

**DIAGNOSIS AND TREATMENT, EPIDIMIOLOGY ANOMALIES IN THE
DEVELOPMENT OF THE AURICLE**

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Annotation: Anomalies in the development of the auricle are a group of congenital pathologies that are characterized by deformity, underdevelopment, or the absence of the entire shell or parts of it. Clinically, it can be manifested by anotia, microtia, hypoplasia of the middle or upper third of the cartilage of the outer ear, including folded or fused ear, lop-eared, split lobe and specific anomalies: "satyr's ear", "macaque's ear", "Wildermuth's ear". Diagnosis is based on anamnesis, objective examination, sound perception assessment, audiometry, impedancometry or ABR test, and computed tomography. The treatment is surgical.

Key words: satyr's ear, macaque's ear.

Abnormalities in the development of the auricle are a relatively rare group of pathologies. According to statistics, their frequency in various parts of the world ranges from 0.5 to 5.4 per 10,000 newborns. Among people of the Caucasian race, the prevalence rate is 1 per person. 7 000 – 15 000 babies. In more than 80% of cases, violations are sporadic. In 75-93% of patients, only 1 ear is affected, including the right ear in 2/3 of cases. Approximately one-third of patients with malformations of the auricle are combined with bone defects of the facial skeleton. In boys, such anomalies occur 1.3-2.6 times more often than in girls.

Defects of the external ear are the result of violations of intrauterine development of the fetus. Hereditary malformations are relatively rare and are part of the genetically determined syndromes: Nager, Treacher-Collins, Konigsmark, Goldenhar. A significant part of the anomalies in the formation of the ear shell is due to the influence of teratogenic factors. The disease is provoked:

- Intrauterine infections. They include infectious pathologies from the TORCH group, the pathogens of which are able to penetrate the hematoplacental barrier. This list includes cytomegalovirus, parvovirus, treponema pallidum, rubella, rubella virus, types 1, 2 and 3 of herpes virus, toxoplasma.
- Physical teratogens. Congenital anomalies of the auricle are potentiated by ionizing radiation during X-ray examinations, prolonged exposure to high temperatures (hyperthermia). Less often, radiation therapy for cancer, radioactive iodine acts as an etiological factor.
- Bad habits of the mother. Relatively often, a violation of the intrauterine development of a child is provoked by chronic alcohol intoxication, narcotic substances, the use of cigarettes and other tobacco products. Among drugs, the most significant role is played by cocaine.
- Medications. A side effect of some groups of pharmacological drugs is a violation of embryogenesis. These medications include tetracycline antibiotics, antihypertensive medications, iodine- and lithium-based medications, anticoagulants, and hormonal medications.
- Diseases of the mother. Anomalies in the formation of the auricle can be caused by metabolic disorders and the work of the mother's endocrine glands during pregnancy. The list includes the following pathologies: decompensated diabetes mellitus, phenylketonuria, thyroid lesions, hormone-producing tumors.

Pathogenesis

The basis for the formation of anomalies of the ear shell is a violation of the normal embryonic development of mesenchymal tissue located around the ectodermal pocket - I and II gill arches. Under normal conditions, the precursor tissues of the outer ear are formed by the end of

7 weeks of intrauterine development. At the 28th obstetric week, the appearance of the outer ear corresponds to that of a newborn baby.

The influence of teratogenic factors in this time period is the cause of congenital defects of the auricle cartilage. The earlier a negative impact is made, the more severe its consequences. Later injuries do not affect the embryogenesis of the auditory system. Exposure to teratogens for up to 6 weeks is accompanied by severe defects or complete absence of the shell and the outer part of the ear canal.

In clinical otolaryngology, classifications are used based on clinical and morphological changes in the auricle and adjacent structures. The main goals of dividing pathology into groups are to simplify the assessment of the patient's functional capabilities, the choice of treatment tactics, and the decision on the need and expediency of hearing replacement. The R. Tanzer classification is widely used, which includes 5 degrees of severity of auricular anomalies:

- I – anotia. It is a total absence of tissues of the shell of the outer ear. As a rule, it is accompanied by atresia of the auditory canal.
 - II-microtia or complete hypoplasia. The auricle is present, but it is very underdeveloped, deformed, or lacks certain parts. There are 2 main options:
 1. Option A-a combination of microtia with complete atresia of the external ear canal.
 2. Option B-microtia, in which the ear canal is preserved.
 - III-hypoplasia of the middle third of the auricle. It is characterized by underdevelopment of anatomical structures located in the middle part of the ear cartilage.
 - IV-underdevelopment of the upper part of the auricle. It is morphologically represented by three subtypes:
 1. Subtype A-collapsed ear. There is an inflection of the curl forward and down.
 2. Subtype B-ingrown ear. It is manifested by the fusion of the upper part of the posterior surface of the shell with the scalp.
 3. Subtype C – total hypoplasia of the upper third of the shell. The upper parts of the whorl, the upper pedicle of the protivozavitka, the triangular and navicular fossa are completely absent.
 - V – lop-eared condition. A variant of congenital deformity, in which there is a fascination with the angle of application of the auricle to the bones of the cerebral part of the skull.
- The classification does not include local defects in certain areas of the shell-the curl and earlobe. These include Darwin's mound, the "satyr's ear", and a bifurcated or enlarged lobe. Also, it does not include a disproportionate increase in the ear due to cartilage tissue – macrotia. The absence of these variants in the classification is due to the low prevalence of these defects in comparison with the above-mentioned anomalies.

Symptoms

Pathological changes can be detected already at the time of birth of the child in the delivery room. Depending on the clinical form, the symptoms may differ significantly. Anotia is manifested by agenesis of the shell and the opening of the auditory canal – in their place there is a shapeless cartilaginous tubercle. This form is often combined with malformations of the bones of the facial skull, most often – the lower jaw. In microtia, the shell is represented by a vertical roller shifted forward and up, with a lobe at the lower end. With different subtypes, the ear canal may persist or be overgrown.

Hypoplasia of the middle of the auricle is accompanied by defects or underdevelopment of the curl leg, tragus, lower leg of the protivozavitka, bowl. Anomalies in the development of the upper third are characterized by "bending" of the upper edge of the cartilage outward, its fusion with the parietal tissues located behind. Rarely, the upper part of the shell is completely absent. The auditory canal is usually preserved in these forms. With lop-eared ears, the outer ear is almost completely formed, but the contours of the shell and anti-scall are smoothed out,

and the angle between the skull bones and the cartilage is more than 30 degrees, which is why the latter is somewhat "protruding" outwards.

Morphological variants of earlobe defects include an abnormal increase in comparison with the entire shell, its complete absence. When bifurcation occurs, two or more flaps are formed, between which there is a small furrow that ends at the level of the lower edge of the cartilage. Also, the lobe can grow to the skin located behind.

Anomaly of the curl development in the form of a Darwin's tubercle is clinically manifested by a small formation in the upper corner of the shell. With the "satyr's ear", there is a sharpening of the upper pole in combination with a smoothing of the curl. With the "macaque ear", the outer edge is slightly enlarged, the middle part of the curl is smoothed or completely absent. "Wildermuth's ear" is characterized by a pronounced protrusion of the anti-curl above the level of the curl.

Complications

Complications of anomalies in the development of the auricle are associated with untimely correction of deformities of the auditory canal. The presence in such cases of pronounced conductive hearing loss in childhood leads to deafness or pronounced acquired disorders of the articulatory apparatus. Cosmetic defects negatively affect the child's social adaptation, which in some cases causes depression or other mental disorders.

Stenosis of the lumen of the external ear impairs the removal of dead epithelial cells and earwax, which creates favorable conditions for the vital activity of pathogenic microorganisms. As a result, recurrent and chronic external and middle otitis media, meningitis, mastoiditis, and other bacterial or fungal lesions of regional structures are formed.

Diagnostics

The diagnosis of any pathology in this group is based on an external examination of the ear area. Regardless of the variant of the anomaly, the child is referred for consultation with an otolaryngologist to exclude or confirm violations on the part of the sound-conducting or sound-receiving apparatus. The diagnostic program consists of the following studies::

- Assessment of auditory perception. Basic diagnostic method. It is carried out with the help of sounding toys or speech, sharp sounds. During the test, the doctor evaluates the child's response to sound stimuli of different intensity in general and from each ear.
- Tonal threshold audiometry. It is shown to children older than 3-4 years, which is due to the need to understand the essence of the study. With isolated lesions of the external ear or their combination with pathologies of the auditory ossicles, the audiogram shows a deterioration in sound conduction while maintaining bone conduction. With concomitant abnormalities of the cortical organ, both parameters are reduced.
- Acoustic impedancometry and ABR test. These studies can be conducted at any age. The aim of impedancometry is to study the functional capabilities of the tympanic membrane and auditory ossicles and to detect a malfunction of the sound-receiving apparatus. If the study is not sufficiently informative, an ABR test is additionally used, the essence of which is to assess the response of the central nervous system structures to a sound stimulus.
- CT scan of the temporal bone. Its use is justified in cases of suspected severe malformations of the temporal bone with pathological changes in the sound-conducting system, cholesteatoma. Computed tomography is performed in three planes. Also, based on the results of this study, the question of the feasibility and scope of the operation is decided.

The main method of treatment is operative. Its goals are to eliminate cosmetic defects, compensate for conductive hearing loss and prevent complications. The selection of the technique and scope of the operation is based on the nature and severity of the defect, the presence of concomitant pathologies. The recommended age of the intervention is 5-6 years. By

this time, the formation of the auricle is over, and social integration does not yet play such an important role. The following surgical techniques are used in pediatric otolaryngology::

- Otoplasty. Restoration of the natural shape of the auricle is performed in two main ways – using synthetic implants or an autograft taken from the cartilage of the VI, VII or VIII rib. Tanzer-Brent surgery is being performed.
- Meattimpanoplasty. The essence of the intervention is the restoration of patency of the auditory canal and cosmetic correction of its entrance opening. The most common method is the Lapchenko method.
- Hearing replacement. It is advisable for severe hearing loss, bilateral damage. Classic prostheses or cochlear implants are used. If it is not possible to compensate for the conductive hearing impairment with the help of meattimpanoplasty, devices with a bone vibrator are used.

Prognosis and prevention

The prognosis for health and cosmetic results depend on the severity of the defect and the timeliness of surgical treatment. In most cases, it is possible to achieve a satisfactory cosmetic effect, partially or completely eliminate conductive hearing loss. Prevention of anomalies in the development of the auricle consists of planning pregnancy, consulting a geneticist, rational medication intake, quitting bad habits, preventing exposure to ionizing radiation during pregnancy, timely diagnosis and treatment of diseases from the group of TORCH infections, endocrinopathies.

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