Chapter 151: Developmental Abnormalities of the Ear

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The pinna, external auditory canal, tympanic membrane, middle ear cleft, ossicles, and bony and membranous labyrinth provide a continuity of anatomic and functional pathways for the introduction of sound into the central nervous system. Developmental abnormalities of these structures impair hearing and frequently present an abnormal appearance as well.

The various congenital abnormalities of the pinna are considered elsewhere. One group of such anomalies, the microtias, occurs in mild to severe forms and is almost always associated with abnormalities of the external auditory canal, either atresia or, less commonly, stenosis. This chapter focuses on abnormalities of the external auditory canal, their relationship to the other ear structures, and their treatment.

External auditory canal atresia was described by the Mesopotamians around 2000 BC (Ballantyne, 1894), and several cases have also been documented from the New World (Hodges et al, 1990).

Congenital auditory canal atresia is a unilateral problem in approximately 70% of cases (De La Cruz et al, 1985), and when unilateral, it affects the right ear more frequently than the left (Jafek et al, 1975). Approximately 60% of cases affect males (De La Cruz, 1985; Jafek et al, 1975). Bilateral cases may be either symmetric total atresia or unilateral atresia with contralateral stenosis.

Embryology

To understand the myriad of deformities that can occur with congenital atresia, understanding the embryology is necessary. Of primary concern are defects of the middle ear and external ear that develop from the first branchial cleft and pouch.

The external ear develops from the upper end of the first and second branchial arches and from the upper end of the fist branchial cleft. In the sixth intrauterine week the auricle develops from six knoblike hillocks situated around the primitive meatus. The hillock destined to become the tragus is derived from the second branchial arch. The other five hillocks arise from the first branchial arch. By the end of the third month a primitive auricle has completed its development.

During the second month a solid core of epithelium migrates inward from this rudimentary pinna toward the first branchial pouch. This core of epithelium is the precursor of the external canal. The first branchial pouch grows outward to form the middle ear cleft. The plaque of tissue formed where these two meet ultimately forms the eardrum. At the same time the malleus and incus are taking shape from the upper end of Meckel's cartilage (the first branchial arch); the stapes forms from the upper end of Reichert's cartilage (the second branchial arch). All the ossicles attain their final shape by the end of the fourth month. By the end of the seventh to eight month, the expanding middle ear cleft surrounds the ossicles and covers them with a mucous membrane. The external auditory canal starts to hollow out and takes it shape during the sixth month. Its medial end starts to canalize by the end of the

seventh month. As this canalization takes place, the developing mastoid becomes separated from the mandible, causing growth of the mastoid to take place posteriorly and inferiorly. This posterior and inferior development carries the middle ear and facial nerve to its normal position (Figs. 151-1 and 151-2). Development may be arrested at any point from the very beginning of tympanic bone development deep over the middle ear (Figs. 151-3 and 151-4) to a stage in which the canalization extends completely to the outer meatus (Fig. 151-5). The possible range of development, then, is from a complete lack of tympanic bone to the normal state.

Classification Systems

Numerous classification systems have been described for congenital ear malformations, some were based on external ear configurations and others on surgical or pathologic findings. All suffer the disadvantage of being too simplistic for what is a complex expression of many variables that may be involved and are relevant considerations for surgical treatment. Among others, these variables include external ear configuration, type of atresia (atresia or stenosis), presence or absence of a tympanic membrane or external canal cholesteatoma, location of facial nerve, degree of mastoid pneumatization, type and extent of ossicular abnormalities, bony labyrinthine abnormalities, and associated congenital malformations.

A practical system that is currently used by many authors is Colman's modification (1971) of Altman's classification (1955), which is based on external auditory canal patency, middle ear pneumatization, and degree of difficulty of surgical procedures. Abnormalities are divided into three groups: mild, moderate, and severe.

Group I: mild aplasia. A meatus and drum exist but are often abnormally small. The ossicles are usually fixed, and the stapes is usually abnormal. Surgery is relatively simple; transcanal surgery can be performed, usually with good results.

Group 2: moderate aplasia. The pinna is severely deformed, the external auditory canal is absent, and numerous middle ear anomalies exist.

Group 3: severe aplasia. The external canal is completely absent, pneumatization is poor, and cochlear anomalies often exist.

Surgery for group 1 abnormalities is much less complicated than for groups 2 and 3 since the location of the external auditory canal is already established, no skin grafting is necessary, and a tympanic membrane is already present. However, an abnormal location for the facial nerve is much more common in group 1 patients than in the general population and should be assumed to be present. There is also an increased likelihood for abnormalities of the ossicles and inner ear structures in patients with stenosis (especially if the opposite ear is atretic). Nonetheless, surgical results for these patients are superior to those for groups 2 and 3, and this chapter focuses primarily on the more complex aplasia and reflects the senior author's experience with over 250 cases.

Anatomic Relationships and Problems Encountered in Major Aplasias

External auditory canal and middle ear

An absent or underdeveloped tympanic bone maximizes the anatomic distortion expected in the relationships of the middle ear position, access to and exposure of the middle ear and oval window, the course of the facial nerve, placement of the external meatus, and grafting of the tympanic membrane. Mastoid development and any residual of the tympanic bone become extremely important. Since the condyle of the mandibule articulates directly on the anterior surface of the mastoid bone, poor mastoid development or complete lack of tympanic bone prohibits access to the middle ear. Any reconstructed ear canal is bounded by the middle fossa dura superiorly, the sigmoid sinus posteriorly and inferiorly, and the temporomandibular joint anteriorly. In a nonpneumatized mastoid the room is inadequate for establishment of an external canal. These findings have most often been noted in combination with other anomalies, such as Treacher Collins syndrome or other severe maxillofacial dystocias.

The developing tympanic bone separates the mastoid from the temporomandibular joint in an anteroposterior direction. When the tympanic bone does not develop, the middle ear is tucked in medial and posterior to the glenoid fossa. This situation limits access to the entire anterior half of the middle ear.

Since the external canal and meatus must angle around the glenoid fossa to approach the middle ear, and the pinna or remnant is usually set low, the site of the new meatus must usually be moved posteriorly and superiorly. This movement of the pinna in reconstruction causes the natural scarring and healing process to place stress in an anteroposterior direction, causing narrowing or collapse of the meatus in that direction.

Facial nerve abnormalities

As the tympanic bone develops, it forces the facial nerve (CN VII) to grow inferiorly (accounting for the long descending portion of the facial nerve) and to exit beneath the tympanic bone before coursing anteriorly and superiorly to bifurcate in the parotid (Fig. 151-6). Since CN VII can cross the mandible at its ramus (rather than at the head of the condyle), it can assume a more gradual medial-to-peripheral plane. If the tympanic bone does not develop, CN VII may abruptly turn anteriorly at the descending genu and cross the posterior and inferior aspect of the middle ear (Fig. 151-7). Visualization of the round window may be impossible if it lies beneath the facial nerve, and visualization of the oval window may be extremely limited because it lies within this acute angle (Fig. 151-8). Lack of a bony fallopian canal with an exposed facial nerve may further limit the maneuverability around the stapes.

Anomalies of the facial nerve in the middle ear occurred in approximately 20% of these cases. The following abnormalities were observed in this series and are listed in decreasing order of frequency: lack of a bony fallopian canal covering the facial nerve in the middle ear, bulging of the nerve over the oval window, the nerve running inferior to the oval window, a bifid facial nerve, and a facial nerve having the appearance of the chorda tympani.

Ossicular abnormalities

The long process of the malleus is frequently curved inward, stubby and shortened, attached to the anterior canal wall by fibrous tissue or bone, or absent entirely. The short process is often larger than normal and fused to the atresia plate. Frequently the malleus and incus are not fully developed and may be a fused mass of bone.

Stapes abnormalities are also frequently present. A unipolar stapes, a stapes with a misplaced head, and absence of the crura have all been noted. Lack of development of the stapes footplate is not unusual. Any of these abnormalities means that some sort of ossicular reconstruction must be undertaken. Although major aplasias are usually recognized in the first few weeks or months life, there is a gradation of deformities, and many children with minor aplasia are not diagnosed until they fail a screening test on preschool evaluation.

Associated malformations of inner ear

Although the inner ear develops separately and at a different time from the middle ear and external ear canal, inner ear abnormalities occur rather commonly in association with congenital atresia. Reports in the literature vary, saying that radiographic techniques or surgery can identify abnormalities of the cochlea and semicircular canal coexisting with external canal atresia in 15% to 20% of cases. Fortunately the incidence of sensorineural hearing loss in association with these abnormalities does not approach these percentages. In our own series some degree of sensorineural loss is associated with about 6% of patients undergoing surgery for atresia.

Evaluation of Patient

Because microtia and external auditory canal atresia are very visible abnormalities, physicians are consulted within the first few weeks of an affected child's life by parents who may not yet have accepted that they have a child with a congenital abnormality. Parental support and reassurance are as important as a knowledgeable explanation of the plan of action to investigate the problem at hand.

When other congenital abnormalities are also present, a complex sorting of priorities of evaluation and treatment is necessary by a team of involved experts from many disciplines. This is frequently accomplished by a craniofacial anomalies clinic or similar team.

In the more common situation the physical abnormalities are restricted to the shape of the pinna and the absence of an auditory canal. The abnormal cosmetic appearance is immediately apparent to the parents, and they quickly and vigorously pursue the road to the best possible cosmetic appearance for the child's ear or ears. An understanding of the magnitude of the consequences of aural atresia and the maximal conductive hearing loss it causes is more difficult for parents to grasp. Finally, the parents must be made aware of not only the obvious cosmetic problem and the deleterious effects of bilateral aural atresia on hearing and language development, but also (1) the potential medical hazards associated with aural atresia such as otitis media and cholesteatoma and (2) all the complications that can develop from those processes when they are obscured by a stenotic or atretic external auditory canal.

At the first visit the parents should be informed that by far the most important initial aspect of management is an immediate assessment of hearing capability in both ears (whether one or both ears are affected). After determining that auditory function is present, immediate bone conduction amplification is imperative, as is the use of language stimulation to facilitate optimum language acquisition. The education of the parents in the first several visits should establish (1) the need for accurate hearing evaluation, (2) the consequences of this type of hearing loss, and (3) the mechanisms (amplification and stimulation of language) to minimize those consequences. Questions about surgery for either microtia or hearing restoration are downplayed until the importance of the foregoing is understood completely.

History and physical examination

A thorough history of the pregnancy and delivery should be obtained. One should question the parent about the presence of illnesses during the pregnancy, drugs that were used, and any history of family members with ear problems. If other family members have had aplasias or if other abnormalities are evident, genetic counseling is advisable.

On physical examination one should note whether a major deformity of the pinna exists, since counseling on plastic reconstruction may be necessary. One should also note the type of meatus and canal remnant that may exist; a rounded blunt meatal opening usually means there is no tympanic bone. A small pinpoint opening or fistula may mean a partially developed tympanic bone. In this case the opening may be prone to infection, and trapped epithelium may develop into a cholesteatoma. One should palpate the temporomandibular joint area for location of the new meatus. One should also note the condyle, and mastoid tip development. In older children, tip development usually means the mastoid will be well pneumatized.

Audiometric evaluation

Modern audiometric techniques and equipment have proven quite helpful in the management of congenital ear malformations. The auditory brain stem response (ABR) is used routinely. This technique can be performed with the child asleep or under sedation in the first few weeks of life. In unilateral cases one can measure the cochlear response of the apparently normal ear.

In bilateral cases one may not be able to obtain responses to air-conducted stimuli, but bone-conducted stimuli can usually evoke easily discernible responses. These may represent either neural activity from both cochleas or only from the better hearing ear. To determine if only one cochlea has hearing (for examplem, when one ear is anacusic), multiple recording electrodes are used while bone-conducted stimuli are delivered first to one mastoid and then to the other. Normally, with unilateral stimulation and simultaneous bilateral recording, an ipsilateral wave I is evident along with the waves that normally follow it, whereas the contralateral side has no wave I but only the later waves. Therefore stimulation of the mastoid of an intact cochlea evokes an ipsilateral wave I, and stimulation of the mastoid of a non-hearing ear does not (Jahrsdorfer et al, 1985).

If necessary, ABR can be done with the child under general anesthesia. These findings allow the child to be fitted with a hearing aid without the necessity of radiologic examination.

In unilateral atresias one can also use ipsilateral acoustic reflexes to help to determine if hearing and reflexes are normal in the unaffected ear. These examinations should be performed in the first few months of life. Routine air and bone conduction can usually be obtained with play audiometry by the age of 2.5 to 3 years or even earlier.

Radiographic examination

In general, radiographic examination is delayed until around 4 years of age and may require general anesthesia for the immobility necessary to accomplish an adequate examination. Axial and coronal plane high-resolution computed tomography (CT) scans at 1 to 1.5 mm slice thicknesses in the bone review algorithm are optimum.

When there is risk of external canal cholesteatoma (for instance, with a pinpoint canal) and symptoms suggest repeated infections, CT should be done earlier, as it should when there are suggestions that otitis media in the atretic or stenotic ear may have caused complications such as facial paralysis, meningitis, or suppurative labyrinthitis. However, in the great majority of cases, there is no need to obtain the radiographic examination until 4 years of age or later.

High-resolution CT scanning is essential as an adjunct for possible surgical intervention. It can provide information about thickness and character of the atresia plate, degree of pneumatization, size of the middle ear cleft, course of the facial nerve, state of development of the three ossicles, amount of room between the sigmoid sinus and the mandibular condyle, extent of tympanic bone development, and whether the air cell system contains air. Close coordination between the radiologist and the otologist makes the interpretation of the images more meaningful and useful.

Management Before Surgical Reconstruction

The primary management consideration is to establish satisfactory hearing that allows normal speech and language development to occur. With unilateral atresia, one must establish the existence of normal hearing levels in the apparently normal ear. When this has been determined, one should reassure the parents that normal speech and language should develop, and the child should be observed at regular intervals to make certain this is occurring. In addition, as he is older, pure-tone levels can be established through conventional audiometry.

In bilateral atresia, after one has established the cochlear reserve, a hearing aid should be fitted as soon as is feasible. If no canal remnant exists, this is usually a bone-conduction aid. Fitting can be accomplished as early as 1 month of age. The bone conduction oscillator can be coupled to either a body style hearing aid or, more commonly, to a behind-the-ear style. In very young infant, bone remodeling may occur around the site of the oscillator, and therefore side-to-side adjustments in its position are advisable to prevent permanent unsightly indentation. Initially, hearing aid use may be limited to short periods of time during communication routines. The audiologist will work with the parents to select appropriate headbands or bonnets so that the oscillator can be placed and maintained in the desired

position.

When the child reaches 8 to 10 months of age, behavioral audiometry can be initiated to evaluate the child's hearing sensitivity and the benefit obtained from the bone conduction hearing aid. In the interim, use of the hearing aid will encourage the child's speech and language development. Periodic medical observations are continued to be sure no complications of the atresia are developing.

Surgical Correction

General considerations

With unilateral atresia, many surgeons do not operate until the patient is an adult and expresses a desire for surgery, and even then, the surgery is elective. There are several considerations to support this approach. Since completely normal hearing is rarely obtained, the hearing improvement noticeable to the child is not dramatic. After surgery, some of the ears require cleaning, frequent follow-up visits, and the avoidance of water after surgery. Occasionally more than one operation is necessary, and rarely a complication may occur. However, on the other hand, improvements in CT have allowed better selection of cases, lowering the risk for complications or repeat surgery, and most patients are able to withstand water contamination of the reconstructed external auditory canal without problems.

With bilateral atresia, most surgeons recommend operating on the second side 1 year after successfully constructing an external auditory canal on the first. The restoration of bilaterally symmetric hearing is very satisfying to patients, who appreciate the change after the second ear almost as much as that after the first.

Timing

With the impressive improvements in the appearance of reconstructed auricles, most families desire surgery to improve both appearance of the auricle and hearing. There is a long history of disagreement regarding timing of these two aims, since one may compromise the other, and multiple operations are necessary. However, some situations seem straightforward.

Surgery for correction of the atresia alone should be first when medically necessary, that is, when otitis media with a complication is present behind the atresia plate or when a cholesteatoma is present. The otologist in these rare cases should consult the surgeon involved with auricular reconstruction before surgery in order to use an incision that will have the least chance of jeopardizing future reconstructive surgery.

Reconstruction of the pinna should precede that for the atresia in unilateral cases. In some cases, cooperating surgeons can combine the atresia portion with one of the stages of pinna reconstruction. Even when the operations are sequential and not combined, communication and cooperation between the two surgeons are appropriate for the most advantageous placement of incisions.

In the great majority of the bilateral cases, auricular reconstruction should precede or coincide with one of the stages of atresia surgery and usually takes place at around 6 years of age with stages 3 months apart. Both pinna and atresia reconstruction should be completed in one ear before starting the second. Size of the particular child will affect choice of time for surgery.

Occasionally, educational development is suboptimal despite good resources and efforts in a child with bilateral microtia and atresia who has ears that are very favorable for restoration of hearing. It may be appropriate to proceed with atresia surgery first at 4 to 5 years of age, with the consultation of the plastic surgeon and parental understanding of the risks as well as the possible benefits.

Obviously, when atresia repair for hearing improvement is not possible, no conflict exists.

Approach

A postauricular approach is used, keeping the incision well away from the glenoid fossa and mastoid tip. (In a primary case with a minimally deformed pinna, this is about 1.5 cm behind the postauricular fold.) The temporalis fascia and linea temporalis are identified, and the soft tissue and cartilaginous remnants of the pinna are closely undermined anteriorly. The periosteum is reflected over the mastoid, and self-retaining retractors are placed. Any tympanic bone remnant visible at this junction of the glenoid fossa and mastoid is identified. The mastoid cortex and cells are then removed from the peripheral area, closely hugging the tegment tympani or the middle fossa dura down to the area of the mastoid antrum. At this point, identifying the horizontal canal or the ossicular remnants in the fossa incudis is usually easy.

Care should be taken not to touch the ossiclesor any remnant of tympanic bone with the cutting burr, since this may transmit the high-speed vibrations into the inner ear and damage the cochlea. The facial nerve and ossicular chain are identified, and the incudostapedial joint is separated (to protect the inner ear against noise trauma). The remainder of the bone is then removed over the temporomandibular joint, and the remainder of tympanic bone or tympanic plate is removed down to the level of the ossicles.

The middle ear is commonly found medial to the temporomandibular joint, which makes it impossible to retain the ossicular chain, since the malleus remnant is almost touching the glenoid fossa. As much bone as possible is removed over the joint and around the facial nerve to expose the maximum amount of middle ear. Frequently the posterosuperior portion of the middle ear is all that can be exposed. A bony shelf of new annular ledge should be fashioned anteriorly and inferiorly, and the graft can then be tucked between the mucosa and annular ledge to anchor it anteriorly.

Controversy exist as to whether the entire mastoid should be exenterated or whether normal residual cells can be left. The obvious advantages of a smaller bony canal are that it results in less area to clean and debride, does not leave an unsightly size to the external canal, and heals more rapidly. The disadvantages are that the surgeon must work through a more restricted space and encounters more difficulty in identifying structures such as facial nerve

and ossicles in approaching the middle ear. Also, a graft over residual cells may develop retractions or chocolate cysts in the cells. In most cases, mastoidectomy is not necessary.

Repositioning ossicles or using cartilage to assist in rebuilding the ossicular chain is usually necessary. The use of tissue glue is extremely helpful in maintaining the integrity of the chain. A fascial graft obtained from the temporalis muscle is then used to graft the area of the middle ear. The graft should not be brought out the anterior canal wall but should be tucked beneath the new annular sulcus and draped posteriorly over the middle ear area.

The use of split-thickness skin to line the entire reconstructed external canal leads to rapid epithelialization and lack of reaction and helps to some degree in maintaining the patency of an ear canal. Rapid attachment of the skin anteriorly at the junction with the graft helps to prevent graft migration. The central portion of the tympanic membrane graft should not be covered with this skin, so that a thin layer of epithelium can be allowed to migrate over the fascia graft, as is done in lateral grafting techniques. All cartilage and redundant soft tissue should be trimmed away from the meatal opening. This allows the skin to attach to the bony meatus and helps prevent soft tissue stenosis of the meatus. Expandable cellulose wicks are used to hold the graft in position during the immediate postoperative period.

Complications

Infection and cholesteatoma

Mastoid and middle ear infections masked by an atretic canal are of concern, because the pathways of least resistance to the infection, the tympanic membrane and external auditory canal, are blocked by an unyielding atresia plate, which prevents egress of any infectious material under pressure. This can increase the likelihood of other complications from the pus under pressure. We have seen patients with serous otitis media, meningitis, and facial paralysis from tympanomastoid suppuration behind an atresia plate. One of us (James Crabtree) has explored two cases of facial paralysis secondary to acute or subacute mastoiditis with rapid recovery of the facial paralysis in both.

Cholesteatomas may develop in a partially formed external canal. Epithelium may be trapped behind an atretic or stenotic meatus and gradually enlarge until it breaks into the mastoid or middle ear and becomes symptomatic. In most cases these are manifested by a small pinpoint atretic canal, with intermittent drainage or with an associated abscess in the area of the meatus. We consider these to be acquired cholesteatomas, although technically one could consider them to be primary cholesteatomas. This process occurs in approximately 5% of operated ears.

Infection in the reconstructed ear may occur as acute otitis media or as serous otitis media. The infection should be treated as in any normal or reconstructed ear; occasionally putting a tube through the graft is necessary to ventilate the ear after an acute inflammatory reaction. Occasionally the graft breaks down over a cell, allowing an opening into the mastoid and serous or mucoid drainage. This may require revision if it does not respond to antibiotic therapy.

Stenosis

All authors mention the problems of restenosis. Some quote an incidence of restenosis following reconstruction as high as 40%; others state that this is a very minor problem. The use of a large meatus, creation of a large cavity, removal of all redundant tissue and cartilage around the meatal opening, and tight closure of the meatal opening with skin grafts all reduce the likelihood of restenosis. Several authors have suggested that the incidence of restenosis is inversely related to the experience of the surgeon. Restenosis with bone, however, does occur in a certain percentage of cases and is much more likely to occur in the group 3 solid or poorly pneumatized mastoids.

Any method of reducing postoperative infection or reaction is of benefit. In children who have a tendency for keloid or scar formation, triamcinolone injections are beneficial. Most authors no longer stent the immediate surgical reconstruction because most stents cause irritation and reaction with additional fibrosis. The use of soft stents with the injection of triamcinolone may, however, be advantageous in the meatus that is beginning to stenose.

Hearing loss

Sensorineural hearing loss can occur if one is drilling away the tympanic bone without first disarticulating the ossicles. Every precaution should be taken not to touch the intact chain with the burr because it may result in damage to the high-frequency responses.

Conductive hearing loss may occur after reconstruction if a repositioned ossicle is touching any part of the bony fallopian canal or the bony walls of the newly constructed canal. Bony fixation has been reported in all tympanoplastic techniques. A graft may not attach well at the newly created anulus and may migrate externally. The possibility of this happening can be reduced by tucking the graft anteriorly beneath the mucosa of the newly formed annular ledge and by allowing the graft to retract posteriorly into the newly created mastoid cavity.

Facial nerve injury

Improvements in CT scanning using both axial and coronal views have allowed much better preoperative assessment of the course of the fallopian canal, and, armed with this knowledge, the surgeon can anticipate how much bone can be safely removed in order to create adequate space for a tympanic membrane and an external auditory canal. During surgery, use of a facial nerve monitor can give warning that the nerve is at hand before any damage has occurred. Using dilute epinephrine solution without any lidocaine when injecting for hemostasis eliminates the possibility of accidently anesthetizing the facial nerve so it cannot respond to stimuli. Younger surgeons particularly may place too much emphasis on the newer technology.

One must also remember that the nerve is usually superior and external to the normal course outside of the temporal bone, as previously described. In raising flaps or moving the external ear, one must therefore be very cautious to keep from injuring the facial nerve by the use of retractors or cauterization.

Hearing Results

Most group 1 atresias give good results. Few problems with graft migration or stenosis occur. Hearing results usually depend on some type of ossicular reconstruction in the middle ear and 80% to 90% of the results are within 20 to 25 dB of the preoperative bone-conduction thresholds.

Group 2 and 3 atresias, however, have successful long-term hearing results in only 65% to 70% of the cases. Despite initial hearing improvement, about 10% of these regress over a period of 1 to 2 years.

A smaller number (6%) of these cases have abnormal bone-conduction thresholds and therefore may not reach serviceable hearing. If, however, the patient has a successfully reconstructed external auditory canal, he or she can frequently go from a bone-conduction aid to an air-conduction aid, which is much more acceptable. A revision operation is necessary in approximately 30% of the reconstructed ears because of recurrent stenosis, graft migration, or the lack of adequate hearing improvement.

Summary

In summary, reconstruction of congenital atresia is both difficult and intriguing for the surgeon. Restoring a child's hearing is one of the most gratifying of all experiences in otologic surgery.