A pilot study of the relationship between Down's syndrome and hearing loss

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ABSTRACT

Objective: Hearing loss is one of the most common disabilities in children with Down's syndrome. The objective of this study is to investigate the incidence and types of hearing loss in Down's syndrome patients.

Methods: Twenty-six subjects with Down's syndrome aged between 2 and 17 year old were evaluated during the year 1998, referred randomly by the Pediatric Department and the Down's Syndrome Centre. Screening audiological procedures were used to evaluate these patients, which include behavioral audiometry, play and pure tone audiometry, tympanometry and ABR, which is the measurement of the activity of the auditory pathway structures from the distal auditory nerve to the midbrain using clicks or tonepips.

Results: The study showed that 35% were found to have normal hearing compared to 4% found to have sensorineural hearing loss, while the majority of patients were found to have a conductive hearing loss (50%).

Conclusion: Hearing evaluation of children with Down's syndrome revealed a high prevalence of conductive hearing loss. This study proposed that this might be attributed to eustachian tube dysfunction and middle ear ciliary malfunction. Further diagnostic studies need to be carried out.

Keywords: Down's syndrome, conductive hearing loss, eustachian tube dysfunction, ciliary malfunction.

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Down's Syndrome (DS) is a genetic disorder that results from a chromosomal abnormality due to trisomy for all or a large part of chromosome 21 and is one of the most common forms of mental disability.[1] Down's syndrome usually presents with a constellation of medical problems including a high incidence of otitis media. Muscular hypotonia and associated eustachian tube dysfunction may be a more important cause of middle-ear disease than impaired immune function.[2]

Hearing loss is one of the most common disabilities in children with Down's syndrome[3] but little agreement exists regarding the incidence or nature of the hearing problem. No doubt, much of the confusion in the literature regarding the incidence or type of hearing loss in Down's syndrome is due to the difficulty in evaluating this patient population and to the lack of early screening programs. The present pilot study was designed to investigate the type of hearing loss in Down's syndrome patients with the hope of designing more comprehensive studies in the future.

Methods. *Subjects.* Twenty six subjects with Down's syndrome aged between 2 and 17 years were evaluated during the year 1998. These patients were referred to audiometry from the pediatric clinics and the Down's syndrome Center.

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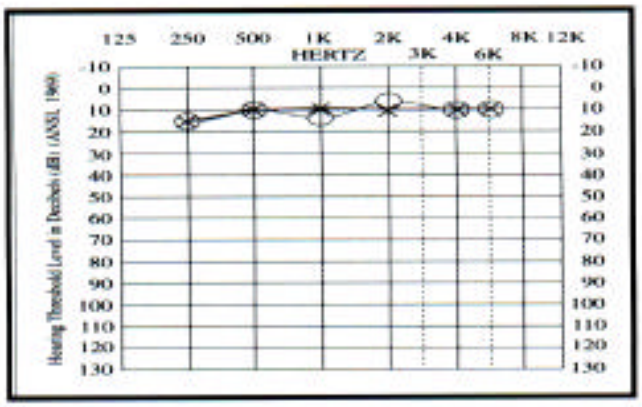

Figure 1 - Normal hearing.

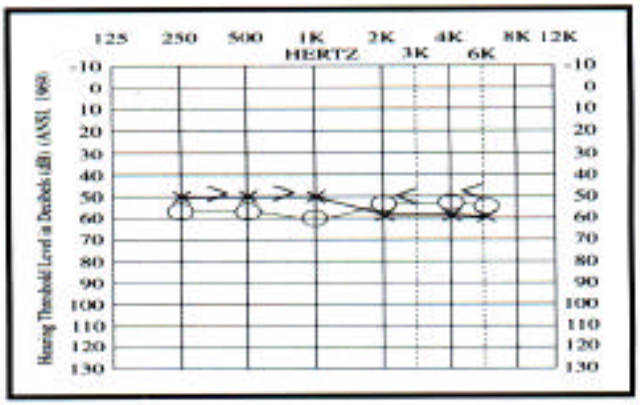

Figure 2 - Moderate sensorineural hearing loss.

Screening procedure. Initial otoscopy was carried out to rule out any accumulated cerumen or discharge before starting the audiological evaluation. Middle ear pressure was checked using tympanometry to evaluate the status of the middle ear. Middle ear pressure was accepted to be normal if it was found to be within 0 to 170 dapa. Audiometry measurements "pure tone and play" consisting of air conduction in the frequency range 250Hz - 8KHz and tone conduction between 250Hz - 4KHz were used. An average of air conduction less than 20 dbHL was considered normal. Visual reinforcement audiometry with 20db across the frequency range 250Hz - 6KHz was implemented as a criteria for normality. However, auditory brainstem response (ABR) was in conjunction with the above measurements in 5 cases to confirm the findings.

Results. Audiometric evaluation was carried out on 26 subjects with DS referred from the Down's Syndrome Center and from consultants in the Department of Pediatrics. The results showed that 18 patients (35%) were found to have normal hearing loss (Figure 1), 34 patients (65%) had hearing loss. Of these, 4% with moderately severe sensorineural hearing loss (Figure 2), 50% with mild to moderate conductive loss (Figure 3) and 11.5% with moderate to severe mixed loss (Figure 4).

Discussion. Severe hearing loss and external pinna malformations are both common in DS children.[4] Numerous reports from the literature reveal that 60-80% of DS children have middle ear involvement resulting in hearing deficit. Evidence indicates that most hearing loss in DS is conductive in nature.[4,5]

Analysis of the results showed that a high percentage of DS children have conductive hearing loss, therefore, one may speculate that the malformation of the middle ear and the eustachian tube muscular system may be playing a role in causing such high incidence of hearing loss. It was proposed that a combination of increased eustachian tube muscular activity and passive nasal over pressure were responsible for the eustachian tube opening, consequently, ventilation of the middle ear.[6]

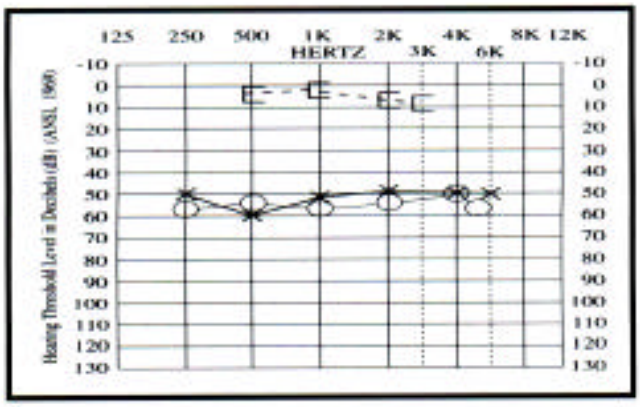

Figure 3 - Moderate conductive hearing loss.

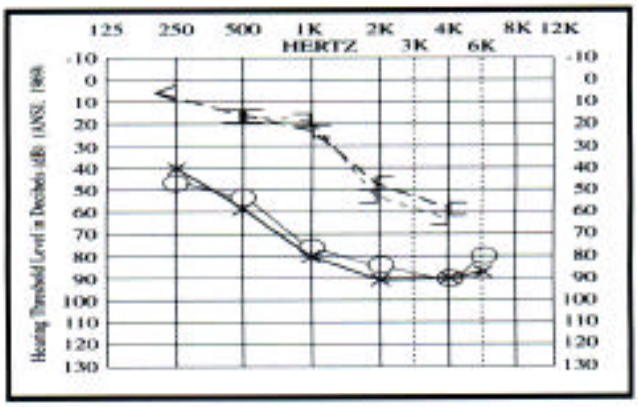

Figure 4 - Mixed hearing loss.

Since DS children have anatomical malformation in their eustachian tube muscular activity, it was proposed that this in part may account for the observed high incidence of conductive loss.

Recent reports have also indicated that hearing loss may contribute to decreased intelligence quotient (IQ) and difficult rehabilitation that is seen in most individuals with DS.[7] A recent study found significant hearing loss in 90% of DS patients compared to 50% in non-DS group and no hearing loss in controls. Also, the major cause of hearing loss in DS patients was conductive, while hearing loss in non-DS was sensorineural.[4] Our findings in this study are in agreement with the above studies which confirm the high percentage of conductive hearing loss in DS patients.

However, this pilot study needs to be verified by testing a large number of DS patients with wide age range, in order to correlate the incidence of conductive hearing loss with different age groups.

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