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EVALUATION OF HEARING IMPAIRMENT IN PATIENTS WITH DOWN SYNDROME

Contributions:
A Study design/planning
B Data collection/entry
C Data analysis/statistics
D Data interpretation
E Preparation of manuscript
F Literature analysis/search
G Funds collection

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Abstract

Introduction: It is estimated that the prevalence of hearing loss in children with Down syndrome (DS) ranges from 38% to 82%. However, the diagnosis of hearing loss in DS is difficult, due to an impaired ability to cooperate for subjective tests. Thus, objective tests such as impedance audiometry, otoacoustic emissions, and auditory brainstem responses (ABRs) may be more appropriate. In this study objective methods without anesthesia were used to determine the amount, type, and prevalence of hearing loss in people with DS.

Material and methods: The study included 39 subjects with DS, ranging in age from 1 year to 27 years. Hearing tests in DS subjects were performed during physiological sleep or while awake. Otoscopic examination was performed in all DS subjects. If no abnormalities were seen, tympanometry, otoacoustic emissions, and recording of ABRs were attempted.

Results: Objective hearing tests showed that the DS group had various types of hearing disorders. Based on all objective tests carried out in all ears ($n = 78$), the following diagnoses were made: normal hearing, 36 ears (46%); cochlear hearing loss, 27 ears (35%); conductive hearing loss, 8 ears (10%); suspected deafness, 7 ears (9%). Based on wave V thresholds, the following degrees of hearing loss were established: normal hearing, 36 ears (46%); 21–40 dB nHL, 21 ears (27%); 50–60 dB nHL, 8 ears (10%); 70–80 dB nHL, 6 ears (8%); > 80 dB nHL, 7 ears (9%).

Conclusions: In previous studies of DS subjects by other authors, conductive hearing loss predominated, followed by sensorineural hearing loss. In the present study, more sensorineural than conductive hearing loss was diagnosed. In most cases, the diagnosis of the type of hearing disorder was based on the ABR result, and other tests such as tympanometry and otoacoustic emissions played a supporting role. Due to the often limited cooperation of the patient, the DPOAE test was difficult to perform.

Key words: otoacoustic emissions • ABR • hearing disorders • Down syndrome (DS) • objective hearing tests • impedance audiometry

OCENA ZABURZEŃ SŁUCHU U PACJENTÓW Z ZESPOŁEM DOWNA

Streszczenie

Wstęp: Szacuje się, że odsetek występowania ubytków słuchu u dzieci z zespołem Downa (ZD) wynosi od 38% do 82%. Jednak diagnostyka słuchu u osób z ZD nie jest łatwa z uwagi na utrudnioną możliwość współpracy podczas wykonywania testów subiektywnych wymagających odpowiedzi ze strony pacjenta. Dlatego wydaje się, że badania obiektywne słuchu, takie jak: audiometria impedancyjna, otoemisje akustyczne, słuchowe potencjały wywołane pnia mózgu (ABR), mogą być optymalnym rozwiązaniem w przypadku diagnozowania pacjentów z zespołem Downa. Celem pracy było określenie wielkości, rodzaju i częstości poszczególnych ubytków słuchu u osób z ZD za pomocą metod obiektywnych bez konieczności stosowania anestezji.

Materiał i metody: W badaniach wzięło udział 39 osób z ZD w wieku od 1 roku do 27 lat. Testy słuchu u osób z DS wykonano podczas fizjologicznego snu pacjenta lub na jawie. Badanie otoskopowe wykonano u wszystkich osób z ZD. Jeśli nie stwierdzono nieprawidłowości w obrębie przewodu słuchowego zewnętrznego oraz błony bębenkowej, podejmowano próbę wykonania tympanometrii, pomiaru emisji otoakustycznych oraz rejestracji ABR.

Wyniki: Analiza wyników obiektywnych badań słuchu wykazała, że w badanej grupie pacjentów występują różnego rodzaju zaburzenia słuchu. Na podstawie analizy wyników wszystkich obiektywnych badań słuchu, przeprowadzonych we wszystkich uszach ($n = 78$), postawiono następujące rozpoznania: norma słuchowa – 36 uszu (46%), niedosłuch ślimakowy – 27 uszu (35%), niedosłuch przewodzeniowy – 8 uszu (10%), podejrzenie głuchoty – 7 uszu (9%). Analiza wielkości ubytków słuchu, przeprowadzona na podstawie progu fali V, pokazała następujące częstości ubytków słuchu: norma słuchowa – 36 uszu (46%), 21–40 dB nHL – 21 uszu (27%), 50–60 dB nHL – 8 uszu (10%), 70–80 dB nHL – 6 uszu (8%), >80 dB nHL – 7 uszu (9%)

Wnioski: W pracach innych autorów dotyczących osób z zespołem Downa dominował niedosłuch przewodzeniowy, a następnie odbiorczy. W niniejszej pracy w badanej grupie rozpoznano więcej niedosłuchów typu odbiorczego niż przewodzeniowego. W większości przypadków rozpoznanie rodzaju zaburzenia słuchu miało miejsce na podstawie wyników badania ABR, a pozostałe badania, takie jak tympanometria czy otoemisje akustyczne, odgrywały rolę pomocniczą. Z uwagi na często utrudniony kontakt z pacjentem badanie DPOAE było trudne do przeprowadzenia.

Słowa kluczowe: otoemisje akustyczne • ABR • zaburzenia słuchu • zespół Downa (ZD) • obiektywne badania słuchu • audiometria impedancyjna

Introduction

A special group of patients in whom objective methods have an important place in the diagnosis of hearing disorders are those with various genetically determined syndromes, including Down syndrome (DS). DS is the most common human genotype pathology. An extra autosomal chromosome at position 21 is responsible for the typical features of the disease [1–3]. As emphasised by numerous authors, hearing diagnosis in people with DS is not easy, due to an impaired ability to cooperate when performing various tests in behavioral audiometry [4–8]. Thus, objective methods are better suited to this group of people [6,7,9–11], although significant difficulties can still be encountered. Some of the problems relate to unwillingness to do the test, psychomotor overexcitability, and difficulty with ear anatomy, such as narrowing of the external auditory canal [2,12].

One possible cause of the delay in intellectual, cognitive, and language development in DS children may be impaired hearing. It is estimated that the prevalence of hearing loss in children with DS ranges from 38% to 82% [13]. Facial defects and abnormal course and narrowing of the Eustachian tubes in individuals with DS are responsible for an increased incidence of otitis media, pharyngitis, and sinusitis. A narrow external auditory canal can make it difficult to visualise the eardrum with a standard speculum [2,12]. Generalised muscular hypotonia can lead to dysfunction of the flexor muscle of the soft palate, thereby increasing the risk of acute otitis media and chronic exudative inflammation; autoimmune dysfunction also contributes to recurrent upper respiratory tract infections, which can also lead to conductive hearing loss [14,15]. Intrapiromkul and colleagues [16] pointed out anomalies in the structure of the inner ear in newborns with DS, such as distortion of the bony islets of the lateral semicircular canal, narrow internal auditory canals, narrowing of the cochlear nerve canal, dilation of the semicircular canals, and widening of the vestibular aqueduct [16]. Many researchers consider that the most common type of hearing loss in children with DS is conductive, but others point to mixed or sensorineural hearing loss [4,7,11,17–22]. A number of authors believe that hearing loss in DS children is the result of disorders of the middle ear [4,7,18,19,21,23]. It has been suggested that the causes of hearing loss in DS children include thicker earwax, frequent exudative otitis media, and ventilation or mechanical abnormalities of the middle ear [24,25].

In DS patients, the prevalence of hearing loss due to middle ear dysfunction has been found to decrease with age, but the incidence of sensorineural hearing loss increases from childhood to adulthood [18,26–28]. In many cases, subjective testing is not possible, due to a lack of cooperation on the part of the DS patient or a failure to understand the procedure and instructions during the test, thus giving a false audiological result. Thus, objective hearing tests have long been used in the diagnosis of hearing disorders in DS subjects [7,9,11,17,29].

The purpose of this study was to determine, without applying anesthesia, the amount, type, and prevalence of hearing loss in people with DS using objective methods.

Material and methods

The study included 39 subjects (78 ears) with DS aged from 1 year to 27 years (mean 10.7 ± 5.2 years), made up of 19 boys and 20 girls. More than 75% of the subjects were between the ages of 5 and 15. Statistical analysis was performed with a Student's *t*-test for independent samples, and showed no statistically significant differences in the mean age of the two groups.

To take the tests, the caregiver of the DS subject had to sign a consent form. Hearing tests in DS subjects were performed during physiological sleep or while awake, either at the child's home or at a special school. The time taken to perform all examinations on one DS patient ranged from 1 to 3 hours. Shorter examination times were observed in patients tested during physiological sleep. Otoscopic examination was performed on all DS patients. If no abnormalities were found in the external auditory canal and tympanic membrane, then tympanometry, otoacoustic emissions, and recording of auditory brainstem responses (ABRs) were attempted.

Tympanometry was carried out using an OTOflex 100 device (GN Otometrics, Tasstrup, Denmark) to rule out middle ear abnormalities. A frequency of 226 Hz at 85 ± 1.5 dB SPL was used, and the pressure range was between + 200 and – 400 daPa. Tympanograms were analysed based on Jerger's classification. Recording of distortion product otoacoustic emissions (DPOAEs) was then performed using the ILO 6 system (Otodynamics Ltd., London). For measuring DPOAEs, the probe was placed in the external ear canal and two tones of frequencies f1 and f2 were emitted (f2/f1 = 1.22). The frequencies ranged from 1 kHz to 8 kHz in half-octave steps. The intensity of the tones was 65/55 dB SPL. A signal-to-noise ratio of > 3 dB was used as a criterion for the presence of a DPOAE.

ABRs were evoked by click stimuli from a Vivosonic Integrity V500 device (Vivosonic Inc. Toronto, Canada). Electrodes were placed on the forehead and mastoid processes. The recording bandwidth was 0.03–3 kHz. Stimuli were presented with alternating polarity through Sennheiser HDA 300 in-ear headphones. The stimulus repetition rate was 37/s, with a response analysis time of 10 ms. The number of averages ranged from 500 to 2000, depending on the number of muscle artifacts, the amplitude of the response, and the intensity of the stimulus. The test

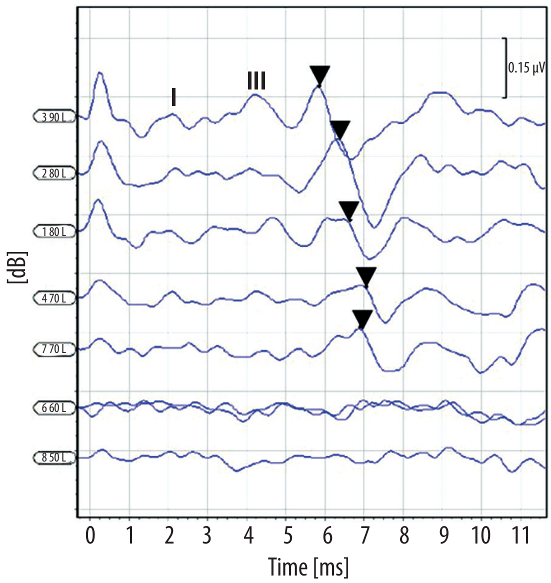

Figure 1. Example of a recording of ABRs in the left ear of a DS patient who was diagnosed with a 70 dB nHL cochlear lesion. Arrowheads mark wave V peak. The patient had a type A tympanogram

was performed using a procedure in which intensity progressed downwards in 10 dB steps from 90 dB nHL. After a recording, the peaks of waves I, III, and V were determined using a cursor, and the latency determined. A latency–intensity function was then plotted, and the hearing threshold was defined as the lowest stimulus intensity at which wave V appeared in the recording. For normal hearing, a hearing threshold criterion for ABR responses of 20 dB nHL was used.

The selection of data for analysis was guided by the principle that the ABR recordings should be of high quality, enabling reliable determination of the wave V peak and its threshold. All objective tests used in the study were non-invasive and painless. As a criterion of statistical significance, a 95% confidence level ($p < 0.05$) was chosen.

Results

Figure 1 shows an example of a recording of ABRs in the left ear obtained in a DS patient who was diagnosed with a cochlear deficit of 70 dB nHL. In this example, the patient had a type A tympanogram.

Table 1 summarises the percentage of ears in which each type of objective hearing test were performed. Tympanometry and ABR tests were performed in 100% of ears, while DPOAE testing was performed in only 63% of ears.

From the 78 ears, **Table 2** summarises the percentage of each type of result obtained with objective testing. In tympanometry, type A tympanograms were the most common, followed by type B, type C, and then type As. The percentage of tympanograms that suggested the presence of conductive hearing loss was 53%. A normal DPOAE signal was recorded in only 28% of ears. Due to difficult

Table 1. Percentages of ears in which each type of test was performed ($n = 78$)

Test type	Percent of ears in which test was performed
Tympanometry	100%
DPOAE	63%
ABR	100%

Table 2. Frequency of each type of result in the entire group of ears ($n = 78$)

Type of test and result	Percentage
Tympanometry	
tympanogram type A ($n = 40$)	47.5%
tympanogram type As ($n = 9$)	11.5%
tympanogram type C ($n = 16$)	15.4%
tympanogram type B ($n = 13$)	25.6%
As + C + B ($n = 38$)	52.5%
DPOAE	
normal ($n = 22$)	28.2%
absent ($n = 27$)	34.6%
not measured ($n = 29$)	37.1%
ABR	
normal ($n = 36$)	46.2%
conductive ($n = 8$)	10.2%
sensorineural ($n = 27$)	34.6%
residual hearing ($n = 7$)	9.0%

cooperation on the part of the subject, it was not possible to perform a DPOAE test in 100% of the ears. Patients often made loud noises during the test, which interfered with DPOAE measurements. In contrast, a normal ABR test result was present in 46% of ears. The test indicated sensorineural hearing loss in 35% of ears, and conductive hearing loss in 10% of ears.

Table 3 summarises the results of impedance audiometry and DPOAEs in the group of ears with normal ABR responses. In these ears, a normal tympanogram was present in 72% of ears, while normal DPOAEs were present in 28% of ears.

In the group of ears in which the ABR results indicated hearing loss (**Table 4**), a normal tympanogram was present in 52% of ears, while a normal DPOAE signal was present in 26% of ears.

In the group of ears in which an ABR latency–intensity plot indicated a conductive-type hearing loss (**Table 5**), 3 ears had type C tympanograms, and the remaining 5 had type B tympanograms. In 4 ears, the DPOAE test could

Table 3. Frequency of each type of tympanometry and DPOAE results in the group of ears with normal ABR responses (*n* = 36)

Type of test and result	Percentage
Tympanometry	
tympanogram type A (*n* = 26)	72.2%
tympanogram type As (*n* = 3)	8.3%
tympanogram type C (*n* = 3)	8.3%
tympanogram type B (*n* = 4)	11.2%
As + C + B (*n* = 8)	27.8%
DPOAE	
normal (*n* = 14)	27.8%
absent (*n* = 14)	27.8%
not measured (*n* = 8)	44.4%

Table 4. Frequency of each type of tympanometry and DPOAE results in ears in which ABRs indicated cochlear damage (*n* = 27)

Type of test and result	Percentage
Tympanometry	
tympanogram type A (*n* = 14)	51.9%
tympanogram type As (*n* = 5)	18.5%
tympanogram type C (*n* = 5)	18.5%
tympanogram type B (*n* = 3)	11.1%
As + C + B (*n* = 13)	48.1%
DPOAE	
normal (*n* = 8)	25.9%
absent (*n* = 9)	33.3%
not measured (*n* = 11)	40.8%

not be performed, and in 3 ears there was no response in the DPOAE test.

Table 6 summarises the percentage of the various types of tympanogram among subjects whose hearing loss was diagnosed on the basis of an ABR examination. For those cases with a normal ABR test result, normal tympanograms were present in 72% of ears. The other types of tympanograms occurred much less frequently. In ears in which the ABR test result indicated a cochlear disorder, a normal tympanogram was also most common, but B and C type tympanograms were also quite common. In ears in which the ABR test result indicated a conductive-type disorder, no normal tympanograms were found (only types B and C).

Only 50 ears (64%) were able to have both tympanometry and DPOAE tests performed. Furthermore, in only 20 ears (40%) were the diagnoses consistent with those made on the basis of the ABR test. Based on an analysis of the results of all objective hearing tests conducted in all ears (*n* = 78), the following diagnoses were made which are presented in **Table 7**.

Table 5. Results of tympanometry and DPOAE in ears in which ABRs indicated a conductive type hearing loss (*n* = 7)

Type of test and result	Percentage
Tympanometry	
tympanogram type A (*n* = 0)	0.0%
tympanogram type As (*n* = 0)	0.0%
tympanogram type C (*n* = 3)	42.9%
tympanogram type B (*n* = 4)	57.1%
C + B (*n* = 7)	100.0%
DPOAE	
normal (*n* = 0)	0.0%
absent (*n* = 3)	42.9%
not measured (*n* = 4)	57.1%

A combined analysis of all the results of objective hearing tests showed that there was no need to correct any diagnosis made solely on the basis of the analysis of the wave V threshold and the latency–intensity function of ABRs. Analysis of the levels of hearing loss based on wave V threshold is presented in **Table 8**.

In the analysed group, ears with a normal threshold and with minor hearing loss (up to 40 dB nHL) predominated. In total, there were 73% of this type of ear. Hearing thresholds of 50 dB nHL and larger accounted for 27%. The results indicate that at least 30% of those examined had a mixed hearing loss (in 30% patients with sensorineural hearing loss, the tympanograms were incorrect).

Discussion

Objective hearing tests are important for diagnosing hearing status in young children and patients in whom reliable audiometric test results cannot be obtained. A special group of patients for whom objective methods are central are patients with Down syndrome. In these patients, audiometric methods fail in most cases, so objective methods are necessary.

Despite their advantages, objective hearing tests also have limitations. First and foremost, the patient needs to be calm. Otherwise, recordings can be strongly disturbed by acoustic and muscle artifacts, making the results unreliable. This problem greatly affects DS patients.

Objective hearing tests have long been used to diagnose hearing disorders in people with DS [7,9,11,17,29]. It should be noted, however, that practically all these works used various types of anesthetics, and even general anesthesia, when testing. Such an approach is also common in Polish audiology centers. In the present work, we wanted to examine DS subjects without the use of anesthesia in order to demonstrate that, with an appropriate amount of time and organisation, reliable objective hearing tests can be performed. We found that due to lack of cooperation from some of the patients – due to mental retardation and motor hyperactivity, among other factors – it was impossible to perform DPOAE tests on 100% of them. Patients

Table 6. Percentage of each type of tympanogram in subjects diagnosed by ABR with different types of hearing loss

Diagnosis based on ABR examination	Tympanogram type			
	A	As	C	B
Normal (*n* = 36)	26 (72.2%)	3 (8.3%)	3 (8.3%)	4 (11.2%)
Sensorineural (*n* = 27)	14 (51.9%)	5 (18.5%)	5 (18.5%)	3 (11.1%)
Conductive (*n* = 8)	0	0	3 (37.5%)	5 (62.5%)
Deafness suspected (*n* = 7)	0	1 (14.3%)	5 (71.4%)	1 (14.3%)

Table 7. Diagnoses based on the analysis of the results of all objective hearing tests conducted in all ears

Diagnosis	Number of ears	Percentage
Normal hearing	36 ears	46.1%
Sensorineural hearing loss	27 ears	34.6%
Conductive hearing loss	8 ears	10.3%
Deafness suspected	7 ears	9.0%

who were tested while awake often made loud noises during DPOAE testing, so that DPOAE signals suffered strong acoustic interference.

Each of the objective methods evaluates different parts of the auditory pathway. Each test helps form a picture of what is wrong with the hearing system. Of the three objective methods, only the ABR method allows differential diagnosis of hearing disorders. The other methods are less specific. For this reason, the emphasis in this paper has been on the results obtained by the ABR method, and we recommend its central role.

The results of previous studies have shown a predominance of conductive hearing loss, followed by sensorineural hearing loss. However, in the present study, more sensorineural than conductive hearing loss was diagnosed. The difference probably relates primarily to the age structure of the subjects studied. If the material is dominated by young children then the proportion of conductive disorders will be higher [9,29]. If, on the other hand, the subjects are mostly adults then there will be more sensorineural defects, as found in [30].

The results of this paper indicate that there was never a need to amend a diagnosis made solely on the basis of the analysis of wave V threshold and the latency–intensity function of ABRs. In other words, ABRs should be regarded as the primary method, and other methods are only supplementary.

Table 8. Incidence of different levels of hearing loss based on ABR wave V threshold

Level of hearing loss	Number of ears	Percentage
Normal hearing	36 ears	46.1%
30–40 dB nHL	21 ears	26.9%
50–60 dB nHL	8 ears	10.3%
70–80 dB nHL	6 ears	7.7%
> 80 dB nHL	7 ears	9.0%

Our own studies, as well as those of other authors [7,9,18,31,32], have shown that not all objective tests can be performed in every case, but under good testing conditions (e.g. while asleep or performing quiet tasks), ABRs can usually be performed. In most cases, the diagnosis of a hearing disorder can be based on an ABR test result, with other tests such as tympanometry, wideband tympanometry (WBT), or otoacoustic emissions playing only a supporting role [7,9,18,30,32,33].

However, despite the fact that the ABR test was able to make a good diagnosis, adding other objective tests does provide greater certainty and gives better insight into the functioning of individual sections of the auditory pathway, even if the disorders are minor.

Conclusions

Based on our data from 39 subjects with Down syndrome, the following conclusions can be made.
1) The ABR method is crucial for diagnosing hearing loss objectively.
2) It is possible to perform ABR testing without anesthesia.
3) DPOAE testing is often difficult to perform, due to poor cooperation on the part of the patient.

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