

Congenital Aural Atresia

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Introduction

- ❧ Birth defect characterized by hypoplasia of EAC, middle ear, and inner ear
- ❧ Rate of occurrence 1 in 10,000 to 1 in 20,000 live births
- ❧ Unilateral atresia - 3 times more common than bilateral, right side and male more common
- ❧ Severity can range from mild to severe
- ❧ Otologist should create functional conductive pathway for sound while preserving facial nerve and labyrinth function

Embryology

- ✎ Inner, middle, and external ear develop independently
- ✎ First and second branchial arches fuse to form the six hillocks which fuse to form the auricle
- ✎ External ear canal and tympanic membrane form from first branchial cleft
- ✎ Initially represented by a solid core of epithelial cells that extends down to the tympanic ring and first pharyngeal pouch
- ✎ Absorption of the epithelial cells begins in seventh month of gestation and proceeds medial to lateral

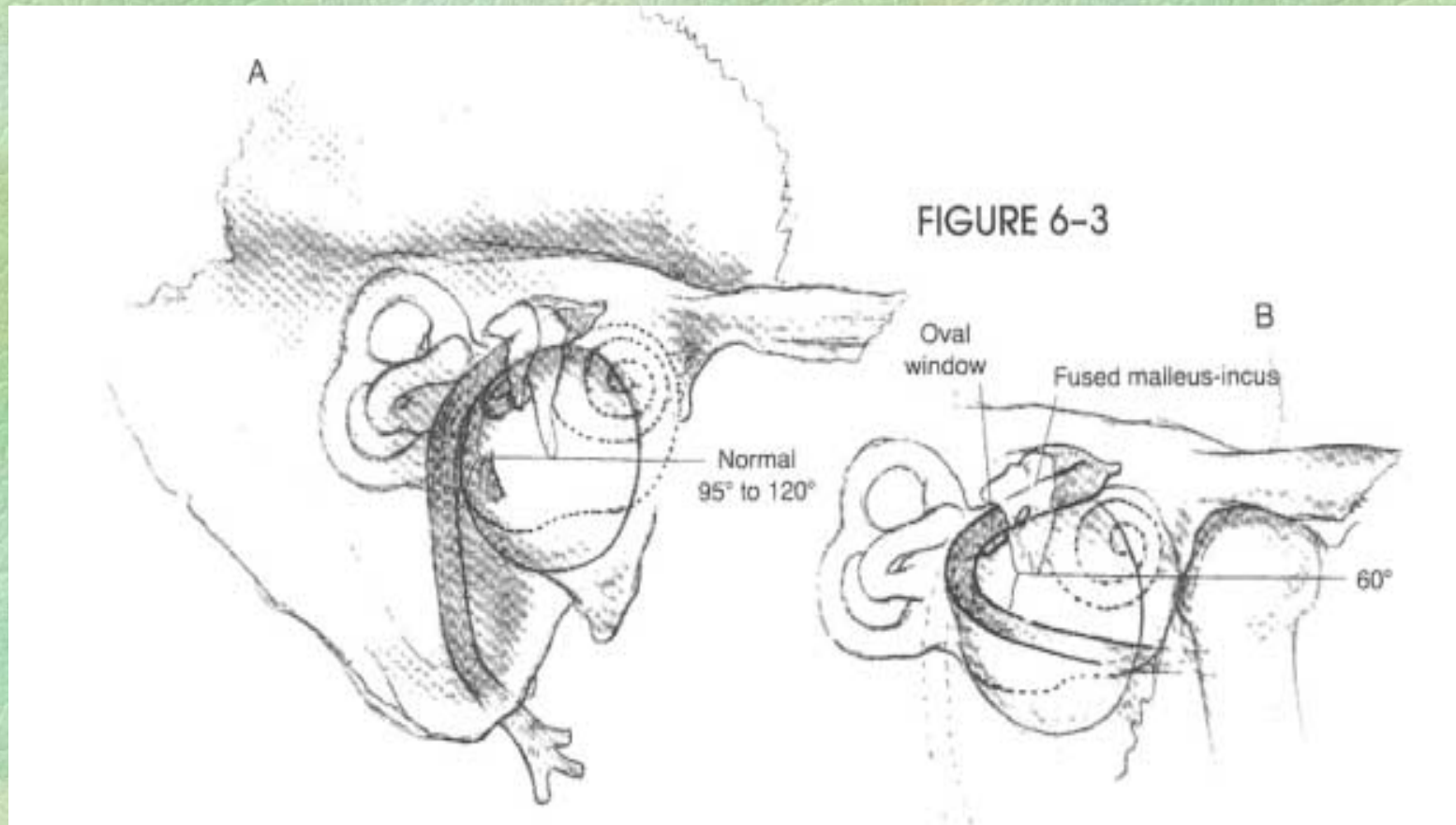
Embryology

- ✎ Medial portion of EAC formed by tympanic bone which ossifies to form the tympanic ring and osseous ear canal
- ✎ Eustachian tube, middle ear, and mastoid air cells are derived from the first pharyngeal pouch
- ✎ Pneumatization of the mastoid - late embryological event
- ✎ Pharyngeal pouch grows outward to form the middle ear cleft
- ✎ Ossicular chain development begins within first 4 weeks
- ✎ Ossicles (except for footplate) come from the first and second branchial arches

Embryology

- ✎ Bony fusion of incudomalleolar joint in atresia
- ✎ Malleus is more deformed and head is fixed to atretic plate
- ✎ Stapes footplate from otic capsule, usually normal
- ✎ Facial nerve develops from acoustico-facial primordium
- ✎ It begins more anterior and superior than in the adult
- ✎ Nerve found to be displaced in 25% to 30% of cases
- ✎ Typically makes acute angle at the second genu and crosses the middle ear in a more anterior and lateral position
- ✎ Correlation between the degree of microtia and the extent of facial nerve abnormality has been noted

Facial Nerve Course



Embryology

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Classification

- ❖ Altmann developed classification in 1955
- ❖ Three groups
- ❖ Based on anatomical variations

Group I:	Small external ear canal, hypoplastic tympanic bone and tympanic membrane with a normal or contracted middle ear space. Ossicles may be normal or malformed.
Group II:	The canal is absent, with an atretic plate present. The middle ear space is small. The malleus and incus are fixed and malformed.
Group III:	Includes those with absent external auditory canal with a contracted or absent middle ear space. The ossicles may be absent.

Classification

- ✿ De La Cruz modified this system using only Altmann groups 2 and 3 for more practical surgical guidelines
- ✿ Divided them into major and minor categories
- ✿ Minor malformations:
 - normal mastoid pneumatization, oval window, and inner ear
 - reasonable relationship between facial nerve and oval window
- ✿ Major malformations:
 - poor pneumatization, abnormal or absent oval window
 - abnormal facial nerve course, abnormal inner ear

Classification

- ✎ Schuknecht - system based on surgical observations
- ✎ **Type A or meatal atresia:**
 - limited to fibrocartilagenous portion of canal
 - predisposed to cholesteatoma formation
- ✎ **Type B or partial atresia:**
 - narrowed bony and cartilaginous portions, ossicular deformities
 - TM smaller and part replaced with bony septum
- ✎ **Type C or total atresia:**
 - absent bony canal, ossicular malformations, missing TM, pneumatized mastoid
- ✎ **Type D or hypopneumatic total atresia:**
 - same as type C, but with poor mastoid pneumatization

Classification

- ❧ Jahrsdoerfer developed point rating system
- ❧ Based on anatomic variations on CT scan
- ❧ Used to select surgical candidates

TABLE 6-2. Grading System of Candidacy for Surgery of Congenital Aural Atresia

PARAMETER	POINTS
Stapes present	2
Oval window open	1
Middle ear space	1
Facial nerve normal	1
Malleus-incus complex present	1
Mastoid well pneumatized	1
Incus-stapes connection	1
Round window normal	1
Appearance of external ear	1
Total available points	10

RATING	TYPE OF CANDIDATE
10	Excellent
9	Very good
8	Good
7	Fair
6	Marginal
≤5	Poor

From Jahrsdoerfer RA, Yeakley JW, Aguilar EA, et al.: Grading system for the selection of patients with congenital aural atresia. *Am J Otol* 13(1): 6-12, 1992.

Classification

- ❧ Chiossone (1983) presented classification based on location of the glenoid fossa
- ❧ **Type I:** fossa is normal
- ❧ **Type II:** fossa is moderately displaced
- ❧ **Type III:** fossa overlaps the middle ear
- ❧ **Type IV:** type III & poor mastoid pneumatization
- ❧ Types I and II good surgical candidates, type III tendency for lateralization, type IV no surgery

Patient Evaluation

- ✎ Most cases evident at birth
- ✎ All patients require formal assessment
- ✎ Assess neurological milestones
- ✎ Evaluated overall craniofacial development - first and second branchial arches
- ✎ Assess degree of aural development, presence of tympanic membrane, atresia vs. stenosis
- ✎ Assess for facial nerve weakness
- ✎ Assess overall hearing status and need for amplification

Patient Evaluation

- ✎ CT scan of the temporal bone to assess surgical candidacy
- ✎ Scanning can wait until age 4 or 5 unless SNHL is present
- ✎ Decision to operate depends primarily on development of middle ear
- ✎ Evaluate course of facial nerve, development of cochlear and vestibular labyrinths on CT
- ✎ CT needed to evaluate for cholesteatoma formation
- ✎ Surgery should be delayed until pneumatization of the temporal bone is complete

Medical Management

- ❧ Audiologist can provide amplification and assistive auditory devices, lip reading instruction, special education, and parental guidance
- ❧ Preferential seating in school
- ❧ Early amplification in patients who need it and in bilateral atresia children is essential
- ❧ Patients with unilateral atresia and normal hearing in the other ear need no medical intervention
- ❧ Controversy exists in correction of unilateral atresia cases

Management

- ❧ Concern over unilateral atresia is the degree and predictability of hearing improvement that can be achieved, potential lifetime care of mastoid cavity, and risk to the facial nerve
- ❧ Some surgeons will delay until adulthood
- ❧ Improvement in hearing to 25dB eliminates the handicap of unilateral hearing loss
- ❧ Recent trend is to operate on properly selected patients at the age of 5 or 6 because of the importance of binaural hearing
- ❧ Bilateral atresia should be operated on to restore hearing so amplification is no longer needed
- ❧ Usually operate as child approaches school age
- ❧ Patients with cholesteatoma should always undergo surgery

Surgical Candidates

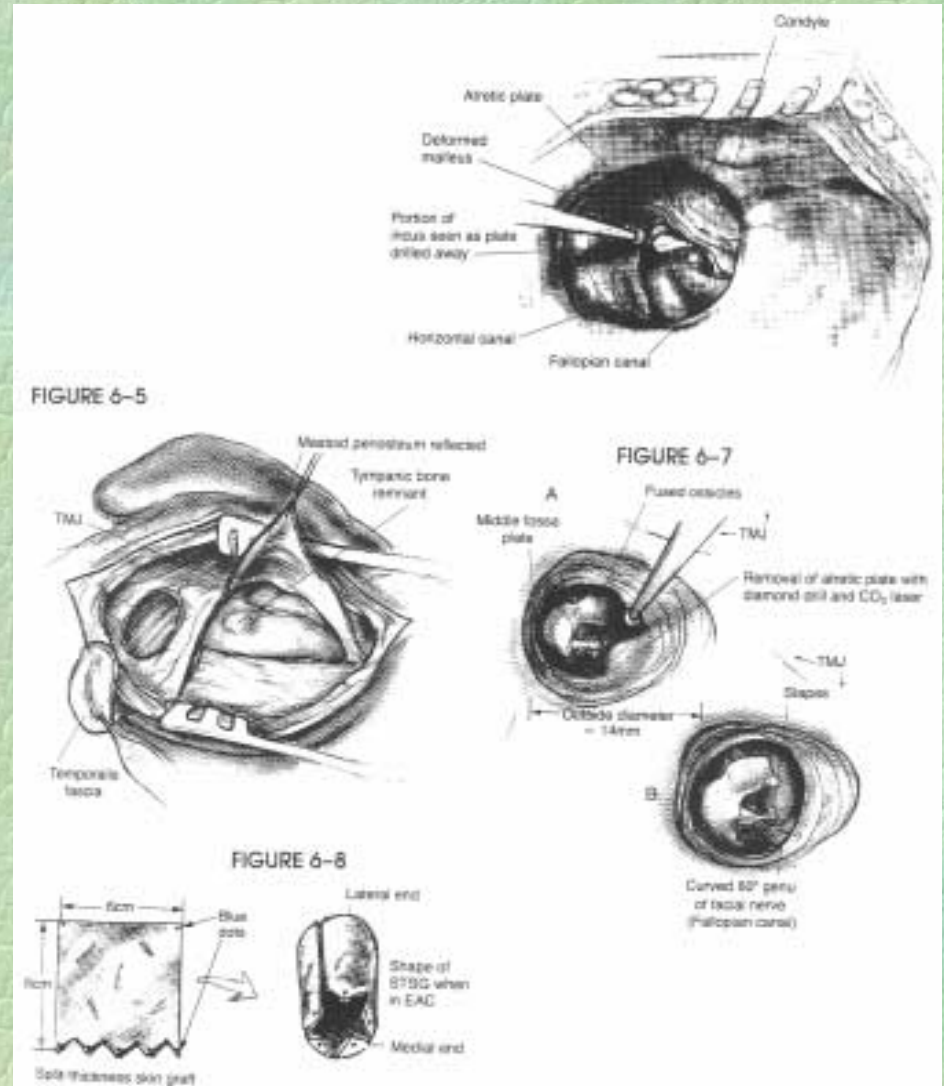
- ❖ Normal bone conduction thresholds and good speech discrimination with no inner ear abnormalities
- ❖ Patients with CT scan revealing little or no middle ear and mastoid pneumatization are not surgical candidates

Surgical Correction

- ✂ Two basic surgical approaches:
 - Mastoid approach
 - Anterior approach
- ✂ General anesthesia with no paralysis
- ✂ Facial nerve monitoring used on all cases
- ✂ Skin graft site and ear prepped and draped
- ✂ Postauricular incision made taking care not to expose grafted cartilage
- ✂ Periosteum is elevated exposing the mastoid cortex and temporomandibular joint

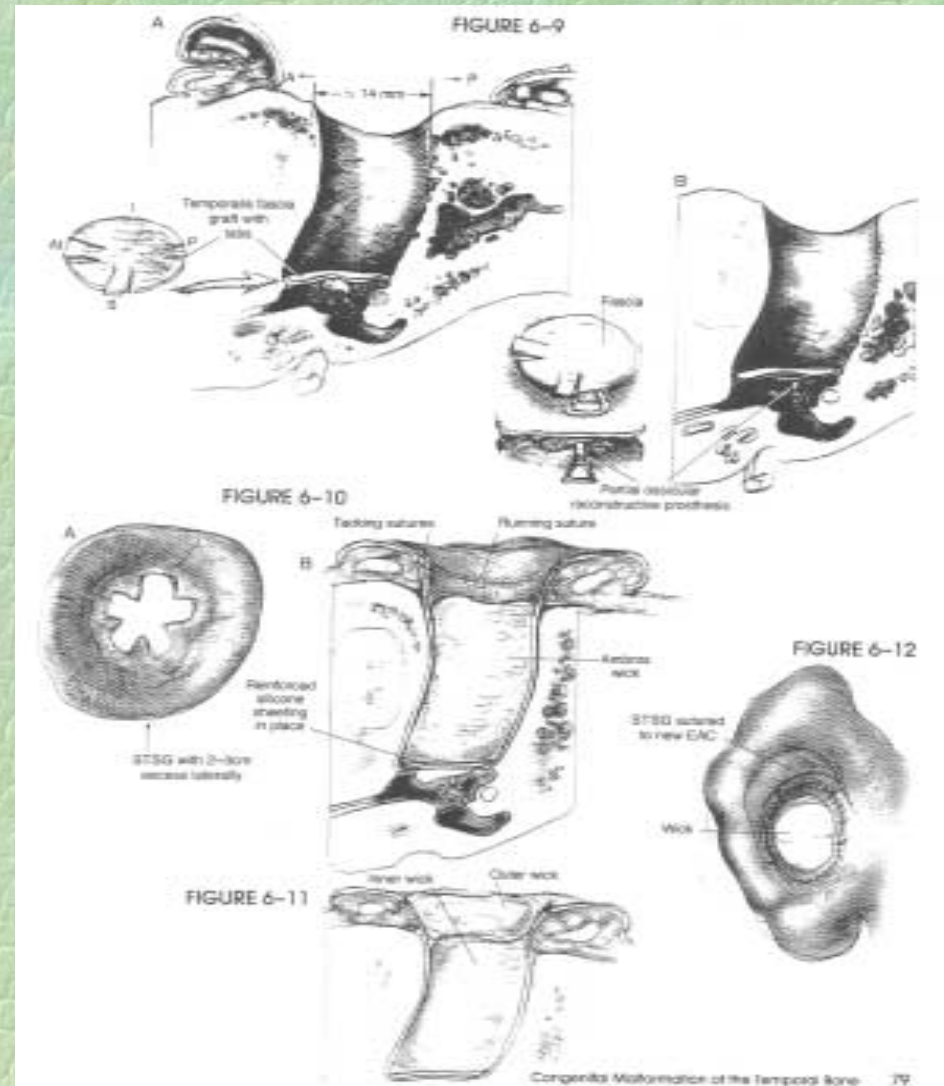
Anterior Approach

- Drilling - middle cranial dura superior landmark and TMJ anterior landmark
- Dissection-anterior and medial
- Epitympanum and fused malleus-incus identified
- Facial nerve always medial to ossicular mass in middle ear
- Facial nerve vulnerable when the canal enlarged in the posterior and inferior direction



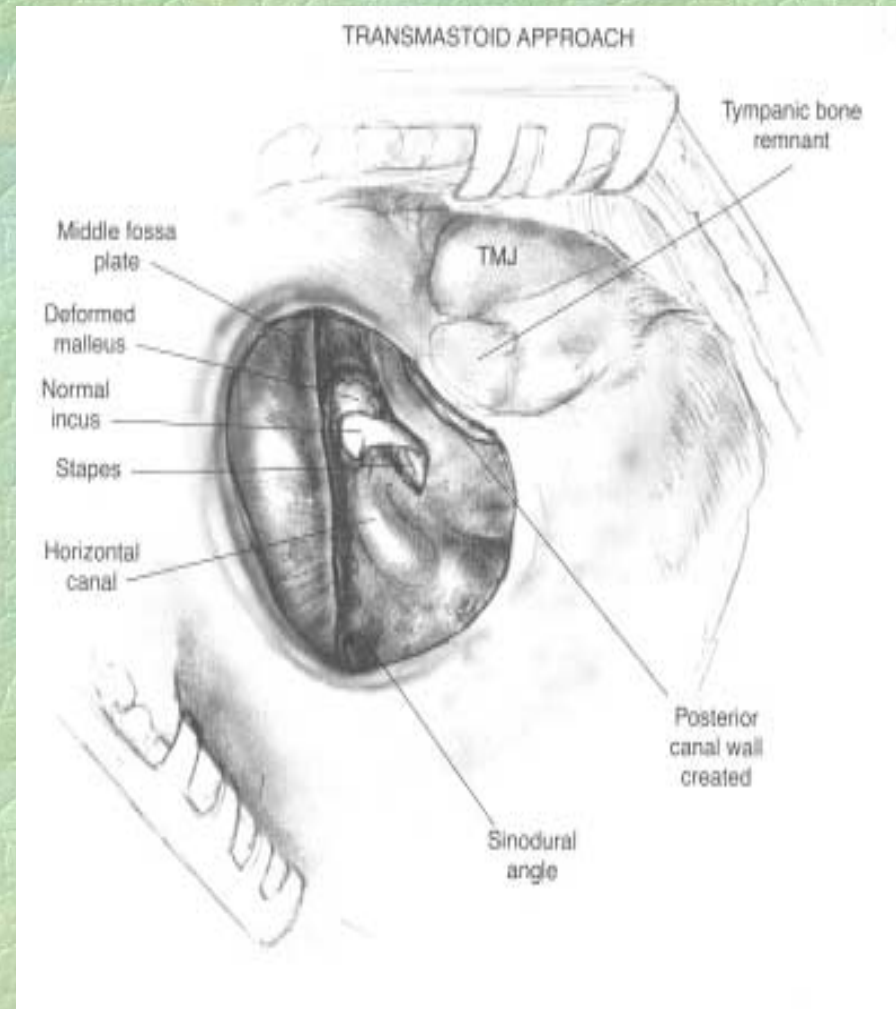
Anterior Approach

- Canal -1.5 times normal
- Ossicular mass dissected free of atretic plate (2-3mm)
- Fibrous ligaments -CO2 laser
- Ossiculoplasty
- Fascia -tabs cut, over ossicles
- Skin graft (6x6cm) lines canal
- Canal packed
- Meatoplasty - debulk soft tissue, twice normal size, limit subQ tissue, check alignment, suture skin graft, pack



Transmastoid Approach

- Not used for many years
- Exposure of glenoid fossa and mastoid cortex
- Drilling -sinodural angle followed medially to the antrum
- Identify -LSC, malleus-incus complex, and atresia plate
- Facial ridge is lowered and creation of EAC with CWD technique
- Bone pate or soft tissue used to obliterate cavity before grafted



Postoperative Care

- ✿ 5 day course of antistaph antibiotics
- ✿ Mastoid dressing removed postoperative day 1
- ✿ At three weeks packing is removed
- ✿ Repack the canal if necessary
- ✿ Audiogram taken at 6-8 weeks
- ✿ If meatus is narrowing - dilate with wicks

Results

- ✿ Initial postoperative hearing level of 30dB or better in 50-75%; hearing level of 20dB or better in 15-50%
- ✿ Schuknecht - 30dB in 50%; 30% with 20dB or better
- ✿ De La Cruz - 53% with 20dB or better in two series
- ✿ Jahrsdoerfer - 65% had PTA of 30dB or less; more recent series with 71% with hearing level 25dB or better
- ✿ Lambert - 67% patient had SRT of 30dB or better and mean improvement in hearing was 30dB

Complications

- ❧ **Labyrinthine injury** -HFSNHL caused by direct manipulation of ossicles and acoustical energy from the drill
- ❧ **Facial nerve injury** -abnormal development of temporal bone places the nerve at risk, temporary paresis seen when nerve has been transposed to gain access to oval window, minimize risk of injury by using nerve monitor and adhering to surgical guidelines
- ❧ **Stenosis** -develops in as many as 25% of patients, make large enough meatus to start, prevent granulation, watch for shifting of pinna, treat with secondary meatoplasty
- ❧ **Canal infections** -incidence increased because normal keratin migration gone and protective secretions absent, anterior approach minimizes debris accumulation, widely patent meatus key
- ❧ **Persistent or recurrent conductive hearing loss** -caused by inadequate mobilization of ossicular mass, joint discontinuity, fixed stapes, lateralization of graft

Case Presentation

- ❧ 7 year old male with a history of right ear microtia and atresia is referred from your friend the plastic surgeon to see if his hearing can be improved.
- ❧ History of congenital microtia and atresia, no ear infections, normal developmental milestones, does well in school, but the kids tease him.
- ❧ No family history, PSHX - ear reconstruction surgeries, no other medical problems

Case Presentation

- ❖ Physical examination reveals complete atresia of the right auditory canal with buried cartilage in good position
- ❖ No craniofacial abnormalities
- ❖ Facial nerve intact and symmetrical
- ❖ Weber lateralizes to the right, Rinne AD-BC > AC; AS-AC > BC

Audiogram

