Understanding Atresia, Microtia and the Baha® System

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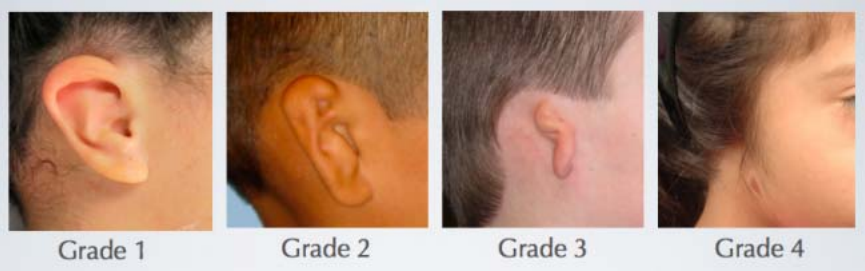
Agenda

- Atresia vs. Microtia
- Softbands
- Baha Implants
- Family Experiences

Terminology

- Microtia – refers to a spectrum of deformities of the external ear;
  - Grade I – slightly smaller ear with majority of structures present
  - Grade II – greater deficiency of ear structure as a missing lobule or helix
  - Grade III – classic 'peanut' deformity
  - Grade IV – anotia (total absence of the external ear)



Microtia

- Prosthetics (best with anotia)
  - VistaFix – attached similar to a Baha Implant
  - Traditional prosthetic – glued on
- Surgical Reconstruction
  - Average of 3 to 4 surgeries
  - Rib Graft Surgical Technique
    - 1920s
    - Uses the body's own biological tissues (rib cartilage and skin)
    - Brent Technique (Dr. Burt Brent) – average 3 stages (sometimes 4)
    - Nagata Technique (Dr. Satoru Nagata) - 2 stages
  - MEDPOR® Reconstruction
    - Uses synthetic material but earlier ages for reconstruction
  - Age requirements vary from surgeon to surgeon but vary between 5 to 8 years of age; primarily because of the growth of the child

Terminology

- Atresia - is the absence or closure of the external auditory ear canal; sometimes called aural atresia
  - The incidence of congenital aural atresia is approximately 1 in 10,000 to 20,000 live births
  - Unilateral atresia occurs three to five times more commonly than bilateral atresia
  - Males are more often affected than females
  - Right ear is more common in unilateral cases
  - Common Syndromes: Treacher-Collins, Goldenhar's, Crouzon's, Mobius', Klippel-Feil, Fanconi's, DiGeorge, VATER, CHARGE and Pierre Robin

Atresia Classification

- Altmann's classification
  - first reported in 1955 but still widely utilized
- Group I – small EAC, hypoplastic temporal bone and TM, a normal or small middle ear cleft and normal or mildly deformed ossicles
- Group II - absent EAC, an atretic plate, a small middle ear space and fixed and malformed ossicles
- Group III – absent EAC, a severely contracted or absent middle ear space, and absent or severely malformed ossicles.

Altmann, F. (1955). Congenital atresia of the ear in man and animals. Annals of Otology Rhinology Laryngology, (64): 824-858.

Atresia Classification

Jahrsdoerfer 1992

- This system establishes a score, 1 to 10, based upon findings of high resolution CT scans
- Open oval window, width of the middle ear cleft, facial nerve course, malleus-incus complex, mastoid pneumatization, incudostapedial continuity, round window patency and auricle appearance = one point
- The presence of a stapes = two points
- The final score is used to predict the likelihood of successful atresia repair
- A score of 8 out of 10 correlates to an 80% chance for restoration of hearing to normal or near-normal levels defined as speech reception thresholds (SRT) between 15 and 25 dB
- Cases with a score of less than or equal to 5 are generally not considered for surgical intervention

Jahrsdoerfer, RA, Yeakley, JW, Aguilar, EA, Cole, RR, Gray, LC (1992). Grading system for the selection of patients with congenital aural atresia. American Journal of Otology, (1) 13:6-12.

Atresia Repair

- Candidacy
  - Facial nerve complications
  - Hearing outcomes
  - Patency of the repair
  - Age of patient
- High-resolution CT imaging has greatly impacted the determination of atresia repairs
  - Condition of the ossicles
  - Presence or absence of round and oval windows
  - Course of the facial nerve
  - Degree of pneumatization of the middle ear and mastoid

Atresia Repair Complications

- Facial nerve injury
- Tympanic membrane lateralization
- Restenosis
- Ossicular refixation
- SNHL
- Canal cholestoma

Atresia Repair

The percentage of cases with a successful outcome from atresia repair reported in the literature varies greatly!

Tollefson (2006)

- 14 specialized centers
- 595 total patients
- 48% (288 patients) had acoustic transmission of $\leq$ 30 dB

Tollefson, T (2006). Advances in the treatment of microtia. Current Opinion in Otolaryngology & Head and Neck Surgery, 14:412-422.

Atresia Repair

Lambert 1998

- Majority of research is citing the hearing results obtained soon after surgical repair
- An important concern is the stability of the hearing results over time
- Compared early postoperative hearing levels <1yr post-op to levels after longer follow-up (average 2.8 yrs)
- 60% of cases had hearing levels of 25dB or better and 70% were at 30 dB or better in the early post-operative period
- This diminished to 46% and 50% with longer follow-up
- Nearly one third of cases required revision surgery most commonly for restenosis of the EAC or lateralization of the TM
- After revision surgery, hearing levels of 25 dB or less were achieved in 50% of cases and levels of 30 dB or less in nearly two thirds of cases
- He also commented that of those patients with an exceptional result after the primary surgery (hearing level 10-20 dB) 83% maintained this outcome over longer periods of follow-up

Lambert, PR (1998). Cogenital aural atresia: stability of surgical results. Laryngoscope, 108:1801-1805.

The Baha System

- Osseointegrated devices have treating conductive and mixed hearing loss since 1977
- Works through direct bone conduction
- Sound is conducted through the skull bone bypassing the outer and middle ear and stimulating the cochlea
- Contains three parts:
  - Internal titanium implant
  - External attachment
  - Detachable sound processor

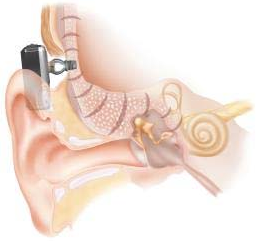
The Baha System

Softbands

- No age restrictions
- Bilateral CHL
- Unilateral CHL
- Unilateral or bilateral Softbands available

Implants

- FDA recommendations for implantation - age 5 years of age or older
- Bilateral or unilateral implants



Softband Data

- Retrospective study of Baha patient charts of infants and children 2002 to 2006
- 20 infants and children
- Age: 8 months to 16 years (mean age = 5.04 years)
- Inclusion criteria was:
  (a) Bilateral symmetrical conductive hearing loss
  (b) Fit unilaterally with Baha Softband

Softband Data

Condition	500 Hz	1000 Hz	2000 Hz	4000 Hz
Unaided SF				
M (SD)	61.2 (9.2)	60.2 (11.7)	56.4 (13.6)	54.8 (13.9)
95% CI	57.6, 64.8	55.6, 64.8	51.8, 61.7	49.4, 60.2
Aided SF				
M (SD)	20.2 (3.1)	18.8 (3.6)	17.8 (4.8)	20.2 (4.7)
95% CI	19.0, 21.4	17.4, 20.2	16.1, 19.5	18.4, 22.0
Functional gain				
M (SD)	41.0 (6.1)	41.4 (8.1)	38.6 (9.2)	34.6 (9.2)
95% CI	38.6, 43.4	38.2, 44.6	35.0, 42.2	31.0, 38.2
Paired *t* test statistic for unaided-aided values	20.29	17.45	13.47	11.7
p value	<.001	<.001	.001	.001

* dB = decibel; HL = hearing level; SF = soundfield; M = mean; SD = standard deviation; CI = confidence interval.

Nicholson, N. Christensen, L, Dornhoffer, J. Martin, P, Smith-Olinde, L. (2011).Verification of speech spectrum audibility for pediatric Baha Softband users with craionfacial anomalies. Cleft Palate-Craniofacial Journal, (48)1: 56-65.

Baha System vs. Traditional BC Aids

Retrospective study of 10 subjects:

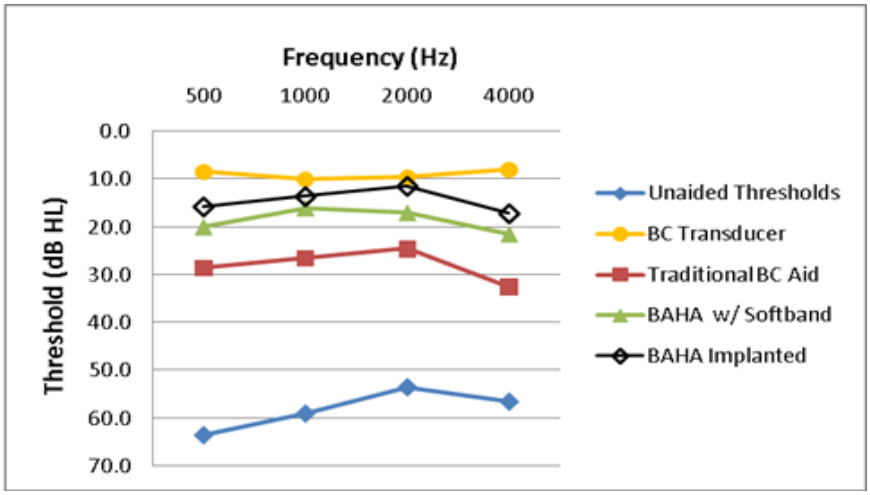
- Age: 6 months to 18 years of age
- Congenital bilateral conductive hearing loss
- Initially fit with a traditional bone conduction hearing aid
- Fit unilaterally with a Softband
- Implanted unilaterally with the Baha System
- Unaided and aided soundfield thresholds available for four frequencies from 500 Hz to 4000 Hz

Christensen L, Smith-Olinde L, Kimberlain J, Richter G, Dornhoffer J (2010). Comparison of traditional bone-conduction hearing aids with the Baha system. Journal of the American Academy of Audiology. (21)4:267-273

Results

- Bone conduction transducer provides the most gain of any device tested
- The implanted Baha System provided second highest amount of functional gain
- Softband results provided the third amount of highest functional gain
- Traditional bone conduction hearing aids provided the least amount of functional gain
- There is some overlap among devices at 1000 Hz, but at no other frequency.
- Implanted Baha System has statistically as much gain as a bone conduction transducer at all frequencies tested
- Implanted Baha System provides statistically more gain at 500 Hz than the Baha attached to a Softband
- Traditional bone conduction hearing aid provides significantly less gain than all the other devices at all frequencies with the exception of the Baha Sound Processor with Softband at 2000 Hz

Christensen L, Smith-Olinde L, Kimberlain J, Richter G, Dornhoffer J (2010). Comparison of traditional bone-conduction hearing aids with the Baha system. Journal of the American Academy of Audiology. (21)4:267-273



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Decisions

- The ultimate decision maker is the family – provided there are no medical complications to contradict either option
- Audiologists must know all the facts and research (atresia, atresia repair, and Baha Softbands and implants) in order to help each family individually
- As in most cases, one size does not fit all
- When working with children, it is imperative that families understand the urgency of fitting amplification
- EHDI 1-3-6

But what if it's *just* unilateral?

- Kesser, Krook, and Gray
- Compared unilateral SNHL to unilateral atresia for academic performance
- 40 atresia patients
- None repeated a grade
- 65% needed some resources
  - 12.5% used a hearing aid
  - 32.5% used a FM system in school
  - 47.5% had an IEP
  - 45% were in speech therapy
- Conclusions: unilateral CHL/atresia has an impact on academic performance in children; not as a profound impact as the unilateral SNHL

Kesser, B, Krook, K, Gray, L. (2013). Impact of unilateral conductive hearing loss due to aural atresia on academic performance in children. Laryngoscope, (123) 9:2270-2275.

Family Discussions

- Interviews with 3 families
  - Family 1 – unilateral atresia/microtia currently 4 years old
  - Family 2 – bilateral atresia 6 years old
  - Family 3 – Goldenhar syndrome with bilateral atresia/microtia; 18 years old
- Discussed these main topics
  - Diagnosis
  - Treatment Options
  - Professionals
  - Internet
  - Initial Treatments
  - The Future
  - Early Amplification
  - Advice for New Parents

Family 1

Diagnosis

- Left sided unilateral microtia and atresia.
- Shortly after our son's birth, we were told our son did not pass his hearing evaluation in his left ear because there was no ear canal. We were told he needed to be referred to an ENT.
- We did not know the name for the condition he had when we left the hospital, but we quickly educated ourselves and discovered the medical names when searching on the internet after returning home from the hospital.
- When my son was around 8 months old we were scheduled with the ENT.

Family 1

Treatment Options

- Because of the kind of people we are, we sought information and researched on the internet prior to our son's first ENT appointment. The ENT appointment confirmed what we suspected, Grade 3 microtia with atresia. We had already read about possible surgeries to reconstruct the inner and outer ear. Our local ENT does perform the canalplasty surgery and we discussed a little about this at the first appointment. The ENT also mentioned the Baha System and connected us with a knowledgeable audiologist at out first visit. We set up a two year follow up appointment with the ENT and we were then closely followed by the audiologist.

Family 1

Treatment Options

- **We as parents have been the driving force behind educating ourselves on options available nationwide in regards to microtia and atresia**. I mean no disrespect to any physicians/professionals, but I feel we as parents have been the best at looking at the whole picture of our child instead of just the individual conditions. For example our local ENT is very knowledgeable about atresia, but for our son we are dealing with microtia as well. For my son's dual conditions our decisions are interconnected. If we were to choose to reconstruct my son's outer ear using the rib graft method we would wait until the four rib-graft reconstruction surgeries were complete and then the canalplasty could be completed. If we were to choose to reconstruct my son's outer ear using the Medpor method he would have the canalplasty first and then the outer ear reconstruction would be completed. Another option would be to have a prosthetic outer ear made. We also have the option of leaving my son's ear as he was born or using any of the outer ear reconstruction methods and implant the Baha System.

Family 1

Professionals

- My son's audiologist was our main person to explain the process of obtaining a hearing aid, benefits of a hearing aid, and at what age he would be appropriate for the hearing aid. Since the option of a canalplasty does not occur for a few years we were focused on obtaining a hearing aid (Baha System) at first and other decisions were put on hold for a while. We as parents knew that aiding our son was not going to hurt him in any way so we were very interested in obtaining the Baha System for him.

Family 1

Internet Information

- We were able to find an on-line support group (yahoo group) specific for microtia/atresia and we found a wealth of information on that forum that spurred other research on our own. We also found out about very well respected doctors that have dedicated most or all of their individual practices to microtia repair or atresia repair. Through the yahoo group we found out about a conference specific to microtia and atresia issues that was being held in Texas. We loaded up the car with our then nine month old son and went to hear from the experts in the field and learn first hand some of the options available from some of the top physicians/surgeons performing microtia/atresia reconstruction in the nation. We were not naïve that the surgeons were soliciting potential business for themselves, but it was also educational and informative for us as parents.

Family 1

Initial Treatments

- Since my son has atresia with microtia his only option for a hearing aid was a bone conduction device. We were provided with a loaner processor while my son's processor was ordered.
- The audiologist's advice was to wait until my son was holding his head up well on his own before trying the Baha Sound Processor on a Softband. My son was fitted with the Baha System (Softband) around the age of seven months.

Family 1

The Future

We have made no final decisions other then to fit our son with a Baha Sound Processor on a Softband. As our son gets closer to an age he could undergo reconstruction surgery we are even more hesitant to move forward with any life long decisions for our son. In the four and a half years since our son was born the medical community has made great strides with "growing" ears in labs and we do not want to rush into anything at this time. Our next step would be for our son to have a CT scan that would be read /interpreted by the ENT to decide if he would even be a candidate for a canalplasty. We would then have to make a decision about how to move forward regarding outer and inner ear reconstruction. At this time we are in a holding pattern. Our son wears his Baha Sound Processor on the Softband headband on a daily basis and we are pleased with his progress. We have also not ruled out the possibility of implanting the post for the Baha device and forgoing a canalplasty altogether. Part of the skin used for outer ear reconstruction is often times around the existing ear structure so it is best to not "scar" the skin by implanting the Baha implant until outer ear reconstruction has taken place. There are no easy decisions for a family.

Family 1

Early Amplification

- I am happy we choose to aid our son especially so young. He is used to wearing and hearing from the Baha Sound Processor and it is not a big deal to him. Don't get me wrong, there are still times he does not want to wear the Baha and we do not fight with him too often about it.

Family 1

Advice

- I would tell a new parent congratulations on the baby and that things will be ok. There is a grieving process that comes with anything not being as one expects when a child is born. It is very hard to try and explain to family members and friends when you do not fully understand the condition at first. Having a good support group can help new parents deal with things that might come up. I also found it very helpful to find an on-line community/support with other parents that had been there before. Before my son was a year old we also found Arkansas Hands & Voices. Even though most of the people I have connected with the through the local chapter of Hands & Voices do not have a child with microtia/atresia they are still a source of support and friendship in raising a child with a hearing loss. You will have to get used to questions from strangers especially with a hearing aid attached to a Softband. One of the strangest questions I got in a store was, "Is that a tracking device?" I have learned to have a thicker skin and to educate others about my son's condition. My son has even become his own advocate at age four and a half. The other day when a friend's son asked me about my son's Baha Sound Processor, I explained it was a hearing aid that helped him hear out of that side. My son chimed in with his own explanation saying, "My ear doesn't have an opening on that side."

Family 2

Diagnosis

- We first learned of his atresia at his first doctors visit. He had failed the hearing test at the hospital but no one looked at his ears. He was about 2 weeks old. He has atresia – no microtia – in both ears.
- He was around 2 months old when we first saw the ENT.

Treatment Options

- We were told the option for his age was the Softband Baha and that he should be aided as soon as possible. He got his Baha at 4 months. We were also told that repair surgery might be a possibility when he was close to 5 years but if he was not a candidate for repair surgery the Baha System could be implanted.

Family 2

Professionals

- The audiologist and ENT both worked together to guide us through the process.

Internet

- I did lots of research online in the beginning.

Treatment

- He has had atresia repair on both sides. We are very pleased with our choice. His hearing on the right side is now near normal with only a slight impairment on the left side in the higher frequencies. He has started kindergarten and no longer needs a hearing aid. Recovery was easy and he has had no problems not even an ear infection at this point. We spent 5 years worrying and research what would be best and I could not have asked for a better surgery experience. We meet with ***several doctors*** before making a decision on surgeons.

Family 2

Advice

- My advice for other families: ***Aid has soon as possible***. We have meet families that decided to wait and their children had some language delays. Our son only has some mild articulation issues and I believe his language success is because he was aided so early. Stay on top of the early care with audiology and ENT. ***The Baha Sound Processor will soon be just part of getting dressed.*** We made sure he wore it all waking hours so he never fussed about wearing it. When he started talking that would be the first thing he asked for in the morning "my Baha".

Family 3

Diagnosis

- When she was born she was transported to another hospital with a NICU because of her facial paralysis and deformed ears. We didn't know anything about the condition until she was born.

Treatment Options

- The options we were given for treatment were that she needed both microtia and atresia repair. It was two separate doctors – one to repair the outside and one to repair the inside – but neither would operate until age 8.

Early Amplification

- Around a year old she saw an ENT. ***She saw audiology sooner when she was a couple of months old.*** We tried a regular hearing aid first. But it was too hard to keep a regular hearing aid over her ear, so at one year old she started using a bone conduction aid.

Family 3

Professionals

- ***We heard about the Baha surgery from the audiologist.***
- The ENT guided most of her treatment. But once they did CT scans before her microtia/atresia repairs, they felt the Baha was the better option over atresia repair.

Treatments

- The microtia repair was unsuccessful - “a mess”. She is now a senior in high school. She has a Baha System implanted on one side but because of the jaw location and lack of bone on the other side she could not have a Baha System implanted there.
- Because the microtia repair was unsuccessful, we chose to have another surgery to remove the microtia repaired ears and have a VistaFix ear on the same side as the implanted Baha components and a regular prosthetic ear on the other side with poor bone structure.

Family 3

Advice

- We were not happy with the microtia repair. She had lots of pain and surgery to end up with prosthetic ears. We wish we would have just done that first and saved her the pain.

The Future

- Overall we are happy in the end. We are ok with the Baha but have had complications because of keloids. Overall it’s been a good experience for her with the Baha System.

Conclusions

- Know the options for families
  - It is possible you will be the only professional that can help guide them through this process
- Know what surgical techniques are available near your patients and across the globe – you never know what families will want to pursue
- Know the audiological outcomes for each option – our job as audiologists is to make sure our patients get the best audiological outcomes possible
- Do not wait on surgical decisions to start auditory intervention

QUESTIONS?

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