ASPO Clinical Practice Guidance Sheet: MICROTIA and EAR CANAL ATRESIA

“It is extremely stressful for a parent to have a child with microtia and atresia. Families often do not know how to navigate through the treatment options for the hearing and reconstructive needs of the child in a coordinated fashion. As pediatric otolaryngologists, we often play a large role in the care of this child, yet, no guidance about the management of microtia and atresia has been discussed. This clinical practice guidance sheet gives healthcare professionals a framework and timeline to help parents in the care of their child with microtia and atresia.”-Mai Thy Truong MD, panel co-author

What is microtia?
-Microtia is a congenital deformity of the outer ear or pinna where the ear is smaller, underdeveloped, and misshapen. Microtia ears vary in appearance, with four grades, the most common is grade 3, consisting of a tiny peanut-shaped lobe. Microtia occurs about 1 in 10,000 births, though rates can vary depending on ethnic background. In 90% of cases, it affects only one ear, usually the right ear, and is more common in males. Microtia is often associated with absence of the ear canal (called canal atresia or aural atresia), or an extremely narrow ear canal (canal stenosis).

What is canal atresia?
-Canal atresia is the absence of the bony ear canal, and is often present with microtia. A CT scan can help differentiate between canal atresia and canal stenosis, when there is an extremely narrow bony ear canal. The presence of canal stenosis may not be obvious on physical exam of the outer ear, when the meatus is very tiny and/or hidden by microtic skin remnants.

Why is this guidance sheet important?
• To standardize the work up for a newborn with microtia/ataresia
• To standardize hearing screening recommendations of a newborn/child with microtia/ataresia
• To review hearing amplification options (worn and implanted) for microtia/ataresia
• To review reconstructive options for microtia and the timeline each option entails
• To review issues concerning ear canal reconstruction, especially in coordination of timing of microtia reconstruction

Significant points made in the Cotton-Fitton Panel, “Current concepts and controversies in Microtia and Canal Atresia.”
Panelists: Mai Thy Truong, MD, Kay Chang, MD; Siva Chinnadurai, MD; Yi-Chun Carol Liu, MD; David Zopf, MD
1. The work up of a newborn with Microtia/Atresia
- Clinicians should perform a complete history and physical exam of the infant, looking for associated syndromic findings
- **Hearing exam**: The infant should undergo newborn hearing screening, but should continue with a sleep deprived ABR within the first 3 months of life to decrease the need for a sedated ABR and expedite the option for hearing amplification. The ABR is important both to establish that the inner ear is functioning on the microtia ear and also that the non-microtia ear has normal hearing from 500-4000 Hz.
- **Imaging studies**: A CT scan of the head or temporal bone is generally not recommended in the newborn for the diagnosis of canal atresia or the work up of the hearing loss. This can be reserved for just before hearing reconstructive surgical options are discussed (approximately age 5 years) or if there is any clinical concern for cholesteatoma and/or infection. Clinical concern for cholesteatoma, which is most often seen in conjunction with severe canal stenosis, may warrant a DWI (Diffusion Weighted Imaging) MRI.
- Many groups recommend a renal ultrasound in the newborn period to rule out abnormalities of the kidney which are associated with more syndromic causes of microtia.
- **Consultations**: Audiology, Geneticist if the child appears to be syndromic, Otolaryngologist/Reconstructive surgeon. These can be made as an outpatient though many parents are comforted to meet with an otolaryngologist after birth.
  *An older child with hemifacial microsomia may require consultations to address issues with mandibular hypoplasia and malocclusion. This may be through a Craniofacial clinic, plastic surgeon or oral surgeon.

2. Hearing device options and recommendations for use in an infant/child with Microtia/Atresia
- **Worn hearing device options include**: Bone anchored hearing devices such as Baha® or Ponto device on a softband. The ADHEAR device is worn on adhesive gel pad. In children > 5 years old, the Baha® SoundArc may be worn on the posterior aspect of the head as an alternative to the headband.
- Clinicians should encourage early use of worn bone conduction devices for infants with CHL.
- In cases of bilateral microtia/atresia, infants should be fit by 6 months.
- In cases of unilateral microtia/atresia, infants should have a counseling consultation with a pediatric audiologist to discuss bone conduction options, and encourage early use (by 9 – 18 months) for possible early adaptation. Although this age child is seldom exposed to challenging hearing environments, it is the experience of most centers that successful use of bone conduction aids in school aged children tends to be correlated with earlier fitting and adaptation.
- Parents should aware that there may be challenges with insurance coverage for bone conductive hearing devices in unilateral microtia.
- Parents should be counselled that toddler years can be challenging for successfully using worn devices, and a goal of 2-4 hours a day use is encouraged, until child is more tolerant.
- **Implanted hearing devices include**: percutaneous osseointegrated implants (Baha® Connect, Ponto) and transcutaneous magnetic systems (Baha® Attract, Sophono) which are FDA
approved at 5 years age. The active implant with direct drive stimulation (BoneBridge) is FDA approved at 12 years age.

- Care should be taken to work in collaboration with the reconstructive surgeon before an implant is placed as to not jeopardize the fascial flaps which may be integral for microtia repair. It is often preferred that the implant is done after microtia reconstruction, and that the superficial temporoparietal fascia is spared during implant placement.
- If placement must be done before microtia reconstruction, it should be placed in the posterior temporal region. Doppler of the superficial temporal artery at the time of surgery is recommended for safe placement of the implant posteriorly.

3. Reconstructive options for microtia, and the timeline of each option

-Ear prosthetic: An ear prosthetic can be made by an anaplastologist with experience in microtia, and can be attached to the remnant ear by adhesives at an early age. Factors involved in age to initiate prosthetic use is the developmental age to be able to care for the prosthetic, not pull it off, and is often considered at the start of school. Bone anchored devices can be implanted to allow for a more secure attachment of the prosthetic after the age of 5 years.

-MEDPOR® Ear Reconstruction: MEDPOR® reconstruction utilizes a porous high-density polyethylene implant covered with a well vascularized fascial flap, typically the superficial temporoparietal fascia, which is then covered in skin grafts. This is typically done as a single stage surgery. The earliest MEDPOR reconstruction can be done is age 3-5 years.

-Autologous cartilage reconstruction: Costal cartilage from ribs 6-9 can be used to create a framework with is typically covered by the local skin of and around the microtia remnant. This surgery is completed in 2-4 stages, depending on the surgeon’s technique. The earliest cartilage reconstruction can be performed is dependent on the size of the child, the amount of rib cartilage available, the surgeon’s technique, and the grade of microtia. Typically, this is age 5-9 years.

*Advantages and disadvantages exist for all reconstructive options. Microtia surgery is a demanding surgery. Each technique has risks, particularly for wound complications. Parents should be guided to meet the experienced surgeon to determine the best reconstructive option for the individual needs of the child.
*Often a prosthetic is considered after failure of other reconstructive techniques.

4. Different techniques using autologous cartilage for microtia reconstruction

-Burt Brent Technique: This technique uses autologous costal cartilage to classically reconstruct the ear in 4 stages.

1st stage: Costal cartilage is harvested and carved. The cartilage framework is then placed under the retro and peri-auricular skin of the remnant.
2nd stage: Transposes the lobule to the framework.
3rd stage: Elevation of the ear with the use of a skin graft placed in the posterior aspect of the ear.
4th stage: Creation of a tragus from contralateral conchal cartilage, and deepening the conchal bowl of the construct.
Satoru Nagata/Francoise Firmin Techniques: These techniques use autologous costal cartilage to classically reconstruct the ear in 2 stages.

1st stage: Costal cartilage is harvested and carved. The cartilage framework is then placed under the retro and peri-auricular skin of the remnant, with concurrent lobule transposition.

2nd stage: Elevation of the ear with the use of banked pieces of costal cartilage from the 1st stage, skin grafts, and often vascularized fascial grafts.

5. Optimal timing for ear canal reconstruction, depending on the microtia reconstruction technique planned
- Microtia technique planned: Timing of ear canal surgery
  - Ear Prosthetic: ear canal surgery can be done at any time.
  - MEDPOR® reconstruction: ear canal surgery is ideally done BEFORE or at the time of microtia reconstruction (concurrent surgery). Canal surgery after MEDPOR® risks exposure/extrusion of the implant and/or infection which is poorly tolerated by MEDPOR® implants.
  - Autologous costal cartilage reconstruction: ear canal surgery is ideally done AFTER microtia reconstruction. This is to maintain the viability of the overlying local skin which covers the cartilage construct in a tissue envelope.

6. Hearing results for ear canal reconstruction, controversies of ear canal reconstruction
- Hearing outcomes: Average hearing gain from atresiaplasty is 24 dB, compared to 38 dB from bone anchored devices. Atresiaplasty achieves < 30 dB PTA or ABG in ~70% patients if hearing assessed < 6 months after surgery, however this deteriorates to ~50% in patients assessed > 12 months after surgery.
- Patients with canal stenosis receiving surgery obtain hearing results approximately 10 dB better than patients with canal atresia.
- Considerations for patients with canal reconstruction: swimming may lead to issues of recurrent ear drainage (7-24%). There is a concern for the rate of recurrent canal stenosis (11-35%) postoperatively. Parents need to be educated on the required maintenance and care of the reconstructed ear canal, which may require regular cleaning.
- Ear canal reconstruction may be necessary when there is concern for cholesteatoma.

7. What is a general timeline for parents with a child with microtia/atresia?
Age: Intervention recommended for the hearing needs of the child
Newborn: Hearing screening
1-3 months: Sleep deprived ABR
6-9 months: Behavioral audiogram
3-12 months: Consultation on hearing devices (bilateral cases should be fit for aid by 6 months of age)
12 months-5 years: Annual audiological testing and surveillance
5 years: Consider CT scan for hearing implants, canal reconstruction candidacy. Consider waiting for surgery to implant bone anchored device until after or coordinated with concurrent microtia reconstruction.

Age: Intervention recommended for microtia reconstruction
Newborn: Consultation with reconstructive surgeons for parental counselling regarding reconstructive options for the future
3-5 years: Earliest age for consideration for MEDPOR® reconstruction. Any ear canal surgery should be done BEFORE or concurrently with MEDPOR reconstruction.
5 years: Earliest age for osseointegrated 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: Earliest age for autologous cartilage reconstruction. Any ear canal surgery is ideally done AFTER microtia reconstruction.

REFERENCES:


