VISUAL IMPAIRMENT

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1. Definitions

International criteria for visual impairment take into account optimally corrected visual acuity at the best eye and visual fields. ICD-10 and WHO definitions do completely overlap.

Table 2 Definitions of visual impairment

ICD-10

	normal	**low vision**	**blindness**
corrected visual acuity*	≥1/3	<1/3 and ≥3/60	<3/60
at the best eye (Snellen values)**			
visual field from the fixation point	≥30	<30 and ≥10	<10

WHO

	corrected visual acuity* at the best eye **	
	<	3
moderate low vision	6/18 (0.3)	6/60 (0.1)
severe low vision	6/60 (0.1)	3/60 (0.05)
social blindness	3/60 (0.05)	1/60 (0.02)
blindness	1/60 (0.02)	light perception
total blindness	no light perception	

* In people with intellectual disability the best accepted correction is meant.

** If monocular measurement of the visual acuity is not feasible, binocular visual acuity is measured.

Visual acuity in young children and in adults with severe intellectual disability is measured by means of grating acuity methods, usually at a distance of 55 cm. These acuities are not expressed in Snellen values but in cycles per degree. Grating acuity values can not be directly used as optotype acuity values. Correlations are presented in table 3.

Visual impairment exists at a visual acuity lower than 10 cycles/degree.

Table 3. Correlation table of visual acuities measured in english feet, meters, cy/degree and min. arc

Snellen, metric	Snellen eng. ft	Spatial frequency cy/dg	min. arc
6/6	20/20	30	1
6/12	20/40	15	2
6/18	20/60	9.8	3
6/24	20/80	8	4
6/30	20/100	6	5
6/60	20/200	3	10
6/120	20/400	1,5	20
6/180	20/600	1,0	30
6/240	20/800	0,75	40
6/360	20/1200	0,50	60
6/480	20/1600	0,35	80

Near vision

Visual acuity is usually assessed at 40 cm, preferably measured in the crowded version. Children and adults with intellectual disabilities do often prefer a shorter distance. If they are tested with pictures, Landolt rings or Snellens E, they will be asked to read or match the pictures at a distance of 30 cm. Impairment for near vision may then be defined as:

Moderate low vision < 0.3 and >0.1

Severe low vision <0.1

People with a severe or profound intellectual disability are more dependent on near vision than on vision at a distance. Therefore, in this group, assessment of near vision is of central importance. In praxis, vision in this group is already usually measured with acuity cards at a distance of 55 cm.

Other aspects of visual impairment

In some instances persons will have normal visual acuity and visual fields but still have a functional visual impairment, as a result of impaired color vision, contrast sensitivity or light sensitivity or of oculomotor dysfunction. Prevalences might be higher in the population with an intellectual disability than in the general population, but sofar, epidemiological data are completely lacking.

Impairment and disability

The consensus committee is aware, that the actual disability caused by a visual impairment is also influenced by other factors, such as the moment of diagnosis and treatment, cooperation of parents and other caretakers, proper use of spectacles and methods of alternative communication, auditive

and motor handicaps, the socio-emotional development and the level of the intellectual disability. As a result, less severe levels of visual impairment may lead to more severe disability in people with an intellectual disability. Practical experiences show that *dual sensory impairment* has an increased prevalence in the population with intellectual disabilities, especially in people with a severe or profound intellectual disability and in ageing people. Expertize on diagnosis and on interventions in people with a severe intellectual as well as a double sensory impairment is still very limited.

2. What are the advantages of early identification of visual impairment and the consequences of late identification in children and adults with an intellectual disability?

The primary justification for early identification of visual impairment in infants relates to its impact on the psychomotor development, cognitive achievements and social/emotional development. Reduced visual function or blindness at birth or during early childhood interferes with early psychomotor development of the child, which is essential for cognitive and social development, whereas, without expert support and education, parent-child interactions will tend to be inadequate. Therefore, early childhood visual impairment is likely to have additional harmful effects on the social, cognitive, emotional and communicative development in this already handicapped group. Delayed identification and management of visual impairment may impede the child's ability to adapt to family and community life and may mimic or cause behavioural problems or may enlarge the experienced intellectual disability.

The effects on communication and social functioning of early identification and treatment or rehabilitation of acquired visual loss in older children and adults with an intellectual disability have not been assessed. It can be argued that these effects are even larger in a population with an intellectual disability. Apart from that, they have the same right on diagnosis and treatment of sensory loss as other people have. Identification requires active screening in this group.

3. Which children and adults should be screened for visual impairment and when?

Visual screening in infants

Children with a developmental delay are at risk for a congenital visual impairment. They may be expected to benefit from a standardized infant vision screen during the first years of life as is used routinely in well-baby clinics. However, apart from an increased risk of eye abnormalities, a different

pattern of abnormalities than in regular child health care is to be expected in the group with developmental delay, with higher frequencies of retinal pathology and cerebral visual impairments (Bleeker-Wagemakers, 1981; Schenk-Rootlieb et al, 1992). Moreover, many children with a developmental delay are under pediatric supervision and do not participate in regular routine infant screening programs. Therefore, by Warburg & Riise (1994) as well as in the Dutch consensus (NVAZ, 1997), *referral for ophthalmological evaluation is recommended for all children in whom a developmental delay is suspected, irrespective of outcomes of infant vision screens*. Ophthalmological diagnosis may also enhance the etiological diagnosis of the developmental delay. Further, *immediate postnatal checking for congenital cataract and follow-up for the detection of strabism and refractive errors is recommended in infants with Down's syndrome.*

Screening in older children

A number of children will develop visual loss during early childhood, but parents, teachers and other caregivers do not always recognize this as such because of the different communication of children with a developmental delay. Firstly, parents should be informed of the increased risk of visual impairment in their child and *parental concern about vision should always prompt diagnostic assessment*. Another necessary approach includes ongoing *active evaluation of visual function at routine child health monitoring* using formal assessment tools. Any failure at screening or lack of cooperation should prompt referral for further diagnostic evaluation.

Screening in adults

One of the most striking features of studies of visual impairment in adults with an intellectual disability, is the lack of self-report. Marked visual impairment may be just accepted or expressed as inactivity, irritability, inflexibility, autistiform behaviour, refusal to walk or self-injurious behaviour, often directed at the eyes. The visual loss is often not recognized as such by the caretakers and by consequence does not receive medical attention. As a result, authors assessing residents of group homes report visual loss or ocular problems in 51-68% (Wilson & Haire, 1990; Beange et al, 1995). These problems had been previously undiagnosed and unexpected in many cases. Therefore, active screening of visual functions, using formal tools, is strongly recommended. Preliminary research has shown that adults with an intellectual disability by other causes than Down's syndrome develop age-related visual impairment at comparable ages as non-handicapped people (Evenhuis, 1995; Schrojenstein Lantman-de Valk et al, 1997). As such, *screening in adults with previously normal visual function should start at the age of 45 years*. Adults with Down's syndrome are at risk for premature age-related visual loss (cataract, keratoconus), necessitating an additional screening around the age of 30 years.

At risk groups

Congenital

1 Down's syndrome (cataract)

2 Inborn errors of metabolism (e.g. mucopolysaccharidoses)

3 Specific syndromes (e.g. Leber's congenital amaurosis, Batten-Spielmeyer-Vogt, Bardet-Biedl syndrome)

Pregnancy and birth

4 Intra-uterine infection: rubella, cytomegalovirus, syphilis, toxoplasmosis, herpes, foetal alcohol syndrome

5 Asphyxia

6 Prematurity (cerebral hemorrhagia, artificial respiration)

Late-onset ophthalmological conditions

7 Down's syndrome

8 Self-injurious behaviour, directed at or near the eyes?

9 Old age

Late onset cerebral visual impairment

10 Meningitis

11 Significant head trauma

12 Brain tumour

13 Asphyxia by near-drowning or near-sudden infant death

4. What are the advantages and disadvantages of the current diagnostic system for visual impairment in different countries?

An overall disadvantage is, that many children with an intellectual disability do not participate in the regular child health monitoring system.

The consensus panel is also aware of the special problems in low-budget countries. Prevalences of childhood ophthalmological disease, visual impairment and blindness in these countries may be increased due to infectious diseases, lack of vitamin A and oxygen retinopathy, whereas screening techniques may not be generally available.

Early identification of early childhood visual impairment

In several countries, a.o. Sweden, Canada and the Netherlands, standardized screening of visual

function in infants by means of observation of visual attention and fixation and external assessment of eye structures and eye movements, is applied by public health officers (Lennerstrand, 1995). In this way, many cases of strabism could be detected and treated earlier and thus amblyopia prevented, whereas serious ocular and visual conditions may be detected earlier. In the Netherlands, a large study to evaluate effects of such a screening programme is in progress. Until now, effects in children with a developmental delay have not been assessed.

Diagnosis of visual impairment in older children

Many children with an intellectual disability can normally participate in existing screening programmes for pre-school and school children. However, not all children with an intellectual disability do visit schools, whereas children with a severe intellectual disability, autism or behavioural problems may not be able to cooperate.

Diagnosis of age-related and other progressive visual loss in adults

Many general practices include adults with an intellectual disability, often living in special homes. Experiments with annual health checks of these people by general practitioners or nurse-practitioners, including assessment of visual function, are taking place in Britain. Most adults with a mild or moderate intellectual disability can be assessed with routine methods. However, most general practitioners and nurse-practitioners will lack the equipment, time and experience to apply special methods to a non-cooperative population (Kerr, 1994; Evenhuis et al, 1997).

5. What are the current methods to assess visual functions in children and adults with an intellectual disability?

Generally used charts for visual acuity measurement

Visual acuity estimates using *letter, picture or symbol charts* can be performed without difficulties in most older children and adults with mild and many people with moderate intellectual disabilities (developmental age over 4-5 years).

Adapted methods for visual acuity measurement

Adapted methods are available for visual acuity measurement, such as the *Stycar and LH tests, Kay pictures and Osterberg chart* (Sheridan, 1981; Hyvärinen et al, 1992; Osterberg, 1965), providing matching cards for those who are not able to name the letters or symbols, and single letters or symbols for children that are easily distracted. The same symbols can be used for distance and near vision testing. Visual acuity measurements with such methods take 5-15 minutes per

person, dependent on the developmental level and the effective concentration. They can used reliably in people with developmental ages of 3-4 years and over.

Near visual acuity tests

If possible, the same methods are used as for visual acuity measurement at distance. Visual acuity for near is usually assessed at 40 cm, preferably in the crowded version.

Visual fields

According to the definition of visual impairment, screens of visual function should include assessment of visual fields. In children and adults with moderate or severe intellectual disability, assessment by means of perimetry can often not be performed reliably. In this group, visual fields can only be assessed globally by means of a confrontation method, and subtile peripheral as well as central defects can not be detected.

Visual screening in infants, young children, and people who are unable to perform on optotype tests

Visual screening of infants and young children may consist of structured observation of the eyes, visual attention and fixation, eye position and eye movements, whereas grating acuity is to be measured by a *preferential looking method, e.g. the Teller or Cardiff acuity cards* (Mohn & Van Hof-van Duin, 1986; Mackie & McCulloch, 1995; McCulloch et al, 1996; Hertz BG & Rosenberg J, 1992). The acuity card method has been evaluated for use in handicapped children and can be applied for vision screening of children and adults with severe to profound intellectual disabilities. Reproducible visual acuity estimates are obtained with this method in 80-85% of young children and non-cooperative adults (Mackie & McCulloch, 1995). Its application requires special training and experience.

Additional diagnostic methods to be used in epidemiological studies

Contrast sensitivity

Impaired contrast sensitivity may add considerably to the level of functional visual impairment. In people with intellectual disability, it can be tested by e.g. the Hiding Heidi test. Although this is not a screening method, it is included in functional assessment procedures and it might be applied in epidemiological studies, especially in people with neurological motor impairment and an increased risk of optic atrophy.

Refraction measurement

Although refraction measurement is not a screening method, it should be part of the diagnostic battery in epidemiological studies, to obtain best corrected visual acuity values. Objective refraction measurement can be performed in a majority of children and adults with an intellectual disability.

Visual Evoked Potentials (VEP)

VEP are definitely not a method for screening purposes. In the past, VEP have frequently been used by specialized centres for visual acuity determination in young and handicapped children. Reliable application of this method requires highly specialized apparatus and advanced techniques for data-management and analysis, as well as specially trained technicians (Apkarian, 1994a,b). The sweep VEP technique (Tyler et al, 1979), albeit technically considerably simpler (and now readily available at reasonable cost) is inadequate for assessment of visual acuity and visual pathway integrity and development. This statement is applicable for children, in general, and for people with intellectual disability, in particular.

Therefore, VEP is now mainly applied for objective specialist diagnostic assessment.
The applicability of the different methods for vision screening in children and adults with an intellectual disability, according to the developmental age, is summed up in Table 4.

Table 4. Diagnostic methods for vision assessment screening in people with an intellectual disability

Method	**applicable for developmental age (yrs)**
Ocular inspection, eye movements, visual attention and fixation	> 0.2
Visual fields (confrontation method)	> 0.2
Teller/Cardiff acuity cards	> 0.2
(Visual Evoked Responses (VEP)	> 0.2)
Objective measurement of refractive errors	> 0.5
Hiding Heidi	> 0.2
Stycar single letters (distance and near)	> 3-4
LH single symbols + matching (distance and near)	> 2
Children's picture chart	> 3-4
Tumbling E	> 4-5
Stycar (distance and near)	> 4-5
LH chart + matching (distance and near)	> 2.5-3
Logmar charts (including Snellen) with letters and numbers	> 6

6. What is the preferred model for visual screening and follow-up?

The need for universal screening of ophthalmological abnormalities in young children with a developmental delay, as well as follow-up at pre-school and school age and active screening for age-related visual loss in adults has been explained above. This has resulted in the following proposed screening protocol (Table 5).

Table 5 Screening protocol for visual impairment in children and adults with an intellectual disability

People with an intellectual disability in principle should participate in the regular national health monitoring system. Referral is necessary in case of failure and in case of insufficient cooperation.

1. Age 0-3 years

Aim: detection of congenital ophthalmological abnormalities and cerebral visual impairment

- Referral of all children with suspected developmental delay to an experienced ophthalmologist for specialistic evaluation.
- Postnatal referral of infants with Down's syndrome for detection of congenital cataract.

Initiative: pediatrician, public health officer.

2. Age 3/4 years

Aim: detection of early childhood visual loss, strabism and refraction errors

Screening of all children who are not under ophthalmological control (at first specialist assessment no or non-progressive abnormalities) by means of observation of the eyes, eye movements, visual attention and fixation, visual fields and visual acuity measurement by means of acuity card procedure.

In case of abnormalities (at ocular inspection, visual acuity lower than according to age curve), referral to ophthalmologist.

Initiative: pediatrician, youth health physician, general practitioner.

3. Age 6/7, 12 and 18 years

Aim: detection of later childhood visual loss, strabism and refraction errors

Screening of all children who are not under ophthalmological control, by means of assessment of visual fields and visual acuity measurement by means of Snellen chart, tumbling E, children's picture chart, Stycar or LH cards, or an acuity card procedure, dependent on the developmental level.

In case of abnormalities (visual loss as compared to earlier assessments, discrepancy of near and distant vision, visual acuity ≤ 0.5 or ≤ 13 cycles/degree), referral to ophthalmologist.

Initiative: youth health physician, school physician or nurse, district team physican.

1. Adults

Aim: first detection of visual impairment

- Screening by means of global assessment of visual fields and visual acuity measurement in all adults who have never been assessed.

Aim: detection of age-related visual loss

- Adults with refractive errors: checks of visual acuity (with glasses) every 5 years.
- Adults with Down's syndrome: visual acuity measurement around age 30 years for detection of age-related cataract and degenerative changes in keratoconus.
- Screening of all adults from age 45 years onwards every 5 years by means of global assessment of visual fields, visual acuity measurement, and measurement of ocular pressure.

Initiative: district team physician, general practitioner or nurse-practitioner, institute physician, ophthalmologist.

7. What are the important directions for future reseach on assessment of visual impairment?

- Large-scale epidemiological studies (a.o. on etiology, frequency, severity and risk factors) of visual impairment in the population with an intellectual disability.

- Association of visual impairment and specific syndromes.
- Development of checklists for behavioural signs of visual impairment and innovative behavioural tests that are applicable for visual screening programmes.
- Development and evaluation of methods for visual field assessment in people with an intellectual disability.
- Evaluation of methods for measurement of ocular pressure in adults with an intellectual disability.
- Studies of (cost-)effectiveness of various procedures and different settings for visual screening.
- Evaluation of the feasibility of the proposed screening protocol.

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