Evaluation and Management of Aural Atresia; Review Article

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

Review Article

ABSTRACT

The absence of a patent ear canal, which can be acquired or congenital, is known as Aural Atresia. The most common cause of acquired Aural Atresia is an inflammatory response to trauma or otologic surgery. Although rare, acquired Aural Atresia is most commonly found after external ear trauma, such as car accidents, gunshot wounds, or recent otologic surgery. The main treatment of Aural Atresia is surgical, also the invention of bone anchored hearing devices provide greater alternative solution, each methods has its advantages of disadvantages, we hope In the future of developing more effective treatment options.

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1. INTRODUCTION

The absence of a patent ear canal, which can be acquired or congenital, is known as Aural Atresia. The most common cause of acquired Aural Atresia is an inflammatory response to trauma or otologic surgery. Congenital Aural Atresia is an abnormality of the external auditory canal in newborns that causes significant conductive hearing loss. The inner, middle, and outer ear are the three sections of the human ear. The auricle and external auditory canal make up the outer ear, commonly known as the external ear. By guiding sound waves to the tympanic membrane and middle ear, the external ear is essential for hearing [1].

CAS is characterised by Cole and Jahrsdoerfer as an EAC with a diameter of 4 mm or less that frequently occurs in association with grade 1 and grade 2 microtia [2]. Congenital Aural Atresia (CAA) is a congenital ear deformity that affects the development of the external ear canal (EAC) to varying degrees. One in 10,000 to 20,000 babies is affected. Male dominance and right ear dominance are both visible; CAA is usually unilateral [3].

Conduction hearing loss, induced by ear canal blockage and ossicular chain defects, is the most common symptom of CAA. The CAA is one of the most difficult surgeries to do, and surgical outcomes are unpredictable. In 1883, Kiesselbach attempted the first surgical correction, which resulted in facial-palsy. Jahrsdoerfer described important aspects of a new surgical approach in the 1970s and claimed positive results. The International Microtia and Atresia Workgroup (IMAW) issued international recommendations for functional ear reconstruction in patients with microtia and CAA in a multidisciplinary team environment that agreed on the intended treatment objectives in 2019 [3-6].

The number of patients with CAS is low, and our understanding of the condition is still centered on Cole and Jahrsdoerfer [7]. Patient age and stenosis size are two essential factors to consider while dealing with CAS1. Patients with a stenosis of 2 mm or less are at a high risk of developing cholesteatoma and should be operated on [8]. There have been no large-scale investigations of the clinical aspects and long-term consequences of CAS to yet, and no research have detailed the critical parameter of determining the diameter of the EAC [2,8-14].

2. EPIDEMIOLOGY

Congenital Aural Atresia affects one out of every 10,000 to 20,000 babies born. In the majority of cases, it is unilateral, with a strong preference for the right ear, and males are 2.5 times more likely than females to be affected. It's frequently associated with variable degrees of microtia, which could be related to the severity of the underlying middle ear malformation. Children with unilateral Aural Atresia usually have normal speech development and hearing in the unaffected ear [1,15-17].

The canal status, hearing, and postoperative success of 24 patients with acquired atresia of the external auditory canal were studied retrospectively: Acquired stenosis is more frequent in men, with a male-to-female ratio of 2-3:1, and it appears to be a condition that affects young adults. The main etiological reasons of acquired ear canal stenosis were previous ear surgery (54.2%) and external ear trauma (45.8%). The majority of these etiological factors include mastoidectomy and traffic accidents [18].

Patients with CAS who did not have cholesteatoma often had meatoplasty at the age of six, when they were mature enough to grasp the reasoning behind the treatment and help with postoperative care. Except for patients under the age of six years, age was not an exclusion factor in CAS patients with cholesteatoma, and there was no significant difference in cholesteatoma production between age groups [2].

3. ETIOLOGY

Although rare, acquired Aural Atresia is most commonly found after external ear trauma, such as car accidents, gunshot wounds, or recent otologic surgery. Canal stenosis and atresia have been reported in the context of neoplastic alterations and idiopathic inflammatory processes in rare cases. As a result, acquired auditory atresia is caused by a post-inflammatory state [1].

Congenital Aural Atresia's specific cause is unknown, however it's likely complex. It is caused by a disruption of normal embryological development that begins as early as the sixth week of pregnancy. The development of the first pharyngeal cleft, which is important for the development of the external auditory canal, is hampered by this disruption. This disruption happens most of the time at random, but it has

been linked to a number of syndromes, including Goldenhar, Treacher Collins, and Crouzon [1].

4. DEVELOPMENT, ANATOMY AND CLASSIFICATION

Bone and cartilage make up the external ear. The bony EAC is located within the temporal bone and houses vital structures like the face nerve and carotid artery. Beginning in week 4 of pregnancy, the ossicles begin to differentiate. By week 8, the previously united mass has separated into the malleus and incus. The first branchial arch produces the Meckel cartilage, the head and neck of the malleus, and the body and short process of the incus. The Reichert cartilage, malleus manubrium, long process of the incus, and stapes superstructure are all formed by the second branchial arch by week 16 of pregnancy. The external auditory canal is formed at birth by the medial bony tympanic ring and the lateral membrane cartilaginous area. Until the child is 4 to 5 years old, the bony tympanic ring expands into a cylinder (lengthening). The development of the middle and external ear shares a common notion with CAA classification. Stenosis (type A), partial atresia (type B), and total atresia (type C) are the three types of CAA [3].

The numerous classification schemes offered for CAA are based on a physical examination, imaging, or surgical repair findings. In 1955, Altmann established a method of grading based on the degree of the anomaly. There is some section of the external auditory canal (EAC) present in Group 1 (mild). There is no EAC and a constricted middle ear in Group 2 (moderate), and there is no EAC and a severely hypoplastic or missing middle ear space in Group 3 (severe). The purpose of more recent categorization methods has been to delineate which patients are the best candidates for surgical repair using high resolution computed tomography (HRCT) imaging findings. Jahrsdoerfer's grading system has become a popular tool for clinicians for deciding surgical candidacy [19-22].

5. EVALUATION

Within the first few months of life, every child with an ear malformation should receive a full hearing evaluation. Because both conductive and sensorineural hearing loss can occur in either ear, auditory brainstem testing is usually the initial step in the evaluation of individuals with unilateral or bilateral Aural Atresia. Although conductive hearing loss accounts for up to 90% of hearing loss, up to 15% of individuals may have sensorineural hearing loss, which necessitates further investigation [1].

Hearing must be checked to rule out middle and inner ear problems. Objective hearing tests (e.g., auditory brainstem response or auditory steady state response test) should be conducted as soon as possible in patients with bilateral abnormalities. The prognosis of CAA repair is influenced by the preoperative hearing threshold. Preoperative computed tomography (CT) is required to detect the facial nerve, ossicles, otic capsule, degree of pneumatization, and key arteries, as well as to evaluate the Jahrsdoerfer grading system's middle and inner ear components. A score of more than 7/10 indicates that the surgery will be successful [3].

Jahrsdoerfer grading system:

- Stapes bone (2)
- Oval window open (1)
- Middle ear space (1)
- Facial nerve (1)
- Malleus-incus complex (1)
- Mastoid pneumatization (1)
- Incus-stapes connection (1)
- Round window (1)
- External ear (10
- Total possible score (10)

A history, physical examination, family history, complete audiometry, and HRCT imaging are all used to evaluate a patient with CAA. The CT scan can be used to assess the exterior, middle, and inner ears, as well as check for cholesteatoma. To determine air and bone conduction thresholds, pure tone and speech audiometry should be used. The sensorineural acuity level (SAL) test should be used to determine correct bone conduction thresholds in each ear to account for the masking challenge in patients with bilateral Aural Atresia. Electrophysiologic examination of cochlear function is critical when a kid comes as a newborn, especially in patients with binAural Atresia. The kid is a candidate for hearing rehabilitation if the cochlea appears to be working normally, as determined by auditory brainstem response testing (ABR) or bone conduction thresholds. Children with bilateral auditory atresia should be placed with bone conducting hearing devices as soon as feasible to allow for speech and language development during their formative years [19].

6. TREATMENT

Surgical correction is the gold standard treatment for acquired Aural Atresia. However, management of persistent inflammation in the early stages is controversial, as some otologists urge antibiotic/anti-inflammatory medication while others contend that delaying surgical intervention leads to medial canal fibrosis advancement [1].

6.1 Surgical Treatment Methods

Cremers, et al. [23] categorized the surgical procedures used to treat CAA and conductive hearing loss during the 1970s into three categories. Because of the significant risk of consequences, including sensorineural hearing loss, fenestration of the lateral semicircular canal for stapedial fixation has been abandoned. The freshly formed tympanic membrane is connected to the head of the stapes with type 3 tympanoplasty. Many surgeons now prefer canalplasty because of the little disruption to neighbouring structures and the simplicity with which it may be combined with auricular restoration [3].

Due to concerns about the use of the cartilaginous rib graft in the setting of microtia-atresia, auricular reconstruction should come before atresia correction. Rib cartilage does not usually become solid enough for grafting until the age of 5 or 6. Furthermore, by the age of 5 to 6, the patient's ears normally reach 85 percent maturity. Patients benefit from bone conducting hearing aids prior to surgical treatment. For a variety of reasons, the optimal age for surgery in bilateral Aural Atresia is around 6 years of age. One rationale is to allow the eustachian tube to mature and the temporal bone to pneumatize, preventing middle ear issues. Another reason is that patients now understand and comply with postoperative instructions. The development of an external canal cholesteatoma, a known complication that can result in irreversible damage to the middle ear, is an exception to this age [2,24,25,7].

Surgical technique for CAS is modified meatoplasty with endoaural-conchal incision. With long-term follow-up, a study found a steady hearing outcome. Longer follow-up in CAA cases, on the other hand, was associated with a decline in hearing outcomes. This disparity could be due to the postoperative state of the EAC, TM, and ossicles. Another intriguing discovery was that following surgery, BC hearing thresholds increased by 5 to 6 dB at 0.5, 1 and 2 kHz [2].

Meatoplasty is a successful surgical treatment for CAS, and long-term follow-up revealed a stable hearing outcome. The Jahrsdoerfer score had an effect on postoperative ABG, while age had no bearing on surgical indication [2].

7. METHODS COMPARISON

In a Study: 24 patients with acquired atresia of the external auditory canal were retrospectively evaluated regarding their canal status, hearing, and postoperative success. During the procedure, endaural incision is made in 79.2 percent of cases and postauricular incision is performed in 20.8 percent of cases. Transcanal (70.8 percent), transmastoid (20.8 percent), and combination (8.4 percent) surgical procedures are the most common. In most cases, the atretic plate is seen at the bony-cartilaginous junction (37.5%) and in the cartilaginous canal (33.3%); the bony canal is only involved in a few cases. The ear canal is lined with preserved healthy canal skin, split- or full-thickness skin grafts, or pre- or postauricular skin flaps, but retained healthy canal skin is recommended [18].

6.9-46 percent of patients who received atresiaplasty to reduce the airbone gap and improve hearing required ossiculoplasty to repair ossicular chain defects (e.g., incudostapedial joint discontinuity, poor lateral chain and/or stapes fixation, and lack of the incus). The sort of ossicular repair utilised may have an impact on the final hearing result. Total ossicular replacement prosthesis placement was linked to poorer outcomes than partial ossicular replacement prosthesis insertion. After partial ossicular replacement prosthesis insertion, the audiological result was comparable to that after intact native chain reconstruction [3,26,27,28].

In another study that was done Between 2007 and 2012, 75 CAA patients and 50 CAS patients had congenital meatoplasty with canalplasty and tympanoplasty. Results: Paired comparison studies found no significant differences in preoperative ABG between CAA and CAS, but significant differences in postoperative ABG, ABG, the number of ABG 30 dB, and ABG 10 dB. In 61.3 percent of CAA patients and 20% of CAS patients, complications such as postoperative stenosis, bone regrowth, external aural canal (EAC) infection, EAC eczema, total deafness, and lateralization of the tympanic

membrane (TM) were seen. Patients with CAA and CAS had significantly different complications [2].

Furthermore, owing of the increased incidence of tympanic membrane lateralization during healing, Ahn, et al. discovered that longer prostheses were related with worse hearing results. Because hearing loss is common during combined ossiculoplasty/atresiaplasty, the neo-annulus should be implanted as far medially as possible and the prosthesis length should be kept as short as possible. Overall short- and long-term hearing improvements of 30.5 and 22.2 dB were found in a retrospective study of 283 atretic ears treated with the anterior technique. Others have observed similar improvements (about 25 dB). The hearing improvement was 23.35 dB when a transmastoid technique was employed to treat 33 ears, which is identical to the data above. Hearing, sound localization, and hearing in noisy environments all improved [5,29,30,31].

In people with CAS, eatoplasty combined with canalplasty and tympanoplasty can produce dependable and long-lasting favourable hearing results with a low risk of serious consequences. The presence and preoperative state of patients' TM and EAC skin aided in improving hearing outcomes and lowering the risk of complications. The ultimate hearing outcomes and consequences, on the other hand, necessitated stronger indications for CAA patients [2].

8. BONE ANCHORED HEARING DEVICES

A bone conducting hearing aid can be worn around the head on a hard metal band or a soft band, or a bone anchored hearing device can be implanted in the calvarium, in addition to atresia correction. These osseo integrated titanium implants were initially launched in the 1980s and use a percutaneous abutment to connect the implant to a hearing aid (bone oscillator). The BAHA® (Cochlear Americas, Centennial, CO) and the Ponto® (Cochlear Americas, Centennial, CO) are two commercially marketed bone anchored hearing aid systems (Oticon Medical Corp., Somerset, NJ). In the United States, these devices are FDA-approved for children aged 5 and up. A BAHA® or Ponto® softband, a bone conducting hearing aid worn on a band that is not attached to an osseo integrated implant, may be utilized in younger children [19].

9. CONCLUSION

The most common cause of acquired Aural Atresia is an inflammatory response to trauma or otologic surgery. Congenital Aural Atresia is an abnormality of the external auditory canal in newborns that causes significant conductive hearing loss. The cognital cause of the disease is mainly genetic, evaluation of the patient must be done carefully befor the operation, currently Jahrsdoerfer grading system, is the most widely used grading system. While the main treatment of Aural Atresia is surgical, the invention of bone anchored hearing devices provide greater alternative solution, each methods has its advantages of disadvantages, we hope In the future of developing more effective treatment options.

CONSENT

It is not applicable.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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