

## Tympanometric and TOAEs Results of Kuwaiti School Children with Down Syndrome

Aseel Al-Meqbel\*

Department of Speech and Hearing Therapy, Kuwait University, Kuwait

\*Corresponding author: Aseel Al-Meqbel, Department of Speech and Hearing Therapy, Kuwait University, Kuwait, Tel: 24983412; E-mail: [aseel.m@HSC.EDU.KW](mailto:aseel.m@HSC.EDU.KW)

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### Abstract

**Objective:** Conductive hearing loss is one of the most common disabilities in children with Down Syndrome (DS). The primary aim of this study is to evaluate the results from tympanometry and transient otoacoustic emissions (TOAEs) in children with DS in special schools.

**Methods:** We investigated the results of tympanometry and TOAEs testing for a group of 57 children with DS aged 7.1-16.2 years.

**Results:** Type A tympanograms were found in 4 participants, type C1 in 2 participants, type C2 in 15 participants, and type B in 33 participants. TOAE was present in six and absent in 48 participants.

**Conclusion:** A combination of tympanometry and TOAE is a fast, efficient and objective evaluation of middle ear and outer hair cell functions in children with DS. This test battery successfully examined over 80% of the children with mild to moderate intellectual impairment, who would be difficult to test using behavioral methods.

**Keywords:** Tympanometry; Transient otoacoustic emissions Down's syndrome; Conductive hearing loss and Eustachian tube dysfunction

### Introduction

Down Syndrome (DS) is a genetic disorder resulting from trisomy of chromosomal abnormality 21 affecting 1 in every 1000 live births [1,2]. DS is one of the most common forms of mental disability [2]. DS commonly presents with a group of medical issues including a high incidence of otitis media (OM). Muscular hypotonia and associated eustachian tube dysfunction may be a more important cause of middle-ear disease than impaired immune function.

Conductive involvement frequently arises in children with DS, with recurrent episodes of otitis media or middle ear dysfunction not uncommon [3]. Previous reports have documented that DS usually associated with a high incidence of otitis media and eustachian tube dysfunction that may be an underlying cause of middle ear disorders [4]. External pinna defects are also regularly found in this population [5]. These otolaryngologic anomalies complicate the diagnosis and classification of hearing deficit. Evaluating the tympanic membrane in young children with DS is made more difficult by stenosis of the external auditory canal and increased rates of cerumen impaction. Middle ear abnormalities, including eustachian tube dysfunction, remnant tissue in the middle ear cavity, and ossicular malformations, are frequently seen and contribute further to the higher rate of conductive hearing loss reported in this population [6].

Hearing screening for DS population using behavioural test batteries is difficult and unreliable responses could be obtained due to their intellectual impairment. Therefore, it is highly recommended hearing screening should be done using objective methods such as tympanometry or/and TOAE.

Children with DS characteristically have ears similar to those of neonates, with a small ear canal. The similar ear morphologies, and the established sensitivity of 1000 Hz tympanometry in neonates, suggest that the 1000 Hz probe tone may be a more accurate measure of middle ear function in the DS population. Clinical experience has also shown many false positives using the 226 Hz probe tone with DS participants, indicating limited static admittance when middle ear effusion actually does not exist [7]. We therefore used the 1000 Hz probe tone in the current study.

Despite widespread awareness that children with DS are particularly susceptible to hearing disorders, no previous study has investigated the audiological status of Kuwaiti children with DS, and the audiological status of such children in special schools in Kuwait is unknown. Consequently, these children with DS may be disadvantaged by delayed identification of their hearing disorder.

### Method

This cross-sectional study investigated children with DS at two special schools in Kuwait, employing a screening assessment to examine the children's hearing status. Approval from the school's ethics committee was obtained before any data collection commenced.

### Participants

Fifty-seven students with DS (25 males and 32 females), aged from 7.1 years to 16.2 years (mean age 12.9 years; standard deviation 2.7 years) were recruited from two schools. All had mild to moderate intellectual impairment, no student was wearing a hearing aid or cochlear implant, and there were no previous records of their hearing status. Ambient noise levels within the rooms were in the range of 34 to 36 dB A (mean=36.3 dB A).

## Procedure

We first conducted otoscopic investigation on each participant to detect occluding cerumen and investigate the status of outer ear canal and tympanic membrane. This preliminary procedure ensured that tympanometry and TOAEs results were not negatively influenced by obstruction, wax occlusion, or collapse of the external auditory canal.

We then performed tympanometry using a MADSEN OTOflex 1000 tympanometer, Otometrics to evaluate the middle ear status. The instrument was calibrated to ANSI S3.39-1987 standards [8] and Jerger's [9] system was used to classify the tympanometric results (Table 1).

| Tympanogram | Middle Ear Compliance (ml) | Middle Ear Pressure (daPa) |
|-------------|----------------------------|----------------------------|
| A           | 0.3 to 1.5                 | +50 to -100                |
| As          | <0.3                       | +50 to -100                |
| Ad          | >1.5                       | +50 to -100                |
| B           | -                          | -                          |
| C1          | 0.3 to 1.5                 | -101 to -200               |
| C2          | 0.3 to 1.5                 | -200                       |

Note: Based on data from Jerger [9].

**Table 1:** Tympanogram types.

Type B or C2 tympanograms were regarded as failures, while type A or C1 tympanograms were considered passes [10].

Transient otoacoustic emission (TOAE) screener test was then performed using OToRead system Interacoustics Denmark to measure the TOAE responses. The instrument was calibrated to the manufacturer's recommended standards.

Ears with type B tympanograms associated with TOAE failure were suspected to have middle ear effusion [11]. All children were referred to the otology and audiology center for diagnostic assessment if they did not pass either tympanometry or TOAE in either ear.

## Results

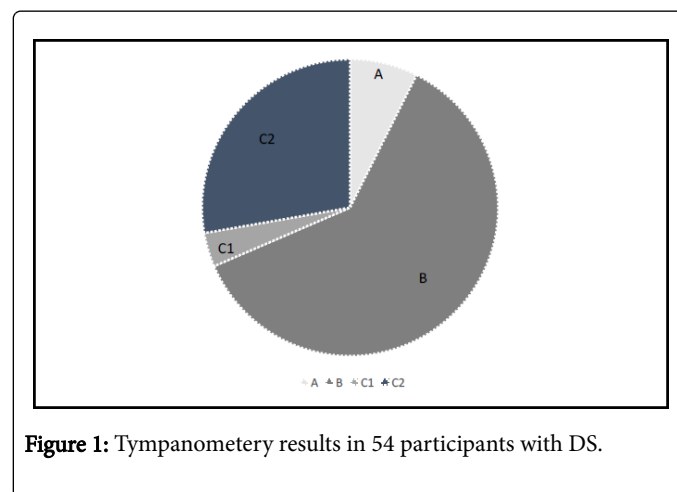
Following otoscopic examination, three participants were referred for medical consultation and wax removal, and were excluded from the study (Table 2).

| Tympanogram | TOAE |      |
|-------------|------|------|
| Fail        | Fail | Pass |
| B           | 33   | -    |
| C2          | 15   | -    |
| Pass        | -    | -    |
| C1          | -    | 2    |
| A           | -    | 4    |
| Total       | 48   | 6    |

**Table 2:** Tympanometric and TOAEs results in 54 participants.

Six participants (10.5%) were found to have normal findings for combined tympanometry and TOAEs battery. The remaining 48 participants (84.2%) had abnormal findings. However, no DS students were identified by the caregivers or school staff as having a diagnosed hearing impairment. That means those students have undiagnosed hearing problems at the time of testing.

Tympanometry and TOAE could be performed in 54 participants bilaterally. For tympanometric results Type A was found in four participants (7.4%), Type C1 was found in two participants (3.7%), type C2 in 15 participants (27.7%), and type B in thirty-three participants (61.1%). High failure rate was noted from the TOAEs results obtained from our DS participants. TOAEs were present in six participants and absent in 48 (Figure 1).



**Figure 1:** Tympanometry results in 54 participants with DS.

## Discussion

Hearing evaluation on children with developmental delay such as DS using behavioral tests is always accompanied by difficulties in obtaining reliable and valid responses. Combined objective testing using tympanometry and TEOAE is a feasible protocol for screening school children with DS. The results of this study showed abnormalities of the external, middle and inner ear in children with DS, which contribute to the hearing loss in this population. Over 80% of the hearing loss is conductive as judged by the type B tympanograms. In children much of this is due to otitis media with effusion, and is therefore amenable to medical and surgical intervention [12]. However, 4-20% of hearing loss in this population is due to sensorineural hearing loss [13]. Until relatively recently this type of hearing loss has been treated only by amplification and non-oral communication techniques.

Analysis of the results from our study showed that a high percentage (84.2%) of children with DS have conductive hearing loss. One could therefore speculate that the high incidence of middle ear problems in children with DS may be due to the deformity of the middle ear and the anatomical malformation in their eustachian tube muscular activity. This could also account for the high occurrence of conductive hearing loss. Remarkably, our results show that the vast majority of participants failed the combined tympanometry and TOAEs battery. This finding shed light for immediate hearing screening programs in special schools, especially for those children with DS. Our findings concur with previous studies that have shown that 60-80% of children with DS have middle ear problems resulting in hearing disorders. Most

such hearing deficit appears to be conductive [5,14]. Early detection and management of hearing deficits is, therefore, essential for the child with DS because the vast majority of children acquire speech and language primarily by hearing.

Reliable and valid responses are generally difficult to obtain when conducting behavioral tests and diagnostic hearing assessment on children with developmental delay and DS. Combining objective testing using tympanometry and TOAE is a feasible protocol for screening school children with DS.

## Conclusion

A combination of tympanometry and TOAE is a fast, efficient and objective evaluation of middle ear and outer hair cell functions in children with DS. This test battery successfully examined over 80% of the children with mild to moderate intellectual impairment, who would be difficult to test using behavioral methods. The combination of failed tympanograms and absence of TOAEs indicated a high prevalence of conductive hearing loss in children with DS, possibly due to middle ear pathologies. Therefore, tympanometry and TOAE technology could be employed as tools for screening children with DS. Further diagnostic study is needed once middle ear pathology is resolved.

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