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Round Window Stimulation with an Implantable Hearing Aid (Soundbridge®) Combined with Autogenous Reconstruction of the Auricle – A New Approach

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Key Words
Implantable hearing aid · Active middle ear implant · Round window · Ear malformation · Hearing loss · Auricle · Microtia · Atresia

Abstract
Background: Congenital malformations of the auricle are often combined with atresia of the outer ear canal and malformations of the ossicles, representing aesthetic as well as functional deficits. Optimal treatment should therefore address both aspects equally. This report describes a new approach, combining the reconstruction of the auricle with implantation of an active middle ear hearing aid, stimulating the round window membrane. ***Method:*** A 33-year-old male patient, with bilateral ear microtia, fibrous atresia of the external ear canals and malformation of the ossicles due to Treacher Collins-Franceschetti syndrome was included in the study. In stage one, the cartilage framework of the new auricle, made of autogenous rib cartilage, was fabricated and implanted. During stage two, the auricle was elevated, a retro-auricular sulcus was formed and a Vibrant MED-EL Soundbridge® device was implanted. The transducer was coupled to the round window membrane. ***Results:*** Both functional and aesthetical results were favourable. Aided thresholds were between 15 and 30 dB in the frequency range of 0.75–6 kHz, monosyllabic word understanding at 65 dB SPL increased from 0 to 80%. ***Discussion:*** Combining aesthetic and functional rehabilitation, autogenous reconstruction of a new auricle together with the implantation of an active middle ear hearing aid, coupled to the round window membrane, is a promising new approach.

Introduction

Severe congenital malformations of the auricle are often combined with atresia of the outer ear canal, either bony or fibrous, partial or complete, a missing eardrum and malformations of the ossicles, representing aesthetic as well as functional deficits. Optimal treatment of combined congenital ear malformations should therefore address both aspects equally.

For aesthetic rehabilitation, reconstruction of the auricle using a framework fabricated from autogenous rib cartilage transplants together with regional skin and free skin grafts is a well-established technique [1–4] that can achieve excellent results if performed by experienced surgeons.

Alternatively, epithetic replacement of the auricle, coupled to bone anchors, is available, e.g. in patients with a severely compromised local skin situation or in those who are unwilling to undergo a more extensive surgery

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[5]. However, recent studies have established a high degree of subjective satisfaction and psychological acceptance of reconstructions with autologous cartilage, even if the results are not always perfect [6].

On the other hand, treatment of functional deficits in malformed ears with a conductive hearing loss is still not satisfactory in all respects.

Surgical reconstruction of the outer ear canal is often difficult, requires a long-term follow-up and complications like restenosis or recidivating infections are not uncommon [7]. Adjacent malformations of the ossicles are also difficult, if not sometimes impossible to reconstruct, especially if the stapes is severely malformed. Under these circumstances, surgical attempts of reconstruction might be risky or contraindicated.

Even if ears to reconstruct are carefully selected, e.g. by using preoperative scales like the Jahrsdoerfer scores [8], functional results remain extremely variable [9, 10]. In the large majority of reconstructions, a residual conductive hearing loss of 20–40 dB will persist [11, 12]. Postoperatively, this situation would require the fitting of hearing aids to obtain optimal benefits of the ear, a task which is very difficult as ear moulds are often not well tolerated in reconstructed ear canals due to accumulation of debris and recidivating infections. Also, wearing a behind-the-ear hearing aid might be impeded by an insufficiently deep retro-auricular sulcus. Thus, the search for alternative methods of treatment that give more reliable, predictable and satisfactory results seems justified.

Active middle ear implants, also referred to as implantable hearing aids, have been developed for the treatment of sensorineural hearing loss and have been successfully used in large series of patients [13–15]. They provide acoustic amplification and transmission of sound energy by coupling a vibratory element directly to the ossicular chain. Two systems are available, a partially implantable device, the MED-EL Vibrant Soundbridge®, and a fully implantable device, the Otologics® MET, which is presently introduced in the EU. The Soundbridge consists of an external audio processor and an implantable part, the vibrating ossicular prosthesis (VORP). The active vibrating element is a small electromagnetic element, called the floating mass transducer (FMT) that is normally attached to the long process of the incus with an intact ossicular chain. It transmits the vibrations to the stapes footplate.

In malformed ossicular chains, this specific way of coupling is in general difficult or impossible because abnormal anatomy prevents correct placement and fixation to the chain and the ossicular chain is often immobile. However, alternative methods of coupling exist: the FMT might be coupled directly to the stapes footplate, if it is mobile and if the oval window niche is accessible as well as sufficiently large.

Based on computer model calculations and first clinical results reported by Colletti et al. [16], stimulation of the cochlea by coupling of the FMT to the round window membrane (RWM) may be a promising second alternative. Round window stimulation can be regarded as a reversal of the normal pathway of activation of the basilar membrane.

The use of an active implantable hearing aid offers a promising perspective of a reliable and reproducible functional rehabilitation of middle ear malformations that may avoid many complications of surgical reconstruction of an outer ear canal, eardrum and ossicular chain and circumvent the disadvantages of conventional hearing aids in reconstructed outer ear canals.

We have therefore initiated a study to evaluate the possibility of using an implantable hearing aid for functional restoration in conjunction with the reconstruction of the auricle using autologous rib cartilage. The aim of this report is to demonstrate the feasibility and outline the surgical procedures that are used.

Methods

Patient

A 33-year-old male subject with Treacher Collins-Franceschetti syndrome, congenital, bilateral severe conchal type microtia, and fibrous atresia of the outer ear canal was selected for the study. The bony canal was partially present. He had a transposition of the microtial ear in his childhood and bilateral osteotomies of the mandibula. Preoperative audiometric assessment showed bilateral conductive hearing loss of 55–60 dB; bone conduction thresholds were normal in pure-tone audiometry. Speech understanding was evaluated using the Freiburg test of speech understanding with numbers and monosyllables. Speech discrimination was 50% on the left side and 30% on the right ear at 95 dB HL; at speech levels of 65 dB HL, monosyllable understanding was 0%. The patient did not wear hearing aids.

Preoperative CT scans are shown in figures 1 and 2. They reveal bony partial meatal atresia, and a pneumatised middle ear with a rudimentary incus/malleus complex, partially fixed to the lateral attic wall (fig. 1). The oval window is present, but empty, the inner ear is normal, and the round window has a normal configuration (fig. 2).

Implantable Hearing Aid (Vibrant MED-EL Soundbridge)

The Vibrant MED-EL Soundbridge consists of an external audio processor and the implantable part, the VORP.

The audio processor is equipped with a microphone and the electronics that are essentially identical to the SIGNIA® hearing

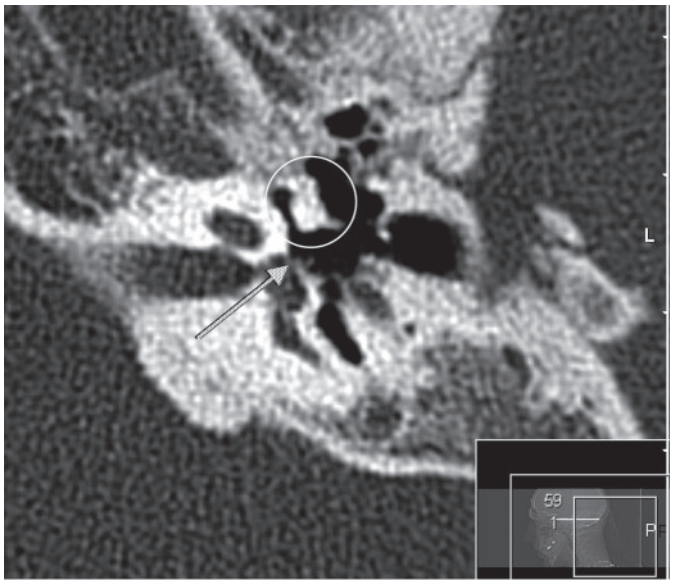

Fig. 1. Preoperative CT scan with empty oval window niche (arrow) and incus/malleus complex (circle).

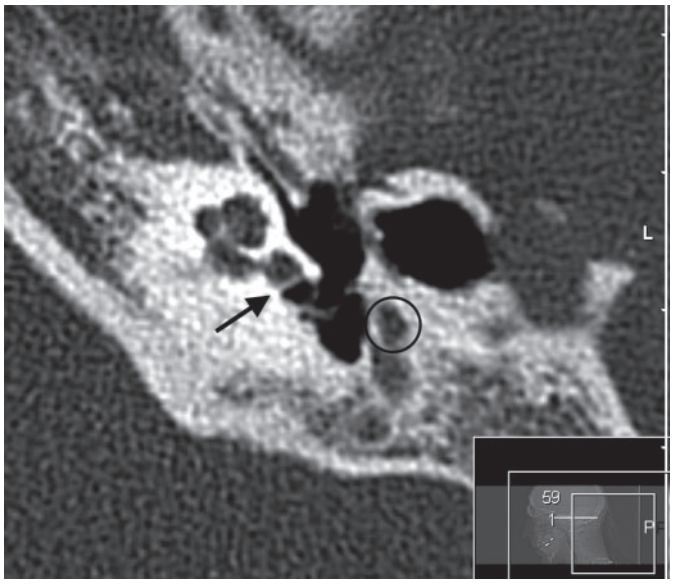

Fig. 2. Preoperative CT scan, showing the intact round window (arrow) and the facial nerve (circle).

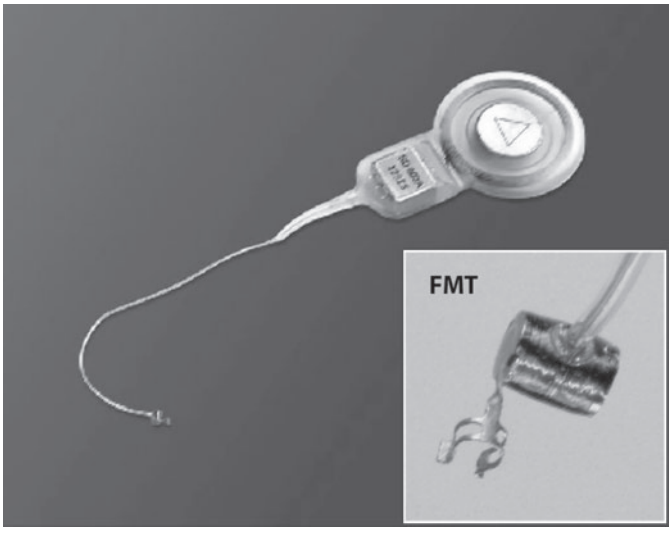

Fig. 3. VORP with receiver coil, demodulation electronics and FMT.

aid technology. The processed and amplified signal is transmitted transcutaneously via a radiofrequency link to the VORP. The electronic chip of the VORP demodulates the signal and transfers it to the FMT by isolated and silicone-coated gold wires (fig. 3). The FMT has a diameter of 1.4 mm and a length of 2.1 mm. Normally, it is crimped to the long process by an attachment clip. For this specific application, the clip was cut by a steel wire scissor.

Surgical Procedures

The surgical procedure used in this case consists of three essential steps and two stages.

In the first stage, the rib cartilage is harvested and the auricle is formed from the autologous cartilage and implanted in a subcutaneous pocket in its definite position.

During the second stage, the implantation of the VORP and placement of the FMT onto the RWM is performed as step two of the procedure. In step three, the newly formed auricle is elevated and a retro-auricular sulcus is created. It is of great importance that these steps are well coordinated. All incision lines of the skin and fascia or muscle have to be carefully planned in order not to challenge the blood supply of the skin region over the new auricle or the temporoparietal fascial flap, vascularised by branches of the temporal artery that is potentially needed. Figure 4 shows the skin incision line for stages one and two as well as the incision of the galea/periost and the temporal fascial graft in stage two.

Stage 1: Formation of the Auricle

An auricle framework was created in the first operation with the use of autogenous rib cartilage harvested from the 6th to the 9th rib, based on the technique described by Nagata [2]. The new auricle consists of a base plate constructed from ribs 6 and 7 with a carved scapha and triangular fossa. The anterior and posterior crura are reinforced by application of a y-formed cartilage. The 9th rib was used to form the helix of the new auricle. After creating the new auricle, we placed it in a subcutaneous pocket of the mastoid plane.

The remaining pieces of rib cartilage were stored under the thoracic skin to be placed under the base plate of the framework to construct the retro-auricular sulcus in the next operation (for a detailed description, refer to Staudenmaier et al. [4]).

Stage 2: Implantation of the VORP and Elevation of the Auricle

Two months after implantation of the framework, the new auricle was raised to construct the retro-auricular sulcus. As a small skin dehiscence over the posterior part of the auricle had formed due to poor vascularisation of the skin, this step was combined with the preparation of a temporoparietal fascial flap to cover the sulcus.

The skin incision line of stage one was prolonged in the posterior direction to allow for the implantation of the VORP. A second skin incision perpendicular to the extended retro-auricular incision line allowed for the preparation of the temporoparietal fascial flap. The periosteal incision was carried out posterior to the temporalis muscle in order to preserve the vascularisation of the temporoparietal fascial flap (fig. 4). A broadly based flap was dissected and elevated; the nutritional arteries could not be identified as they were probably ligated during previous osteotomy of the mandibula. The framework was elevated and carefully deflected anteriorly to allow access to the middle ear.

To assess the middle ear and the round window, two routes are possible. The choice should be made according to the individual anatomical situation as determined by clinical examination and high-resolution CT scan.

In cases with a well-pneumatised mastoid, the middle ear can be approached after cortical mastoidectomy, identification of the antrum, and the semicircular canal as well as the incus (if present), via a posterior tympanotomy, eventually sacrificing the incus bridge.

Alternatively, if the mastoid is poorly pneumatised, the middle ear can be approached following the course of the atretic external ear canal. This approach has been chosen in the present case. The soft tissue was removed and the bony channel was enlarged until the middle ear could be entered with ease. Using navigated control based on the preoperative CT scans may be helpful [17].

An abnormal course of the facial nerve is frequent in malformed ears and has to be identified on the preoperative CT scans as well as intraoperatively. Facial nerve monitoring should be employed to avoid accidental facial nerve injury.

After the middle ear was opened, the ossicles and the oval window niche were identified. We found a severely hypoplastic stapes that was partially fixed in a small oval window. The round window niche was exposed. The overhanging rim of the oval niche had to be removed until at least two thirds of the RWM were clearly visible. Mucosal folds may sometimes be present laterally to the RWM and be mistaken for the RWM. They have to be removed carefully until the RWM can be clearly identified. Exposing the RWM has to be performed with greatest care to avoid rupture of the membrane and noise trauma of the cochlea. We recommend giving 250 mg of Solu-Decortin intravenously to obtain protection of the cochlea against noise trauma.

A bony bed was drilled for placement of the electronic housing of the VORP and a subperiostal pocket was prepared to lodge the antenna coil. A tie-down suture was anchored to the bone. If no mastoid is present, it is necessary to drill a circular recess anterior to the implant to receive eventual surplus of the wire. A groove in the posterior part of the ear canal was drilled to accommodate the wire. The VORP was placed in the prepared bed (fig. 5) and fixed with the tie-down suture. The attachment clip of the FMT that normally serves to crimp it to the incus had to be cut using a wire scissor or a cutting forceps. The FMT was covered with a piece of fascia to protect the RWM and serve as coupling material and placed in close contact to the RWM. To secure the FMT, we placed the wire FMT in the bony groove of the ear canal (or alternatively in the facial recess) and fixed it with bone paté, approximately 2 mm away from the FMT. Bone paté was obtained by mixing sampled bone dust with fibrinogen that is activated in situ by addition of thrombin.

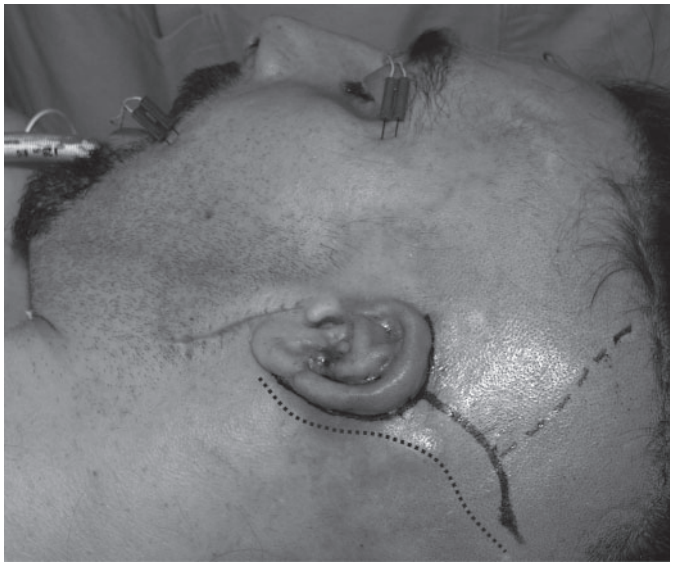

Fig. 4. Incision lines for the second stage of auricular reconstruction and implantation of the VORP. The dotted line indicates the incision of the periost. Facial nerve monitor needles are visible.

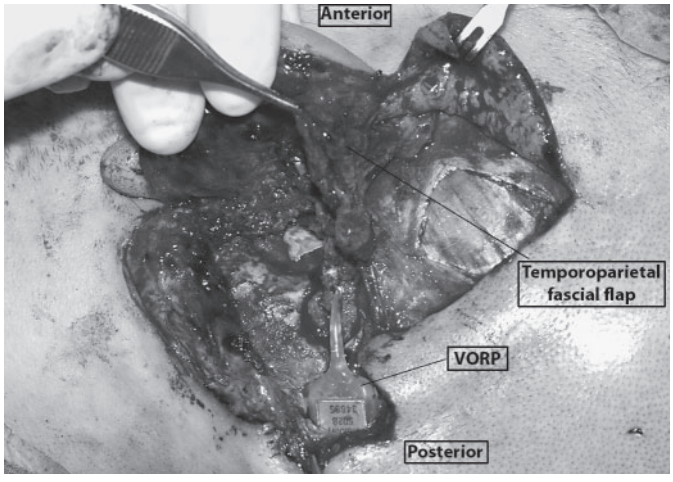

Fig. 5. Intraoperative situs with implanted VORP, elevated auricle and elevated temporoparietal fascial flap.

Fig. 6. Postoperative result with the new auricle and audio processor of the VORP.

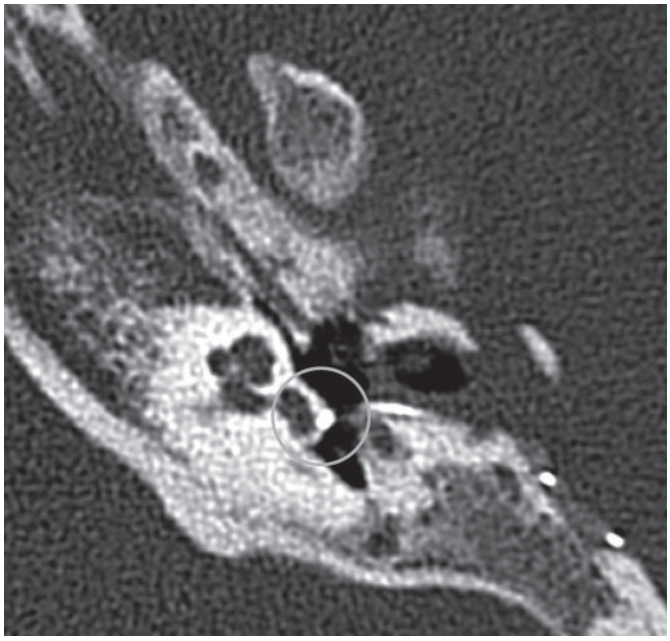

Fig. 7. Postoperative CT scan with FMT placed at the round window (circle).

The wire placed in the bony groove of the ear canal was completely covered with bone paté. In addition, we placed a fascia graft to protect the wire against possible extrusion or infection.

Excess wire was lodged in the bony recess anterior to the VORP. The fascia and periostal layer were sutured over the implant.

By placing a rib cartilage wedge, harvested for creating the framework and stored under the thoracic skin during the first step, under the base plate of the framework, the auricle was elevated to obtain a satisfactory projection of the new pinna. The second step of auricular reconstruction was accomplished by covering the cartilage wedge and the posterior surface of the framework with the temporoparietal flap, deflected downwards. Using V-Y skin plasty, the retro-auricular skin was closed to form the medial layer of the retro-auricular sulcus; a full-thickness skin graft of the thoracic skin covered the lateral part of the sulcus.

Results

After primary reconstruction of the right auricle in the standard technique described above, the first stage of reconstruction of the left auricle was performed. Slight problems of skin perfusion led to a small skin dehiscence over the middle part of the auricle, requiring additional cover by a vascularised temporoparietal fascial flap during stage two. Wound healing was complete and the VORP could be activated 6 weeks postoperatively (fig. 6). A postoperative CT scan demonstrates the FMT in correct position in the round window niche (fig. 7).

After first fitting, bone conduction thresholds were essentially unchanged compared to preoperative values. Aided thresholds with the Soundbridge showed lower amplification up to 500 Hz, but satisfactory thresholds between 15 and 30 dB in the frequency range of 750 Hz to 6 kHz (fig. 8). Speech discrimination increased from 0 to 80% at 65 dB HL (fig. 9). His scores of 80% monosyllabic word understanding might reflect the fact that the patient was not a native German speaker, thus needing higher presentation levels to obtain 100% speech discrimination. When the VORP was activated, the patient reported to hear a soft buzzing sound that was initially bothering but continuously decreased over the following days and disappeared after reducing the gain of the amplification. We attribute this phenomenon to the fact that – given the patient's completely normal inner ear function and good coupling of the FMT to the inner ear – he was able to perceive some internal processor noise generated by the VORP. Results with the VORP are stable over 3 months of postoperative follow-up.

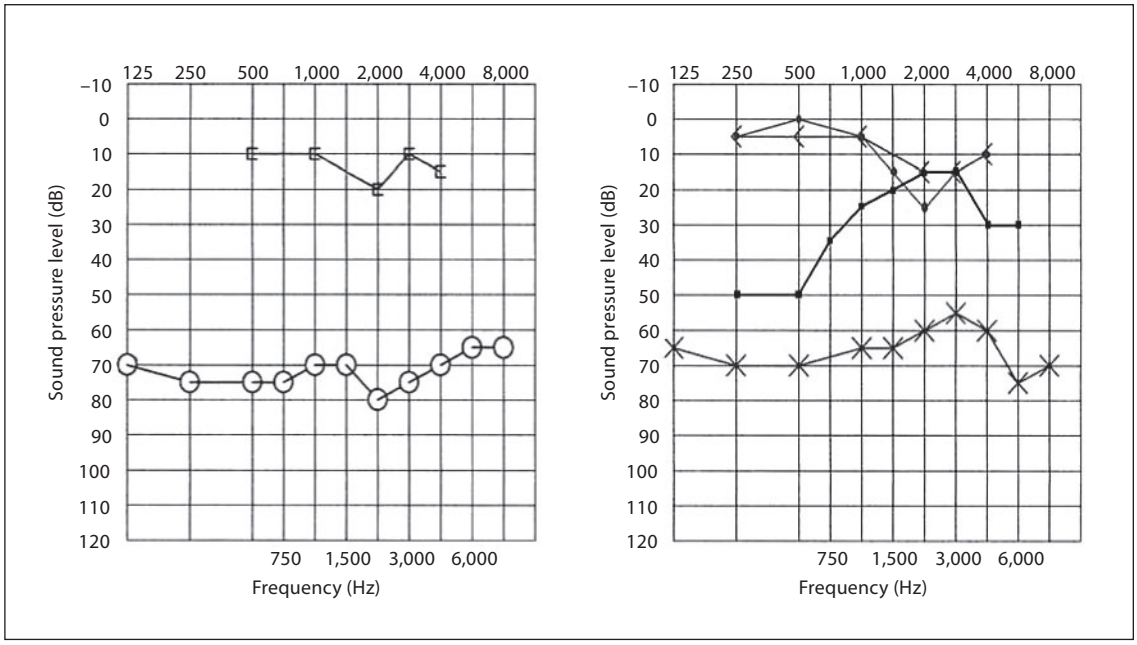

Fig. 8. Pure-tone audiometry with bone conduction ([= right ear; < = left ear, preop., ● = left ear, postop.), air conduction (○ = right ear; × = left ear) and aided thresholds with Soundbridge (■).

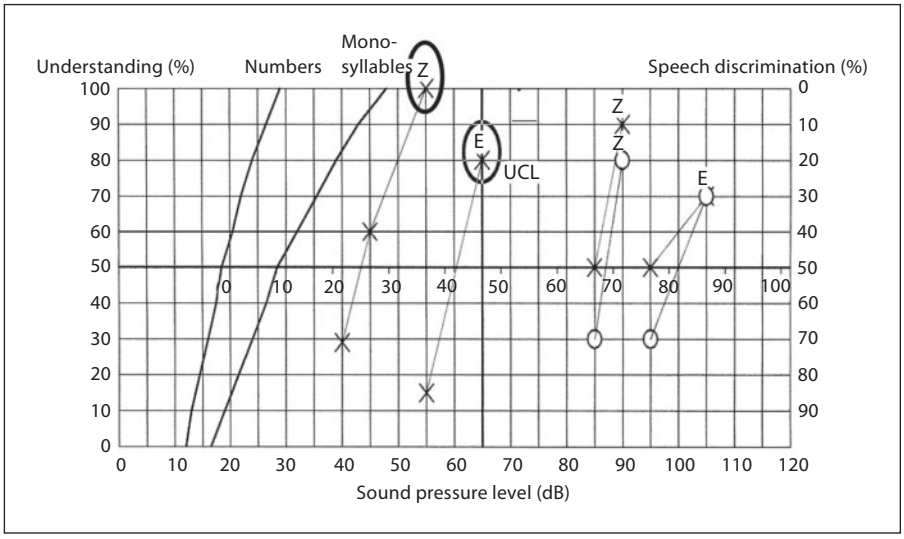

Fig. 9. Speech audiometry: preoperative values for monosyllables (E, left and right ear) and numbers (Z, left and right) and postoperative aided values with Soundbridge (E and Z, circled values).

Discussion

Functional and aesthetic rehabilitation of a combined malformation of the auricle, outer ear canal and the middle ear still remain a challenging surgical task. Reconstruction of the auricle can be performed with autogenous rib cartilage in a two-stage procedure [4]. In experienced hands, excellent results can be obtained. If the vascularisation of the local skin is compromised, e.g. by previous surgery, this technique can be combined with a pedicled fascia temporalis flap to provide additional vascularisation to the cartilage framework and the skin. Ep-

ithetic replacement of an auricle fixed with bone anchors is a valuable alternative, e.g. in the elderly; however, epithetics have to be replaced in regular intervals and the psychological acceptance of autogenous reconstructions is better as they form an integral part of the body, offering a definite answer in favour of the reconstructive approach.

Functional reconstruction, on the contrary, still remains an unsolved question. Bone-anchored hearing aids can be used for restoring hearing on the malformed ear. However, the bone anchors – albeit often well tolerated – may cause local infection and skin reaction in about 9–33% of patients [18, 19]. They require daily meticulous care, and the rate of extrusion is reported to be around 2–5% [20]. Also, bone conduction only offers limited directional hearing, as both cochleae are stimulated simultaneously.

Surgical reconstruction of the outer ear and restoration of the ossicular chain is a possible alternative. A new outer ear canal is drilled, either using an endaural approach following the normal anatomical course of the outer ear canal, or using a transmastoid approach, forming a modified radical cavity. This approach can be used if the mastoid is well pneumatised. The creation of an outer ear canal can also be combined with the reconstruction of the auricle [21]. In a two-staged procedure, the ear canal is formed with autologous rib cartilage, stabilised by a silicone stent and secondarily epithelialised with a skin graft.

However, in all of these techniques, restenosis of the ear canal, thickening, scarring or blunting of the new eardrum and infections are still common problems. In addition, reconstruction of a functional ossicular chain is often difficult, if not impossible, if the stapes is severely malformed or absent, or if the oval window niche is missing or the footplate fixed. Jahrsdoerfer et al. [8] have described a scoring system, underlining the significance of the situation of the stapes. Jahrsdoerfer scores of at least 7–8/10 are commonly recommended for the attempt to perform a reconstructive surgery. Thus, only a limited number of malformed ears may be eligible for reconstructive surgery.

But even in this selected material and in the best of hands, postoperative long-term residual conductive hearing loss mostly remains between 20 and 40 dB, requiring the postoperative use of hearing aids to make full use of the functional value of the reconstructed ear. Chandrasekhar et al. [11] report on the follow-up of 92 congenital aural atresiaplasties performed. Closure of the air-bone gap to less than 30 dB was achieved in 60% of primary surgeries and 54% of revisions. The most common complications were external auditory canal stenosis and lateralization of the tympanic membrane. External auditory canal stenosis due to bony regrowth was seen in 12% of primary cases and 11.5% of revisions; soft tissue stenosis was seen in 10% of primaries and 4% of revisions. Tympanic membrane lateralization was seen in 9% of primary surgeries and in 15% of revisions. A consensus report states that atresia repair surgery is worthwhile if proper patient selection is made by the use of stringent audiological and radiological criteria and state-of-the-art surgery is performed. Review of the literature demonstrated that even in the hands of the best surgeons a mean hearing gain of only 20–25 dB is achieved in atresia type II, and 30–35 dB in type I. Therefore, surgical reconstruction should only be done in the more favourable cases where postoperative hearing of <25–30 dB is attainable. Less favoured patients should be helped with bone-anchored hearing aids, as this type of surgery does not interfere with the future use of new techniques [9].

The use of hearing aids is difficult in reconstructed ear canals, as ear moulds are often not well tolerated and prone to recidivating infections and accumulation of debris.

The current report demonstrated the feasibility of functional hearing restoration using an implantable hearing aid (Vibrant MED-EL Soundbridge). We were able to show that the coupling of the FMT to the round window can give an excellent transmission of the vibratory energy to the inner ear and the basilar membrane following an original idea by Colletti et al. [16].

In conclusion, the implantable hearing aid (Vibrant MED-EL Soundbridge) with an active element, the FMT coupled to the RWM, is a promising alternative to reconstructive surgery of the normal air conduction pathways, which often remains unsatisfactory. It can be combined with a reconstruction of the auricle, using autogenous rib cartilage.

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