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Surgery or implantable hearing devices in children with congenital aural atresia: 25 years of our experience

Andrea Jovankovičová [a,*], Roman Staník [b], Samuel Kunzo [a], Lucia Majáková [a], Milan Profant [c]

[a] Pediatric ENT Department, Children's Univ. Hospital, Limbova 1, 833 40 Bratislava, Slovak Republic
[b] Pediatric ENT Department, Children's Univ. Hospital, Limbova 1, 833 40 Bratislava, Slovak Republic
[c] Department of ORL HNS, University Hospital Bratislava, Antolská 11, 851 07 Bratislava, Slovak Republic

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ABSTRACT

Objectives: Congenital aural atresia and ear deformities have been the subject of serious discussions for centuries. These malformations are associated with significant aesthetic and functional problems. Outcome of the surgical solution is rarely optimal. Despite the gradual improvement of surgical techniques the surgery still remains associated with very limited short-term and mainly long-term functional outcome. Therefore, the priority treatment in modern otology becomes implantable devices—BAHA, Bonebridge and active middle ear implants.

Methods: The functional and aesthetic outcomes of aural atresia reconstruction performed at Pediatric ENT Department of Children's University Hospital were retrospectively evaluated and compared with the results prospectively obtained from implantable hearing devices (BAHA, Vibrant Soundbridge, Bonebridge), which have been implanted in patients with aural atresia at Department of ORL HNS, University Hospital Bratislava.

Results: Aural atresia reconstruction has been performed in 34 patients during last 25 years. Results of the surgery could be viewed as excellent only in three patients (gain above 30 dB). Air conduction threshold has decreased after the surgery in seven patients, and in two cases total deafness occured after the surgery. Patients gain on average 12 dB in auditory threshold after surgery.

Hearing devices were implanted to the group of 11 children in order to improve their hearing. All of them were the patients with bilateral aural atresia. After implantation a significant improvement in hearing threshold occurred in all children (30–35 dB on average). Together with results of air conduction threshold in patient with aural atresia before and after surgery and implantation we also present a standard deviation.

Conclusion: The functional outcome of implantable hearing devices in patients with bilateral aural atresia clearly dominates over the traditional reconstructive surgery. Aesthetic results in pinna deformity management remain a major concern for patients and parents. Implantable epithesis bring promising results. Since there is no universal solution to this disorder, the final selection of the treatment is upon the patient. Patients should opt for the most suitable solution through consultation with the surgeon, after clarifying the advantages and disadvantages of each option.

1. Introduction

Congenital aural atresia is a congenital defect characterized by hypoplasia or aplasia of the external auditory canal, often associated with microtia and middle ear anomalies and occasionally with the inner ear anomalies [1]. Incidence of this malformation is 1 in 10 000 live births. It is mostly associated with a variety of syndromes and disorders (Treacher-Collins, Goldenhar, hemifacial microsomia) [1,2].

Congenital aural atresia affects children in two specific ways. The first is severe conductive hearing loss, which if not corrected, may cause speech development delay. The second is the pinna malformation resulting in feelings of inferiority and problems of integration in the social environment.

* Corresponding author. Tel.: +421 2 59371422.
E-mail address: jovankovicova@gmail.com (A. Jovankovičová).

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Pure-tone audiometry and HRCT of temporal bones are crucial for diagnosis and selection of the suitable type of treatment [3,4]. There are several options in the aural atresia management. In the unilateral case with normal contralateral hearing, the treatment can be completely avoided or postponed until adult age respecting personal decision of patient. In case of bilateral aural atresia aided hearing from early age (6 months) is necessary to ensure hearing and speech development. Surgical treatment in bilateral atresia is usually realized at the turn of preschool and school age. The introduction of implantable hearing devices (BAHA, active middle ear implant) in recent years has changed the timing of early surgical intervention. However, there is still an issue with aesthetic part of the malformed external ear which has a significant impact on the psyche of the young patient [1,5].

2. Patients and methods

A total of 94 patients with congenital aural atresia were treated at the Pediatric ENT Department, Children's Univ. Hospital in Bratislava in the period from July 1989 to May 2014. All children patients with aural atresia were referred to the Pediatric ENT Department by their general practitioners or regional ENT specialists in order to solve their anomaly.

ENT examination, hearing tests (pure tone audiometry in complying children, ASSR in young children and non-complying children; speech audiometry was not perfomed) and CT scan of the temporal bones were realized in all patients. The exact treatment of the diagnosis was selected according to the above results. Aural atresia surgery was realized only to the patients who complied with all necessary requirements (conductive hearing loss, proper ear anatomy, age, parents' approval, mental status, comorbidity). Since the year 2007, children with bilateral aural atresia have been referred to the Department of ORL HNS, University Hospital in Bratislava for implantation of hearing device (BAHA, Vibrant Soundbridge, Bonebridge). Aural atresia surgery is therefore no longer realized in those patients. Children with unilateral aural atresia are left without any surgical intervention and make decision about the exact treatment when adult.

Children were divided into three groups:

- Patients that underwent aural atresia surgery until the year 2007 (34 patients: 12 females and 22 males).
- Patients with implantable hearing devices treated since the year 2007 (11 patients: 6 females and 5 males).
- Patients without surgical intervention (49 patients: 21 females and 28 males).

Patients were retrospectively and prospectively evaluated. Functional hearing results of aural atresia management in the first two groups (conventional surgery vs. implantable hearing devices) were evaluated.

2.1. Aural atresia surgery

Transmastoid approach has been done in majority of the patients. It is not problematic and with less risk of n. facialis injury in the cases of a good mastoid pneumatization compared to the anterior approach through the ear canal. Transcanal approach was chosen only in patients with good anatomical conditions or soft atresia (stenosis). In case of auditory ossicles malformation, tympanoplasty was realized.

3. Results

Unilateral aural atresia dominated over bilateral atresia (total of 94 patients; 125 ears with aural atresia; 63 patients with unilateral aural atresia—40 cases with right-sided defect and 23 cases with left-sided one; 31 patients with bilateral aural atresia). In the majority of patients various grades of auricular malformations were present—grade III microtia was most frequent (67 cases), grade II microtia was found in 20 cases, while grade I microtia occurred in 7 cases only. Anotia was observed in 3 patients. Pinna without any deformities was found in 23 ears out of the totaly 125 ears examined.

3.1. Groups of patients

Patients in this study were divided into three groups according to the type of aural atresia treatment.

3.1.1. Group 1: Conventional aural atresia surgery

Aural atresia surgery was done in 34 children (11 with bilateral aural atresia and 23 with unilateral aural atresia, Fig. 1) out of which 4 children underwent complete bilateral aural surgery. CT scan without anomaly of inner ear was a crucial condition in all patients who underwent conventional surgery.

The average age of the patients solved with atresiaplasty was in the case of unilateral atresia 7.3 years, while 4.9 years in the case of bilateral atresia.

Jahrsdoerfer score system was used to indicate the patients for surgery (surgery in unilateral aural atresia $\geq$7 points, bilateral aural atresia 5–6 points). CT scans of temporal bones before the surgery are shown in Table 1.

General clinical condition of the patients was also taken into account (comorbidity, mental status, pure-tone audiometry thresholds).

Average audiometric results in patients with aural atresia surgery are shown in Table 2.

The hearing (air conduction threshold) improved in early postoperative period 10–20 dB in average (mean gain of 12 dB).

Definitive hearing improvement could be seen 4 to 6 months after the surgery, when anatomical conditions in the tympanic cavity stabilize. In this time frame we found deterioration of air-conduction threshold level (5–30 dB, mean 12 dB) in all patients. Permanent improvement of hearing has been evident in 62% of the patients, however in rest of the children the hearing got worse in comparison with pure-tone audiometry before surgery. This decrease was associated with lateralization of new tympanic membrane in 9 cases indicated for revision surgery.

Out of the total 38 operated ears in 31 cases (81.5%) was found a compact bony atresia plate of varying thickness. Eardrum was completely missing in 27 operated ears. In 11 patients eardrum was present under the bone. The most common middle ear

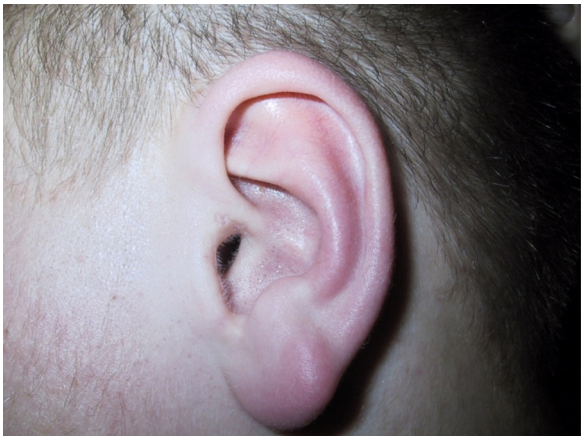

Fig. 1. External auditory meatus in patient 10 years after atresiaplasty for bilateral soft ear canal atresia.