ORIGINAL ARTICLE

# Otological Disorders in Down's Syndrome

**S Satwant, MS (ORL-HNS), K N Subramaniam (Deceased), FRCS, N Prepageran, FRCS, R Raman, MS, Mohd Amin Jalaludin, FRCS**

Department of Otorhinolaryngology, Faculty of Medicine, Universiti Malaya, 50603 Kuala Lumpur

**Summary**

**Objective**: to assess if children with Down's Syndrome have a higher prevalence of otological abnormality compared to their normal counterparts in Malaysia.

**Methodology**: Thirty children with Down's Syndrome and normal children underwent otoscopic ear examination and Impedance test in the ENT outpatients clinic in University Hospital, Kuala Lumpur, Malaysia.

**Results**: The study showed that children with Down's Syndrome had higher otological disorders. Forty four percent had impacted wax compared to 14.4% in normal children. Twenty one percent of ears in the study group had retracted drums compared to 6.6% of control. Fifteen percent of ears in the study group had middle ear effusion compared to 3.4% in controls, 55% had a type B tympanogram compared to 8.3% in controls and 73.4% had auditory canal stenosis compared to 14.4% in controls.

**Conclusion**: Children with Down's Syndrome, thus have a higher incidence of otological disorders.

*Key Words:* Down's Syndrome, Otological disorders

## Introduction

Down's Syndrome accounts for 1% of all congenital mental retardation. Mental retardation is the only consistent feature in all children with this syndrome[1]. Hearing and speech deficiencies feature commonly in these patients, thus are particularly vulnerable to impaired learning resulting from a hearing loss and handicaps of speech retardation.

It has been well established that children with Down's Syndrome experience a high incidence of persistent conductive hearing loss due to canal stenosis, cerumen impaction, serous otitis media and cholesteatoma. Since this hearing problem may be a primary cause of the spoken communication skills in these children far below their cognitive abilities, assertive management is required.

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*Corresponding Author: N Prepageran, Department of Otorhinolaryngology, Faculty of Medicine, Universiti Malaya, 50603 Kuala Lumpur*

## Materials and Methods

This prospective study was carried out in the ENT outpatient in University Hospital, Kuala Lumpur between May 1997 to August 1998; a period of 18 months. Thirty children with Down's Syndrome whose age range from 9 months to 10 years and 30 age matched normal children were seen with parental consent. The children with Down's Syndrome were from a day school program in the Kiwanis School for Down's Syndrome children in Klang.

Two days a week were reserved to see these children; one for the initial visit and a subsequent one for follow-ups.

Parents were explained regarding the various aspects of examination and consent were obtained. The control group was obtained from Paediatric wards and all the selected patients were free from upper respiratory tract infections at least 2 weeks prior to examination.

Children undergoing chemotherapy or radiotherapy were excluded from this study. The assessment protocol consists of an otoscopic examination followed by Tympanometry with a hand held Microtymptype II tympanometer. Those who have ear wax were prescribed sodium bicarbonate ear drops and their parents were given instructions regarding the technique of instilling the ear-drops. They were reviewed again in a week's time, where the wax was removed manually where possible or syringing was carried out.

Otoscopic findings were classified as normal or abnormal based on the presence of middle ear effusion without the prior knowledge of the tympanometry results. Impacted cerumen was classified as complete or partial. Complete impaction was defined as total blockage of the ear canal where the tympanic membrane cannot be visualized.

Partial obstruction was defined as visualization of part of the tympani membrane. (Bopping Balachanda: The Human Ear Canal 1987). After removal of cerumen and ruling out tympani membrane perforation tympanometry was carried out.

The following criteria were used to diagnose middle ear effusion:

1. Retracted tympani membrane.
2. A hair line or bubble visible through tympani membrane.
3. A darker, lustreless tympani membrane as compared to the other ear.
4. Tympani membrane mobility where possible.

Tympanometry was carried out after otological examination and the parameters measured were the ear canal volume and type of tympanogram i.e. A, B or C.

They were plotted automatically by recording compliance values in cubic (mmho) across a pressure range of -200 to 400mmH20. The results were classified according to Jerger's classification (1970);

Type A: normal compliance at a normal middle ear pressure of +/- 100 mmH20
Type B: Non-compliant middle ear system
Type C: Normal compliance of negative pressure of greater than -100 H20.

The external auditory canal is considered to be stenosed if the Vea (external ear volume) is less than 0.4 ml for ages below 10. (Margoliss and Heller 1987, American Speech and Hearing Association 1990).

Once the clinical criteria were satisfied, the results were compared to the control group. Statistical calculations were used employing chi- square analysis with Yates correction method. The calculations were done based on software

obtained from Center for Disease Control (CDC), Atlanta, USA and p less than 0.05 value was taken as significant.

## Results

The results of the study was divided into a few categories:

1. Age distribution of children with Down syndrome.
2. Prevalence of impacted cerumen
3. Middle ear otoscopy
4. Tympanometry results
5. Prevalence of canal stenosis

*Age distribution of children with Down's Syndrome:*
Of the 30 children included in the study, 24 were Malays, 4 Chinese and 2 were Indians. The average maternal conception was 35 years. There was no history of consanguinity noted. Eight (27%) children were below 24 months of age, 6 (20%) between 25 to 48 months, 12 (40%) between 49 and 72 months, 3 (10 %) between 73 and 96 months and 1 child (3%) was between 97 and 120 months.

*Prevalence of impacted cerumen:*
Forty four percent of children with Down's Syndrome had impacted wax on examination compared to 14.4% of the control group. This is statistically significant ($X^2$ =11.86, p=000.6 Yates correction).

*Middle ear otoscopy:*
Only 38 (63.3%) of 60 ears in the Down's Syndrome children had normal middle ear otoscopy as compared to 54 (90%) in the control group. About 13 (22%) of 60 ears in Down's Syndrome children were found to have retracted tympani membrane compared to 4 (6.6%) of the control group. These were statistically significant ($x^2$=12.00, p=0.002).

About 9 (15.1%) ears of the study group had middle ear effusion as compared to 2 (3.4%) in the control group. These were also statistically significant.

*Tympanogram Results:*
Tympanogram results revealed that in the study group, 55% had type B non-compliant tympanogram.

Only 8.3% of the control group had type B tympanograms. Only 31.6% of the study group had type A tympanogram as compared to 86.6% in the control group. About thirteen percent of the study group had type C tracings while 5.1% had similar results in the control group. These were also significant statistically. ($X^2$ =38.24, p=0.001). This is shown in Table I.

**Table I: Tympanometry**

| | Down's Syndrome No. of ears (percentage) | Control No. of ears (percentage) |
|---|---|---|
| **Type A** | 19 (31.6%) | 52 (86.6%) |
| **Type B** | 33 (55%) | 5 (8.3%) |
| **Type C** | 8 (13.4%) | 3 (5.1%) |

*External auditory canal stenosis:*
Table II shows the result of the prevalence of external auditory canal stenosis. A canal diameter of less than 0.7 mm as defined by the American Speech and Hearing Association was taken as the standard baseline to define canal stenosis. Seventy three percent of the study group was found to have stenoses canal as compared to 14.4 % in the control group. This was again statistically significant.

**Table II: Canal Stenosis < 0.4 CC**

| | Down's Syndrome No. of ears (percentage) | Control No. of ears (percentage) |
|---|---|---|
| Normal | 14 (23.3%) | 52 (86.6%) |
| Canal Stenosis | 46 (76.7%) | 8 (13.4%) |

## Discussion

Down's Syndrome is usually diagnosed clinically using anatomical features However, in newborn, as many as 25% of the cases are overlooked in initial diagnosis[1]. Characteristic facial features provide the most recognizable clues. An abnormally shortened pinna is a relatively consistent feature in a newborn[2]. Comparison between longitudinal dimensions of the pinnae in the Down's Syndrome population and normal children shows Down's Syndrome children measuring more than two standard deviation below normal[2]. The external ear especially osseocartilaginous junction, is often narrow to the point of stenotic thus precluding adequate tympanic membrane examination by the primary care physician.

Hypotonia is a major, recognizable diagnostic sign of importance. Thus, it seems reasonable to suspect that that the generalized hypotonia might extend to the tensor veli palatini muscle and cause eustachian tube dysfunction. This condition combined with a narrow external auditory meatus might result, therefore, in a high incidence of undetected middle ear effusion in these children. In view of the higher prevalence of ear pathology and tympanometric abnormalities, it is not surprising that the incidence of hearing impairment was also much higher in the Down's Syndrome group[3,4].

Conductive hearing loss occurs most commonly secondary to congenital ossicular aberrations[5]. The high incidence of conductive hearing loss these children experience is usually a scenario of four conditions they exhibit more than normal pediatric population; stenosis of the external auditory canal, cerumen impaction, serous otitis media and cholesteatoma.

The most severe cases of external ear canal stenosis are usually found in the younger children. The stenosis, usually bony in nature is due to immature closure of the tympanosquamous sutures, creating an hour glassed shaped canal that impedes visualization of the canal and tympani membrane. This prevents free secretion of cerumen and provides a niche in which cerumen accumulates and impede air conduction hearing.

Stenosis is less common in older children for the thickened tissue recedes over a period of time. Persistence of ear canal stenosis after the age of 3 might benefit from surgical canal reconstruction. Dennis G Pappas (1994) presented a study where aggressive treatment was instituted for otological management and to develop spoken communication in children with Down's Syndrome[6]. This consists of reconstruction of the external ear canal, amplification technology and auditory- verbal therapy. They were able to produce age appropriate oral language development in the Down's Syndrome children.

It is thus of extreme importance for the primary care physician that the otological aspect of a child with Down's Syndrome is looked into and appropriate therapy carried out[7,8]. To label a Down child as slow or mentally backward without considering his hearing impairment is unfortunate.

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