

Microtia and Congenital Aural Atresia: Clinical Review and Guidance for Primary Health Care Providers

Anthony R Theile, DO, FAAP; Mark E. Chariker, MD, FACS

INTRODUCTION

Microtia (pronounced mī-krō'shē-ə) is a rare birth defect involving the abnormal development of the outer ear with the end result being a malformed auricle. Congenital aural atresia (CAA) is the failed development of the external auditory canal (EAC), which is routinely seen with microtia.¹ The severity varies widely and may involve one or both ears.

CASE

A term baby boy weighing 8 pounds 4 ounces was born to a 27-year-old gravida 2 and para 1 Caucasian female via spontaneous vaginal delivery. No complications during pregnancy were noted. Prenatal labs were unremarkable with no reports of tobacco, alcohol, or substance abuse. Maternal medications include prenatal vitamins. Apgar scores were 8¹ and 9⁵. The pediatrician at delivery noted a normal physical exam with exception for the ears. The baby had severely malformed ears with very minimal auricles and no visible external auditory canals.

As the pediatrician at the delivery, what is your plan of care? This may presumably be the first time you have ever seen microtia in a baby. What do you tell the parents? The worst fear for any expecting parent is that the health or welfare of their baby is at risk. A visible birth defect like microtia (classic type) is something that cannot be easily disguised and bears social stigmata. The parents often feel that they are responsible for causing the deformity. This is almost never the case. The discussion with the parents needs to be factual, alleviate anxiety/guilt and, most importantly, offer reasonable expectations. The authors aim to provide the general practitioner with the tools needed to deal with these challenging patients and their families.

Dr Theile is Perinatal/Neonatal fellow at the University of Louisville, and Dr Chariker is with the Aesthetic Plastic Surgery Institute in Louisville. Corresponding author: Anthony R. Theile, DO, FAAP; Perinatal/Neonatal fellow, University of Louisville, 571 S. Floyd St, Suite 342, Louisville, KY 40202; e-mail: arthei02@gwise.louisville.edu.

EPIDEMIOLOGY AND ETIOLOGY

Microtia is not very common with an incidence range from 1 in 5,000 to 1 in 20,000 births.^{1, 2} Unilateral microtia and CAA are much more common than bilateral defects (10%), with an increased frequency on the right side. Boys are affected more frequently than girls, and whites and blacks tend to be least affected compared to Hispanics and Asians.³

The cause of microtia is still largely unknown, but most authorities would agree that the etiology is multifactorial. The external ear begins to develop in the sixth week of gestation and some reports implicate the drugs thalidomide and isotretinoin (Accutane) as causative agents.⁴⁻⁶ Most of the cases of microtia and CAA are isolated but can occur with other syndromes, most notably Goldenhar syndrome (oculoauriculovertebral dysplasia). Cervical vertebral defects are common, although associated neurologic symptoms are uncommon. Hereditary etiologies are infrequent.²

GRADING SYSTEM

Since microtia occurs along a spectrum of severity, a classification system was developed to better grade the irregularities.

MICROTIA AND CONGENITAL AURAL ATRESIA

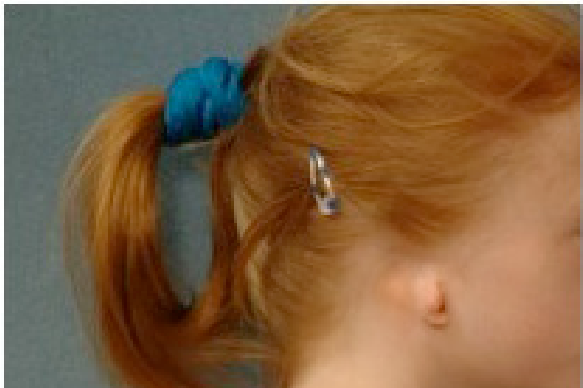


Figure 1. Type II microtia (different patient from the case presented)

Several systems exist but generally Type I microtia implies a relatively normal appearing ear but smaller in size. Type II is a smaller and more abnormally shaped ear while Type III is the classic “peanut” appearing ear. Type IV has been regarded as complete absence of the ear called anotia. All types can have a stenotic or absent external auditory canal.^{1, 5}

CASE CONTINUATION: Specialist care in infancy

The baby was taken to the newborn nursery for routine care. The baby’s pediatrician had never seen this before and did not know what to tell the parents. This, of course, enhanced their anxiety and so, after the baby was discharged, they went to see another pediatrician, and they were ultimately referred to a third doctor. Fortunately, the third doctor was an experienced ENT and informed the parents that most children with microtia have normal inner ears and that corrective surgery was definitely an option. An auditory brainstem response (ABR) test was performed demonstrating normal bilateral nerve function. The baby was fitted with hearing amplification devices at age 5 months for his conductive hearing loss.

INITIAL EVALUATION

When a baby is found to have microtia, the parent’s primary concern will be focused on the child’s ability to hear. If the microtia is isolated and no other major congenital anom-

alies are present, the first thing that needs to be ascertained is the hearing capability, because about 10% to 15% will have a sensorineural hearing loss.¹ However, it is very rare to find patients with microtia who are completely deaf and most patients with unilateral microtia and atresia have normal hearing in the contralateral ear. These children adjust to life with monaural hearing and often do not require hearing aid devices. The biggest challenge the children face will be localization of sounds in a noisy environment. Most children with bilateral microtia have a conductive hearing loss and implementation of hearing aid devices should be initiated within the first month of life.² To promote optimal developmental outcomes, speech therapy is sometimes required, especially in those patients with bilateral microtia.

A computed tomography (CT) scan is required to assess for middle and inner ear anomalies, however, the optimal time in which to obtain the CT scan is still debated. Some will obtain the CT in the first months of life because of the 4% to 7% chance of cholesteatoma formation in the atretic ear; however, this can take years to become problematic.^{1, 7} Others will wait until the inner ear structures are fully mature to obtain the first head CT, around 4 to 6 years of life. At this time one could more accurately assess if the patient is a candidate for the “drill out” procedure. The drill out procedure consists of carving out a canal through the bone in which sound conductance is better achieved. Candidacy for atresia surgery is based on a 10-point scoring system including criteria such as ossicle location and facial nerve position.⁸ For example, a score of 7/10 would be 70% chance of achieving normal or near normal hearing.

CASE CONTINUATION: CT scan at 2 years of age

The baby had his first CT scan of the inner ear at 2 years of age. It revealed bilateral ear atresia with a significant amount of middle ear hypoplasia. His score on the right ear was 6/10 and his left ear was 7/10. The inner ear structures were normal.

PSYCHOSOCIAL IMPACT

After the issues surrounding the child's ability to hear have been addressed, the parents will focus on the physical deformity. Most parents want the ear(s) fixed immediately so their child can live a normal life. However, surgical correction cannot occur until the child reaches school age (surgical correction discussed later). This news will not sit well with most families, and the parents will need counseling to understand that even with surgery, the ears may never look completely normal and their child's hearing will never be 100%.²

In the first few years of life, the child is unaware of his/her deformity. It is not until children reach the age of about three years that he/she develop the understanding that he/she are not like their peers. It can be harder for the child with unilateral microtia because one ear is clearly different than the other. Parents should be truthful with the child, and explain that he/she was born with a different ear and that it can be fixed later. Treating him/her like any other child is the best approach.² Other children will often make hurtful remarks about the child's microtic ear(s) and this affects the whole family. The years leading up to the corrective surgery can be hard not just for the child but the whole family as well. Providing anticipatory guidance for the parents should be a principal concern for the primary care practitioner. A clear understanding of the disorder and how it and treatment options affect developmental milestones is critical.

CORRECTIVE SURGERY

The previous standard has been the 3 or 5 stage technique championed by Dr. Burt Brent.⁹ Today, many surgeons have elected to use the ear cartilage or a plastic material called Medpore (Porex Surgical, Inc, College Park, Georgia) covered by a tissue flap harvested from under the scalp known as the temporal parietal fascial flap. This is done in one or two stages. These procedures are generally performed as an overnight stay in the hospital or as an outpatient procedure (See Figures 1-3).¹⁰



Figure 2. Medpore graft covered with fascial flap and skin graft sewn in place and a suction drain is placed last.

Starting the process of ear reconstruction is dependent on several factors including the method of reconstruction, the anatomic age of the child and the developmental maturity of the child. The ideal age to start the repair is prior to starting formal education around 5 or 6 years old, but the firmness of the rib cartilage used for the reconstruction sometimes occurs a few years later. In addition, around 6 years of life the *normal* ear is almost fully developed so that one can construct the *new* ear with reasonable symmetry. Therefore, the procedure is usually performed between ages 5 to 9.²

Selection of a surgeon with extensive experience is important due to the technical difficulty in learning this procedure. In reference to the ear canal, this is addressed at a later time. Most often the deformity is unilateral and the hearing function is normal on the normal ear side. In these cases where there is normal hearing on the opposite side, a pseudocanal is made to reflect a canal (for cosmetic appeal) without drilling the bone, which puts the facial nerve at risk for injury.^{1, 2}

CASE CONTINUATION: Repair procedures in middle childhood

The patient had five surgeries performed over 18 months by Dr Burt Brent. He had the autologous rib cartilage repair starting at age 7.5 years. No complications occurred with any of the operations. At the age of 13, he had

MICROTIA AND CONGENITAL AURAL ATRESIA

the "drill out" procedure done in his left ear only, performed by Dr Robert Jahrsdoefer, a renowned otolaryngologist. He wears a tiny, well-disguised hearing aid that fits in his "man-made" ear canal.



Figure 3. Preoperative and postoperative views

SUMMARY

Congenital microtia and aural atresia are relatively uncommon anomalies for the general practitioner, but when they do appear, provide much angst to the family. Swift reassurance needs to be given to the parents as well as providing accurate expectations right after birth. The psychosocial ramifications for the whole family can be significant and must be addressed. In the initial evaluation, one must screen for other syndromes as well as evaluate the child's hearing and provide speech therapy or hearing aid devices in a timely fashion. Choosing an experienced surgeon is equally as important because the margin of error is small. A classic case of bilateral microtia was presented along with illustrations of a newer surgical technique.

CASE CONTINUATION: Adult follow-up

The patient growing up learned to read lips to make up for his hearing deficiency. His language skills are normal and indistinguishable from his peers. He is now 28 years old, recently received his PhD in molecular biology, and is happy with the cosmetic appearance of his ears. I know this because he is my brother.

I would like to acknowledge Dr John Roberts, Dr Paula Radmacher, and Dr Deborah Davis.

REFERENCES

1. Kelley PE, Scholes MA. Microtia and congenital aural atresia. *Otolaryngol Clin North Am.* Feb 2007;40(1):61-80, vi.
2. Brent B. The pediatrician's role in caring for patients with congenital microtia and atresia. *Pediatr Ann.* Jun 1999;28(6):374-383.
3. Beahm EK, Walton RL. Auricular reconstruction for microtia: part I. Anatomy, embryology, and clinical evaluation. *Plast Reconstr Surg.* Jun 2002;109(7):2473-2482; quiz following 2482.
4. Quatela VC, Thompson SK, Goldman ND. Microtia reconstruction. *Facial Plast Surg Clin North Am.* May 2006;14(2):117-127, vi.
5. Tollefson TT. Advances in the treatment of microtia. *Curr Opin Otolaryngol Head Neck Surg.* Dec 2006;14(6):412-422.
6. Jahn AF, Ganti K. Major auricular malformations due to Accutane (isotretinoin). *Laryngoscope.* Jul 1987;97(7 Pt 1):832-835.
7. De la Cruz A, Hansen, MR. Reconstruction surgery of the ear: auditory canal and tympanum. In: Cummings CW FP, Harker LA, et al, ed. *Otolaryngol Head and Neck Surgery.* 4th ed. Philadelphia: Mosby; 2004:4439-4444.
8. Jahrsdoefer RA, Yeakley JW, Aguilar EA, Cole RR, Gray LC. Grading system for the selection of patients with congenital aural atresia. *Am J Otol.* Jan 1992;13(1):6-12.
9. Brent B. Technical advances in ear reconstruction with autogenous rib cartilage grafts: personal experience with 1200 cases. *Plast Reconstr Surg.* Aug 1999;104(2):319-334; discussion 335-318.
10. Romo T, 3rd, Presti PM, Yalamanchili HR. Medpor alternative for microtia repair. *Facial Plast Surg Clin North Am.* May 2006;14(2):129-136, vi.