

# Early intervention for child with visual impairment

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Discuss the role of early intervention for children with visual impairments

## **Introduction**

The whole area of visual impairment is a complex and difficult field. Children may either be born with a degree of visual impairment or they may acquire visual impairment at some stage after birth. Some defects may be comparatively obvious and easily detectable other may be very subtle and not obvious for some time.

Visual defects may occur as a single lesion or may be part of a larger spectrum of congenital or acquired problems. They can be directly referable to the eye itself, as in the case of infantile cataracts, or may be as a result of more diffuse trauma such as cerebral palsy or perhaps a genetic error of metabolism or even infections such as meningitis. In this essay we intend to consider the role and value of early intervention together with an assessment of the value of screening which is obviously part of the same consideration.

The mechanism of examination of the issue will be by means of a critical review of some of the relevant literature which has been recently published on the subject

## **Screening**

There are many definitions of screening. Perhaps one of the best for our purposes comes from Wald (1)

" The systematic application of a test or enquiry, to identify individuals at sufficient risk to benefit from further investigation or direct preventive

action, amongst persons who have not sought medical attention on account of symptoms of that disorder"

Screening is a common practice in many areas of the NHS. Whenever it is discussed, it is usually accompanied by prolonged discussions relating to cost-effectiveness and efficacy. With specific regard to visual impairment, screening for conditions that can produce visual impairment at an early age is utterly essential because of the development in early life of the visual processing pathways in the visual cortex (see below). (2)

There is a window of opportunity for correction, which rapidly closes depending on the nature and severity of the visual impairment. Because of the dire implications for vision in later life, the cost effectiveness of such screening procedures are seldom applied in this area. This does not mean to say that considerations of efficacy are not valid (see below), but simply that it is not possible to put an appropriate value on a person's sight. (3)

Clearly the purpose of a screening programme is to try to identify those individuals who may be at risk of developing a potentially treatable condition. It is not a diagnostic service. There will usually be both false positives and false negatives. The importance of the National Screening programme is to identify those individuals who would benefit from further specialist assessment.

A good place to start is the paper by Rahi (4). This study was designed to consider the efficacy of the screening programme in detecting a

comparatively straightforward, although not necessarily easy to detect, lesion – the congenital and infantile cataract.

The study was a cross sectional design study with an entry cohort of nearly 250 children under the age of 15 yrs. The object of the exercise was to ascertain the proportion of these children who were detected and treated at 3 months and 1 year of age.

The significance of this study is that it highlights either the difficulty of diagnosis (or possibly the inefficiency of the system) as the results were poor by any interpretation. The paper itself is quite detailed and comprehensive, but the results that are relevant to our considerations in this essay are that only 35% of congenital cataracts were diagnosed at the routine new-born examination and only another 12% had been diagnosed by the time of the 6-8 week examination.

Only 57% of the cohort had been seen and assessed by an ophthalmologist by the time they were 3 months old and a further 33% had not been examined or assessed by the time that they were one year old. The authors comment that their study showed that the prime reason for getting an assessment was because of the carer's concerns in about 40% of all cases. We shall consider the importance of early assessment, accurate diagnosis and appropriate treatment later in this essay, but it is clear from these figures that all three of these eventualities are denied to a very substantial proportion of children and infants with possible adverse consequences for their subsequent visual acuity.

In all areas of medicine, we should ideally work from a rational and verifiable evidence base. (5)

Given the fact that we can point to evidence that shows that a significant proportion of children with one (at least) visual defect are not routinely detected, we should examine the evidence base for the assumption that early detection is important.

For the sake of clarity we will initially confine our considerations to the circumstance of infantile cataract. We do know that infantile cataract is an important and potentially avoidable cause of visual handicap.(6) The resultant stimulus deprivation of the optic tract and visual cortex (7) caused by the inability of the retina to receive normal images, because of the distortion caused by the cataract, produces various degrees of amblyopia.(8) There is a substantial body of evidence to show that in order to optimise the eventual outcome, particularly with the denser forms of cataract, that corrective surgery needs to ideally have been carried out before the age of three months.(9)

Because of the developmental importance of the early visual stimulus it is reasonable to assume that the earlier that corrective treatment can be implemented, the better the result is likely to be (10)

Although we have initially considered the impact of early screening for the condition of infantile cataract, it follows that other conditions can equally well be screened with the same rationale for early treatment. If we accept that early treatment is the “ gold standard” in childhood visual impairment

(11), then it is possible to predict some populations of high risk births that will clearly need increased surveillance. Many of the genetic disorders which can cause visual impairment can be predicted (at least in statistical terms) and the children specifically assessed at birth (12)

The majority of the papers examined with regard to infantile cataract are of the same opinion that early treatment is vital to secure any hope of reasonable visual acuity. The evidence base for some other conditions of visual impairment is nowhere near as clear, and in some places, frankly contradictory.

If we consider the implications for other conditions of visual impairment in childhood we should consider the paper by Clarke MP (13) which specifically considers the efficacy of treatment of a unilateral visual impairment in the 3-5 yr. old age range. This is particularly relevant to our considerations here because the trial itself was well constructed and has a meaningful outcome. In broad terms, nearly 200 children who were identified as having a degree of unilateral visual impairment were allocated into two groups.

One group had “ appropriate treatment” the other had no treatment. The authors note that all children had treatment after the six month observation period. the children who received “ full treatment” with glasses universally had better visual acuity than those who did not receive treatment.

Interestingly, the mean treatment effect between the two groups was only one line on the Snellen chart. The degree of improvement was proportional to the degree of original impairment. The specific conclusions of this particular study are worth quoting verbatim:-

Treatment is worth while in children with the poorest acuity, but in children with mild (6/9 to 6/12) unilateral acuity loss there was little benefit. Delay in treatment until the age of 5 did not seem to influence effectiveness.

It is worth considering these conclusions in more detail as they have distinct relevance to the need for early intervention. The authors point out that there is a tendency for amblyopia to undergo a degree of spontaneous improvement which is consistent with the results of another trial (14).

It is also fair to point out that other trials do not concur. Simons K (15) suggest that untreated amblyopia will deteriorate with time. The authors feel that, on balance, they recommend the continued wearing of glasses until the age of 7, even if the visual acuity returns to normal before this time, to prevent the development of refractory amblyopia. (16)

When the authors compared the results of their study with children from districts who did not receive pre-school screening, the follow up study showed that deferring their treatment did not limit their potential for improvement and, very significantly, it nearly halved the number of children that needed to wear eye patches at all. (17)

On the basis of this evidence the authors felt able to conclude that it is the acuity at presentation rather than the chronological age of the child, that is the most important determinant of eventual outcome. This is consistent with a similar study by Hardman-Lea SJ (18) They actually quantified this by stating that:-

Children with a moderate acuity loss of 6/18 or worse showed a clear cut response to treatment, which itself arguably justifies screening to identify and treat these children. In contrast, children with mild acuity loss, who represent over half those identified with unilateral acuity impairment at screening in this and other studies, received little benefit from either treatment. This level of impairment, though often excluded from studies, is still commonly treated in routine clinical practice. We argue that children with 6/9 in only one eye should no longer constitute screen failures and do not justify treatment, even with glasses.

Rather disturbingly the trial threw up one (probably statistical) anomaly:-

The glasses group with moderate initial acuity, in whom patching treatment was deferred, showed no overall gain in acuity at post-trial follow up. While this is probably a random effect, it raises the question whether prior refractive correction might in some way limit the effectiveness of subsequent patching.

Clearly this cannot be regarded as based on firm evidence, but raises the spectre that early treatment may actually be detrimental.

If we consider a more technically sophisticated study (19) Weiss A et al (2004) that looked at visually evoked potentials (VERs) in amblyopic children and compared the ages of instigation of treatment (patching) and the detectable effects on the VERs. This is an extremely complex paper but careful weighing and critical analysis of the results shows that, as far as amblyopic children are concerned, the critical window for demonstrating and



exploiting cortical neuronal plasticity extends up to the age of ten.( also 20). Although this paper specifically does not comment on the fact, an earlier paper by the same author (21) points to the fact that the plasticity, and therefore adaptability, progressively diminishes from about the age of five onwards

The PEDI Group (22) complicate the findings further with their contention that treating amblyopic children in the 3-7 yr. age range did not produce significantly different clinical outcomes when compared to an older age range

Screening, in general terms, has been overhauled by the National Screening Committee (23) which has sought to apply the classic Wilson & Junger (24) criteria to all aspects of NHS screening. Within the recommendations of this body, various specialist organisations have produced their own guidelines.

In the UK, the need for early visual impairment screening is recognised. Clearly this is different from being done efficiently in all cases. The most authoritative guidance that is currently available in this country is that which is based on the recommendations of the two national working parties who produced a joint report. The Royal Colleges of Ophthalmologists and Paediatrics and Child Health (25)

The current recommendations include an inspection of the eyes together with an evaluation of the red reflex at birth and then a fuller assessment which would include an examination for the presence of squint and visual behaviour generally at about 6-8 weeks. (26)

Later on in childhood there are other specified screening procedures which are designed to detect abnormalities such as strabismus, abnormalities of colour vision and reduced visual acuity although an examination of the literature would suggest that the pick up rate is surprisingly small (27). This particular author suggests this is mainly because the vast majority of cases are brought to the attention of the primary healthcare teams by the carers before screening is carried out.

In this essay we have conducted a brief overview of some of the relevant literature in the field of early detection of visual impairment. The results are disappointingly confusing. Some areas appear to have a fairly clear cut and universal agreement, others seem to produce well constructed studies that offer seemingly mutually exclusive results. (28).

The area of the infantile or congenital cataract appears to be one of those areas where there is fairly universal agreement that early treatment is beneficial, but the biggest stumbling block appears to be the comparative inability to pick up or detect the abnormality in a clinical screening setting.

Although we have not presented firm evidence, as it is not directly relevant to our discussions, there is also the problem that surgical intervention, although obviously helpful in terms of preserving vision, may actually have a down side that iatrogenic glaucoma is a possibility in later life. (10)

The converse situation appears to apply to the amblyopic patient, or the patient with strabismus. We have presented evidence that appears to be frankly contradictory. Although it appears easier to detect these

abnormalities in the older child there is considerable disparity in opinion about whether early treatment is either beneficial, or in the case of one of the papers presented, even helpful.

It is clearly difficult to form an opinion with any sort of firm evidence base in these circumstances. Although it is reassuring to read a paper and find that the authors call for “ more research to be done” in that particular area, it does not help those practitioners currently working in the clinical field, to come to a firm view on whether early treatment is either indicated, useful or even necessary in these particular circumstances.

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