Management of Microtia and Atresia
Multi-Institutional Residency Education Program

NO DISCLOSURES

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Before
After
Before
After
Multi-Institutional Residency Education Program

Goals

- Work up of a Newborn with Microtia/Atresia
- Review Hearing Screening Protocols
- Discuss Hearing Amplification options
- Examine Hearing and Reconstructive Surgical Options

ASK QUESTIONS!  ACTIVE DISCUSSION!
What famous Rock Star has Microtia and atresia?

Paul Stanley from KISS
Microtia and Atresia

Microtia
- congenital deformity of the outer ear or pinna where the ear is smaller, underdeveloped, and misshapen.
- 1 in 10,000 births
- 90% of cases, it affects only one ear, usually the right ear, and is more common in males

Ear Canal Atresia
- absence of the bony ear canal

Ear Canal Stenosis
- Presence of extremely narrow bony ear canal
Grades of Microtia

Type I

Type II

Type III

Type IV

Grades of Microtia
Embryology of the external ear
CASE 1

*3 day old M with Right Microtia and Atresia*

- Do you see them in the newborn period?
- What do you discuss with the parents?
- Diagnostic work up outside of audiology?
- Consultations?
SYNDROMES ASSOCIATED WITH MICROTIA

1. Treacher-Collins
2. Goldenhar (OAV), hemifacial microsomia, Craniofacial Microsomia
3. Townes-Brocks Syndrome -(renal-ear-anal-radial syndrome (REAR))
4. Branchio-Oto-Renal Syndrome (BOR)
5. CHARGE syndrome
Work up of a Newborn with Microtia

**Imaging**
- CT is *not* recommended at birth
- Renal ultrasound *is* recommended

**Consultations**
- Pediatric Otolaryngologist
- Geneticist (Craniofacial Panel)
- Reconstructive surgeon for microtia
- Reconstructive surgeon for hemi-Facial Microsomia
- Pediatric Audiologist
CASE 1

NEW BORN HEARING SCREEN:

Right Microtia ear - Refer
Left normal ear - Pass

› What is the next step in hearing testing?
› Do you have a time frame?
CASE 1

Sleep Deprived ABR at 4mo age

Right Microtia ear - Maximal CHL
Left ear - Normal hearing

- Do you recommend a hearing aid? What kind?
Bone Anchored Hearing Devices: Worn devices

BAHA®/PONTO on Softband

Minimal Risk

Use as a bridge until a surgical procedure can be done

Well suited for infants and toddlers, ALL AGES
Bone Anchored Hearing Devices: Worn devices

- Adhear (Med-El)- All Ages
- Adhesive retained
Bone Anchored Hearing Devices: Worn devices

BAHA® SoundArc- for use in Childhood
• “Why should I bother with a hearing aid if he will just get an ear canal?”

• “Won’t the hearing aid call attention to his little ear?”

• “Can I just wait until he starts school to use it?”

• “Won’t he do fine with the one good ear?”

- Case-control study: 6-12-yo with UHL compared with sibling controls (n = 148)

- Outcome measure: Scores on the Oral and Written Language Scales (OWLS)

- Results: Children with UHL had worse scores than their siblings
  - language comprehension (91 vs 98; P < 0.003)
  - oral expression (94 vs 101; P < 0.007)
  - oral composite (90 vs 99; P < 0.001)

- **Children with UHL more likely to have:**
  - an individualized education plan (odds ratio: 4.4 [95% confidence interval: 2.0 – 9.5])
  - received speech-language therapy (odds ratio: 2.6 [95% confidence interval: 1.3 – 5.4])
  - Helps explain retention rate 37% compared to 3% in normal hearing peers (Bess et al 1998)

• Retrospective review: 74 pts (48 R atresia, 19 L atresia, 7 bilat atresia)

• Outcome measure: Rates of speech and/or language delay, prevalence of speech therapy and educational interventions, parental report of psychosocial problems

• Results:
  • High rates of speech therapy (86% among bilat, 43% among unilat)
  • Reports of school problems more common with R atresia (31% R, 11% L, 0% bilat)
  • Educational interventions common in all groups (33% R, 21% L, 43% bilat)

• Children with unilat atresia may be at greater risk of speech and learning difficulties than previously appreciated, similar to children with unilat SNHL

- Case control survey (132 to families of children w/ unilat atresia, 48 to unilat SNHL)
- Atresia patients: none repeated a grade, but 65% needed some resources: 12.5% use HA, 32.5% use FM system at school, 47.5% had IEP, 45% in speech therapy.

<table>
<thead>
<tr>
<th>TABLE III. Academic Progress.</th>
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<tbody>
<tr>
<td></td>
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<tr>
<td>Resource</td>
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<td>-----------------------------</td>
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<tr>
<td>Repeated a grade</td>
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<tr>
<td>Any resource</td>
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<tr>
<td>Amplification</td>
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<tr>
<td>Speech therapy</td>
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<tr>
<td>FM system</td>
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<tr>
<td>IEP</td>
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<tr>
<td>Special education</td>
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<tr>
<td>Behavior problem</td>
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</table>

- Unilateral CHL due to aural atresia has an impact on academic performance in children, although not as profound when compared to children w/ unilat SNHL.
CASE 2

3 day old M with bilateral Microtia and Atresia

- What age do you fit for hearing aid?

- Recommend Unilateral or bilateral Aids?
SUMMARY

Audiological Work up Time Line

Birth

Newborn Hearing Screening
- Usually REFER in involved ears
- Referral to Pediatric Audiology by 3 months age

Before 6 Months Age

Sleep deprived ABR
- ABR with both AC and BC toneburst thresholds from 500-4000 Hz

By 6-12 months Age

Hearing aid consultation and for fitting
- Meet with pediatric audiology to discuss and try different passive “WORN” bone anchored aids.
CASE 3

2-year-old with unilateral microtia and atresia starts refusing to wear his bone anchored device on a softband

What do you counsel parents?
Case 4: A 5-year-old has been wearing the BAHA device daily. What are the implantable devices available?

- What device do you implant most frequently?
- What surgical considerations do you have to consider before implanting?
Audiologic Options for Microtia/Atresia Patients

• Passive bone conduction aids
  Non-surgical
  - adhesive retained
  - softband retained
  Surgical
  - percutaneous implant retained
  - BAHA
  - PONTO

• Transcutaneous magnet retained
  - Sophono (children > 5)
  - BAHA Attract

• Active bone conduction aids
  – Vibrant Soundbridge (> 18 yo)
  – Bone Bridge (> 12 yo)
  – Osia system (> 12 yo)
BAHA Attract/Sophono
- Transcutaneous Magnet retained aid

BAHA Connect/ Ponto
- Percutaneous Implant retained aid
Bones Anchored Hearing Devices: Implanted devices

Active Bone Conduction Implants

• BoneBridge (FDA approved 12yrs+)

Source: MedEl Corp
Bone Anchored Hearing Devices: Implanted devices

Active Bone Conduction Implants

• Vibrant SoundBridge (FDA 18yrs+)

Source: MedEl Corp
Cochlear Osia System

• Piezo electric transducer sends sound wave energy via osseointegrated implant to inner ear
• FDA approved 12 years and older
Outcomes in short: At the 3 months follow up the patients demonstrated statistically significant improvements in aided thresholds at all measured frequencies compared to the unaided condition. More importantly, they also showed statistically significant improvements in hearing thresholds above 2,000 Hz compared to the Baha Power Sound Processor on Softband, Figure 1.

Figure 1. Aided thresholds with the Osia System at 3 months compared to unaided and pre-operative test with a Baha Power Sound Processor fitted on a Softband.
Regarding improvement in speech recognition in noise, the Osia System clearly outperformed the unaided condition and the non-surgical solution by an average of 13.3 dB and 7.2 dB respectively, Figure 3.

Figure 3. Speech recognition in noise with the Osia System at 3 months compared to unaided and pre-operative test with a Baha Power Sound Processor fitted on a Softband.
Surgical considerations for placement for Bone Anchored Implants

Figure 26. Combined Medpor surgery and bone anchored implants.
Other guidance or resources for parents?

- Do you give any words of advice?
- Any resources?
Multifactorial Cause of Microtia?

1. **Neural Crest Cell (NCC) migration disturbance**: which can happen from heterogeneous reasons
   - Treacher Collins: *gene mutation* in protein Treacle leads to depletion of NCC precursors.
   - *Retinoids* and diabetic *embryopathy*: associated with NCC apoptosis

2. **Vascular Disruption**
   - Occlusion, vasoconstriction, or underdevelopment of vascular structures of developing tissues

3. **High Altitude**
   - 5X higher prevalence of microtia in *Quito, Ecuador* (located at 9,350 ft) compared with countries in low altitudes of South America.
   - other large high altitude cities of South America, La Paz (Bolivia) and Bogota (Colombia) with similar rates
   - altitude associated with chronic hypoxia

4. **Genetic**

Case 5: 3-day old M born with right microtia and atresia. Parents want to know options for the microtia repair. What about an ear prosthetic?

- Do many of our patients have or want a prosthetic?
Ear Prosthetics

How they can attach
- Adhesives
- Bone anchored Device (Clips)
- Bone anchored Magnets

Advantages
- Life Like ear
- Less surgical risk

Adhesive-Retained Auricular Prosthesis
Ear Prosthetics

Disadvantages

• Fear of ear falling off
• Poor self image of an ear that comes on and off
• Needs to be replaced every 5 years due to fading and wear and tear on material
• Anaplastologist not covered by insurance
• Color match can be poor
• There can be an obvious line or poor blending of the ear with patients skin
• All issues with bone anchored abutments and skin overgrowth or infection
Case 5: 3-day old M born with right microtia and atresia. Parents want to discuss Medpor Reconstruction.

- Advantages?
- Disadvantages?
- At what age can your undergo this reconstructive surgery?
**Medpor**: high-density porous polyethylene implant for microtia reconstruction
Medpor Reconstruction

Medpor Reconstruction

“CAM”- Combined Microtia and Atresia

Advantages

• Does not rely on the patients costal cartilage, so can performed earlier age, as early as 3 years.
• Can be done completed in 1 surgery
• Has a reliable size and shape due to mold used
• Cosmetically reliable
• Thinner and “floppier”

https://www.drjohnreinisch.com/photogallery
Medpor Reconstruction

**Disadvantages**

- Insensate
- Lifelong risk of exposure, infection and extrusion
- Difficult to treat infection due to foreign body material
- Once the implant is exposed, risk of implant loss
- Technique exhausts some of the local tissue that would typically be used to repair the ear
- Uniform “cookie – cutter” ear
- Some children cannot sleep on that ear due to compromise of the TPF flap with pressure.
- *Technically challenging technique, difficult learning curve*
Case 5: 3-day old M born with right microtia and atresia. Parents want to discuss Cartilage Reconstruction.
Advantages

- Autologous cartilage used heals with the patient.
- Sensate
- Good vascular supply: bleeds, and infections are treatable with antibiotics.
- Reconstruction is stable for a lifetime
- Cartilage construct can be personalized for the microtia ear
Cartilage reconstruction

Disadvantages

- Requires 2-4 surgeries (depending on technique)
- Timing of surgery is typically 7-10 years of age when the child has a robust rib cage for the cartilage graft
- Donor site scar and pain at the time of surgery
- Technically challenging surgery for the surgeon, demanding for wound healing

http://earsurgerymd.com/autologous-reconstruction
Case 6: Parents have decided on cartilage reconstruction. What are the differences in techniques?

1. Classic Burt Brent 4 stage technique
2. Nagata 2 stage technique
3. Firmin 2 stage technique
**Brent Technique**

**Stage 1**
- Cartilage harvest
- Creation of framework
- Place under local skin

**Stage 2**
- Transposition of the lobule

**Stage 3**
- Elevation of construct

**Stage 4**
- Creation of Tragus
- Deepen the conchal bowl
Stage 1
- Cartilage harvest
- Creation of framework
- Place under local skin with transposed lobule

Stage 2
- Elevation of ear
Preoperative appearance

Post-operative appearance
Firmin Technique:
2 staged modified Nagata

Stage 1
- Cartilage harvest
- Creation of framework
- Place under local skin with transposed lobule

Stage 2
- Elevation of ear
Firmin Systematic approach to Auricular Reconstruction

Skin Approach: 4 types

Framework: 3 Types

Projection Pieces: 3 Types

Elevation Technique: 4 methods
Firmin
Case 7: 7 yo with a conchal type (Grade 2) microtia with an ear canal: What is the ideal type of microtia repair?

- Advantages/Disadvantages of Medpor?
- Advantages/Disadvantage of Cartilage?
Case 8: 5yo with bilateral microtia, canal atresia, a craniofacial syndrome, small for age: what is the ideal type of microtia repair?

- Advantages/Disadvantages of Medpor?
- Advantages/Disadvantage of Cartilage?
Case 9: 7yo with unilateral microtia and canal atresia, parents wants to be considered for canal atresia repair.

- What are the factors you consider for the canal reconstruction?
- Does it matter the type of microtia repair they are considering for timing of canal surgery?
- How do you counsel them on the hearing benefits?
- What do you discuss in terms of the risks of ear canal reconstruction?
Atresioplasty

- candidacy

- normal hearing uncommon
  - “good” result is hearing at 20-25 dB HL

- complex surgery

- risk of facial paralysis

- frequent restenosis
Hearing Outcomes of Atresia Surgery Versus Osseointegrated Bone Conduction Device in Patients With Congenital Aural Atresia: A Systematic Review

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Pubmed Search
(canal OR aural OR auricular) AND atresia AND (surgery OR reconstruction OR Aid OR conduction OR implant) AND (hearing OR audio*)
465 articles

Review of Abstracts
107 articles

41 articles
38 English
3 non-English

Excluded: 352 articles
• Animal studies
• Middle ear implants
• Acquired aural atresia
• Published prior to 1975

Atresia Surgery
35 articles
1617 operated ears

BAI
6 articles
112 operated ears

Excluded: 36 articles
• EAC stenosis alone
• Hearing outcomes other than SRT, PTA, HG or ABG
• Multiple paper by same author
• < 5 patients in series
• Unable to translate
## Atresiaplasty Results

<table>
<thead>
<tr>
<th>Time</th>
<th>% SRT ≤30dB</th>
<th>% PTA ≤30dB</th>
<th>Average HG</th>
<th>% ABG ≤30dB</th>
</tr>
</thead>
<tbody>
<tr>
<td>Any</td>
<td>73.8%</td>
<td>60.3%</td>
<td>24.1 dB</td>
<td>69.0%</td>
</tr>
<tr>
<td>&lt;6m</td>
<td>78.4%</td>
<td>71.7%</td>
<td>25.5 dB</td>
<td>71.2%</td>
</tr>
<tr>
<td>&gt;12m</td>
<td>61.5%</td>
<td>57.2%</td>
<td>23.5 dB</td>
<td>51.6%</td>
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</table>
### Comparison of Hearing Outcomes

<table>
<thead>
<tr>
<th>Surgery</th>
<th>% SRT ≤30dB</th>
<th>% PTA ≤30dB</th>
<th>Average HG</th>
<th>% ABG ≤30dB</th>
</tr>
</thead>
<tbody>
<tr>
<td>Atresiaplasty</td>
<td>73.8%</td>
<td>60.3%</td>
<td>24.1 dB</td>
<td>69.0%</td>
</tr>
<tr>
<td>BAHA</td>
<td>N/A</td>
<td>95.9%</td>
<td>38.0 dB</td>
<td>98.2%</td>
</tr>
</tbody>
</table>

Positive publication bias - probably better than the average result
Only successful experienced surgeons w/ more extensive case series tend to publish, so these data do not include less experienced surgeons with smaller case series and presumably worse outcomes
Membranous atresia and/or bony stenosis

- Cole & Jahrsdoerfer, Laryngoscope 1990
  - Reviewed 600 congenital ear malformations
  - 54 ears with canal stenosis and/or membranous atresia
  - 91% of pts > 12 yo with stenosis < 2 mm presented with cholesteatoma
My Results

- 24 patients with membranous atresia and/or bony stenosis
- Preop mean thresholds: $64 \pm 4$
- Postop mean (500, 1000, 2000 hz): $22 \pm 14$
- 6 month postop ABG: $16 \pm 13$
- Closure to < 20 dB in 83%
- 6 (25%) required revision surgery to meatus
Microtia and Atresia

Microtia Reconstruction Time Line

Birth

Consultation with reconstructive surgeons for parental counselling regarding reconstructive options for the future

3-5 years Age

Earliest age for consideration for MEDPOR® reconstruction. Any ear canal surgery should ideally be done BEFORE or concurrently with MEDPOR reconstruction.

5 years Age

Earliest age for osseo-integrated anchor for Prosthetic use. Otherwise Prosthetics can be worn with adhesive prior to that. Ear canal surgery can be done at any time.

5-9 years Age

Earliest age for autologous cartilage reconstruction. Any ear canal surgery is ideally done AFTER microtia reconstruction.
**Microtia and Atresia**

**Audiological Work up Time Line**

- **Birth**
  - New Born Hearing Screening
    - Usually REFER in involved ears
    - Referral to Pediatric Audiology by 3 months age

- **Before 6 Months Age**
  - Sleep deprived ABR
    - ABR with both AC and BC toneburst thresholds from 500-4000 Hz

- **By 6-12 months Age**
  - Hearing aid consultation and for fitting

- **12 months-5 years**
  - Annual audiological testing and surveillance

- **5 years**
  - Consider CT scan for hearing implants, canal reconstruction candidacy. Hearing surgeries should be appropriately coordinated with the type of microtia reconstruction planned.
Case 11: What 3D printing technology is being utilized in microtia surgery?

- How close are we to having a 3D printed ear?
- How are 3D printed models used to assist surgery?
3D printed models designed for each patient based on the non-microtia ear
THANK YOU for being ALL EARS!

https://earcommunity.org/microtiaatresia/emotional-support/dont-blame-yourself-as-a-
REFERENCES:


Weblinks for photos used

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