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Congenital aural atresia: Bone-anchored hearing aid vs. external auditory canal reconstruction

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ABSTRACT

Objective: To compare the audiologic outcome and feasibility of bone-anchored hearing aid (BAHA) and external auditory canal reconstruction (EACR) surgeries in pediatric patients presenting a congenital aural atresia (CAA).

Methods: A retrospective chart review of 40 patients operated in our tertiary pediatric care center between 2002 and 2010 was performed. 20 patients underwent EACR, whereas another 20 patients were implanted with a BAHA device. Air conduction (AC), bone conduction (BC), pure tone average (PTA) and speech discrimination score (SDS) were compared preoperatively, and hearing gain (HG) postoperatively at 6 and at 12 months at frequencies of 500, 1000, 2000 and 4000 Hz. Operative time, complications and associated microtia were documented as well. EACR patients were graded retrospectively upon Jahrsdoerfer's classification.

Results: Preoperative AC were significantly different between groups, at 500, 1000 and 2000 Hz but not at 4000 Hz. BAHA group compared postoperatively to EACR group showed significantly a superior HG of 46.9 ± 7.0 dB ($p < 0.001$) and of $39.8(7) \pm 7.2(6.9)$ dB ($p < 0.001$) at 6 months and at 1 year, respectively.

Moreover, aided air thresholds from the EACR group revealed an audiologic status similar to those of the BAHA group patients, at 6 months and one year postoperatively. Both groups had a similar evolution of their BC, as well as of the incidence of complications. We report one case of transient facial paralysis in the EACR group. Total operative time is significantly lower ($p < 0.001$) for a BAHA implantation (56 ± 21 min) than for EACR surgery (216 ± 174 min). No preoperative or postoperative correlation (Pearson correlation test; $p > 0.05$) was found between patient's Jahrsdoerfer's score and their audiologic outcome. HG does not seem to be influenced by the presence of microtia.

Conclusion: EACR, although constituting an attractive option, does not give acceptable results alone. It can however, when combined to conventional air conduction hearing aids, provide excellent audiologic outcomes comparable to BAHA. BAHA implantation is a reliable, safe and efficient therapeutic option that allows a significantly better audiologic outcome when compared to unaided EACR for patients with CAA.

1. Introduction

Congenital aural atresia (CAA) is a rare malformation of the external auditory canal (EAC). It results from an abnormal embryologic development of the first branchial arch. Whereas most of the cases are sporadic, it is sometimes associated with syndromes like Treacher Collins or with chromosomical abnormalities like 18q deletions [1]. There is a frequent association with microtia. This entity is encountered in about 1 in 10,000 births, predominantly in males (2.5:1 male to female ratio), and usually occurs unilaterally [2].

In 1883, Kiesselbach performed the first documented attempt of surgical correction for this anomaly [3]. The surgical reconstruction option is indeed a challenging therapeutic option, considering the lack of landmarks and the altered anatomy of facial nerve and middle ear [4]. Moreover, it carries many risks for complications, including iatrogenic injury to the facial nerve, EAC restenosis and recurrent otitis externa [5]. The development of grading and classification systems for EAC atresia such as Jarhrsdoefer's classification has helped in the preoperative evaluation and outcome expectations [6]. However, the bone conduction concept advanced by Tjellstrom in 1980 brought an interesting alternative to the management of this entity [7]. This led to the bone-anchored hearing aid (BAHA) system that is now

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widely used in developed countries [8]. The BAHA is a surgically implantable system that functions through direct bone conduction rather than via the middle ear.

Multiple studies have assessed long-term audiologic results and complications of each of the treatment options. However, none has clearly compared in one series strictly the audiologic outcome of both strategies nor stated the superiority of one option towards the other. Of note that Evans et al. in 2007 indeed studied both strategies, but mostly comparing the complications and cost-effectiveness of these therapeutic options [5].

Therefore, the purpose of this study is to determine which option would be, for EAC atresia, the treatment of choice regarding the audiologic gain and feasibility. In order to do that, we examined retrospectively our series with both treatment options by comparing and analysing the initial audiologic status of EAC atresia, the postoperative hearing improvement, the complications and the total operating time.

2. Patients and methods

We performed a retrospective review and analysis of the medical records of 40 patients (43 ears) who were treated for their EAC atresia in our pediatric tertiary care center, between 2002 and 2010. These patients underwent either surgical EACR ($N = 20$), or BAHA implantation ($N = 20$).

The decision-making algorithm used in our institution (Fig. 1) was based primarily on the initial degree of the EAC stenosis. If the stenosis was complete, as visualized on the temporal bone computerized tomography (CT) scan (Fig. 2a), BAHA implantation was preferred. However, if the patient presented a partial stenosis (Fig. 2b), we took into account the degree of the ossicular chain malformation. Strong deformation of the ossicles led to BAHA implantation whereas patients with mild malformations and for whom we could not initially offer a conventional hearing aid were considered as a potential candidates for EACR with or without ossiclar chain reconstruction. These patients could benefit from a conventional hearing aid after the EACR. Discussion with the family also oriented the therapeutic choice.

We used 3 mm ($N = 6$) and 4 mm ($N = 14$) auto-tarauding fixtures for the BAHA patients group. As for the EACR group, 10 out of 20 patients needed ossicular reconstruction simultaneously to the EACR procedure. Of note that microtia was present for 14 patients in the BAHA group and for 7 patients in our EACR group. Three patients had a bilateral external auditory canal atresia, two of them underwent bilateral EACR and one had bilateral BAHA implantation. We took in consideration only the first operated ear for those patients.

Moreover, patients from the EACR group were graded retrospectively upon Jahrsdoerfer's classification (Table 1) to assess the correlation between the malformation severity score and their audiologic outcome. Jahrsdoerfer's score, is a potential radiologic indicative factor of the degree of malformation.

2.1. Surgical techniques

Detailed description of the BAHA and EACR surgery can be found elsewhere [9–12]. A single-stage procedure was performed in all our cases of BAHA implantation. A period of 4 months was allowed for osteointegration before BAHA processor fitting. As for reconstructive surgery, the endaural approach was preferred.

2.2. Main outcome measures

Evaluation of the hearing improvement for both treatments was done by comparing the preoperative and the postoperative hearing tests. Hearing thresholds at 500, 1000, 2000 and 4000 Hz frequencies were studied for both groups. Pure tone average (PTA) values was defined as the average of AC thresholds at 500, 1000, 2000 and 4000 Hz. Bone conduction (BC), air conduction (AC), PTA and speech discrimination score (SDS) values were compared. We assessed the hearing improvement of BAHA recipients and EACR patients at 6 months and at one-year follow up. Furthermore, regarding the possibility of EACR patients having had revision surgeries, only the final audiogram results were chosen for comparison. The speech discrimination gain was assessed and compared for each group as well. Total operating time for BAHA and EACR, including revision surgeries, were also gathered.

Postoperative and intraoperative complications were recorded for each treatment. For the EACR group, we documented the presence or absence of the following most commonly encountered complications, as mentioned through literature: infection, tympanic membrane lateralisation, bony or soft tissue meatal or canal restenosis, facial nerve paralysis, sensorineural hearing loss (SNHL), skin graft failure, wound dehiscence (<1 cm), postauricular hematoma, scar contracture and hypertrophy, and temporomandibular joint pain and trauma. Intraoperative complications assessed were: dura mater lesions, facial nerve and tympanic membrane injury. For the BAHA group, we assessed the presence or absence of: local infection or inflammation, failure to osteointegrate, skin overgrowth, fixture loss and flap necrosis as the postoperative complications. Whereas for the intraoperative complications in this group we identified the presence of haemorrhage or cerebrospinal fluid leak through the drilling site.

2.3. Statistical analysis

Variance analysis was conducted, with repeated measures for the intra-subjects frequency factor, as well as for the inter-subjects

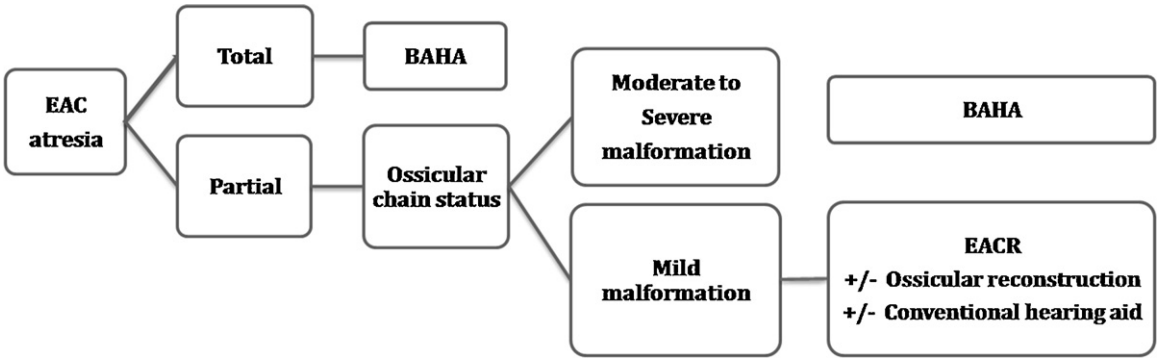

Fig. 1. Decision-making algorithm used in our institution. EACR: external auditory canal reconstruction; BAHA: bone-anchored hearing aid.