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Implantation of the Vibrant Soundbridge in a Case of Bilateral Malformation of the Middle and External Ear

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Data Interpretation D
Manuscript Preparation E
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Conflict of interest: None declared

Patient: Female, 13-year-old
Final Diagnosis: Bilateral congenital external and middle ear malformation
Symptoms: Hearing loss
Medication: —
Clinical Procedure: —
Specialty: Otolaryngology

Objective: Congenital defects/diseases

Background: Here we present a case of Vibrant Soundbridge implantation in a 13-year-old girl with bilateral aural atresia of the external ear canal. In this instance, we attached the device's floating mass transducer (FMT) to a mobilizable complex of the incus and malleus, which functionally connected to the short process of the incus.

Case Report: The article presents a case study of a patient with a congenital defect of the middle and external ear and conductive hearing loss, who was referred for middle ear implantation. Tonal audiometry revealed bilateral moderate to severe hearing loss with a 30 to 50 dB air-bone gap. After making a sufficiently wide antromastoidectomy, it became apparent that implantation of the MedEl Bonebridge hearing aid was not possible because of an overhanging dura. The short process of the incus was then visualized and, by drilling the bone laterally and anteriorly, the incus and malleus were found to have formed a conglomerate, firmly fused to the anterior wall of a rather small tympanic cavity. By removing the bony adhesion, mobility of the ossicular chain was restored. The MedEl Vibrant Soundbridge could then be implanted by attaching its FMT to the incus-like conglomerate.

Conclusions: Restoration of ossicular chain mobility was achieved, and the patient's hearing was improved by implanting the Vibrant Soundbridge hearing aid. Speech audiometry 1 month later showed improved hearing. Implantation of the Vibrant Soundbridge following ossiculoplasty may be a feasible solution in cases of bilateral congenital defect of the middle and external ear.

Keywords: Ear, Middle • Hearing Loss • Tympanoplasty

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/929933

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Background

Over the past 50 years, many publications have focused on reconstruction methods of congenital critical stenosis or atresia of the external ear canal [1-6]. Owing to frequent complications from this type of surgery, the middle ear implant is becoming a valuable option for improving hearing with a variety of devices including the BAHA Connect, BAHA Attract, Ponto, Medtronic Sophono, Bonebridge, and Vibrant Soundbridge [4,5,7]. The Vibrant Soundbridge device was initially intended for patients with sensorineural hypoacusis, but its use was later extended to mixed and conductive hearing loss of various origins [1,4,8]. The typical point of attachment for the floating mass transducer (FMT) is the long crus of the incus or stapes (in cases with preserved anatomy) or the round or oval window if the middle ear has been destroyed [5,9]. A few years ago, another coupler was introduced that could be applied to the short process of the incus, an arrangement which allows the surgeon to simplify and shorten the procedure of implantation by making a posterior tympanotomy redundant, thereby helping to safeguard the facial nerve and chorda tympani [10,11].

Here we present a case of Vibrant Soundbridge implantation in a child with bilateral aural atresia of the external ear canal. In such cases, there is no existing standard treatment. In this instance, we attached the device's FMT to a mobilized conglomerate that then acted on the short process of the incus.

Case Report

The case described is of a 13-year-old girl with congenital malformation syndrome associated with chromosomes X and 18, who was admitted for surgical treatment to the oto-rhino-laryngosurgery clinic for a bilateral congenital defect of the external and middle ear. Otolaryngologic examination revealed a relatively normal auricle, funnel-shaped stenosis of both external ear canals in the cartilaginous portions, and atresia in the bony portions. Pure tone audiometry showed bilateral symmetric mixed hearing loss (**Figure 1**). From the age of 2 years, the child had been provided with bone conduction hearing aids on a headband, on both sides. Because of the discomfort caused by constant pressure, poor sound quality, especially in the high-frequency range, the child did not accept the devices.

A computed tomography scan identified major hypoplasia of the middle ears with bilateral atresia of the external ear canals. Temporal bone assessment revealed poor suitability for a Bonebridge implantation (**Figure 2**). Bilaterally, there were shadows of the ossicular chain indicating conglomeration of the first and second ossicles, which were fused to the anterior wall and the roof of the small tympanic cavity. Both stapes were relatively normal, and the oval and round windows occupied their normal anatomical positions. Owing to serious deformity of the middle ear, it was not feasible to reconstruct the external ear canal.

Based on the results of simulation tests, the patient was referred for middle ear implantation. According to the patient's own reports, it was likely that the left side would benefit most.

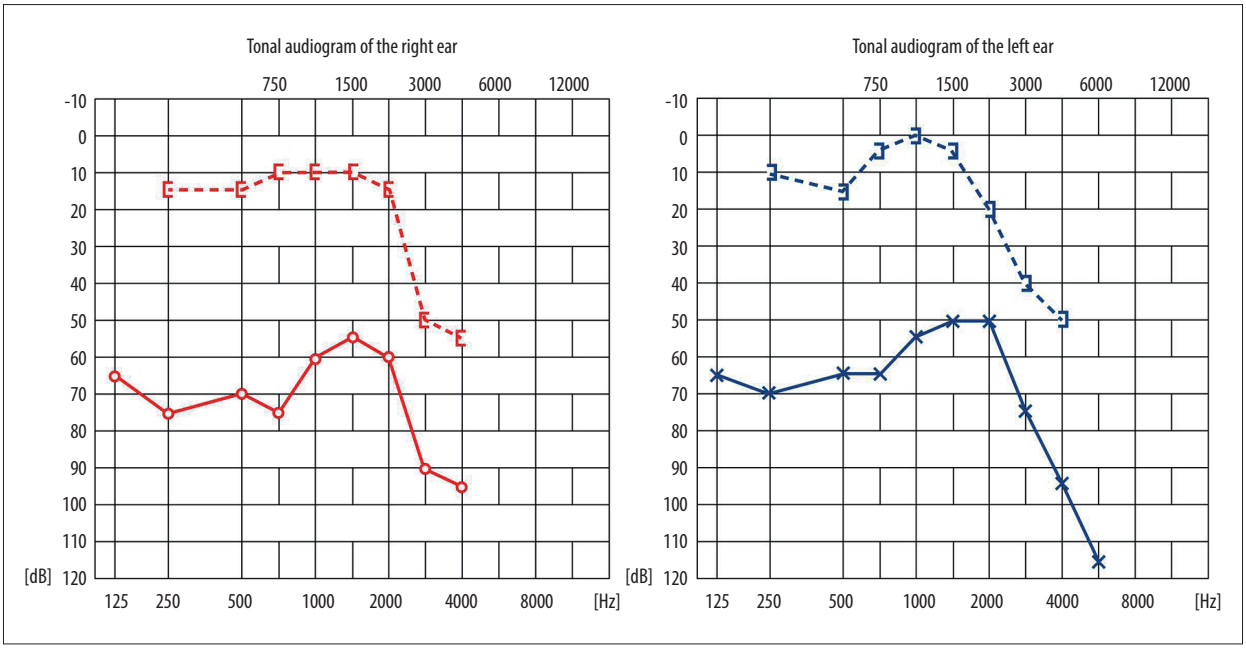

Figure 1. Pure tone audiometry result.

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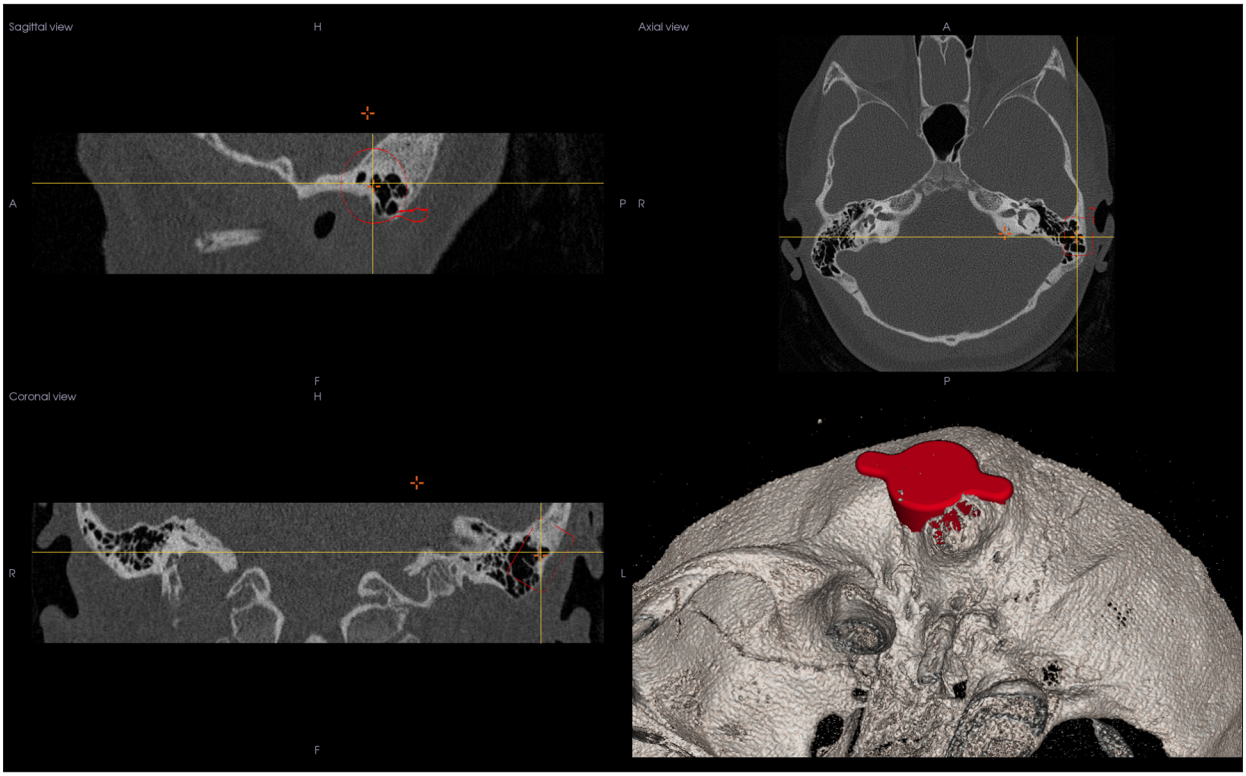

Figure 2. Computed tomography temporal bone assessment.

After performing a limited antromastoidectomy, the surgical site was determined as unsuitable for implanting the Bonebridge device because of an overhanging dura mater. Therefore, a small epitympanic recess was opened, through which it was possible to visualize a portion of the ossicular chain: it resembled the short process of the incus and was immobile. After drilling the bone anteriorly and laterally, a complex of the first and second ossicles was revealed, a mass which was partially fused to the bony wall and constituted the front, side, and roof of a small tympanic cavity.

The mobility of the ossicular conglomerate was restored by removing the bony adhesion. The conglomerate, resembling the long crus of the incus, was attached to the mobile stapes. However, at this site, where the malleus handle is normally visible, a major malformation was evident. These conditions pointed to possible implantation of the Vibrant Soundbridge VORP 503 with an FMT and incus-SP-coupler (MedEl, Innsbruck, Austria) which might be attached to the portion resembling the short process of the incus. Therefore, on the surface of the temporal bone squama, a cavity was formed for the internal part of the device, which was affixed with 2 screws, and the FMT was attached to the ossicular conglomerate (**Figure 3**).

Closure of the surgical site was performed in the normal manner, with subcutaneous and cutaneous sutures. Implant activation was 1 month after surgery, and improvement in hearing

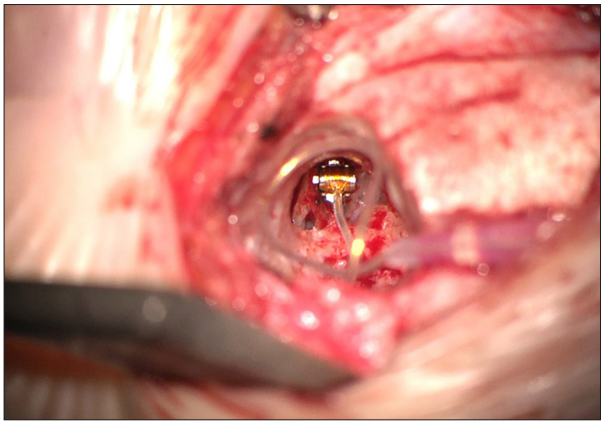

Figure 3. Method of attaching the Vibrant Soundbridge VORP 503 with the floating mass transducer and incus-SP-coupler in the temporal bone.

was confirmed at 1 and 12 months after implantation, using free-field audiometry (**Figure 4**).

Discussion

Currently, most authors prefer middle ear implants over traditional reconstruction methods [5,7], which produce generally unsatisfactory results and comorbid complications. It is now thought that reconstruction of the external ear canal can be

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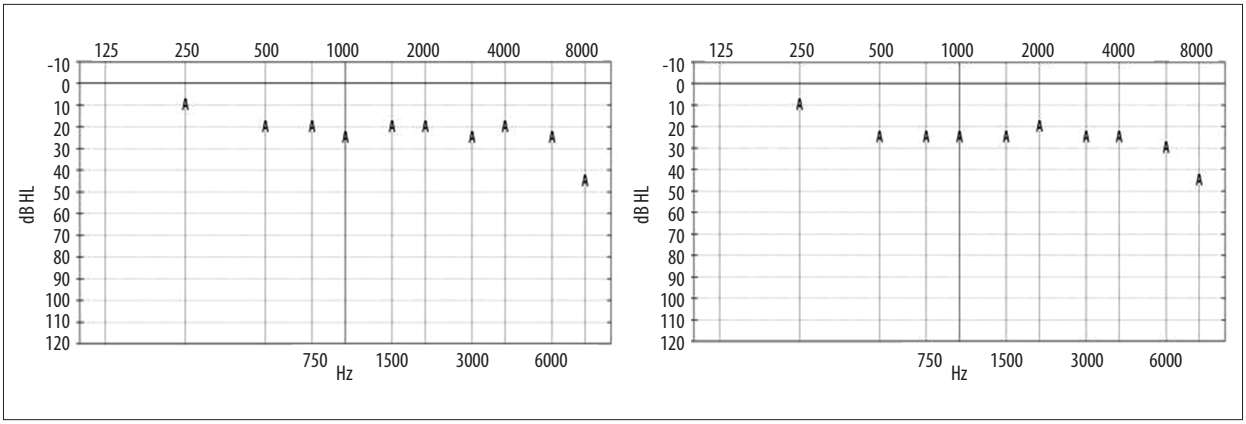

Figure 4. Free-field audiogram at 1- and 12-month follow-up.

successfully done only in cases where there is minor deformity, the tympanic cavity is of normal size, and it is possible to intraoperatively confirm that the ossicular chain is fully mobile [2]. Although there is sometimes the opportunity to rebuild the auricle (for cosmetic effect), in most cases, a reconstructed external ear canal usually becomes re-occluded. Malformation of the external and middle ear also tends to be associated with resistance to hearing improvement. Thus, patients with bilateral aural atresia usually require alternative methods of treatment.

The simplest and most popular way to improve hearing is with a bone conduction hearing aid. It is crucial for the good speech development of young children with bilateral malformations to encourage them to wear such devices [12]. However, device usage tends to be limited, especially in small children, because of the need to maintain a constant pressure on the skull, a circumstance that leads to bone deformity and poor appearance in the long term [3,12].

A more advanced way to boost perception of sound is to employ a bone-anchored hearing aid (eg, Cochlear BAHA, Oticon Ponto) or a transcutaneous bone conduction implant (Medtronic Sophono, Cochlear BAHA Attract), which give better and more stable performance and are more comfortable to wear. However, because of anatomical restrictions, these devices cannot be used in children under 5 years of age. In open skin devices, problems may occur with integration with abutting bone, abnormal skin reactions, and even irregular temporal bone development. According to some authors, this type of implant may require surgical intervention in up to 40% of children [13]. The use of bone-anchored hearing aids is limited in patients with external ear canal atresia and microtia, in whom aural reconstruction may be considered in the future [3].

Over the last several years, a good solution for cases of bilateral atresia has been the Vibrant Soundbridge system, which now has an increased number of advocates [4,5,7]. However, because of small patient numbers, there are currently no clear recommendations regarding the technical details of the procedure. Depending on the level of deformity of the natural structures of the ear, the FMT may be attached to different anatomical elements, most often the long crus of the incus or the stapes [8]. To bypass a defective ossicular chain in cases of congenital malformation, another site for attachment is the round window [5]. However, because of its anatomical variability, the round window is much less accessible, from a surgical point of view, than the ossicular chain. In addition, drilling next to the round window may cause sensorineural hearing loss [14], and direct stimulation of the round window by the FMT is not always possible. According to some authors, insertion of a fascia between the round window membrane and the FMT can worsen sound conduction, especially at low frequencies [15,16]. Attaching the FMT to the ossicular chain seems to be a better option, and there is also a lower risk of its translocation. Generally, most authors find that revision operations are less common in cases of attaching the FMT to the ossicles than in those attaching to the round window [10].

Another point of attachment used recently is the short process of the incus. Such an attachment needs to be considered because it is an easier surgical approach than other approaches [10]. In 3 children with microtia and absence of the external ear canal, Célérier et al obtained good functional results from implanting the Vibrant Soundbridge device by attaching the FMT to the short process of the incus. Similar to our experience, they found that the procedure was relatively simple and did not need a posterior tympanotomy, which poses a danger to the facial nerve in congenitally malformed ears [17].

In our present case, the short process was chosen owing to difficult anatomical conditions. After excess bony adhesions were removed, the ossicular chain was mobilized, and attachment of the FMT was considered stable. In our view, attachment of the FMT to the short process of the incus appears to be a good and workable solution; however, more research on a larger group of patients over a longer term is required.

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Another type of middle ear implant which can be considered for children over 5 years old with microtia is the Bonebridge [4,18]. Comparing it to the solutions mentioned above, the significant advantages of the Bonebridge would be lack of skin problems and stable positioning [4,11,13]. However, implantation of this device is possible only when there is sufficient space in the temporal bone, which can present as a problem in the abnormal mastoid process that frequently occurs in microtia [18]. A promising tool for preoperative assessment can be 3D high-resolution computed tomography rendering, which enables better recognition of key anatomical features [19].

Because of overhanging dura, the Bonebridge device could not be implanted in our patient. An alternative retrosigmoid approach for Bonebridge application requires neurosurgical experience and is not practiced in our center [20].

Considering the advantages and disadvantages of all implantable devices, our recommended and primary choice is the Bonebridge, when anatomical features are suitable. The next choices would be the BAHA Attract or Vibrant and, finally, the BAHA Connect or Ponto. However, each patient's case should be analyzed separately, while considering the experience of the medical center and audiological, anatomical (including possible skin reaction), surgical conditions, and preferences of the patient.

Conclusions

The MedEl Vibrant Soundbridge system was successfully used in a case of bilateral external ear canal atresia, with malformation of the middle ear. Mobilization of the hypoplastic sound conduction apparatus enabled effective attachment of the FMT to the remaining element, which resembled the short process of the incus. However, additional research in a larger population of patients with congenital malformations of the middle and external ears is needed to confirm the suitability of the Vibrant Soundbridge in such cases.

Conflicts of Interest

None.

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