




# WORLD SKULL BASE E-LEARNING MATERIAL

## Microtia

# Microtia

<b>Microtia</b>	
<i>Classification and external resources</i>	
	
Unilateral grade III microtia (more often affecting the right ear).	
<b>ICD-10</b>	Q17.2 <sup>[1]</sup>
<b>ICD-9</b>	744.23 <sup>[2]</sup>
<b>OMIM</b>	600674 <sup>[3]</sup>
<b>DiseasesDB</b>	29876 <sup>[4]</sup>
<b>eMedicine</b>	ped/3003 <sup>[5]</sup>

**Microtia** is a congenital deformity where the pinna (external ear) is underdeveloped. A completely undeveloped pinna is referred to as anotia. Because microtia and anotia have the same origin, it can be referred to as **microtia-anotia**.<sup>[1]</sup> Microtia can be unilateral (one side only) or bilateral (affecting both sides). Microtia occurs in 1 out of about 8,000–10,000 births. In unilateral microtia, the right ear is most commonly affected. It may occur as a complication of taking Accutane (isotretinoin) during pregnancy.<sup>[6]</sup> Perhaps one of the better-known people to suffer from microtia is KISS guitarist and frontman Paul Stanley.

## Classification

There are four grades of microtia:

- Grade I: A less than complete development of the external ear with identifiable structures and a small but present external ear canal
- Grade II: A partially developed ear (usually the top portion is underdeveloped) with a closed [stenotic] external ear canal producing a conductive hearing loss.
- Grade III: Absence of the external ear with a small peanut-like vestige structure and an absence of the external ear canal and ear drum. Grade III microtia is the most common form of microtia.
- Grade IV: Absence of the total ear or anotia.

## Medical treatment

The goal of medical intervention is to provide the best form and function to the underdeveloped ear.

### Hearing

Typically, testing is first done to determine the quality of hearing. This can be done as early as in the first two weeks with a BAER test (Brain Stem Auditory Response Test).<sup>[7][1]</sup> At age 5–6, CT or "CAT Scans" of the middle ear can be done to elucidate its development and clarify which patients are appropriate candidates for surgery to improve hearing. For younger individuals, this is done under sedation.

The hearing loss associated with congenital aural atresia is a conductive hearing loss—hearing loss caused by inefficient conduction of sound to the inner ear. Essentially, children with aural atresia have hearing loss because the sound cannot travel into the (usually) healthy inner ear—there is no ear canal, no eardrum, and the small ear bones (malleus/hammer, incus/anvil, and stapes/stirrup) are underdeveloped. "Usually" is in parentheses because rarely, a child with atresia also has a malformation of the inner ear leading to a sensorineural hearing loss (as many as 19% in one study).<sup>[8]</sup> Sensorineural hearing loss is caused by a problem in the inner ear, the cochlea. Sensorineural hearing loss is not correctable by surgery, but properly fitted and adjusted hearing amplification (hearing aids) generally provide excellent rehabilitation for this hearing loss. If the hearing loss is severe to profound in both ears, the child may be a candidate for a cochlear implant (beyond the scope of this discussion).

Unilateral sensorineural hearing loss was not generally considered a serious disability by the medical establishment before the nineties; it was thought that the afflicted person was able to adjust to it from birth. In general, there are exceptional advantages to gain from an intervention to enable hearing in the microtic ear, especially in bilateral microtia. Children with untreated unilateral sensorineural hearing loss are more likely to have to repeat a grade in school and/or need supplemental services (e.g., FM system – see below) than their peers.<sup>[9][10]</sup>

Children with unilateral sensorineural hearing loss often require years of speech therapy in order to learn how to enunciate and understand spoken language. What is truly unclear, and the subject of an ongoing research study, is the effect of unilateral conductive hearing loss (in children with unilateral aural atresia) on scholastic performance. If atresia surgery or some form of amplification is not used, special steps should be taken to ensure that the child is accessing and understanding all of the verbal information presented in school settings. Recommendations for improving a child's hearing in the academic setting include preferential seating in class, an FM system (the teacher wears a microphone, and the sound is transmitted to a speaker at the child's desk or to an ear bud or hearing aid the child wears), a bone conducting hearing aid, or conventional hearing aids. Age for BAHA implantation depends on whether you are in Europe (18 months) or the US (age 5). Until then it is possible to fit a Baha on a softband<sup>[11][12]</sup>

It is important to note that not all children with aural atresia are candidates for atresia repair. Candidacy for atresia surgery is based on the hearing test (audiogram) and CT scan imaging. If a canal is built where one does not exist, minor complications can arise from the body's natural tendency to heal an open wound closed. Repairing aural atresia is a very detailed and complicated surgical procedure which requires an expert in atresia repair.<sup>[1]</sup> While complications from this surgery can arise, the risk of complications is greatly reduced when using a highly experienced otologist. Atresia patients who opt for surgery will temporarily have the canal packed with gelatin sponge and silicone sheeting to prevent closure. It must be stressed that many surgeons believe that ear canal reconstruction is unnecessary and overcomplicated and that very good hearing is possible with modern hearing aids which can be hidden under the skin.

In cases where a later surgical reconstruction of the external ear of the child might be possible, positioning of the Baha implant is critical. It may be necessary to position the implant further back than usual to enable successful reconstructive surgery – but not so far as to compromise hearing performance. If the reconstruction is ultimately successful, it is easy to remove the percutaneous BAHA abutment. If the surgery is unsuccessful, the abutment can be replaced and the implant re-activated to restore hearing.

## External ear

The age when outer ear surgery can be attempted depends upon the technique chosen. The earliest is 7 for Rib Cartilage Grafts. However, some surgeons recommend waiting until a later age, such as 8–10 when the ear is closer to adult size. External ear prostheses have been made for children as young as 5.

For auricular reconstruction, there are several different options:

1. *Rib Cartilage Graft Reconstruction*: This surgery may be performed by specialists in the technique.<sup>[13]</sup> It involves sculpting the patient's own rib cartilage into the form of an ear. Because the cartilage is the patient's own living tissue, the reconstructed ear continues to grow as the child does. In order to be sure that the rib cage is large enough to provide the necessary donor tissue, some surgeons wait until the patient is 8 years of age,<sup>[13]</sup> however, some surgeons with more experience with this technique may begin the surgery on a child aged six.<sup>[14]</sup> This surgery varies from two to four stages depending on the surgeon's preferred method. The major advantage of this surgery is that the patient's own tissue is used for the reconstruction.
2. *Reconstruct the ear using a polyethylene plastic implant (also called Medpor)*: This is a 1–2 stage surgery that can start at age 3 and can be done as an outpatient without hospitalization. Using the porous framework, which allows the patient's tissue to grow into the material and the patient's own tissue flap, a new ear is constructed in a single surgery. A small second surgery is performed in 3–6 months if needed for minor adjustments. This surgery should only be performed by experts in the techniques involved.<sup>[1]</sup>
3. *Ear Prosthesis*: An auricular (ear) prosthesis is custom made by an anaplastologist to mirror the other ear.<sup>[15]</sup> Prosthetic ears can appear very realistic. They require a few minutes of daily care. They are typically made of silicone, which is colored to match the surrounding skin and can be attached using either adhesive or with titanium screws inserted into the skull to which the prosthetic is attached with a magnetic or bar/clip type system. These screws are the same as the BAHA (bone anchored hearing aid) screws and can be placed simultaneously. The biggest advantage over any surgery is having a prosthetic ear that allows the affected ear to appear as normal as possible to the natural ear. The biggest disadvantage is the daily care involved and knowing that the prosthesis is not real.

## Related conditions

Aural atresia is the underdevelopment of the middle ear and canal and usually occurs in conjunction with microtia. Atresia occurs because patients with microtia may not have an external opening to the ear canal, though. However, the cochlea and other inner ear structures are usually present. The grade of microtia usually correlates to the degree of development of the middle ear.<sup>[1]</sup> Microtia is usually isolated, but may occur in conjunction with hemifacial microsomia, Goldenhar Syndrome or Treacher-Collins Syndrome.<sup>[1]</sup> It is also occasionally associated with kidney abnormalities (rarely life-threatening), and jaw problems, and more rarely, heart defects and vertebral deformities.<sup>[1]</sup>

## References

- [1] <http://apps.who.int/classifications/icd10/browse/2010/en#/Q17.2>
- [2] <http://www.icd9data.com/getICD9Code.aspx?icd9=744.23>
- [3] <http://omim.org/entry/600674>
- [4] <http://www.diseasesdatabase.com/ddb29876.htm>
- [5] <http://www.emedicine.com/ped/topic3003.htm>
- [6] Pretest self assessment and review for the USMLE, pediatrics, 12th edition, question 84, general pediatrics

## External links

- Craniofacial Anomalies Program ([http://children.photobooks.com/directory/profile.asp?dbase=main&setsize=5&pic\\_id=0083419](http://children.photobooks.com/directory/profile.asp?dbase=main&setsize=5&pic_id=0083419)) at Children's Hospital Boston
- Microtia Repair (<http://www.microtia.us.com/>)

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- Atresia Repair — University of Virginia (<http://www.medicine.virginia.edu/clinical/departments/otolaryngology/atresia/>)
  - Microtia Reconstruction at Cedars-Sinai (<http://www.cedars-sinai.edu/Patients/Programs-and-Services/Microtia/>)
  - Bennun RD, Mulliken JB, Kaban LB, Murray JE (December 1985). "Microtia: a microform of hemifacial microsomia". *Plast. Reconstr. Surg.* **76** (6): 859–65. doi: 10.1097/00006534-198512000-00010 (<http://dx.doi.org/10.1097/00006534-198512000-00010>). PMID 4070453 (<http://www.ncbi.nlm.nih.gov/pubmed/4070453>).
  - Goldenhar Family Support Group (UK) (<http://www.goldenhar.org.uk/>)
  - Microtia Australia (<http://microtiaaustralia.org.au/>)
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