Chapter 116 – Congenital Malformations of the Middle Ear

Arpita I. Mehta,
Yael Raz

The various manifestations of middle ear malformations provide an interesting clinical challenge for physicians. In the adult population undergoing exploratory tympanotomy for presumed otosclerosis, such anomalies may be unexpected. The otologic surgeon must be prepared to deal with unanticipated findings such as partial or complete absence of components of the ossicular chain, lateral chain fixation, a persistent stapedial artery, abnormal course of the facial nerve, or absence of the oval window. Knowledge of the embryologic origins of middle ear structures is useful in understanding the various anomalies that arise and anticipating associated defects. For example, branchial arch anomalies may accompany defects of the ossicular chain, which is derived in part from the first two arches. Additionally, the stapes and facial nerve develop simultaneously.[1] An inferiorly displaced facial nerve may prevent contact between the developing stapes and the inner ear, thereby inhibiting further development of the stapes and preventing proper induction of the oval window.[2] Thus, an abnormal course of the facial nerve should be suspected when anomalies of the stapes or oval window are encountered.

Middle ear malformations should always be considered in the differential diagnosis for children with conductive hearing loss. Congenital ear deformities are estimated to occur in 1 in 11,000 to 15,000 individuals.[3,4] Isolated anomalies of the middle ear with a normal auricle and external auditory canal account for only a small percentage of these deformities.[5] Approximately one quarter of congenital middle ear anomalies occur in the setting of a disorder such as Treacher Collins, Klippel-Feil, or branchio-oto-renal syndrome.[6] In an adult patient, congenital middle ear anomalies should be suspected when hearing loss has been present since childhood, thus making otosclerosis unlikely, and the history is negative for otitis media.

This chapter focuses on the operative management of isolated middle ear malformations. Management of aural atresia is addressed in Chapter 111, and microtia repair is discussed in Chapter 85.

PATIENT SELECTION

Depending on the nature of the middle ear anomaly, the resultant hearing loss can be purely conductive or mixed, unilateral or bilateral, and range from mild to profound. Whereas bilateral middle ear anomalies are often recognized in early childhood, in the past, unilateral middle ear abnormalities were frequently not identified until a school-aged child received a screening hearing test. However, with the recent adoption of universal newborn hearing screening tests by most states, the age at identification of children with developmental middle ear abnormalities can be expected to decrease significantly. Non–otitis-related unilateral conductive hearing loss should be managed conservatively until the child is at least 5 to 7 years of age. The age at which surgery is appropriate must be tailored to each patient and depends on eustachian tube function and the reliability of the results of behavioral audiometric testing. In the setting of significant bilateral hearing loss, amplification should be pursued as soon as possible. Given the significant possibility that stapedial fixation will be encountered, it is not advisable to proceed with surgery while there is a significant risk of otitis media. Stapes surgery at an early age presents an unnecessary risk of perioperative labyrinthitis with resultant sensorineural hearing loss. Surgery is not warranted unless the hearing loss is in the moderate to severe range, with a speech reception threshold greater than 35 and a pure-tone average greater than 30 dB.[7] Once the child reaches an age when hearing can be reliably tested and is beyond the time period of increased vulnerability to otitis media, stapes surgery can be performed quite safely.[8,9]

Conductive hearing loss should always be confirmed with tuning fork testing before proceeding with surgical intervention. Children who are unable to respond reliably to tuning fork testing will probably not prove reliable for behavioral air and bone conduction testing. It is safer to provide amplification until the audiometric test results are reliable. Surgery for conductive hearing loss in the setting of a normally formed ear canal is an elective procedure. The hearing loss can be effectively managed with hearing aids, and it is imperative that parents understand the alternatives to surgery before proceeding with exploration in a pediatric patient, particularly for anomalies that are associated with significant risk to the facial nerve or inner ear (i.e., congenital stapedial fixation or absence of the oval window). Some authors argue that it is not appropriate to proceed with surgical management of unilateral conductive hearing loss not attributable to otitis media until the child can participate in the informed consent process.

When stable conductive hearing loss has been documented in children, the possibility of congenital fixation of the stapes must be entertained. This disorder is caused by failure of differentiation of the lamina stapedialis into the annular ligament, which results in ankylosis of the stapes footplate. The chief risk associated with fenestration of the footplate in such cases is the occurrence of a perilymphatic gusher. Although the true incidence of a perilymphatic gusher is unknown, it has been reported to be significantly higher than the incidence seen in otosclerosis (<1%).[10] Congenital stapedial footplate fixation can occur in the setting of an X-linked familial disorder characterized by progressive mixed hearing loss associated with inner ear anomalies, including Mondini's malformations and dilatation of the internal auditory canal (Fig. 116-1). The excessive flow of perilymph that follows opening of the footplate in these children is believed to be due to either a patent cochlear aqueduct or a defect in the lateral end of a dilated internal auditory canal. Mutations in the gene encoding the POU3f4 transcription factor have been implicated in at least one form of X-linked progres-sive mixed hearing loss.[11,12] The presence of mixed loss, particularly in males, should alert the surgeon to a potential gusher. Although some surgeons have obtained successful hearing results, it is generally conceded that computed tomography (CT) evidence of a widened cochlear aqueduct or internal auditory canal should be considered a relative contraindication to surgery.

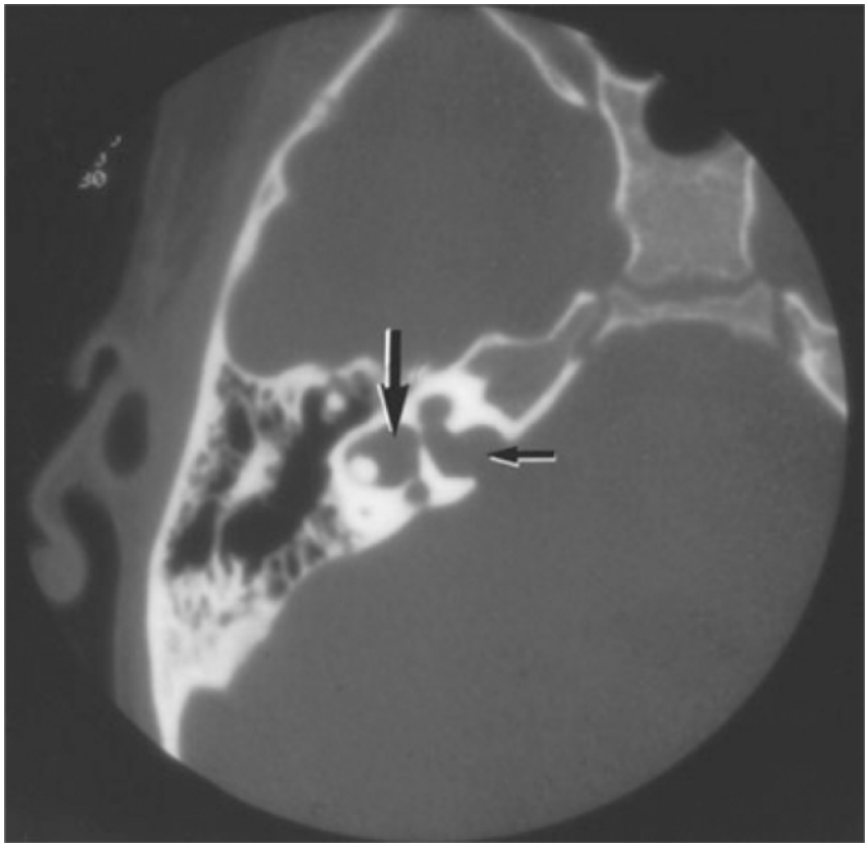


Figure 116-1 Computed tomography scan of a dysplastic inner ear showing a dilated and tortuous internal auditory canal (*small arrow,* internal auditory canal; *large arrow,* Mondini's deformity).

Superior canal dehiscence syndrome, in which the bone overlying the superior semicircular canal is absent, can be manifested as conductive hearing loss and probably accounts for a fraction of the patients whose air-bone gap fails to close after stapes surgery.[13] The conductivehearing loss persists postoperatively because it is not secondary to stapes fixation. Rather, the conductive loss is due to bony deficiency in the otic capsule and the resultant “mobile third window.” This problem can be avoided by confirming the absence of a middle ear reflex before proceeding with middle ear exploration. Ossicular fixation should result in an elevated or absent stapedial reflex, whereas in superior canal dehiscence syndrome, this reflex will be preserved. Another situation in which caution is advised is an adult patient with conductive hearing loss and a history suggestive of Meniere's disease. Stapes surgery in the setting of endolymphatic hydrops is associated with a higher risk of sensorineural hearing loss and dizziness, which may be related to the proximity of the distended saccular epithelium to the stapes footplate.

PREOPERATIVE EVALUATION

History and Physical Examination

A thorough history should be elicited from the patient or caregivers, or both. In children it is important to note the onset of symptoms to help differentiate a congenital disorder from the juvenile onset of an acquired disorder. Prenatal history, school performance, social interactions, and language development should be assessed. Previous otologic history, particularly the frequency of otitis media and requirement for pressure equalization tubes, should be assessed. The history will often be limited in a pediatric patient, but when possible, it should address otologic symptoms such as otalgia, otorrhea, tinnitus, or vertigo. A careful review of symptoms may reveal associated cardiac, renal, or other medical problems suggestive of a syndromic diagnosis. A family history of hearing loss is important to address.

Physical examination should include inspection for other developmental anomalies, particularly those originating in the first and second branchial arches. Children with disorders affecting the mandibular arch (i.e., Treacher Collins syndrome) are at risk for airway compromise, so appropriate precautions must be taken in the operating room. Identification of even minor flaws in the pinna or surrounding area, such as preauricular tags and pits, should alert the surgeon to a higher probability of an anomaly. Conductive hearing loss in a normal-appearing ear in a patient with contralateral atresia or microtia should be assumed to represent an abnormality of the middle ear. Occasionally, ossicular anomalies, especially those involving the malleus, may be visible on otomicroscopy. However, most often the otoscopic examination is unremarkable. Weber and Rinne tests must always be included and the results should be consistent with the audiometric findings.

Audiologic Testing

Identification and treatment of significant hearing loss in the pediatric population are critical for appropriate development of speech and language. Assessment of hearing loss can be challenging in this population because of the limitations inherent in patient histories and the questionable reliability of audiologic testing. Because conductive hearing loss is so commonly associated with otitis media, hearing loss caused by an ossicular malformation is occasionally inappropriately attributed to recurrent infection and eustachian tube dysfunction. It is important to confirm closure of the air-bone gap once a child's middle ear has been ventilated to avoid overlooking a congenital anomaly.[7] Audiologic testing is performed in an age-appropriate fashion. Children as young as 2 years old can be trained, with repeated testing, to respond reliably to masked air and bone conduction tests.[7] Tympanometry should be included as well because even the most astute clinician can sometimes miss a middle ear effusion, particularly in a poorly cooperative child. When appropriate, acoustic reflexes should be determined as well.

Progressive versus Stable Hearing Loss

An important determination in children is the distinction between progressive and stable hearing loss. Progressive conductive loss in the absence of significant otoscopic findings is most often compatible with a diagnosis of childhood otosclerosis. Alhough unusual, especially before the age of 12 years, this condition may be handled with the same surgical techniques as those used to perform stapes surgery in adults (see Chapter 117). Several authors have attested to the safety of stapes surgery in children.[14] Congenital fixation of the stapes, in contrast, carries some increased risk for sensorineural deafness and requires additional preparation. Otosclerosis may be diagnosed by documentation of progressive impairment in hearing, whereas congenital stapedial fixation is stable. The family history may provide an important clue in establishing this diagnosis, and CT may identify characteristic radiolucency of the otic capsule.

Imaging

Developments in radiologic evaluation of the temporal bone have facilitated the management of middle ear malformations. Careful evaluation of radiographic information will allow the surgeon to assess the chance of successful hearing restoration and provide specific details for informed patient consent. Dilatation of the

internal auditory canal or enlargement of the cochlear aqueduct can be identified preoperatively and may lead the surgeon to recommend conservative management (Fig. 116-1). All pediatric patients with conductive hearing loss not attributable to otitis media merit high-resolution temporal bone CT. Radiologic imaging is not indicated in an adult patient undergoing exploration for conductive hearing loss unless there is strong suspicion of a congenital middle ear malformation or neoplasm.

SURGICAL APPROACH

Most of the operative techniques used to correct middle ear malformations are similar to or modifications of tympanoplasty methods. Procedures that are successful in middle ear reconstruction after eradication of chronic disease are highly adaptable in dealing with a great variety of congenital ossicular problems. Various approaches to reconstruction of the conductive apparatus can be used, with technical details varying according to surgeon preference and the individual patient's anatomy. Thus, a “cookbook” approach to surgical correction of middle ear abnormalities can be difficult, and surgeons will ultimately develop their individual preferences for materials and reconstructive approaches. Materials must be available in the operating room to adequately replace or reconstruct any ossicular abnormality. Incus sculpting is often a viable option in these cases. An assortment of partial and total ossicular replacements, as well as stapes prostheses, should also be kept on hand. The Skeeter drill should be available for incus sculpting, as well as possible drill outs for thickened stapes footplates. Depending on surgeon preference for stapes procedures, CO_2, argon, or other otologic lasers can provide less traumatic access to the vestibule.

Regardless of the material chosen for reconstruction, the basic principles for reconstructing the middle ear conductive apparatus include the use of appropriately sized material in an adequately aerated middle ear and a stable, columnar mechanism for conduction of sound from the most lateral mobile structure (capitulum or footplate) to the malleus or tympanic membrane. Appropriate sizing of the reconstruction requires patience and experience—short prostheses are prone to slippage, whereas prostheses that are excessive in length may sublux into the vestibule. The more angled the reconstruction, the higher the chance for slippage. Some prosthetic materials (i.e., titanium) require placement of a small cartilage graft between the prosthesis and the tympanic membrane to prevent extrusion. Cartilage can be harvested from the tragus or concha via a small incision. With a postauricular incision, often a necessity in a pediatric patient, conchal cartilage is preferred because it can be harvested without a second incision.

Anesthesia

In an adult it is preferable to perform exploratory surgery under local anesthesia with intravenous sedation whenever possible. Local anesthesia results in the advantages of less bleeding, cost-effectiveness, postoperative analgesia, quicker patient mobilization, reduced risk for aspiration, the ability to test hearing during surgery, and immediate feedback regarding potential vestibular injury.[15] Young children require general anesthesia. The surgeon must balance the increased latitude provided by deeper anesthesia with the loss of feedback information from the patient. If the course of the facial nerve is found to be significantly aberrant on preoperative CT, general anesthesia with the use of a facial nerve monitor may prove safer than local anesthesia with sedation.

Elevation of a Tympanomeatal Flap and Exploration of the Middle Ear

Regardless of the mode of anesthesia, four-quadrant injection of a local anesthetic in combination with epinephrine is useful in minimizing bleeding during elevation of the tympanomeatal flap. When planning the tympanomeatal flap, one must always ensure that the superior limb of the tympanomeatal flap extends far enough superiorly to allow considerable removal of the scutum in the case of a lateral ossicular chain procedure (Fig. 116-2). In adults a transcanal approach typically provides adequate exposure. Younger children may require a postauricular approach. Once the middle ear is entered, each component of the ossicular chainis palpated independently. It is important to recognize that multiple defects may coexist—as highlighted by the increased incidence of lateral ossicular chain fixation identified on re-exploration after unsuccessful stapes surgery.[14]

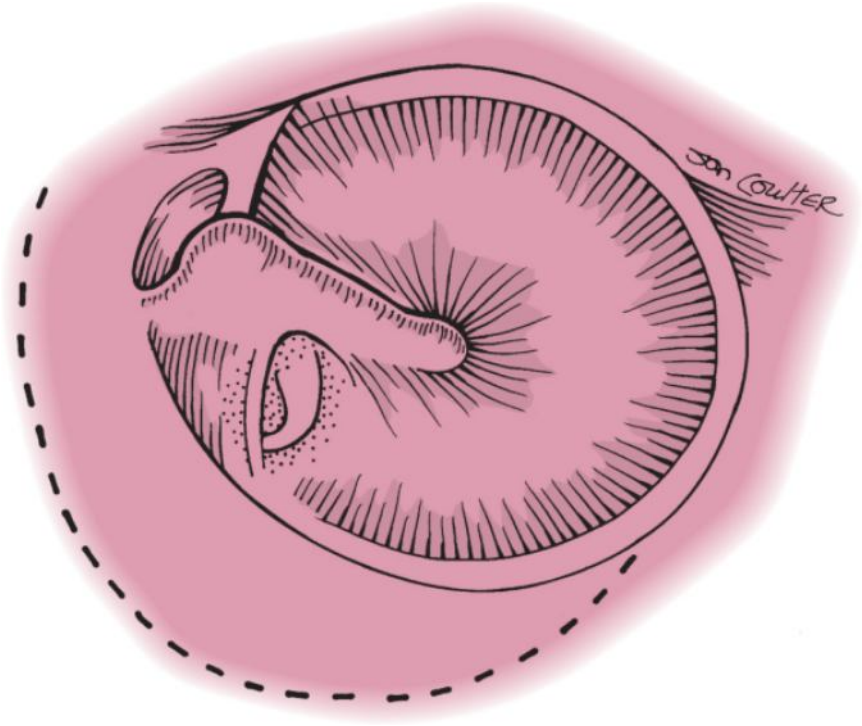


Figure 116-2 Correct incision for middle ear exploration to evaluate for conductive hearing loss.

Anticipating an Aberrant Facial Nerve

In approaching a malformed ear, identification of the facial nerve is critical. The preoperative CT scan should be reviewed carefully and the course of the nerve delineated. The surgeon must be familiar not only with the usual landmarks for the tympanic segment of the facial nerve but also with the most common aberrant courses. Dehiscence of the horizontal portion of the facial nerve is such a common finding that it may be considered a normal variant. Usually, the surgeon need only recognize the condition and avoid trauma to the nerve. Overhanging of the nerve may, on the other hand, present some difficulty in accessing the oval window (Fig. 116-3). The facial overhang can be managed by gentle retraction of the nerve while working in the oval window area. A stapes prosthesis may be placed in close contact with the nerve and may occasionally even cause a small indentation in the epineurium. When this abnormality is combined with shortening of the long process of the incus, the situation is more difficult. It can usually be solved by bending a crimp-on prosthesis to fit around the facial nerve (Fig. 116-4).

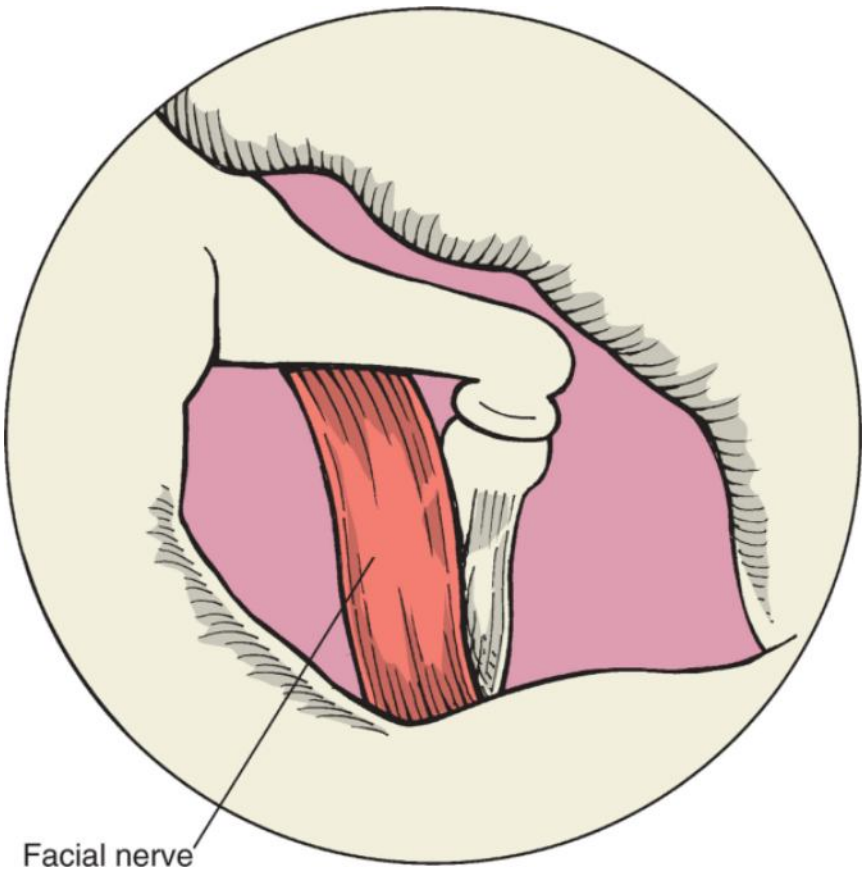


Figure 116-3 Dehiscent and overhanging facial nerve.

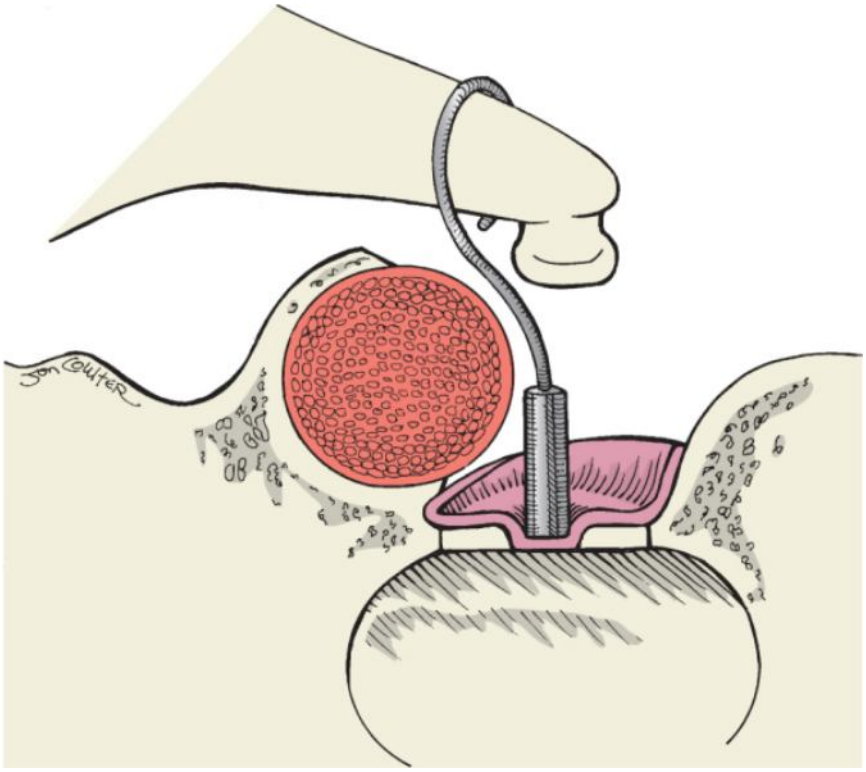


Figure 116-4 Use of a fashioned crimp-on piston prosthesis to circumvent an overhanging facial nerve.

The facial nerve may subdivide into several branches and in some cases may not traverse the bony fallopian canal. Occasionally, its course across the oval window prevents surgical access to the vestibule; to avoid injury to the facial nerve, these cases should be abandoned in favor of amplification. Anterior deviation of the vertical portion of the facial nerve is usually associated with congenital aural atresia, but it may exist alone; it is identified as it courses across the hypotympanum. In a series of 94 patients with congenital malformations of the ear (50 of whom had anomalies confined to the middle ear), Jahrsdoerfer reported an aberrant course of the facial nerve in 12 cases.[16] Most commonly, the tympanic segment of the nerve was displaced inferiorly and obscured the oval window. The nerve was often dehiscent in the middle ear. The free-floating facial nerve was mistaken for fibrous tissue in two cases previously operated on elsewhere and resulted in injury to the nerve.

Modifications in the anatomy of the chorda tympani nerve are common. This nerve may be larger or smaller in diameter than normal and may be displaced inferiorly or superiorly relative to the posterosuperior canal wall. On occasion, the chorda tympani may enter the middle ear from a position lateral to the annulus of the tympanic membrane. In this situation, the nerve travels beneath the skin of the posterior canal wall for a short distance and may be injured on elevation of the tympanomeatal flap.

In cases in which a middle ear malformation is suspected, intraoperative facial nerve monitoring provides a definite advantage. Unfortunately, these defects are often not suspected preoperatively. When unexpected questions arise regarding the location of the facial nerve, disposable stimulators can be used. If the intraoperative findings suggest that facial function may be jeopardized, the procedure should be terminated, appropriate imaging studies obtained, and formal nerve monitoring used in a subsequent procedure.

The following discussion reviews middle ear anomalies in accordance with the likelihood of their appearance. Congenital anomalies of the incus or malleus in the setting of a mobile stapes (i.e., epitympanic fixation or ossicular discontinuity) are quite amenable to reconstruction, as is congenital fixation of the stapes footplate in the setting of a normal incudomalleolar complex. However, when stapedial fixation occurs inthe context of an abnormal lateral chain, the chance for closure of the air-bone gap drops significantly.[14,17,18] Aplasia or severe dysplasia of the oval or round window carries an even worse prognosis for restoration of hearing, and controversy exists regarding whether reconstruction should be attempted at all in this situation. However, some authors have reported successful reconstruction of even these severe malformations.[16]

Defects of the Stapes

Although abnormalities of the stapes are the most frequently encountered of all ossicular defects, the variations seen are most often minor and inconsequential. Differences in the tilt and shape of the superstructure of the stapes are common. Hough has comprehensively reviewed these many variations.[19] The size of the obturator foramen and the shape of the crura often vary. Even defects as significant as a monopedal stapes are clinically unimportant unless hearing loss is present (Fig. 116-5).

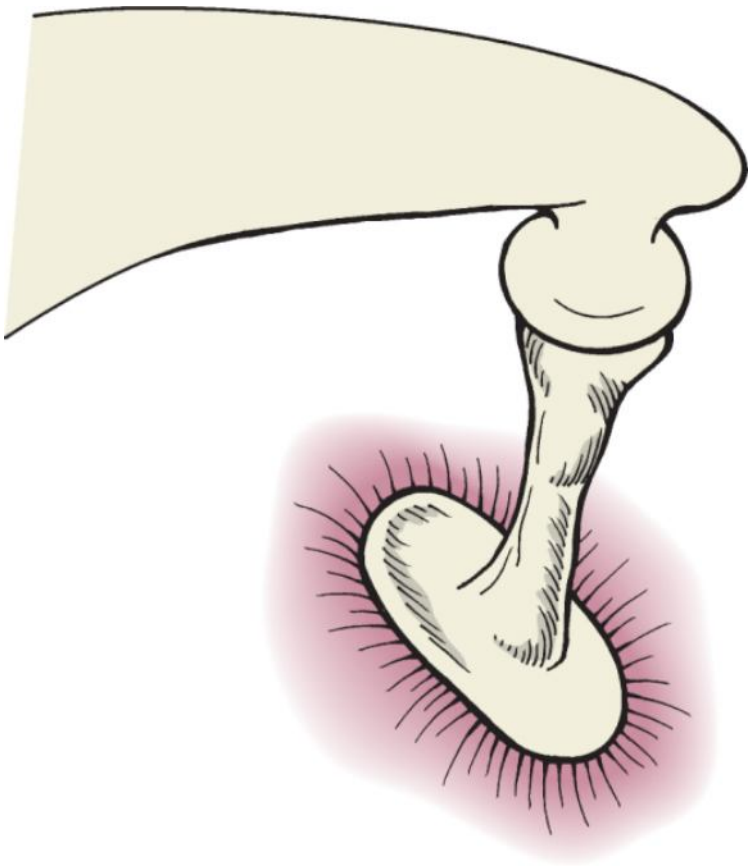


Figure 116-5 Monopedal stapes superstructure.

Abnormalities requiring surgical correction are related to stapes fixation and incomplete articulation of the stapes with the incus. Bony bridges may connect the superstructure to the promontory (Fig. 116-6) or the facial nerve, or the stapes tendon may be ossified. These bony bridges probably represent failure of detachment of the developing stapes from Reichert's cartilage (second branchial arch).[20] These bridges may be separated by fracturing them or dividing them with a CO_2 laser. Occasionally, the superstructure of the stapes is separated from the footplate (Fig. 116-7). The defective arch should be removed and replaced with a prosthesis. Reconstruction can proceed with a total ossicular prosthesis or a stapes prosthesis, depending on the mobility of the footplate.

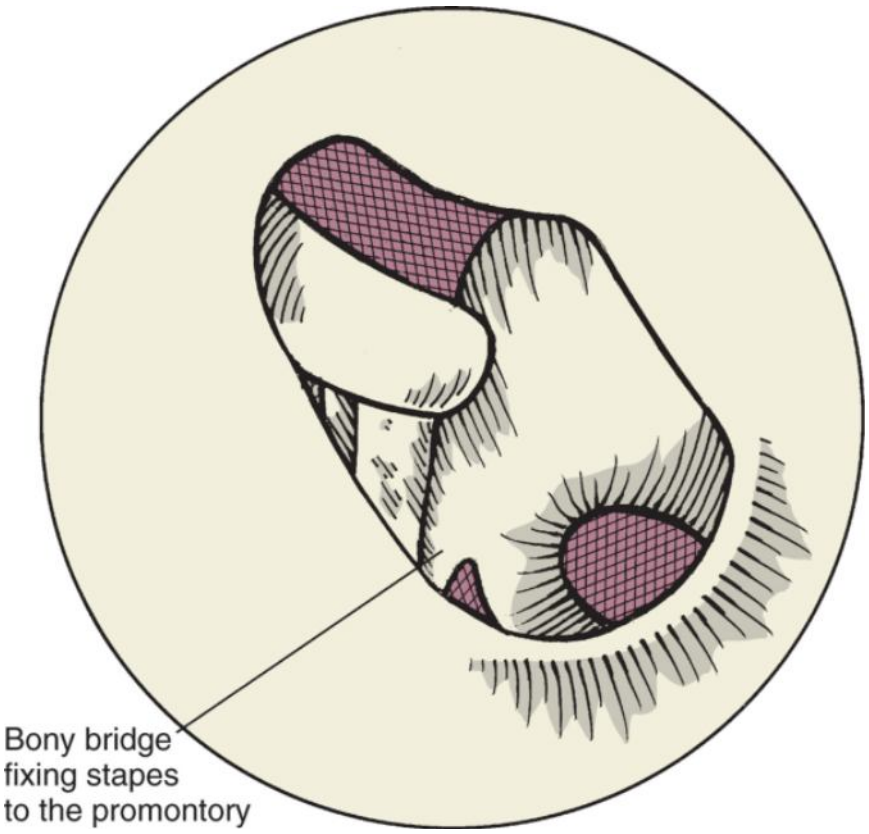


Figure 116-6 Bony attachment fixing the stapes to the pro-montory.

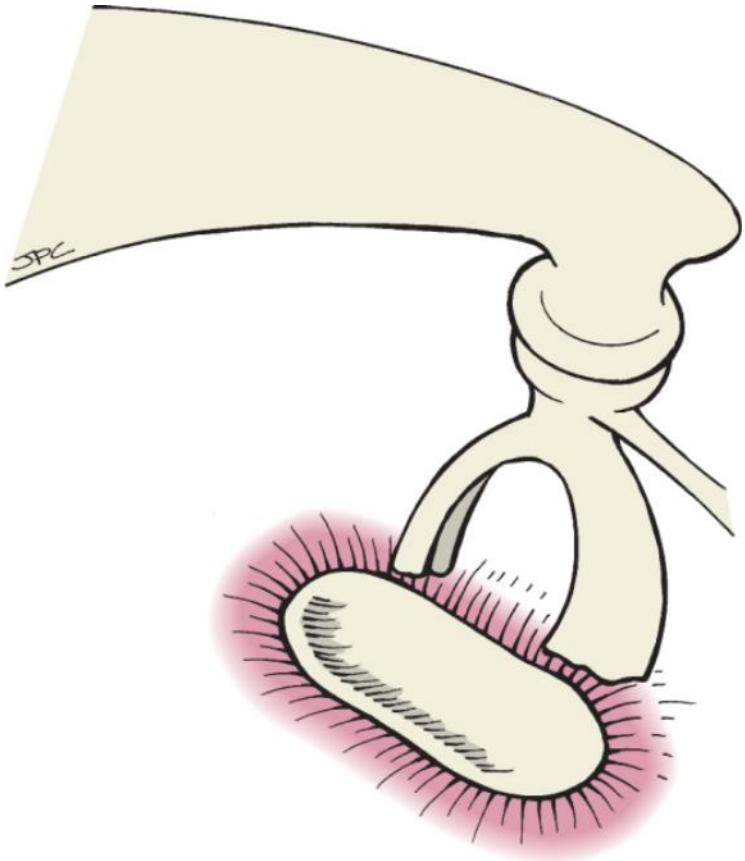


Figure 116-7 Congenital failure of the stapedial superstructure to articulate with the footplate.

Footplate thickness varies but is important only relative to fixation. Congenital fixation is recognized as a blending of the bone of the footplate with the surrounding otic capsule and the absence of a visible annular ligament (Fig. 116-8). Congenital fixation of the stapes may occur as a sporadic abnormality or in the setting of various syndromes. The fixed stapes is often deformed. In one series, one half the cases of stapedial footplate fixation were accompanied by associated anomalies of the incudomalleolar complex.[18] Although the reported results of stapes surgery in the setting of isolated stapedial fixation are quite good, the presence of associated abnormalities of the lateral ossicular chain suggests a less favorable chance for a satisfactory hearing result. In a series from De la Cruz and associates, the air-bone gap was closed to within 10 dB in 71% of patients with normal incudomalleolar complexes as opposed to only 12.5% of those with

anomalies involving the lateral chain.[14] The drop in hearing results can be attributed to the decreased stability of the reconstruction when the prosthesis must extend from the footplate to the malleus or tympanic membrane. The thickness of the footplate may necessitate a drill-out procedure. This is performed in much the same manner as for obliterative otosclerosis. The oval window is gradually saucerized with a microdrill until the footplate is “blue-lined.” At this point, fenestration is carried out in the usual manner, followed by stapes replacement with an appropriate prosthesis. Total absence of the stapes is rare, as is true of the other ossicles.

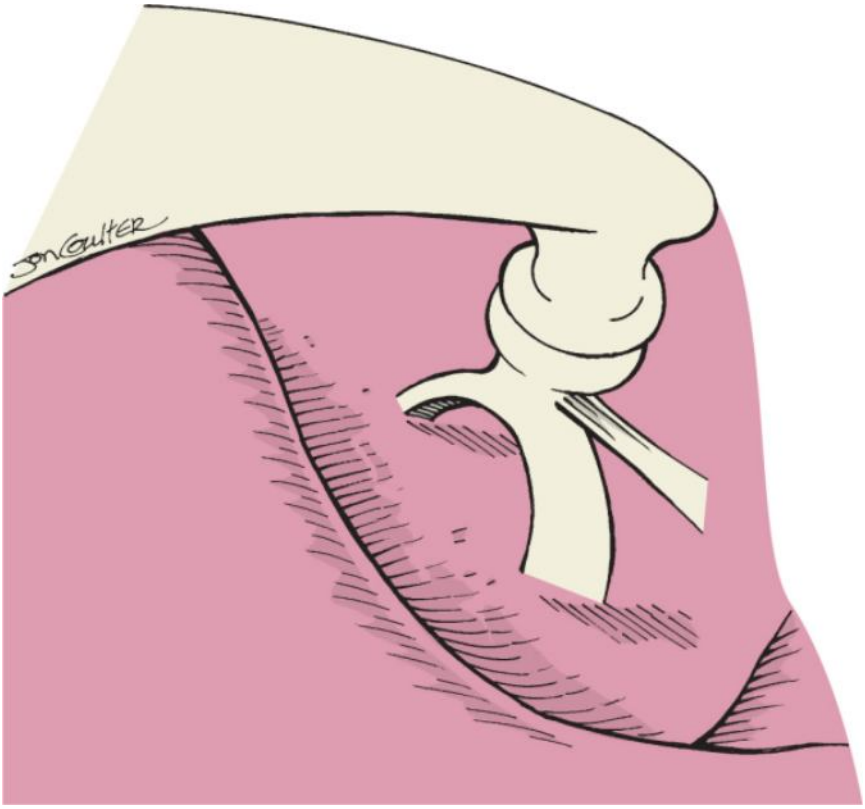


Figure 116-8 Congenital footplate fixation with absence of the annular ligament.

Handling a Perilymphatic Gusher

Excessive flow of perilymph on accessing the vestibule is more common in the setting of congenital stapedial fixation than otosclerosis.[10] Perilymph flow may be terminated by elevating the head of the table and packing the oval window with connective tissue. This situation must be handled with extreme patience. It is critical to avoid suctioning the vestibule. On occasion, a lumbar drain is necessary for controlling the flow of fluid at the oval window; however, it should be performed only if conservative measures fail. The patient may experience a spinal headache postoperatively. The preoperative CT scan should be reviewed carefully for the presence of a dilated internal auditory canal with inadequate definition of the habenula perforata, enlargement of the cochlear aqueduct, or the presence of a Mondini malformation, all of which suggest a risk for a perilymphatic gusher and may warrant management with amplification in lieu of surgical reconstruction.

Defects of the Incus

Hypoplasia or fibrous replacement of the long or lenticular process is the most common defect of the incus (Fig. 116-9). The appearance is much the same as with incus erosion after chronic otitis media with drum retraction. Complete lack of an incudostapedial joint is rare. These defects are most easily remedied by incus interposition, provided that mobility of the malleus and stapes is normal. This time-tested technique is associated with little risk of extrusion or absorption. There are many variations in modeling the incus for use, and surgeons have their own preferences. Figure 116-10 illustrates two options for fashioning and positioning the incus, depending on the anteroposterior distance between the malleus and stapes. Another option in dealing with this problem is a partial ossicular reconstructive/replacement prosthesis (PORP).

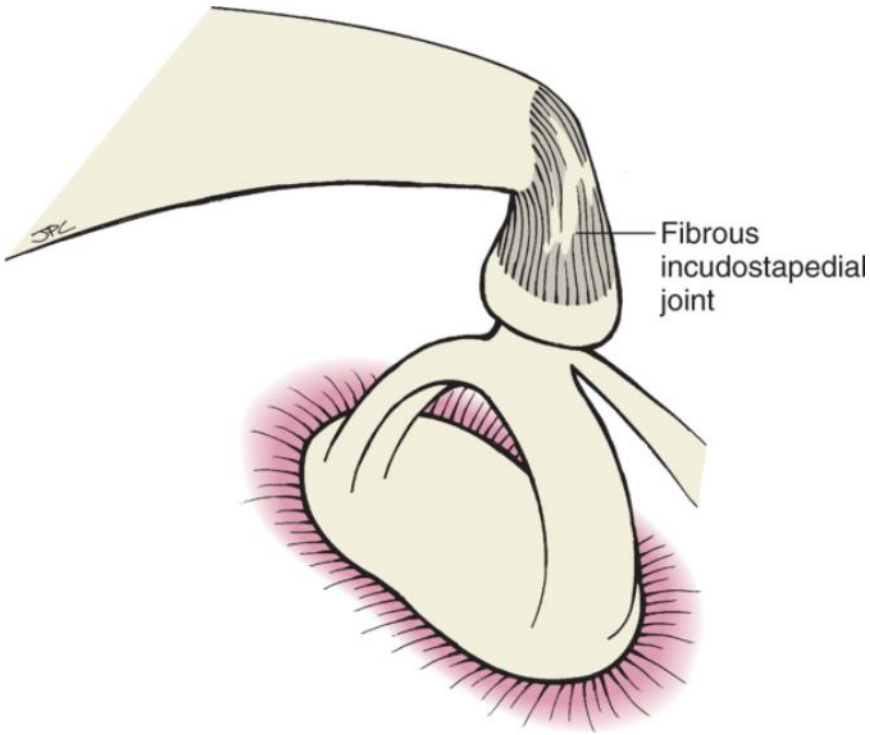
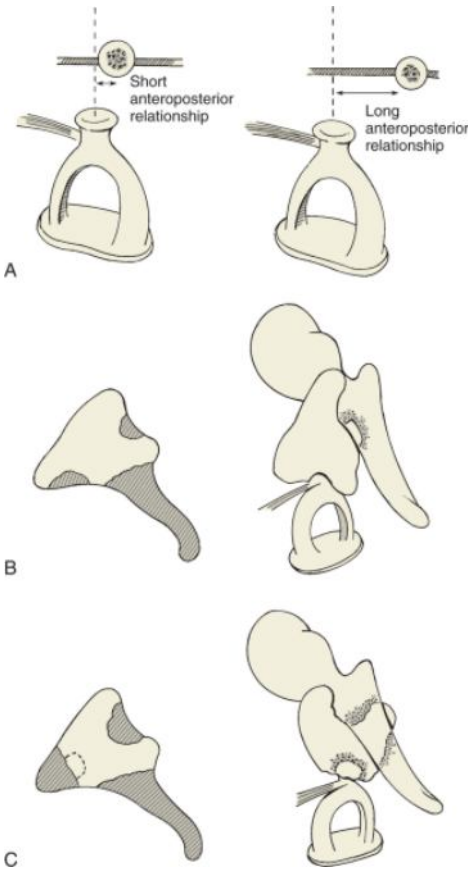


Figure 116-9 Fibrous incudostapedial joint.



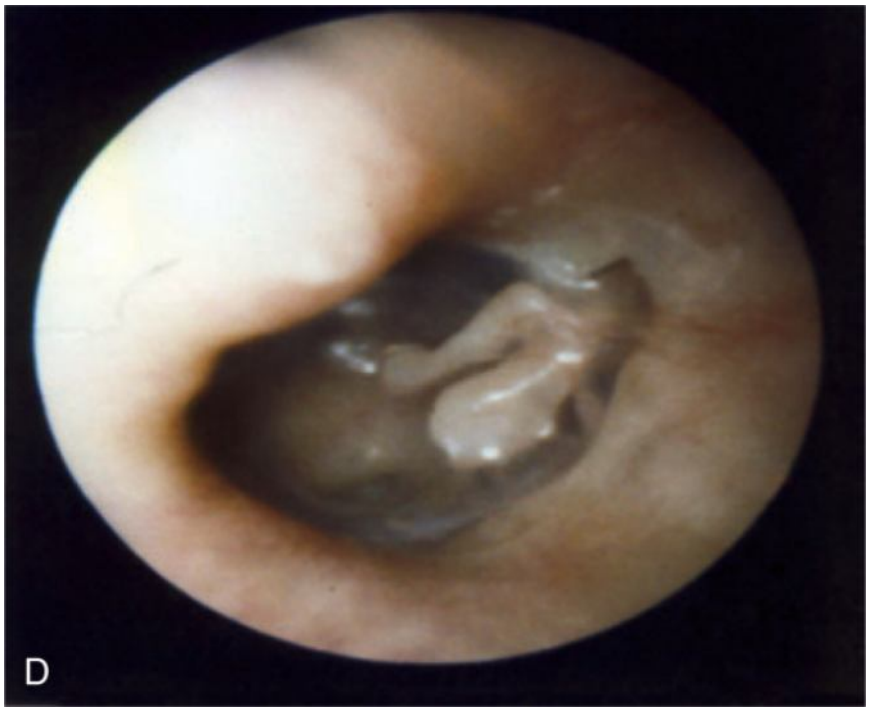


Figure 116-10 **A,** Variation in the anteroposterior (AP) dimensions between the malleus and the stapes head. **B,** Method of interpositioning the incus when the AP distance is long. **C,** Method of interpositioning the incus when the AP distance is short. **D,** Postoperative photograph of a left tympanic membrane after incus interposition.

Fixation of the incus may occur secondary to bony attachments within the attic or from the horizontal semicircular canal. Failure of complete development of the incudomalleal joint (fusion) is common in aural atresia, but it may also occur as an isolated finding (Fig. 116-11). Minor spurs or projections sometimes emanate from the long process of the incus, and rarely, a bony bridge may connect the long process to the malleus handle. The latter problem is easily solved by separation of the bony ridge from the malleus with a malleus nipper. Fixation occurring in the epitympanum, however, should be corrected by separation of the incudostapedial joint, removal of the incus, and interposition of the incus between the stapes and the handle of the malleus. In this instance, mobilization procedures are associated with a significant risk of refixation.

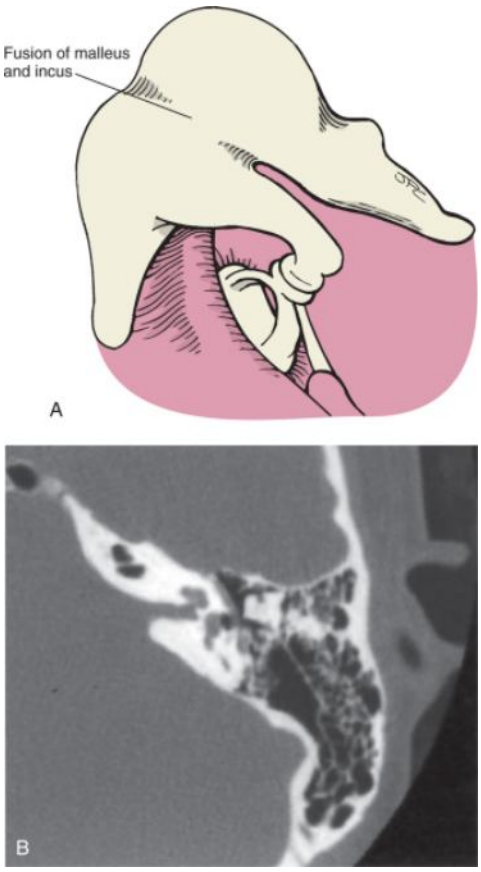


Figure 116-11 **A,** Fusion of the malleus and the incus. **B,** Radiographic appearance of malleoincudal fusion.

Variations in angulation and size of the distal portion of the long process are common in the setting of a congenital malformation. These aberrations may affect the use of certain stapedial prostheses, particularly the bucket-handle variety. A prosthesis with a large well may be necessary, and failure to engage the bucket handle may result.

Defects of the Malleus

Developmental anomalies of the malleus are the most uncommon of the ossicular defects. Complete absence of this structure is extremely rare and has been described only in the presence of multiple abnormalities. Separation of the manubrium from the neck and head of the malleus has occasionally been reported,[21] and the manubrium is also sometimes twisted or rotated anteriorly or posteriorly from its normal position. Absence or discontinuity of the malleus handle (Fig. 116-12) constitutes a major conductive hearing loss that is most easily handled by removing the incus and placing a PORP from the stapes to the tympanic membrane. Preservation of the incus in these cases makes reconstruction difficult. Use of a PORP ensures that the center of the tympanic membrane is used to provide the maximal driving force to the ossicular chain. The authors' choice of prosthesis is a hydroxyapatite PORP with a flexible shaft. Another rare defect is absence of contact between the malleus handle and the tympanic membrane (Fig. 116-13). This defect can be addressed by a lateral tympanoplasty with placement of the graft medial to the malleus handle. Alternatively, the incus can be removed and a PORP used to bridge between the stapes and the tympanic membrane.

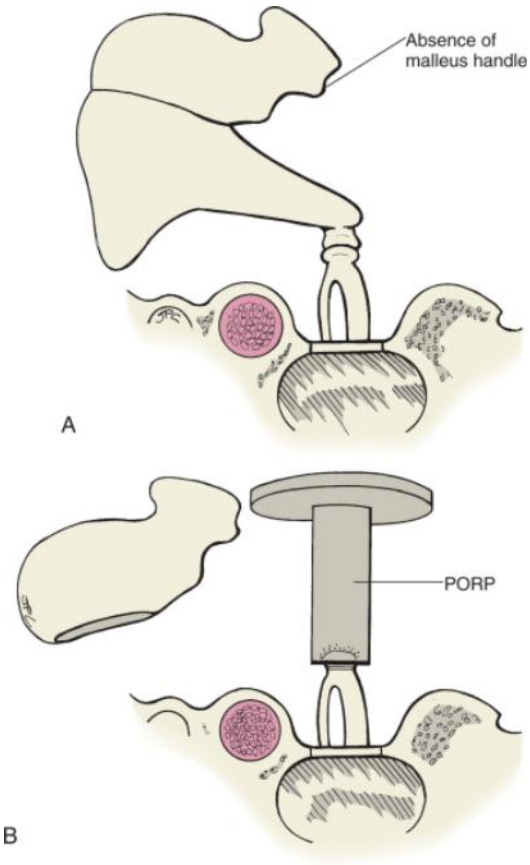


Figure 116-12 **A,** Absence of the malleus handle. **B,** Repair with a partial ossicular replacement prosthesis (PORP).

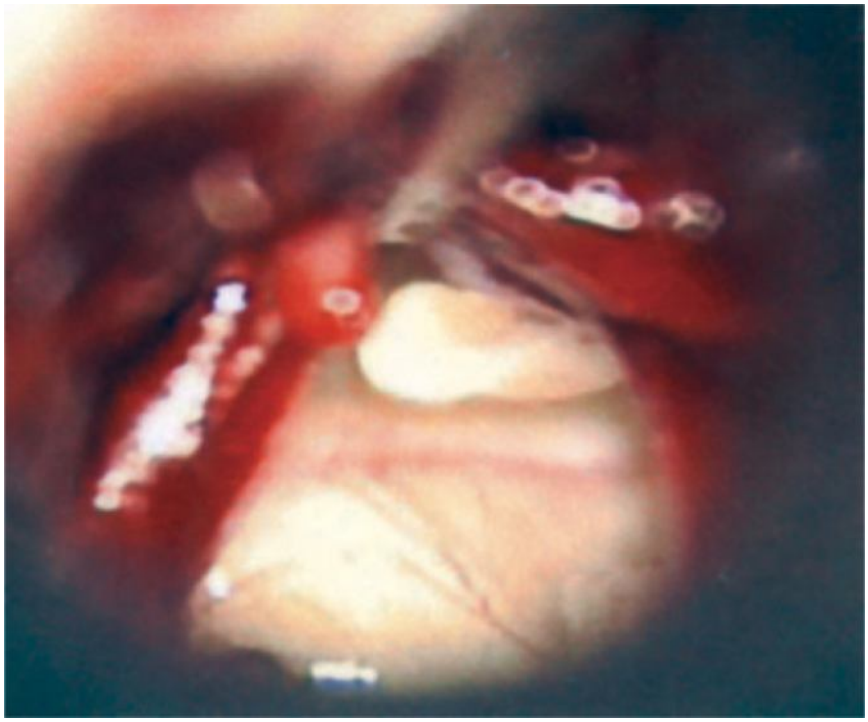


Figure 116-13 The long process of the malleus has failed to attach to the tympanic membrane.

Fixation of the head of the malleus occurs in about 1 in every 100 ears explored for conductive hearing loss (Fig. 116-14).[21] Bony attachments to the scutum may be present superiorly or laterally; fusion to the incus was described earlier. Early attempts at correction by merely fracturing these attachments were met with a relatively high incidence of refixation. For this reason, the head should be separated from the malleus with malleus nippers or a microdrill after separation of the incudostapedial joint. The incus is then removed, sculpted, and interposed between the stapes and malleus. Thus, lateral fixation of the ossicular chain is actually managed in the same fashion regardless of whether the malleus or the incus is involved.

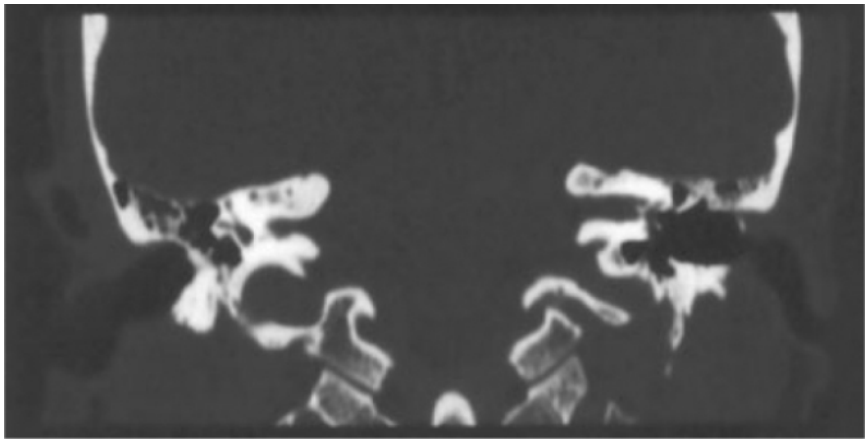


Figure 116-14 Computed tomography scan revealing fixation of the left malleus head.

Vascular Abnormalities

Small remnants of the stapedial artery are frequently noticed in the course of middle ear exploration. These tiny vessels usually present no problem, and the small amount of bleeding that they engender will cease spontaneously. Large remnants of the stapedial artery require more careful attention (Fig. 116-15). Access to the footplate and even its removal can usually be accomplished by working anterior or posterior to these vessels. Ligation of large vessels with clips or laser hemostasis should be avoided because of potential injury to the facial nerve. The persistent stapedial artery substitutes for the middle meningeal artery, and the foramen spinosum is congenitally absent.[22]

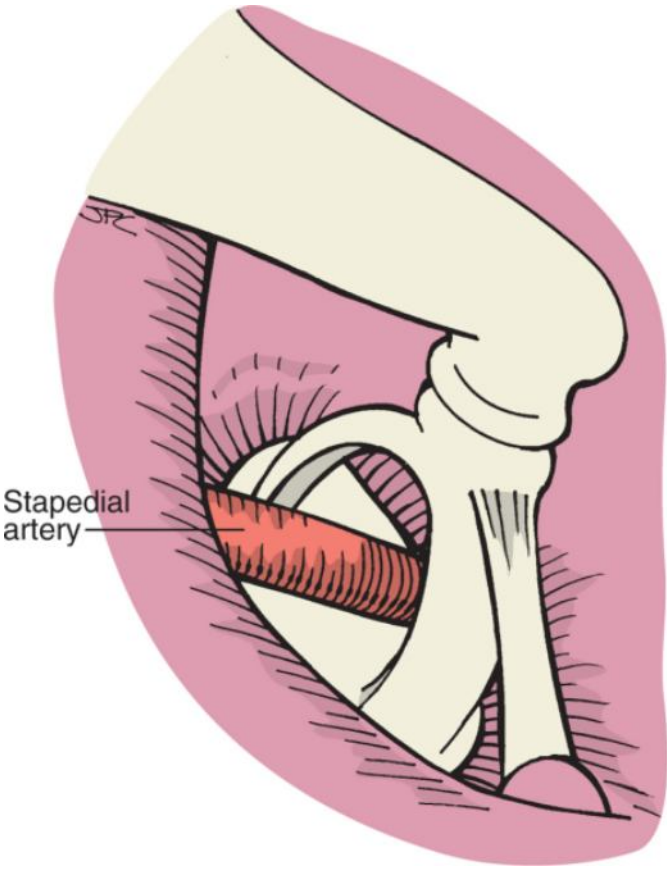


Figure 116-15 Stapedial artery remnant coursing through the obturator foramen of the stapes.

The presence of a high jugular bulb is relatively common and may be associated with dehiscence into the middle ear. The jugular bulb is most often defined as high riding when the most cephalad portion of the bulb reaches beyond the level of the floor of the internalauditory canal.[23] Occasionally, a high-riding bulb may contain a diverticulum, which projects even further cephalad (Fig. 116-16). This anomaly is usually asymptomatic and does not require intervention. If the high-riding bulb is symptomatic (pulsatile tinnitus, conductive hearing loss), it may be skeletonized and held inferior to the round window by a graft of tragal cartilage or cortical bone.

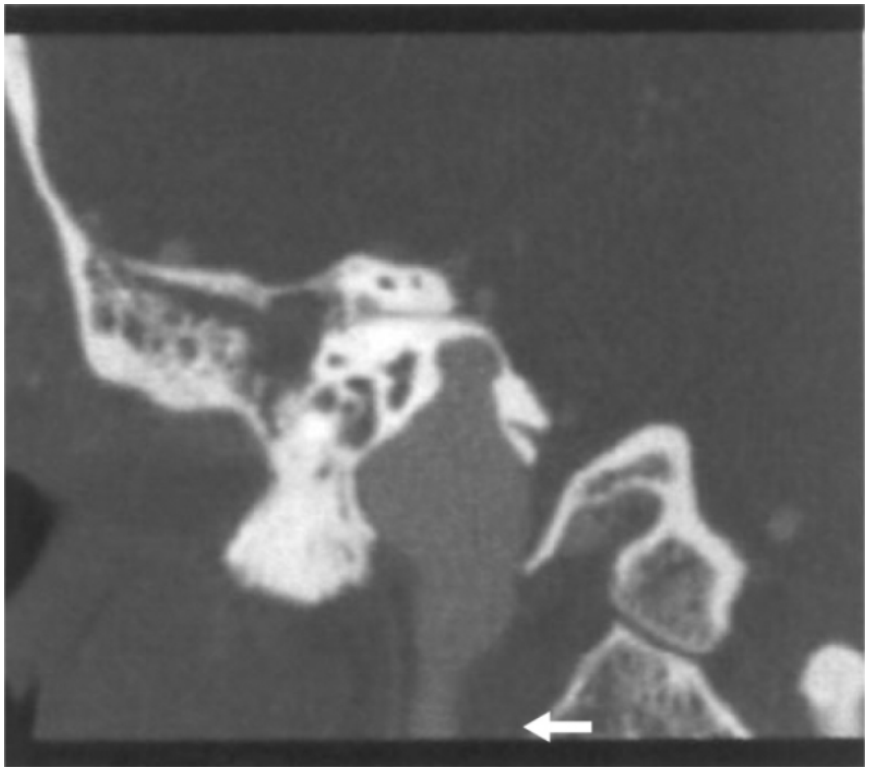


Figure 116-16 Contrast-enhanced coronally reconstructed computed tomography scan demonstrating a high-riding jugular bulb with a small diverticulum arising from the superior surface *(arrow)*.

The rarest of vascular abnormalities is an aberrant carotid artery. The normal petrous carotid turns from its vertical course to a horizontal and medial trajectory. This genu is normally anteromedial to the cochlea. An aberrant carotid artery results from inadequate development of the cervical internal carotid artery. Blood flow is redirected through the normally very small inferior tympanic branch of the ascending pharyngeal artery and to the caroticotympanic artery.[24] The aberrant vessel courses through the middle ear and appears on CT as a tubular soft tissue mass along the promontory that enters the horizontal carotid canal through a dehiscence in the lateral carotid plate (Fig. 116-17). An aberrant carotid artery is not amenable to surgical therapy. The chief danger with this entity is failing to recognize its true identity or mistaking it for a glomus tumor. Misdiagnosis with puncture or biopsy of the vessel can result in significant hemorrhage and possibly neurologic sequelae.[25]

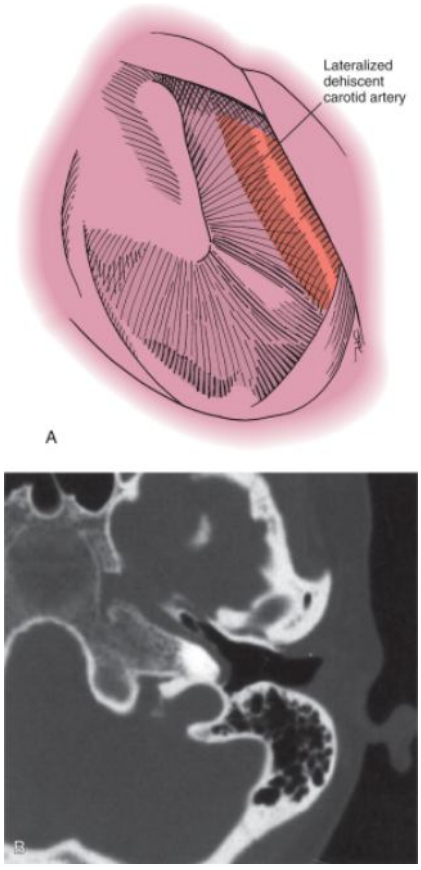


Figure 116-17 **A,** Aberrant carotid artery as seen through the tympanic membrane. **B,** Computed tomography scan demonstrating an aberrant internal carotid artery.

Round Window

Complete absence of the round window is perhaps the rarest of middle ear defects. As the otic capsule ossifies during development, a cartilaginous ring prevents bony deposition at the site of the nascent round window membrane.[26] Absence of the round window is presumed to be secondary to improper formation of this cartilaginous ring. Jahrdoerfer reported successful hearing outcomes in three patients with congenital oval window aplasia who had no apparent round window, so this finding does not necessarily represent a contraindication to ossicular reconstruction.[16] Fenestration of the promontory to create a round window is not advisable because of a significant risk of sensorineural hearing loss.

Other Lesions

Congenital cholesteatomas are discussed in Chapter 126. These lesions are usually seen transtympanically or occasionally discovered at the time of myringotomy, but they are rarely encountered unexpectedly in the course of middle ear exploration. Choristomas are unusual lesions consisting of heterotopic tissue and may be found in the middle ear as well. In both instances, treatment requires complete surgical ablation followed by reconstructive measures common with tympanoplasty. Other soft tissue lesions, such as hemangiomas and adenomas, are rare.

POSTOPERATIVE MANAGEMENT

Postoperative management of patients undergoing surgery for congenital middle ear abnormalities is generally the same as that after tympanoplasty with reconstruction. The exception is a perilymph gusher. In these cases the head is maintained at an elevation of at least 30 degrees for 48 hours. When perilymphatic drainage continues postoperatively, a lumbar subarachnoid catheter is inserted, and 50 mL of cerebrospinal fluid is removed every 8 hours in adults and older children. Correspondingly, younger children require withdrawal of lesser amounts. Minimal hydration is maintained, and the catheter can usually be removed within 48to 72 hours. Strenuous activity is forbidden for 2 to 3 weeks. The usual precautions against blowing the nose and avoidance of water in the ear are given.

Packing in the external canal consists of a small silk sleeve held in place over the tympanomeatal flap and supported with several small cotton balls or Merocel spheres moistened with antibiotic ointment. This material is removed 1 week after surgery, but water is avoided until the canal is completely healed. Audiometry is performed when it can be determined that the flap is completely healed and there is no longer fluid or blood within the middle ear, typically within 3 to 4 weeks postoperatively. Antibiotics are not routinely administered after middle ear surgery, and analgesics are necessary for several days only.

COMPLICATIONS

Complications after surgical correction of middle ear anomalies include hearing loss, vertigo, perilymphatic drainage, and facial paralysis. Treatment of perilymphatic drainage has been addressed.

In the early postoperative period, hearing loss may usually be assessed with a tuning fork. Conductive loss is common because of packing in the canal and middle ear effusion, but sensorineural loss may follow a drill-out procedure or one associated with a perilymph gusher. Stopping the fluid leakage is the most effective way to prevent permanent hearing loss. Steroids have no proven value in these cases but are often administered in divided doses of 60 to 80 mg of prednisone daily in adults.

Mild to moderate vertigo can follow any middle ear procedure but is most common after opening of the oval window. Severe vertigo, especially when not present intraoperatively under local anesthesia, is unexpected and may signal a serious problem. The surgeon should consider a fistula, subluxation of the footplate, or a prosthesis that extends too far into the vestibule. Sensorineural hearing loss often accompanies this complication. If one of the listed causes is believed to be possible, surgical exploration should be carried out immediately to correct the problem.

Temporary facial paralysis may result from diffusion of local anesthetic toward the stylomastoid foramen or, less commonly, from the use of topical anesthesia in the middle ear in the presence of dehiscence in the fallopian canal. An appropriate period of observation will soon delineate these cases. When facial weakness persists, the surgeon must question whether the nerve was exposed and injury may have occurred. This situation is unusual after middle ear surgery but is more common after mastoidectomy or procedures for aural atresia. When paralysis is unexpected and without explanation, re-exploration is mandatory to rule out disruption of the nerve.

Acknowledgments

The authors would like to acknowledge Dr. Donald B. Kamerer for his major contribution to the first edition of this chapter and Dr. Barton Branstetter for generously sharing radiologic images of congenital temporal bone malformations.

PEARLS

- A chance encounter with a middle ear abnormality is a probability for every otologic surgeon.
- Middle ear malformations should always be considered in the differential diagnosis of children with conductive or mixed hearing loss.
- Preoperative CT should be obtained in all pediatric patients with conductive hearing loss not attributable to otitis media to fully assess the middle ear for an aberrant course of the facial nerve and abnormalities of the ossicular chain.
- A variety of materials should be available so that the otologic surgeon is equipped to deal with any unexpected finding in the course of middle ear exploration for conductive hearing loss.
- If a perilymph gusher is encountered, avoid suctioning at the fenestra, elevate the head of the bed until the flow stops, and pack the oval window with connective tissue.

PITFALLS

- Middle ear anomalies are accompanied by a higher risk of an aberrant facial nerve and require vigilance.
- Stapes surgery should not be performed when there is radiologic evidence of a widened internal auditory or cochlear aqueduct.
- Failure to recognize vascular anomalies such as an aberrant carotid and attempted myringotomy or biopsy may result in significant complications —temporal bone CT allows this entity to be recognized readily.
- Inappropriate exploratory tympanotomy can be avoided by confirmation of the absence of stapedial reflex before surgery.
- Unexpected postoperative vertigo should raise concern for a possible fistula, subluxation of the footplate, or a prosthesis that extends too far into the vestibule.