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# Is community screening for amblyopia possible, or appropriate?

Amblyopia is the commonest cause of acquired unilateral visual loss in children under the age of 6 years. Vision screening programs of widely varying nature currently exist in many communities and the recent development of photoscreening technology has resulted in proposals that this become an integral part of the screening armamentarium. People have focused on whether it is *possible* to detect precursors of amblyopia or the condition itself in younger children. What is lacking, however, is a consideration of whether it is *appropriate* that this occur as part of community screening programs. This annotation considers the current level of understanding about amblyopia, its natural history and treatment, and whether or not well recognised criteria for screening are met.

Amblyopia refers to subnormal visual acuity in an ophthalmoscopically normal eye, due to abnormal visual experience early in life. The commonest forms are usually unilateral. Strabismus is the major cause of amblyopia, which can also be due to anisometropia (a significant difference in the refractive error of the two eyes), ametropia (a large refractive error, particularly hypermetropia, in both eyes that frequently leads to a convergent squint), deprivation (such as with congenital cataract or corneal opacity), or a combination of the above. Amblyopia is the most common cause of acquired unilateral visual loss in children under the age of 6 years.

This discussion of whether amblyopia is a suitable target for community screening has been generated by both the increasing and necessary interest in preventive childhood community health measures, and by the propensity for screening programs to be proposed and established without adequate data to develop a clear understanding of how effective they are likely to be. The criteria by which a potential screening program should be judged, whatever the condition to be detected, are well recognised.[1] These criteria are often overlooked altogether, or only considered in part, in the establishment of community health screening programs. Vision screening programs of widely varying nature exist in many communities. Consistent goals are the prevention of amblyopia, or the early detection and treatment of amblyopia and any associated ophthalmological abnormalities. In this article we review critically whether such programs are appropriate, by considering how well the condition of amblyopia fulfils each of the following criteria for a screening program.

## (1) The condition being sought should be an important health problem for the individual and for the community

This is linked in part to the prevalence of the condition, as well as to the effect it has on the individual and on the community. The range of amblyogenic and refractive states in a specific community is not well known. Amblyopia is most commonly reported to have a prevalence of about 2%.[2] The rate is even higher with prematurity, developmental delay, and where there is a positive family history.[3] However, reported prevalence varies among studies according to the populations examined, and tests and criteria used. The prevalence is much higher than some well accepted screened-for conditions such as cystic fibrosis,[4] so this certainly should not be an impediment to setting up a screening program for amblyopia. Despite being so common, however, there is little known about the long term consequences of amblyopia – vital knowledge if one is to be able to determine the importance of the condition.[5]

The most commonly given reason for wanting to prevent amblyopia is to ensure that there are two functioning eyes, and so that the risk of blindness in the event of trauma to an eye, or as a result of one of the many processes that occur with advancing age, is minimised. The only reported estimate for the incidence of blindness is about two to three times that of non-amblyopic individuals,[6] yet the reliability of this estimate must be viewed with caution because of extrapolation of data from a tertiary centre to the community. Certainly it is intuitive to believe that two eyes are better than one, but apart from the increased risk of total blindness, what other negative effects are there of failing to screen for or not detecting amblyopia? Even if amblyopia cannot be completely avoided, correction of refractive errors in the better eye and in the worse eye can have positive benefits through optimising normal visual function and visual function if the better eye is subsequently impaired.

There are some – but not many – occupations from which amblyopes are excluded.[7] As well, surgery for senile cataracts is generally delayed for as long as possible in subjects with ocular comorbidities because of the risk of blindness associated with the procedure.[8] Cataract surgery is a very common surgical procedure performed in the United States and Australia, and current research is seeking to confirm the impression that cataract surgery leads to an improved quality of life.[9] Delay of such a procedure, as well as the increased concern that patients will have that they will completely lose their eye sight, is likely to lead to significant morbidity, but this has not been confirmed or quantitated.

The relationship between visual disorders and learning difficulties is one area that has received considerable interest, particularly because learning difficulties are so common, and because of the popularity of unproved therapies such as optometric training. While it would appear that defects in acuity, refractive errors, problems of accommodation-convergence, and instability of eye dominance can make the reading task more difficult, and are often clustered within individuals, there is notable lack of strong evidence that these factors inevitably lead to a learning disability.[10 11] In other words, amblyopia has not been shown to cause learning difficulties. It should be conceded, however, that it may contribute when other factors are suboptimal, and the influence of amblyopia and/or strabismus on school performance (through influencing self perception and peer relationships, or sporting performance, for example) has yet to be determined.

Other possible negative effects of amblyopia and its treatment do not appear to have been well studied. It is not known, for example, the extent of any psychosocial maladjustment associated with amblyopia, whether self esteem is diminished, and whether long term functional outcomes such as employment record and degree of criminality are increased. These are important longterm outcomes that must be considered as possible consequences of a chronic childhood condition,[12] though admittedly it may be difficult to separate out the effects of amblyopia from the many other possible contributing factors.

## (2) There should be an acceptable form of treatment for patients with recognisable disease, or some form of useful intervention should be available (for example, genetic advice)

Can amblyopia be reversed or prevented? While the answer is yes, this must be a qualified yes. There are still a number

of issues around the form treatment should take (physical or pharmacological occlusion treatment, or strabismus surgery for example), and it is not clear whether intervention could be successful for all potential amblyopes or not. Much of the research would suggest that a number of children may not be treatable. In part this is related to the degree of progression of the amblyopia, as well as the development of the central nervous system (and therefore age at which treatment is started), but there also appears to be some other individual factors – plasticity we will call them – that as yet have not been clearly defined. Some children simply respond to treatment better than others, and this is more than just a compliance effect. Further research is needed to determine what proportion of children detected at various ages are treatable. The proportion of children for whom visual acuity subsequently deteriorates after successful treatment (in one study reported to be 55%),[13] the degree of such deterioration, and whether it can be predicted is another area requiring research.

On the other hand, even if treatment of the amblyopia itself is not successful, there are benefits from the early detection of amblyopia, as mentioned in the previous section, and as well the ability to provide appropriate education about eye protection strategies. On the latter point, however, there is lack of research to show whether, on an individual or on a community level, this is in fact helpful.

**(3) The natural history of the condition, including its development from latent to declared disease, should be adequately understood**

While the general concept of progression from predisposing factors such as strabismus (where present) to amblyopia is understood, the natural history of specific risk factors such as anisometropia is not so clearly known.[5] For example, it is not known why some children progress and others do not, nor why some children develop amblyopia with smaller differences in refractive error. Nor is it clear whether an early refractive error of itself necessarily represents a visual deficit. Regardless of whether there is a suitable screening test, in order that guidelines for referral be maximally effective (that is, optimising sensitivity and specificity) it is required that such factors be known.

**(4) There should be a recognisable latent or early symptomatic stage**

This is related to the detection of conditions which predispose to the development of amblyopia, such as strabismus and refractive errors, and is very much an issue of the effectiveness of the screening test used. As indicated in the preceding section, however, it is not clear what proportion of children so detected in fact do need treatment. Nor is it known what factors, other than age, determine when amblyopia becomes no longer treatable. It is thus very difficult to predict which children will develop amblyopia, and thus renders problematic the issue of a recognisable latent or early symptomatic stage.

**(5) There should be a suitable test or examination for detecting the disease at an early or latent stage, which should be acceptable to the population**

Cochrane and Holland have laid down characteristics of what they perceive to be the ideal screening test.[14] It should be:

1. Simple, quick, and easy to interpret – capable of being performed by paramedical and other personnel. These factors will determine the reliability of the test and the cost of administration.
2. Acceptable to the public, as participation in screening programs is voluntary. The perceived benefits of the detection of a condition are balanced against factors such as the inconvenience and discomfort involved in the process of screening, confirmation of diagnosis, and treatment.
3. Accurate, that is give a true measurement of the attribute under investigation. Often there is some compromise between a screening test and a diagnostic one, and the training and experience of test administrators will influence this component.
4. Repeatable. This involves the components of observer variability (both within and between tests), subject variability, and test variability. By their nature screening tests are usually performed on a large scale and therefore a number of test administrators are involved.
5. Sensitive – the proportion of those with the condition who are detected – and specific – the proportion of those failing the screening process who don't have the condition. As it is not usually possible to have a screening test that has the highest sensitivity and highest specificity all at the one time,[15] a compromise is needed. The nature of the compromise depends on factors such as the seriousness of the disease, consequences of it being missed, how treatable it is if detected, and the effects of false positive results both on the individual and community. These factors appear not to have been addressed for amblyopia screening, though it is not unreasonable to place sensitivity above specificity on purely clinical grounds.

It is now appropriate to apply these criteria to screening for amblyopia. This process relies on detection of either a predisposing condition, such as squint or refractive error, or of a diminished visual acuity in the affected eye – in other words amblyopia already present. In infants and young children particular problems are encountered because of variable compliance with screening procedures, and developmental factors influencing whether a meaningful test can be performed (these two issues can be, but are not always, inter-related). At this stage, there is no suitable test for the routine screening of visual acuity in the child aged less than 3·5 years.[16] Tests are hampered by inadequate norms, low success rates, low reliability, and long test times.[17] Many of these problems are related to the developmental limitations of having a child respond to different visual stimuli in a manner that indicates that he or she is able to visualise and differentiate such stimuli.

In the older preschool child (from about 3·5 years) there is some evidence that orthoptists are able to screen for amblyopia with acceptable degrees of sensitivity, specificity and positive predictive value,[18 19] though others still dispute the ability of a true screening process, such as using Snellen charts, to assess adequately visual acuity in the 3·5 year old.[16 20]

The more training and experience a screener has, the better in general they will manage the young and/or non-compliant child, and the more able they will be to obtain accurate results,[21] however, comparisons between screening carried out by orthoptists and by 'generic' health care providers need to be viewed carefully for potential confounders. Firstly, invariably the orthoptists are using a much more extensive battery of tests, debatable sometimes whether in fact they are screening.[18 22] Secondly, it is not always made clear whether conditions under which the screening is carried out are the same for both orthoptists and others.[18] Thirdly, the amount of training of non-orthoptists is often either not specified or potentially inadequate.[22 23] As a consequence, it is not clear whether vision screening by orthoptists is necessarily more cost

efficient or effective. Furthermore, screening by orthoptists or other specialists is often not practical in terms of availability of personnel.

In the infant and young child, one is therefore reliant on the detection of strabismus, refractive errors, or abnormalities of the eye such as cataracts. The only published trial of a screening program for amblyopia in the young child showed that while the prevalence of amblyopia in the control group was greater two years after screening, this difference was not statistically significant.[24] Some relatively recent technological advances, video photorefraction for example, may change this, but are yet to be fully evaluated. The advantage with photorefraction is that minimal cooperation of the child is required, it is quick (less than one minute to get an adequate image), and that paramedical and other personnel can be trained to perform the screening. As long as the process by which photorefractive screening is carried out user friendly (that is, not requiring significant travelling or long waits), it is likely to be acceptable to the public. The unknowns are related to its accuracy, repeatability, sensitivity, and specificity in a community screening situation.

**(6) Facilities should be available for diagnosing and treating the patients detected by the programme**

Even if one assumes that at present all people with amblyopia are detected at some stage, resources for diagnosis would need to be increased if an early screening program were in place as the difficulty of performing formal assessments increases as the age of the referred child decreases. The younger child may need to be evaluated over a number of sessions. On the other hand, it is not clear whether the resources required for appropriate treatment would need to be increased if more children were detected at an earlier age. While treatment of the younger child is technically more difficult (compliance once again is a major issue), if the treatment was more successful and less time consuming, money might be saved in the management of each individual.

**(7) There should be an agreed policy on whom to treat**

As the natural history of progression from predisposing factor to amblyopia is not well known, it is not possible at present to differentiate those who will progress from those who will not. Furthermore we cannot adequately determine before starting treatment which children are likely to respond. Consequently, it is difficult to sustain a policy that would omit to treat all children who failed screening and who had an abnormality detected on full ophthalmological examination. This would potentially result in unnecessary treatment for many.

**(8) The treatment at the presymptomatic stage of the disease should favourably influence its course and prognosis**

Potentially, amblyopia is preventable and/or reversible if detected early in its course. This is not the case for all individuals, however. Once present, and with increasing age, amblyopia becomes less treatable,[25 26] though there are reports of adult amblyopes regaining vision in the affected eye.[27] While it is not unreasonable to suppose that early detection would improve the overall response rate to treatment, it is not known what proportion of children untreatable at say 5 years may have been treatable at 2 years, and so on. It is also not known for example, to what extent detection at 12 months may offer advantage over detection at 18 or 24 months.

The treatment of amblyopia is focused on correction of strabismus where appropriate, correction of significant refractive errors, and occlusion. The first is not without potential side effects, and occlusion is often a traumatic process for the child and family. Negative effects of this struggle may outweigh any positive benefit, especially in those who are unlikely to respond.

**(9) The cost of case finding, which should include cost of diagnosis and treatment, should be economically balanced in relation to (a) possible expenditure on medical care as a whole and (b) the cost of treatment if the patient does not present until the disease reaches the symptomatic stage**

Amblyopia is generally said to be not treatable once fully developed, usually by the time a child is in the middle primary school years (though a small proportion may respond to treatment at a later age).[26] One therefore either detects at an early age those conditions known to predispose to amblyopia, or picks amblyopia itself up at an early age.

The costs of failure to diagnose a condition such as amblyopia are hard to determine, even assuming that treatment up to say school age was invariably effective. They must be heavily influenced by the costs incurred due to trauma to the good eye, and processes resulting in acquired secondary ocular comorbidity (such as cataracts), as well as the likelihood of these occurring.

The cost of a vision screening program needs to include not only the personnel and equipment costs of carrying out the screen itself, but also the costs in running and supervising the program, in assessing those children referred for further evaluation (and it is not clear what proportion will end up not requiring treatment), and in ongoing treatment (including those cases where it is not successful).

**(10) Case finding should be a continuous process, not a once and for all project**

There are many vision screening programs targeting preschool children currently in existence, yet there is little scientific evidence for their continuation. In *Health For All Children*, it is stated that there is no justification for formal screening for visual defects as part of preschool health surveillance – a reflection of the current state of the art with regards to vision screening rather than denial of the significant potential health gains that could be made.[16]

If it is going to be effective at all, vision screening is most likely to be so when carried out as one component of an ongoing and complete child health surveillance program. Despite the current technical difficulties in providing appropriate means of early detection of vision problems, many countries such as the UK do have the public health framework required to support a vision screening program.

## Summary

Photoscreeners are becoming increasingly available and are being widely used to screen for visual abnormalities in young children.[28 29] However, consideration of accepted criteria for screening programs indicates there is still much further research that needs to be carried out before amblyopia screening could be recommended as a routine component of a community health surveillance program – an adequate description of the potential consequences of an individual developing amblyopia has yet to be provided and the natural history of the condition and factors that

determine the effectiveness of treatment have yet to be fully described. While there is the promise of technology that satisfies specific test requirements, this still needs to be trialed in community settings and community trials are required before it will be possible to determine whether the costs that will be incurred in carrying out routine screening and in providing the resources for treatment are warranted.

In conclusion, the development of new and possibly more effective technology for screening is only part of the answer to the amblyopia question. While superficially this technology makes screening for amblyopia a possibility, we do not know at this stage whether or not it is appropriate.

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