

Congenital Aural Atresia

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Embryology

- * External Auditory Canal
 - * first branchial groove
 - * starts as a solid core of epithelial cells, which undergoes absorption in a medial to lateral direction
 - * if this is arrested can result in a normal TM with bony external ear canal assoc with an atretic or very stenotic or membranous canal predisposing to canal cholesteatoma

- * Medial portion of the EAC

- * formed by tympanic bone and begins to ossify in the 3rd month

- * forming tympanic ring and osseous portion of EAC

- * Malformation produces atretic bone at the level of the tympanic membrane, resulting in atresia of the ear canal

Mastoid and Middle Ear

- * ET, middle ear, mastoid air cells: First pharyngeal pouch
- * Ossicles (except for vestibular portion of stapes footplate): First and second branchial arches
- * Membranous labyrinth is derived from the ectodermal otocyst, so hearing and vestibular function should be normal
- * Stapes footplate from otic capsule and is usually normally developed

Facial Nerve

- * Typically see bony dehiscence of the fallopian canal
- * Acute angle at the second genu
- * More **anterior** and **lateral** direction
- * Places the nerve at higher risk when drilling in the posterior inferior portion of the new ear canal
- * Nerve may lie just deep to the tragus as it exits the skull, making it susceptible to injury if undermining the auricle
- * There is a correlation between degree of microtia and extent of facial nerve abnormality

Classification of Microtia

- * Grade I-minor malformation
 - * auricle smaller than normal, but with all parts discernable
- * Grade II
 - * auricle represented by a curving or vertical ridge of tissue
- * Grade III
 - * only a small rudimentary soft tissue structure present

Ombredanne's Criteria

- * Major Malformation
 - * EAC and TM usually absent, and stenosis prevents visualization of the medial aspect of the ear canal
 - * ME space is reduced, malleus and incus are deformed, fused, and fixed to the atretic bone
 - * Severe cases-ME space is hypoplastic, and ossicles are rudimentary or absent. Dehiscence or displacement is expected in most major malformations
 - * Commonly seen with grade II and III microtia with normal inner ear function

* Minor Malformation

- * CHL secondary to absence or deformity of one or more ossicles, or fixation of the ossicular chain
- * **Abnormalities of the stapes** may be more severe in minor malformations
- * ME space and TM are normal, EAC is patent
- * Dehiscence or displacement of FN may occur
- * Pinna usually normally developed or grade I microtia

Audiometry

- * If canal patent can do electrocochleography
- * Patients with bilateral atresia difficult to test secondary to masking dilemma
- * Cannot assume that cochlear function is normal bilaterally even if inner ear normal on CT
- * Need bone conduction auditory brainstem response testing
 - * Wave I of ABR is generated by the distal portion of the auditory nerve, with minimal crossover to the contralateral ear
 - * Best measured by a recording electrode ipsilateral to the stimulated side

CT

- * Necessary for all patients being considered for surgery
- * In cases of stenosis can evaluate for possible cholesteatoma formation
- * Axial-body of malleus and incus, IS joint, round window
- * Coronal-stapes, oval window, vestibule
- * Both projections needed to evaluate the facial nerve

Medical Management

- * Unilateral Atresia
 - * If normal hearing in contralateral ear, no immediate intervention necessary
 - * Preferential seating in school
 - * Atresia-bone conduction HA
 - * Stenosis-air-conduction HA
- * Bilateral Atresia
 - * need early amplification with bone conduction HA

Surgical Management

- * Reluctant to operate on unilateral cases until adulthood
 - * hard to predict degree of hearing improvement, potential lifetime care of mastoid cavity, risk of injury to the facial nerve
 - * Need to improve hearing threshold to 25dB or greater to eliminate handicap of unilateral hearing loss--can be achieved in about 50% of carefully selected patients
- * Goal in bilateral atresia is to restore sufficient hearing so amplification no longer needed
 - * better ear (by CT evaluation) is selected for initial procedure

Selection Criteria

- * Patients typically have a 10dB residual conductive deficit after repair
- * SN function should normal in operative ear, and normal to near-normal in contralateral ear to avoid operating on better hearing ear
- * Mostly determined by CT evaluation
- * In unilateral cases, only ideal candidates are selected
- * In bilateral cases, minimal criteria are a middle ear of at least one half normal size, and presence of ossicular mass
- * Only 60% of patients with aural atresia are surgical candidates

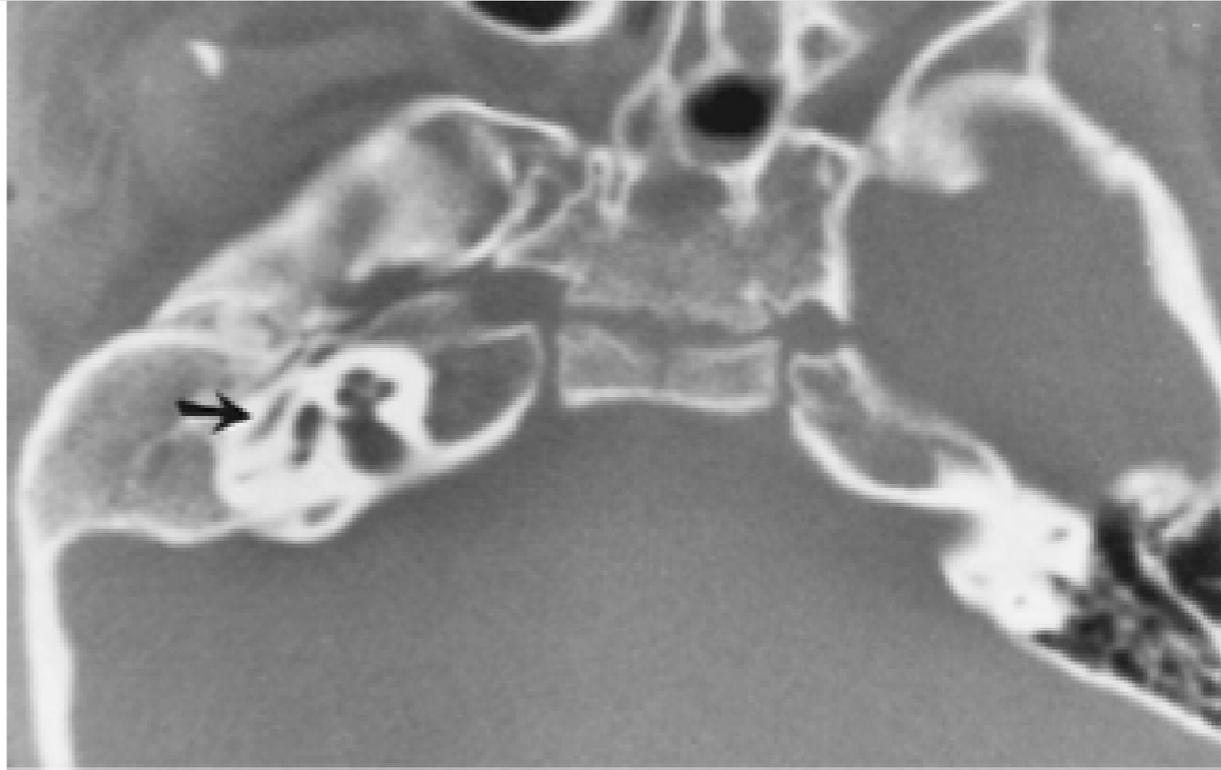
- * Often CT findings can be predicted by physical exam--poor middle ear development seen more frequently in patients with craniofacial abnormalities than those with isolated aural atresia
- * Treacher Collins--often have bizarre middle ear findings
- * The better developed the auricle, the larger and better developed the middle ear

Jahrsdoefer Grading System

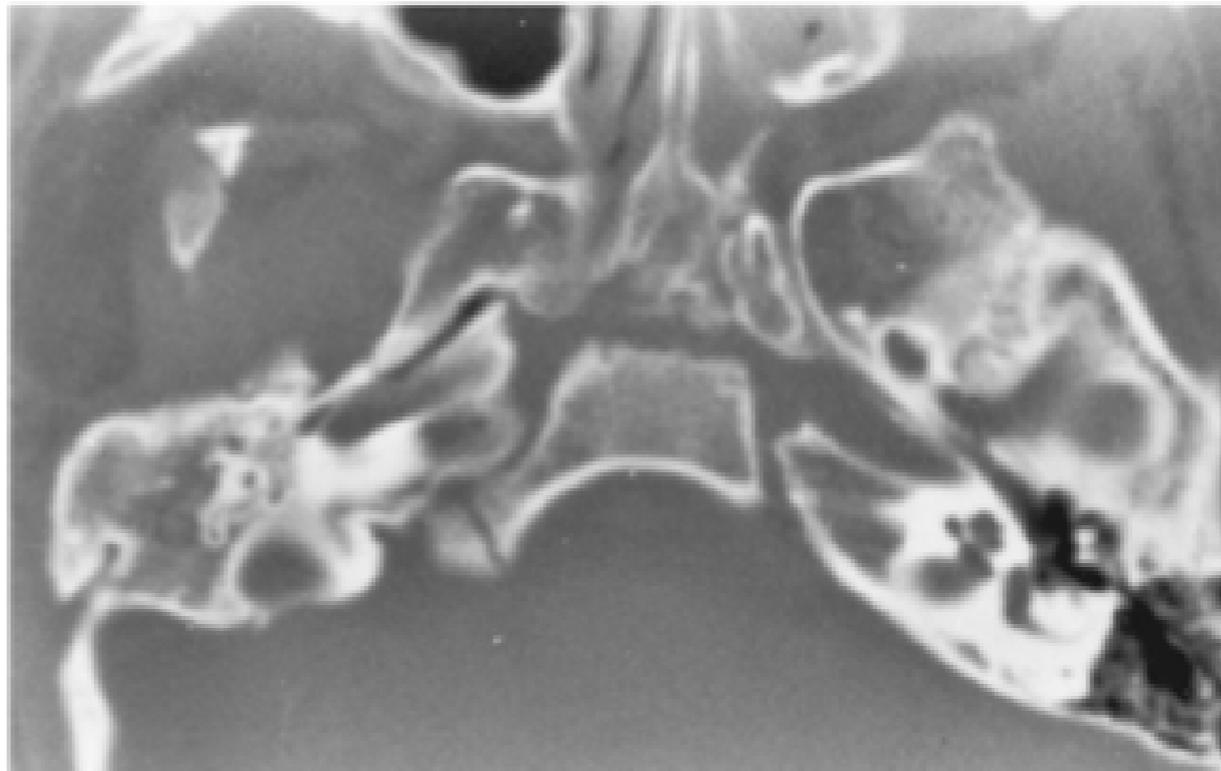
- * Total points:
 - * 8-good prognosis
 - * 7-fair prognosis
 - * 6-marginal candidate
 - * 5-poor prognosis

Parameter	Points
Stapes Present	2
Oval window patent	1
Middle ear space	1
Facial Nerve	1
Malleus/Incus Complex	1
Mastoid Pneumatization	1
Incus-Stapes Connection	1
Round Window	1
External Ear Appearance	1

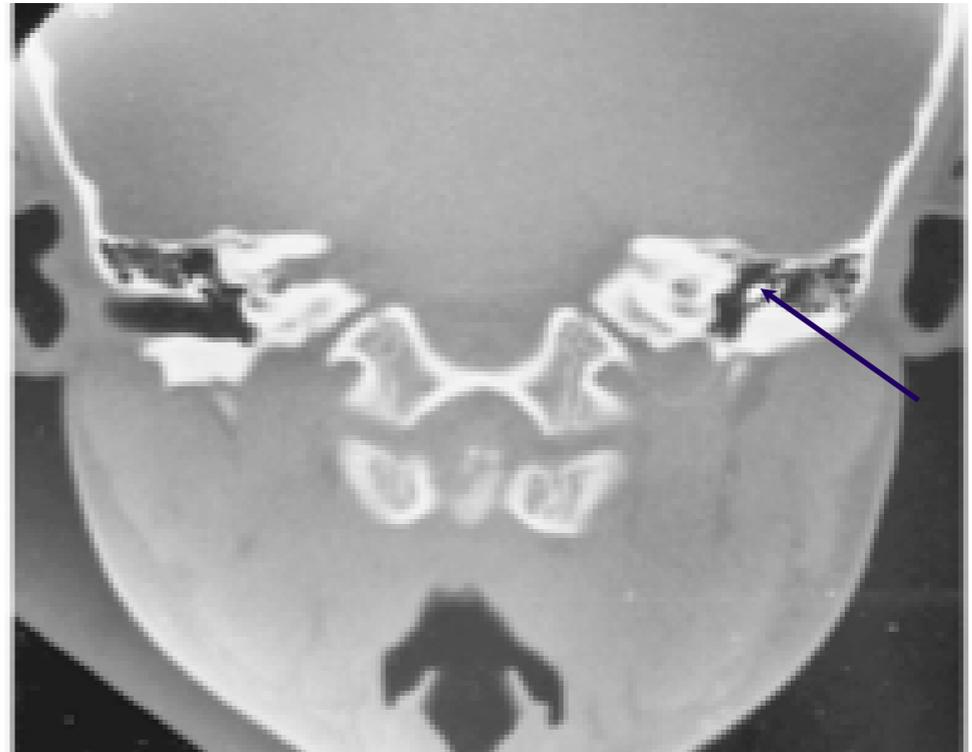
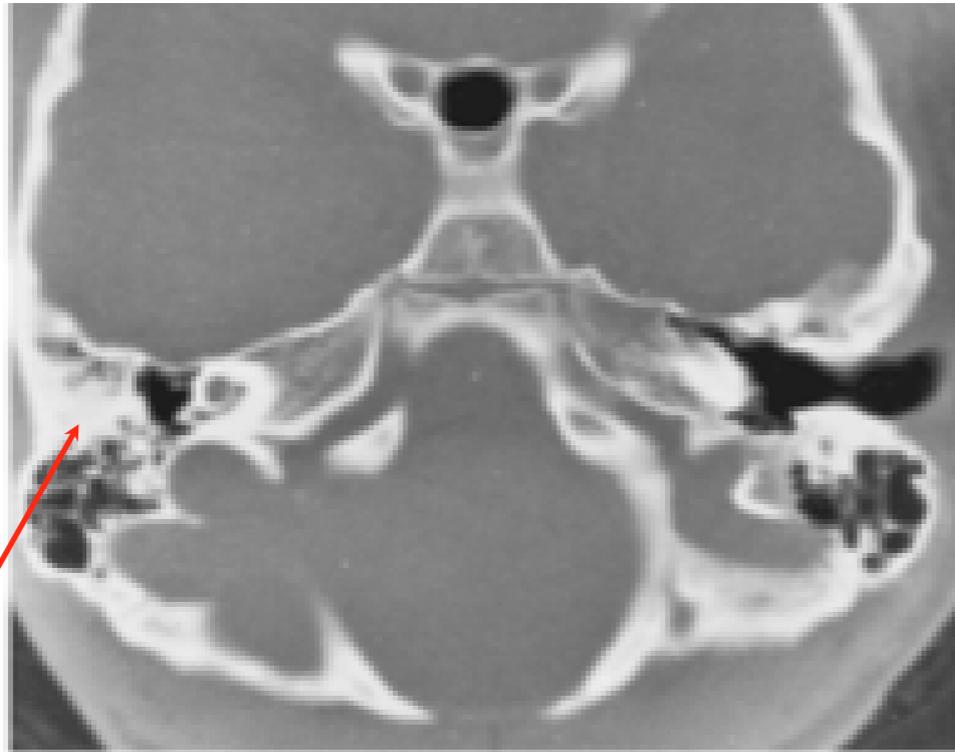
NOT A
SURGICAL
CANDIDATE



A



B



CANDIDATES FOR SURGERY

Timing of Surgery

- * Early as 6 to 7 years old, and microtia repair, if needed should be well underway, ensuring an unoperated field without compromised blood supply for the implanted auricular framework.
- * Typically atresia surgery is performed midway through the microtia repair, after the auricular framework has been implanted and the lobule transposed, but prior to reconstructing the tragus or elevating the auricle.
- * The auricle can be repositioned to align the meatus and EAC

Cholesteatoma

- * Patients with stenosis are at higher risk for cholesteatoma formation
- * Cole and Jahrsdoefer: of 50 pts with average canal diameter of 4mm or less, 50% developed cholesteatoma
- * Patient age and exact canal size were important variables: no cholesteatoma in age less than 3, and bone erosion and middle ear involvement not seen in patients under 12
- * Most cholesteatomas developed in canals 2mm or less
- * Commonly present with drainage or postauricular fistula track

Management Protocols in Stenosis

- * Patients with significant stenosis that prevents adequate cleaning of EAC and visualization of TM should have CT by age 5
- * If no cholesteatoma and favorable middle ear findings, can proceed with canal and middle ear surgery
- * If middle ear unfavorable, can offer canaloplasty
- * If parents opt for non-surgical management, follow with serial CTs every few years
- * Periodic CTs not indicated in patients with a completely atretic ear canal, as cholesteatomas are rare in this patient population

Surgical Technique

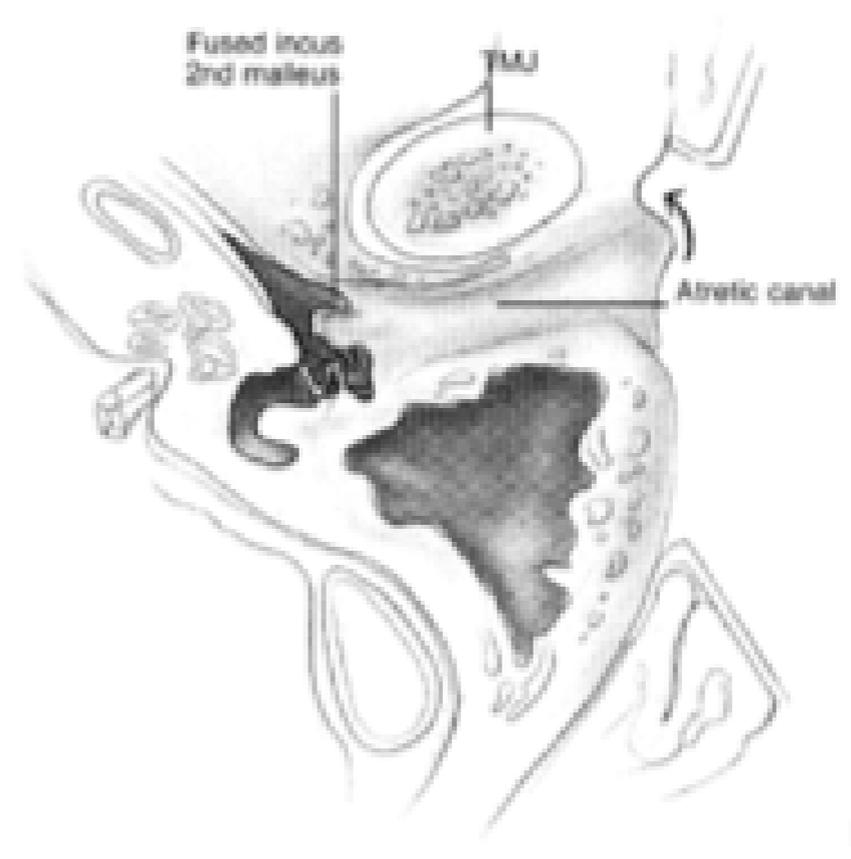
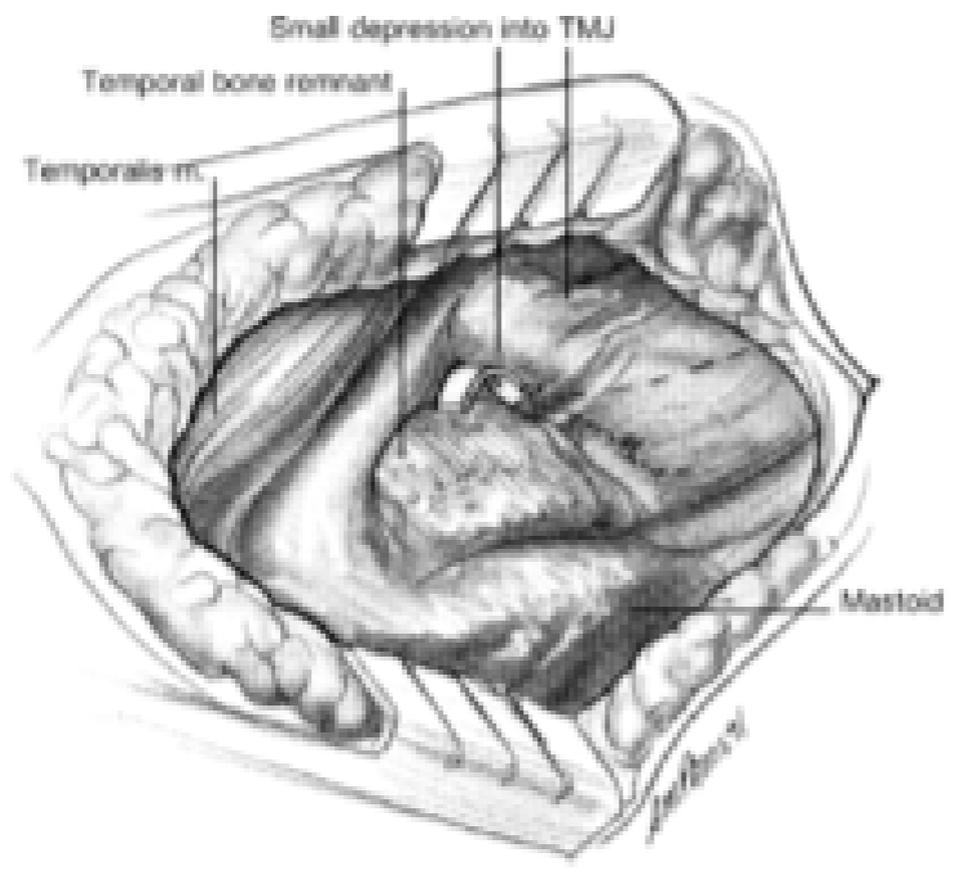
- * Two surgical approaches
 - * Mastoid
 - * Sinodural angle identified and followed to antrum, facial recess opened and IS joint separated. Atretic bone is then removed
 - * Anterior (popularized by Jahrsdoefer)
 - * Exposure of mastoid air cells is limited, and drilling is confined to an area defined by TMJ anteriorly MCF dura superiorly, and mastoid air cells posteriorly
 - * Large mastoid cavity is avoided, there is less surgical manipulation in the area of the mastoid segment of the facial nerve, and the more cylindrical canal better facilitates a STSG

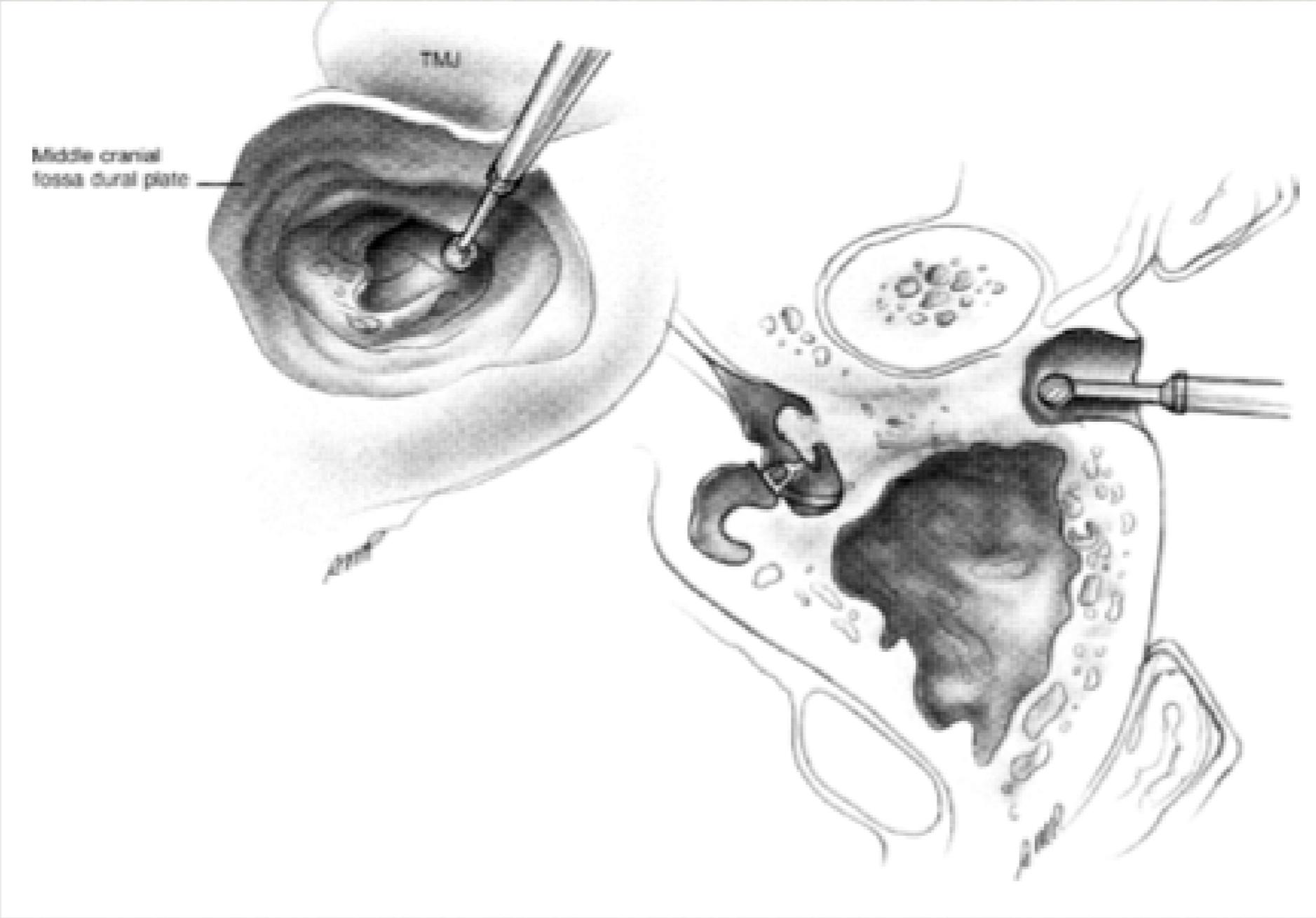
Incision

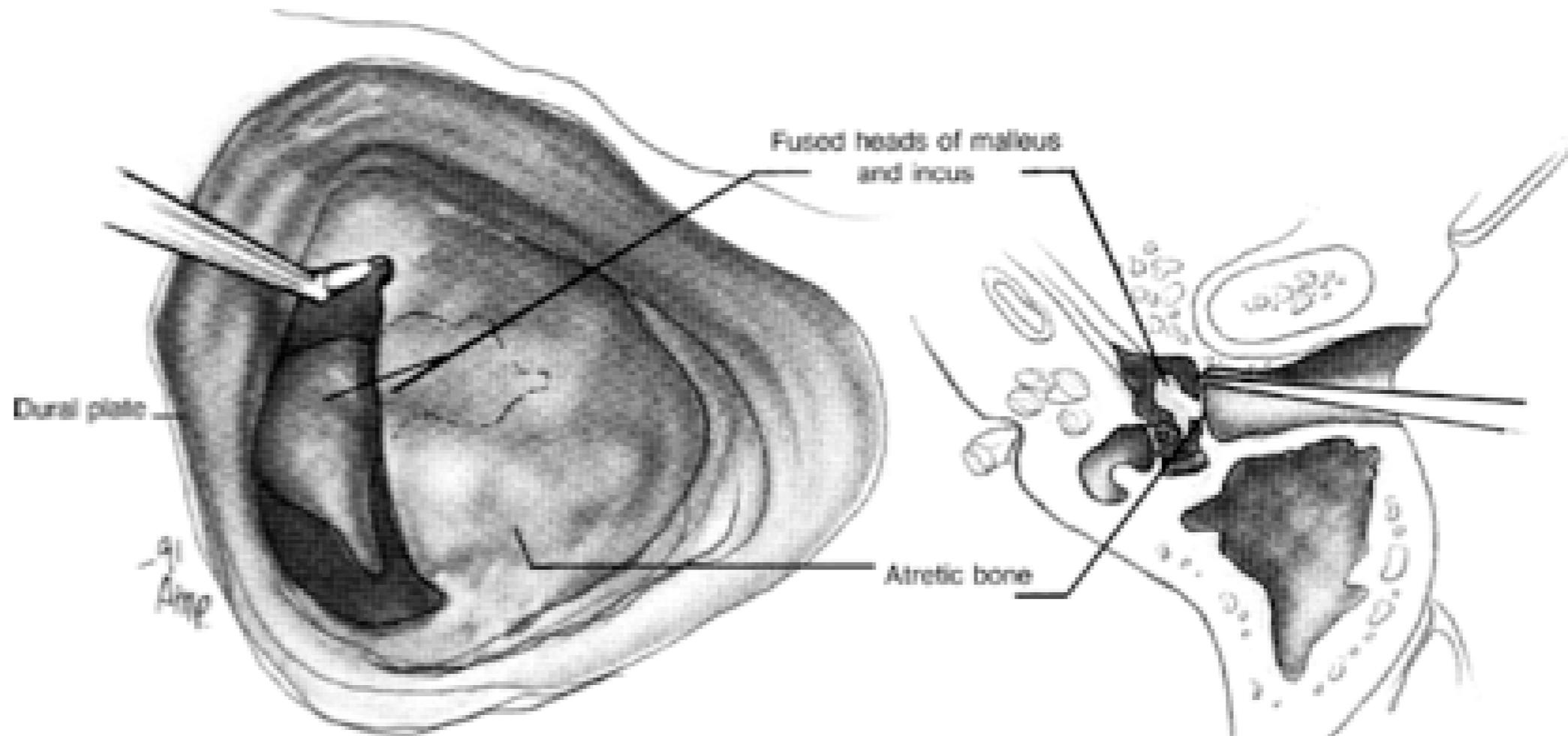
- * Post-auricular
- * Soft tissues elevated anteriorly until a depression is encountered
 - * In most major malformations, this is the TMJ, but occasionally a stenotic bony ear canal is encountered
 - * Manipulation here should be limited as the facial nerve frequently exits the skull into the glenoid fossa

Drilling a Canal

- * Most atretic ears without a tympanic bone remnant
 - * Sufficient space between the glenoid fossa anteriorly and mastoid air cells posteriorly for a canal
- * If tympanic remnant present, the atretic bone directs the drilling for the EAC
- * Concentrating the drilling superiorly along the MCF protects the facial nerve because it will always lie medial to the ossicular mass in the epitympanum
- * Because of the acute angle of the facial nerve at the 2nd genu, it is vulnerable to injury as the EAC is opened in a posteriorinferior direction
- * In the middle ear cavity, the nerve may lie lateral and be anteriorly displaced

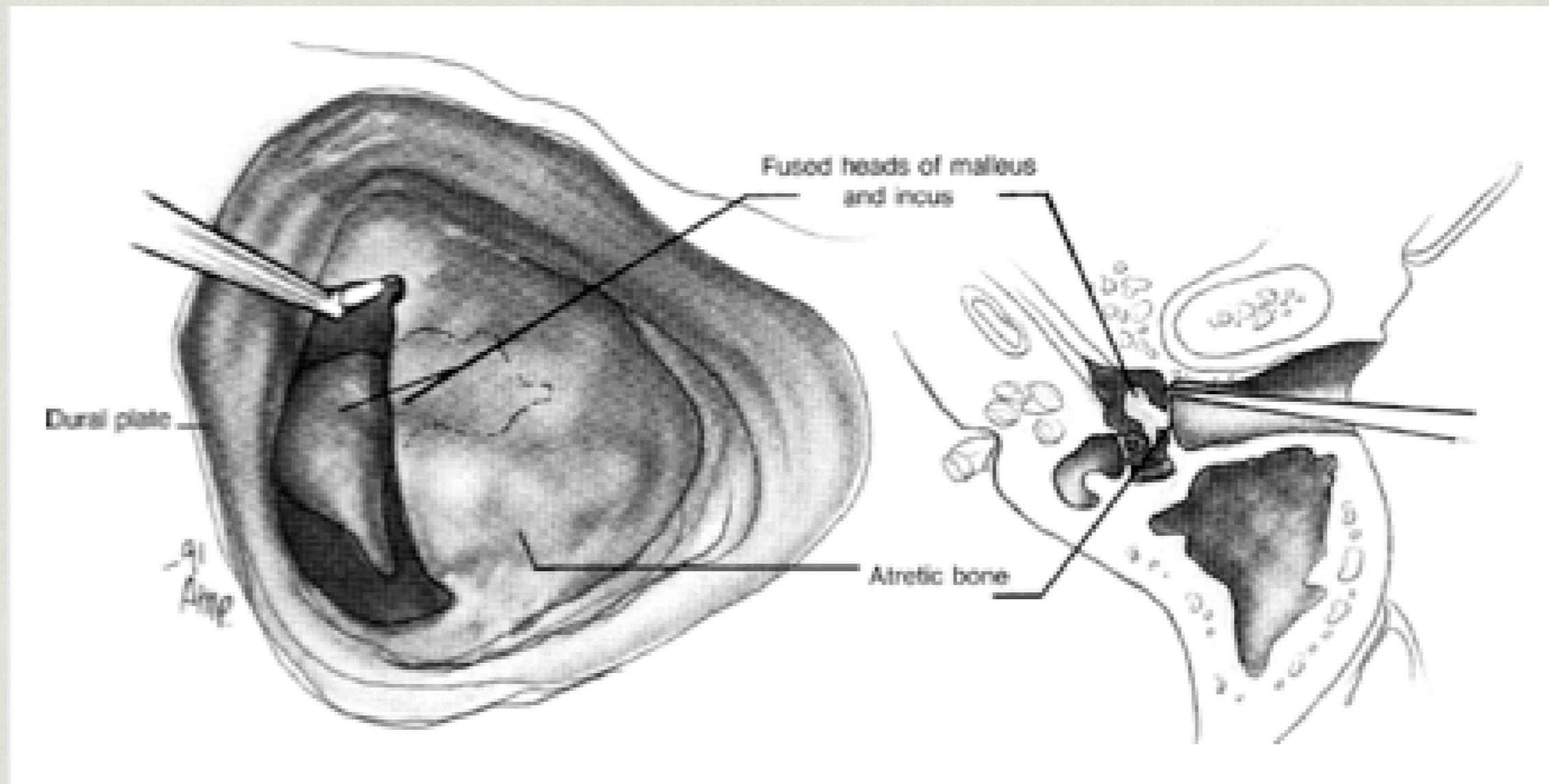


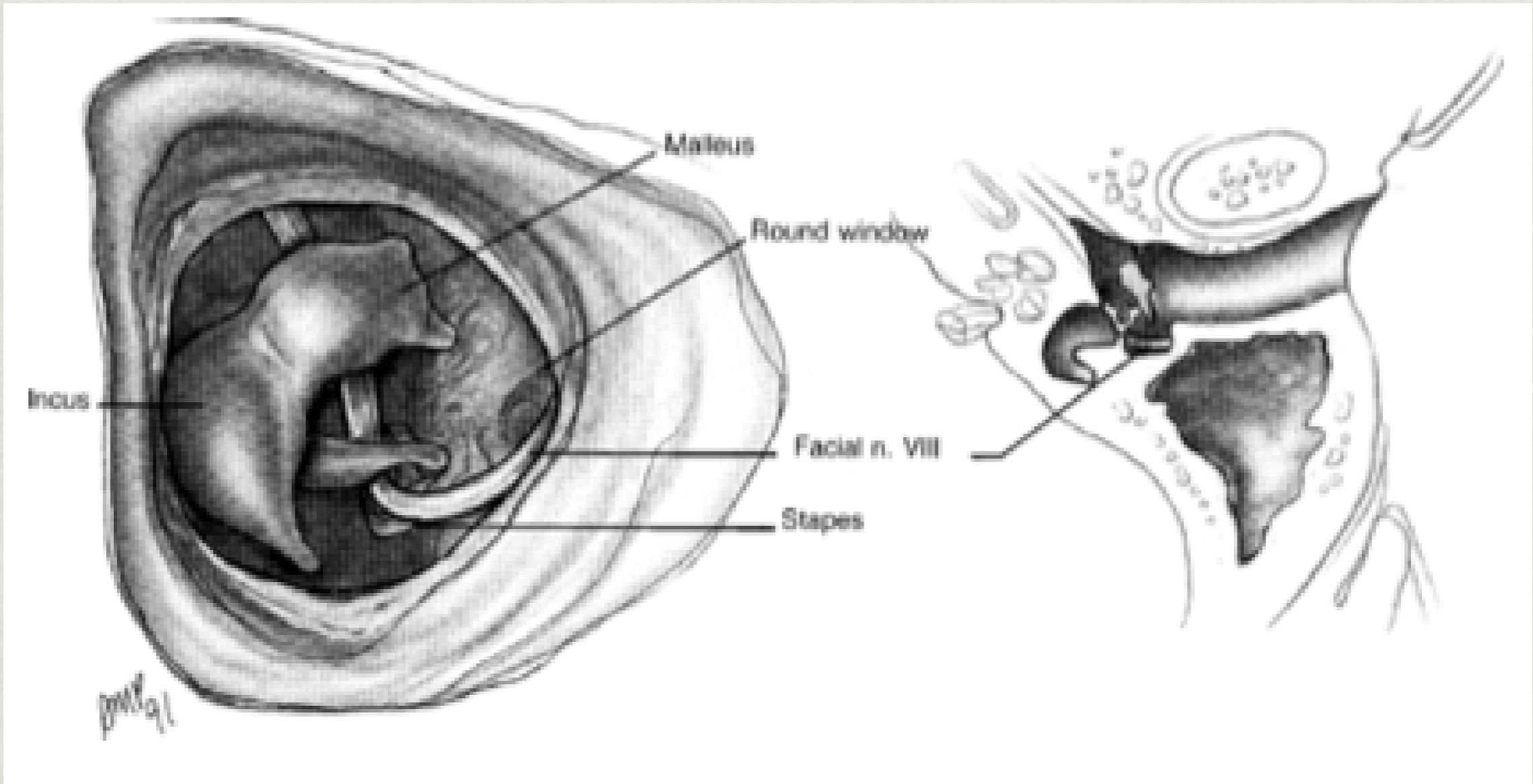




Exposure of Ossicular Chain

- * Malleus neck or manubrium often fused to atretic bone
- * In order to free the ossicular chain, the overlying bone is carefully thinned then removed
- * Limit drilling or manipulation of ossicles to protect to the inner ear
- * Except for fossa inudis, which may be left intact, bone should be completely removed around the ossicles, leaving at least 2-3 mm space
- * Atretic bone is removed leaving the ossicular mass centered in the new canal
- * To ensure proper draping of STSG, the canal walls should be smooth and without ledges lateral to the ossicular mass





Middle Ear Surgery

- * Stapes often obscured by contracted middle ear cavity, malformed lateral ossicles, or overlying facial nerve
- * Enough usually seen in order to assess the IS joint
- * Stapes is often small, with a delicate misshapen crura, but normal oval window and stapes footplate are usually found
- * Lateral ossicular chain, although deformed, is often mobile and left in place for better hearing results

Tympanic Membrane Grafting

- * Thin fascia graft is placed over the mobilized ossicular chain
- * Manubrium of malleus often deformed making anchoring the graft beneath the ossicular chain difficult
- * Absence of TM graft remnant or annulus predisposes to lateralization
 - * To prevent this the graft can be tucked under the anterior and superior edges of the bony wall or by covering the fascia graft with the STSC of the canal and then placing a Silastic button which has been contoured to the canal

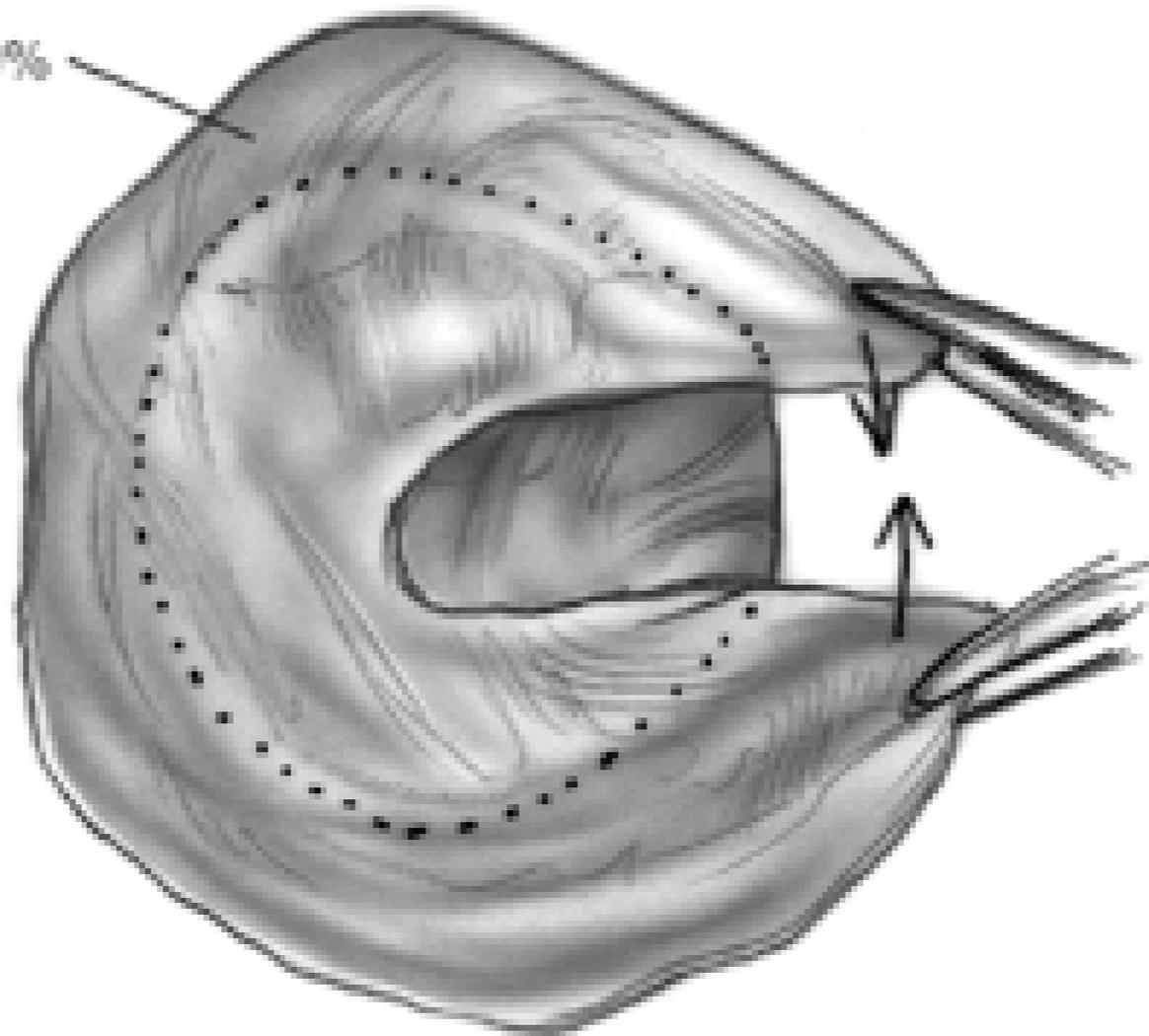
Meatoplasty

- * Auricle undermined and soft tissue debulked at approximate area of meatus, preventing future stenosis
- * Extensive undermining can place the facial nerve at risk as prior auricular reconstruction may have caused scarring and tethering of the extratemporal facial nerve in a more superficial location

Skin Grafting

- * 0.01-inch thick, pie-crusting, graft from thigh or upper arm used to line the canal
- * Typically shaped like a hexagon 4x6cm with small wedges taken from the medial portion to facilitate placement at the TM
- * Overlaps the fascia graft, which helps facilitate epithelialization of the neo-TM
- * Sialastic disc placement helps prevent lateralization
- * Packing then placed

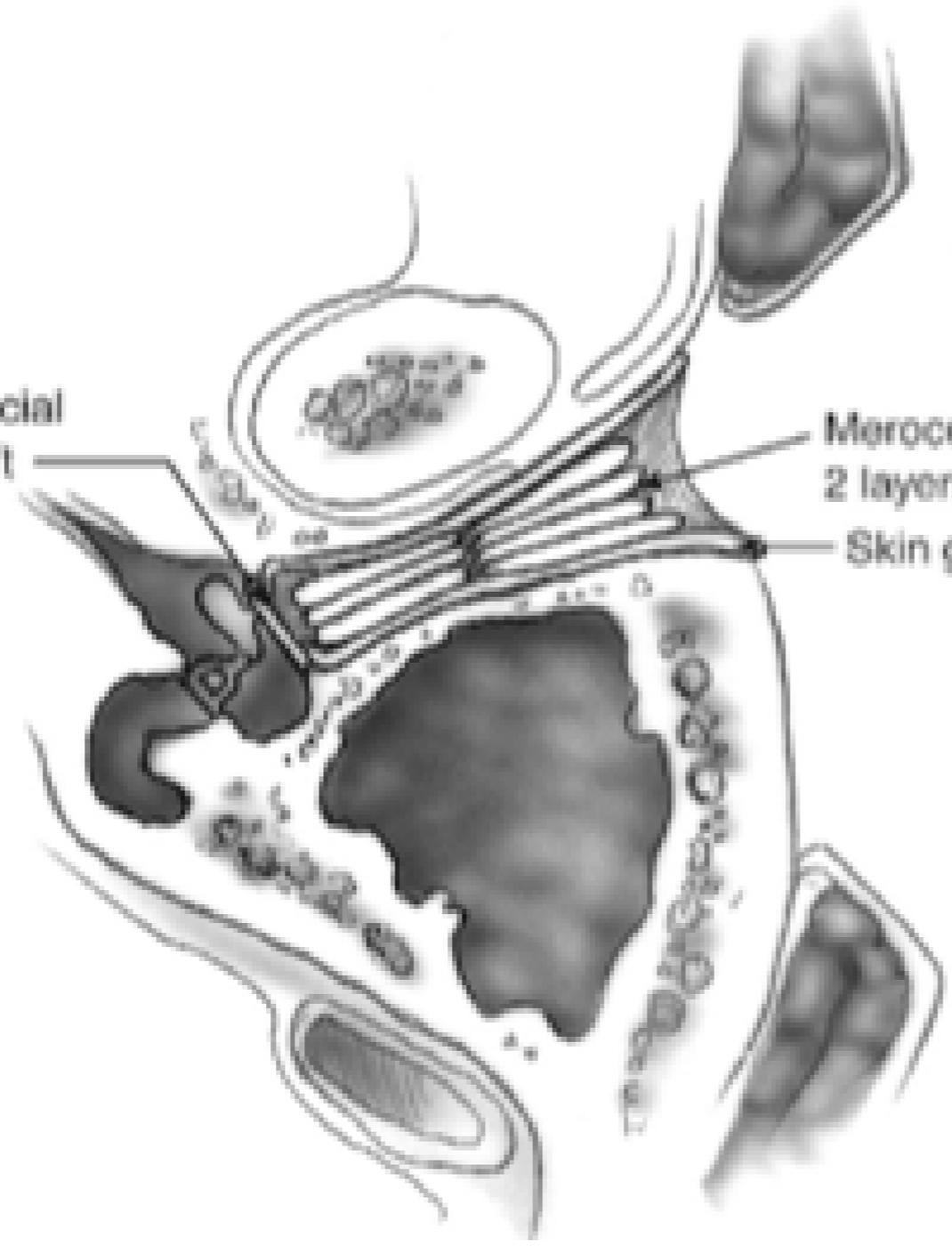
**Skin graft
overlaps 80%
of TM**



Fascial
graft

Merocel wicks
2 layers

Skin graft

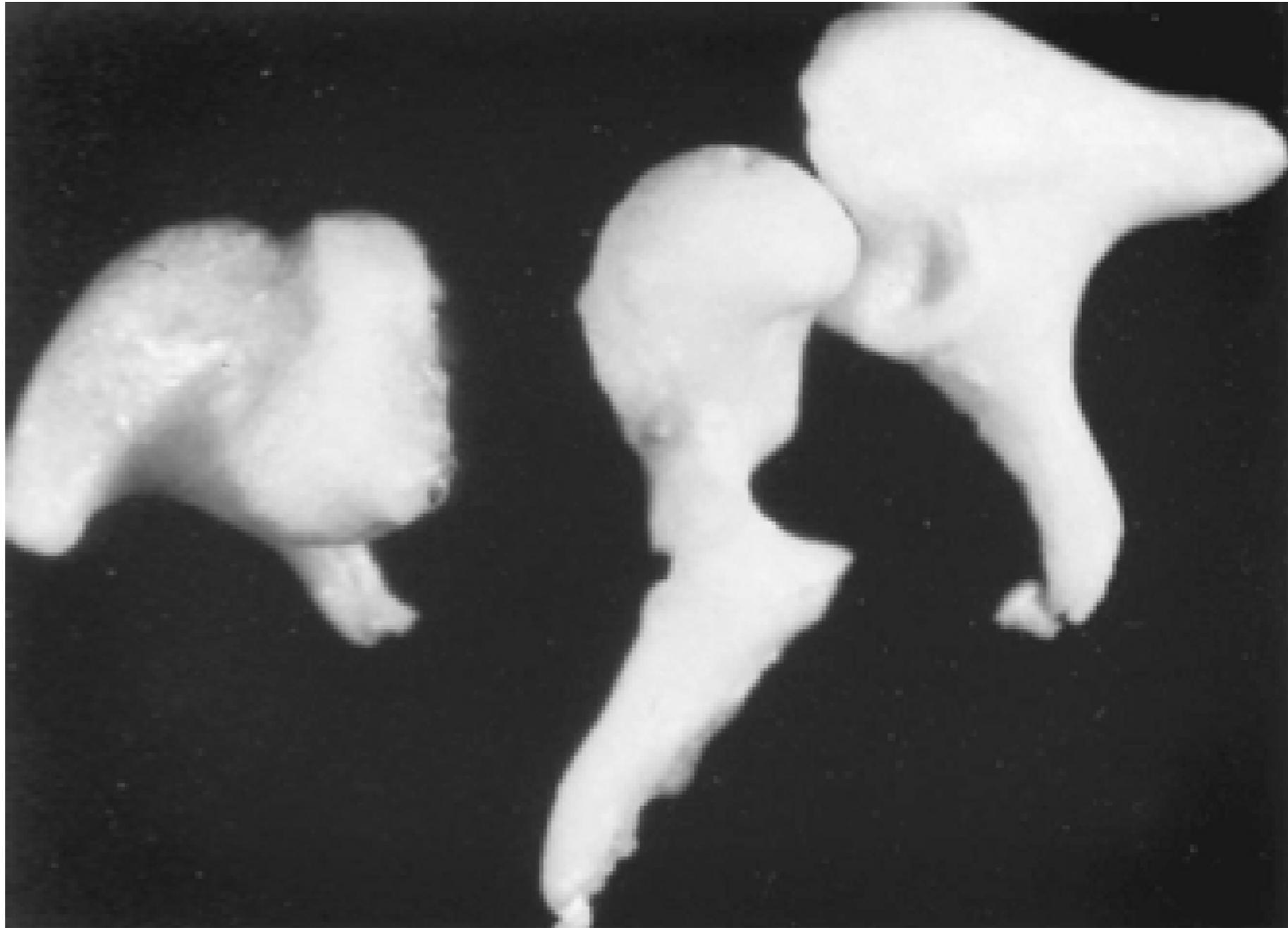


Postoperative Care

- * POD 10 packing and silastic disc are removed
- * If granulation tissue present, antibiotic-soaked gelfoam is placed in the canal and the patient is given drops for 7-10 days

Ossicles

- * Malleus/Incus Complex-malleus more deformed than the incus and has a short manubrium
- * Ossicular fixation usually between malleus neck or shortened manubrium and atretic bone
- * Fixation of stapes, or severe deformity, more likely to be seen in patients with a patent EAC than in those with major atresia



Hearing Results

- * Initial postoperative hearing level of 30dB or better can be achieved in approximately 50% of cases
- * Most of these patients continue to have improved SRT with long-term follow-up

Complications

- * Serious: SNHL and facial nerve injury
- * Other: Canal stenosis, chronic infection, recurrent CHL
- * Many of these patients require revision surgery (approximately 1/3). Stenosis and lateralization are the most common problems encountered.

Labyrinthine Injury

- * Anterior approach limits exposure of mastoid air cells and the horizontal SCC.
- * High-frequency SNHL noted in some patients postoperatively, but loss in the speech frequencies is rare.
- * Avoid by limiting drilling near the ossicles or excessive manipulation

Facial Nerve Injury

- * Facial Nerve monitoring is necessary
- * Usually occurs with transposition of the facial nerve to gain access to the oval window
- * Jahrsdoefer and Lambert: 1% incidence in 1000 cases
- * Concentrate drilling superiorly along MCF dural plate, with caution when enlarging the canal in the posterior-inferior direction
- * Nerve can also be injured in the extratemporal segment when making the postauricular incision or undermining the auricle

Canal Stenosis

- * Occurs in 25% of cases
- * If a large meatus is made, then not an issue
- * Significant stenosis can trap squamous epithelium and causing infection
- * Can attempt to dilate the canal with soft or hard stents, but usually ineffective, and a secondary meatoplasty with STSF is necessary
- * Pinna can become displaced, usually anteriorly or inferiorly, causing a malalignment of the meatus and bony canal. A permanent suspension suture can be placed from the framework of the auricle to the mastoid periosteum or to a hole in the mastoid cortex

Chronic Infection

- * Normal migration of keratin debris is absent in the skin-grafted ear canal as well as protective secretions from sebaceous and apocrine glands
- * Higher incidence of canal infections
- * Widely patent meatus and membranous canal are required for aeration and cleaning
- * Most patients not restricted from water activities

Conductive Hearing Loss

- * Persistent or recurrent
- * Most common negative outcome in aural atresia surgery
 - * Inadequate mobilization of the ossicular mass from the atretic bone
 - * Unrecognized IS joint discontinuity
 - * Fixed stapes footplate
- * Recurrent CHL usually from refixation or TM lateralization

BAHA

- * Bone anchored hearing aid is an alternative, especially for marginal or poor surgical candidates
- * Osseointegrated titanium fixture with a sound processor attached percutaneously
- * Allows more efficient transfer of sound than with bone conduction HA

Conclusion

- * Goal in congenital ear surgery is to create a functional pathway by which sound have reach the cochlear fluids
- * Challenging surgery requiring thorough knowledge of the anatomic variations, audiometric and radiographic findings in these patients
- * With proper patient selection in cases of major atresia, it is possible to achieve a hearing level of 25 dB or better in approximately 50-70% of patients