Introduction
1. Atresia of ear canal with middle ear anomaly can occur in isolation or with microtia or craniofacial dysplasia
2. Otolaryngologist can expect to be called on to evaluate this defect several times in the average career

Epidemiology
1. 1/10000-20000
2. Bilateral in 1/3 cases
3. Male >Female
4. right side > left side
5. Degree of auricular malformation usually correlates with degree of middle ear deformity
6. Inner ear abnormalities incidence is low

Embryology
1. Auricle
   a. Begins at 4th week of gestation to 12th weeks
   b. 1st and 2nd branchial arches
   c. Hillock of His fusion: 1=>tragus  2=> helical crus  3=> helix  4=>anti-helix  5=>anti-tragus  6=> lobule
   d. Lies in the relative adult position at 20wks
2. External canal
   a. Begins at 8\textsuperscript{th} week of gestation
   b. From first branchial cleft
   c. Epithelial cells solid core medial migration
   d. Recanalizing at 6\textsuperscript{th} to 7\textsuperscript{th} month of gestation, medial to lateral (most inner, middle and outer ear are well differentiated)
   e. Canal cholesteatoma if external canal process arrested

3. Ossicles
   a. 1\textsuperscript{st} and 2\textsuperscript{nd} brachial arch (except vestibular portion of stapes footplate)
   b. Meckel’s cartilage(1\textsuperscript{st} branchial arch): Malleus head and neck, Incus body and short process
   c. Reichert’s cartilage(2\textsuperscript{nd} branchial arch): Malleus handle, Incus long process, Stapes suprastructure, tympanic surface of footplate
   d. Stapes footplate is usually developed (formed from otic capsule)
   e. It is uncommon to encounter fixed stapes footplate in major congenital ear formation, but the suprastructure is commonly deformed

\textbf{FIGURE 128.3.} Origin of the ossicles—two interpretations.
4. Labyrinth
   a. 3<sup>rd</sup> week of gestation
   b. Invagination of otic placode to form otic
   c. 6<sup>th</sup> week of gestation: semicircular canal
   d. 8<sup>th</sup> week of gestation: Utricle and saccule
   e. 7-12 week of gestation: Cochlea
   f. Sensorineural function and vestibular function should be normal

5. Isolated branchial arch or cleft deformity is possible but usually combined

6. Facial nerve
   a. 4-5 weeks of gestation
   b. Abnormality is common in aural atresia
   c. Typically, it makes an acute angle at 2<sup>nd</sup> genu
   d. The nerve crosses the middle ear cavity in more anterior-lateral direction=>
      exit into glenoid fossa
**Classification**

1. **Schuknecht**
   
   - **Type A:** meatal atresia limited to fibrocartilagenous portion of canal
   
   - **Type B:** partial atresia with narrowed bony and cartilaginous canal
   
   - **Type C:** Total atresia with absent bony canal, ossicular malformation, missing tympanic membrane and pneumatized mastoid
Type D: Same as type C, but poor mastoid pneumatization

2. **Jahrsdoerfer**
   a. Based on HRCT
   b. Score related to success of surgery

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Points</th>
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<tbody>
<tr>
<td>Stapes present</td>
<td>2</td>
</tr>
<tr>
<td>Oval window open</td>
<td>1</td>
</tr>
<tr>
<td>Middle ear space</td>
<td>1</td>
</tr>
<tr>
<td>Facial nerve normal</td>
<td>1</td>
</tr>
<tr>
<td>Malleus-incus complex present</td>
<td>1</td>
</tr>
<tr>
<td>Mastoid well-pneumatized</td>
<td>1</td>
</tr>
<tr>
<td>Incus–stapes connection</td>
<td>1</td>
</tr>
<tr>
<td>Round window normal</td>
<td>1</td>
</tr>
<tr>
<td>Appearance of external ear</td>
<td>1</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Rating</th>
<th>Type of candidate</th>
</tr>
</thead>
<tbody>
<tr>
<td>10</td>
<td>Excellent</td>
</tr>
<tr>
<td>9</td>
<td>Very good</td>
</tr>
<tr>
<td>8</td>
<td>Good</td>
</tr>
<tr>
<td>7</td>
<td>Fair</td>
</tr>
<tr>
<td>6</td>
<td>Marginal</td>
</tr>
<tr>
<td>≤5</td>
<td>Poor</td>
</tr>
</tbody>
</table>
3. De la Cruz
   a. Major:
      i. Poor pneumatization
      ii. Absent or abnormal oval window
      iii. Abnormal horizontal facial nerve
      iv. Anomalous inner ear

   b. Minor
      i. Normal mastoid pneumatization
      ii. Normal oval window
      iii. Reasonable oval window-facial nerve relationship
      iv. Normal inner ear
**Audiologic evaluation**

a. Patient with unilateral microtia and CAA usually have normal hearing in the contralateral ear
b. Conductive hearing loss: 80%-90% of pts, Threshold is expected to be 40-60 dB
c. Sensorineural hearing loss: 10%-15%
d. Non-microtic ear passes the newborn hearing screen, additional test can be done until 6-7 months old

1. ABR
   a. within 3 months of birth
   b. Air conduction: maximal at 60db
   c. Bone conduction: usually normal
   d. If bilateral hearing loss => Bone conduction hearing aid and speech therapy

2. BOA
   a. ABR unilateral hearing loss or normal
   b. Performed at 6 months old
   c. Hearing aid is not needed traditionally

**Computed tomography**

1. Age 5-6 years old
2. Axial: malleus, incus, IS joint, Round window
3. Coronal: Stapes, oval window, vestibule
   a. Middle ear and mastoid pneumatization
   b. Ossicles anatomy
   c. Inner ear morphology
   d. Course of facial nerve

**Surgical candidate**

1. Absolute requirement
   a. normal inner ear
   b. normal cochlear function
2. Jahrsdoerfer score > 6
3. Bilateral aural atresia cases
4. Best ear should be first
5. Unilateral aural atresia could be delayed to adult and only those ideal candidates
6. Only 60% patients are candidates
7. Associated with other syndromes (Goldenhar, Treacher-Collins, Crouzon…): not surgical candidates
Timing of surgery
1. Unilateral: could be delayed to adult
2. Bilateral: 6 y/o
3. The surgery for microtia should be at least two months prior

Surgical technique
1. Transmastoid approach: rare
2. Anterior approach
   a. Post-auricular incision
   b. Subcutaneous tissue and periosteum raised to glenoid fossa
   c. Drilling continues anteriorly and medially to the epitympanum
   d. Drill until new canal about 10mm
   e. Ossicular reconstruction
   f. Tympanic membrane is created
   g. Split thickness skin graft is used to line the new EAC
   h. Meatoplasty is created to augment the opening
   i. Reduction in diameter of new canal by 30% is normal post-operatively

Surgical results
Laryngoscope, 116: October 2006
Analysis of Long-Term Hearing Results after the Surgical Repair of Aural Atresia
Sun O. Chang M.D. et al
1. 50% of patients are candidates
2. Jahrsdoerfer score 6 or >6
3. Success: AB gap <30 at 6 months and 3 year follow up
4. 1/3 fail to achieve desirable results

Otology and neurotology, 28:54-60, 2006
Congenital aural atresia: review of short and long term surgical results
G. Paul Digoy and †Roberto A. Cueva
1. Short term < 1yr  Long term > 1yr
2. Successful hearing: post-op ABG 20dB or less, SRT 30dB or less, PTA 30dB or less
3. 1/2 fail to achieve successful result
Complications of congenital aural atresia repair
1. Lateralization of tympanic membrane graft (22-28%)
2. Stenosis of Canal (8-20%)
3. temporomandibular joint pain and trauma (2%)
4. Facial nerve damage (1%)
5. Sensorineural hearing loss (2-5%)

Medical treatment
1. Bone conduction hearing aid

2. Bone anchored hearing aid (BAHA)
   a. Unilateral hearing loss: to gain sound localization and improved speech perception in noise
   b. No interference to the normal ear function
   c. For Bilateral congenital aural atresia inoperable, application of bilateral BAHAs is the only means for them to receive binaural hearing
   d. 2-10 y/o
   e. Surgically-implanted, percutaneous titanium screw fixture
   f. Osseointegrates into the temporal bone

3. BAHA surgery
   a. One stage surgery or two stage surgery
   b. Complications limited: 1. local infection / inflammation (7.5%)
      2. failure to osseointegrate (2.5%)
Results of the Bone-Anchored Hearing Aid in Unilateral Hearing Loss
Jack J. Wazen, MD; Jaclyn Spitzer, PhD; Soha N. Ghossaini, MD; Ashutrosh Kacker, MD; Anne Zschommler

TABLE I.
Preoperative Hearing Thresholds (in dB Hearing Level, HL).

<table>
<thead>
<tr>
<th>Frequency (Hz)</th>
<th>250</th>
<th>500</th>
<th>1000</th>
<th>2000</th>
<th>4000</th>
<th>8000</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean</td>
<td>68.89</td>
<td>68.30</td>
<td>68.89</td>
<td>74.40</td>
<td>70.44</td>
<td>97.30</td>
</tr>
<tr>
<td>SD</td>
<td>16.7</td>
<td>19.53</td>
<td>17.99</td>
<td>25.30</td>
<td>25.30</td>
<td>17.90</td>
</tr>
</tbody>
</table>

SD = standard deviation.

TABLE III.
Mean Gain* in Hearing Thresholds (dB).

<table>
<thead>
<tr>
<th>Frequency (Hz)</th>
<th>Mean</th>
<th>Range</th>
</tr>
</thead>
<tbody>
<tr>
<td>500</td>
<td>41.9</td>
<td>5–80</td>
</tr>
<tr>
<td>1000</td>
<td>60.3</td>
<td>40–100</td>
</tr>
<tr>
<td>2000</td>
<td>49.4</td>
<td>15–75</td>
</tr>
<tr>
<td>4000</td>
<td>45.6</td>
<td>20–80</td>
</tr>
</tbody>
</table>

*Preoperative unaided thresholds—postoperative BAHA-aided thresholds.
BAHA = bone-anchored hearing aid.

% Correct

PRE
POST

S1  S4  S11  S16  S21  S23  S27
4. BAHA soft-band
   a. For children too young for BAHA

   b. ![](image1)

   ![](image2)


**Canal atresia: “Surgery or implantable hearing devices?”**

<table>
<thead>
<tr>
<th></th>
<th>Reconstructed</th>
<th>BAHA 2-stage</th>
<th>BAHA 1-stage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hearing gain</td>
<td>17.3 dB per ear</td>
<td>31.8</td>
<td>31.8</td>
</tr>
<tr>
<td>Cost</td>
<td>$51506</td>
<td>$42449</td>
<td>$28341</td>
</tr>
<tr>
<td>Cost/dB</td>
<td>$2909</td>
<td>$1238</td>
<td>$826</td>
</tr>
</tbody>
</table>

**Conclusions**

1. Surgery of congenital aural atresia is challenging
2. The audiologic evaluation and image study are important
3. Immediate amplification if needed
Bibliography


3. Sun O. Chang, MD; Byung Yoon Choi, MD; Dong Gu Hur, MD, Analysis of the Long-Term Hearing Results after the Surgical Repair of Aural Atresia Laryngoscope, 116: October 2006


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