Original Article

Audiologic Screening of People with Down Syndrome in Special Schools in Surulere Lagos, Nigeria

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Abstract

Background: Down syndrome (DS) is associated with a high incidence of ear pathologies and hearing loss. There is a paucity of information on the audiology of people with DS particularly those in special schools in Nigeria. This study aimed to compare pneumatic otoscopy with tympanometry for middle ear screening and pure tone audiometry (PTA) with distortion product otoacoustic emission (DPOAE) for hearing screening in the different age groups of people with people DS.
Methodology: People with DS in two special schools for people with intellectual disability in Surulere, Lagos state were selected based on the schools' record and their phenotype. Otoscopy, pneumatic otoscopy, tympanometry, screening PTA and screening DPOAE were performed on the subjects.
Results: 52 subjects aged 6 to 36years and an M: F ratio of 1.5:1 were studied. 82.7% of subjects had wax impaction. The prevalence of otitis media with effusion (OME) by pneumatic otoscopy was 57.8% and 40.2% by tympanometry and the same number of subjects performed both tests successfully. Screening PTA and DPOAE were successfully performed on 32.7% and 94.2% of the subjects respectively. Fail rate of 29.4% by PTA and 65.3% by DPOAE were recorded.
Conclusion: Regular ear check-up in people with DS is essential for the detection and treatment of ear wax, OME and hearing impairment which is highly prevalent in them. Pneumatic otoscopy is recommended for the detection of OME in all the age groups. Screening PTA and OAE are recommended for adults and children and none co-operative adults hearing screening respectively.
Keywords: Down Syndrome; Nigeria; Screening Audiology; Special Schools.

Introduction

Down Syndrome (DS) is a common chromosomal abnormality in life born children.[1] DS is typically associated with a wide array of dysmorphic features and congenital malformations many of which are in the head and neck region.[2] DS is the most common genetic cause of intellectual disability.[3] High incidence of middle ear infections and hearing loss has been reported in people with DS in literature.[4] This has been attributed to the varied congenital malformations of their ear among other factors. These ear malformations include; small pinna in children resulting in poor sound localization and concentration, stenotic external auditory canal in infants and toddlers making ear examination difficult and predisposing them to cerumen impaction, small eustachian tube causing poor aeration of the middle ear cavity with resultant

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Access this article online
Quick Response Code:
Website: www.nigerianmedjournal.org

How to cite this article: Asoegwu CN, Abdulazeez AA, Nwawolo CC. Audiologic Screening of people with Down syndrome in special schools in Surulere Lagos, Nigeria. Niger Med J 2021;62;(6):312-317

chronic middle ear disease with all culminating in conductive hearing loss. Inner ear dysplasia causing sensorineural hearing loss is also reported in some.[5,6] Hearing loss has been reported in 50% to 90% of people with DS.[7-9] The predominant type of hearing loss in people with DS is conductive although they are more likely to have sensorineural hearing loss than their peers.[10,11]

Pneumatic otoscopy and tympanometry are some of the recommended diagnostic tools for otitis media with effusion. Pneumatic otoscopy is affordable, easily available and accurate in experienced hands but requires patient co-operation. Tympanometry is expensive and not easily available particularly in resource-challenged countries but preferred in children 2-24months and none co-operative patients. Assessment of hearing acuity in people with DS can be problematic and there is no general agreement as to the preferred test method. Pure tone audiometry (PTA) is the ideal test for detecting hearing impairment but the subject's ability to understand instructions and to respond to the heard sound by carrying out certain actions are required. Oto-Acoustic Emission test (OAE) assesses the integrity of the hearing pathway. It does not require active participation from the subject and is, therefore, suitable for very young children and subjects with intellectual disability.

There is a paucity of published local data on the hearing profile of people with DS generally and specifically for those in special schools in Nigeria. The objective of this prospective study was to compare pneumatic otoscopy with tympanometry for middle ear screening and pure tone audiometry with DPOAE for hearing screening in different age groups of people with DS in special schools. The result will guide physicians in the choice of the type of test method to be used in the follow-up hearing screening of people with DS in special schools in Nigeria.

Methods

This is a study on people with DS living in two institutional settings in Surulere, Lagos State, Nigeria. Ethical clearance approval number 669 was obtained from the Lagos University Teaching Hospital Health Research Ethics Committee and permission from the management of the selected institutions. Using national guidelines for nontherapeutic research involving minors and decisional impaired individuals, informed consent was obtained from the adults where possible, their legally authorized representative which included their parents/guardians/class teachers and minor assent for those ≤ 12 years where applicable.

All subjects with Down syndrome in the selected schools who consented to be part of the study were recruited. Information on their bio-data was obtained from their class register. All screening assessments were carried out in their schools.

Assessment protocol consisted of otoscopic examination followed by ear syringing for the removal of earwax and debris when present, pneumatic otoscopy, tympanometry, screening pure tone audiometry (PTA) and screening Distortion Product Oto-Acoustic Emission (DPOAE).

Pneumatic otoscopy for assessing the mobility and position of the tympanic membrane was carried out with Reister 2050 Otoscope. Tympanometry was performed using Amplivox otowave tympanometer with a probe tone frequency of 226Hz, the intensity of 85dB and pressure range of +200 to -300. Tympanogram tracing was classified based on Jerger's classification where type A- normal middle ear condition, type B- fluid in the middle ear and type C- negative middle ear pressure.[12] The presence of OME in this study was confirmed by Type B and C tympanogram.

Screening manual PTA was performed using Kamplex AS7 screening audiometer with circum-aural air conduction earphone. PTA test was at the frequencies 0.5, 1, 2 and 4 kHz with the screening level of 30 dB HL. Response at all frequencies in both ears was pass while non-response in one or more frequencies in either or both ears was fail. Screening DPOAE was carried out with MAICO ERO·SCAN-OAE V100.01. REFER in any ear is regarded as failed screening.

Data was collated and descriptive statistical analysis was carried out using Statistical Product and Service Solution (IBM SPSS-21) for windows. The result was presented in text, tables and figures.

Results

A total of 74 Down Syndrome subjects were recruited, 52(70.3%) completed the study while 22(29.7%) did not complete the study. There were 31(59.6%) males and 21(40.4%) females with a male: female ratio of 1.5:1. Age range was 6 years to 36 years with a mean age of 19.13±8.74. The study population was divided into 3 age groups; namely: children (age 6 – 11years), adolescents (age 12 – 19 years) and adults (20 years and above). 11(21.2%) were children, 20(38.5%) were adolescents while 21(40.4%) were adults.

Wax impaction was observed in 43(82.7%) subjects, bilateral in 35(81.4%) and unilateral in 8(18.6%) of the subjects. Other otoscopic findings are shown in table 1.

Pneumatic otoscopy and tympanometry were performed on all the subjects except one child who had bilateral TM perforation. Immobile TM was noted in 30(58.8%) subjects while 21(41.2%) subjects had mobile TM bilaterally. Type A tympanogram was recorded in 28(54.9%) subjects bilaterally. Types B and C tympanogram were recorded in 23 (45.1%) subjects. The details of the tympanometry and pneumatic otoscopy results among the study subjects in the three age groups are presented in table 2.

Pure tone screening audiometry was successfully carried out on 17/52 (32.7%) of the study subjects made up of 7/20 (35%) adolescents and 10/21 (47.6%) adults. 5 out of 7 (71.4%) adolescents and 7 out of the 10 (70%) adults that were screened passed giving a pass rate of 70.6%. This audiometric screening could not be carried on subjects within the children age group due to non- cooperation.

Screening DPOAE was however successfully carried out on 49 (94.2%) of the subjects. 3 (5.8%) subjects (1 in each age group) were uncooperative. Overall, 65.3% of the study subjects failed the screening. This consists of 100% of the children, 68.4% of the adolescents and 45% of the adults as illustrated in figure 1.

Table 1: Otoscopic Findings among participants in Various Age Groups

Findings	Children(11) n (%)	Adolescents(20) n (%)	Adults (21) n (%)	Total (52) n (%)
Wax Impaction	10 (90.9)	19 (95.0)	14 (66.7)	43 (82.7)
Otorrhea	1 (9.1)	1 (5.0)	1 (4.8)	3 (5.8)
TM Perforation	1 (9.1)	- -	- -	1 (1.9)
TM Retraction	7 (63.6)	12 (60.0)	11 (52.4)	30 (57.7)

Table 2: Pneumatic otoscopy and Tympanometry results among study subjects in various Age groups

Age Group	Pneumatic Otoscopy		Tympanogram		
	Mobile TM n (%)	Immobile TM n (%)	Type A n (%)	Type B n (%)	Type C n (%)
Children	3 (15.0)	17 (85.0)	4 (20.0)	12 (60.0)	4 (20.0)
Adolescents	20 (50.0)	20 (50.0)	24 (60.0)	14 (35.0)	2 (5.0)
Adults	20 (47.6)	22 (52.4)	33 (78.6)	5 (11.9)	4 (9.5)
Total	43 (42.2)	59 (57.8)	61 (59.8)	31 (30.4)	10 (9.8)

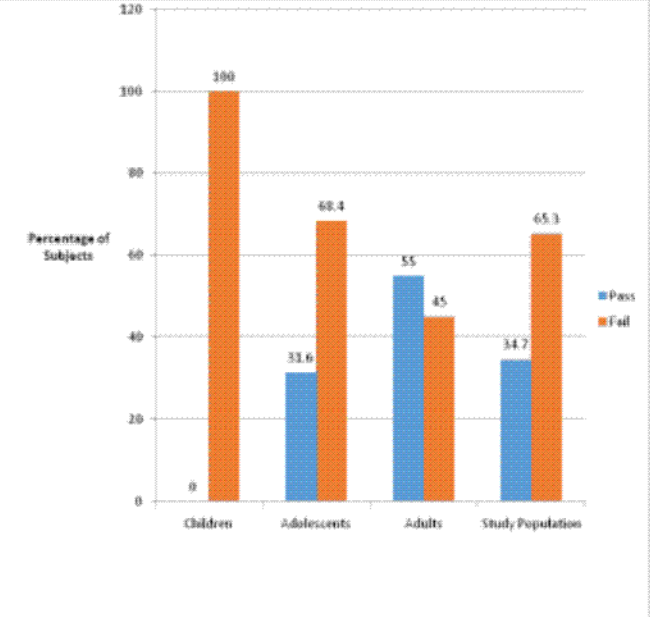

Figure 1: Screening DPOAE Results among study subjects in various age groups

Discussion

Wax impaction was highly prevalent in the general study subjects. The prevalence of wax impaction in the children group in this study of 90.0% is much higher than 46.7% reported among normal school children of similar age in Nigeria.[13] It is also higher than the 14.4% - 40.0% prevalence reported in studies among children with DS in Malaysia and Greece.[14,15] The prevalence of wax impaction in the adult group was 59.5%. There is a paucity of data in the literature to compare this with. It was noted that

the first-ever ear examination by a doctor for the majority of the study subjects was during the research. Thus, the absence of hospital visit for a routine checkup and ear cleaning in the study subjects could be responsible for the recorded high prevalence. A major sequelae of impacted earwax is conductive hearing loss, which can be deleterious to linguistic, social, and intellectual development, particularly for individuals with mental retardation.[16]

An equal number of the study population underwent both pneumatic otoscopy and tympanometry tests successfully. The overall prevalence of OME using immobility of the tympanic membrane on pneumatic otoscopy was 57.8% and using types B and C on tympanometry it was 40.2%. Though the rates appear close, there was no statistical analysis of this difference to determine significance. However, a previous study reported a similar predictive value from both pneumatic otoscopy and tympanometry.[17] This study was not able to confirm this statistically. Thou both tests are suitable for screening for OME, there is disparity between the cost and availability of both test methods with pneumatic otoscopy being cheaper and more readily available. Pneumatic otoscopy can be done by any physician but with minimal training and clear diagnostic guidelines. Tympanometry, on the other hand, requires an audiologist or audiometrician who are not readily available in developing countries. The prevalence of OME in the children age group in this study is 70% - 80.0% and is similar to the prevalence reported in children with DS in literature.[18-20] It is, however, higher than 15.9% - 25.2% reported prevalence in their normal peers and 43.7% prevalence in children that attend daycare in Nigeria.[21-23] The prevalence of OME among adults by tympanometry in this study was 28.2% and is consistent with 18.7% prevalence from a study on adults with DS.[24] Untreated OME in children with DS has been known to persist into early adulthood as in this study population.[19]

The challenges of performing PTA on people with DS which has been well documented was also noted in this study with a successful test rate of only 32.7%.[7,17] Age was a determining factor in the ability to perform PTA test in this study as all those successfully tested were either adolescents or adults (≥ 12 years). Reports from previous studies vary on the appropriate chronological age for people with DS to successfully undergo behavioral auditory assessment like pure tone audiometry. Roizen et al. and Maurizi et al reported three and a half years and eight years respectively.[18,25] A combination of the subject's mental age and developmental level have been advocated as a good determinant of the audiological procedures to be employed.[15,26]

The low prevalence rate of hearing loss of 29.4% detected by PTA test in this study can be attributed to the limited number of subjects that were screened particularly that the children could not be screened. Screening with DPOAE was successful in 94.2% of the study subjects, making it a good hearing screening tool for people with DS. Objective screening methods are preferred for people with intellectual disability. The prevalence of hearing loss by DPAOE of 65.3% in this study is consistent with 50% - 90% prevalence of hearing loss that has been previously recorded in DS people in the literature.[7-9,27,28] The reason for a 100% prevalence of hearing loss in the children age group could be partly due to the high prevalence of OME in this group. OME has been noted to be responsible for conductive hearing loss in 80% of DS patients in a study.[29] The prevalence of hearing loss among the adults agrees with 51% to 74% recorded in the studies of hearing loss in adults with DS.[24,30] High-frequency hearing thresholds loss that begins in the second decade of life in people with DS could be responsible for the noted prevalence.[31] A hearing loss prevalence pattern that reduces with increasing age was recorded in this study.

Conclusion

The prevalence of wax impaction, OME and hearing loss in the people with DS in the present study was high. This may impact negatively on the learning, communication and rehabilitation of people with DS. There is, therefore, a need for regular ear check-up, hearing screening for early detection and treatment of the ear pathologies. Pneumatic otoscopy which is readily available and affordable is recommended to primary caregivers for the detection of wax impaction and OME in all age groups. PTA is recommended for hearing screening in adults while screening OAE is recommended for children and none co-operative adults.

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