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The Results of Hearing Screening at the Children with Down Syndrome in Andijan Region of Uzbekistan

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Abstract

This article presents the primary results of targeted hearing screening among children with Down syndrome, as one of the most common pathologies at risk of developing various forms of hearing loss. As a result of the screening according to the data of delayed otoacoustic emission and otoacoustic emission distortion product, out of the total number of children in 41.5 of cases, objective tests showed the presence of auditory dysfunction, i.e. the answer is refer. Thus, we found it expedient to carry out hearing screening in children with Down syndrome in order to prevent hearing loss, and secondary forms of delayed psychoverbal development associated with it.

Index terms— down syndrome, auditory screening, hearing loss, children with down syndrome, objective methods of audiometry.

1 Introduction

According to the European Down Syndrome Association (EDSA), 80% of people with Down syndrome of various ages have some form of hearing impairment, however, there are also cases of sensorineural hearing loss. Epidemiological studies show that the prevalence of moderate to profound hearing loss in children, including sensorineural hearing loss and conductive hearing loss, is up to 6:1000, with 10% of children having profound hearing loss [1].

Children with Down syndrome are at higher risk of hearing loss than their normally developing peers. According to the literature, 1.4 per 1000 newborns and 5 per 1000 children aged 3 to 7 years have hearing loss (Centers for Disease Control and Prevention, National Center for Birth and Developmental Defects, Division of Birth Defects and Developmental Disabilities 2013).

According to numerous studies, up to 20% of children may have sensorineural hearing loss caused by defects in the development of the inner ear and auditory nerve. They have narrow nasal passages, a small mouth, and tongue deviation. However, conductive hearing loss is observed in up to 70-78% of children with Down syndrome. The pathogenesis of conductive hearing loss is based on the anatomy and physiology of the maxillofacial skeleton, nasopharynx. Thus, the horizontal location of the Eustachian tube in combination with muscular hypotension that occurs in diabetes causes frequent diseases of the middle ear, in particular exudative otitis media (EOM).

Modern pediatric audiology today allows for an objective assessment of hearing from the first days of a child's birth [2,3,4]. Such objective audiological tests as short-latency evoked auditory potentials (SAEP), delayed otoacoustic emission (DOAE), MultiASSR allow to obtain hearing thresholds in children regardless of age and psychological state. An important factor is the age aspect. It is well known that the so-called "second signaling system" is active in children under 3 years of age, and by the age of 5 it weakens in its physiological significance [5,6,7]. The preschool age of the child is therefore considered to be the most relevant and vulnerable in terms of hearing and further associated speech development, both for children from the general group and even more so for children with Down syndrome. Thanks to objective audiometry tests, hearing diagnosis in this age group of children has become possible, even in the presence of concomitant neuropsychiatric diseases [8,9]. Thus, early diagnosis of hearing impairment in children with Down syndrome will allow not only adequate treatment and correction in a timely manner, but also to prevent STDs associated with auditory dysfunction, which in turn will guarantee an improvement in the quality of life of children with this pathology. However, the type and degree of

hearing loss remain unexplored. Moreover, pathological changes in the central parts of the auditory analyzer in children with Down syndrome have not been studied and studied, which is reflected in the impairment of speech intelligibility with the preserved and normal functioning of its peripheral parts.

Thus, the fact of early diagnosis of hearing in children with Down syndrome is socially and economically justified, determining the timely correction, the effectiveness of subsequent rehabilitation in order to prevent secondary forms of delayed psychoverbal development in this group of children.

2 II.

3 Materials and Methods

The examination included 50 children with confirmed Down's syndrome, registered in the neurological dispensary, as well as in the departments of neurology of the Andijan regional multidisciplinary children's center and at the department of neurology of the clinic of the Andijan State Medical Institute. At the same time, the age of the children was t: 0-1 year 5 children (3 boys and 2 girls), 1-3 years 24 children (12 boys and 12 girls), 3-5 years 8 children (5 boys and 3 girls), 5-7 years 7 children (4 boys and 3 girls), 7-10 years 3 children (2 boys and 1 girl), 10< years 3 children (1 boy and 1 girl). The genetic analysis performed confirmed: 47 children had trisomy in 21 pairs (94%), 2 children had a mosaic form (4%), 1 child had a translocation form, 27 boys and 23 girls (2%). At the first stage, auditory screening was carried out using DOAE based on the Otoread clinical screening audiometer (Interacoustics) In this case, broadband acoustic clicks presented with a repetition rate of 20-50/s served as stimuli. DOAE registration criteria: the ratio of the emission power to the background noise power in three or more frequency bands, which was at least 3 dB. After reaching the required signal-to-noise ratio, the computer automatically stopped the work, issuing the "PASS" indicator -"test passed", if there was no answer after 1000 repetitions, the "REFER" indicator appeared -the test was not passed.

At the second stage, children with the result "failed" underwent complex objective audiometry under natural sleep -SAEP both in air and bone conduction, stationary evoked potentials -Multi-ASSR, game audiometry, and also acoustic impedancemetry. In-theear telephones with pre-fitted ear tips served as the source of sound stimuli. Cup silver chloride electrodes were used to record brain responses. The electrodes were fixed on the nasal region (ground electrode), on the border of the scalp (reference electrode) and in the region of the mastoid processes on the right and left (active electrodes). In the studies, the interelectrode resistance did not exceed 5 k Ω , which was achieved by preliminary treatment of the patient's skin and the use of special conductive gels. Various types of stimuli were used during the ABR, including an acoustic click with a duration of 100 ms, tone signals with a frequency of 1000, 4000, 2000 and 500 Hz, a broadband Chirp stimulus, and octave-filtered (frequency specific) Chirp stimuli. For analysis, it is recommended to record at least 2 runs (graphs) for each intensity value (to avoid false interpretation of artifacts).

4 III.

5 Results and Discussion

Out of 50 children with Down syndrome, DOAE showed the result of "refer", i.e. the response "failed" was registered in 24 children, which indicated the presence of auditory dysfunction in comparison with the control group. The absence of registration of DOAE at the same time on both sides was observed in 11 children (32.3%), and unilateral in 13 children (67.6%).

Accordingly, these 24 children with Down syndrome were subsequently referred for a comprehensive hearing diagnosis using objective audiological tests.

Behavioral (playing) tone audiometry was performed only in adult children older than five years and with convincing evidence of the adequacy of their response to the examination, providing reliable results only in 8 children (21.6%).

Impedance tests are quite informative, but their use has been limited by the high incidence of stenosis of the external auditory canal. The analysis of the obtained tympanograms was carried out according to the international Jerger classification. According to the data of the conducted tympanometry at 226 Hz, type "A" tympanogram, i.e. as normal, were obtained in 28 ears (28%). Type "B" was identified in 16 ears (16%), and type "As" was registered in the remaining 4 ears (%).

Tympanometry data at 1000 Hz in the same group of patients revealed type "B" in 23 ears (23%), type "A" in 15 ears (15%) and type As in 10 ears (10%). Stapedial reflexes were often absent even in the presence of type A tympanogram. This fact can be explained by severe weakness (hypofunction) of the tubal muscles.

Data on SAEP (CHIRP) detection of the V peak at a level above 30dB was shown in 11 ears (11%), 45-50dB in 28 ears (28%), making up the majority, in 2 ears (in one child with Down syndrome) was determined at the level 90-95 dB, which corresponded to bilateral sensorineural deafness. In the remaining 9 ears (9%), normal hearing threshold values were determined.

However, a comparative analysis of the SAEP data and the conducted tympanometric data showed greater information content and comparability when performing 1000 Hz typometry. Thus, according to our research and the data of foreign researchers on this issue, it is 1000 Hz tympanometry that is the most diagnostically reliable and valuable in this group of patients. Namely, it is necessary to take into account the features of the anatomy

and physiology of the nasopharynx, the structure and pathology of the middle ear, as well as the maxillofacial skeleton in children with Down syndrome. The authors describe hypotheses: a narrow external auditory canal and an immature cartilaginous structure of the auricle reduce the level of sound transmitted to the tympanic membrane and middle ear system. Different types of fabrics show specific levels of resistance to each frequency, resulting in different levels of sound transmission. In addition, the sound compliance is determined by the mass of the structures of the middle ear and the effect of the rigidity of the skeletal system. The mass of the tympanic membrane, bone components (hammer, anvil, stirrup) determine mass reactions. The flexibility of the tympanic membrane, the bony transmission mechanism, and the tension of the perilymphatic fluid on the footrest of the stirrup determine the stiffness element. When considering the anatomical and physiological conditions, the adult middle ear is defined as a system with adjustable stiffness at low frequencies, while the infant middle ear system is a mass-dominated system and requires more pressure. In the case of children with Down syndrome, this mechanism is more persistent and has almost the status of a constant in all age groups: a specific horizontal location of the Eustachian tube, prolonged presence of mesenchymal tissue in the tympanic cavity, stenosis of the external auditory canal, hypoplasia of the mastoid process, nasal passages, small mouth, language deviation. IV.

6 Conclusion

Thus, according to the data obtained, the following can be done:

1. In children with Down syndrome, complex objective hearing diagnostics can reveal even latent (minimal hearing impairments), which requires further audiological monitoring;
2. The incidence, type, and degree of auditory dysfunction is often unrelated to the genetic type of Down syndrome;
3. The most common form of hearing loss in children with Down syndrome is conductive hearing loss (39%), which is determined by objective tests and confirmed by tympanometry at 1000 Hz;
4. Taking into account the anatomical and physiological features of the structure of the ENT organs in children with Down syndrome, tympanometry at 1000 Hz shows greater information content and sensitivity than at 226 Hz.

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- [Grigoriev Konstantin Ivanovich et al. ()] ‘Down syndrome: comorbidity and program goals in the work of a pediatrician with such children’. Grigoriev Konstantin Ivanovich , Olga Vykhristyuk , Donin Igor Filippovna , Zavadenko A Mikhailovich , A N Zavadenko . *Grigor’ev Konstantin Ivanovich, Vykhristyuk Ol’ga Filippovna, Donin Igor’ Mihajlovich*, 1-2, 2017. 2017. 15 p. . (Trudnyj pacient)
- [Shott (ed.) ()] *Hearing loss in children with Down syndrome // International Journal of Pediatric Otorhinolaryngology*, S R Shott . 2.?. 1-6. Shott S.R (ed.) 2000. 2001. 5 p. . (Down syndrome: Common pediatric ear, nose and throat problems //Down Syndrome Quarterly)
- [Balkany et al. ()] ‘Ossicular abnormalities in Down’s syndrome’. T J Balkany , R E Mischke , M P Downs , B W Jafek . *Otolaryngology: Head and Neck Surgery* 1979. 87 p. .
- [Madjidova Ya and Salikhova ()] ‘Possible pathogenetic mechanisms of progression and the occurrence of intellectual and cognitive impairment in patients with Down syndrome’. . N Madjidova Ya , S M Salikhova . *International Journal of Psychosocial and Rehabilitation* 2020. 24 (1) p. .
- [Marcell ()] ‘Relationships between hearing and auditory cognition in Down’s syndrome youth’. M M Marcell . 10.3104/reports.54. <http://dx.doi.org/10.3104/reports.54> *Downs Syndr Res Pract* 1995. 3 (3) p. .
- [Marcell ()] ‘Relationships with between hearing and auditory cognition in Down’s syndrome youth’. M M Marcell . *Down Syndrome Research and Practice* 1995. 2003. 3 (3) p. .
- [Baranova Yu ()] ‘Samsonova // Proceedings of the XVI Congress of Pediatricians of Russia’. . A Baranova Yu . *Samsonova // Materialy XVI Kongressa pediatrov Rossii «Aktual’nye problemy pediatrii*, 2012. 2012. 80. (Actual problems of pediatrics. Baranova YU.A. s soavt., N. A. Benis,)
- [Hunter et al. ()] ‘Wideband acoustic immittance in children with Down syndrome: prediction of middle-ear dysfunction, conductive hearing loss and patent PE tubes’. L L Hunter , D H Keefe , M P Feeney , D K Brown , J Meinzen-Derr , A M Elsayed , J M Amann , V Manickam , D Fitzpatrick , S R Shott . 10.1080/14992027.2017.131455. <https://doi.org/10.1080/14992027.2017.131455> *International journal of audiology* 2017. 56 (9) p. .
- [Novikov et al. ()] ‘YU. I. Barashnev «Sindrom Dauna. Mediko-geneticheskij i social’no-psihologicheskij portret’. P V Novikov , I Yu , P V Barashnev ; Novikov . *Rossijskij vestnik perinatologii i pediatrii*, 2007. 2007. 52 p. . (Medical-genetic and socio-psychological portrait)