amblyopia at the first visit. Eight per cent (18) of units would discharge a child who was found to have no strabismus or amblyopia at the first visit, while 77% (164) review some children and 15% (31) review all.

In the 174 orthoptic units continuing to follow up some children, criteria for further follow-up included family history ($n = 90$), parental concern ($n = 70$), poor co-operation ($n = 50$) and age of child ($n = 45$) (Table 5). Where age was used as a criterion for further review, 22 units continued until the age of 3.5 years, 21 units until 2.5–3 years, three units until 2 years and 19 units failed to specify a particular age for discharge. Criteria for follow-up in units reviewing all children were: obtaining an accurate visual acuity ($n = 12$), a normal visual acuity ($n = 16$) and a normal orthoptic assessment ($n = 13$).

Part 4 enquired about whether an entirely orthoptist/optometric service had been considered for the management of children suspected of having strabismus or amblyopia. Reasons for not wanting such a system were then asked for. Forty-seven (22%) units in the UK have an entirely orthoptic/optometric management plan. One hundred and twenty-six units did not feel that this system was appropriate stating that orthoptists were not qualified to refract or conduct fundoscopy ($n = 71$, 56%), no optometrist service was

Table 5 Criteria used for the continued follow-up of children following a normal orthoptic assessment. Units are divided into those that continued to follow up all children and those that only reviewed some

	Reviewed all	Reviewed some
Family history	4	90
Accurate visual acuity	12	26
Parental concern	3	70
Poor co-operation	2	50
Age	7	45
Refractive error	4	12
Normal visual acuity	16	26
No evidence of a heterophoria	0	3
Normal orthoptic assessment	13	20
History of strabismus	0	2

available ($n = 57, 45\%$), and resistance from ophthalmologists ($n = 49, 39\%$) (Table 6). No unit volunteered resistance from orthoptists.

Part 5 enquired about the involvement of an ophthalmologist in the management of children with amblyopia or strabismus. Ophthalmologists are involved in the initial assessment of children in 145 units. Thirteen of these units only saw an ophthalmologist again if they failed to respond to treatment ($n = 7$), needed surgery ($n = 5$), or had poor visual acuity ($n = 1$). Thirteen units only involved an ophthalmologist if surgery was required. Thirty units involved an ophthalmologist if the response to orthoptic treatment was poor: defined as a visual acuity of 6/9–6/18 ($n = 23$), or 6/24 or worse ($n = 7$) in the amblyopic eye. Twenty-two of the 47 units who had an entirely orthoptist/optometric management plan also had an alternative system in which an ophthalmologist was involved at the initial visit.

In retrospect, we appreciate that certain sections of the questionnaire contained ambiguities that were not identified in the initial piloting of the questionnaire. These sections have been excluded from analysis and are not presented here.

Discussion

This study highlights diversity in two aspects of the assessment and management of amblyopia and strabismus: first in the systems in place for assessment and management, and second in the techniques used to assess infants and young children suspected of having these conditions.

Children suspected of having amblyopia or strabismus enter the ophthalmic service predominantly by three routes: preschool screening, referral of an asymptomatic child from an at-risk group (eg, ex-preterm, neuro-developmental delay and a child with a disability), and symptomatic referral. The last named can arise from a number of community or hospital healthcare sources.

Table 6 Reasons for not wanting to adopt an entirely orthoptic/optometric run system

	<i>Number</i>
Insufficient number of qualified orthoptists	10
Orthoptist not qualified in refraction and fundus examination	71
No optometrist service available	57
Resistance from ophthalmologists	49
Resistance from optometrists	10
Parental expectation to be seen by a doctor	18
Not safe	19
Other	0

Once within the ophthalmic system how the child is assessed and subsequently managed also varies. The ophthalmic team contains a number of professions (ophthalmologist, orthoptist, optometrist and nurse) whose roles differ across the country according to local practices, as shown in this survey. Given these differing routes of referral, service structure and the diversity of the population, it is not surprising that many ophthalmic departments have more than one assessment and management scheme. Indeed this is entirely appropriate. For instance, the healthy young strabismus suspect is ideally assessed by an orthoptist working with an optometrist, whereas the child with neurodevelopmental delay requires a full ophthalmic evaluation. Children within at-risk groups are reviewed as clinically indicated and they will not be considered further.

The ensuing discussion focuses on the issue of what the management plan should be for the healthy infant or child who has no evidence of strabismus or amblyopia at an initial assessment. Our survey showed that only 8% of units felt justified in discharging these children at the first visit, while the majority continued to monitor children, in particular those with a family history, refractive error and parental concern. This review process places considerable demands on the family and ophthalmic resources, so it is now pertinent to consider the role of these risk factors in determining the need for follow up. The links between these three factors and strabismus are well known, however their predictive value is less certain.²³ The questions arise: How can we determine at risk children from a single evaluation? What tests are required to identify risk factors and at what age are these risk factors most significant?

This survey sought information about the techniques of assessment of the infant and child suspected of having amblyopia and/or strabismus. Here we will focus on the measurement of visual acuity and refractive state. Measurement of visual acuity in young children is the Achilles heel of paediatric ophthalmology, fully reflected in this study by the wide range of tests being utilised. Acuity testing at the age of 2 years results in a high rate of false positives (86.3%) compared with that at the age of 3 years (54.7%).²⁴ It is well established that grating tests such as the acuity card procedure underestimate the depth of amblyopia compared to tests of recognition acuity.^{25,26} To measure amblyopia precisely, a crowded optotype test is necessary. Although accurate Snellen acuities are obtained in 80% of children at the age of 3 years, this increases to 90% by 3.25 years.¹⁸ In 1976 Bailey and Lovie developed the logMAR test as an alternative to the Snellen chart in order to surmount its

All Health Visitor and GP referrals for suspected strabismus over 9 months of age:
(excluding neurodevelopmental delay and expreterm children)

<p>Positive family history / Continued parental concern Positive cover test* Abnormal ocular movements* Visual Acuity of <6/6 &/or 1 line difference at ≥ 2.5 years** Stereopsis reduced*** Fusion absent**** Any other pathology Refraction (cyclopentolate 1%)***** Myopia Hyperopia $\geq +2.50$DS Astigmatism ≥ 1.00 Anisometropia >0.75 DS</p>	}	<p>If yes to any of these refer to Children's Eye Clinic. If no to all of these then discharge</p>
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*Decision about which cover test and eye movement abnormalities require referral is a clinical judgement.

**Visual Acuity: Single optotype letters are used. If unable to do this then use Kay's.

***Stereopsis: < 3 years <550 secs (lang). ≥ 3 years <100secs.

****Fusion: <3 years +/- to 20 dioptres; ≥ 3 years ≥ 20 dioptre PFR.

*****Refractive values allow for distance alone – nothing allowed for cycloplegia.

Figure 1 Fast track clinic protocol used at the Children's Eye Service, St Mary's Hospital, London.

shortcomings, ie irregular progression of letter sizes, variation in the number of letters on each line and unequal legibility of letters used.^{27–32} Accordingly, logMAR charts are now routinely employed in clinical vision scientific research and increasingly in routine clinical practice.³³ Detection rates for amblyopia using the logMAR test are twice those of Snellen charts, and reproducibility is superior to Snellen-obtained measures, yet only 25% of UK units have taken to using them.^{27,28,29,33,34} It is universally recognised that 'objection to occlusion' is of great value in identifying the presence of amblyopia in very young children. However, attempts to precisely quantify the visual deficit in a standard clinical setting, in children under about 2.5 years, is fraught with difficulty and is of debatable value, because testing generates many spurious results. Yet, our survey showed that such measurements are attempted in 171 units for children under 1 year of age and 186 units for children of 1–2.5 years of age. There is a need to rationalise visual acuity testing in young children for the purpose of identifying and quantifying amblyopia. In the meantime, the need for the continued review of children who are otherwise normal, on the basis of visual acuity alone should be questioned.

The association between hypermetropia and the

development of strabismus is well documented.^{35–38} However, studies linking refractive error (eg astigmatism, spherical error and anisometropia) with strabismus and amblyopia show that because of refractive changes during childhood, more than one measurement is required to determine a child's risk of developing amblyopia or strabismus.^{35,37} This is due to the process of emmetropisation which is largely complete by the age of 2 years but may continue until 4 years of age.^{35,36} The management of a child, with what is considered to be a borderline error under the age of 4, is controversial due to uncertainty over the significance of these findings. Relative risks have been assigned to certain refractive errors, however in terms of screening, hypermetropia in the order of 3.5 spherical dioptres or 2 dioptres in one meridian, are both risk factors for amblyopia.^{35–39} Increasing astigmatism, oblique astigmatism and presence of strabismus have the highest relative risk.^{36,39,40} The benefit of early detection and treatment of refractive errors has also been the subject of considerable debate,^{41–43} with evidence both for and against early correction.^{41,44} Thus, the appropriate management of a child with a refractive error depends on the age of the child, the type of refractive error and the presence or absence of strabismus.

The incidence of strabismus increases to 17.6% (vs 3–4%) when a positive family history is elicited.³⁵ The risk of developing esotropia in patients with a positive family history is increased four-fold in the presence of hypermetropia.³⁶ Rosner and Rosner showed that given superficial instruction, parents can detect 66–76% strabismus,^{45–47} although in the absence of strabismus (as in anisometropia) only 17% of children with amblyopia were noticed by their parents.⁴⁸ While the predictive value of family history, in the absence of other amblyopia risk factors, is not known, a positive family history of amblyopia is probably a valid reason on its own for reviewing the amblyopia and strabismus suspect. The healthy child without a positive family history, or evidence of refractive error, strabismus or amblyopia on initial assessment is most unlikely to develop amblyopia or strabismus.

This survey has highlighted several professional issues. While it is recognised that local issues influence professional roles, it is unlikely that they account for all the variations reported herein. Forty-seven units have a completely orthoptic/optometric run system, but many units were reluctant to adopt such a system. Reasons given included othoptists not being trained in refraction, lack of optometry services and resistance from ophthalmologists. Most units still prefer to have a consultant ophthalmologist involved at the start of treatment—usually to confirm the absence of ocular pathology. Ophthalmologists tend to be involved at the commencement of treatment, when there is a poor therapeutic response, and if surgery is to be considered. Ophthalmologists are relatively infrequently involved in children with suspected strabismus and the management of amblyopia and accommodative esotropia.⁴⁹

Studies have shown that orthoptists are the most effective screeners of strabismus and amblyopia^{20,21,50,51} and given suitable training they are also able to refract.⁵² While the debate on preschool vision screening continues, primary screening backed up by secondary orthoptic/optometric assessment has been shown to substantially decrease the number of inappropriate referrals to hospital clinics.^{33,51,53–58} Where this occurred, only 14% were referred to the hospital clinic, of whom 76% were referred back to the community for subsequent full or partial management.⁵⁴ Such schemes have also demonstrated that children are referred earlier for treatment (average age decreased from 6.6 years to 5 years) and that the relationship between social deprivation and the age of presentation of asymptomatic amblyopia is abolished.¹²

Conclusion

There is considerable variation across the country in the way that we assess and review children suspected of harbouring strabismus and/or amblyopia. Therefore many children continue to be followed up despite having no ophthalmic abnormality. This places a considerable burden on the NHS and is of no benefit to the child. Further work is necessary to determine the value of individual tests at different ages, and only then can clear management criteria be drawn up. Hypermetropia, especially if unilateral, and a positive family history are risk factors for amblyopia and strabismus, and in their absence, the review of a child is probably not indicated. While the diagnostic value of simple qualitative tests is not in doubt, the detailed measurement of visual acuity for the amblyopia and strabismus suspect under 2.5 years of age, in a routine clinical setting, is doubtful.

A number of fundamental questions still remain about the methodology of screening: efficacy, validity and cost effectiveness. While it is recognised that further research is required, several centres have already introduced models of practice that are community based and utilise orthoptic optometric teams.⁵⁴ Our unit, which serves an area without community screening, uses the following protocol to diagnose strabismus and amblyopia: identify at risk children, and discharge as soon as possible those who are not at risk—so minimising unnecessary follow up visits (as shown in Figure 1).

There are a bewildering number of systems in place for the assessment and management of amblyopia and strabismus and their simplification would facilitate the development of cohesive children's ophthalmic services. It is our opinion that, while the full ophthalmic team should be involved in developing integrated care pathways, much of the assessment and subsequent management can be undertaken by orthoptists working with optometrists.

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