

IASSID INTERNATIONAL CONSENSUS STATEMENT

**EARLY IDENTIFICATION OF
HEARING AND VISUAL IMPAIRMENT
IN CHILDREN AND ADULTS
WITH AN INTELLECTUAL DISABILITY**

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Introduction

For a number of reasons, a considerable amount of undiagnosed hearing and visual impairment is to be expected in children and adults with intellectual disability. Although epidemiological data for this population are still scarce, the prevalence of early childhood hearing and visual impairment can be expected to be higher than in the general population, since many causes of congenital and acquired childhood hearing and visual impairment, such as intrauterine infections, perinatal asphyxia, inborn errors of metabolism, meningitis and brain tumours, may be a common cause of cerebral damage as well (Rijn, 1989). Indeed, studies of people with an intellectual disability in institutes and in the community invariably show high prevalences of hearing and visual impairment (Jacobson, 1988; Wilson & Haire, 1990; Schenk-Rootlieb et al, 1992; Warburg, 1994; Beange et al, 1995; Mul et al, 1997; Van Schrojenstein Lantman-de Valk et al, 1997). On the other hand, in some cases, e.g. people with autism, a false-positive diagnosis of sensory impairment may be made as a result of abnormal responses to subjective test methods. Further, age related hearing and visual impairments can be increasingly expected as a result of the growing life expectancy in this population (Schrojenstein Lantman-de Valk et. al., 1994; Evenhuis, 1995). Moreover, hearing loss as a result of impacted ear wax is a frequent problem in adults with intellectual disability (Crandell & Roeser, 1993). Children and adults with Down's syndrome are an at risk group for congenital cataract and severe refractive errors (Pires da Cunha et al, 1996; Woodhouse et al, 1996), childhood conductive hearing loss, (Brooks et. al., 1972; Roizen et. al., 1993), early presbycusis (Buchanan, 1990; Evenhuis et. al., 1992), premature age-related cataract (Eissler & Longenecker, 1962) and degenerative changes in keratoconus (Völker-Dieben et al, 1993).

The above studies corroborate experience which shows that the visual or hearing loss is seldom spontaneously mentioned by patients, and remains frequently unrecognized by parents, teachers and staff of homes. During the past 10-15 years, screens of sensory functioning, with adapted or objective methods in populations of adults with intellectual disability, either living in institutes or in the community, have revealed substantial numbers of unidentified hearing and visual impairments and a large number of not properly treated impairments (Aitchison et al, 1990; Buchanan, 1990; Evenhuis et al, 1992; Warburg, 1994;

Kwok et al, 1996; McCulloch et al, 1996). Therefore, active diagnostic screening is important in this population.

However, with the tendency of providing the same care for people with an intellectual disability as for people without an intellectual disability, medical care is usually only provided when people or their caregivers ask for it (Wilson & Haire, 1990). Apart from this, specialized skills and equipment, necessary to overcome diagnostic and therapeutical problems, especially in those with moderate and severe intellectual disabilities, are not always known or accessible.

During recent years, in several countries initiatives have been taken to improve this situation. The Welsh Health Planning Forum issued a protocol for health gain in people with an intellectual disability, including active detection and monitoring of visual and hearing impairments in children as well as adults (Welsh Health Planning Forum, 1992). The Scandinavian ophthalmologists Warburg & Riise (1994) published suggestions for regular screens of vision and eye disease at recommended ages. In the Netherlands, national consensus was developed on early detection, diagnosis and treatment of hearing and visual impairment in children and adults with an intellectual disability (Evenhuis et al, 1996; NVAZ, 1997). There is a clear need for an international consensus on methods and models for early identification of sensory impairment in this population.

On September 11-14, 1997, the IASSID Special Interest Research Group on Health Issues together with the Dutch association of physicians providing care for people with an intellectual disability (NVAZ) convened in Leiden, the Netherlands, to develop an international consensus on early identification of hearing and visual impairments in children and adults with an intellectual disability. The conference brought together general practitioners and psychologists for intellectual disability, and specialists in audiology, otolaryngology, ophthalmology, pediatrics, neurology, speech and hearing sciences and epidemiology.

On June 8, 1998, the same group convened again in Manchester, United Kingdom, to discuss and conclude on last topics.

A consensus was reached based as much as possible on scientific evidence and further on additional clinical expertise. The panel prepared a draft statement in response to the following key questions:

1. What are the definitions of hearing and visual impairment and their severity?
2. What are the advantages of early identification of hearing and visual impairment and the consequences of late identification in children and adults with an intellectual disability?
3. Which children and adults should be screened for hearing and visual impairment and when?
4. What are the advantages and disadvantages of the current diagnostic system in different countries?
5. What are the current methods to screen hearing and visual functions in children and adults with an intellectual disability?
6. What is the preferred model for hearing and visual screening and follow-up?
7. What are the important directions for future research?

The panel is aware of the vast differences among countries around the world concerning risk factors and prevalences of ophthalmological and otological disease in the general population, and the availability of screening techniques and possibilities for intervention. However, the panel was of the opinion that this should not keep her from formulating recommendations for a screening procedure, based on current technological insights.

The consensus on hearing and on visual impairment, respectively, will be dealt with in two separate parts. Guidelines for further functional assessment, treatment and guidance of people with an intellectual disability and a visual and/or hearing impairment will be developed in a next stage.

Conclusions

Epidemiology

1. Prevalences of congenital and early childhood visual and hearing impairment are increased in children with an intellectual disability, as compared with children with normal intellectual abilities.
2. Children with Down's syndrome, with cerebral palsy and with a severe or profound intellectual disability are the largest special risk groups.
3. As a result of increasing life expectancy in the population with an intellectual disability, absolute numbers of adults with an intellectual disability and an age-related visual and/or hearing impairment are also increasing.
4. Age-related visual and hearing impairment start earlier in adults with Down's syndrome.
5. Impacted ear wax resulting in conductive hearing loss has increased prevalences in children and adults with an intellectual disability, as compared to people with normal intellectual functions.

Delayed identification

6. Identification and intervention of visual and hearing impairment are considerably delayed in this group because of underreporting by the persons and their caregivers, with increased chances of additional negative effects on the communication and social functioning and behavioural problems.

Diagnosis

7. Visual and hearing functions in cooperative children and adults can be screened by youth health physicians with methods that are normally used in children and adults without an intellectual handicap.
8. Visual and hearing function in insufficiently or non-cooperative individuals can be assessed with objective methods (OtoAcoustic Emissions, Auditory Brainstem Responses) or adapted subjective methods (whispered speech, adapted methods of subjective tone and speech audiometry, adapted methods of visual acuity measurement and assessment of other visual functions), requiring special expertise and equipment.

Recommendations

1. We recommend routine referral of all infants with an intellectual disability for a specialist audiological and ophthalmological evaluation as early as possible.
2. We recommend immediate postnatal checking for congenital cataract in children with Down's syndrome and active follow-up for the detection of strabism and refractive errors.
3. We recommend that all older children with an intellectual disability normally participate in routine health monitoring of hearing and vision, and that parental concern about hearing or vision always prompts diagnostic assessment.
4. We recommend screening of visual and hearing functions in all children and adults with an intellectual disability that have never been assessed or of whom no diagnostic data are available.
5. We recommend routine screening of all adults with an intellectual disability for age-related visual or hearing loss at age 45 years and every 5 years thereafter.
6. We recommend an extra vision check at age 30 years for adults with Down's syndrome, as well as screening of their hearing function every 3 years throughout life.
7. We recommend that district referral systems are developed for diagnosis, treatment and intervention of visual and hearing impairment in children and adults with an intellectual disability and for screening of non-cooperative individuals.
8. We recommend scientific studies of visual and hearing impairment in people with an intellectual disability, aimed at epidemiological aspects, innovative diagnostic methods and effectiveness of screening procedures.

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