

The evaluation of the usefulness of objective methods for testing of hearing in patients with Down syndrome

Abstract

Introduction

The group of patients with genetically conditioned disorders, including patients with Down syndrome, is a special group of patients, which is characterized by significant position of objective methods in diagnosis of hearing disorders. Taking such features of objective methods as their non-invasiveness, painlessness, short examination time and high sensitivity and specificity it seems that they are particularly indicated also in examination of Down syndrome patients, which are characterized by the fact that it is very hard, if not impossible, to conduct audiometric examinations with them.

Different authors report that in case of Down syndrome patients difficulties may arise in performing impedance audiometry and otoacoustic emissions. Among others the above is the result of the specific structure of ear canal, denser earwax, and frequent otitis media with effusion (OME). Clinical experience indicates that the method free of those limitations is the auditory evoked potentials (AEP) method. Within the group of AEPs the most significant ones for clinical practice are the auditory brainstem responses (ABR). Nevertheless even this method can encounter significant difficulties in performing the examination, due to lack of test acceptance or excessive motor activity.

Taking the aforesaid into account we must assume that we will not be able to perform all objective tests for every case, but we can still expect to perform, provided that we use optimal conditions for examinations, at least the auditory brainstem responses test. We can thus assume, that for majority of cases the diagnosis of hearing disorders will be based on ABR examination, and the remaining test will have an auxiliary function only.

The ABR diagnostic of hearing disorders is based on the analysis of the V wave threshold, the course of latency-intensity function of wave V, and the time intervals of waves I-III and III-V. That is why the analysis of conductivity in auditory nerve and brainstem, and the development of benchmark graphs for latency-intensity function are crucial issues.

Objective of the work

The main objective of the present work was to evaluate the applicability of objective methods, and ABR in particular, to diagnostics of hearing of Down syndrome patients.

The detailed objectives of the work were as follows:

- analysis of the neuronal transmissions in auditory nerve and brainstem in case of DS patients in comparison with persons with normal hearing and no neurological disorders
- analysis of the latency-intensity function in case of Down syndrome patients
- evaluation of the degree and type of hearing disorders in DS patients
- evaluation of the applicability of respective objective methods to DS patients
- evaluation of the possibility of diagnosing hearing disorders of DS patients with use of ABR and remaining methods

Material and method

Material included 39 persons with Down syndrome (DS group), aged 1 to 27 (average 10.7 ± 5.2 years), including 19 boys and 20 girls, and 112 persons (58 girls and 63 boys) with normal hearing (aged 1 to 35, average age 9.8 ± 5.7 years), who consist the control group – the Group N. Hearing tests of the Down syndrome patients were conducted during physiological sleep or during waking, at children's home or special educational needs school. The persons from control group were examined at the Institute of Physiology and Pathology of Hearing in Warsaw, during waking stage or physiological sleep. The control group was selected from persons with normal otoscopic examination results, normal audiogram (whenever this was performed) and normal results of the respective objective hearing tests. Otosopic examinations were also performed for all Down syndrome patients. The interviews with parents or caretakers of Down syndrome patients revealed that in no case there was a reliable result of audiometric examination at hand. These persons were also not diagnosed for hearing disorders, although in many cases periodic hearing conduction disorders were reported.

In the group of Down syndrome patients tympanometry was performed with use of Otometrics OTOflex 100 device, whereas in control group the same device or Madsen Zodiac 901 were used. The tympanograms were analyzed pursuant to Jerger classification. The transiently-evoked otoacoustic emission TEOAE test was performed in both groups with use of ILO 6 system (Otodynamics Ltd., London). We applied the criterion of signal to noise separation of over 3 dB for evaluation of OAE signal. The examinations of click evoked auditory brainstem responses were performed in both groups with use of the same Vivosonic Integrity V500 devices, for intensity ranging from 20 to 80 dB nHL. The biological amplified band ranged from 30 to 3000 Hz. The stimuli were presented with alternating polarity with use of Sennheiser HDA 300 headphones. The stimulus repetition rate was 37/s and the analysis time was 10 ms. Depending on the number of myogenic artifacts the averaging number was ranged from 500 to 2000. For responses recorded for 80 dB nHL the peaks of I, III, V waves were marked with cursor and the interpeak-intervals were calculated. For responses recorded for

intensity ranging from 70 dB nHL to threshold the peak of wave V was marked and then the latency-intensity function determined.

Results

The average value of the I-III interval for the group of Down syndrome patients was 2.07 ± 0.13 ms and was significantly shorter ($p < 0.05$) than in control group (2.19 ± 0.14). The values of III-V intervals in both groups showed no significant differences ($p > 0.05$). In both groups the average values of the I-III interval were significantly longer than the average values of the III-V interval.

Due to the fact that the I-III time interval was shorter in the group of Down syndrome patients we compared the latency-intensity function characteristics of subjects with normal hearing and with Down syndrome, who had a wave V threshold that was not higher than 20 dB nHL (DS NI, $n = 36$ ears) and had a normal tympanogram.

The slope of latency-intensity function of subjects with Down's syndrome was steeper than that of subjects with normal hearing. This fact means that in case of differential diagnosis of hearing disorders based on latency-intensity function graph we should utilize the latency-intensity function graph of the DS NI group. Based on average latency-intensity function graph for this group and with aid of the reports by Kochanek (2002) concerning the influence of different severities of cochlear impairment on the shape of latency-intensity function graph we determined reference graphs for conductive and cochlear impairments of different severities. Based on those reference graphs we determined the types of hearing loss in the group with Down syndrome with a wave V threshold in excess of 20 dB nHL. The analysis of latency-intensity function graphs in this groups showed that 34,6% cases were that of cochlear impairment and 10,2% that of conductive disorders. In no ear prolonged I-III and III-V intervals were diagnosed.

In the analyzed group of Down syndrome patient ears with normal threshold and slight hearing loss (up to 40 dB nHL) were predominant, with a total of 73% of such cases. Hearing losses in excess of 50 dB nHL and larger accounted for 27% of cases.

100% of ears were correctly diagnosed with tympanometry, 62,8% with otoacoustic emission, and 100% with ABR examination. The most frequent tympanograms were that of A type, followed by B and then C and As. The percentage of different types of tympanograms that suggested the presence of conductive hearing disorders 52,5%. Normal otoacoustic emission test result was recorded in 29.5% cases only. Normal ABR test result was recorded in 46,2% of cases. The analysis of all cases showed that just 50 ears (64,1%) were successfully diagnosed with both test – tympanometry and otoacoustic emission. But only in case of 20 ears (40%) these diagnoses were concurrent with diagnoses set on the basis of ABR exam. The total analysis of all results of objective hearing tests demonstrated that there was no need to correct the diagnoses made solely on the basis of the threshold of wave V and the course of intensity-latency function of auditory brainstem responses.

Conclusions

Based on the tests performed we conclude as follows:

1. Neuronal transmission in auditory nerve is faster in case of DS patients than in case of persons with no hearing loss.
2. The auditory brainstem neuronal transmission in the Down syndrome group are the same as in control group.
3. The slope of latency-intensity function of subjects with Down's syndrome was steeper than that of subjects with normal hearing. .
4. Different characteristic of changes of wave V latency in the function of click intensity in case of Down syndrome patients with normal wave V threshold, compared to the control group indicates different representation of cochlea activity for the same amplitudes in both groups.
5. Within the analyzed group of Down syndrome patients ears without hearing loss were predominant.
6. Cochlear hearing loss was predominant in case of Down syndrome patients with wave V thresholds in excess of 20 dB nHL.
7. The auditory brainstem responses examination is essential for objective diagnosis of hearing loss of Down syndrome patients.