HEARING IMPAIRMENT

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References on hearing impairment

1. Definitions

Hearing loss is defined as **the averaged hearing loss at 1000, 2000 and 4000 Hz**, measured by pure tone audiometry. This definition, although different from WHO/ISO definitions, is currently used in the Netherlands because of its higher relevance to speech discrimination. It also corresponds to the standard proposed by the British Association of Otolaryngology and British Society of Audiology (1983). High frequency hearing loss has to be taken into account in this population, because with this type of loss, speech understanding is specifically hampered in noisy circumstances. People with an intellectual disability are frequently exposed to higher noise levels in homes and daycare centres.

Hearing impairment is considered to be present as the hearing loss as defined above is 25 dB or more at the best ear.

Definitions of the degrees of hearing impairment are presented in Table 1. The consensus committee is aware that these definitions are usually related to the loss at 500, 1000 and 2000 Hz, but decided to redefine them in relation to 1000, 2000 and 4000 Hz.

Table 1 Degrees of hearing impairment

	loss (dB)
mild	25 - 30
moderate	30 - 60
severe	60 - 90
profound	90 - 120
(sub)total deafness	over 120

The consensus committee is aware, that the actual handicap by a hearing impairment is also influenced by other factors, such as the moment of diagnosis and treatment, monaural hearing, co-operation of parents and other caretakers, proper use of hearing aids and methods of augmentative and alternative communication, the level of noise in daily life, visual and other handicaps, the social-emotional development and the level of the intellectual disability. Many people with intellectual disability suffer from both hearing and visual or motor impairment. As a result, less severe levels of hearing impairment may lead to more severe handicap, if they appear in people with an intellectual disability, as compared to people without an intellectual disability.

In people with a severe or profound level of intellectual disability, apart from the actual hearing loss, other aspects may influence intervention decisions, such as pre-speech functions, the noise level in the home or daycare centre and the feeling of security.

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

The primary justification for early identification of hearing impairment in infants relates to its impact on speech and language acquisition, cognitive achievements and social/emotional development. Reduced hearing acuity during infancy and early childhood interferes with the development of speech and language skills, because it is likely that the child will not receive adequate auditory, linguistic, and social stimulation required for speech and language learning, social and emotional development, and that family functioning will suffer accordingly (NIH, 1993). It can be argued that these effects are even larger in a population with an intellectual disability. Therefore, early childhood hearing 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 hearing 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 hearing loss in older children and adults with an intellectual disability have not been assessed. In principle, they have the same right on diagnosis and treatment of sensory loss as other people have.

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

Infants

According to an NIH consensus statement on early identification of hearing impairments (NIH, 1993), "it is clear that the earliest possible identification of hearing-impaired children is optimal for effective intervention to improve communication skills, language development, and behavioural adjustment. Identification of all children with hearing impairment at birth is ideal. As a practical matter, the cost of universal screening has been prohibitive. Attempts

have been made to limit costs by focusing neonatal testing on those at highest risk. Unfortunately, research shows that this approach misses 50 percent of children who are eventually diagnosed with severe to profound hearing impairment." Indeed, according to a recent report from the British National Coordinating Centre for Health Technology Assessment, 400 out of about 840 children that are born every year in Britain with clinically significant permanent hearing loss, will have been missed by the age of 18 months by current detection methods (Abbasi, 1997). Therefore, universal screening for all infants within the first 3 months of life was strongly recommended in this American consensus statement. Recently, it was also recommended in Britain that the National Screening Committee should urgently address the need for a national neonatal screening programme for congenital hearing impairment (National Coordinating Centre for Health Technology Assessment, 1997). Children with a developmental delay are at risk for congenital hearing impairment. *Therefore, if universal neonatal hearing screening would be implemented, children with developmental delay would undoubtedly benefit. As long as this is not the case, we strongly recommend hearing screening of all infants in whom developmental delay becomes apparent, as early as possible*.

Older children

A number of children will develop hearing loss during early childhood, but parents and teachers 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 hearing impairment in their child and *parental concern about hearing should always prompt diagnostic assessment*. Another necessary approach includes ongoing *active evaluation of hearing function at routine child health monitoring* using formal assessment tools. Any failure at screening should prompt referral for further diagnostic evaluation.

Adults

One of the most striking features of studies of sensory impairment in adults with an intellectual disability, is the lack of self-report. Marked hearing impairment may be just accepted or expressed as inactivity, deterioration of speech, irritability, inflexibility or autistiform behaviour. The sensory 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 hearing loss in 25-42% (Wilson & Haire, 1990; Beange et al, 1995; Mul et al, 1997). These problems were previously undiagnosed and were not considered by

caretakers in up to 80% of cases (Mul et al, 1997). Therefore, active screening, 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 hearing 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 hearing function should start at the age of 50 years*. Adults with Down's syndrome are at risk for conductive hearing loss as a result of ongoing middle ear infections as well as premature age-related hearing loss (presbyacusis). Therefore, *ongoing screening of hearing function during life is recommended in adults with Down's syndrome.*

At risk groups

At risk groups for hearing impairment, for whom active diagnostic evaluation should be considered, are presented in Table 2.

Table 2. At risk groups for hearing impairment

Congenital

1. Family history of hereditary sensorineural hearing impairment
2. Craniofacial anomalies
3. Inborn errors of metabolism (e.g. mucopolysaccharidoses, mucolipidoses and Refsum's disease)
4. Autism

Pregnancy and birth

5. Intra-uterine infection: rubella, cytomegalovirus, syphilis, toxoplasmosis, herpes
6. Maternal ototoxic drug use
7. Low birth weight (less than 1500 grams)
8. Hyperbilirubinaemia at a level requiring exchange transfusion
9. Asphyxia

Late-onset

10. Bacterial meningitis (especially pneumococcal)
11. Recurrent or persistent otitis media with effusion
12. Anatomic deformities that affect eustachian tube function
13. Ototoxic drug use
14. Head trauma associated with loss of consciousness or skull fracture
15. Down's syndrome
16. Automutilation of the ears (eardrum perforations, chain luxations, leakage)

14. Noise (work, walkman)

17. Old age

Impacted ear wax

Occlusion of external ear canals by impacted ear wax is a frequently overlooked cause of conductive hearing loss in children and adults with an intellectual disability. Crandell & Roeser (1993) found a considerably higher incidence of excessive cerumen and cerumen impaction in adults with an intellectual disability (28%) than in the general adult population (2-6%), both in people with and without Down's syndrome. Associated conductive hearing loss varied from 15 to 45 dB. Moreover, a strong propensity for recurrent cerumen impaction was found. Therefore, all children and adults with an intellectual disability should be checked once or twice a year for impacted ear wax; people with Down's syndrome, ageing people and people wearing hearing aids might be checked more frequently.

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

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

Early identification of congenital hearing impairment

Currently, infant screening of hearing function is usually done by means of behavioural reflex audiometry (Ewing test, distraction test). The average of detection of significant permanent hearing impairment in British children, as reviewed by Davis, is 26 months in those children screened by health visitors, with intervention around 32 months (Abbasi, 1997). Detection in children with a developmental delay can be expected to be specifically hampered: because reliable application of behavioural reflex audiometry is strictly limited to the developmental age of 7-13 months, it can often not be reliably performed in these infants.

Recently, attention has turned to the measurement of *otoacoustic emissions* as a fast, inexpensive, noninvasive test of cochlear function (Kemp & Ryan, 1990; Probst et al, 1991). Since 1993, several promising experiments with OAE screening of newborns have taken place, both in hospital wards and as a part of routine infant health care. In Rhode Island (USA), Copenhagen County (Denmark) and Rotterdam (the Netherlands) large-scaled screening programmes are in progress (Kok et al, 1993; Oudesluys-Murphy et al, 1996).

Identification of acquired childhood hearing impairment

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

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

In many countries, routine hearing screening of adults is not available or only in case of reporting of hearing loss. The general experience is, that many adults with an intellectual disability are not active in this respect. 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, including assessment of sensory function, are taking place in Britain. Many adults with a mild or moderate intellectual disability can be assessed by normal *screening audiometry*. However, many general practitioners lack the equipment for screening audiometry and most of them will lack the equipment, time and experience to apply special methods to non-cooperative people (Evenhuis et al, 1997).

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

Pure tone and speech audiometry

Many older children and adults with a mild or moderate intellectual disability are able to cooperate reliably with pure tone and adapted methods of speech audiometry, if necessary after a conditioning procedure (play audiometry). If available and evaluated, methods of speech audiometry using pictures or miniature toys, developed for young children, can be applied. Practical experience in this group is, that these methods can be applied in most people with developmental ages of around 4 years and more. Often, more than one measurements are required to obtain reliable results.

Adapted methods of subjective audiometry

Subjective audiometry in children and adults with developmental ages lower than around 4 years requires specially trained and experienced audiologists, audiology assistents or speech and hearing therapists (Fulton & Lloyd, 1975). They may be able to apply play audiometry in persons with developmental ages of 2-3 years onwards. Two or three measurements are usually required to obtain reproducible results, because of the slower understanding, delayed and less consequent reactions, or behavioural problems. Methods using specific conditioning are *air puff audiometry* (Fulton & Lloyd, 1975; Lancioni & Coninx, 1995) and *TOUCH-audiometry*, which uses tactile stimuli and has originally been developed for children with autism (Verpoorten & Emmen, 1995). The use of visual stimuli to reinforce correct reactions (*visual reinforcement audiometry*) may even allow measurement of hearing thresholds in people with developmental ages from 10-12 months onwards.

In infants and people with profound intellectual disability (developmental ages lower than 1 year) *behavioural observation audiometry* may be applied (Mc Kormick, 1995; Moore, 1995). This method is time-consuming, its reliable interpretation requires a considerable experience, and it gives only a global impression of the hearing function. For practical reasons, visual reinforcement and behavioural response audiometry are not the methods of choice for screening.

Whispered speech

Although screening of the hearing function by means of whispered speech is often considered obsolete since the availability of screening audiometry, this may be a useful, easy and low-cost screening test when audiometry is not available or co-operation is refused. People with developmental ages over 5-6 years are usually able to repeat a series of words, whispered at a distance of 3 meters. The development of a Dutch whispered speech picture chart for use with older children and adults with intellectual disability is in progress. First evaluation in a group of 48 adults with a mild or moderate intellectual disability and a monaural or binaural hearing impairment and a control group of 43 persons with normal hearing has shown, that such a chart is applicable in people with developmental ages of around 3-6 years, resulting in a sensitivity for hearing losses of 25 dB and more of around 90%. Only some mild losses were missed. Specificity was lower (70-75%) as a result of behavioural and concentration problems (Evenhuis, personal communication). Similar charts can be constructed for other languages.

OtoAcoustic Emissions (OAE) and impedance audiometry

OAE is a quick and reliable method for hearing screening in older children and adults with an intellectual disability (Gorga et al, 1995; Maurizi et al, 1995). Because it is to be expected that results will often be influenced by middle ear pathology in these age-groups, routine application of impedance audiometry is recommended.

Several experiments are now taking place in the Netherlands with screening of (mostly adult) institutionalized populations with an intellectual disability by means of OAE, accompanied with impedance audiometry to detect middle ear problems. Dependent on the use of either click-evoked or distortion product OAE and the developmental level, such screening takes 10-20 minutes. These first experiments suggest, that click evoked OAE may be more easily disturbed by an unfavourable signal-noise ratio than distortion product OAE. First analysis shows, that 40-45% of adults are to be referred for specialist assessment because of middle ear pathology and/or inner ear hearing loss (personal communications by conference members).

Auditory Brainstem Responses (ABR)

During the last 10 years, measurement of high-frequency hearing thresholds by means of ABR has been succesfully applied in children and adults with moderate to profound intellectual disabilities, that is in those that can not perform subjective audiometry reliably (Evenhuis et al, 1992; Maurizi et al, 1995). The test takes 30-60 minutes, depending on the hearing threshold and on the cooperation of the tested person. Practical experience learns, that oral sedation is required in around 30% of this group to avoid restlessness or anxiousness. Since ABR is a time-consuming method, requiring expensive equipment and specifically trained personel, this method is not fit for screening.

A limitation of both OAE and ABR is, that impairment of the function of the middle and inner ear is detected, but not abnormal function of cerebral auditory pathways. To date, knowledge of effects of cerebral hearing impairment is very limited.

The applicability of the different methods for hearing assessment in children and adults with an intellectual disability, according to the developmental age, is summed up in Table 4.

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

method	**applicable for developmental age (yrs)**
OtoAcoustic Emissions (OAE)	> 0
Auditory Brainstem responses (ABR)	> 0
Behavioural observation audiometry	> 0
Pure tone audiometry with visual reinforcement	> 1
Whispered speech	> 3
Pure tone (play) audiometry	> 3-4

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

The need for universal screening for hearing impairment in young children with a developmental delay, as well as follow-up at pre-school and school age and active screening for age-related and other progressive hearing loss in adults has been explained above. This has resulted in the screening protocol as proposed below.

Table 5 Screening protocol for hearing 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 hearing impairment

Screening of all children with suspected developmental delay with OtoAcoustic Emissions (OAE).

In case of abnormalities referral to an audiologist for auditory brainstem response (ABR) and/or behavioural response audiometry, if necessary after ENT treatment of middle ear pathology or obstruction in the external ear canal.

Initiative: pediatrician, youth health physician, ENT specialist.

2. Age 3/4 years

Aim: detection of early childhood hearing loss

Screening with OAE of the following groups:

- children with Down's syndrome
- children with recurrent or chronic middle ear infection
- children from families with progressive hearing loss.

In case of abnormalities referral to audiologist, if necessary after ENT treatment, ABR or adapted subjective audiometry.

Initiative: pediatrician, ENT physician, youth health physician, general practitioner.

3. Age 6/7, 12 and 18 years

Aim: detection of later childhood hearing loss

Screening of all children who are not under audiological control with screening audiometry, whispered speech at 3m distance or OAE.

In case of abnormalities referral to audiologist, if necessary after ENT-treatment, for pure tone and speech audiometry or ABR. More than one measures are usually required to obtain reliable results.

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

4. Adults

Aim 1: detection of previously unidentified childhood hearing impairment

- Hearing screening of all adults who have never been assessed.

Aim 2: detection of noise-exposure hearing loss

- Annual screening of all adults who are frequently exposed to noise (> 80 dB) with screening audiometry, whispered speech at 3m distance or OAE.

Aim 3: detection of age-related hearing loss

3A. Adults with an intellectual disability in general:

- Hearing screening of all adults from age 50 onwards every 5 years by means of screening audiometry, whispered speech at 3m distance or OAE.

3B. Adults with Down's syndrome:

- Hearing screening every 3 years throughout life by means of screening audiometry, whispered speech at 3m distance or OAE.

Initiative: district team physician, general practitioner, institute physician, ENT

7. What are the important directions for future research on assessment of hearing impairment?

- Large-scale epidemiological studies (frequency, severity, cause and risk factors) of hearing impairment in the population with an intellectual disability.
- Large-scale study of efficacy of early identification and intervention.
- Development of innnovative behavioural tests that are applicable for hearing screening programmes.
- Studies to allow comparison of various techniques and procedures for hearing screening.
- Evaluation of different settings on the effectiveness of screening procedures.
- Evaluation of the feasibility of the proposed screening protocol in low-budget countries.

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