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CROSS-SECTIONAL STUDY

Study of Auditory Function Changes in Patients with Down Syndrome

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Abstract

The aim of this study: Is to determine the incidence and pattern of auditory function disorders in patients with Down syndrome. In addition to describe factors that affect the severity of hearing loss.

Methods: The study involved 30 patients with Down syndrome between 3-18 years-old who had visited ENT clinic in Tishreen University Hospital (TUH) during the period from June 2021 to June 2022. A detailed history was taken, a comprehensive examination of the ear, nose and throat, and auditory tests were taken in each patient case, and the results of these tests were recorded for all patients.

Results: The percentage of hearing loss was 53.3%, as Conductive hearing loss was the most frequent type with 81.25%, sensorineural hearing loss was 12.5% and mixed hearing loss was 6.25%. The percentage of mild hearing loss was 15.6% but we didn't notice any case with profound hearing loss. The cerumen impaction was the most prevalent cause of hearing loss among patients with a rate of 33%, while cases of serous otitis media accounted for about 13.3%, and no case of acute suppurative otitis media was recorded in this study.

Conclusion: Hearing deficiency is an important problem in patients with Down syndrome. The accurate diagnosis, early and effective treatment, periodic audiological tests in Down syndrome patients is crucial in detecting problems and managing them in the best way in order to avoid affecting their life style.

Keywords

Down syndrome, Hearing loss, Tympanometry, PTA

Abbreviations

PTA: Pure Tone Audiometry, DS: Down syndrome, ET: Eustachian Tube, TM: Tympanic Membrane, OME: Otitis Media with Effusion, AOM: Acute Otitis Media, CSOM: Chronic Suppurative Otitis Media

Introduction

Down syndrome is the most common genetic syndrome in humans generally [1]. Individuals with this syndrome are considered an important part of society and they have the full right to obtain good medical care and live a normal life. This Syndrome was first described in detail by an English doctor, John Langdon Down, in 1866 [2].

Individuals with DS often have a characteristic facial appearance [3] and an increased risk of developing several medical conditions which include heart defects [4], gastroesophageal, neurological, hematological and otolaryngological and vision problems [5]. Disorders of the ears, nose and throat are common among DS children due to midfacial hypoplasia, contracted nasopharyx, Et dysfunction [6] and several anatomical, pathological ad immunological characteristics [7].

Otologic problems such as EAC stenosis, cerumen impaction, OME, CSOM with cholesteatoma [8] and inner ear dysplasia [9]. The prevalence of hearing loss has been established at 35% or more in different populations [10,11].

Although most hearing loss in DS can be attributed to conductive pathology, some patients have sensorineural and mixed hearing loss [12].

As a result, children with DS should have audiological evaluation in the first month of life if it is available and the every 6 months. If a child fails his hearing tests, the ENT doctor can determine the cause and the appropriate medical or surgical treatment [13].

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Objective

The objective of this study is to determine the incidence and pattern of auditory function disorders in patients with Down syndrome. In addition to describe factors that affect the severity of hearing loss.

Patients and Methods

A cross-sectional study involved 30 patients with Down syndrome between 3-18 years-old who had visited ENT clinic in Tishreen University Hospital (TUH) during the period from June 2021 to June 2022.

Tests included pure-tone and Impedance audiometries which were carried out by ENT specialists.

Hearing loss was diagnosed based on the World Health Organization's classification for children, defined according to the average thresholds in the 0.5 kHz, 1 kHz, 2 kHz, and 4 kHz frequencies [14].

This classification defined an average of 16-25 dB as slight; 26-40 dB as mild; 41-55 dB as moderate; 56-70 dB as moderately severe; 71-90 dB as severe; more than 90 dB as profound. In the impedance audiometer, curves were classified as type A, B, C, As or Ad.

Type A Tympanograms show a clear peak around atmospheric pressure and are common in subjects without hearing disorders. If a type A Curve displays a shallow peak, it is classified as AS (common in otosclerosis or reduced Compliance). If a type A curve displays a very sharp peak, it is classified as AD, which is observed in dislocation of ossicles or those with flaccid eardrum or scarring. Type B curves are basically shallow across the pressure range and are typical in cases of effusion or fluid in the middle ear. Type C tympanogram shows a negative peak pressure below -100 daPa, indicating negative pressure in the middle ear which associated with Eustachian tube disorders [15].

Exclusion criteria

Children with familiar hearing loss history, neonates, previous ears surgery ad Down syndrome with autism.

Results

The sample of patients in our study included 30 patients, of whom 22 were males (73.3%), and 8 females (26.6%). The ages of the patients in the study sample ranged between 3 and 18 years, with an average age of about 8.7 years. After taking a detailed medical history, clinical examination and appropriate audiological tests, we found that hearing loss was the most common symptom among the study sample with a percentage of 33.3%, followed by a sense of heaviness in the ear with a percentage of 26.6%.

The cerumen plug was the most common cause of hearing loss by up to 33%, Otitis media with effusion 13.3%, while no case of acute suppurative otitis media was recorded within the study sample (Table 1).

The results of the Tympanometry showed that type A is the most common with a rate of 65%, followed by type B with a rate of 17.3%, and the percentage of type C was about 13.8%, while no cases of type Ad were recorded within the study sample (Table 2).

The results of pure-tone audiogram (PTA) for the 16 patients over the age of 7 years showed normal results in 8 patients (50%), while bilateral hearing loss was found in 6 patients (37.5%), and unilateral in two patients only (12.5%) (Table 3).

When analyzing audiograms and hearing thresholds according to the classification of the World Health Organization for children and according to the examined ear, it was found that the hearing thresholds are normal for 56.25%, and the form of Mild hearing loss (26-40 dB) is 15.6%, which is the most common pattern, while no cases of Profound hearing loss were found within the study sample.

By analyzing the previous results according to age groups and the results of audiological tests, it was found that the rate of hearing loss among the study sample of

Table 1: Distribution of the study population according to Otologic examination (number of ears n = 60).

Ear Examination	n (%)
Normal	18 (30%)
Cerumen impaction	20 (33.3%)
EAC stenosis	3 (5%)
Absence of light cone	3 (5%)
Retracted TM	6 (10%)
OME	8 (13.3%)
AOM	0 (0%)
CSOM	2 (3.33%)

Table 2: Distribution of the study population according to tympanometry results.

Tympanometry		N (%)
Normal (Type A both ears)		17 (56.6%)
Pathological	bilateral	8 (26.66%)
	unilateral	4 (13.33%)
Excluded		1 (3.33%)

Table 3: Distribution of the study population according to PTA results.

PTA results	n (%)
Normal	8 (50%)
Hearing loss	
Bilateral	6 (37.5%)
Unilateral	2 (12.5%)
Type of hearing loss (PTA)	n (%)
CHL	5 (62.5)
SNHL	2 (25%)
Mixed	1 (12.5%)

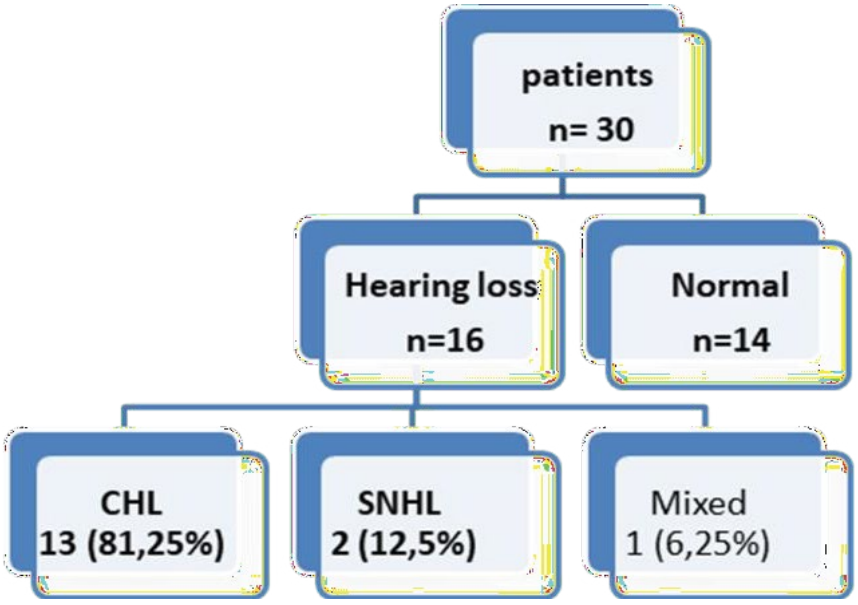

Figure 1: Hearing loss in patients.

people with Down syndrome was approximately 53.3%, and conductive hearing loss constituted approximately 81.25% of all cases, making it the most common type of hearing loss in DS patients (Figure 1).

Discussion

In this study, the ages of Down syndrome patients ranged between 3-18 years, while we find that most of the international studies conducted in America and China included children from the moment of birth [16,17].

The percentage of hearing loss in our study was 53.3%, which is a significant percentage and close to the percentage of hearing loss in both the Colombian study and the Utah University study. While the study of the International Center for Deafness in South Carolina recorded the highest rate of hearing loss among patients with Down syndrome, at a rate of 84.6% [18].

Conductive hearing loss is the most prevalent pattern among patients with Down syndrome in our study and all international studies, whether American, Colombian or Chinese [19,20].

The results of the impedance audiometry showed a high prevalence of type B and C curves due to ET dysfunction ad midfacial dysplasia. These results were found in many several international studies too.

Previous studies had shown this sort of tendency and related it to the characteristics of children with DS, such as Eustachian tube dysfunction, ossicular chain abnormalities, and other middle ear alterations.

Conclusion

This study is the first of its kind in Syria and it showed a high prevalence of hearing loss in patients with DS. The high prevalence of conductive hearing loss underpins the need for control and follow-up in this population. Follow-up should adhere to the recommendations of international organizations and encourage parental compliance.

Study Limitations

- The exceptional circumstances that the country is going through, which negatively affected the access to a larger sample size that supports the results of this research.
- The privacy of patients with Down syndrome and the difficulty of communication necessary to conduct audiological tests.

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Disclosures

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