CONGENITAL AURAL ATRESIA
27th Alexandria International Combined ORL Congress
Alexandria, Egypt   April 8, 2009

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Epidemiology

- 1 in 10,000 - 20,000 births
- Unilateral to bilateral 3:1
- 10% associated with recognizable synd.
- Inherited in 5% of non-syndromic cases
Congenital Atresia EAC

18q 22.3-18q23 Region
Long arm Deletion

Congenital Aural Atresia
239 Patients - 302 Ears

141 Males
98 Females
108 Right
61 Left
70 Bilateral
Embryology

EXTERNAL EAR

- The external ear develops from the first and second branchial arches and the interposed first branchial cleft.
Auricle

- 4 weeks → condensation from 1st and 2nd arches form the hillocks of Hiss
- Hillocks fuse to form the auricle
- Adult configuration reached by 20 weeks
At 2 months, the 1st branchial cleft migrates medially to abut the lateral aspect of the 1st branchial pouch.

These 2 structures are then separated by mesenchyme.

A **solid core** of epithelium migrates from the rudimentary auricle to the lateral end of the first pharyngeal pouch and forms the tympanic plate.
First pharyngeal pouch

EAC

- Recanalization starts at the 6th month
- Proceeds in a medial to lateral direction
- Arrest at any stage leads to different degrees of atresia
**Tympanic Ring**

- The tympanic ring develops from four foci of ossifications.
- The mastoid grows in a posterior inferior direction, *carrying with it the middle ear structures and the facial nerve*.

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**Congenital Aural Atresia Surgery**

*Facial nerve variations*
Middle Ear

- Ossicles derive from the Meckel’s and Reichert’s cartilages
- This process starts at the second month and matures by the fourth month
- Lining of the middle ear derives from the 1st pharyngeal pouch, a process which is complete by the end of seventh month
STAPEDIAL ARTERY

- Artery of the 2nd branchial arch
- Appears during the 4th-5th week
- It forms the obturator foramen of the stapes
- It atrophies during the 3rd month
- Its dorsal branch forms the middle meningeal a.
- Its persistence -> absence of the foramen spinosum
Atresia Evaluation
Audiology

- Cochlear function: Pure Tone audiometry
- Complete atresia causes 50 to 60 dB CHL
- Bone line usually normal
- Bilateral cases -> masking dilemma
  - Wave I of ABR can provide ear specific information
IMAGING

AURAL ATRESIA

IMAGING

• Pneumatization
• Inner Ear
• Facial Nerve – Foot Plate
ARE AURAL ATRESIA CLASSIFICATIONS USEFUL?

ALTMAN
JAHRSDOERFER
OMBREDANNE
Jahrsdoerfer Scale

- Stapes present: 2
- External ear appearance: 1
- Middle ear space: 1
- Mastoid pneumatization: 1
- Malleus-incus complex: 1
- Incus stapes connected: 1
- Facial nerve course: 1
- Oval window open: 1
- Round window open: 1

ATRESIA Classifications

There was no relationship between hearing levels, the degree of middle ear aeration, facial nerve abnormalities or severity of the microtia.

ATRESIA CLASSIFICATION

**Minor**
- Normal pneumatization
- Normal inner ear
- Normal FP/oval window
- Normal facial nerve

**Major**
- Poor pneumatization
- Inner ear malformation
- Absent oval window
- FP/Facial nerve abnormality
INOPERABLE

(Atresiaplasty)
BAHA
Mixed and Conductive Hearing Loss
GOALS

- Improvement of hearing
- Long term patency
- Long term stability of hearing
CONGENITAL ATRESIA SURGERY

Pre-school age (6 yo)

- Post-op Patient Cooperation
- Audiometry (proper masking)
- Easier Radiological Evaluation

Indications

- Enough Pneumatization
- Bilateral Cases
- Functioning Cochlea
ATRESIA - SURGERY

Indications - **Unilateral Cases**

- Cholesteatomas
- Minor Malformations
- Adults, Children?
CONGENITAL ATRESIA

First Operation? Why?

1- Auricular Reconstruction
2- Atresiaplasty / Tympanoplasty

Auricular Reconstruction

Reconstructive Surgery or Prosthesis?
MICROTIA RECONSTRUCTION
SURGICAL COMPLICATIONS
ATRESIA PLASTY

Technique
Congenital Aural Atresia Surgery

Surgical Technique
Atresia - Primary Cases

Cholesteatomas = 14%
# ATRESIA

## HEARING- LONG TERM FOLLOW-UP

**Primary Operations (N = 107)**

<table>
<thead>
<tr>
<th>Post-op AB Gap</th>
<th>%</th>
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<tbody>
<tr>
<td>&lt;10 dB</td>
<td>15</td>
</tr>
<tr>
<td>&lt;20 dB</td>
<td>53</td>
</tr>
<tr>
<td>&lt;30 dB</td>
<td>73</td>
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</tbody>
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High-tone SNHL 5

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**COMPLICATIONS**
### Complications

<table>
<thead>
<tr>
<th>Condition</th>
<th>Primary (n=87)</th>
<th>Revision (n=29)</th>
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<tbody>
<tr>
<td>Soft tissue EAC stenosis</td>
<td>7 (8.0%)</td>
<td>1 (3%)</td>
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<tr>
<td>Bony EAC stenosis</td>
<td>6 (6.9%)</td>
<td>2 (7%)</td>
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<tr>
<td>Ossicular chain refixation</td>
<td>10 (11.5%)</td>
<td>2 (7%)</td>
</tr>
<tr>
<td>SNHL &gt; 30 dB</td>
<td>9 (10.3%)</td>
<td>0</td>
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<tr>
<td>Dead Ears, Facial Paralysis</td>
<td>0</td>
<td>0</td>
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</table>
Congenital Aural Atresia Surgery

Present Surgical Technique

- Dissection of ossicles with laser
- Fascia graft - tabs, Gelfim disc
- STSG - 0.008 inches,
- .020 Silastic sheets in EAC
- Merocel wick
**Conclusions**

- Early evaluation (ABR)
- Bone conduction hearing aids
- C.T. Coronal & axial views
- Surgery at age 6
- Split thickness skin graft
- Prevent lateralization
- Prevent High Tone SNHL Laser