American Journal on Intellectual and Developmental Disabilities

Global Data on Ear and Hearing Screening in an Intellectual Disability Population

--Manuscript Draft--

Manuscript Number:	AJIDD-D-20-00117R1
Article Type:	Research Report
Keywords:	Deafness; Down syndrome; hearing loss; intellectual disability; Otolaryngology
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Manuscript Region of Origin:	BELGIUM
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Abstract

Intellectual disability (ID) and hearing loss are frequent comorbid conditions, although otological problems often go unnoticed until picked up by screening. In the hearing program of Special Olympics (SO), athletes with ID are screened for otological problems. By retrospective analysis of all SO meetings between 2007 and 2017, more than 100,000 screenings could be included. Cerumen impaction was found in 40.7%, middle ear problems in 29.5% of those who failed hearing screening, and hearing loss confirmation in 26.9%. Prevalences for different world regions and country income groups are provided. The results emphasize the high prevalence of hearing loss in this ID population. Awareness among health care workers and active screening are required to reduce health disparities among this disadvantaged population.

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Introduction

Intellectual disability (ID) is defined by the World Health Organization (WHO) as a significantly reduced ability to understand new or complex information and to learn and apply new skills, resulting in a reduced ability to cope independently (WHO Europe, 2010). It starts before adulthood with a lasting effect on development, and is associated with impaired intelligence and social functioning. Intellectual disability is a frequent condition estimated to affect more than 1% of the world population, being higher in children compared with adults, and in low- and middle-income countries compared with high-income countries (Maulik et al., 2011). The health status of people with ID is generally poor, shown by increased morbidity and premature death among them (Tracy & McDonald, 2015). Different contributors might explain this poor health status: a higher number of health problems in persons with ID, some of which syndrome related, decreased self-care and health perception, less measures towards disease prevention, communication problems, encountered barriers in health care use, lack of specific medical knowledge in their environment, lack of contextual knowledge in health care providers… (Sharby, Martire & Iversen, 2015; Marks et al., 2018).

When focusing on audiological problems in people with ID as one of the items of health status, a high need for action becomes apparent. The combination of prevalences of ID (around 1%) (WHO Europe, 2010) and permanent hearing loss (6.1%) (Duthey, 2013) in the general population seems not in line with results of epidemiological studies, showing 8.2% of ~~hearing-impaired~~ children with hearing loss having ID (Szymanski et al., 2012) and generally 30% of adults with ID exhibiting hearing loss, with even up to 100% in certain subgroups (Meuwese-Jongejeugd et al. 2006). Moreover, hearing loss in persons with ID often remains undetected without active hearing screening, demonstrated by the latter study in which hearing loss had not been diagnosed prior to screening in about half of the participants (Meuwese-Jongejeugd et al., 2006). ~~The burden of untreated hearing loss is high and persists due to~~ Not only the lack of awareness and poor diagnosis ~~amongst others~~add to the high and persistent burden of untreated hearing loss, but also the lack of epidemiological data, social and medical disparities, underuse of hearing devices, noise exposure and aging (Duthey, 2013). In addition to hearing loss, excessive and/or impacted cerumen is common in the ID population compared with the general population (Crandell & Roeser, 1993), as well as middle ear problems, especially in the subset of people with Down syndrome (DS) (Maris et al., 2014).

In order to improve the health status in children as well as in adults with ID, the international organization of Special Olympics (SO), a global sports organization for athletes with an ID that is represented in over 180 countries, provides a complimentary health program for persons with ID ~~people~~ in addition to their sports competitions (Special ~~Olypics~~Olympics – Inclusive Health, ~~2020~~2021). During sports events, athletes with ID can voluntarily participate in a health screening program, including physical therapy screening, podiatry, optometry, oral health, emotional health, prevention and nutrition, and audiology. In the latter, known as the SO Healthy Athletes - Healthy Hearing program, outer and middle ~~ears~~ear status and hearing of athletes are screened by trained professionals and students in health care. Follow-up recommendations are provided. The aim is to identify specific otological problems and hearing loss in people with ID, to refer ~~to~~for appropriate medical care, to increase access to hearing health care for this population, and to raise awareness of the hearing concerns of people with special needs, including difficulties in diagnosis and access to treatment options. Results of these screenings from a limited number of athletes have already been published, each demonstrating a high occurrence of sensorineural hearing loss, middle ear problems and ~~ear wax~~earwax impaction (Hey et al., 2014; Hild et al., 2008; Kumar Sinha et al., 2008; Neumann et al., 2006; Pradhan, Stormon & Lallo, 2019; Starska & Lukomski, 2006). The largest study to date in this population is that of Herer (Herer, 2012), studying 9,961 international athletes and confirming these high prevalences of otological and audiological problems. In his study, the lack of demographic data of the included athletes in order to better interprete the results was mentioned. Apart from age and gender, the country of origin seems of importance as health conditions among different countries and regions might be highly variable and the prevalence of hearing loss proved to differ among different regions, being highest in South and East Asia and sub-Saharan Africa (Duthey, 2013).

~~In this~~ The purpose of the study~~, we want~~ is to add to existing knowledge by studying the results of over 100,000 hearing screenings in persons with ID ~~people~~ at SO and address the limitations of previous studies by including demographics of the participants. In the discussion, ~~we will compare our~~ the results will be compared with available data from other groups of persons with ID ~~people~~ in order to examine how hearing loss is represented for the whole ID population. ~~Our~~The ultimate aim is to increase awareness among caregivers, health care practitioners and policy-makers about the high prevalence of otological and audiological problems in the ID population.

Materials and Methods

Data Collection

The ~~presented~~present data were obtained at worldwide national and international sporting events of SO between 2007 and 2017 (included). During these events, the Healthy Athletes health screening program made up of different health disciplines, including otological and audiological screening, was offered to the athletes on a voluntary basis. All participants had an ID and their minimal age was 8 years. No data about ID type or severity were available. Prior to participation, written informed consent was obtained by the athletes and/or their legal guardian. In addition, oral consent was obtained from the athlete at the start of the screening. All data were de-identified and made available by SO International in view of this report. After quality control with incomplete or unclear items considered missing, data were transferred to an SPSS database (SPSS Statistics version 26.0, IBM Corp, Armonk (NY), USA). Regarding the demographics, age was categorized in decades, and countries of origin were classified according to the SO regions (Figure 1). Moreover, the countries were grouped into the four World Bank income groups (low, lower-middle, upper-middle, and high) (World Bank, 2019). Statistical analysis included chi square test with the application of Bonferroni correction where appropriate. In full accordance to the Helsinki Declaration, the Ethics Committee of the author's institution approved the retrospective analysis (EC 115-2017/MF).

Hearing Screening at Special Olympics

The SO Healthy Hearing program uses an international standardized screening protocol developed and well referred to in literature by SO International (Herer & Montgomery, 2006). This protocol was strictly followed by trained professional volunteers (audiologists, speech/language pathologists, medical doctors) supported by trained health care students, and is summarized in Figure 2~~.~~; the updated manual can be found online (Special Olympics –

Healthy Hearing, 2021). Registration of demographic information (gender, age, nationality) was performed at check-in~~ was followed by handheld~~. Handheld otoscopy was subsequently performed and ~~eventual~~ cerumen removal, ~~the latter being~~if necessary and after approval of the athlete, was performed at many ~~but not all~~ screening events.

Next, hearing screening by ~~a~~ distortion product otoacoustic emissions (DPOAE) ~~test~~testing was performed. DPOAE reflect the integrity of the outer hair cells of the inner ear, can be easily and rapidly recorded and are sensitive to hearing loss. Consequently, DPOAE testing is frequently used in screening setting, such as universal newborn hearing screening programs. It was performed in a quiet space, recorded between between 2 and 5 kHz ~~was performed~~ independent of cerumen presence, and resulted in ~~pass~~'pass' or 'no ~~pass~~pass' for each ear. A pass was given when a signal-to-noise ratio of 6 dB or more was obtained for at least three out of the four tested frequencies. If a pass on both ears was obtained, the test protocol was terminated.

In case DPOAE testing resulted in no pass in one or both ears, tympanometry (screening for middle ear problems) and pure tone audiometry (PTA, hearing confirmation) were performed. ~~Tympanometry~~Tympanometry is a short test to evaluate the middle ear function including the tympanic membrane and ossicles by creating air pressure variations. It led to a normal or abnormal result, with normal signifying an admittance between 0.20 and 2.00 mmho, middle ear pressure between +20 and -200 daPa and an ear canal volume between 0.60 and 2.00 ml.

~~PTA also~~PTA is a subjective test measuring ear-specific hearing thresholds using supra-aural headphones. It was also performed in a quiet room and led to pass or no pass based on screening at 2 and 4 kHz at 25 dB HL, after conditioning with a more intense sound. Only in case of no pass at one or both frequencies, determination of air and bone conduction thresholds at different frequencies was offered to the athlete if available, in order to further elaborate the hearing loss and to discriminate between conductive and sensorineural hearing loss. At check-out, results were discussed with the athlete and guardian and a report with the findings and follow-up suggestions was issued. Referral was provided based on a no pass on PTA screening for one or both frequencies in one or both ears, or based on otoscopic or tympanometric findings (excessive cerumen or pathological findings, abnormal middle ear) in the primary language of the athlete. The type of eventual hearing loss (conductive/mixed or sensorineural) was determined based on otoscopic and tympanometric findings as well as on air and bone conduction thresholds if available.

Results

Demographics

Data of 106,369 screenings conducted between 2007 and 2017 (included) were available. Of these, 37.8% were of female athletes and 62.2% were male. The majority of athletes (70.3%) were aged 8-29y, with 40.8% younger than 20y. Distribution among the different SO regions can be found in Figure 1. According to the World Bank country-specific data, 75.1% of the athletes came from a high-income country, whereas the percentages for low-income, lower-middle-income and upper-middle-income countries were 0.6%, 7.6% and 16.7%, respectively. The distribution of gender and age among the different regions and income groups is provided in Table 1. In all regions, less women than men participated at the hearing screening, with the highest representation of women in North America (40.8%) and Africa (38.1%). Regarding age, the youngest group (<20y) was the largest in all regions, especially in Africa (76.2%). The middle-aged and older adults were best represented in North America (19.2% ≥40y) and Europe/Eurasia (15.9%).

Screening Otoscopy and DPOAE Testing

Even after eventual removal of cerumen~~ (only if the athlete agreed and rather reluctant because of suboptimal conditions without suction aid or otomicroscope),~~, 40.7% had at least one ear canal that was not clear, with 24.6% partially blocked (tympanic membrane only partially visible due to cerumen impaction) and 16.1% compeletely blocked (no tympanic membrane visible due to cerumen impaction). More left ear than right ear cerumen obstruction was observed (21.2% versus 20.8% partial obstruction and 11.9% versus 11.6% complete obstruction for left and right ear respectively, p<0.001). In addition to cerumen, foreign bodies such as ~~earbuds~~the tip of an earbud, parts of toys and pencil points were also removed from the ear canal.

Subsequent DPOAE testing yielded a pass for both ears in 47.4% of the participants, so in 52.6% at least one ear failed this screening. Again, more refers were seen in left ears than in right ears (45.5% versus 43.7~~%),~~%, $p<0.001$), which is ~~in relation~~consistent with the otoscopy as ear canal obstruction ~~might influence~~will impact DPOAE ~~test~~screening results. Of interest, DPOAE testing resulted in a pass in 54.4% of the athletes with clear ear canals, in 41.0% with partially blocked ear canal and in 31.0% with completely blocked ear canal. Figure 2 depicts the screening protocol together with the main prevalences of the screening results.

Elaboration of Screening Failures

The athletes not having a pass on DPOAE for one or both ears (52.6%) were subsequently tested with tympanometry. A normal tympanogram for both ears was obtained in 64.4% of the referred participants, whereas a no pass was found in 27.3% and 27.7% of the tested right and left ears respectively (right and left combined: 35.6% of the referred participants). After comparison of these results with the otoscopy findings, middle ear problems were suspected based on abnormal tympanometry in 29.5% of the referred participants (21.9% right ears and 22.0% left ears).

In addition to tympanometry, PTA screening was performed in the ears with DPOAE failure to confirm or refute the DPOAE result. In contrast with the DPOAE data, 48.9% showed normal hearing on PTA screening, but hearing loss was confirmed in 51.1% of the referred participants (42.5% right ears and 45.2% left ears, only DPOAE screening failure ears were measured), which is 26.9% of the total group of participants.

Based on a combination of the previous tests and air and bone conduction threshold testing in a selection of athletes, the origin of hearing loss could be estimated with 67.8% of the ~~hearing-impaired~~ participants with hearing loss exhibiting permanent sensorineural hearing loss and 32.2% conductive hearing loss or at least a conductive component.

Reasons for Referral

~~A first~~The initial reason for referral was ~~the~~cerumen obstruction of the ear ~~canals~~canal. In all participants with partial or complete obstruction in at least one ear (40.7%), follow-up was advised. A second reason was a middle ear problem as shown by an abnormal tympanogram not explained by ear canal findings during otoscopy (29.5% of DPOAE referred athletes). A third referral reason was the confirmation of hearing loss, irrespective of the type (26.9%). As a combination of the above-mentioned findings is frequent, the total referral rate for participants was 57.7%. Referral rates for these reasons were compared with the demographics. Gender differed significantly for cerumen obstruction (39.0% refer in women, 41.8% in men, $p<0.001$) and overall referral rate (56.4% refer in women, 58.4% in men, $p<0.001$), but not for middle ear problems (29.5% refer in women failing DPOAE, 29.5% in men failing DPOAE, $p=0.95$) and hearing loss (26.7% refer in women, 27.0% in men, $p=0.29$). Comparison of the age groups for the different referral reasons is shown in Figure 3, as well as the comparison of SO regions and World Bank income groups (World Bank, 2019) for referral reasons. Significant differences for each of the referral reasons among the different age groups are found, as well as among the SO regions ans World bank income groups, even after Bonferroni correction ($p<0.001$ for group comparisons).

Discussion

~~Otological~~Ear and ~~audiological~~hearing problems ~~seem to be~~are common in the ID population, as shown by the results of this study. Significant cerumen impaction was present in more than 40%, with total obstruction in 16%. Occasionally, in addition to cerumen, foreign bodies needed to be removed out of the external ear canals. Middle ear problems, including chronic otitis media, abnormal middle ear pressure, and tympanic membrane perforation or retraction, were detected in about 30% of those who failed hearing screening by OAEs. Hearing loss, based on screening with OAEs and subsequent PTA screening, proved to be present in 27%. Of course, some of these are ~~supposed~~thought to be temporary conductive losses based on the cerumen impaction or middle ear problems, but in two out of three ~~hearing-impaired~~ participants with confirmed hearing loss, permanent sensorineural hearing loss was ~~most likely~~detected by pure tone audiometry. The results shown refine the numbers mentioned in previous reports of screening in a limited number of SO athletes. In these studies, cerumen impaction ranged from 33% to 53%, middle ear problems from 6% to 21%, and hearing loss from 2% to 38% (Herer, 2012; Hey et al., 2014; Hild et al., 2008; Kumar Sinha et al., 2008; Neumann et al., 2006; Pradhan, Stormon & Lallo, 2019; Starska & Lukomski, 2006). Sensitivity and specificity of the SO approach has proven highly sensitive and specific in the detection of hearing loss (Hild et al., 2008).

Certain demographics seem to influence the results of ~~ear and hearing~~otologic and audiologic outcome. Female athletes were less prone to cerumen impaction compared to male athletes, but middle ear problems and hearing loss did not differ among women and men. With advancing age, a stepwise increase in cerumen impaction (from 38% to 47%), middle ear referral (from 13% to 25%) and hearing loss (from 17% to 69%) can be seen. In the oldest age group (≥60 years of age), the referral rate irrespective of the etiology was 84%. Regional differences were also apparent: cerumen impaction was highest in East Asia and North America (both 42.2%), whereas middle ear referrals were higher in Asia Pacific and Europe/Eurasia (both 16%). Hearing loss was highest in North America (29%) and Asia Pacific (27%), especially in comparison with Latin America (15%). South and East Asia, together with sub-Saharan Africa, are regions known to have a higher prevalence of hearing loss both in children and adults, mainly because of environmental factors such as infections and noise exposure (Duthey, 2013). The high prevalence of hearing loss among athletes in North America can be explained by the higher number of older participants in this region compared with other regions. Taking into account the income groups, cerumen impaction and middle ear referral had the highest occurence in low-income countries (46% and 19% respectively). A possible explanation might be the inclusion of a higher number of SO events in high-income countries, which implies that several athletes might have participated at several events over time and consequently have been treated during a previous screening. ~~We~~It might also ~~hypothesize~~be hypothesized that the health care availability contributes to this difference, again pointing towards the importance of screening at such events. In contrast, hearing loss was highest in high-income countries (29%), but again the higher age of the partipants in these countries should be taken into account.

The population studied in this report is rather specific: SO participants are ~~sportive~~able to participate in sporting events and have usually mild or moderate ID. In order to assess the representativeness of ~~our~~the numbers for the whole ID population, a comparison with other ID groups is necessary. Previously, the SO population was compared with a group of ~~ID~~ children and adolescents with ID of special needs schools, showing no significant difference in hearing loss prevalence (Hey et al., 2014). The younger age in the school study might contribute to the absence of difference, as the prevalence of hearing loss is influenced by age to a high degree and by ID severity to a lower degree (Evenhuis et al., 2001). In a group of institutionalized children with profound ID, hearing loss was present in 32% (Stein et al., 1987). The prevalence of bilateral hearing loss in an institutionalized ID population with a higher age was 47%

(Evenhuis et al., 2001). Finally, a large study in ~~ID~~ service users with ID, in which measures to approach representativeness for the whole ID population were taken, concluded a weighted population hearing loss prevalence of 30.3% (Meuwese-Jongejeugd et al., 2006). Based on the above, ~~our~~the numbers of hearing loss are in the same order compared with previous studies and might be an underestimation of the actual prevalence given a rather young profile of SO participants with a milder ID grade than the general ID population. Most studies do not report on the type of hearing loss. ~~Our~~The observed ~~ratio~~distribution of conductive versus sensorineural hearing loss (32% versus 68%) is similar to the scarce data in literature (38% versus 62%) (Stein et al., 1987), although the latter being in a pediatric ID population. A limitation of the current study inherent to its setting is the audiometric tests being performed in a separate quiet room with the lowest ambient noise level and thus not in a soundproof booth. This might especially impact threshold determination and thus type and degree of hearing loss.

A specific subgroup of persons with ID that is mentioned in several studies because of its higher hearing loss prevalence, is DS (Evenhuis et al., 2001; Meuwese-Jongejeugd et al., 2006). The elevated occurence of hearing loss can be attributed to both middle ear problems and sensorineural hearing loss. At certain age intervals, otitis media peaks to 60% in ~~DS~~ children with DS (Maris et al., 2014). Sensorineural hearing loss is low among younsters with DS (De Schrijver et al., 2019) but increases with age to 65% of the ~~DS~~ adults with DS, especially in the higher frequencies. The high-frequency hearing loss of DS resembles premature aging of the hearing system (Picciotti et al., 2017). ~~We do not have information~~Information of the proportion of ~~DS~~ athletes with DS in ~~our~~the studied population is lacking, but ~~neither have~~there are no arguments for a participation bias in this respect.

With these results, ~~we want to stress~~ health issues and more specifically the high prevalence of hearing loss and otologic problems in persons with ID~~.~~ are stressed. Screening is important as self-report is lower and hearing loss might be masked by the appearance of the ID when not actively searched for. Diagnosis is often more difficult to establish in these individuals~~,~~ (Diefendorf et al., 2017), albeit no reason to refrain from proper testing. Patience, adaptation to the situation and professional experience in testing people with special needs can help in obtaining an accurate diagnosis~~. However,~~ and should be adopted in ~~a prestation driven~~ the health care ~~system this can be difficult. Objective testing, especially~~setting. Especially in persons with severe to profound ID, objective testing might be considered~~.~~, such as auditory brainstem response audiometry in which brainstem signals are recorded in response to sound without needing the person's active cooperation. Although the burden of hearing loss in persons with ID ~~people~~ and the societal effect of treatment is difficult to estimate, appropriate individual treatment should be initiated upon diagnosis. Treatment failure, especially rehabilitation with hearing aids, is possible and service provision to this specific group must be patient-tailored (Meuwese-Jongejeugd, Verschuure & Evenhuis, 2007). More research on the outcome following screening failure and on the assessment of treatment effect with its determinants is required.

In conclusion, health care workers involved in ID should be aware of the high prevalence of hearing loss in this population. Because of low self-report and masking by comorbidities, active screening ~~seems~~is mandatory. The health program of SO is able to screen a large number of athletes with ID, confirming the increased need for active otological and audiological follow-up in the far majority of the participants.

References

Crandell CC & Roeser RJ (1993). Incidence of excessive/impacted cerumen in individuals with mental retardation: A longitudinal investigation. Am J Ment Retard, 97(5): 568-574.

De Schrijver L, Topsakal V, Wojciechowski M, et al (2019). Prevalence and etiology of sensorineural hearing loss in children with down syndrome: A cross-sectional study. Int J Pediatr Otorhinolaryngol, 116: 168-172.

Diefendorf AO, Corbin KR, Trepcos-Klinger R, et al (2017). Audiologic considerations for children with complex developmental conditions. In Tharpe AM & Seewald R (Eds.), Comprehensive handbook of pediatric audiology (2nd ed., pp. 609–628). Plural: San Diego (CA, USA).

Duthey B (2013). Background paper 6.21 - Hearing loss. WHO: Geneva (Switzerland).

Evenhuis HM, Theunissen M, Denkers I, et al (2001). Prevalence of visual and hearing impairment in a Dutch institutionalized population with intellectual disability. J Intellect Disabil Res, 45(Pt 5): 457-464.

Herer GR (2012). Intellectual disabilities and hearing loss. Commun Disord Q, 33(4): 252-260.

Herer GR & Montgomery JK (2006). Special Olympics Healthy Hearing Program: Guidelines for standardized screening procedures, 2nd ed. Washington (DC): Special Olympics.

Hey C, Fessler S, Hafner N, et al (2014). High prevalence of hearing loss at the special olympics: is this representative of people with intellectual disability? J Appl Res Intellect Disabil, 27(2): 125-133.

Hild U, Hey C, Baumann U, et al (2008). High prevalence of hearing disorders at the Special Olympics indicate need to screen persons with intellectual disability. J Intellect Disabil Res, 52(Pt 6): 520-528.

Kumar Sinha A, Montgomery JK, Herer GR, et al (2008). Hearing screening outcomes for persons with intellectual disability: a preliminary report of findings from the 2005 Special Olympics World Winter Games. Int J Audiol, 47(7): 399-403.

Maris M, Wojciechowski M, Van de Heyning P, et al (2014). A cross-sectional analysis of otitis media with effusion in children with Down syndrome. Eur J Pediatr, 173(10): 1319-1325.

Marks L, Wong A, Perlman S, et al (2018). Global oral health of athletes with intellectual disabilities. Clinical Oral Investigations, 22: 1681-1688.

Maulik PK, Mascarenhas MN, Mathers CD, et al (2011). Prevalence of intellectual disability: a meta-analysis of population-based studies. Res Dev Disabil, 32(2): 419-436.

Meuwese-Jongejeugd A, Verschuure H & Evenhuis HM (2007). Hearing aids: expectations and satisfaction of people with an intellectual disability, a descriptive pilot study. J Intellect Disabil Res, 51(Pt 11): 913-922.

Meuwese-Jongejeugd A, Vink M, van Zanten B, et al (2006). Prevalence of hearing loss in 1598 adults with an intellectual disability: cross-sectional population based study. Int J Audiol, 45(11): 660-669.

Neumann K, Dettmer G, Euler HA, et al (2006). Auditory status of persons with intellectual disability at the German Special Olympic Games. Int J Audiol, 45(2): 83-90.

Picciotti PM, Carfì A, Anzivino R, et al (2017). Audiologic assessment in adults with Down syndrome. Am J Intellect Dev Disabil, 122(4): 333-341.

Pradhan A, Stormon N & Lalloo R (2019). Oral and aural problems in Australian Special Olympics athletes. Spec Care Dentist, 39(5): 478-484.

Sharby N, Martire K & Iversen MD (2015). Decreasing health disparities for people with disabilities through improved communication strategies and awareness. Int J Environ Res Public Health, 12(3): 2201-3316.

Special Olympics – Healthy Hearing [Internet]. Washington (DC): Special Olympics [cited 2021 April 19]. Retrieved from: resources.specialolympics.org/health/healthy-hearing/

Special Olympics - Inclusive health [Internet]. Washington (DC): Special Olympics [cited ~~2020 October 8~~2021 April 19]. Retrieved from: www.specialolympics.org/our-work/inclusive-health/

Starska K & Lukomski M (2006). Realization of International Healthy Hearing Program in Poland--hearing evaluation in participants of Special Olympics. Adv Med Sci, 51: 197-199.

Stein LK, Kraus N, Ozdamar O, et al (1987). Hearing loss in an institutionalized mentally retarded population. Identification by auditory brainstem response. Arch Otolaryngol Head Neck Surg, 113(1): 32-35.

Szymanski CA, Brice PJ, Lam KH, et al (2012). Deaf children with autism spectrum disorders. J Autism Dev Disord, 42(10): 2027-2037.

Tracy J & McDonald R (2015). Health and disability: partnerships in health care. J Appl Res Intellect Disabil, 28(1): 22-32.

WHO Europe - Definition: intellectual disability [Internet]. Geneva (Switzerland): World Health Organization [cited 2020 October 8]. Retrieved from: https://www.euro.who.int/en/health-topics/noncommunicable-diseases/mental-health/news/news/2010/15/childrens-right-to-family-life/definition-intellectual-disability/

World Bank [Internet]. Washington (DC): World Bank Data Team; [updated 2019 July 1; cited 2020 October 8]. Retrieved from: https://blogs.worldbank.org/opendata/new-country-classifications-income-level-2019-2020/

Tables

Table 1: Distribution of gender and age among the different Special Olympics regions and World Bank country income groups. The absolute numbers are shown, followed by the percentage of gender or age group for the specific region/income group.

Figures

Figure 1: Distribution of athletes over Special Olympics regions.

Figure 2: Summary of screening protocol in black, and prevalences for the different findings in grey.

Figure 3: Prevalences of the different reasons for referral compared with age groups (panel A), Special Olympics regions (panel B) and World Bank income groups (panel C).

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Intellectual disability (ID) is defined by the World Health Organization (WHO) as a significantly reduced ability to understand new or complex information and to learn and apply new skills, resulting in a reduced ability to cope independently (WHO Europe, 2010). It starts before adulthood with a lasting effect on development, and is associated with impaired intelligence and social functioning. Intellectual disability is a frequent condition estimated to affect more than 1% of the world population, being higher in children compared with adults, and in low- and middle-income countries compared with high-income countries (Maulik et al., 2011). The health status of people with ID is generally poor, shown by increased morbidity and premature death among them (Tracy & McDonald, 2015). Different contributors might explain this poor health status: a higher number of health problems in persons with ID, some of which syndrome related, decreased self-care and health perception, less measures towards disease prevention, communication problems, encountered barriers in health care use, lack of specific medical knowledge in their environment, lack of contextual knowledge in health care providers… (Sharby, Martire & Iversen, 2015; Marks et al., 2018).

When focusing on audiological problems in people with ID as one of the items of health status, a high need for action becomes apparent. The combination of prevalences of ID (around 1%) (WHO Europe, 2010) and permanent hearing loss (6.1%) (Duthey, 2013) in the general population seems not in line with results of epidemiological studies, showing 8.2% of children with hearing loss having ID (Szymanski et al., 2012) and generally 30% of adults with ID exhibiting hearing loss, with even up to 100% in certain subgroups (Meuwese-Jongejeugd et al. 2006). Moreover, hearing loss in persons with ID often remains undetected without active hearing screening, demonstrated by the latter study in which hearing loss had not been diagnosed prior to screening in about half of the participants (Meuwese-Jongejeugd et al., 2006). Not only the lack of awareness and poor diagnosis add to the high and persistent burden of untreated hearing loss, but also the lack of epidemiological data, social and medical disparities, underuse of hearing devices, noise exposure and aging (Duthey, 2013). In addition to hearing loss, excessive and/or impacted cerumen is common in the ID population compared with the general population (Crandell & Roeser, 1993), as well as middle ear problems, especially in the subset of people with Down syndrome (DS) (Maris et al., 2014).

In order to improve the health status in children as well as in adults with ID, the international organization of Special Olympics (SO), a global sports organization for athletes with an ID that is represented in over 180 countries, provides a complimentary health program for persons with ID in addition to their sports competitions (Special Olympics – Inclusive Health, 2021). During sports events, athletes with ID can voluntarily participate in a health screening program, including physical therapy screening, podiatry, optometry, oral health, emotional health, prevention and nutrition, and audiology. In the latter, known as the SO Healthy Athletes - Healthy Hearing program, outer and middle ear status and hearing of athletes are screened by trained professionals and students in health care. Follow-up recommendations are provided. The aim is to identify specific otological problems and hearing loss in people with ID, to refer for appropriate medical care, to increase access to hearing health care for this population, and to raise awareness of the hearing concerns of people with special needs, including difficulties in diagnosis and access to treatment options. Results of these screenings from a limited number of athletes have already been published, each demonstrating a high occurrence of sensorineural hearing loss, middle ear problems and earwax impaction (Hey et al., 2014; Hild et al., 2008; Kumar Sinha et al., 2008; Neumann et al., 2006; Pradhan, Stormon & Lallo, 2019; Starska & Lukomski, 2006). The largest study to date in this population is that of Herer (Herer, 2012), studying 9,961 international athletes and confirming these high prevalences of otological and audiological problems. In his study, the lack of demographic data of the included athletes in order to better interprete the results was mentioned. Apart from age and gender, the country of origin seems of importance as health conditions among different countries and regions might be highly variable and the prevalence of hearing loss proved to differ among different regions, being highest in South and East Asia and sub-Saharan Africa (Duthey, 2013).

The purpose of the study is to add to existing knowledge by studying the results of over 100,000 hearing screenings in persons with ID at SO and address the limitations of previous studies by including demographics of the participants. In the discussion, the results will be compared with available data from other groups of persons with ID in order to examine how hearing loss is represented for the whole ID population. The ultimate aim is to increase awareness among caregivers, health care practitioners and policy-makers about the high prevalence of otological and audiological problems in the ID population.

Materials and Methods

Data Collection

The present data were obtained at worldwide national and international sporting events of SO between 2007 and 2017 (included). During these events, the Healthy Athletes health screening program made up of different health disciplines, including otological and audiological screening, was offered to the athletes on a voluntary basis. All participants had an ID and their minimal age was 8 years. No data about ID type or severity were available. Prior to participation, written informed consent was obtained by the athletes and/or their legal guardian. In addition, oral consent was obtained from the athlete at the start of the screening. All data were de-identified and made available by SO International in view of this report. After quality control with incomplete or unclear items considered missing, data were transferred to an SPSS database (SPSS Statistics version 26.0, IBM Corp, Armonk (NY), USA). Regarding the demographics, age was categorized in decades, and countries of origin were classified according to the SO regions (Figure 1). Moreover, the countries were grouped into the four World Bank income groups (low, lower-middle, upper-middle, and high) (World Bank, 2019). Statistical analysis included chi square test with the application of Bonferroni correction where appropriate. In full accordance to the Helsinki Declaration, the Ethics Committee of the author's institution approved the retrospective analysis (EC 115-2017/MF).

Hearing Screening at Special Olympics

The SO Healthy Hearing program uses an international standardized screening protocol developed and well referred to in literature by SO International (Herer & Montgomery, 2006). This protocol was strictly followed by trained professional volunteers (audiologists, speech/language pathologists, medical doctors) supported by trained health care students, and is summarized in Figure 2; the updated manual can be found online (Special Olympics –

Healthy Hearing, 2021). Registration of demographic information (gender, age, nationality) was performed at check-in. Handheld otoscopy was subsequently performed and cerumen removal, if necessary and after approval of the athlete, was performed at many screening events.

Next, hearing screening by distortion product otoacoustic emissions (DPOAE) testing was performed. DPOAE reflect the integrity of the outer hair cells of the inner ear, can be easily and rapidly recorded and are sensitive to hearing loss. Consequently, DPOAE testing is frequently used in screening setting, such as universal newborn hearing screening programs. It was performed in a quiet space, recorded between between 2 and 5 kHz independent of cerumen presence, and resulted in 'pass' or 'no pass' for each ear. A pass was given when a signal-to-noise ratio of 6 dB or more was obtained for at least three out of the four tested frequencies. If a pass on both ears was obtained, the test protocol was terminated.

In case DPOAE testing resulted in no pass in one or both ears, tympanometry (screening for middle ear problems) and pure tone audiometry (PTA, hearing confirmation) were performed. Tympanometry is a short test to evaluate the middle ear function including the tympanic membrane and ossicles by creating air pressure variations. It led to a normal or abnormal result, with normal signifying an admittance between 0.20 and 2.00 mmho, middle ear pressure between +20 and -200 daPa and an ear canal volume between 0.60 and 2.00 ml.

PTA is a subjective test measuring ear-specific hearing thresholds using supra-aural headphones. It was also performed in a quiet room and led to pass or no pass based on screening at 2 and 4 kHz at 25 dB HL, after conditioning with a more intense sound. Only in case of no pass at one or both frequencies, determination of air and bone conduction thresholds at different frequencies was offered to the athlete if available, in order to further elaborate the hearing loss and to discriminate between conductive and sensorineural hearing loss. At check-out, results were discussed with the athlete and guardian and a report with the findings and follow-up suggestions was issued. Referral was provided based on a no pass on PTA screening for one or both frequencies in one or both ears, or based on otoscopic or tympanometric findings (excessive cerumen or pathological findings, abnormal middle ear) in the primary language of the athlete. The type of eventual hearing loss (conductive/mixed or sensorineural) was determined based on otoscopic and tympanometric findings as well as on air and bone conduction thresholds if available.

Results

Demographics

Data of 106,369 screenings conducted between 2007 and 2017 (included) were available. Of these, 37.8% were of female athletes and 62.2% were male. The majority of athletes (70.3%) were aged 8-29y, with 40.8% younger than 20y. Distribution among the different SO regions can be found in Figure 1. According to the World Bank country-specific data, 75.1% of the athletes came from a high-income country, whereas the percentages for low-income, lower-middle-income and upper-middle-income countries were 0.6%, 7.6% and 16.7%, respectively. The distribution of gender and age among the different regions and income groups is provided in Table 1. In all regions, less women than men participated at the hearing screening, with the highest representation of women in North America (40.8%) and Africa (38.1%). Regarding age, the youngest group (<20y) was the largest in all regions, especially in Africa (76.2%). The middle-aged and older adults were best represented in North America (19.2% ≥40y) and Europe/Eurasia (15.9%).

Screening Otoscopy and DPOAE Testing

Even after eventual removal of cerumen, 40.7% had at least one ear canal that was not clear, with 24.6% partially blocked (tympanic membrane only partially visible due to cerumen impaction) and 16.1% compeletely blocked (no tympanic membrane visible due to cerumen impaction). More left ear than right ear cerumen obstruction was observed (21.2% versus 20.8% partial obstruction and 11.9% versus 11.6% complete obstruction for left and right ear respectively, $p<0.001$). In addition to cerumen, foreign bodies such as the tip of an earbud, parts of toys and pencil points were also removed from the ear canal.

Subsequent DPOAE testing yielded a pass for both ears in 47.4% of the participants, so in 52.6% at least one ear failed this screening. Again, more refers were seen in left ears than in right ears (45.5% versus 43.7%, $p<0.001$), which is consistent with the otoscopy as ear canal obstruction will impact DPOAE screening results. Of interest, DPOAE testing resulted in a pass in 54.4% of the athletes with clear ear canals, in 41.0% with partially blocked ear canal and in 31.0% with completely blocked ear canal. Figure 2 depicts the screening protocol together with the main prevalences of the screening results.

Elaboration of Screening Failures

The athletes not having a pass on DPOAE for one or both ears (52.6%) were subsequently tested with tympanometry. A normal tympanogram for both ears was obtained in 64.4% of the referred participants, whereas a no pass was found in 27.3% and 27.7% of the tested right and left ears respectively (right and left combined: 35.6% of the referred participants). After comparison of these results with the otoscopy findings, middle ear problems were suspected based on abnormal tympanometry in 29.5% of the referred participants (21.9% right ears and 22.0% left ears).

In addition to tympanometry, PTA screening was performed in the ears with DPOAE failure to confirm or refute the DPOAE result. In contrast with the DPOAE data, 48.9% showed normal hearing on PTA screening, but hearing loss was confirmed in 51.1% of the referred participants (42.5% right ears and 45.2% left ears, only DPOAE screening failure ears were measured), which is 26.9% of the total group of participants.

Based on a combination of the previous tests and air and bone conduction threshold testing in a selection of athletes, the origin of hearing loss could be estimated with 67.8% of the participants with hearing loss exhibiting permanent sensorineural hearing loss and 32.2% conductive hearing loss or at least a conductive component.

Reasons for Referral

The initial reason for referral was cerumen obstruction of the ear canal. In all participants with partial or complete obstruction in at least one ear (40.7%), follow-up was advised. A second reason was a middle ear problem as shown by an abnormal tympanogram not explained by ear canal findings during otoscopy (29.5% of DPOAE referred athletes). A third referral reason was the confirmation of hearing loss, irrespective of the type (26.9%). As a combination of the above-mentioned findings is frequent, the total referral rate for participants was 57.7%. Referral rates for these reasons were compared with the demographics. Gender differed significantly for cerumen obstruction (39.0% refer in women, 41.8% in men, $p<0.001$) and overall referral rate (56.4% refer in women, 58.4% in men, $p<0.001$), but not for middle ear problems (29.5% refer in women failing DPOAE, 29.5% in men failing DPOAE, $p=0.95$) and hearing loss (26.7% refer in women, 27.0% in men, $p=0.29$). Comparison of the age groups for the different referral reasons is shown in Figure 3, as well as the comparison of SO regions and World Bank income groups (World Bank, 2019) for referral reasons. Significant differences for each of the referral reasons among the different age groups are found, as well as among the SO regions ans World bank income groups, even after Bonferroni correction ($p<0.001$ for group comparisons).

Discussion

Ear and hearing problems are common in the ID population, as shown by the results of this study. Significant cerumen impaction was present in more than 40%, with total obstruction in 16%. Occasionally, in addition to cerumen, foreign bodies needed to be removed out of the external ear canals. Middle ear problems, including chronic otitis media, abnormal middle ear pressure, and tympanic membrane perforation or retraction, were detected in about 30% of those who failed hearing screening by OAEs. Hearing loss, based on screening with OAEs and subsequent PTA screening, proved to be present in 27%. Of course, some of these are thought to be temporary conductive losses based on the cerumen impaction or middle ear problems, but in two out of three participants with confirmed hearing loss, permanent sensorineural hearing loss was detected by pure tone audiometry. The results shown refine the numbers mentioned in previous reports of screening in a limited number of SO athletes. In these studies, cerumen impaction ranged from 33% to 53%, middle ear problems from 6% to 21%, and hearing loss from 2% to 38% (Herer, 2012; Hey et al., 2014; Hild et al., 2008; Kumar Sinha et al., 2008; Neumann et al., 2006; Pradhan, Stormon & Lallo, 2019; Starska & Lukomski, 2006). Sensitivity and specificity of the SO approach has proven highly sensitive and specific in the detection of hearing loss (Hild et al., 2008).

Certain demographics seem to influence the results of otologic and audiologic outcome. Female athletes were less prone to cerumen impaction compared to male athletes, but middle ear problems and hearing loss did not differ among women and men. With advancing age, a stepwise increase in cerumen impaction (from 38% to 47%), middle ear referral (from 13% to 25%) and hearing loss (from 17% to 69%) can be seen. In the oldest age group (≥60 years of age), the referral rate irrespective of the etiology was 84%. Regional differences were also apparent: cerumen impaction was highest in East Asia and North America (both 42.2%), whereas middle ear referrals were higher in Asia Pacific and Europe/Eurasia (both 16%).

Hearing loss was highest in North America (29%) and Asia Pacific (27%), especially in comparison with Latin America (15%). South and East Asia, together with sub-Saharan Africa, are regions known to have a higher prevalence of hearing loss both in children and adults, mainly because of environmental factors such as infections and noise exposure (Duthey, 2013). The high prevalence of hearing loss among athletes in North America can be explained by the higher number of older participants in this region compared with other regions. Taking into account the income groups, cerumen impaction and middle ear referral had the highest occurence in low-income countries (46% and 19% respectively). A possible explanation might be the inclusion of a higher number of SO events in high-income countries, which implies that several athletes might have participated at several events over time and consequently have been treated during a previous screening. It might also be hypothesized that the health care availability contributes to this difference, again pointing towards the importance of screening at such events. In contrast, hearing loss was highest in high-income countries (29%), but again the higher age of the partipants in these countries should be taken into account.

The population studied in this report is rather specific: SO participants are able to participate in sporting events and have usually mild or moderate ID. In order to assess the representativeness of the numbers for the whole ID population, a comparison with other ID groups is necessary. Previously, the SO population was compared with a group of children and adolescents with ID of special needs schools, showing no significant difference in hearing loss prevalence (Hey et al., 2014). The younger age in the school study might contribute to the absence of difference, as the prevalence of hearing loss is influenced by age to a high degree and by ID severity to a lower degree (Evenhuis et al., 2001). In a group of institutionalized children with profound ID, hearing loss was present in 32% (Stein et al., 1987). The prevalence of bilateral hearing loss in an institutionalized ID population with a higher age was 47% (Evenhuis et al., 2001). Finally, a large study in service users with ID, in which measures to approach representativeness for the whole ID population were taken, concluded a weighted population hearing loss prevalence of 30.3% (Meuwese-Jongejeugd et al., 2006). Based on the above, the numbers of hearing loss are in the same order compared with previous studies and might be an underestimation of the actual prevalence given a rather young profile of SO participants with a milder ID grade than the general ID population. Most studies do not report on the type of hearing loss. The observed distribution of conductive versus sensorineural hearing loss (32% versus 68%) is similar to the scarce data in literature (38% versus 62%) (Stein et al., 1987), although the latter being in a pediatric ID population. A limitation of the current study inherent to its setting is the audiometric tests being performed in a separate quiet room with the lowest ambient noise level and thus not in a soundproof booth. This might especially impact threshold determination and thus type and degree of hearing loss.

A specific subgroup of persons with ID that is mentioned in several studies because of its higher hearing loss prevalence, is DS (Evenhuis et al., 2001; Meuwese-Jongejeugd et al., 2006). The elevated occurence of hearing loss can be attributed to both middle ear problems and sensorineural hearing loss. At certain age intervals, otitis media peaks to 60% in children with DS (Maris et al., 2014). Sensorineural hearing loss is low among younsters with DS (De Schrijver et al., 2019) but increases with age to 65% of the adults with DS, especially in the higher frequencies. The high-frequency hearing loss of DS resembles premature aging of the hearing system (Picciotti et al., 2017). Information of the proportion of athletes with DS in the studied population is lacking, but there are no arguments for a participation bias in this respect.

With these results, health issues and more specifically the high prevalence of hearing loss and otologic problems in persons with ID are stressed. Screening is important as self-report is lower and hearing loss might be masked by the appearance of the ID when not actively searched for. Diagnosis is often more difficult to establish in these individuals (Diefendorf et al., 2017), albeit no reason to refrain from proper testing. Patience, adaptation to the situation and professional experience in testing people with special needs can help in obtaining an accurate diagnosis and should be adopted in the health care setting. Especially in persons with severe to profound ID, objective testing might be considered, such as auditory brainstem response audiometry in which brainstem signals are recorded in response to sound without needing the person's active cooperation. Although the burden of hearing loss in persons with ID and the societal effect of treatment is difficult to estimate, appropriate individual treatment should be initiated upon diagnosis. Treatment failure, especially rehabilitation with hearing aids, is possible and service provision to this specific group must be patient-tailored (Meuwese-Jongejeugd, Verschuure & Evenhuis, 2007). More research on the outcome following screening failure and on the assessment of treatment effect with its determinants is required.

In conclusion, health care workers involved in ID should be aware of the high prevalence of hearing loss in this population. Because of low self-report and masking by comorbidities, active screening is mandatory. The health program of SO is able to screen a large number of athletes with ID, confirming the increased need for active otological and audiological follow-up in the far majority of the participants.

References

Crandell CC & Roeser RJ (1993). Incidence of excessive/impacted cerumen in individuals with mental retardation: A longitudinal investigation. Am J Ment Retard, 97(5): 568-574.

De Schrijver L, Topsakal V, Wojciechowski M, et al (2019). Prevalence and etiology of sensorineural hearing loss in children with down syndrome: A cross-sectional study. Int J Pediatr Otorhinolaryngol, 116: 168-172.

Diefendorf AO, Corbin KR, Trepcos-Klinger R, et al (2017). Audiologic considerations for children with complex developmental conditions. In Tharpe AM & Seewald R (Eds.), Comprehensive handbook of pediatric audiology (2nd ed., pp. 609–628). Plural: San Diego (CA, USA).

Duthey B (2013). Background paper 6.21 - Hearing loss. WHO: Geneva (Switzerland).

Evenhuis HM, Theunissen M, Denkers I, et al (2001). Prevalence of visual and hearing impairment in a Dutch institutionalized population with intellectual disability. J Intellect Disabil Res, 45(Pt 5): 457-464.

Herer GR (2012). Intellectual disabilities and hearing loss. Commun Disord Q, 33(4): 252-260.

Herer GR & Montgomery JK (2006). Special Olympics Healthy Hearing Program: Guidelines for standardized screening procedures, 2nd ed. Washington (DC): Special Olympics.

Hey C, Fessler S, Hafner N, et al (2014). High prevalence of hearing loss at the special olympics: is this representative of people with intellectual disability? J Appl Res Intellect Disabil, 27(2): 125-133.

Hild U, Hey C, Baumann U, et al (2008). High prevalence of hearing disorders at the Special Olympics indicate need to screen persons with intellectual disability. J Intellect Disabil Res, 52(Pt 6): 520-528.

Kumar Sinha A, Montgomery JK, Herer GR, et al (2008). Hearing screening outcomes for persons with intellectual disability: a preliminary report of findings from the 2005 Special Olympics World Winter Games. Int J Audiol, 47(7): 399-403.

Maris M, Wojciechowski M, Van de Heyning P, et al (2014). A cross-sectional analysis of otitis media with effusion in children with Down syndrome. Eur J Pediatr, 173(10): 1319-1325.

Marks L, Wong A, Perlman S, et al (2018). Global oral health of athletes with intellectual disabilities. Clinical Oral Investigations, 22: 1681-1688.

Maulik PK, Mascarenhas MN, Mathers CD, et al (2011). Prevalence of intellectual disability: a meta-analysis of population-based studies. Res Dev Disabil, 32(2): 419-436.

Meuwese-Jongejeugd A, Verschuure H & Evenhuis HM (2007). Hearing aids: expectations and satisfaction of people with an intellectual disability, a descriptive pilot study. J Intellect Disabil Res, 51(Pt 11): 913-922.

Meuwese-Jongejeugd A, Vink M, van Zanten B, et al (2006). Prevalence of hearing loss in 1598 adults with an intellectual disability: cross-sectional population based study. Int J Audiol, 45(11): 660-669.

Neumann K, Dettmer G, Euler HA, et al (2006). Auditory status of persons with intellectual disability at the German Special Olympic Games. Int J Audiol, 45(2): 83-90.

Picciotti PM, Carfi A, Anzivino R, et al (2017). Audiologic assessment in adults with Down syndrome. Am J Intellect Dev Disabil, 122(4): 333-341.

Pradhan A, Stormon N & Lalloo R (2019). Oral and aural problems in Australian Special Olympics athletes. Spec Care Dentist, 39(5): 478-484.

Sharby N, Martire K & Iversen MD (2015). Decreasing health disparities for people with disabilities through improved communication strategies and awareness. Int J Environ Res Public Health, 12(3): 2201-3316.

Special Olympics – Healthy Hearing [Internet]. Washington (DC): Special Olympics [cited 2021 April 19]. Retrieved from: resources.specialolympics.org/health/healthy-hearing/

Special Olympics - Inclusive health [Internet]. Washington (DC): Special Olympics [cited 2021 April 19]. Retrieved from: www.specialolympics.org/our-work/inclusive-health/

Starska K & Lukomski M (2006). Realization of International Healthy Hearing Program in Poland--hearing evaluation in participants of Special Olympics. Adv Med Sci, 51: 197-199.

Stein LK, Kraus N, Ozdamar O, et al (1987). Hearing loss in an institutionalized mentally retarded population. Identification by auditory brainstem response. Arch Otolaryngol Head Neck Surg, 113(1): 32-35.

Szymanski CA, Brice PJ, Lam KH, et al (2012). Deaf children with autism spectrum disorders. J Autism Dev Disord, 42(10): 2027-2037.

Tracy J & McDonald R (2015). Health and disability: partnerships in health care. J Appl Res Intellect Disabil, 28(1): 22-32.

WHO Europe - Definition: intellectual disability [Internet]. Geneva (Switzerland): World Health Organization [cited 2020 October 8]. Retrieved from: https://www.euro.who.int/en/health-topics/noncommunicable-diseases/mental-health/news/news/2010/15/childrens-right-to-family-life/definition-intellectual-disability/

World Bank [Internet]. Washington (DC): World Bank Data Team; [updated 2019 July 1; cited 2020 October 8]. Retrieved from: https://blogs.worldbank.org/opendata/new-country-classifications-income-level-2019-2020/

Tables

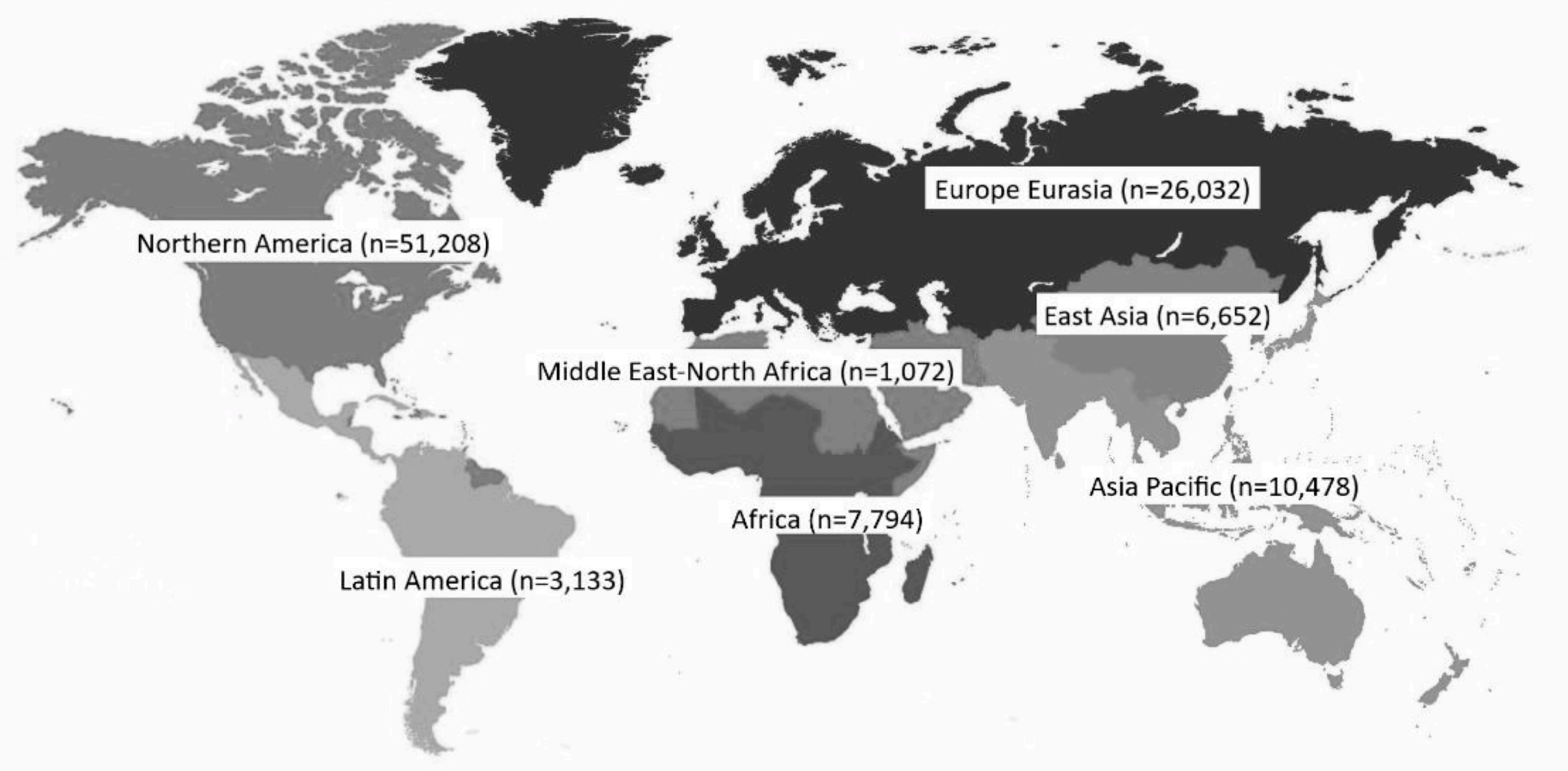
Table 1: Distribution of gender and age among the different Special Olympics regions and World Bank country income groups. The absolute numbers are shown, followed by the percentage of gender or age group for the specific region/income group.

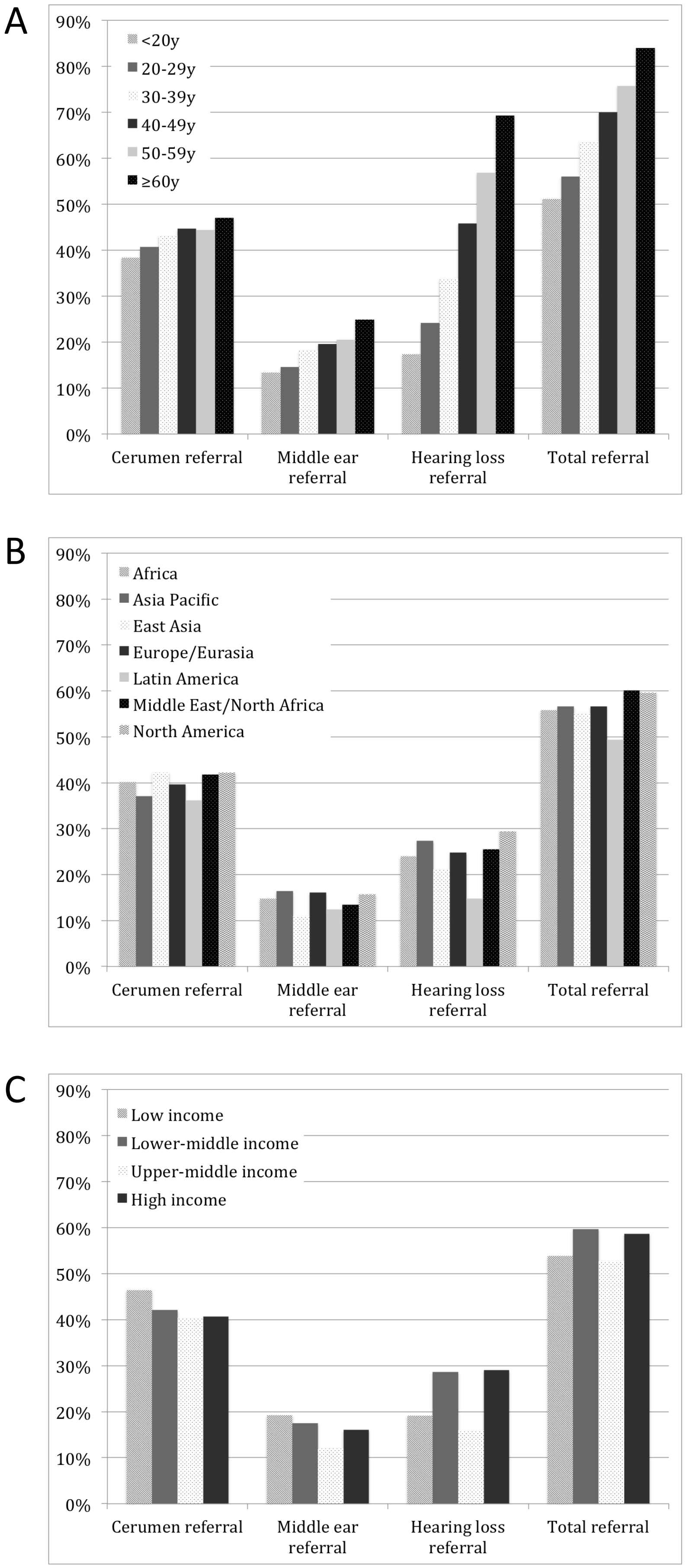
Figures

Figure 1: Distribution of athletes over Special Olympics regions.

Figure 2: Summary of screening protocol in black, and prevalences for the different findings in grey.

Figure 3: Prevalences of the different reasons for referral compared with age groups (panel A), Special Olympics regions (panel B) and World Bank income groups (panel C).


Europe Eurasia (n=26,032)
Northern America (n=51,208)
East Asia (n=6,652)
Middle East-North Africa (n=1,072)
Asia Pacific (n=10,478)
Africa (n=7,794)
Latin America (n=3,133)


A
90%
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B
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Europe/Eurasia
Latin America
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North America
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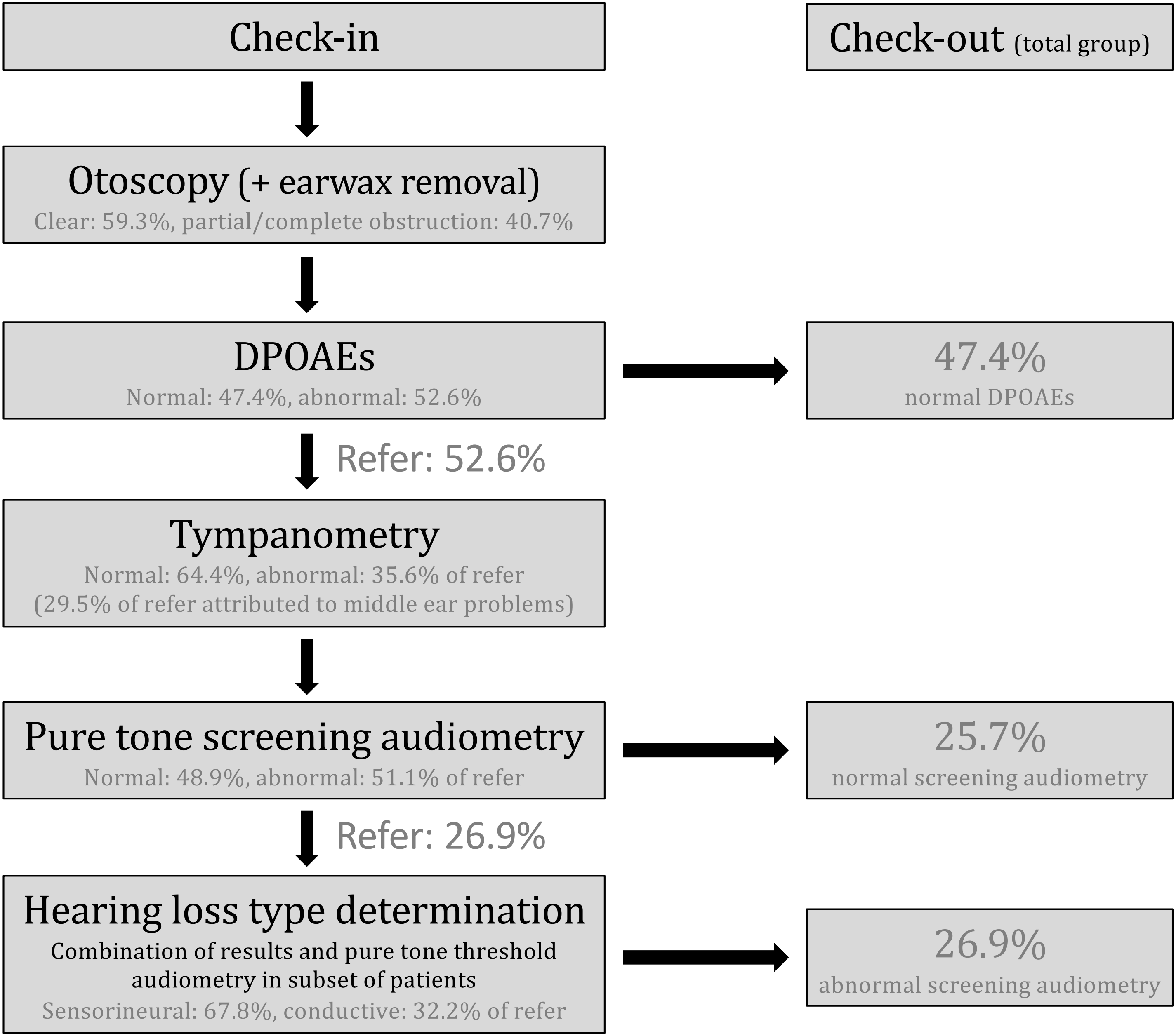
Check-in
Otoscopy (+ earwax removal)
Clear: 59.3%, partial/complete obstruction: 40.7%
DPOAEs
Normal: 47.4%, abnormal: 52.6%
Refer: 52.6%
Tympanometry
Normal: 64.4%, abnormal: 35.6% of refer
(29.5% of refer attributed to middle ear problems)
Pure tone screening audiometry
Normal: 48.9%, abnormal: 51.1% of refer
Refer: 26.9%
Hearing loss type determination
Combination of results and pure tone threshold audiometry in subset of patients
Sensorineural: 67.8%, conductive: 32.2% of refer
Check-out (total group)
47.4%
normal DPOAEs
25.7%
normal screening audiometry
26.9%
abnormal screening audiometry

Table 1: Distribution of gender and age among the different Special Olympics regions and World Bank country income groups. The absolute numbers are shown, followed by the percentage of gender or age group for the specific region/income group.

	Gender n=105,609 (760 missing)		**Age** n=104,342 (2,027 missing)					
	Women (37.8%)	**Men (62.2%)**	**<20y (40.8%)**	**20-29y (29.5%)**	**30-39y (15.1%)**	**40-49y (8.6%)**	**50-59y (4.5%)**	**≥60y (1.6%)**
Africa (7.3%)	2,917 (38.1%)	4,733 (61.9%)	5,430 (76.2%)	1,204 (16.9%)	272 (3.8%)	129 (1.8%)	68 (1.0%)	26 (0.4%)
Asia Pacific (9.9%)	3,588 (34.3%)	6,874 (65.7%)	5,121 (50.2%)	3,057 (29.9%)	1,085 (10.6%)	603 (5.9%)	251 (2.5%)	93 (0.9%)
East Asia (6.3%)	2,110 (31.8%)	4,516 (68.2%)	4,479(67.6%)	1,681 (25.4%)	320 (4.8%)	105 (1.6%)	35 (0.5%)	8 (0.1%)
Europe/Eurasia (24.5%)	9,219 (35.6%)	16,682 (64.4%)	9,281 (36.0%)	8,069 (31.3%)	4,338 (16.8%)	2,463 (9.5%)	1,227 (4.8%)	420 (1.6%)
Latin America (2.9%)	1,092 (34.9%)	2,039 (65.1%)	1,357 (43.9%)	1,106 (35.8%)	465 (15.0%)	110 (3.6%)	34 (1.1%)	18 (0.6%)
Middle East/North Africa (1.0%)	334 (31.3%)	733 (68.7%)	504 (47.4%)	467 (43.9%)	77 (7.2%)	12 (1.1%)	2 (0.2%)	1 (0.1%)
North America (48.1%)	20,710 (40.8%)	30,062 (59.2%)	16,363 (32.5%)	15,178 (30.1%)	9,192 (18.2%)	5,543 (11.0%)	3,045 (6.0%)	1,103 (2.2%)
Low income (0.6%)	181 (29.1%)	440 (70.9%)	430 (73.1%)	135 (23.0%)	20 (3.4%)	2 (0.3%)	0 (0.0%)	1 (0.2%)
Lower-middle income (7.6%)	2,978 (36.9%)	5,097 (63.1%)	5,411 (71.3%)	1,826 (24.0%)	287 (3.8%)	55 (0.7%)	10 (0.1%)	4 (0.1%)
Upper-middle income (16.7%)	6,186 (35.1%)	11,454 (64.9%)	10,736 (62.0%)	4,730 (27.3%)	1,299 (7.5%)	339 (2.0%)	137 (0.8%)	63 (0.4%)
High income (75.1%)	30,625 (38.6%)	48,648 (61.4%)	25,958 (32.9%)	24,071 (30.5%)	14,143 (17.9%)	8,569 (10.9%)	4,515 (5.7%)	1,601 (2.0%)