Hearing stimulation of the pediatric patient with congenital aural atresia: Surgical and audiological evaluation of 38 patients

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The aim of this work is to stress the importance of and discuss the timing and options for the treatment of congenital aural atresia (CAA), including non-surgical alternative treatment modalities and amplification, and to report the audiological and surgical results of a series of patients. Thirty-eight children with CAA were evaluated with regard to hearing and anatomical anomalies accompanying CAA: the state of the ossicles and the facial nerve, postoperative complications and audiological results. The ages of the patients ranged between 4 and 18 years, with a mean of 10 years. All underwent surgical treatment; 32 had unilateral atresia, while 6 had bilateral atresia. The mean follow-up duration was 7 months. The facial canal was dehiscent in 36.8% of cases. In 70.2% cases, the malleus and incus were present as an ossicular mass, fixed and attached to the atretic bone. The stapes was normal in 97.3% of the patients; in 2.7% the suprastructure was deformed. The success rate, defined as an air-bone gap of 20 dB or less, was 63.1% in this series of patients.

If atresia is bilateral, very early hearing stimulation to prevent the maldevelopment of children's speech and cognitive skills is of the utmost importance. In unilateral cases, surgery may be postponed until early adulthood, when the patient is able to make his/her own decision and cooperate in the treatment and postoperative aspects.

Key words: congenital aural atresia, air-bone gap, bone mass, hearing aids.

Congenital aural atresia (CAA) is characterized by aplasia or hypoplasia of the external auditory canal (EAC), often associated with abnormalities of the auricle (microtia), the middle ear and, occasionally, the inner ear structures[1]. Management of CAA must begin with early identification and discussion of the developmental, surgical and educational issues specific to each child. A team approach to management is essential. In unilateral atresia with a contralateral normal-hearing ear, surgery may be postponed until the patient is mature enough to decide for him/herself. Depending on the communication skills achieved with bone-conduction hearing aids (BCHA) and

osseointegrated hearing aids, the child with bilateral atresia may need special education from an early age to maximize speech and language development[2]. Surgery for bilateral CAA is generally postponed until 5 to 7 years of age, for the better cooperation of the patients in audiometric evaluation as well as in postoperative care, and for completion of mastoid pneumatization[3].

The main purpose of CAA surgery is to provide patients serviceable hearing and a new external auditory canal[4]. The techniques of canalplasty, meatoplasty, tympanoplasty and ossiculoplasty have improved considerably, but surgical correction of congenital aural atresia is still one

of the most difficult and challenging procedures performed on the ear[1,4]. The complexity of this surgery requires a good knowledge of the surgical anatomy of the facial nerve, oval window and inner ear as well as their congenital variations[3]. Because of this, the management team should consider all treatment modalities, which include both surgical and nonsurgical alternatives. The aim of this work is to report the efficacy of EAC atresia reconstruction in children in terms of surgical and audiological results, and to provide information about nonsurgical alternative treatment modalities and amplification to health care professionals who deal with children with CAA.

Material and Methods

Thirty-eight children who were operated in the Hacettepe University Department of Otolaryngology due to CAA between 1995 and 2012 were included in the study, which was approved by the Hacettepe University Ethics Committee. The patients' medical records were evaluated for age, sex, ENT examination results (total Jahrsdoerfer score, and the specific point value [0-1-2] assigned to each anatomical structure on the basis of the Jahrsdoerfer grading scale)[5] and surgical and audiological findings. The duration of follow-up ranged from 3 to 9 months, with a mean duration of 7 months. Preoperative computed tomography was conducted for each patient in order to identify the situation of the middle ear cleft, ossicles and facial nerve. All patients were operated via the anterior approach; in all patients, endaural incision and temporalis muscle fascia as a graft material were used. For the lining of the newly formed EAC, a split-thickness skin graft, harvested in most cases from the lateral thigh, was used. Information was obtained from the patients' preoperative records, operative notes, audiograms and postoperative office visits. All patients underwent audiometric testing before surgery and were typically seen approximately 3 months after surgery for a full audiometric follow-up, with many subsequent office visits as well.

Surgical technique

We use endaural incision (Fig.1) and protect the external ear canal skin flap. By extensively drilling out the deformed or absent EAC, we create a new, wide EAC. The tegmen tympani is identified and followed to the epitympanum, where the ossicles are identified. Usually, the malleus and incus are found as a single block. This is removed and the graft is laid on the stapes. Temporalis fascia is used as graft material to create a new tympanic membrane; a split-thickness skin graft and EAC skin are used to completely cover the EAC. After the skin graft is used to line the new ear canal, which is lying over the fascia, antibiotic-soaked gelfoams are used to fill the canal. Skin, subcutaneous tissue and cartilage are removed to create the new, wide meatus. The lateral edge of the skin graft is sutured to the meatal skin, and a wick tamponade is then placed in the outer portion of the EAC.

Data Analysis

In our study, pure-tone air and bone-conduction thresholds were obtained before and after the surgery, with thresholds at 500, 1000, 2000, and 4000 Hz used to calculate the pure-tone averages. Air and bone thresholds from the same test were used to calculate the air-bone gap (ABG). A successful hearing result was defined as a postoperative ABG of 20 dB or less.

Results

There were 18 male and 20 female patients; their ages ranged between 4 and 18 years, with a mean of 10 years. Thirty-two patients had unilateral atresia; 6 had bilateral atresia (Table I). All patients included in the study were scored 6-8 on the basis of their preoperative Jahrsdoerfer grading (5 of the patients had a score of 6, 11 had a score of 7, and 22 had a score of 8). The malleus and incus appeared normal in 11 (29.8%) cases. In the other 27 (70.2%) cases, the malleus and incus were present as an ossicular mass, fixed and attached to the atretic bone (Fig. 2). The stapes was normal in 37 (97.3%) cases; suprastructure deformation was found in 1 (2.7%) case (Table II). In none of the cases was the stapes footplate absent or fixed. In 11 (29.8%) patients, the graft was laid on the malleus, keeping the ossicular chain intact; in 3 cases (7.8%), there was no connection between the incus and the stapes. This connection was reconstructed using bone cement, and the graft was laid on the malleus. In the 27 (70.2%) patients where the malleus-incus block was removed, the graft was laid on the stapes. The facial canal was dehiscent in 14 (36.8%) cases, and the

facial nerve followed an unexpected route in 6 (15.7%) patients (Table III). In preoperative PTAs (pure tone audiograms), the average bone threshold was found to be 32.5 dB and the average air threshold 58.4 dB; the preoperative average ABG was 25.9 dB. The postoperative average bone threshold was found to be 25.7 dB and average air threshold 44.2 dB, with a postoperative average ABG of 18.5 dB (Table IV). According to the postoperative PTAs, 24 (63.1%) patients' ABGs were 20 dB or less. Pre- and postoperative audiological findings according to patients' Jahrsdoerfer scores are summarized in Table V.

There were 17 (44.7%) cases of restenosis. Fourteen of these patients had granular tissue in the EAC; this tissue was removed through revision surgery. Three of them had bone collapse of the EAC, which was removed by drilling to create a new and wider EAC. Three (7.8%) patients had temporary facial nerve paralysis, and in one patient cholesteatoma formation in the middle ear was seen as a complication. Graft perforation was observed in the postoperative follow-up of 2 (5.2%) patients.

Discussion

The incidence of congenital aural atresia ranges from 1/10,000 to 1/15,000 and can be bilateral in up to 33% of cases. A multidisciplinary approach is mandatory for successful management of these cases. The overall goals are to achieve a cosmetically acceptable external ear and to create a functional pathway for sound from the external ear to the cochlea while preserving facial nerve and labyrinth function[5]. Medical care should be focused on language development. Understanding the impact of hearing loss caused by the atretic ear canal is important. Consideration of the educational impact of hearing impairment in the treatment plan for each child is mandatory. The importance of restoring early hearing in these patients cannot be underestimated. Once the indication arises regarding the potential for hearing loss, early hearing testing and subsequent early placement of hearing aids greatly facilitate language outcomes. Some centers perform ABR testing as soon as feasible and attempt to place hearing aids by the age of 6 months at the latest[6]. Bilateral atresia usually requires surgery to restore hearing.

Before surgery, and as early as possible, patients should be fitted with appropriate amplification, such as bone-conduction hearing aids. Surgical intervention in unilateral or bilateral EAC cases, followed with appropriate hearing stimulation, is generally performed around the age of 5-7 years. The actual anatomical configurations of the external auditory canal have been demonstrated to affect the gain distribution and contribution to audition[6]. Not all patients with unilateral atresia require surgery. Hearing, learning and social issues must be considered for audiologic and surgical decision making.

Difficulties of Surgery

Usually an endaural incision (Fig.1) modified to form an S-shape or to create a superiorly based skin flap is chosen. To determine the location of the incision, the root of the zygomatic bone, the mandibular condyle, the anterior border of the mastoid and the mastoid tip are palpated and identified[7]. The EAC should be constructed on the same plane as the contralateral EAC. In the anterior approach, drilling of the EAC is done between the temporal line and the temporomandibular joint[8]. The atresia plate is drilled with a diamond burr or with a curette until the ossicles are exposed[8-10]. After removal of the atresia plate, the bone around the ossicles is drilled to enlarge the entrance to
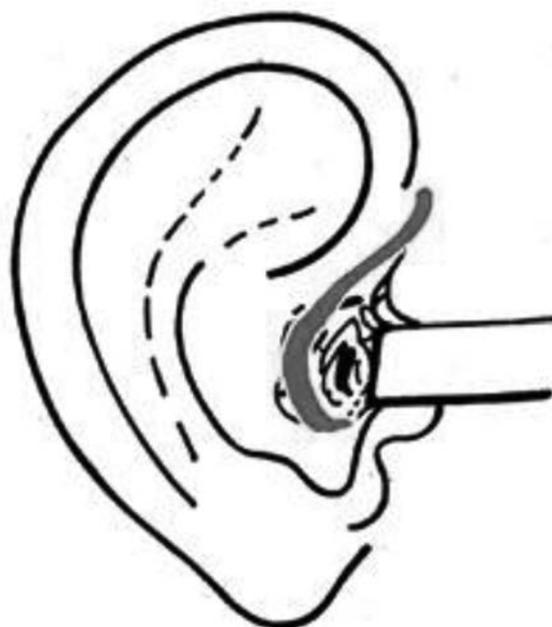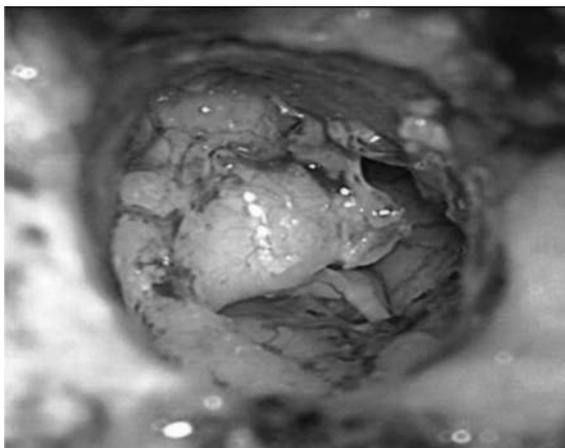

Fig. 1. Endaural incision

Fig. 2. Perioperative malleus-incus mass

the middle ear cleft. If the ossicles are mobile and functional, they should be preserved. If there is fusion and a nonfunctioning malleus and incus, they should be removed[11]. In the study group, the most common ossicular finding was fusion of the malleus and incus with a normal stapes; 70.2 % of patients had a malleus-incus block, which we removed (Fig 2.). The graft was laid on the mobile stapes in all of the patients in our series.

To determine the ossicular status, the tympanic cavity should be widely exposed to view the oval and round windows; for this, the inferior part of the atresia plate is removed with a curette.

It should always be kept in mind that the facial nerve may follow an unexpected route. The degree of microtia is parallel to the anomalous route of facial nerve[12]. In our group, the facial canal was dehiscent in 14 (36.8%) cases, and the facial nerve followed an unexpected route in 6 (15.7%) cases. In severe anomalies, the vertical segment of facial nerve is located anteriorly. If the middle ear cleft is narrow, the chance of damaging the facial nerve is higher[13]. The tympanic nerve on the promontorium is located vertically, and when followed superiorly points to the cochleoriform process, which is located just anterior to the facial canal[14].

The air-bone gap cannot be closed totally in all aural atresias, and there is also the chance of sensorineural hearing loss. De La Cruz et al[15]. defined a successful hearing result as a postoperative ABG of 30 dB or less. They found a success rate (ABG<30 dB) of 57.9% of patients, with a postoperative average ABG

Fig. 3. Bone-anchored hearing aid for CAA

of 18.5 dB[17]. We defined a successful hearing result as a postoperative ABG of 20 dB or less. On this basis, our success rate was 63.1%, with the hearing test taking place three months postoperatively on average. This rate can be expected to deteriorate with a longer follow-up period. Carfrae et al[16] reported that Jahrsdoerfer grading scores of 6 or more are essential for successful resulst after, surgery in patients with CAA. All of the patients in our study had scores from 6 to 8 on the Jahrsdoerfer grading scale, and no significant difference was observed between the various scores in terms of audiological findings.

A vibrating ear drum and a sufficiently wide EAC should be constructed. If the stapes is fixed, it should not be removed in the first operation. If the malleus and incus are fixed to the atresia plate or to the surrounding structures, they should be removed. In this study, 70.2% of patients had a malleus-incus block; it was removed, and the graft was laid on the mobile stapes. Direct contact with the malleus during the drilling of the atresia plate may cause sensorineural hearing loss. An iatrogenic fistula in the semicircular canals or the stapes footplate will most likely result in total sensorineural hearing loss. No sensorineural hearing loss was observed in our experience. Allografts should be avoided, since there is a high risk of foreign body reaction. The malleus and incus may be replaced on the stapes, or a graft may be laid directly on the stapes, forming a membranostapediopexy. In 3 cases (7.8%) there was no connection between the incus and the stapes. A connection was established using glass ionomer bone cement and was laid on the malleus. A graft was laid on

the stapes, forming a membranostapediopexy, in 27 patients. An annulus should be formed to insert the graft underneath to avoid lateralization; for the same reason, the skin graft should cover the fascia graft[17]. Restenosis of the new ear canal and cheloids at the meatus affect the hearing result. We found that 44.7% of patients developed restenosis. Fourteen of them had granular tissue at the EAC; this tissue was removed through revision surgery. This was related to the incision technique, but 3 of the affected patients had bone collapse or regrowth of the EAC, which was removed by drilling to create a new and wider EAC. This issue may be related to genetic factors. Use of a split- rather than a full-thickness graft assists in epithelization and decreases restenosis[18].

Because of the high restenosis rate, osseointegrated hearing aids might be considered as a first line of treatment. Therefore, families should be consulted prior to atresia surgery and given the right to choose between different treatment modalities. As mentioned above, congenital aural atresia poses significant challenges to surgical remediation. Both bone-anchored hearing aids (BAHA) and, recently, the Vibrant Soundbridge (VSB) have been considered as alternatives or adjuncts to conventional atresiaplasty. As traditional atresiaplasty and BAHA may not be fully satisfying solutions. VSB implantation offers another suitable alternative in CAA patients, can be performed with preservation of cochlear and facial nerve function, and has stable results in the near term[19]. The BAHA surgical implantation procedure is simple, but requires compliance with certain basic principles to minimize the postoperative complication rate (Fig. 3). Many surgical variants concerning the skin incision have been described, all designed to reduce these complications. BAHAs clearly provide better hearing comfort for patients with conductive hearing loss after failure of conventional hearing aids or in patients unsuitable for rehabilitation surgery. However, skin complications frequently occur around the implant abutment and are a source of discomfort and inconvenience for patients (repeated visits, surgical revision, delayed installation of the processor). Despite good results on osseointegration and limited skin reaction with percutaneous bone conductors, there remains room for improvement. Especially

in children, adverse events with percutaneous bone conductors might occur more frequently. Transcutaneous bone conductors, if powerful enough, can provide a solution that minimizes adverse events and implant loss. In children with large bilateral conductive hearing losses, bone conductors are the first and, customarily, even the sole treatment option. The benefits of bilateral application have demonstrated its merits, particularly in cases of bilateral conductive hearing loss, but its cost is quite high. The application of bone conductors in children from as young as 3 months is possible by connecting the hearing device to an elastic softband. From the age of 4 years, the cranium is thick and mature enough to allow safe titanium implantation for percutaneous devices[19].

Conclusion

Hearing stimulation of a child with EAC atresia is very important. Early amplification of the child with bilateral atresia via hearing aids such as bone-conducted or bone-anchored hearing aids should be performed immediately. Surgical reconstruction of EAC atresia should not be planned until 5 to 7 years of age, for better cooperation of patients with audiometric evaluation and postoperative care, as well as completion of mastoid pneumatization. Unilateral atresia surgery may be postponed until adulthood, when the patient is able to make his/her own decision. The aim of surgery should be to construct a vibrating ear drum and a sufficiently wide EAC. To avoid complications, surgery for aural atresia should be performed by experienced surgeons. If anomalies are always kept in mind, a functional result with fewer complications may be attained.

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