

**THE INCIDENCE OF HEARING LOSS IN DOWN'S SYNDROME: A CLINICOAUDIOLOGICAL STUDY**Satish S. Raju<sup>1</sup>, Thanzeemunisa<sup>2</sup>**HOW TO CITE THIS ARTICLE:**

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**ABSTRACT: CONTEXT:** Ear, nose, and throat (ENT) problems are common in individuals with Down syndrome. High prevalence of ENT disease in Down syndrome is observed due to structural and functional abnormalities of ear, nose and throat and immune system of which hearing loss is one. Hearing loss early in life leads to language disabilities. Appropriate screening would allow hearing losses to be detected early so that early treatment can be initiated. **AIM:** To assess type and degree of hearing loss in cases of Down syndrome. **SETTING AND DESIGN:** Cross sectional study at tertiary referral centre. **MATERIALS AND METHODS:** Children clinically diagnosed as Down Syndrome (DS) aged between 3 to 20 years were evaluated. A detailed history, clinical examinations and hearing assessments such as audiogram, tympanometry and brainstem evoke response audiometry (BERA) were used to evaluate these patients. **RESULTS:** A total of 60 patients (120 ears) were enrolled for this study. This study showed that the commonest type of hearing loss were conductive in 68 ears (56.7%), mixed in 4 ears (3.3%). The other 48 ears (40%) had normal hearing. Most of them fall in the mild level of conductive hearing loss. **CONCLUSION:** Conductive hearing loss is a common feature (56.7% of ears) in DS children.

**KEYWORDS:** Down syndrome, Tympanometry, Brain stem evoked response audiometry (BERA).

**INTRODUCTION:** The incidence of Down Syndrome (DS) is approximately 1 in 700 live births<sup>1</sup>. There are studies concerning a high prevalence of hearing loss in individuals with DS. Anatomic anomalies of the eustachian tube, and the typically stenotic ear canals of children with DS, predispose them to congenital and acquired otologic problems. There is high prevalence of otitis media with effusion in these children as indicated by tympanometry.<sup>2</sup> Mucoïd secretions accumulates in the middle ear and stops the ossicles from vibrating freely, therefore reducing hearing levels. There is limited published data of Down syndrome in developing countries. Therefore the objective of this study was to assess the type of hearing loss and degree of hearing loss in Down syndrome in a tertiary Hospital.

**MATERIALS AND METHODS:** Cross sectional study done from January 2014 to May 2015 on 60 patients clinically diagnosed as Down syndrome aged 3 to 20 years.

**AIM OF STUDY:** To assess the type and degree of hearing loss in Down syndrome.

**Inclusion Criteria:** Down's syndrome cases aged 3 to 20 years.

**Exclusion Criteria:** Down's syndrome cases having active ear discharge.

**METHODOLOGY:** After a thorough prenatal, natal, developmental and treatment history, a detailed general physical examination was done. Detailed Ear examination and Otomicroscopy was carried out to look for external canal stenosis, accumulated cerumen or ear discharge. If wax present, it was removed. The status of tympanic membrane was assessed, whether perforation was present, or

## ORIGINAL ARTICLE

tympanic membrane was normal or retracted or whether effusion was seen through tympanic membrane was noted.

Nose, midface and throat examination (Mouth, teeth and jaw, presense of cleft lip and palate) was done.

Tympanometry was done in all patients. The tympanometry results were classified as type A, B, C, As and Cs. Type A is normal, type B is related to the presence of middle ear fluid and type C is related to Eustachian tube dysfunction. As indicates a type with low compliance and Cs is C with low compliance.

In children aged 3 to 5, BERA was done to estimate hearing thresholds. The test is based upon utilizing auditory stimuli to evoke an electrical response generated from the cochlea to the upper brainstem. This test provides an effective measure of the integrity of the auditory pathways up to the upper brainstem level. Above 5yrs those children who cooperated for test, pure-tone audiometry was done and those children who did not cooperate for test and had moderate to severe mental retardation were taken up directly for BERA. Audiological tests like pure-tone audiometry were conducted at our hospital. Patients were referred to private centres for Tympanometry and BERA.

The type of hearing loss was defined as conductive, sensorineural or mixed. The degree of hearing loss was defined as normal (< 25 dB), mild (26-40 dB), moderate (41-70 dB), severe (71-90 dB) and profound (>90 dB).<sup>3</sup> The average hearing threshold was calculated by averaging the threshold at 500 Hz, 1 kHz and 2 kHz.

**RESULTS:** In our study Down syndrome children were divided into two groups aged 3 to 5 years and 6 to 20 years. There were 19 children in 3 to 5 age group and 41 children in 6 to 20 age group. All children in age group of 3 to 5 were subjected to BERA. Out of 41 children in age group of 6 to 10years, 26 children patient underwent PTA and 15 children in whom PTA could not be done, BERA was done to assess hearing thresholds.

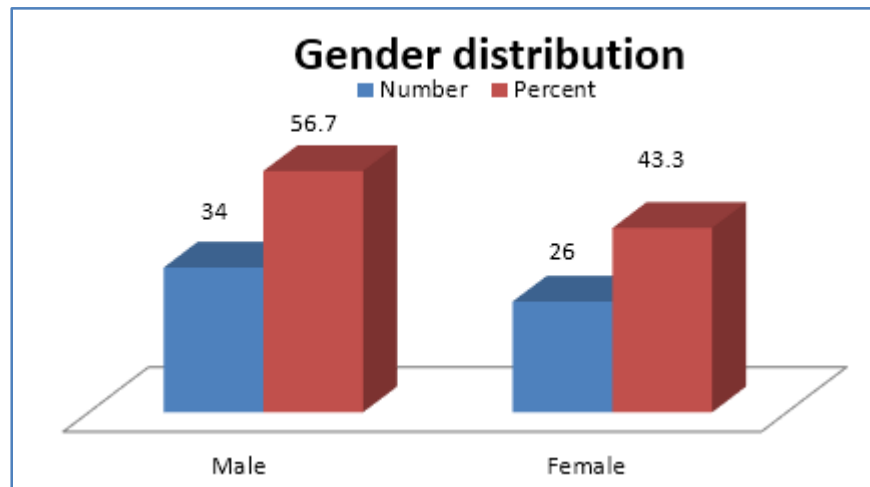
Age in years	Test	Number of patients
3-5	BERA	19
6-20	PTA	26
	BERA	15

**Table 1: Distribution according to age and test done for hearing thresholds (N=60)**

Gender	Number	Percent
Male	34	56.7
Female	26	43.3
<b>Total</b>	<b>60</b>	

**Table 2: Distribution according to gender (N=60)**

## ORIGINAL ARTICLE

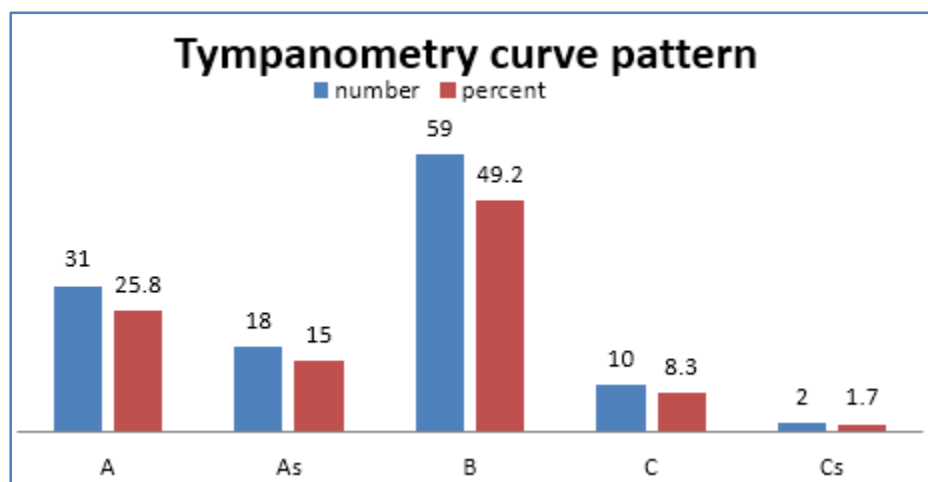


Among the 60 children with DS, 34 (56.7%) were male and 26 (43.3%) were female.

Curve type	Right	Left	Total	Percent
	Number	Number		
A	15	16	31	25.8
As	8	10	18	15
B	31	28	59	49.2
C	5	5	10	8.3
Cs	1	1	2	1.7
<b>Total</b>	<b>60</b>	<b>60</b>	<b>120</b>	

Table 3: Distribution based on tympanometry curve pattern in right and left ear

Out of the 60 patients who underwent tympanometry(120 ears), 59 ears (49.2%) had type B tympanogram, 31 ears (25.8) had A type, 18 ears (15%) had As type, 10 ears (8.3%) had C type and 2 ears (1.7%) had Cs type of curve.



## ORIGINAL ARTICLE

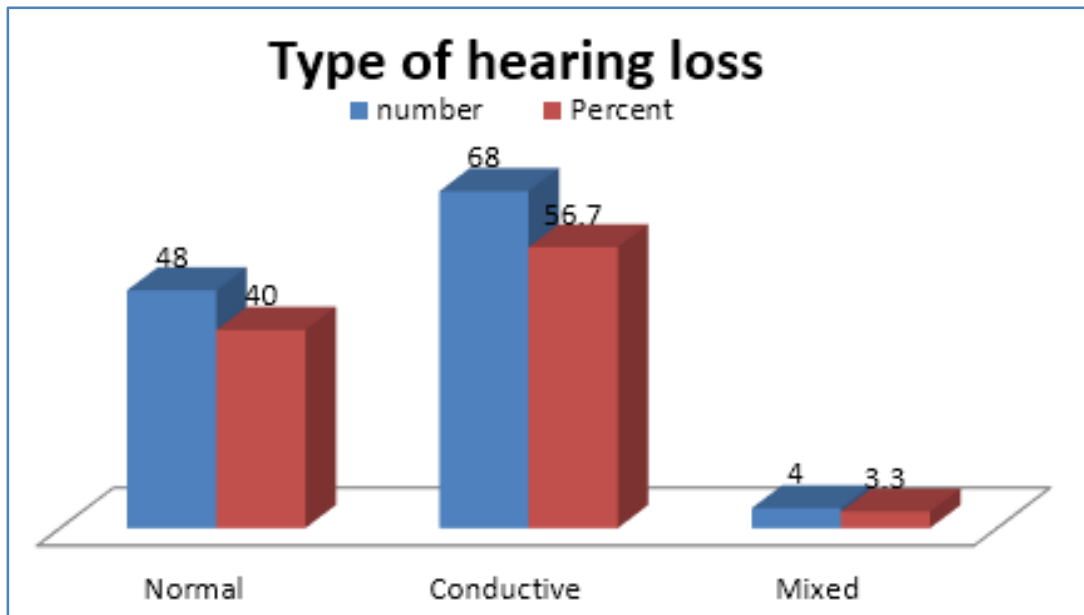
Variable	Number of patients	Percent
Both ears normal	18	30
Hearing loss only in right ear	9	15
Hearing loss only in left ear	3	5
Bilateral hearing loss	30	50
<b>Total</b>	<b>60</b>	<b>100</b>

**Table 4: Distribution according to hearing loss involving one or both ears (N=50)**

In our study out of 60 Down syndrome children tested for hearing, 18 children (30%) had both ears normal, 9 children (15%) had hearing loss only in right ear. 3 children (5%) had hearing loss only in left ear. Thus 20% had unilateral hearing loss. 30 children (50%) had bilateral hearing loss.

Type	Right Ear	Left Ear	Total	Percent
Normal	21	27	48	40
Conductive	36	32	68	56.7
Mixed	3	1	4	3.3
<b>Total</b>	<b>60</b>	<b>60</b>	<b>120</b>	

**Table 5: Distribution according to type of hearing loss in 60 patients (N=120 ears)**

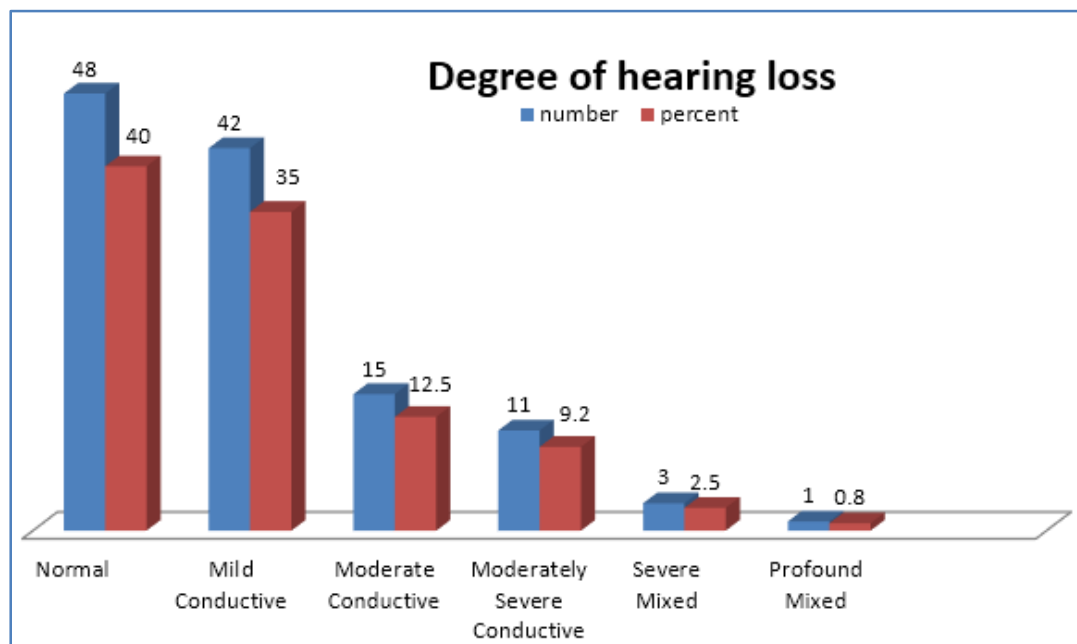


60 children examined for hearing thresholds accounts for 120 ears being examined. 48 ears (21 right and 27 left) constituting 40% showed normal hearing, 68 ears (36 right and 32 left) forming 56.7% showed conductive hearing loss and 4 ears (3 right and 1 left) forming 3.3% of total showed mixed hearing loss.

## ORIGINAL ARTICLE

Type	Number		Total	Percent
	Right ear	Left ear		
Normal	21	27	48	40
Mild Conductive	26	16	42	35
Moderate Conductive	8	7	15	12.5
Moderately Severe Conductive	2	9	11	9.2
Severe Mixed	3		3	2.5
Profound Mixed		1	1	0.8
<b>Total</b>	<b>60</b>	<b>60</b>	<b>120</b>	

Table 6: Distribution according to degree of hearing loss in 60 patients (N=120 ears)



In our study of 120 ears, 48 ears (40%) had hearing within normal limits, 42 ears (35%) of ears had mild conductive hearing loss, 15 ears (12.5%) had moderate conductive hearing loss, 11 (9.2%) of ears had moderately severe hearing loss, 3 ears (2.5%) had severe mixed hearing loss and 1 ear (0.8%) had profound mixed hearing loss.

**DISCUSSION:** Speech and hearing impairment, upper airway obstruction and ENT infections are frequently observed in DS children. Most of these ENT abnormalities are linked to functional and anatomical abnormalities in the ear, nose, and throat and immune system.<sup>4</sup>

Among the 60 children with DS, 34 (56.7%) were male and 26 (43.3%) were female.

In our study, out of 60 Down syndrome children tested for hearing, 18 children (30%) had both ears normal, 12 children (20%) had unilateral hearing loss. 30 children (50%) had bilateral hearing loss. In study by Roizen, 34% percent had normal hearing, 28% had unilateral loss, and 38% had bilateral loss.<sup>5</sup>

## ORIGINAL ARTICLE

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60 children examined for hearing thresholds accounts for 120 ears being examined. 48 ears constituting 40% showed normal hearing, 68 ears (56.7%) showed conductive hearing loss and 4 ears forming 3.3% of total, showed mixed hearing loss.

Of 68 ears with conductive hearing loss, the grading was 42 ears (35%) had mild conductive hearing loss, 15 ears (12.5%) had moderate conductive hearing loss, 11(9.2%) of ears had moderately severe hearing loss.

Of 4 ears with mixed hearing loss, 3 ears (2.5%) had severe mixed hearing loss and 1 ear(0.8%) had profound mixed hearing loss.

In study by Kattan et al, 35% were found to have normal hearing, 34 patients (65%) had hearing loss. Of these, 4% with moderately severe sensorineural hearing loss, 50% with mild to moderate conductive loss and 11.5% with moderate to severe mixed loss.<sup>6</sup>

The possible explanation of these observations are anatomical abnormalities such as chronic nasal obstruction, adenotonsillar hypertrophy, collapse of Eustachian tube and recurrent otitis media as discussed earlier. Another possible explanation of higher incidence of OME is patulous Eustachian tube, weakness of the pharyngeal musculature or poor immunological function among the DS populations. The immune system in DS children develops more slowly, predisposing them to a higher incidence of upper respiratory tract infections and thus middle ear effusion. The Eustachian tubes are often less effective in allowing drainage from the middle ear, as they tend to be narrower in children with DS.

Tympanometry were performed in all of the study population. The purpose of this test was to examine the middle ear compliance. The proportion of abnormal tympanometry in children with DS previously reported was 60-70%.

In our study, out of the 60 patients who underwent tympanometry(120 ears),59 ears (49.2%) had type B tympanogram, 31 ears(25.8%) had A type, 18 ears(15%) had As type,10 ears(8.3%) had C type and 2 ears (1.7%) had Cs type of curve.

In study by affendi et al, 69.64% had type B tympanogram, 19.65% had type C and 10.71% had a type A tympanogram.<sup>7</sup>

**CONCLUSIONS:** Conductive hearing loss is a common feature (56.7%) in DS children. The presentation of hearing loss in DS in our population is similar to previous studies.

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## ORIGINAL ARTICLE

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