

Research

Otoacoustic emission examination results on Down syndrome students

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ABSTRACT

Background: Down syndrome (DS) is a genetic disorder that occurs due to an excess of genetic material on chromosome 21. Down syndrome is associated with various congenital abnormalities and dysfunction of several organs, one of which is hearing. DS patients often experience sensorineural hearing loss (SNHL). Evaluation of (SNHL) in DS patients could be done in several ways, one of which is the Otoacoustic Emission (OAE) examination. **Purpose:** This study aimed to describe the results of the OAE examination in elementary school children, at special need schools in Padang City in 2022. **Method:** This was a descriptive study, using a cross sectional design. Data were collected and processed using consecutive sampling technique. The research subjects who met the criteria were 31 samples. **Result:** The majority of DS subjects received “refer” results 25/31 on OAE examination, and OAE “refer” results occurred in one ear or both ears. The “refer” results of the OAE were greater in male DS patients 20/24 than female 5/7 and more common in the 18–21-year-old age group. **Conclusion:** Persons with DS are more at risk of having SNHL.

Keywords: hearing loss, Down’s syndrome, otoacoustic emission

ABSTRAK

Latar belakang: Sindrom Down (SD) adalah kelainan genetik yang disebabkan oleh adanya kelebihan materi genetik kromosom 21. SD memiliki keterkaitan dengan berbagai kelainan kongenital dan disfungsi beberapa organ tubuh, salah satunya adalah pendengaran. Penderita SD seringkali mengalami gangguan pendengaran sensorineural. Evaluasi gangguan pendengaran sensorineural pada penderita SD dapat dilakukan salah satunya dengan pemeriksaan Otoacoustic Emission (OAE). **Tujuan:** Untuk mengetahui gambaran hasil pemeriksaan OAE pada penderita SD di sekolah luar biasa Kota Padang tahun 2022. **Metode:** Penelitian ini merupakan penelitian deskriptif dengan menggunakan desain potong lintang. Data dikumpulkan dan diolah dengan menggunakan teknik consecutive sampling. Subjek penelitian yang memenuhi kriteria sebanyak 31 sampel. **Hasil:** Mayoritas penderita SD mendapatkan hasil refer 25/31 pada pemeriksaan OAE. Hasil OAE “refer” bisa terjadi pada salah satu telinga maupun kedua telinga. Hasil OAE “refer” lebih banyak terkonfirmasi pada penderita SD berjenis kelamin laki-laki 20/24 dibanding penderita SD berjenis kelamin perempuan 5/7, dan hasil OAE “refer” lebih banyak terkonfirmasi pada kelompok usia 18-21 tahun. **Kesimpulan:** Didapatkan bahwa penderita sindrom Down lebih berisiko memiliki gangguan pendengaran sensorineural.

Kata kunci: gangguan pendengaran, Sindrom Down, otoacoustic emission

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INTRODUCTION

Down syndrome (DS) is a genetic anomaly caused by an overabundance of genetic material on chromosome 21. It is estimated that one baby is born with DS out of 800-1000 live births. In recent years, there has been reported an increase in DS cases in numerous countries, including Indonesia. According to the *Riset Kesehatan Dasar (Riskesdas)* report, DS incidence in Indonesia was gradually increasing. In 2010, it was 0.12%; in 2013, it rose to 0.13%; and in 2018, it reached 0.21%.¹ Unfortunately, there is no precise data on the number of DS patients, nationally nor provincially, in Indonesia.

DS is associated with various congenital abnormalities and dysfunction of several organs, one of which is hearing. Down syndrome sufferers often experience hearing loss, both conductive and sensorineural hearing loss.² Conductive hearing loss is a disorder of sound conduction, in DS sufferers conductive hearing loss can be caused by the ear canal which tends to narrow and has weak muscle tone; while sensorineural hearing loss are disorders that can be caused by malformations of the inner ear and vestibular organs.³ Meqbel⁴ in 2016, reported his study in special schools in the State of Kuwait, that out of the 54 primary school students who underwent the Transient Evoked Otoacoustic Emission (TEOAE) test, there were 48 (88.8%) students who failed the test.⁴ Due to the high prevalence of hearing problems in DS sufferers, it is therefore necessary to evaluate hearing function in DS sufferers at least once a year until they start school, and once every two years after adulthood.⁵

Evaluation of hearing in DS sufferers can be performed in several ways, including using an Auditory Brainstem Response (ABR) examination, or by using an Otoacoustic

Emission (OAE) examination of the Transient Evoked Otoacoustic Emission Test (TEOAE).² ABR is a hearing test that can detect hearing loss at the level of the cochlea, auditory nerve and auditory pathways in the brainstem. The OAE examination is an electrophysiological examination used to assess cochlear function.⁶ The advantages of OAE examination are including easy examination, non-invasive, practicality, objective examination results, and very efficient hearing screening programs.⁷

To ensure healthy language development and enhance the quality of life for DS sufferers, it is necessary to maximize their hearing. To make the right diagnosis and offer the right treatment, hearing screenings for DS patients are required.⁸ It is intended that by tracking the incidence of sensorineural hearing loss in DS patients, public awareness of the threat of hearing loss in this population will rise.

The authors were interested in exploring this because there had not been any research to date on the hearing screening performed on Down syndrome patients in West Sumatra. The reason for using OAE in this study was due to the advantages of this examination, including easy examination, non-invasive, practical, objective examination results, and very efficient for hearing screening programs.

METHOD

This was a cross-sectional descriptive study using consecutive sampling techniques. This research was conducted at six Special Schools in Padang City from March to September 2022. The sample in this study was the entire population that met the inclusion and exclusion criteria. Inclusion criteria consisted of willing and cooperative subjects participating in the study, and of subjects with

clean ear canals. Exclusion criteria consisted of subjects with conductive hearing loss, and conditions in the middle ear that could cause hearing loss. There were 31 samples. The subjects of this study were Down syndrome (DS) sufferers who were examined for otoacoustic emission. The diagnosis of DS was established by chromosomal examination and/or clinical examination. The characteristics of the samples collected in this study were age and gender. The data obtained were studied based on univariate analysis by analyzing the existing research variables descriptively with the help of a computerized program to calculate the frequency distribution.

This research obtained a research permit from the Faculty of Medicine, Andalas University with a research permit number: B-1894/UN16.02.WD1/PP/2022. This research also obtained ethical approval from the Faculty of Medicine, Andalas University with the certificate number for passing ethical review: 696/UN.16.2/KEP-FK/2022. This research obtained permission from the West Sumatra Education Office with letter number: 070/630/PSLB.2022.

RESULT

From March 2022 to September 2022, researchers looked at Otoacoustic Emission (OAE) tests in people with DS at Special Schools in Padang City. They observed 31 students with DS who met the requirements for inclusion and exclusion criteria. In this study, male DS sufferers composed 24 out of 31 samples, and female samples composed just 7 out of 31 samples. The age range of the samples was 5-23 years old, with the largest age range 11-14 years old, as many as 9 samples. The Shapiro-Wilk test was used to analyze the normality of the age data group, and the findings showed that the age data sample was normally distributed, with

a significance value of 0.194 ($p > 0.05$). The mean age of the sample was 15 years old. (Table 1)

The results of OAE examination on 31 samples of elementary school students at Special Schools in Padang City showed that the results of the pass in both ears were 6/31 samples, the results of referrals to one ear were 4/31 samples, and the results of referrals to both ears as many as 21/31 samples. The results of this examination showed that the majority of DS patients who were examined for OAE in this study had sensorineural hearing loss, namely 25 samples from the total sample obtained by the researchers. (Table 2)

Out of the researcher's 31 total samples, male samples were 24 and female samples were 7. The OAE examination results for male samples showed the following information: 4/24 samples had 'pass' results in both ears, 4/24 had 'refer' results in one ear, and 16/24 had refer results in both ears. Female samples had OAE examination results as followed: 2/7 samples had 'pass' results in both ears, no samples had 'refer' result in one ear, and 5/7 samples had 'refer' results in both ears, that indicated both ears needed to be referred for further examinations. (Table 3)

Based on the age of the total sample that the researcher obtained, which was 31 samples, the sample age range was 5-23 years with the largest age group being 11-14 years. The mean age of the sample was 15 years. The results of this study showed that there were 5 samples in the age range of 5-10 years, the results of bilateral OAE examination were 3 samples, there were no samples with unilateral refer OAE results, and 2 samples bilateral refer. There were 9 samples with an age range of 11-14 years, with 1 sample of bilateral passes, 1 sample of unilateral referrals, and 7 samples of bilateral referrals. There were 6 samples with an age range of 15-17 years, with 2 samples of bilateral passes and 4 samples of bilateral referrals. There

were 7 samples with an age range of 18-21 years, none with bilateral passes, 3 samples of unilateral referral OAE examination results,

and 4 samples of bilateral referrals. There are 4 samples with age > 21 years, with the results of all four being bilateral referrals. (Table 4)

Table 1. Subjects' distribution

Characteristics	N	%
Gender		
Male	24	77.4
Female	7	22.6
Age		
5-10 y.o.	5	16.1
11-14 y.o.	9	29.0
15-17 y.o.	6	19.4
18-21 y.o.	7	22.6
>21 y.o.	4	12.9

Table 2. Frequency distribution of OAE examination results

	OAE results						N	%
	Bilateral Pass		Unilateral Refer		Bilateral Refer			
	N	%	N	%	N	%		
Total	6	19.4	4	12.9	21	67.7	31	100

Table 3. Frequency distribution of OAE examination results by gender

Gender	OAE results						N	%
	Bilateral Pass		Unilateral Refer		Bilateral Refer			
	N	%	N	%	N	%		
Male	4	16.7	4	16.7	16	66.7	24	100
Female	2	28.6	0	0	5	71.4	7	100

Table 4. Frequency distribution of OAE examination results by age

Age	OAE results						N	%
	Bilateral Pass		Unilateral Refer		Bilateral Refer			
	N	%	N	%	N	%		
5-10 y.o.	3	60	0	0	2	40	5	100
11-14 y.o.	1	11.1	1	11.1	7	77.8	9	100
15-17 y.o.	2	33.3	0	0	4	66.7	6	100
18-21 y.o.	0	0	3	42.9	4	57.1	7	100
≥21 y.o.	0	0	0	0	4	71.4	4	100

DISCUSSION

The subjects of this study were 31 students with Down syndrome (DS) at special needs schools in Padang City in 2022, who had met the inclusion and exclusion criteria of this study. The result of the study showed that 25 out of 31 subjects with DS got refer OAE results which were an indication of sensorineural hearing loss; with details of 4 subjects with unilateral refer OAE results, and 21 subjects with bilateral refer OAE results. Refer result in one ear of a DS patient is an indication of sensorineural hearing loss in one ear of a DS patient, or known as unilateral SNHL. The most common cause of unilateral SNHL in children is due to structural abnormalities of the temporal bone.⁹ Malformations of the temporal bone occur in the developing primitive auditory ossicles that do not ossify. Research conducted by Ogando et al.¹⁰ in 2013 on 13 subjects with DS, found that there was a distance between the lenticular process of the incus bone and the head of stapes bone. The distance formed in the three incudostapedial joint regions of DS sufferers was significantly different when compared to normal persons. There was a widening of the incudostapedial joint distance in DS sufferers.¹¹ Refer results in both ears indicate that there is a sensorineural hearing loss in both ears of DS sufferers or known as bilateral SNHL. Bilateral SNHL disorders in Down syndrome occur due to anomalies such as reduced spiral ganglion cells in the temporal bone, and inner ear disorders such as Mondini dysplasia.¹² Based on gender, the results of this study concluded that male DS sufferers had a higher percentage of refer results than females, although not too significant. Gender differences have an influence on the otoacoustic emissions produced by vibrations arising from the organ of Corti. Spontaneous otoacoustic emission is generated by the outer hair cells in the absence of external stimulus. Spontaneous otoacoustic emission is detected in 70% of all human hearings; and spontaneous otoacoustic

emission is more likely to be detected in female subject in approximately 80% to 60% in male hearings. The exact mechanism underlying the differences in OAE test results for gender in OAE has to do with prenatal exposure to male androgen hormones.¹³ Experimental findings suggest that greater prenatal exposure to androgens will result in weakened amplifiers in the cochlea, resulting in a weaker OAE production and a slight loss of hearing sensitivity.¹⁴

Based on age, the results of this study concluded that the incidence of abnormal OAE examination results increased with increasing age of DS sufferers. Increasing age of DS sufferers affects the results of OAE examination, and the prevalence of sensorineural hearing loss increases with age.¹⁵ Along with ageing in individuals with Down syndrome, there is a progressive decrease in hearing sensitivity as the frequency increases. Sensorineural hearing loss in DS sufferers occurs 30-40 years earlier than the general population, and is estimated to occur in 12-72% of the DS population. Hearing testing is recommended for people with DS not only in childhood but also in adulthood to quickly identify and implement interventions that can help prevent negative health impacts and improve the quality of life for DS sufferers.¹⁶

The conclusion based on this study was that persons with Down syndrome were more at risk of having sensorineural hearing loss, and the risk of sensorineural hearing loss in DS increased with age, and the number was higher in male. It is highly expected that the community pay attention to detect hearing loss as early as possible in DS sufferers.

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