CONGENITAL STENOSIS OF THE EXTERNAL AUDITORY CANE WITH CHOLESTEATOMA IN CHILDREN

Ibragimov Jasur Nodirovich
Organisation: National children's medical center
Gulyamov Sherzod Bakhramdjanovich
Scientific supervisor: Doctor of Medical Sciences
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Abstract. Congenital stenosis of the external auditory canal in children is a rare but clinically significant pathology, often combined with microtia and middle ear anomalies. One of the most serious complications of this anomaly is the development of cholesteatoma, leading to destruction of the temporal bone and persistent hearing loss. The aim of this study is to analyze modern surgical approaches to the treatment of children with this pathology, including an assessment of indications for surgery, diagnostic criteria and postoperative results.

According to the analysis, hearing improvement after reconstructive surgeries is achieved in 50-75% of cases, but the frequency of cholesteatoma recurrence and the need for revisions remain high, which emphasizes the importance of long-term follow-up. An individualized approach to the choice of treatment and rehabilitation tactics remains a key factor in the successful management of such patients.

Key words: congenital stenosis, external auditory canal, cholesteatoma, children, atresioplasty, canaloplasty, Jahrsdoerfer score, surgical treatment.

Relevance. Congenital external auditory canal stenosis (CEAS) is a rare developmental anomaly of the outer ear, often combined with microtia and middle ear anomalies. An extremely important clinical problem in such patients is the risk of cholesteatoma formation, which can lead to destruction of the temporal bone, persistent conductive hearing loss and intracranial complications. The incidence of congenital atresia/stenosis of the external auditory canal is estimated at approximately 1:10,000-20,000 newborns [1].

Purpose of the study. To analyze modern surgical approaches to the treatment of congenital stenosis of the external auditory canal with concomitant cholesteatoma in children, to evaluate indications for surgery, diagnostic criteria and clinical outcomes (improvement of hearing, frequency of relapses and complications).

Materials and methods. A literature review and analysis of clinical protocols of leading otolaryngology centers were conducted. The main reference points were data on the incidence of cholesteatoma in external ear anomalies, the results of surgical treatment (canaloplasty/ateresioplasty and cholesteatoma sanitation), and recommendations for the use of the Jahrsdoerfer scale to determine indications for reconstructive surgeries. The recommended Jahrsdoerfer threshold for a good surgical prognosis is ≥7 points; at ≤5 points, surgery is usually not indicated due to the low probability of hearing restoration [2,3].

Indications for surgical intervention:

1. The presence of clinically significant conductive hearing loss that interferes with speech development and auditory rehabilitation.

2. The presence or suspicion of canal/retro-canal cholesteatoma (clinical, CT/MRI) is an indication for sanitation to prevent destruction [4,5].

Surgical methods:

1. Radical/sanitary surgery for cholesteatoma (canalectomy, mastoidectomy if the mastoid process is involved) with removal of epidermoid masses.

2. Atresiaplastika / Canaloplasty – restoration of patency of the external auditory canal and, if possible, reconstruction of the ossicular chain (canal-tympanoplasty). Results vary: according to a number of studies, successful hearing restoration (closure of the air-bone gap ≤25 dB) is achieved in 54-75% of cases, while the frequency of hearing improvement is noted in 69-75% of studies; however, the success rate depends on the initial anatomy, the degree of middle ear hypoplasia and the Jahrsdoerfer score [6,7].

Results and discussion. The incidence of cholesteatoma in congenital atresia/stenosis varies in the literature: from less than 5% to 8% and higher in specially selected series (in some series in combination with microtia - up to 43%). This emphasizes the need for screening and early imaging (CT of the temporal bones) in children with congenital anomalies of the outer ear [4,5].

After sanitation of cholesteatoma, long-term dynamics are mandatory: in a number of pediatric series, the frequency of repeated interventions/revisions exceeded 30-50% (depending on the stage and volume of the lesion), which reflects the growing need for follow-up and possible reoperations [8,9]. Compared with prosthetic or bone anchored hearing device (BAHD), reconstructive surgeries on the external auditory canal provide cosmetic and functional effects, but in the long term, the effectiveness of BAHD in a number of studies exceeds the results of reconstruction in terms of hearing indicators and complication rates. The decision on the rehabilitation strategy should be individualized [10,11].

Postoperative management:

– Regular visits every 3-6 months in the first year, then annually; periodic otoscopy, audiometry and control CT as indicated.

– Early detection of recurrent cholesteatoma requires revision surgery. The high rate of re-interventions in pediatric series requires informing parents about the need for long-term follow-up [8,4].

Conclusion. 1. Congenital stenosis of the external auditory canal with cholesteatoma in children is a rare but clinically significant condition that requires a multidisciplinary approach.

2. Surgical treatment is primarily aimed at sanitation of cholesteatoma and prevention of complications; reconstructive surgeries (aterosioplasty/canaloplasty) can be performed with favorable anatomy (Jahrsdoerfer ≥7) with a probability of hearing improvement in 50-75% of cases [2,6].

3. Despite satisfactory short-term results, a significant percentage of patients require repeated interventions; therefore, long-term dynamics and individual choice of the method of auditory rehabilitation (reconstruction vs BAHD) are indicated [4,10].

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