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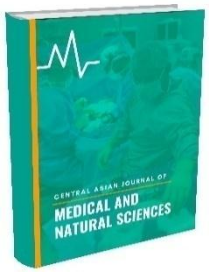

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Results of Management of Patients with Congenital Developmental Defects of the External Ear

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Annotation. Rehabilitation of patients with congenital defects is a challenge for a specialist of any profile. The key to success in this difficult matter is a multidisciplinary approach, a trusting relationship between doctor and patient, and the participation of loved ones. The need for information, support and often surgical intervention is high, some of the procedures required are very complex, and the results obtained are variable. However, to date there is no centralized national program for the rehabilitation of this group of patients and standards for providing them with medical care. This work is an attempt to generalize our experience of monitoring patients with congenital malformations of the external and middle ear, to determine the forms and timing of assistance provided, and to evaluate the effectiveness of treatment.

Key words: external ear, atresia, stenosis.

Introduction. The main types of congenital pathology of the external and middle ear are microtia and atresia/stenosis of the external auditory canal [1].

Microtia is usually unilateral (right-sided microtia is more common). Its incidence is 1 in 10,000 newborns [2]. The random nature of the occurrence of microtia predominates; sometimes (15%) it is part of the genetic syndromes of Goldenhar and Treacher Collins [3].

The incidence of atresia, according to different authors, varies from 0.92 to 1.72 per 10,000 live newborns [4 - 8]. Variability is due to differences in diagnostic standardization between countries and differences in the time periods of studies. The right-sided nature of the process, as with microtia, is more common; men suffer in a greater percentage of cases [9]. There is no clear correlation between the degree of disruption of the structure of the outer and middle ear [5].

Various classifications found in the literature are based on surgical, radiological, histopathological and functional indicators. Some of them can be used for selection for surgical treatment, others are more theoretical in nature. From the point of view of functional surgery, the most interesting are the classifications of Marx, Altman, Market, Cremers (cited from [10]).

Classification of microtia, the simplest: I degree - all anatomical structures, but the form has been changed, ear loss with or without pathology of the external auditory canal; II degree - part of the anatomical structures is damaged or absent, the curl is changed, the lobe is present, often with pathology of the external auditory canal; III degree - the upper part of the auricle is represented by a skin-cartilaginous ridge, the altered lobe is usually combined with atresia of the external auditory canal; IV degree - anotia.

Patients with malformations of the outer and middle ear have two sets of problems: *functional* (it is difficult to wear glasses and hearing aids, hearing is impaired, stenosis causes the formation of cerumen plugs, cholesteatomas, recurrent infections are observed) and *psychophysiological* (decreased self-esteem, impaired socialization, learning difficulties, behavioral disturbance, disturbance of sound localization) [11]. The way to solve them is surgical.

Rehabilitation of patients with underdevelopment of the auricle . In the treatment of patients with congenital defects of the external and middle ear, two main directions can be distinguished - hearing improvement and aesthetic correction.

An important issue is the timing of ear surgery. Some authors believe that reconstruction of the external auditory canal should precede cosmetic surgery [10]. This opinion is based on the fact that success requires a clear understanding of the initial relative position of the temporomandibular joint and the rudiment of the auricle, and also that after auriculoplasty the possibility of manipulating skin flaps and accessing the middle ear will be difficult. However, it has been proven that the auricular framework can be successfully manipulated to create the ear canal and access the tympanic cavity, and performing auriculoplasty primarily ensures that adequate regional blood supply is maintained. In our clinic, if it is necessary to perform both functional and cosmetic interventions, aesthetic correction is carried out at the first stage, then the external auditory canal is formed.

The timing of auriculoplasty may depend on whether the process is unilateral or bilateral, the severity of psychosocial disorders, the degree of microtia, and the method of treatment. For unilateral defects, surgery is performed at the age of 11-12 years after the growth of a healthy auricle has completed to maintain symmetry. With bilateral pathology, the operation can be performed at 8-9 years old, but we are of the opinion that the optimal age is 11-12 years old, when the child develops a more mature self-esteem, personal preferences regarding his appearance, as well as costal the cartilage reaches a sufficient size, which is important when performing plastic surgery using autologous tissue. When using autocartilage, the operation is performed in 2 stages: in the first stage, a cartilage graft is taken and installed in the temporal region, in the second stage, the auricle is retracted using a fragment of cartilage left during a previous operation in a subcutaneous pocket on the chest . Canaloplasty is performed at the abduction stage. The most difficult is the second stage. Initially, a Filatov stem was used to form the postauricular fold, but due to the large thickness of the formed auricle, this practice was abandoned in favor of the use of free split flaps [12].

Implantation of a silicone framework can be performed in one stage. The frequency of use of artificial materials is limited by the high risk of extrusion even with a small defect in the integumentary tissue due to injury due to rapid infection and, as a consequence, the need to remove the implanted element. Implantation of costal cartilage is devoid of this feature, since it has the ability to revascularize, and treatment of open injuries occurs as in the case of injury to the ear's own cartilage. Previously widely used in plastic surgery, allochondral cartilage showed its failure due to pronounced resorption within 2 years after surgery.

It is also possible to create a removable auricle on implanted supports or implanted magnetic retainers. This method involves the complete removal of rudimentary structures of the outer ear and does not allow subsequently choosing another method of rehabilitation. The advantages of this method of

auriculoplasty are a relatively simple, minimally invasive operation and the ability to create a fairly realistic auricle. But there are significant disadvantages: the need to remove the structure every day to toilet a given area, the possibility of losing the outer part when playing sports and active games; There may also be difficulties in masking the transition from the auricle to the skin, especially with seasonal changes in the color of the skin; It is possible that an inflammatory process will develop in the area of the implanted fixing elements, limiting the wearing of the frame until it is completely relieved. In addition, the prosthesis needs to be replaced as it wears out.

Functional rehabilitation . In case of unilateral atresia with normal hearing on the healthy side, the question of hearing-improving measures can be postponed until the child is 12 years of age. Despite publications about impairments in the function of sound localization and speech intelligibility in noisy environments in such patients, in general there are no serious adaptation disorders.

In case of bilateral disease, the earliest possible correction using bone sound conduction devices on a soft bandage is indicated for timely psycho-speech development. In the future, it is necessary to make a choice between implantation of a bone sound conduction system, and in some cases, a middle ear implant, and reconstruction of the external auditory canal and tympanic cavity. Selection of patients for reconstructive intervention requires careful assessment of audiological data and radiological criteria. The most common is the evaluation of computed tomograms on a 10-point scale [13]. We use a 26-point scale that we developed [14], which provides an individual approach and takes into account the different contributions of individual factors to the overall indicator. According to R. Jahrsdoerfer [13], no more than 50% of patients with atresia of the external auditory canal can become candidates for reconstructive surgery, especially for syndromic diseases with craniofacial malformations, since the middle ear is often poorly developed.

More often, canaloplasty is performed for stenosis of the external auditory canal. According to R. Cole and R. Jahrsdoerfer [15], if the width of the bony part of the external auditory canal is less than 2 mm, cholesteatoma develops in children under 12 years of age in 91% of cases. In this case, surgical treatment is performed for children aged 7-8 years or upon detection of cholesteatoma.

There are three main approaches for creating the external auditory canal. The “anterior” approach begins near the temporal line just behind the temporomandibular joint. The cells of the mastoid process are not opened, and the posterior wall of the external auditory canal is also preserved. A passage into the epitympanum opens through the atresia zone. Anatomical landmarks are the dura mater, the roof of the tympanic cavity and the glenoid fossa. This approach avoids injury to the often abnormally located facial nerve. But removal of the atresia zone requires care to avoid acoustic trauma due to noise and vibration when using the bur. “Posterior” access is carried out through anthromastoidotomy. Anatomical landmarks are the meninges of the middle cranial fossa, the sigmoid sinus and the sinodural angle. The atresia zone is removed before the mesotympanum is opened. In the “modified anterior” approach, an antrotomy is first performed to visualize the short process of the incus. Then the bone mass in front of the formed cavity is removed, preserving the bone wall between the mastoid cavity and the formed auditory canal.

We use a modification of the “posterior” approach without a wide mastoidotomy, with exposure of the incus-stapedial joint from the antrum. Due to the high risk of injury to the facial nerve, the intervention is usually carried out with monitoring of the facial nerve, in complex cases using navigation equipment.

The reported best outcome from such interventions is quite variable and depends on the severity of the anomaly. About 30% of patients require hearing aids after surgery. According to K. Teufert and A. de la Cruz [16] and K. Nishizaki et al. [17], up to 30% of cases are accompanied by restenosis of the formed ear canal; according to our data, no more than 12%.

Implantable bone conduction systems . For more than 35 years, implantable bone sound conduction systems have been successfully used in the rehabilitation of patients with congenital anomalies of the outer and middle ear. A big advantage is the possibility of using sound processors of various powers to compensate not only conductive hearing loss, but also mixed hearing loss, which occurs much less frequently in this group of patients with bone conduction thresholds up to 65 dB HL.

The importance of hearing aids for bilateral pathology from a very early age is beyond doubt, but there are still different opinions on this issue. R. Powell et al. [18] reported on performing the operation on a 2-year-old child. In Nijmegen (Netherlands), for a long time, implantation was carried out from the age of 10, although later the age limit was reduced to 5 years. Taking into account the characteristics of bone tissue in children with hearing defects, we install a support for wearing the device starting from 4-5 years of age.

For this purpose, a two-stage surgical procedure was proposed: implantation with installation of a support 4-6 months after completion of the osseointegration process. A single-stage procedure was introduced in 1989, followed by modifications at other major centers [19]. In adult patients, the operation is easily performed under local anesthesia, in children - under general anesthesia. The entire process is unified thanks to a specially developed set of tools.

In our department, the operation is performed in one stage. At this time, this is a safe option, taking into account the constantly developing technology for the production of titanium implants, which ensures a reduction in engraftment times, as well as the ability to control the dynamics of stability thanks to OsstellTM equipment (Sweden).

Among the large number of options for approaching the implantation site, types and shapes of incisions, the most reliable, in our opinion, is an arcuate incision away from the point of removal of the support above the skin, providing the opportunity to manipulate more freely in the surgical field and select the implantation site with maximum bone thickness immediately during surgery, ensuring good healing of postoperative wounds with minimal complications from soft tissues. Thanks to the hydroxyapatite coating of modern supports, the need to excise the soft tissue of the skull in the area of implantation has disappeared, which provides a good cosmetic result.

Recently, for conductive hearing loss, implantation of closed systems with magnetic pads without removing the supports above the skin has been used. However, for the mixed nature of the pathology, supports derived through the skin remain relevant.

Management of patients at the stage of surgical treatment . In addition to competent selection and an individualized approach to choosing a method of rehabilitation, it is necessary to take into account a number of features of patient management at the stage of surgery and in the early postoperative period. Since the goal of treatment is not to eliminate life-threatening conditions, surgery can only be performed if the patient's general somatic status is good. Children with syndromic pathology are characterized by structural features of the respiratory apparatus, especially with hemifacial microtia, which can complicate intubation for surgery under anesthesia. There is a need to consider the possibility of intubation using fiber optics and extubation in the intensive care unit.

When performing reconstructive operations on both the auricle and the external auditory canal, the average bed-day is 12 days, during which physiotherapeutic treatment, care of surgical wounds, and "nursing" of skin grafts are carried out.

When using implanted bone sound conduction systems, the length of hospital stay can be reduced to 3-4 days if outpatient care for the postoperative wound is possible after discharge. If not, then the period of hospitalization is 7-9 days. When performing surgery, you need to remember the need to preserve

the maximum volume of intact skin and good vascularization in the temporal region for subsequent auriculoplasty.

In the long-term postoperative period, it is necessary to be able to monitor the patient and solve problems if they arise, since specialists in the regions rarely have sufficient experience in the rehabilitation of patients in this group.

Evaluation of the effectiveness of the treatment . Recently, criteria for quality control of treatment for all pathological conditions have been actively developed. For the rehabilitation of patients with congenital forms of hearing loss, we use objective and subjective indicators. Objective ones include achieving a good functional result according to pure tone threshold audiometry in a free sound field and speech tests, a low frequency of complications in the form of extrusion, local inflammatory reactions. To monitor subjective indicators, we widely use the Glasgow Children Benefit Inventory (GCBI) questionnaire adapted for Russian-speaking countries [20].

As an illustration of monitoring the effectiveness of treatment, we present our study.

56 patients with congenital malformations of the outer and middle ear were examined, with average airborne sound conduction thresholds of 63.9±4.8 dB nHL. Meatotympanoplasty was performed in 32 patients aged 6–17 years. Hearing thresholds improved significantly (to 33.3±10.5 dBnHL) after surgery and decreased slightly at follow-up - to 39.4±5.7 dBnHL (p <0.001). Questionnaire scores using the GCBI ranged from 10.2 to 46.4 points. 24 patients aged 5-17 years were implanted with Baha bone conduction systems. Hearing thresholds improved to 31.2±3.4 dB hHL and remained stable throughout the observation period. GCBI scores ranged from 31.6 to 78.2 points. Thus, the use of bone sound conduction systems has shown greater effectiveness of rehabilitation compared to reconstructive interventions. We also use GCBI to assess the effectiveness after aesthetic correction, where the use of any objective criteria has not been developed and is extremely difficult. When assessing patient satisfaction with surgical treatment, the best results were achieved when using autocartilage - the total score was 44.7, more modest indicators when using a silicone frame - 36 points. The lowest result was noted in the group where allocartilage was used - 28.3 points.

Conclusion _ When a child is born with an anomaly in the development of the outer and middle ear, the main issue is compensation for hearing loss. In case of unilateral damage (normal hearing on the healthy side), rehabilitation measures can be postponed until 10-12 years of age. In case of bilateral hearing loss, prosthetics with bone conduction systems from infancy, implantation of a bone sound conduction apparatus after 4 years, reconstruction of the external auditory canal in case of isolated atresia/stenosis or after 10-12 years in the presence of microtia are indicated.

In case of stenotic auditory canal, canaloplasty is performed at 7-8 years or any age upon detection of cholesteatoma of the external auditory canal, a recurrent inflammatory process. Cosmetic surgery is performed after 10-12 years, when the patient himself can more consciously approach the choice of method; earlier than this age - for psychosocial indications.

Good functional results in hearing replacement with implantable systems, low complexity and minimal invasiveness of the operation, and significant improvement in the quality of life of patients determine the high frequency of their use. But in a number of cases, reconstructive interventions remain relevant. For example, in patients with chronic and oncological diseases in the head and neck area who require regular MRI examinations, since the implanted elements produce a significant artifact during the examination; for social reasons - in the absence of confidence in the possibility of regular hygiene measures and provision of batteries for the sound processor, as well as reluctance to use a hearing aid.

In our country, an economic issue is relevant: the cost of a bone conduction system and its maintenance exceeds the cost of performing reconstructive surgery followed by wearing a conventional hearing aid.

One of the most important omissions remains the lack of support from the state in creating centralized information to specialists and patients about the possibility of rehabilitation of this group of patients, as well as standards of medical care. Various thematic groups on social networks and materials on the websites of medical companies are not able to give a complete picture of all the possibilities and lack an integrated approach and step-by-step implementation of the necessary actions.

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