High prevalence of hearing disorders at the Special Olympics indicate need to screen persons with intellectual disability

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Abstract

Background  Persons with intellectual disabilities (ID) are at increased risk for hearing impairment which often remains undetected. If left untreated, such hearing impairments may worsen the social and communicative problems of these persons. The aims of this study are to determine the prevalence of hearing impairment, to specify type and degree of hearing loss, and to evaluate the sensitivity and specificity of the screening in this population.

Methods  During the German Special Olympics Summer Games 2006, 552 athletes with ID had their hearing screened according to the international protocol of Healthy Hearing, Special Olympics. This screening protocol includes otoscopy, measurement of distortion product otoacoustic emissions, and – if necessary – tympanometry and pure tone audiometry (PTA) screening at 2 and 4 kHz. Additionally, 195 athletes underwent a full diagnostic PTA. The results of the screening and diagnostic PTA were compared.

Results  Of the 524 athletes who completed the screening protocol, 76% passed and 24% failed it. Ear wax was removed in 48% of all athletes. 42% of the athletes were recommended to consult an otolaryngologist or an acoustician. Of the 99 athletes whose screening-based suspicion of a hearing loss was confirmed with diagnostic PTA, 74 had an undetected hearing loss. The correlation (Cramer’s V) between screening and diagnostic PTA was .98. The sensitivity of the screening was 100% and the specificity 98%.

Discussion  The screening reliably detects hearing disorders among persons with ID. The prevalence of hearing impairment in this population is considerably higher than in the general population, and the proportion of undetected hearing impairments is large, even among people with only mild and moderate ID, as examined in this study. Therefore, a screening is highly recommended, and special attention from caregivers and professionals as well as
regular hearing assessment and standard therapy programmes are required for persons with ID.

**Keywords** audiometry, hearing loss, hearing screening, intellectual disability, Special Olympics

**Introduction**

Because of improved socioeconomic conditions, intensive neonatal care and reduced mortality the prevalence of intellectual disabilities (ID) is constantly rising (Cooper et al. 2004). Because of the special health needs of persons with ID, this rise has an impact on primary healthcare services.

Persons with ID are less healthy than the general population (Schmid et al. 2004). For example, syndrome-related disorders such as epilepsy, motor problems and sensory disorders, but also inactivity-related diseases such as osteoporosis and cardiovascular diseases, as well as lifestyle-related problems in the area of nutrition and exercise, are reported (Piachaud et al. 1998; Van Schrojenstein Lantman-De Valk et al. 2000, 2007; Jansen et al. 2004). Many of these problems remain unidentified and untreated, not the least because ID hamper individuals’ ability to communicate their health status and to participate in decisions about their own health and well-being. Moreover, the knowledge of the carers regarding the medical history and the possible health problems of the individuals they are caring for are often insufficient. There is a need for external assistance for people with ID to access healthcare services (Kerr et al. 2003; Van Schrojenstein Lantman-De Valk 2005).

Persons with ID are at an increased risk for hearing impairment (Beange et al. 1999; Van Schrojenstein Lantman-De Valk et al. 2000; Evenhuis et al. 2001). There is a high prevalence for conductive hearing loss caused by chronic middle ear infections and ear wax blocking the canal, and a moderate prevalence for sensorineural and mixed hearing loss. Especially, persons with Down syndrome are at a greater risk for hearing loss (Roizen 1996; Van Allen et al. 1999; Van Buggenhout et al. 1999; Shott et al. 2001; Meuwese-Jongejeugd et al. 2006). Although epidemiological data about the population of intellectually disabled persons have increasingly been reported during the last 10 years, hearing problems frequently remain unidentified and untreated (Beange et al. 1999; Bogardus et al. 2003). Untreated hearing loss may lead to numerous social and psychological problems and a reduced quality of life even in persons without disabilities (Arlinger 2003; Chia et al. 2007). Therefore, the detection and treatment of hearing loss in persons with ID are more urgent.

The study presented here was conducted at the German Special Olympics Summer Games in Berlin in 2006. Special Olympics is the largest worldwide recreational sports programme for persons with ID (Dykins & Cohen 1996). The main aim of Special Olympics is to enable persons with such disabilities to develop physical fitness and to participate in training and competitions. A second aim is to create greater understanding for people with ID in an environment of equality, respect and acceptance.

The Healthy Athletes Program, founded in 1996, is a special health service for Special Olympics athletes. It includes the ‘disciplines’ of Fit Feet (podiatry), FUNfitness (physical therapy), Healthy Hearing (audiology), Health Promotion (information on health), Opening Eyes (optometry), MedFest (general status such as weight, height, blood pressure, etc.) and Special Smiles (dentistry). The objectives of the Healthy Athletes Program are: (1) to improve access and health care for Special Olympics athletes at event-based health screenings; (2) to make referrals to local health practitioners when appropriate; (3) to train healthcare professionals and students in the health professions about the needs and care of people with ID; (4) to collect, analyse and disseminate data on the health status and needs of people with ID; and (5) to advocate improved health policies and programmes for people with ID (Special Olympics 2006). Past examinations within the Healthy Athletes Program revealed that many athletes were medically untreated which indicates serious deficits in the medical care for people with ID in general. The Healthy Athletes Program, therefore, does not only serve the direct health service for the athletes but also provides insights for medical professionals about the prevalence and coincidence of additional physical problems. The screening data are entered into a worldwide database to address the improvement of the medical care for and research about the population of persons with ID.
The Healthy Hearing Program consists of a hearing screening protocol including otoscopy, measurement of otoacoustic emissions, tympanometry and pure tone audiometry (PTA), and special services, such as check-up of hearing aids and fitting of hearing aids when possible. It was performed in Germany for the first time in 2004. Of the 755 participating athletes, more than one-third failed the screening. This was a high failure rate compared with international results (Neumann et al. 2006). In order to evaluate and increase the diagnostic quality of this extensive screening, a diagnostic threshold PTA has recently been added to the Healthy Hearing Program.

The aims of this study are: (1) to determine the prevalence of hearing impairment in intellectually disabled populations more exactly than in former studies (Montgomery 2003; Montgomery et al. 2006; Neumann et al. 2006) by the inclusion of a diagnostic PTA; (2) to specify type and degree of hearing loss; and (3) to evaluate the sensitivity and specificity of the screening by comparing screening and diagnostic PTA.

Method
Participants

Of the 2700 athletes who participated in the German Special Olympics Summer Games 2006, 552 athletes (214 female, 338 male; age 10–69 years, mean 27 years) underwent a hearing screening. The participants had been recruited in schools, job training institutions or sport clubs. Medical exclusion criteria were limited to unstable physical conditions or reasons that would place the athlete at risk for injury (Carek et al. 2002). The diagnosis and severity of the ID were not reported for ethical reasons.

Procedure

The screening was performed on 4 days of sports competitions in three rooms. The PTA room was carpeted for subsonic noise reduction. A team of 28 professional volunteers, including phoniatricians, paediatric audiologists, otolaryngologists, audiologists, acousticians, speech-language pathologists and students of hearing aids acoustics carried out the screening. Most athletes had been informed about the possibility of the Healthy Athletes service in advance. Therefore, many of them came spontaneously single or in groups to participate in the screening. Additionally, several volunteers recruited participants on the spot.

The hearing screening procedure followed the international guidelines from Healthy Hearing (Herer & Montgomery 2006) and was performed at six stations: (1) check-in; (2) otoscopy and ear microscopy; (3) distortion product otoacoustic emission (DPOAE) screening; (4) tympanometry (middle ear) screening; (5) PTA-screening; and (6) check-out. Additionally, there was a diagnostic hearing threshold PTA station and two stations for research projects on DPOAE growth rates and tests of central auditory processing.

All participants were welcomed at the check-in desk at Station 1, and were informed about the course of the tests. The athletes and their caregivers were questioned about former ear and hearing problems. At Station 2, ears were inspected by otolaryngologists or by paediatric audiologists either by otoscopy or, in case of ear wax or suspect findings, by ear microscopy. If ear wax was detected which could hinder further screening, it was removed if possible. The status of the ear canal was documented as to be clear, partially blocked or blocked. The participant failed this station if there was blocking ear wax which was irremovable, or if anomalies of the outer or middle ear were found. The otoacoustic emissions were assessed at Station 3 by measuring DPOAE bilaterally at 2, 3, 4 and 5 kHz with the Cochlea Scan® (Fischer-Zoth Diagnosesysteme GmbH, Germering, Germany) according to the Healthy Hearing Standard protocol. For passing the screening, emissions for at least three of these frequencies had to be detected. The athlete finished screening if Stations 2 and 3 were completed with a pass.

For those athletes who had failed any of these stations, a tympanometry screening and a PTA screening were added. At Station 4, the pressure dependence of the eardrum impedance was registered with the MAICO MI 34 middle ear analyzer® (MAICO Diagnostic, Berlin, Germany). At Station 5, behavioural PTA screening was performed for each ear via headphones with the audiometer MAICO Ks5® (MAICO Diagnostic, Berlin,

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Germany) in audiometric test booths. After an initial training where the athletes were asked to raise their hand after hearing sounds at levels decreasing from 50 dB HL to 25 dB HL, hearing was tested at 2 and 4 kHz at 25 dB HL. The success of the initial training and the test performance were documented.

At each station, pass/fail results and possible recommendations for further testing were documented. Pass and fail criteria of the hearing screening are described in detail in Neumann et al. (2006).

At the check-out desk at Station 6 the athletes received a giveaway as reward. A written recommendation was provided to those athletes who had failed the screening, or to their caregivers. They were advised to consult an otolaryngologist for any of the following reasons: (1) regular check-up for ear wax; (2) problems of the outer ear canal; (3) problems in the middle ear; and (4) audiometric testing.

To quantify the quality of the screening, the screening PTA results were compared with those of a diagnostic PTA at 0.5, 1, 1.5, 2, 3, 4 and 6 kHz. Thus, 101 athletes who had failed the screening and an additional 94 athletes who had passed the screening PTA, performed diagnostic PTA. The latter served also to determine type and degree of hearing loss of the confirmed fail cases. Bone conduction audiometry separates conductive hearing losses from mixed losses, and would be necessary to identify the amount of conduction loss and, thereby, the type of a hearing impairment. A bone conduction audiometry was tried but too often was not feasible with our clientele in the bustling Special Olympics sports atmosphere. Because a maximum conductive hearing loss rarely exceeds an air-bone gap of 40 dB (Murphy 2000), a mixed hearing loss was assumed to be already likely if the PTA threshold was higher than 50 dB and the tympanometry screening failed. It has to be admitted that this cut-off criterion differentiates conductive from mixed hearing loss only sub-optimally.

Results

The hearing screening results of the 552 study participants are summarised in Table 1. For 28 athletes, the screening could not be finished, in 18 cases of those because of non-removable ear wax. Seventy-six per cent of the remaining 524 cases passed the screening. A fail was obtained in 23.5%, but only in 5.7%, a hearing disorder was known before. Sixteen of these athletes (3.1%) had been fitted with hearing aids, but only eight of them wore them during the games.

Table 1 Overall results of the hearing screening of athletes with intellectual disabilities during the German Special Olympic Summer Games 2006

<table>
<thead>
<tr>
<th>Cases</th>
<th>Number (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Screened athletes</td>
<td>552</td>
</tr>
<tr>
<td>Males</td>
<td>338</td>
</tr>
<tr>
<td>Females</td>
<td>214</td>
</tr>
<tr>
<td>Ear wax removal</td>
<td>265</td>
</tr>
<tr>
<td>Completed screenings</td>
<td>524 (100)</td>
</tr>
<tr>
<td>Pass</td>
<td>401 (76.5)</td>
</tr>
<tr>
<td>Fail</td>
<td>123 (23.5)</td>
</tr>
<tr>
<td>Possible cause:</td>
<td></td>
</tr>
<tr>
<td>Sensorineural</td>
<td>65 (12.4)</td>
</tr>
<tr>
<td>Conductive</td>
<td>26 (5.0)</td>
</tr>
<tr>
<td>Combined</td>
<td>32 (6.1)</td>
</tr>
<tr>
<td>Fail bilateral</td>
<td>87 (16.6)</td>
</tr>
<tr>
<td>Fail right ear</td>
<td>17 (3.2)</td>
</tr>
<tr>
<td>Fail left ear</td>
<td>19 (3.6)</td>
</tr>
<tr>
<td>Advised to consult otolaryngologist/</td>
<td>228 (41.3*)</td>
</tr>
<tr>
<td>acoustician, because of</td>
<td></td>
</tr>
<tr>
<td>Ear canal problems</td>
<td>16 (2.9*)</td>
</tr>
<tr>
<td>Middle ear problems</td>
<td>54 (9.8*)</td>
</tr>
<tr>
<td>Suspected sensorineural hearing loss</td>
<td>114 (20.7*)</td>
</tr>
<tr>
<td>Blocking ear wax</td>
<td>146 (26.4*)</td>
</tr>
</tbody>
</table>

* Here percentages of 552 cases (completed screenings plus 18 cases of un-removable blocking ear wax).
For 195 athletes a diagnostic PTA was performed in addition to the screening PTA at 2 and 4 kHz. Altogether 101 athletes who had failed the screening underwent a diagnostic PTA. A hearing loss was confirmed in 99 of them and was bilateral in 69 cases and unilateral in 30 cases. The bilateral hearing loss was mild (<40 dB) in 31% of the respective cases, moderate (40–69 dB) in 49%, severe (70–94 dB) in 17% and profound (>95 dB) in 3%. The unilateral hearing loss was mild in 37% of the respective cases, moderate in 40%, severe in 10% and profound in 13%. In 74% of these athletes the hearing loss was unknown until this screening event and thus untreated. An alarming 11 of the 14 cases with profound or severe hearing loss were undetected so far.

The correlation between PTA screening and diagnostic PTA yielded a Cramer’s V index of 0.98. Only two cases differed because of an outlier at 2000 Hz in the PTA screening. Of the 195 athletes, 96 persons passed and 99 persons failed the screening PTA. Using the outcome of the diagnostic PTA measurements as external reference criterion for the screening results (all screening methods pooled) and bearing in mind the restrictions of such a procedure, the sensitivity of the PTA screening on the basis of these data is 100% (95% confidence interval 98.1% to 100%), and the specificity is 98% (95% confidence interval 95.1% to 100%).

Hearing screenings with a comparable protocol were performed at a series of Special Olympic Games 2005 and 2006, US Games 2006 and 2007 and the Indonesian Games 2006 ranged from 16.3% (International 2006) to 27.4% (USA 2007), with sample sizes ranging from \( n = 295 \) to \( n = 1070 \). The 23.5% fail rate of the current study lies within that range, and is relatively close to the weighted mean of 20.3% or the median of 21.6% from all these games (total \( n = 3316 \)). Hence, from all the above named screenings, including the one reported here, a mean fail rate of 20.8% (total \( n = 3840 \)) results.

### Discussion

The fail rate of 24% of the hearing screening as obtained in this study is in line with the results of other national and international Healthy Hearing screenings. Mild and moderate hearing losses dominated in both screenings where the degree of hearing loss was determined (German National Games 2006, International Games in Nagano 2005). However, the proportion of severe or profound hearing loss was significantly higher at the German National Games 2006 than at the Nagano International Games (Sinha et al. 2006), probably because of sample differences. It may be possible that the populations of participants at national games contain a higher proportion of more severely intellectually disabled persons than the populations at international games. That sample or recruitment differences may account for differences in degree, and type of hearing loss is also suggested by the results of Evenhuis et al. (2001) and Timehin &
Timehin (2004) who reported an about-equal frequency of mild-moderate and severe-profound hearing loss among intellectually disabled people tested in residential homes. Obviously, participants of international games, of national games and persons with ID in residential homes belong to different populations. The majority of the participants of the games were able to comply with the PTA procedure, and such a competence may not be assumed for the same majority of persons in residential homes. Generalisations about prevalences across such populations can only be made with caution, which constitutes a limitation of the present study.

The novel feature of this study is the inclusion of a full diagnostic audiometric test of the hearing threshold, which enables a validation of the screening programme. It was performed with about one-third of the athletes in addition to the hearing screening. Only two of these 195 athletes showed a non-identical result because of outliers in the screening with 2000 Hz. The sensitivity and specificity indices of the screening were so satisfactory that the screening (including a PTA at 2 and 4 kHz) alone can be assumed to already deliver a valid diagnosis given good screening conditions. Thus, the fail rates from national and international competitions, which range from 16.3 to 27%, seem to reveal a realistic picture of the hearing situation of the Special Olympics athletes. Even though some persons with profound ID cannot provide responses in performance audiometry, and have to be diagnosed by objective audiometric methods, for the majority with a developmental age of 4 years or older, the PTA is an adequate screening and diagnostic instrument (Evenhuis et al. 2003). Out of all 524 screening participants in this study, only 10 had problems with the PTA test instructions. One important caveat, however, must be added: a bone conduction audiometry would have been necessary to identify the type of hearing loss with sufficient certainty, but this was not possible under the on-site conditions.

About one-fourth of the population with ID had a confirmed hearing loss, which is considerably more than the 4–6% in the general population of comparable age (Hesse & Laubert 2005). This problem is corroborated by the fact that occluding ear wax contributes to hearing problems particularly in persons with ID because of their frequent irregularities of ear anatomy, ear wax composition and ear wax expulsion, compared with the general population (Crandell & Roeser 1993). In almost half of the athletes, ear wax had to be removed.

About three-quarters of the bilateral hearing impairments were previously unknown and thus untreated. Reduced hearing is already a severe communication handicap for persons with unimpaired cognitive abilities. Even mild or moderate bilateral or unilateral hearing impairments negatively impact language, scholastic and social abilities in persons without ID (Culbertson & Gilbert 1986). In intellectually disabled persons, however, poor hearing is an even greater obstacle for verbal communication, social orientation and coping with daily problems.

The hearing loss of older persons with ID is particularly serious because the hearing loss associated with intellectual impairment is added on the general hearing loss with aging. The age-related hearing loss of persons with Down syndrome exceeds considerably the age-related hearing loss in the general population, even the one with other ID, and reaches almost 100% after the age of 60 years (Meuwese-Jongejeugd et al. 2006).

The 2006 replication of the German Special Olympics Healthy Hearing screening shows substantially better results than the 2004 event, where 38% of the athletes failed the screening (Neumann et al. 2006), compared with only 24% 2 years later. The difference can be attributed to two facts: (1) compared with 2004, the screening in 2006 was performed under markedly better extraneous noise protection; and (2) by inspection, the proportion of athletes with Down syndrome appeared to be smaller than in 2004. Persons with Down syndrome are at higher risk for hearing impairment than people with other kinds of ID (Evenhuis et al. 2001).

By the inclusion of a diagnostic PTA an immediate and on-the-spot fitting with hearing aids was possible. Ryals (2006, personal communication) reported that a follow-up of Special Olympics athletes who received the recommendation to consult an otolaryngologist or an acoustician resulted in a compliance of only 2%. The main reasons seem to be the reduced ability of persons with ID to report hearing problems and take initiatives for solving such problems. The transmission of information and
responsibility from one caregiver to another one is vulnerable, and caregivers are only indirectly affected by the problem. With an on-the-spot fitting with hearing aids, however, a more robust compliance and hence a greater coverage can be expected because treatment starts immediately and directly, and a further fitting adjustment is more probable if the persons are already equipped with hearing aids.

The considerations mentioned above ought to compel primary care providers for people with ID to arrange regular assessment of the hearing status of their charges. The check-ups should be regulated on national levels and financed by health insurance (Evenhuis 1996). An international consensus on early detection and diagnosis of hearing impairment in people with ID advises screenings in the first year of life, during school age and every 5 years after the age of 50 years (Evenhuis & Nagtzaam 1998). People with Down syndrome should be screened every 3 years during their entire life because of high incidence of middle ear problems, extensive ear wax and combined conductive-sensorineural hearing loss.

Care providers often are not aware of that people with mild and moderate ID, living in the community, having an increased risk of sensory impairments, and therefore, they do not feel responsible for screening or increased care for sensory problems. This study, however, shows clearly that this opinion is not justified. Because hearing loss is generally of high prevalence in people with ID, and is rarely spontaneously expressed by them, a net of prevention, regular examinations and standard therapy programmes, as well as a higher awareness among professionals and caregivers, is required.

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