CLINICAL AND NEUROLOGICAL FEATURES OF CHILDREN WITH CONGENITAL AND ACQUIRED SENSORIAL HEARING LOSS.

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Abstract: Sensorineural hearing loss (NSHL) is one of the diseases, the diagnosis, and prognosis of the development of which do not lose their relevance [1, 4, 5]. The development of the second signaling system of children largely depends on the state of hearing, since through hearing the child receives new information from the outside world. In children with hearing loss, hearing impairment affects their general and mental development, the development of speech and coordination in space [1, 3, 7].

According to the literature, in 82% of cases, hearing loss in children occurs in the first year of life, that is, in the prelingual period or during the formation of speech. Of this number, only 38.5% of disorders are detected in the perinatal period, and only gross hearing impairments appear. However, it should be noted that there are no data on mild to moderate hearing loss. This is due to the late appeal to specialists, as well as the lack of proper attention from pediatricians (in one-third of cases, mild and moderate hearing loss is detected only at the age of 3-7 years).

According to the World Health Organization, the population with hearing impairments has increased by 24% by 2020. The problem of sensorineural hearing loss and deafness in children is currently the focus of the attention of researchers.

According to various authors, congenital sensorineural hearing loss is determined on average in 82–83% of children in the general structure of children with hearing loss. Among the causes that caused the occurrence of NSHL in 41.8% of cases were genetic mutations leading to non-syndromic (48.1%) and syndromic hearing pathology (13.8%). In 5.9%, NSHL was due to intrauterine infections, such as CMV, herpes, toxoplasmosis, and influenza. In 5% of cases, ante- and intranatal fetal hypoxia was detected (5%). Among the causes that caused NSHL were: a deep degree of prematurity - in 3.1% of cases; congenital anomalies in the development of the inner ear - in 3% of cases; anomalies in the development of the outer and middle ear, malformations of the maxillofacial skeleton - 2.7% of cases. Despite numerous studies in which the ototoxic effect of aminoglycizide antibiotics has been proven, in pediatric practice, one can often observe the unjustified use of this group of drugs in pregnant women and young children. In 1% of cases of NSHL in newborns was due to ototoxic drugs during pregnancy. Among the causes of NSHL, hemolytic disease of the newborn was identified (0.7% of cases). [10,11]

The "critical age" for the development of speech, psycho-emotional skills, and the formation of thinking is the age of the child up to two years. The absence of a hearing-speech environment during the first two years of a child's life has an irreversible impact on his further ability to use the potential of his residual hearing. Mental retardation is formed in cases where a hearing disorder in a child was not detected at an early age. At the same time, the physical health of the child is directly dependent on the state of the functional development of the brain.

Keywords: visual impairment, sensorineural hearing loss, neurological disorders.

Purpose: to study clinical and neurological features in patients with sensorineural hearing loss.

Materials and methods: 105 children were examined. Of these, 67 were children with congenital NSHL and 38 were children with acquired NSHL. To characterize the state of the central nervous system, data on the neurological status of the examined patients were used. All patients underwent an extended clinical and neurological examination, SAEP, EEG, and MRI of the brain.

The subjects underwent an audiological examination by a computer audiometric method. Depending on the genesis of the development of hearing loss, the patients were divided into two groups: the 1st group - patients with NSHL that arose as a result of exposure to infectious-toxic factors in the perinatal period (congenital hearing impairment), the 2nd group - with NSHL formed

as a result of exposure to exogenous environmental factors in postnatal ontogenesis, including infectious diseases transferred at an early age (acquired hearing loss).

Registration of the total bioelectrical activity of the brain (EEG) was carried out using a computer encephalograph. When examining patients, short-lateral auditory evoked potentials were recorded on a four-channel computer device "Neuro-MEP". To register SAEP, monoauricular acoustic stimulation was performed using headphones, with a rectangular tone stimulus with a frequency of 10 Hz. The stimulus intensity was selected individually based on 70 dB above the subjective threshold and ranged from 100 to 120 dB.

We analyzed the absolute latencies of peaks I, II, III, IV, V, VI, peak-to-peak intervals I-III, III-V, I-V, peak amplitudes as peak-to-peak I-Ia, III-IIIa, IIIa-IV, IV-V, V- Va, Va-VI, as well as the amplitude ratio of the peaks I-Ia/III-IIIa, I-Ia/V-Va and III-IIIa/V-Va.

Results: The reason for patients to visit a specialist was complaints about the lack of speech and reaction to sounds, and violation of the stages of psychoverbal development, as a result of which sensorineural hearing loss of varying degrees and deafness were detected in these patients.

Hearing loss II degree was diagnosed in 9 (16%) of the examined, III degree - in 11 (17%), and IV degree - in 40 (67%).

Tabl. 1

Pathological condition in children during the neonatal period, %

Pathological conditions	**1 group, n=68**	**2 group, n=37**
Asphyxia during childbirth	36	25*
Hyperbilirubinemia	3	72,5
Prematurity	49*	50
The use of ototoxic drugs	28	67**
Viral infections	13	37
Anemia	19	30

Note: * - p-values are < 0.05 **- p values are < 0.001

Asphyxia during childbirth among the examined groups 1 and 2 occurred in 36 and 25%, respectively, hyperbilirubinemia - in 72.5 and 3%. Ototoxic drugs were used in 67% and 28% of children, prematurity occurred in 49 and 50%.

Most children with NSHL were found to have diffuse organic neurological symptoms, as well as central insufficiency of the VII and XII pairs of cranial nerves, anisoreflexia, and revitalization of tendon reflexes.

In the examined patients, individual analysis of the parameters of the SAEP waves was carried out, provided that the intensity of the stimulating signal was counted from the hearing threshold of each patient (dB SL).

Tabl. 2

Latent periods of ABR waves when measuring the sound pressure value from the hearing threshold (dB SL) in persons with sensorineural hearing loss, M±m

dB	**Waves SAEP**		
	I	III	V
10	3,34±0,07 (80)	6,04±0,10 (80)	8,32±0,13 (80)
20	1,98±0,04 (80)	4,32±0,14 (80)**	6,26±0,13 (80)**
30	1,91±0,03(80)**	4,17±0,08 (80)**	5,96±0,09 (80)**
40	1,88±0,02 (80)**	3,94±0,06 (80)**	5,96±0,09 (80)**
50	1,85±0,03 (80)**	3,86±0,04 (80)**	5,77±0,06 (80)**
60	1,83±0,03 (65)**	3,75±0,04 (65)**	5,69±0,04 (65)**

Note: * - p-values are < 0.05 **- p values are < 0.001

By the results obtained in children aged 2-5 years with hearing impairments, the maximum for the P1 component was recorded in the occipital region. At the same time, in children with congenital sensorineural hearing loss, the registration of the maximum P1 component was detected in the right hemisphere, and in children with acquired it was found in the left area of the brain.

In children aged 3-6 years with NSHL, two maxima of the amplitude values of the N1 component were present in the frontal and left occipital regions. In addition, in these areas of the brain, an asymmetry in the formation of the N1 component was found: in children with acquired NSHL - mainly in the right, in children with congenital - in the left hemisphere.

When studying the results obtained in children with acquired sensorineural hearing loss, a wider display of interhemispheric connections was found than in children with congenital hearing loss or deafness.

Tabl. 3

The bioelectric activity of the brain in groups of patients, %

EEG data	1 group, n=68	2nd group, n=37
Delayed maturation	40,0	28,5
Diffuse changes	30,0*	28,2*
Focal changes	25,0	14,2
Paroxysmal activity	10,0*	21,4
Epileptiform activity	15,0	-
Variant of the age norm	-	-

Note. * - pvalue<0.001; ** - significant EEG differences between groups (p <0.001).

In 69.6% of patients, bilateral EEG asymmetry and asynchronization were detected, and a wide range of changes in the regulatory and organic genesis of mild to moderate severity was diagnosed.

The obtained EEG data in children with congenital NSHL indicate that they have more changes that are significant in the bioelectrical activity of the cerebral hemispheres than in children with acquired NSHL. In 25% of children of the 1st and 14.2% of the 2nd group, focal changes are mainly represented by outbreaks of slow-wave activity.

Tabl. 4

MRI in the examined groups, %

MRI data	1group, n=68	2 group, n=37
Expansion of the subarachnoid spaces	61,1	31,7
Ventriculomegaly	61,1	31,7
Focal lesions of white matter and basal ganglia	27,9	7,3
Periventricular changes	16,8	4,7
Anomaly of development	2	-

In children with congenital NSHL, there was an expansion of the subarachnoid spaces (55.6%), ventriculomegaly (55.6%). Among children with congenital NSHL, developmental anomalies were noted in 2%, which characterizes a violation of the maturation of the nervous tissue in the background of intrauterine lesions.

Conclusions

1. Risk factors for the development of early hearing impairment in children are related to marriage (72.8%), the use of ototoxic drugs during pregnancy, as well as in the perinatal period (70%, 67%), hyperbilirubemia (72.5%).

2. An in-depth examination of children with hearing impairment using clinical, otoneurological, ophthalmic functional studies makes it possible to determine the structure of various factors that lead to hearing impairment and influence the further course of the disease.

3. Sensorineural hearing loss is characterized by disorganization and disruption of the configuration of the component composition of I, III, and V SAEP waves.

4. In children with sensoneural hearing loss, the process of formation of the alpha-rhythmic activity of the electroencephalogram is impaired relative to the control group

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