Surgery for Congenital Ear Malformations

Robert A. Jahrsdoerfer and Bradley W. Kesser

Classification

Congenital malformations of the ear can be broadly classified into two categories:

- **Minor malformations**  Problem is limited to the middle ear
- **Major malformations**  Atresia or stenosis of the external ear canal. Congenital aural atresia means the external ear canal has failed to develop. Congenital stenosis refers to partial development of the external ear canal. Both are usually associated with microtia, although on occasion the external ear may be well formed.

Hearing Evaluation

- Auditory brainstem response (ABR) testing is mandated in a newborn with microtia/atresia. Otoacoustic emissions may be of use in unilateral atresia if the contralateral ear is normal.
- The side with the better cochlear reserve is operated upon to achieve the best postoperative hearing result.
- If there is any doubt concerning hearing, a bone conduction hearing aid should be placed as early as possible.

Preoperative Considerations

Timing of Surgery

- Once a child with unilateral or bilateral atresia has been determined to have one normal hearing ear or has been aided for hearing loss, he or she is allowed to
mature until 5 years of age. At that time, consultation should be had with a plastic surgeon and an otologic surgeon.

Prior to age 5 there is often a degree of immaturity that makes routine postoperative care problematical, often requiring the use of sedation or forcible restraint.

Preoperative Testing

- Audiologic testing should be repeated prior to surgery.
- A high-resolution computed tomography (CT) scan of the temporal bone in the 30-degree axial and 105-degree coronal planes should be obtained. The appearance of the CT scan will be the primary determinant of the patient’s candidacy for atresia surgery. The scan is again reviewed on the day of surgery with emphasis placed on the course of the facial nerve, slope of the tegmen, and depth of the middle ear from the lateral aspect of the skull.

Special Surgical Considerations

Minor Malformations

- Minor malformations typically involve the stapes/oval window/facial nerve axis. The auricle and external ear canal are normal in appearance, or almost so, and the tympanic membrane can be identified.
- When an absent oval window is found at surgery, the choices available to the surgeon are to terminate the operation or to attempt hearing restoration by creating a vestibulotomy. If the latter approach is chosen, there should be a defined oval window area that can then be targeted for drilling, as well as mature ossicular development to enable placement of a prosthesis.
- Once a vestibulotomy is drilled, the opening should be covered with temporalis fascia or other appropriate soft tissue seal. The distance between the new oval window and the ossicle may vary greatly. A wide assortment of prosthesis lengths should be available.
- A bare facial nerve will not infrequently be found to overlie the area where the new oval window should be drilled. In malformations limited to the middle ear, it is not possible to transpose the facial nerve as it is in atresia surgery. A decision must be made to drill above or below the bare facial nerve. Again, a well-defined oval window area helps in this decision. Lacking this, drilling may begin superior to the nerve using a saucerizing technique. If the vestibule is not encountered at a depth of 2 mm, the procedure is terminated because there is a heightened risk of sensorineural hearing loss from surgical violation of the membranous labyrinth.
- If a bare facial nerve obscures the oval window superiorly, a promontorial window may be drilled according to Plester.¹ The promontorial bone should be saucerized down to an intact endosteal membrane. Disruption of the endosteal membrane carries a greater chance of nerve loss from cochlear injury. A prosthesis can be inserted between the endosteal membrane and the malleus handle, or the tympanic membrane if the handle is unfavorable.
- The most important aspect in exploring the middle ear of a patient with a conductive hearing loss is an awareness that one may be dealing with a congenital
malformation. This awareness must include the possibility of finding a bare and displaced facial nerve. Failure to recognize this possibility places the patient at a huge risk of facial nerve injury.

- Congenital primary incus fixation is rare. Fixation of the malleus in the epitympanum is not. There are two surgical methods to correct malleus head fixation:
  - Remove the malleus head and incus, and place an incus strut from the malleus handle to the stapes.
  - Drill away the bony attachment in the epitympanum, and interpose Silastic® sheeting as a barrier to bony refixation.

- Ossicular fixation may also be attributable to a malleus bar. This is a term coined by Nomura to describe a bar of bone running from the malleus neck to the posterior bony annulus. This bony bar will be ~1 mm in thickness and firmly fixes the malleus in place, producing a conductive hearing loss. The chorda tympani nerve frequently runs in a bony groove in the bar and must be considered when drilling away the bar. Freeing the ossicular chain from the malleus bar will usually correct the conductive hearing loss, but not always. If the chain remains fixed, one must search for additional sites of fixation. It is our preference that any suspected congenital middle ear malformation be approached through a postauricular incision. The ossicular anomaly, commonly the stapes/oval window/facial nerve complex, is often concealed by overhanging bone. As much as 3 to 4 mm of bony overhang may need to be drilled away to access the area of concern.

**Major Malformations (Congenital Aural Atresia)**

- Atresia repair should follow microtia repair. The external ear reconstruction should be done by a plastic surgeon or facial plastic surgeon experienced with this challenging surgery. Rib graft reconstruction is best performed when the rib cage has reached satisfactory growth to enable harvesting an amount of cartilage adequate for sculpting into an ear framework. This usually occurs between 6 and 8 years of age for those children with unilateral atresia. In bilateral atresia cases, we will encourage the reconstructive surgeon to operate earlier, thereby allowing the otologic surgeon to do the atresia repair at least in one ear prior to starting school.

- If an alloplastic implant has been chosen for the external ear reconstruction, we recommend atresia surgery before the microtia repair. The child is a noncandidate for atresia repair. Silastic or polyethylene implants may do well until they are exposed, at which time they invariably become infected and extrude. Our major concern is that the child may need revision surgery after an alloplastic implant has been placed, in which case the implant is at great risk for exposure.

- It is our preference that the reconstructive surgeon completely finish the microtia repair prior to atresia surgery. If this is not possible, then the earliest the otologic surgeon should operate is after the rib cartilage has been implanted and the lobe transposed. We prefer to wait 4 to 6 months after the plastic surgeon has finished to begin operating for atresia.

- In cases of bilateral atresia, the better appearing, or higher graded, ear should be operated first. In patients with bilateral atresia, the initial restoration of hearing is a dramatic event. In unilateral atresia with normal hearing in the contralateral ear,
restoration of hearing results in a subtle but meaningful response. Discrimination is improved immediately, and sound localization, though less predictable, usually improves over time.

◆ Surgical Technique

◆ The patient is placed on the operating room table in a slightly reverse Trendelenburg position. A swath of hair ~0.5 to 1 inch in width is shaved from above the ear. Local anesthesia is infiltrated in the postauricular crease, and electrodes are placed for facial nerve monitoring. The anesthesiologist is requested to avoid paralytic agents unless they are needed for induction.

◆ A postauricular approach is used routinely. Temporalis fascia is harvested and allowed to dry. The periosteum overlying the mastoid bone is incised and elevated. A cuff of periosteum is preserved at the level of the glenoid fossa for future use.

◆ If a tympanic bone remnant is identified, drilling should begin there. Most of the time a tympanic bone remnant is absent, and drilling therefore commences in the cribiform area.

◆ The direction of drilling should be a straight approach to the middle ear. Do not indiscriminately drill out the mastoid. The mastoid approach may soothe the anxiety of an insecure surgeon but does little good for the patient. The degree of mastoid pneumatization is noted. Atretic bone is usually found anteriorly and should be tracked medially. Do not pursue mastoid air cells, as this route will lead into the mastoid antrum far posterior to where the surgeon desires to be.

◆ At a depth of 1.5 cm, the atretic plate is usually encountered. Our definition of atretic plate is that layer of atretic bone contiguous to the middle ear. If the atretic plate has not been identified at a depth of 2 cm, the surgeon should backtrack in the dissection because the middle ear may have been bypassed.

◆ It is imperative to maintain a vigil for the facial nerve while drilling. In ~25% of patients the facial nerve will make a sharp turn at the second genu to cross the middle ear in atretic bone and exit into the glenoid fossa. What is not generally appreciated is that the nerve ascends in its course through the middle ear. The facial nerve may be 4 mm more lateral at the round window level than at the oval window. In this situation the nerve is still encased in atretic bone and is at risk of being injured by drilling before the middle ear is ever reached (Fig. 21–1).

◆ The atretic bone is thinned with a diamond bur and carefully picked away to reveal the contents of the middle ear. In complete atresia where there is no tympanic membrane remnant, the malleus handle will be absent. The ossicular mass is attached to the atretic plate at the level of the malleus neck by either bone or periosteum. If there is a bony connection between the malleus neck and the atretic plate, the ossicular mass will be firmly fixed. A periosteal attachment will allow some movement of the ossicles. A floppy ossicular chain indicates a failure of the fused incus–malleus to connect to a stapes. A firmly immobile ossicular mass indicates fixation at least at the level of the atretic plate, although the ossicles may be fixed at more than one site.

◆ The middle ear findings will include an incus–malleus complex that is usually fused, an incus long arm that is often foreshortened and sometimes vertical, an
incudostapedial joint that has failed to develop and that may form a solid bony union between the two ossicles, and a stapes of variable development. There is a 4% incidence of a fixed stapes from failure of the footplate to differentiate.

A chorda tympani nerve is seen in fewer than 50% of the cases, and when present, will typically be situated far inferior in the middle ear. In this location the chorda is difficult to save as it courses through the atretic bone, which must be drilled away to complete the operation. The facial nerve may be bare above the oval window and as previously mentioned is significantly displaced in 25% of the cases.

- Ideally, the best possible condition in which to find the ossicles is to have an intact ossicular chain, although malformed, but which moves as a unit. In ~20% of the cases, the ossicular chain will be discontinuous, requiring ossicular chain reconstruction. Total ossicular replacement prostheses (TORPs) and partial ossicular replacement prostheses (PORPs) of the surgeon’s preference are suitable in reconstructing the ossicular chain.

- Bone peripheral to the ossicular chain is drilled away to center the ossicles in the approximate middle of the new tympanic membrane. The bony bridge connecting the malleus neck to the atretic plate is saved until last. It is then thinned by drilling with a diamond bur and then carefully drilled away at slow revolutions, or vaporized with a laser. Once free, the ossicular chain is highly mobile, and the inner ear is vulnerable to injury from overzealous ossicular manipulation or damage from the drill.

- One must be diligent in removing bone dust from the middle ear, particularly from the oval window niche and the undersurface of the incus–malleus complex. Failure