Modified Surgical Techniques for Combined Congenital External and Middle Ear Malformations

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Abstract

Hypothesis: There is a positive correlation between type and intensity of combined congenital ear malformations and results of surgical reconstruction.

Background: Combined congenital malformations of external and middle ear pose numerous surgical problems and demand a high degree of surgical skill and a planned, staged therapy. The goals of these operations are: a limited number of operations with minimal complications, a good cosmetic result, hearing improvement, an easier use of hearing aids, removal of congenital cholesteatomas, and preservation of the labyrinth and facial nerve.

Methods: The aim of this report is to present the author's experience in treating patients with combined malformations of the external and middle ear. In the period between 1992 and 2001, seventeen patients with this type of congenital malformations were operated upon. The long-standing dilemma of who operates first, the plastic surgeon or the otosurgeon, is solved here by the complete otosurgical treatment with reduced number of operations.

Results: A total of 36 operations were performed on 17 patients. Staged operations were done for the indications of auricular reconstruction (14 patients), and reoperations for meatal restenosis (five patients), or cartilage graft necrosis (one patient). The number of corrective reoperations is 6 (six). Atresia was successfully treated in 14 patients, while in three cases postoperative connective tissue meatal restenosis formed. The results of reconstruction of the auricular malformation were aesthetically good in 13 patients and fair in four patients.

Conclusion: We found a positive correlation between the external ear and middle ear deformity and the results of reconstruction. Two thirds of the patients had a hearing threshold level under 30 dB, while the thresholds in other patients were 30-40 dB. A residual air-bone gap of at least 20 dB was found in all patients.
Introduction

The first written reports concerning ear malformations are found in the ancient era. However, the first confirmed description of ear malformations date from the VII century when Paulus (from Aegina) mentioned the differences between congenital and acquired forms. Treatment consisted of incision and cauterization. Scientifically based reconstruction was first tried in 1920 when Gillies used a costal cartilage graft. Different materials for restoration of delicate auricular shape were used, such as cobalt-chrome mash, acrylic, silastic and composite grafts. The reasons for the plethora of different materials and techniques are the suboptimal aesthetic results and the many operations required to achieve a good outcome when costal cartilage is used. During the last 50 years, development of tympanoplasty techniques gave rise to advances in the treatment of combined malformations of the external and middle ear. In addition, new radiologic and audiologic diagnostic methods led to improved selection and preparation of these patients for surgical reconstruction. 1,2

Embryological development of the auricle includes auricular tubercles derived from the mandibular (the first branchial arch) and hyoid arch (the second branchial arch). The external auditory canal and tympanic membrane are formed from the first branchial pouch. The first and the second branchial arches are important for the formation of the middle ear ossicles, excluding the stapedial footplate. The tympanic part of the temporal bone also calcifies postnatally creating the medial part of external meatus. Meatal atresia is caused by irregularities of this process. Cartilaginous atresia is possible, even if the tympanic ring and tympanic membrane are developed. 1,2

A number of different classifications of ear malformations that are mainly descriptive can be found in the literature. The simplest one, divides deformities into those that are big and little. Big deformities include severe microtia, underdevelopment of the meatus and tympanic membrane, reduced middle ear space, and deformed and fixed middle ear ossicles. Small deformities are characterized by moderate or little deformity of the ossicles, a normal meatus and a pneumatized middle ear space. Auricular malformations can be classified as little (Type I), vertical rudiment of auricle (Type II), and small auricular remnant (Type III). 1

Combined malformations of both structures are very important and difficult to treat. Other deformities, particularly affecting the mandible, can be also be found. Facial canal malformations, such as dehiscence or an irregular course are frequently found in the combined forms. The middle ear ossicles are often fused, with a smaller, deformed malleus and other changes. Reduced pneumatization indicates changes of other middle ear structures. As the inner ear is derived from an ectodermal otocyst, combined malformations of middle and inner ear are rare.

The aim of this work is to present our experience in the surgical treatment of congenital, mixed deformities of the external and middle ear, and to present characteristics of the specific type of reconstruction that was used.

Methods and Materials

A complete ENT examination, auditory evoked potentials and computerized tomography were routinely performed preoperatively. During the ten year period from 1992-2001, 17 patients were operated upon for congenital external and middle ear malformations. There were seven major ear malformations, six atresia and middle ear deformities, and four minor external and middle ear malformations. A total of 36 operations were performed.

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The classical surgical procedure for reconstruction of combined external and middle ear malformations has several stages. In order to improve the results of reconstruction and to avoid the dilemma of who operates first, the plastic surgeon or the otosurgeon, we modified the timing, order and type of surgical interventions. In the first stage, reconstruction of the atresia and middle ear space, with implantation of cartilaginous transplant is performed. Costal rib cartilage is shaped according to a prepared model. This operative stage is the longest, and the most complex. It is performed completely by the otosurgeon. In the next phase, the auricle is elevated, and in final stage, precise reconstruction of certain auricular parts is performed (Table 1 and 2).

Table 1. Integrated Auricular Reconstruction Protocol (IARP), Aguilar EF (1996)

<table>
<thead>
<tr>
<th>STAGE AND TYPE OF OPERATION</th>
<th>SURGEON</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stage I Harvesting and placement of cartilaginous frame</td>
<td>Plastic surgeon</td>
</tr>
<tr>
<td>Stage II Creation of lobule</td>
<td>Plastic surgeon</td>
</tr>
<tr>
<td>Stage III Atresia repair</td>
<td>Oto surgeon</td>
</tr>
<tr>
<td>Stage IV Creation of tragus</td>
<td>Plastic surgeon</td>
</tr>
<tr>
<td>Stage V Elevation of auricle</td>
<td>Plastic surgeon</td>
</tr>
</tbody>
</table>

Table 2. Modified surgical technique for atresia and microtia repair

<table>
<thead>
<tr>
<th>STAGE AND TYPE OF OPERATION</th>
<th>SURGEON</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stage I Harvesting and placement of cartilaginous frame, Atresia repair</td>
<td>Oto surgeon</td>
</tr>
<tr>
<td>Stage II Creation of lobule and tragus</td>
<td>Oto surgeon</td>
</tr>
<tr>
<td>Stage III Elevation of auricle, minor corrections</td>
<td>Oto surgeon</td>
</tr>
</tbody>
</table>

For the reconstruction of meatal atresia and the middle ear space, an anterior incision is used with elevation of soft tissue in two layers. The skin incision should never be placed over the cartilaginous transplant. The atretic plate is visualized and completely removed with a burr. Any malformed ossicles which are found are inspected and their mobility, shape and relation with other structures are analyzed. After surgical enlargement of the external meatus and middle ear space, an adequately large temporalis fascial graft is placed over the ossicular system and borders of the newly created anulus. A meatoplasty with resection of soft tissues and formation of a new ear canal with a 50% bigger diameter than normal are the next steps. Proper position of the auricle and cartilaginous canal is assured.

Reoperations were performed for staged reconstruction of the auricle, or for correction of a suboptimal anatomic or functional outcome. Aesthetic and audiologic postoperative results were analyzed for at least six months.
RESULTS: The applied surgical technique resulted in adequate diameter of the meatus in 61.5%, while in 38.5% restenosis was found. Reoperations were performed in cases of restenosis, and only in one case (7.7%) the surgical result was insufficient. Postoperatively, in four patients, infection of the skin occurred, but was successfully treated with parenteral and local antibiotics (Table 3, 4 and 5).

Table 3. Type of deformity and number of performed operations

<table>
<thead>
<tr>
<th>MIDDLE EAR</th>
<th>CANAL</th>
<th>AURICLE</th>
<th>NUMBER patients</th>
<th>operations</th>
</tr>
</thead>
<tbody>
<tr>
<td>Big deformity</td>
<td>Atresia</td>
<td>Vertical rudiment</td>
<td>7</td>
<td>24</td>
</tr>
<tr>
<td>Small deformity</td>
<td>Atresia</td>
<td>Moderate deformity</td>
<td>6</td>
<td>8</td>
</tr>
<tr>
<td>Small deformity</td>
<td>Atresia</td>
<td>Normal</td>
<td>4</td>
<td>4</td>
</tr>
</tbody>
</table>

Table 4. Type and number of performed operations

<table>
<thead>
<tr>
<th>TYPE OF OPERATION</th>
<th>NUMBER</th>
</tr>
</thead>
<tbody>
<tr>
<td>Combined reconstruction of middle and external ear with harvesting of costal cartilage</td>
<td>7</td>
</tr>
<tr>
<td>Combined reconstruction of middle ear without cartilage graft</td>
<td>10</td>
</tr>
<tr>
<td>Stage II of operation</td>
<td>7</td>
</tr>
<tr>
<td>Stage III of operation</td>
<td>7</td>
</tr>
<tr>
<td>Reoperation</td>
<td>5</td>
</tr>
</tbody>
</table>

Table 5. Results of reconstruction of combined external and middle ear malformations
In all cases with major malformations (microtia gradus III, meatal atresia and middle ear malformation) we found fusion of the malleus and incus with their fixation to atretic plates. The stapes was unchanged, but with significantly reduced visibility during surgery. Removal of the atretic plate enables ossicular mobility and adequate function. In all the cases, a mobile ossicular chain was achieved (Figure 1-5.).

Figure 1. CT of congenital atresia: (Left atretic plate, Right well-developed middle ear space)

Figure 2. Reconstruction of atresia and minor auricular malformation
Figure 3. Atresia repair.

Figure 4. Reconstruction of the atresia and major auricular malformation using cartilage transplant

Figure 5. Intraoperative view on malformed ossicles (M-malleus, I-incus, S-stapes) and atretic plate (AP)
In one case, postoperative partial facial palsy was noticed, but the resolution was spontaneous. Sensorineural hearing loss and postoperative otorrhea were universally absent.

The degree of external ear deformity correlated with the deformity of the middle ear and results of operation. In the cases of major auricular deformities big malformations of ossicles, reduced pneumatization and facial malformations were always present. Audiological results were also worse.

Postoperative audiometry showed hearing thresholds under 30 dB in 66.9% of patients, while in the others it was 30-40 dB. A residual air-bone gap of at least 20 dB was found in all patients (Figure 6).

Figure 6a and 6b. Changes in postoperative hearing as compared to preoperative values, categorized using the Jahrsdoerfer classification. (ABG = air bone gap, PTA = pure-tone average, and hearing gain for different frequencies in kHz).

**Discussion**

Major congenital malformations of the external ear have a significant impact on psychosocial development. They are frequently combined with middle ear malformations and when bilateral they cause hearing loss and defects in speech development. Major malformations demand early evaluation of the patient and a staged reconstruction.
Because of possible congenital cholesteatoma, it is not generally accepted that only bilateral cases should be operated upon. The patients and parents opinion should be carefully taken into account. The ideal condition for operation is absence of facial deformities and an adequate middle ear space. For bilateral atresia, bone-anchored hearing aids are used starting from the 4th month, and operation on the first ear is recommended in the 4th year.

The most frequent occurrence is that the patient approaches the plastic surgeon and otosurgeon. Each surgeon wants to do his part of the operation first, and are usually concerned only for their part of the work.3–5 The objective of reconstruction of the malformed auricle is obtaining a sufficient aesthetic result with maximally reducing the number of operations and avoidance of possible complications. Before the operation, the parents should receive precise information concerning the type and possible results of the planned surgical interventions.

Proper selection of patients with combined external and middle ear malformations is necessary. Audiometry and auditory evoked potentials are needed for the determination of the status of conductive and sensorineural components of the hearing loss and to verify inner ear function needed for surgical reconstruction. Computerized tomography of the temporal bone is performed before the operation in order to define the shape of the cochlear and vestibular labyrinth, size of middle ear space, condition of the middle ear ossicles and the course of the facial nerve. This is especially important when there is suspicion of a congenital cholesteatoma. If unilateral atresia is found together with an extremely reduced pneumatic spaces (50% smaller than the contralateral normal side) that has no visible ossicles, the operation is not recommended because of a poor chance of success. Besides this, cochlear hypoplasia with an enlarged vestibule and lateral semicircular canal indicates a possible pathologic communication of perilymph and CSF.3,6

Congenital aural atresia is a complex problem demanding a multidisciplinary professional team in order to obtain acceptable results. A systematic approach from the beginning to the end is required. Knowledge of the anatomical characteristics of the temporal bone and the possible types of malformations affecting delicate structures is needed. Creation of a proper external meatus, aerated middle ear space with functioning conductive system, preservation of facial nerve and inner ear are the primary surgical goals.2,6

The surgical approach for the repair of atresia can be transmastoid or anterior. In the mastoid approach, the sinodural angle is identified first, than the antrum, facial recess and ossicular chain, with final removal of the atretic plate. The anterior route involves removal of the bone between the temporomandibular joint and the middle cranial fossa. The surgical work in the mastoid is reduced. This approach creates a smaller operative cavity, and thus prevents accumulation of keratin and reduces the need for chronic cleaning after the operation. In addition, manipulation around the facial nerve is significantly avoided. In cases of more severe ossicular deformities, autotransplant of cortical bone, or other reconstructive materials may be required.8

The reporting of audiologic results of combined reconstruction are not standardized in the literature. The success depends on the degree of deformity of the external and middle ear.

Potential complications of staged reconstruction should not be overlooked. They include: Skin necrosis over the cartilaginous transplant, infection of cartilage, necrosis of the lobule, hypertrophic scar formation and meatal restenosis. Meatal restenosis is the most frequent postoperative complication.
Conclusion

Congenital combined malformations of the external and middle ear have important psychological consequences on individual development. When bilateral, abnormalities of the middle ear causes a conductive hearing loss and delayed speech development. Early diagnosis using auditory evoked potentials and computerized tomography is needed for the subsequent surgical reconstruction. Proper selection of patients can result in hearing thresholds up to 30 dB in over two thirds of the patients. Modified surgical reconstruction with a reduced number of operations and one surgeon performing all the interventions gave good aesthetic and functional results with a very low complication rate.

References


