

Guidelines for regular ear and hearing screening in people with an intellectual disability

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Objective

- Persons with ID are less healthy than the general population
- Syndrome-related disorders (epilepsy, motor problems), sensory disorders, inactivity-related diseases (osteoporosis, cardiovascular diseases), lifestyle-related problems (nutrition, exercise)
- Problems remain often unidentified
- ID hampers individuals' ability to communicate their health status and to participate in decisions about their own health
- Knowledge of the care givers regarding the medical history and the possible health problems of their wards are often insufficient (Hild et al., 2008)
- Persons with ID are medically undersupplied in general



Intellectual disability and hearing loss

- High coincidence of ID and hearing loss
- 1.2 to 3% in premature neonates (Lorenz et al, 1998; Robertson et al, 1994)
- 7 years old children - 10% hearing disorders (Cans et al, 2003)
- Adults with ID: 66% hearing disorders (Lowe & Temple, 2002)
- Frequently moderate to severe loss
- Causes: congenital and early acquired hearing impairment or conductive loss (unrecognized chronic middle ear infections, ear wax), eventually superimposed upon presbycusis (Evenhuis, 1995)
- Different kinds of mental disabilities are associated with different prevalence rates of hearing disorders
- Down syndrome: hearing deficits in 28 % (van Schrojenstein Lantman-de Valk et al, 1994) to 73 % (Squires et al, 1986) compared with 8 % to 22 % of persons with other types of intellectual disability

What is known from studies?

Hearing screening programs during Special Olympics games

Special Olympics: worldwide largest program of regional, national and international sport games and trainings for people with ID

Healthy Athletes Program of Special Olympics:

- Screenings for the otological and audiological, visual, statomotor, dental problems and for the general health state of the athletes
- Worldwide database
- Hearing screening of 4477 athletes from 87 countries collected at 36 Special Olympics events between 1999 and 2003 → fails in 30.9% (Montgomery, 2003)
- Even considering false positive screenings, this failure rate remarkably exceeds expected comparable failure rates of non-disabled adolescents and young adults where the prevalence of hearing disorders requiring treatment lies between 2 to 4%, depending on age (Hesse, 2003, Montgomery et al., 2001)
- Studies on ear and hearing disorders in ID people: German Special Olympics Summer Games 2004, 2006, 2008, 2012



Hearing screenings during Special Olympics games

performed by (professional) volunteers

Passage of six stations:

- (1) Check-in: personal data, history
- (2) Otoscopy and ear microscopy
- (3) DPOAE screening (2, 3, 4, 5 kHz) → also TEOAE

For those who had failed any of these stations →

- (4) Tympanometry screening
- (5) PTA (pure tone audiometry) screening for 2 and 4 kHz at 25 dB HL
- (6) Check-out: oral and written recommendations

Optional:

- (7) On-site fitting of hearing aids and individual water-protection ear moulds



Special Olympics Games 2004

Neumann K., Dettmer G., Euler H. A., Giebel A., Gross M., Herer G. Hoth S, Lattermann C, & Montgomery J (2006) Int J Audiol 45:83–90

755 athletes screened: 38% failed the screening in the German SO summer games in 2004, but noisy conditions

High prevalence for conductive hearing loss caused by chronic middle ear infections and blocking ear wax, moderate prevalence for sensorineural and mixed hearing loss



Special Olympics Games 2006

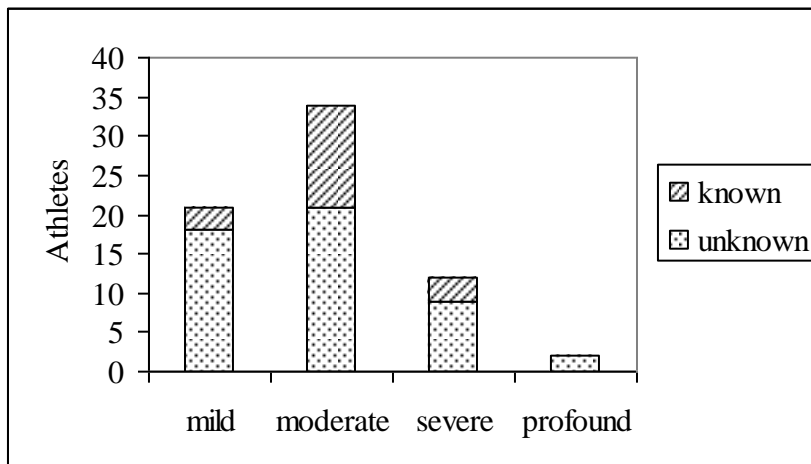
To quantify screening quality, screening PTA results were compared with those of a diagnostic PTA at 0.5, 1, 1.5, 2, 3, 4, and 6 kHz (air conduction)

→ 101 athletes who had failed the screening + 94 athletes who had passed the screening PTA, performed diagnostic PTA



Results peripheral hearing disorders (2006)

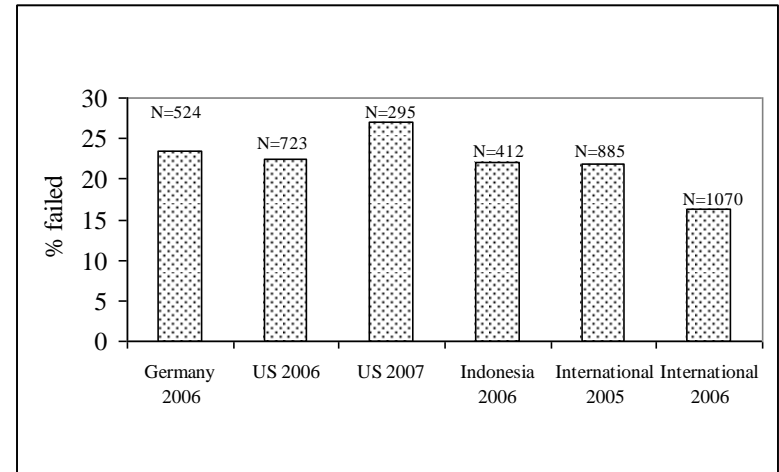
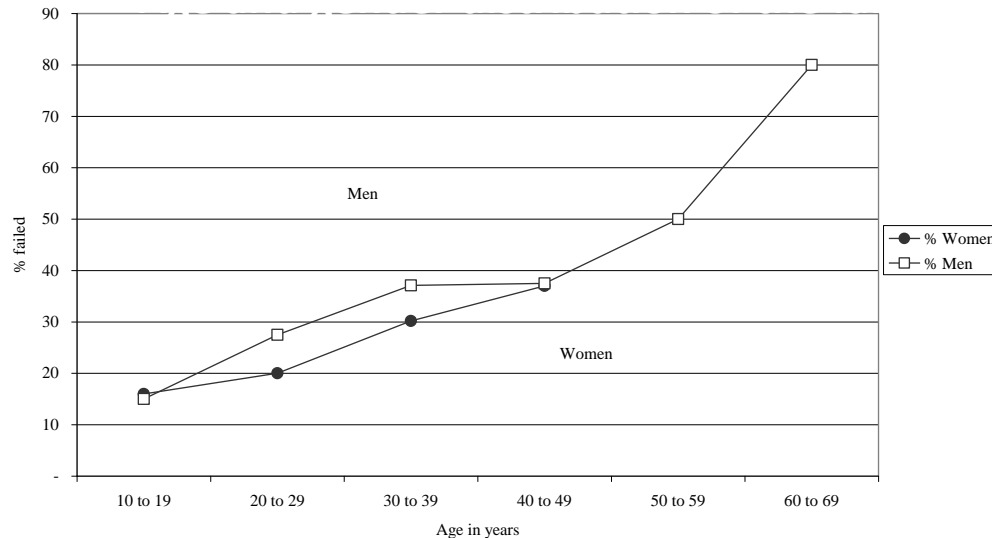
<i>Cases</i>	<i>Number (Percentage)</i>
Total	524 (100%)
Pass	401 (76.5%)
Fail	123 (23.5%)
<hr/>	
Fails	
Total	123 (100%)
Known hearing disorders	30 (24.3%)
Binaural	87 (70.7%)
Unilateral	36 (29.3%)
Sensorineural hearing loss	65 (52.8%)
Conductive hearing loss	26 (21.1%)
Mixed hearing loss	32 (26.0%)



- 42% → advice to consult an otolaryngologist or an acoustician
- 27% needed a regular ear canal control because of blocking ear wax
- 8 athletes wore hearing aids, 8 others did not wear them during the games
- bilateral hearing loss was mild (< 40 dB) in 31% of the fail cases, moderate (40-69 dB) in 49%, severe (70-94 dB) in 17%, and profound (>95 dB) in 3%.
- Of the 99 cases with hearing loss confirmed by a diagnostic PTA, 74% were unknown. An alarming 11 of the 14 cases with profound or severe hearing loss were undetected so far.
- Correlation between PTA screening and diagnostic PTA: Cramer's V index of 0.98 → screening identifies hearing loss reliably

Results peripheral hearing disorders

(Hild et al., J Int Disab Res, 2008)



No gender differences in the fail rates
Quicker aging of hearing in ID subjects

International results of Special Olympics
Healthy Hearing screenings: $\approx 25\%$ fail rate; no significant differences (χ^2 -test)

Cerumen

- High incidence of obturating cerumen in people with ID (Evenhuis, 1995); 28% compared with 2% - 6% in a non-disabled population (Crandell & Roeser, 1993)
- Causes: irregular ear canal shape, deficient self-cleaning mechanism, and digital or instrumental manipulation
- Obturating ear wax → ear canal irritations and subsequent manipulations, conductive hearing loss, reduced benefit from ear tubes and hearing aids
- → Recommendation: regular otolaryngological ear canal cleaning



Reality is...

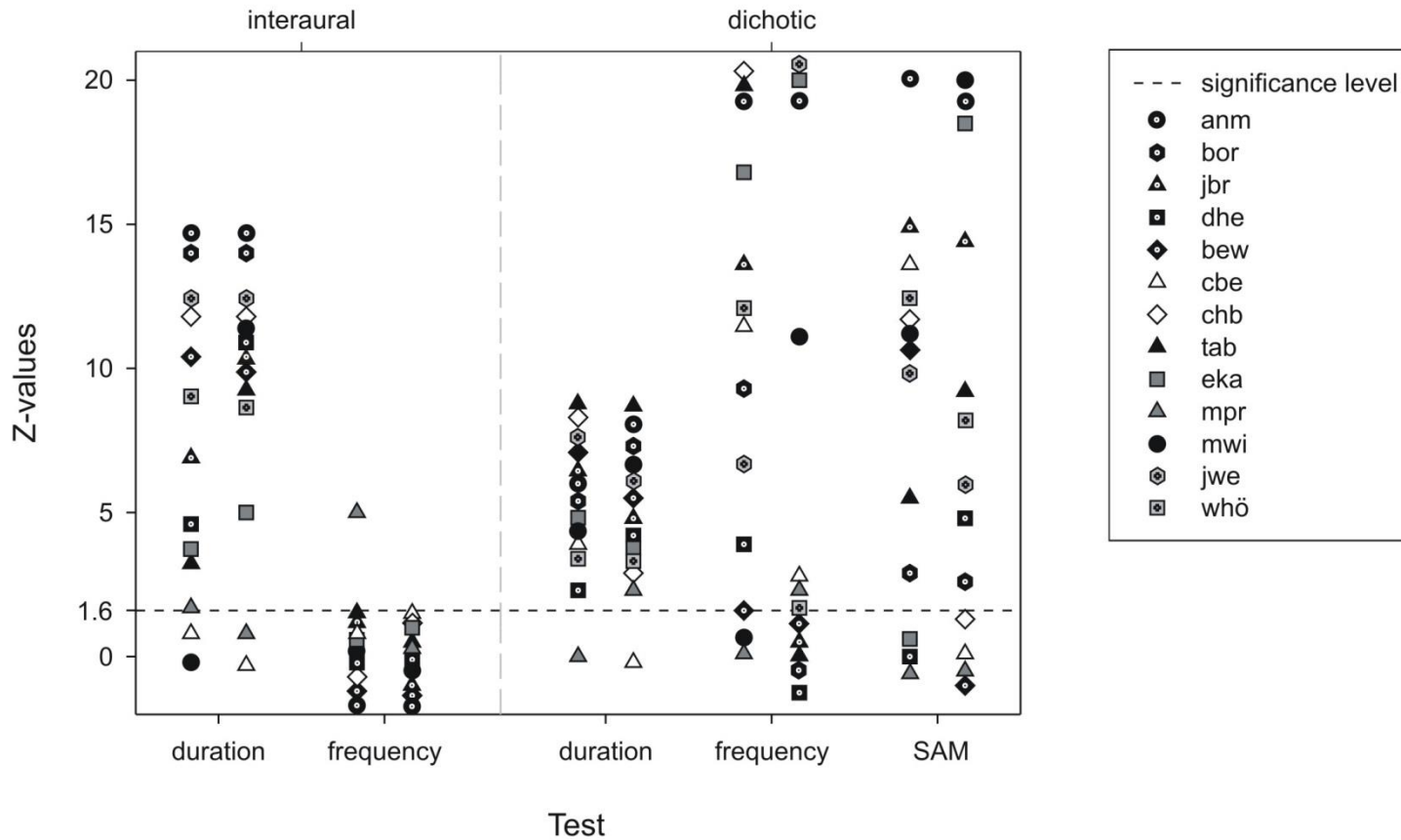
- High proportion of unknown or ignored hearing disorders
- **Alarming: high proportion of athletes with previously undetected profound hearing loss or deafness!!!**
- People with ID generally have communication handicaps at all → ear & hearing disorders should be detected early and treated appropriately
- But: therapy of hearing disorders in ID persons disappointingly deficient
- Hearing aids are rarely used
- Even most athletes with known hearing loss often did not get appropriate therapy
- Most of them had old hearing aids which needed repair or new fitting
- Athlete with a CI: no check-up and hearing rehab because his teacher of the deaf meant that is CI useless for ID people
- Additionally, central auditory processing problems have been shown to be present in all examined athletes with ID, also with no peripheral hearing loss (Neumann)

But...

- Early and continued therapy of hearing disorders is possible and beneficial
- Children with multiple disabilities benefit from CI (Waltzman et al, 2000)
- Medical or surgical treatment of chronic otitis media → normal hearing levels in 98% of Down syndrome children (Shott et al., 2001)
- Even at older ages treatment of these clients possible: after individual habituation training, majority of ID subjects >70 yrs. use hearing aids without difficulties (Evenhuis, 1995)

Difficulties in Central Auditory Processing (Neumann et al. 2013)

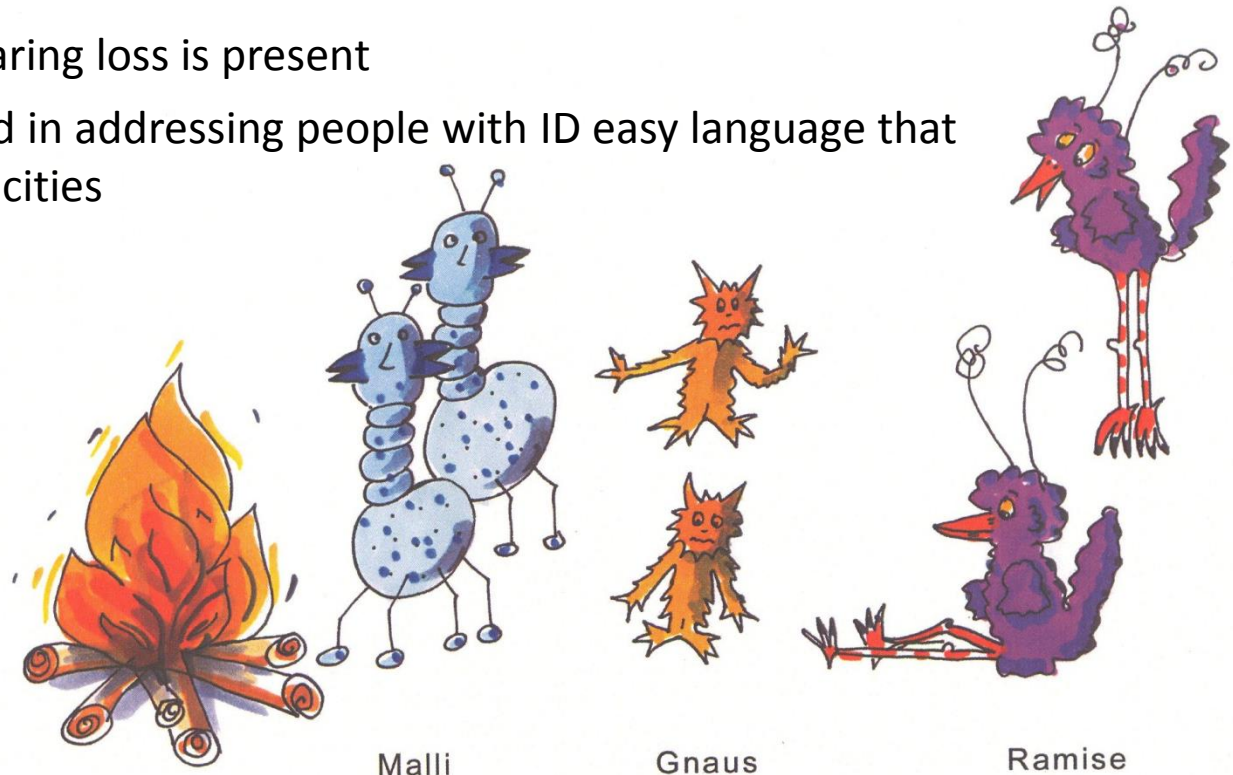
Deficits in temporal signal processing and partly in dichotic frequency discrimination



Language abilities (Neumann et al. 2013)

Grammatical abilities of persons with ID tested by assessing their ability to produce correct plural forms of nonsense nouns

- Level of plural acquisition far below that one of 4-year old children (Zaretsky et al. 2014)
- Language concept in people with ID mostly more simple than one expects from the everyday communication
- They often communicate out of the context or of their empirical knowledge
- Imitate
- Problem worsens if hearing loss is present
- This shall be considered in addressing people with ID easy language that really meets their capacities



Conclusion from screening studies

Peripheral Hearing:

About one quarter fails, in national and international SO hearing screenings
Screening reliably identifies hearing loss

Central auditory processing:

Disturbed in all subjects
Rather disturbed processing of temporal than of frequency information
More processing disturbances on auditory cortex than on brainstem level

Language

A simple language is necessary for communication and awareness for language impairment in people with ID

Because hearing loss is of high prevalence in people with intellectual disabilities and is rarely spontaneously expressed by them, a net of prevention, regular controls, and standard therapy programs as well as a higher awareness among professionals and caregivers is required.

EFAS Guidelines

- Intellectual disabilities (ID) are reported to have a high rate of co-morbidities, including hearing loss
- Prevalence of hearing loss in adults with ID have been estimated at 25-40% (Neumann et al. 2008, Hild et al. 2008)
- Multiple barriers to healthcare access for individuals with ID are widely reported (Hild)
- Despite the high prevalence of hearing loss in individuals with ID, strategies for hearing screening in this population currently varies widely or are, mostly, lacking at all.
- EFAS Working Group on Audiology and Intellectual Disability proposed guidelines for hearing screening across the lifespan for individuals with ID
- These recommendations provide standards for the conduct of an audiological intervention that represents, to the best knowledge of EFAS, the evidence-base and consensus on good practice given the stated methodology and scope of the document and at the time of publication
- Target audience:
 - medical and audiological professionals, e.g. audiologist, ENT, SLP, phoniaticians, pediatric audiologists, GP, paediatricians, psychologists, psychiatrists, pedagogues, and other people involved in ID-care
 - Stakeholders: local health providers, politicians

Definition of Intellectual Disability according to the World Health Organization

Intellectual disability means a significantly reduced ability to understand new or complex information and to learn and apply new skills (impaired intelligence). This results in a reduced ability to cope independently (impaired social functioning), and begins before adulthood, with a lasting effect on development.

Definition of Intellectual Disability according to the American Association on Intellectual and Developmental Disabilities

An Intellectual Disability (intellectual developmental disorder) is a disorder with onset during the developmental period that includes both intellectual and adaptive functioning deficits in the conceptual, social and practical domains. The following 3 criteria must be met (AAIDD 2010):

- Deficits in intellectual functions, such as reasoning, problem solving, planning, abstract thinking, judgement, academic learning and learning from experience, confirmed by both clinical assessment and individualized, standardized intelligence testing
- Deficits in adaptive functioning that result in failure to meet developmental and socio cultural standards for personal independence and social responsibility. Without ongoing support, the adaptive deficits limit functioning in one or more activities of daily life, such as communication, social participation, and independent living, across multiple environments, such as home, school, work and community
- Onset of intellectual and adaptive deficits during the developmental period.

Screening according to WHO criteria

Hearing Screening

- WHO “gold standard” principles that justify a mass screening first described 1968 by Wilson and Jungner.
- These criteria were adapted for hearing screening by Davis et al (1997) in “A critical review of the role of neonatal hearing screening in the detection of congenital hearing impairment.” This seminal work was instrumental in the implementation of newborn hearing screening.

Current Hearing Screening for Individuals with ID

- In 2016 the EFAS Audiology and ID Working Group sent a questionnaire to representatives from 27 European countries regarding current hearing screening provision for individuals with ID. Answers received from 22 countries: Only 2 countries reported having specific hearing screening programs however 7 reported including modifications to existing programs for individuals with ID and 10 reported targeted follow-up screening for individuals with ID for whom no issue was identified on the hearing screen.

Hearing Screen for Individuals with ID beyond the Newborn Period

- Hearing screening beyond the newborn period for individuals with ID necessary
- Need for considered and careful costing of hearing screening for individuals with ID for each healthcare model in which it is being considered being offered

Recommendations on frequency of screenings:
depend on a person's age, if a person has Down syndrome or not, and if a person wears already a hearing device or not

AUDIOLOGICAL CARE	PERSON WITH ID IN GENERAL	PERSON WITH DOWN SYNDROME	PERSON WITH ID ELIGIBLE FOR HEARING DEVICE
EARWAX REMOVAL	Annual	2x/year	2x/year
HEARING SCREENING	Neonatal Hearing Screening	Neonatal Hearing screening	Annual hearing evaluation (at preschool ages 2-4x per year, up to 10 years 2 x per year)
	Annual screening < age 6	2x/ year > age 6	
	Every 3 years from age 6 to 18	Every 2 years from age 6 to 18	
	Every 5 years from age 18 to 50	Every 3 years from age 18 and 35	
	Every 3 years > age 50	Annual > age 35	
	Annually if 8h/ day noise exposure (>80dBA)		

WHO criteria for implementing mass screenings (Wilson & Jungner, 1968)

1. The disorder to be screened for should be an important health problem

- High prevalence of hearing loss (25-40%)
- Impact of hearing loss on quality of life (Chiorba et al 2012)
- Implications of communication difficulties for those with an ID are greater than for the wider population (Wiley & Moeller 2007)
- In addition to the impact of undiagnosed hearing difficulty on the individual themselves, it is reported that behavior that challenges increases (Austin & Jeffrey 2007)

2. There should be an accepted rehabilitation means for cases of identified by the screen

- Awareness of care persons for a hearing problem may improve communication
- Benefits of hearing aids and hearing implants for individuals with IDs reported (Evenhuis et al 1995, Bent et al 2015)

WHO criteria for implementing mass screenings (Wilson & Jungner, 1968)

3. Facilities for assessment, diagnosis and rehabilitation should be available.

- In the event of a screening programme planned, audiology services should be accessible to individuals with IDs.
- For information on country specific Audiology provision see <http://globalaudiology.org/>

4. The disorder should be recognizable at an early stage.

- Progressive hearing loss is more frequently a gradual process than a sudden deterioration, so if a hearing screen is offered sufficiently frequently a hearing impairment could be identified at an early stage.

5. A suitable test should be available (quick, high sensitivity and specificity, easy to interpret)

- A variety of methods have been investigated for use in a hearing screen in this population (Driscoll et al 2002, Anderson et al 2013, Neumann et al. 2006, 2013).
- In light of the wide range of capacity observed in individuals with ID a flexible approach to hearing screening methods should be considered.
- It is strongly advised however that this does not take the form of a carer questionnaire which typically under-reports communication difficulties (Lavis et al 1997).
- Future guidance by the WG is planned regarding methodology.

WHO criteria for implementing mass screenings (Wilson & Jungner, 1968)

6. The screen should be acceptable to the client and their family/carers

- Evidence from high uptake of hearing screening at the Special Olympics would suggest that common methodology used to screen and assess hearing is generally acceptable to this population (Neumann et al. 2006, 2013, Hild et al. 2008).

7. The natural history of hearing impairment in individuals with IDs should be known and understood.

- Likelihood of late-onset and progressive losses is assumed to be higher in the population of those with ID related to the higher incidence of co-morbidities that can result in a coincidental hearing loss
- Literature review exploring the relationship between hearing loss and age and how this differed for individuals with ID (Bent et al. 2015): in addition to the much higher prevalence of hearing impairment in the population with ID, the onset of hearing loss associated with age for the population with ID and in particular those with Down Syndrome occurred considerably earlier than those without; hearing loss is reported in 59–68% of over 50 year olds with IDs and in 62–93% of over 50 year old adults with Down syndrome. This literature review also identified that 70% of adults with Down Syndrome over the age of 40 had significant hearing loss which had been undiagnosed before systematic testing.

WHO criteria for implementing mass screenings (Wilson & Jungner, 1968)

8. There should be an agreed policy on whom to treat as patients with hearing impairments

- See Table before

9. Finding cases of hearing impairment should be viewed as a continuous process

- The recommendation of repeated hearing screens being offered through-out the life span for individuals with a reduced ability to self-report facilitates this principle

10. The incidental harm of screening programs should be small in relation to overall benefits

- Evidence from hearing screening programs at the Special Olympics suggest that common screening methodology causes negligible harm related to overall benefit (Neumann et al. 2006, 2013, Hild et al. 2008).

12. There should be guidelines on how to explain screening results , together with transitional counselling support for family of clients who have been screened positively

- Since the advent of the newborn hearing screening programs, communicating the need for the hearing screen, the details of the process and the outcomes has been developed.
- A range of communication strategies for individuals with IDs have also improved over time.
- It is advised that easy read information about the screen is available and that local policy regarding consent is adhered to before any screening is carried out.

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Comments on these guidelines are welcomed and should be sent to the WG members

Thanks a lot for your attention!

