Aural Microtia and Atresia

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CME Code: MUNQOX
Outline

- Microtia
  - Introduction
  - Classification schemes
  - Clinical evaluation
  - Physiologic/Psychological Impact
  - Reconstruction
- Atresia
  - Introduction
  - Classification schemes
  - Reconstruction
- Conclusion
Microtia Epidemiology

- Incidence of microtia 1-3 in 10,000
  - 58% right sided, 32% left sided, 9% bilateral
  - 63% male, 37% female

- Genetics
  - w/ positive family history
    - AD 9%, AR 90%, X 1%
  - Familial non-syndromal high grade microtia
    - Predominantly AD w/ variable penetrance
Etiologies

- Ischemia
  - In utero obliteration of stapedial a. or hemorrhage into local tissues
- Chromosomal aberrations
  - Goldenhar, Treacher-Collins, 18q deletion, others
- Teratogens
  - Thalidomide, isotretinoin, vincristine, colchicine, cadmium
  - Maternal infections
- Unknown
Development of Classification Schemes

- Marx (1900)
- Rogers (1950)
- Tanzer
- Weerda
- Nagata
- Aguilar (2000)
Classification

- Grade 1
  - Pinna malformed and smaller than normal but with good definition and minor anomalies
Classification

- Grade 2
  - Helix may or may not be fully developed
  - Triangular fossa, scapha, antihelix have much less definition
Classification

- Grade 3
- Pinna essentially absent except for vertical remnant of unorganized elastic cartilage superiorly with relatively well-formed lobule
Classification

- Anotia
  - Total absence of the pinna
**Additional Classification Schemes**

- **Brent**
  - **Classical microtia**
    - Larger lobule with amorphous cartilage remnant
    - Usually w/ EAC absence
  - **Atypical microtia**
    - More recognizable portions of concha, antihelix, tragus and antitragus
    - Upper portion absent
    - EAC may or may not be present
Additional Classification Schemes

- Nagata
  - Lobular remnant type deformity
    - Corresponds to classic type microtia
  - Conchal remnant type deformity
    - Corresponds to atypical type microtia
    - small and large type depending on conchal development
Additional Considerations

- Degree of associated facial hypoplasia
- Auricular dystopia
  - Medial, inferior and anterior displacement of auricle or microtic vestige due to underlying facial skeleton deficiency
  - May also have a dystopic EAC
Initial Evaluation

- Timing: ideally within a few weeks after birth
- H/o exposure to teratogens sought
- Examination:
  - Anomalous ear, normal ear
  - Other possible associated anomalies
  - Hearing status
    - OAE and/or ABR within first 2-3 months
    - Bone conduction usually but not always normal w/ CAA
    - CHL usually maximal due to lack of EAC/ossicle fixation
Imaging and Follow-up

- Renal US
- Xray: C, T, L spine to r/o malformations
- CT T-bone
  - Possibility of reconstruction
  - Presence of cholesteatoma
  - Some obtain at 1 yr, others delay until 5-7 years
- 3D CT scans in those w/ craniofacial microsomia
- Follow in office in 6-12 month intervals
Impact

- **Functional**
  - Hearing in b/l cases
  - Difficulty wearing glasses/hearing aids

- **Psychological**
  - Age < 5 yrs: usually little psychological impact
  - Age > 5 yrs: begins to notice difference between ears
Goals and Expectations

- Goals to emphasize with parents
  - 1\textsuperscript{st}: Maximizing hearing
    - Speech development usually normal in u/l microtia/atresia
    - Frequent otologic evaluations to r/o problems in normal ear
    - Early use of BAHA softband especially for b/l, but also u/l
    - Hearing may improve with recon, perfect hearing unlikely
  - 2\textsuperscript{nd}: Cosmesis
    - An auricle can be created that looks much better than the vestige but it may not be completely normal
- Reconstruction requires several stages
  - Revisions may be necessary
Reconstruction Options

- Observation
- Molding
- Prosthetic replacement of external ear
- Reconstruction with prosthetic framework
- Autologous framework with local tissue/flap coverage
Molding

- Some grade 1 malformations
- Instituted very shortly after birth
Prosthetic Ear Replacements

- Newer osseointegrated anchoring systems
  - Surgical placement of a titanium anchor
  - Magnet system and prosthesis attached
- Bone conduction HA can be built in
- Expensive
- Precludes future recon
Prosthetic Frameworks

- First reported in 1960’s
  - Then abandoned due to erosion/exposure

- Newer medpor framework has seen better results
  - Long-term follow-up needed
Autologous Reconstruction

- First reported in 1930’s, expanded in 1940’s
- Came to forefront with Brent in 1980’s
- Further refinements by Nagata recently

- 3 main elements among all methods
  - Construction and placement of cartilage framework
  - Lobule rotation, conchal excavation and tragus formation
  - Elevation of the pinna
Timing/Sequence of Surgery

- B/l vs u/l microtia w/ or w/o atresia
- Recommended age varies widely
  - Early
    - Psychological impact minimized, hearing function improved at earlier age
  - Later
    - Better cosmetic outcome, patient compliance
- Microtia repair prior to atresia to preserve blood supply
Surgical Planning

- Template for positioning of reconstructed auricle
- Template for cartilage framework
- Low hairline
- Assessment of costal cartilage volume
Relevant Anatomy

- 3 levels or complexes of reconstruction
- Proportions and relative position
Cartilage Framework

- Constructed from costal cartilage fragments from 6th-8th ribs
  - Opinions on ipsilateral vs contralateral differ
- Alterations with conchal remnant microtia
- Cartilage banked for projection
First Stage

- Cartilage framework constructed
- Vary by technique
  - Nagata, Firmin, Bauer: rotation of the lobule and tragal construction at 1\textsuperscript{st}
  - Brent: delay lobule transposition and tragal reconstruction till 2\textsuperscript{nd}
- Amorphous cartilage excised, framework can be spliced into usable portions
Second Stage

- 3-4 months after 1st Nagata and Firmin:
  - Elevation of reconstructed ear

- Brent:
  - Rotation of the lobule, conchal excavation
Third and Fourth Stages

- Brent:
  - construction of tragus
  - elevation of the reconstructed ear

- Cryptotia repair is similar to the 4th stage

- Post-op scar formation can obliterate part of the sulcus and decrease projection
Dystopic Auricle

- Ideal procedure yet to be achieved
- Altered vascular anatomy
- Transposition of microtic vestige
- Framework construction modified due to hypoplastic base
Complications

- Infection
- Cartilage/soft tissue loss
- Chest wall deformity
- Pneumothorax
Aural atresia

- Failure of development of EAC and middle ear
  - Generally have normal cochlear function

- Epidemiology
  - 1/10K-20K live births
  - 3-4 u/l : 1 b/l
  - Right > left
  - Male > female
Classification

- Degree depends on point of interruption of development

- Group 1
  - Normal or stenotic canal with hypoplastic middle ear and mild malformation of the ossicles
Classification

- **Group 2**
  - Fistulous tract or complete atresia of the canal with a bony atretic plate and some degree of malformation of the middle ear structures
Classification

- Group 3
  - Complete ear canal atresia with nonpneumatized mastoid and middle ear

- Atresia commonly coexists with microtia, can occur alone
Goals of Atresiaplasty

- Restoration and stability of hearing
- Maintenance of a patent, skin-lined EAC
Alternatives to Atresioplasty

- Softband bone conduction aid
- BAHA
  - Alterations in technique
- Additional resources for hearing loss
  - Preferential seating, FM system, individualized education program, speech therapy
### Jarhsdoerfer Grading Scale

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

<table>
<thead>
<tr>
<th>PARAMETER</th>
<th>POINTS</th>
</tr>
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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>
<tr>
<td><strong>Total available points</strong></td>
<td><strong>10</strong></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>
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Patient Selection

- Anatomic considerations
  - Malleus-incus complex (MIC) large & directly lateral to the stapes
  - Low lying tegmen
  - Facial nerve obstructing oval window
  - Facial nerve turns anterolateral obstructing attic/middle ear
  - TMJ lateral to middle ear
- Anomaly of the inner ear
Timing of Surgery

- Unilateral
  - Early repair
  - Late repair
- Bilateral
  - Hearing status
  - Favorable anatomy
- Microtia implant
Surgery

- Schuknecht methods
  - Transmastoid
- Jahrsdoerfer anterior approach
  - Standard in atresia surgery today
- Anesthesia concerns
  - Facial nerve monitoring
  - Avoid nitrous oxide
Surgery

- Temporalis fascia graft obtained
- Mastoid periosteal incisions
- Identification of landmarks
- Drill atretic bone
- Enter middle ear in epitympanum
Surgery

- Establish status of ossicles
- Fascia grafting
- Skin grafting
- Meatoplasty
Follow-up

- Periodic debridement
- May swim after 1 month
Complications

- Facial nerve injury
- Sensorineural hearing loss
- Need for revision
Future

- Newer alloplasts
- Tissue engineering
Conclusion
References

References

Thank you

Advisor: Dr. O’Reilly
Questions/Comments