Severe visual impairment: practical guidance for paediatricians

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Abstract

Visual impairment is generally categorised, along with hearing impairment, as a sensory disorder. Vision is arguably the dominant human input sense, and supports the development of cognitive, language and social skills as well as early emotional bonding and later literacy and independent mobility. Vision however also drives the integration of input from other senses and limitations in vision presents the infant with significant challenges in making sense of what is heard or touched. Severe visual impairment consequently has a significant impact on multiple aspects of early development and challenges continue throughout childhood. It is more appropriate therefore to think of severe visual impairment as a potential neurodevelopmental disorder, which requires ongoing input from a range of professionals.

Paediatricians play a key role not only in the early detection of severe visual impairment arising from primary visual disorders but also in the assessment and ongoing management of the developmental consequences. Severe visual impairment may be underdiagnosed in children with complex neurological impairment even in those already known to the Eye clinic. Paediatricians can play a crucial role in identifying possible visual difficulty, during early follow up of those at risk of neurological impairment, or via ongoing review of those with established complex disability.

Keywords blindness; development; developmental setback; functional vision; visual acuity; visual impairment

Definition of visual impairment

Visual impairment is defined according to distance visual acuity measured with both eyes open, once any refractive error has been corrected by spectacle wear (this is known as corrected acuity). A child with poor vision in one eye only is therefore not considered to be visually impaired. Snellen charts and acuity measures have been superseded by logMAR chart design and measures.

LogMAR refers to the logarithm of the minimum angle of resolution. Since an adult with normal vision can recognise letters whose individual components or spaces between components subtend 1 minute of arc at the eye (1/60th of a degree) at 6 m, a normally sighted adult has a Minimum Angle of Resolution of 1. Thus normal vision measured in logMAR is 0.0, since the logarithm of 1 is 0. It follows that a higher logMAR value indicates worse vision. (LogMAR values should not be confused with the decimalised notation of Snellen values used previously in some European countries, where a 6/60 Snellen value was equivalent to an acuity of 0.1).

ICD10 classification of visual impairment

Category	Presenting distance visual acuity	
	Worse than	Equal to or better than
0 Mild or no visual impairment		logMAR 0.5 (6/18)
1 Moderate visual impairment	logMAR 0.5 (6/18)	logMAR 1.0 (6/60)
2 Severe visual impairment	logMAR 1.0 (6/60)	LogMAR 1.3 (3/60)
3 Blindness	logMAR 1.3 (3/60)	logMAR 1.75 (1/60)
4 Blindness	logMAR 1.75 (1/60)	Light perception
5 Blindness	No light perception	

Table 1

ICD10 classifies visual impairment into four broad groups, namely normal, moderate, severe and blindness. However, the use of the word 'blindness' can be misleading since, as indicated in Table 1 this category is subdivided into several levels. A very low level of vision which allows gross detection of visual form, but not detail, may be developmentally useful for a young child, and the word blindness can be very misleading therefore, and particularly for parents. A further difficulty in classification of vision level for children is the requirement for distance acuity measures, which cannot be achieved for very young children.

There are further sources of confusion for the paediatrician who is not yet fully familiar with vision measures, particularly the practice of citing of certification or registration status. Certification is a system originally introduced for adults who required formal confirmation of acquired vision loss, to ensure provision of rehabilitation services. There are two categories, sight impaired and severely sight impaired, based on acuity reduction and/or visual field restriction. Certification may lead to local inclusion on a register of sight impairment (registration) which ensures that a social services assessment of need will be offered. However visual acuity measures may change over time, either reflecting genuine if modest improvement as the visual system matures, or the achievement of more accurate measures as the child's ability to comply with formal testing develops. Vision may also deteriorate in certain conditions such as retinal dystrophy. An acuity measure is more informative than a broad certification category, particularly as the certifying ophthalmologist is permitted discretion in choice of category. Whilst it may be helpful to know if certification has been completed, a clinic letter which states certification category with no further mention of current acuity measures will not provide useful guidance as to the child's current functional level of vision.

A final source of confusion may arise if refractive status is described without accompanying reference to measured acuity.

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Refraction is the process of light bending which achieves image formation within the eye, and is achieved by the lens and corneal curvature combined. Eye ball shape also impacts on whether the image falls on, behind or in front of the retina. Whilst a refractive error can of course influence acuity level, assessment of refractive status does not imply that acuity has been measured, since separate processes are required. Refraction is achieved with retinoscopy and requires minimal cooperation. Refractive status is measured in dioptres; a minus value indicates myopia (shortsightedness) and a positive value indicates hypermetropia (long sightedness.) Correction of a refractive error with glasses may result in normal (corrected) acuity. However corrected acuity may be reduced from normal if there are other abnormalities present. It is important to avoid the assumption that glasses wear achieves normal acuity and that acuity has been measured if refraction has been performed.

Epidemiology

Worldwide, 19 million children are visually impaired, according to WHO criteria; those with reversible conditions include 12 million with uncorrected refractive errors. 1.4 million have severe and irreversible visual impairment. Causes vary according to country income level with potentially preventable causes commoner in middle and low income counties. Globally, the commonest causes of severe visual impairment and blindness include retinal disorders, glaucoma, corneal scarring (primarily due to Vitamin A deficiency), cataracts and neurological causes.

Epidemiological studies in the UK have shown that cerebral causes (e.g. due to hypoxic ischaemic encephalopathy or neonatal hypoglycaemia) contribute to nearly half of the cases (40%) of severe visual impairment and blindness with inherited and acquired retinal disorders due to retinopathy of prematurity and retinal dystrophies, contributing to just under a quarter of the cases (24%). Disorders of the optic nerve (optic nerve hypoplasia and optic atrophy) are slightly less common (23%) and developmental disorders of the whole globe and cornea much less common.

Compared to other conditions affecting development, severe visual impairment is rare in UK children. Congenital causes (such as eye malformations, infantile cataract, early onset retinal dystrophies) are commoner than those with onset in childhood or adolescence (e.g. degenerative retinal conditions). The British Childhood Visual Impairment Study reported an incidence of 4/10,000 live births in the first year of life, rising to 6/10,000 by 15 years of age, and noted that severe visual impairment is accompanied by additional difficulties in up to 77% of children. Comorbid conditions may be a direct consequence of the underlying pathology or be developmental manifestations of the visual impairment itself. Children with disability are also at increased risk of ocular and visual abnormalities and these may remain silent if other problems dominate.

In the UK, as reported in other industrialised countries, visual impairment due to cerebral damage is reported to be the commonest cause of severe visual impairment. However this may be a very heterogenous group of children and may in practice include some children where early poor visual performance may in part be a manifestation of later developmental difficulties.

How does severe visual impairment present?

Abnormal ocular appearance, nystagmus or concerns about poor visual responses are common presentations for severe congenital visual impairment. An abnormal red reflex either during the neonatal examination or on subsequent images may indicate cataract or more rarely, retinoblastoma. However presentation can be delayed for some children if the significance of abnormal eye movements or delay in developing visually directed reaching is not appreciated or other concerns predominate. Some parents may find it hard to judge when eye contact appears to be reduced. Paediatricians should remain alert to the possibility of a severely visually impairing condition and any concern should prompt urgent referral to an ophthalmologist. Diagnosis will require ophthalmological examination which will include fundoscopy, refraction and assessment of visual responses. An electroretinogram may be required to identify retinal abnormalities, since fundoscopy may be normal initially and brain imaging may be helpful if optic nerve hypoplasia is suspected. Electrophysiology is available in specialist centres only and a second opinion from the regional paediatric ophthalmologist may also be required.

Identification of severe visual impairment in children of any age with significant movement limitations may be delayed if poor fixation and reach are wrongly attributed to postural and movement limitations rather than poor vision. Paediatricians should remain aware of the possibility of severe visual impairment in this group even if the child is already known to the ophthalmology clinic, since reduced acuity can also be missed by Eye clinic professionals.

Care pathway for the child with severe visual impairment

Following diagnosis of any condition causing severe visual impairment, a referral should be made by the ophthalmologist to the local specialist teaching service for children with visual impairment and to the community paediatrician, to provide further advice and support for the child and parents. Depending on the condition, further investigations may be indicated. For example, if optic nerve hypoplasia is identified, evaluation of pituitary function is indicated as this may occur as part of septo-optic dysplasia. For any condition likely to have an underlying genetic cause, referral to clinical genetics should be made. Clinical correspondence from initial and follow up consultations should be copied to all those involved in the child's care. The ophthalmologist may offer certification, which may lead to the child's inclusion on a local register (usually held by Social Care) but certification is not intended as a vehicle for ensuring educational and developmental support is provided. However there is misunderstanding about the role of certification even amongst Eye clinic professionals and it may be inappropriately assumed to directly activate appropriate support.

Vision assessment

Vision assessment is useful in determining how much vision is available to help drive key developmental skills, such as reaching for objects or towards the source of sounds. A very low level of vision may therefore be developmentally useful. A brief review of normal visual development provides useful context when considering how vision can be assessed in a paediatric clinic.

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