Combined Aesthetic and Functional Reconstruction of Ear Malformations

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Abstract

Background: Surgery for major malformations of the outer and middle ear involves aesthetic as well as functional aspects. Whereas reconstruction of the auricle with autogenous rib cartilage is well established and has shown favorable results, functional repair using classic reconstructive techniques is possible only in a limited group of patients and the outcome is often unsatisfactory. Active middle ear implants (MEI) offer a promising alternative to reconstructive surgery. Method: Fifteen patients with ear malformations underwent implantation of an active middle ear implant (Soundbridge®), with or without concomitant reconstruction of the auricle. The vibrating element, the floating mass transducer (FMT), was coupled either to the round window, stapes, oval window or incus, according to each individual’s anatomical middle ear situation. Aesthetic as well as functional outcomes were evaluated. Results: Implantation could be integrated into aesthetic reconstruction of the auricle without complications. In 14/15 patients, a satisfactory functional result could be achieved (<30 dB pure-tone audiometry). Neither facial nerve palsy nor inner ear hearing loss was observed after implantation. Conclusion: The versatile form of the FMT of the Soundbridge allows for adaptation of the coupling procedure to the individual anatomical situations. Implantation of a Soundbridge MEI is a valuable option for functional reconstruction of the malformed ear, which may offer more consistent and reliable results than classic reconstructive surgery.

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Classic microtia is in most cases combined with aural atresia of the external ear canal as well as considerable dysplasia of the middle ear, involving the ear drum, the ossicles, the facial nerve and possibly the oval and round window niches. Typically, a functional impairment with conductive hearing loss of around 50–60 dB is present. The reduced hearing ability, especially in patients with bilateral microtia, will challenge speech and language development. Therefore, fitting a bone-conducting hearing aid is mandatory in the first months of the patient’s life to ensure normal development. However, patients with a unilateral malformation may also suffer from functional impairment in addition to the aesthetic problem, e.g. in noisy acoustic environments.
Therefore, treatment of microtia in combination with aural atresia and middle ear malformation has to take into account the functional as well as the aesthetic aspect of the disease.

Siegert et al. [1] evaluated malformed petrous bones in patients with third-degree microtia using high-resolution CT, and found atresia of the external auditory canal in 88% and normal middle ear ossicles in only 3%. The most common dysplasia of middle ear structures is related to the complex of the malleus and incus. In 69%, this had morphological malformations, and in 27% it was absent. Severe abnormalities of the labyrinth were rare in these patients. Moreover, displacement of the facial nerve canal can be expected in about 77% of patients with auricular dysplasia, a fact that has important implications for surgery in these cases.

Both auricle reconstruction and reconstruction of a malformed middle ear with atresia are difficult and challenging operations. While reconstruction of the auricle mostly produces an excellent aesthetic outcome, the indication for operations improving hearing ability by building up the external auditory canal and middle ear should be judged carefully, because functional outcomes vary widely and are often unsatisfactory. The chances of achieving a satisfactory result depend on the individual anatomical situation, which is evaluated preoperatively by high-resolution CT of the petrous bone and can be judged with the help of radiological scores [1, 2]. In general, all authors agree that surgical reconstruction of the ear canal, ear drum and middle ear should only be attempted in patients with favorable anatomical conditions, e.g. a present stapes and sufficiently aerated middle ear. However, even in cases which are eligible to undergo surgical reconstruction, reduced hearing ability requiring an air conduction hearing aid remains at least in more than half of patients, even in the series of most experienced centers. Because of this limited success rate, reconstruction of the external ear canal and the middle ear is commonly only recommended in cases of bilateral microtia.

Active middle ear implants (MEI), also referred to as implantable hearing aids were developed for the treatment of sensorineural hearing loss, and have been successfully used in large series of patients [3]. They provide acoustic amplification and transmission of sound energy by direct coupling of a vibratory element to the ossicular chain. Currently, 2 systems are available, a partially implantable device (the MED-EL Vibrant Soundbridge) and a fully implantable device, which has been introduced in the European Union. The Soundbridge consists of an external audio processor and an implantable part, the vibrating ossicular prosthesis (fig. 1). The active vibrating element is a small electromagnetic element called the floating mass transducer (FMT), which is normally coupled to the long process of the incus with the ossicular chain intact. It transmits the vibrations to the stapes footplate.

In malformed ossicular chains, however, this way of coupling is generally difficult or impossible because anatomical abnormalities may frequently prevent correct placement and fixation to the chain. In addition, the ossicular chain in the malformed ear is often immobile. Fixation of a vibrating transducer to a fixed ossicular chain would probably not transmit enough energy to the inner ear. However, alternative
methods of coupling may exist. Classic experiments by Wever and Lawrence [4] have demonstrated that the transfer of vibratory energy to the inner ear and the subsequent evocation of hearing sensations can be achieved at every point of the cochlea [via ossicles and stapes, directly at the footplate of the oval window, at the round window, or via a newly created third window (fenestration) of the cochlea]. Thus, the various strategies of coupling the FMT to the inner ear fluid will probably result in adequate hearing sensation.

The FMT might be coupled directly to a mobile stapes footplate if the oval window niche is large enough and surgically accessible.

Based on computer model calculations and the first clinical results reported by Colletti et al. [5], stimulation of the cochlea by coupling the FMT to the round window membrane may be a promising second alternative [6]. Round window stimulation may be regarded as a reversal of the normal pathway of activation of the basilar membrane.

Provided that effective and stable coupling of a vibratory transducer can be achieved surgically, active implantable hearing aids offer a promising opportunity for reliable and reproducible functional rehabilitation of middle ear malformations, and may avoid complications of surgical reconstruction and circumvent the disadvantages of conventional hearing aids in reconstructed outer ear canals.

Aesthetic reconstruction of the outer ear is well established, and in general gives very satisfying results. The most common way of auricular reconstruction is the use of autogenous rib cartilage based on the technique described by Nagata [7]. Further development and refinement of the technique [8–11] has made it possible to achieve excellent results in the majority of patients, if performed by experienced surgeons.
In the first step, the cartilage framework is created and transplanted under the skin of the mastoid plane. In a second operation a few months later, the new auricle is raised by placing a cartilage wedge under the base plate of the auricle framework. In order to accomplish the reconstruction, the posterior side of the framework and the cartilage wedge are covered with a full-thickness skin graft. Commonly, a third step is necessary for minor refinements.

In an attempt to combine esthetical and functional reconstruction, we have initiated a study to evaluate the feasibility of using an implantable hearing aid for functional restoration in conjunction with reconstruction of the auricle using autologous rib cartilage. The aim of this report is to demonstrate the feasibility of this technique and outline the surgical procedures that are used.

Method

Preliminary Considerations

Within the group of possible candidates for combined esthetical and functional reconstruction using an active middle ear implant, 3 groups may be differentiated according to the sequence of surgical steps. These are patients who have: complete reconstruction of the outer ear before an MEI is inserted; reconstruction of the outer ear performed at the same time as implantation of an MEI; implantation of an MEI preceding reconstruction of the outer ear.

The timing of the implantation in relation to the auricle reconstruction is of importance. If the implantation is performed prior to the reconstruction, incision lines have to be planned carefully after consultation with the surgeon responsible for the plastic reconstruction to avoid scars that interfere with the reconstructive procedure. If implantation is planned concomitantly to the reconstruction, we have to consider at which stage implantation would be best performed. Integrating the implantation into the reconstructive procedure has the principle advantage of avoiding an additional surgical intervention. The reconstructive surgery comprises 2 principle steps. During the first step, the cartilage framework is built from costal cartilage and placed into subcutaneous tissue. During the second step, the auricle is elevated and retroauricular sulcus is formed. A critical point during the first step is possible skin breakdown or infection. It is possible that implantation at that stage would increase the risk of skin breakdown and infection, since tissue manipulation and tension would be increased, and put the implant at risk. In addition, preparation of the subsequent steps might harm the electrode of the implant. We therefore favor integrating the implantation into the second stage of reconstruction, when the skin and framework have already stabilized and are sufficiently revascularized. However, implantation may also be performed after reconstructive surgery has already been finished. Implantation after completed reconstruction also imposes an additional surgical procedure, but, other than this, it has no major disadvantage if the incisions and manipulation avoid damaging the vascularization of the newly formed auricle.

Preoperative Diagnostics and Preparations for Surgery

- Assessment of auricular deformity and atresia at birth.
- Early postnatal pediatric counseling with a general physical examination to search for other congenital anomalies.
Early evaluation of auditory function in both unilateral and bilateral atresia.

Bone-conduction brainstem-evoked response audiometry for bilateral cases of congenital aural atresia within the first few weeks of life. The incidence of an association between an inner ear abnormality and congenital aural atresia is uncommon, but must be excluded.

Imaging in cases of aural atresia can be postponed until surgery is planned.

For reconstruction of the outer ear canal or middle ear high-resolution CT scans in both the axial and coronal planes are important. The following are critical pieces of information required for possible repair:

- Presence of sufficient space to create a new ear canal;
- Degree of pneumatization of the temporal bone;
- Presence and appearance of the ossicular chain;
- Course of the facial nerve, focusing on the relationship of the horizontal portion to the oval window and the location of the mastoid segment;
- Existence of the oval window and stapes footplate;
- Existence of a round window and its relation to the facial nerve;
- Anatomy of the cochleovestibular system.

Reconstruction of the outer ear canal and middle ear is recommended between the 5th and 10th years of life; reconstruction of the auricle with autogenous rib cartilage is usually possible from age 8 years upwards, when sufficient cartilage is available. Favorable conditions for Soundbridge implantation are:

- Sufficiently pneumatized mastoid process;
- Aerated tympanic cavity;
- Presence of ossicular chain (incus-malleus complex, stapes);
- Detectable oval and round windows;
- Route of the facial nerve that allows access to the target structure for coupling.

If available, 3D reconstruction may be helpful for planning the surgical access. Navigated surgery may be a useful option; however, the authors have not yet used this technique in surgery of the malformed ear.

The operation should always be carried out under facial nerve monitoring to prevent injury to this particular nerve, which is especially at risk in implantation of the malformed ear.

Surgical Procedures

Reconstruction of the Auricle and Preparing the Implant Bed

In the first step, the future position of the auricle is determined by taking into account the hairline and degree of hemifacial dysplasia. If present, the contralateral normal ear is used as a model. A stencil of the normal ear is built, sterilized, and used intraoperatively to adjust the dimensions of the new auricle to the normal contralateral ear. The cartilage is harvested from the 6th, 7th and 8th ribs, and then the cartilage framework is created (fig. 2). This is transplanted into a subcutaneous pocket of the mastoid plane, in the desired position. In the second operation, which is scheduled about 8 weeks after the first operation (after complete healing and recovery of the soft tissue), the auricle will be elevated and the retroauricular sulcus created. For this step, a curved retroauricular incision is used. If implantation of the MEI is planned at this step, the incision is enlarged posteriorly by using an incision line perpendicular to the retroauricular incision (fig. 3a).

The periosteum is incised in a line created with regard to the skin incision, in order to avoid infection of the implant in case of skin suture dehiscence (fig. 3b).

In general, placement of the implant housing should follow the established guidelines of cochlear implant surgery. The position of the housing should be planned with sufficient distance between the posterior rim of the cartilage framework and the anterior rim of the housing to
Fig. 2. Case study: 9-year-old boy with grade III dysplasia. a Preoperative situation. b Cartilage framework built out of autologous rib cartilage. c Aesthetic result after 2 operations. MEI implantation was performed at a later stage.

Fig. 3. Integration of MEI into the second stage of auricle reconstruction: operative procedure. a Incision line. b Preparation of the musculoperiosteal flap and the pocket for the implant housing. c Combined transmastoid/meatal access and placement of the implant. d Positioning the FMT against the round window membrane (access under the facial nerve).
Combined Aesthetic and Functional Reconstruction

avoid later conflict and implant exposure in cases of skin problems at the newly formed retroauricular sulcus. A bony bed for the implant should be drilled at the appropriate position and tie-down sutures should be prepared to hold the implant securely in place. These tie-down sutures can be fixed to the bone or to the surrounding periosteum. After completed placement of the implant housing (fig. 3c) and closure of the periosteal layer, a semi-lunar piece of cartilage is placed under the auricular framework to form the posterior conchal wall and to elevate the new auricle. This cartilage is then covered with a pedicled superficial temporalis flap or a mastoidal musculoperiosteal flap to provide secure soft tissue coverage of the cartilage. Grafting of the rear aspect of the auricle with a thin split-thickness skin graft completes the formation of the retroauricular sulcus.

Surgical Access to the Middle Ear

Based on the preoperative CT scan, surgical access is planned either as a transmastoid procedure (if the mastoid is sufficiently pneumatized), a transcanal procedure involving drilling along the course of the atretic ear canal (if the mastoid is not pneumatized), or a combined procedure using both access routes in combination. The starting point should be determined using intraoperative landmarks, such as a visible mold in the region of the pars tympanica, the horizontal line of the zygomatic arch and the mandibular fossa. They should be related to the CT scan to determine the correct drilling position. Navigated surgery could be a helpful tool.

For the transmastoid approach, the technique is generally similar to the technique used for cochlear implantation, with cortical or complete mastoidectomy, and identification of the middle fossa plate, the sigmoid sinus, the horizontal semicircular canal and the short process of the incus. However, the posterior tympanotomy should not be performed until the facial nerve is clearly identified. Therefore, it may be recommended to open the epitympanum until the incus is clearly observed. Then, the facial nerve can be identified underneath in relation to the oval window and the stapes, and the course of the facial nerve can then be carefully followed visually by opening the facial recess (starting superiorly). As the facial nerve often travels more laterally and anteriorly in its mastoid portion in malformed ears, it may sometimes be necessary to pass underneath the facial nerve to gain access to the middle ear, e.g. the round window (fig. 3d). The middle ear structures, ossicles and oval and round windows are exposed as far as possible to be able to choose the optimal structure for coupling the FMT.

The final results of combined functional and aesthetic reconstruction are shown in fig. 4.
Coupling the FMT to the Vibratory Structures

In malformed middle ears, the incus and malleus often form a synostotic complex that is fixed anteriorly to the pars tympanica of the temporal bone. The long process may be abnormally formed or missing. Therefore, classic coupling of the FMT to the long process of the incus is often impossible or not the best choice.

If a stapes is present, mobile and sufficiently stable to support the FMT, direct coupling to the stapes head is possible. This might be achieved by direct superposition or placement of the FMT parallel to the stapes, fixing the FMT with a bend-over attachment.

If the stapes’ suprastructure is unstable or missing, then direct coupling to a mobile footplate is another option.

If the footplate is not mobile or the round window niche is too narrow, then enlarging the oval window and drilling a fenestration in the round window niche is another option.

In some cases, neither the ossicles nor the oval window may be readily accessible. Placement of the FMT against the round window membrane is another alternative way of coupling the FMT to the inner ear fluid that has been shown to give excellent results.

A visual overview of these possibilities is presented in figure 5.

Patients

In this series, we included 15 patients who underwent MEI implantation, either with or without reconstruction of the auricle. The age at implantation varied greatly, the youngest implantee was 5.7 years old, the oldest was 45 years old. Etiology was idiopathic in the majority of cases, 1 patient had Franceschetti/Treacher-Collins syndrome and 1 patient presented with CHARGE syndrome.

Six of fifteen patients had a normal external ear, and the remainder had grade II–III dysplasia (Weerda classification); 6/15 patients had a normal ear canal, while the others showed subtotal
stenosis or aplasia. Middle ear abnormalities were found in all patients. In 1 patient, who had undergone a previous attempt at reconstructing the external ear canal, a retention cholesteatoma was found intraoperatively during the implantation of the MEI. The cholesteatoma was removed in the same operation.

Results

Table 1 summarizes the operative procedures in terms of reconstruction of the auricle, operative access to the middle ear, coupling of the FMT and global outcome. Coupling to the round window niche was used most frequently (n = 8), followed by coupling to the stapes (n = 4). Due to the limited number of patients and the wide variations in etiology, individual anatomy and target of coupling, it is not possible at this stage to evaluate subsets of patients. In the following, we will therefore refer to the results of the overall group.

Wound healing was complete and without problems in all patients: no infection, suture insufficiency or wound healing problems in the area of the free skin graft were observed.

All but 2 patients obtained satisfactory hearing results after primary placement of the FMT. One patient had to be revised, as primary coupling to the round window niche was not satisfactory. After revision under local anesthesia with replacement of the FMT to the oval window niche, we obtained a satisfactory result. In 1 child, neither placement at the oval window niche nor the round window niche resulted in hearing sensation, although preoperative hearing tests suggested a mixed bilateral hearing loss (severe on the implanted ear, moderate on the contralateral ear) with a moderate inner ear hearing loss at the implanted side. Preoperative tests with a bone-anchored hearing aid were positive, and the child reported hearing on the implanted side. However, neither placement on the oval window niche or round window niche resulted in hearing sensation on the implanted ear.

Figure 6 presents the mean preoperative air conduction thresholds versus postoperative aided thresholds. Preoperative thresholds ranged between 70 and 60 dB. Aided thresholds with the Soundbridge were significantly increased (with a bell-shaped curve) in the main speech frequencies [mean values between 38 dB (0.5 kHz) and 22 dB (1.5 kHz)]. The mean functional gains with the Soundbridge MEI ranged between 20 and 36 dB (fig. 7); in some individual cases, this reached 65 dB. The hearing in the main speech frequency area was optimal, while in the high-frequency area it was sufficient. A weakness in amplification of the low-frequency area could be noted. This is probably due to technical limitations of the FMT (low weight and floating mass). However, in all patients tested so far, speech understanding was significantly increased in comparison to the preoperative situation, and subjective benefits were large. All patients who were successfully implanted continue to use the MEI during all waking hours, including those patients with unilateral malformation. They report increased ease of listening, spatial hearing and the benefit of being able to hear from both sides.
<table>
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RWN = Round window niche; OWN = oval window niche.
Discussion

Functional and aesthetic rehabilitation of 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 2-stage procedure.
In experienced hands, excellent results can be obtained. If the vascularization of the local skin is compromised, e.g. by previous surgery, this technique can be combined with a pedicled auriculotemporal flap to provide additional vascularization to the cartilage framework and the skin. Replacement of an auricle fixed with a bone-anchored prosthesis is a valuable alternative (in the elderly, for example); however, prostheses have to be replaced at regular intervals and psychological acceptance of autogenous reconstruction is better, as the implant forms an integral part of the body and offers a definitive solution.

Functional reconstruction, on the other hand, still holds many unsolved questions. Bone-anchored hearing aids can be used to restore the hearing of the malformed ear. However, the bone anchors, although often well-tolerated, may cause local infection and/or skin reactions in about 10% of patients; furthermore, they require meticulous daily care, and the rate of extrusion varies between 2% in adults and 5% in children. Also, bone-conduction devices only offer 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 by an endaural approach (following the normal anatomical course of the outer ear canal) or a transmastoid approach (forming a modified radical cavity). This approach can be used if the mastoid is well pneumatized. The creation of an outer ear canal can also be combined with reconstruction of the auricle [12]. In a 2-stage procedure, the ear canal is formed with autologous rib cartilage, stabilized by a silicone stent and epithelialized with a skin graft.

However, in all of these techniques, restenosis of the ear canal, thickening, scarring or blunting of the neo-eardrum and infections are still common problems. In addition, reconstruction of a functional ossicular chain is often difficult, if not impossible, when the stapes is severely malformed or absent, the oval window niche is missing or the footplate fixed. Jahrsdoerfer et al. [2] described a scoring system which underlines the significance of the condition of the stapes. Jahrsdoerfer scores of at least 7–8/10 are commonly recommended before an attempt to perform reconstructive surgery. Thus, only a limited number of malformed ears may be eligible for reconstructive surgery [13].

Unfortunately, even when this material is used in the best of hands, postoperative long-term residual conduction hearing loss usually 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. The use of hearing aids is difficult in reconstructed ear canals, as earmolds are often not well tolerated and prone to recidivating infections and the accumulation of debris.

Firmin et al. [14] also combined the construction of the auditory canal with auricle reconstruction, similarly integrating the functional procedure of atresia surgery and tympanoplasty in the second step of correcting the microtia. To line the drilled hole, they use a subgaleal fascial flap to ensure a well-vascularized
ground for the skin graft. After the otological procedure, the subgaleal fascia flap is harvested together with the galeal fascia, used for the reconstruction of the retroauricular sulcus and covering the posterior surface of the auricular framework and the cartilage wedge. After placing a split-thickness skin graft on the subgaleal flap, the new canal and the conchal cavity are filled with Gelfoam for stabilization. This kind of reconstruction also lacks mechanical stability and leads very often to restenosis.

With the use of computer-assisted surgery and facial nerve monitoring in atresia surgery, the risk of injury to the facial nerve is reduced. In a retrospective study, Caversaccio et al. [15] compared intra- and postoperative clinical and audiological findings of atresia surgery using computer-assisted surgery with similar interventions that were applied without computer-assisted surgery. Computer-assisted surgery in congenital bony aural atresia, which is combined with altered petrous bone anatomy and scarcity of surgical landmarks, provides the surgeon with increased safety and accuracy in critical situations. This fact reduced the mean operating time in the evaluated cases by 25 min.

In combination with facial nerve monitoring, the rate of complications (e.g. dysfunction of the facial nerve) can be reduced and the new external ear canal can be maximally enlarged, which minimizes the complication of postoperative restenosis.

**Conclusion**

In the present study, a new technique of combining aesthetic and functional reconstruction has been evaluated. We were able to integrate the implantation procedure into the auricle reconstruction procedure; thus, avoiding the need for a further intervention. Functional results of implantation were satisfactory, obtaining air conduction thresholds of 30 dB or better in the majority of patients, including patients with unfavorable conditions on preoperative CT scan who were not eligible for classic reconstructive surgery. The versatile form of the FMT of the Soundbridge allows for adaptation of the coupling procedure to each individual anatomical situation. It appears to be very suitable for implantation in malformed ears. Implantation of MEI is a valuable option for functional reconstruction of the malformed ear, which may offer more consistent and reliable results than classic reconstructive surgery, and should be evaluated in larger series of patients.
References


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