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CLINICAL PEARL
Diagnosing and Treating Microtia
Mitchell A. Stotland, MD, MS

Classification
Severe hypoplasia of the external ear (microtia) is commonly encountered in any pediatric plastic surgical practice. A useful clinical classification of these anomalies was proposed by Tanzer [1]:

A. Anotia (complete absence of ear)
B. Microtia
    1. constricted (cup or lop) ear
    2. cryptotia (in which the top of the auricle is hidden under the scalp)
    3. hypoplasia of the entire superior third of the auricle
C. Hypoplasia of the middle third of the auricle
D. Hypoplasia of the superior third of the auricle
E. Prominent ears
    1. complete hypoplasia of the auricle (external ear) with atresia of the external auditory canal
    2. complete hypoplasia of the auricle without atresia of the external auditory canal

Lobular microtia.

Epidemiology
Based on analyses of large birth registries of congenital malformations [2-4], the incidence of microtia has been estimated at between 1 in 4,000 to 1 in 12,000 births. Microtia affects about 1.5 times more male children than female, and those with the condition are nearly three times more likely to be of Hispanic descent, and almost twice as likely to be of Asian descent, than to be black or white.

Signs and Symptoms
Microtia is noted on visual inspection at birth as an obvious hypoplastic external ear deformity. A variable extent of middle ear pathology, with associated conductive hearing loss, is expected.

Pertinent History, including Developmental History
Key clinical information should be obtained at the initial visit (typically when the child is an infant): pregnancy and labor complications, maternal drug use or toxin exposure, and family history of craniofacial or other anomalies. A developmental assessment should be made, specifically inquiring about the child's behavior, school performance, social interaction with peers, and self-esteem, to inform expectations of patient cooperation and compliance during the reconstructive process.

Physical Exam
When evaluating a school-age child, clinical findings specific to microtia should be documented.

1. Describe the anatomic extent of the auricular defect in detail. It is useful when communicating findings and in planning reconstruction to outline the components of the auricle that are present or absent (e.g., lobule, tragus, constricted concha, severe helical constriction, etc.). The greater the extent of auricular hypoplasia, the greater the amount of cartilage needed and the more complex the framework fabrication will be.
2. Rule out the presence of additional auricular anlagen (embryologic defects) such as pre-auricular tags, pits, sinus tracts, or other chondrocutaneous remnants that may lie anywhere along the embryologic line from the oral commissure to the temporal region. These additional anomalous structures will require surgical removal.
3. Determine whether there is temporal bone hypoplasia and whether there is soft-tissue hypoplasia on the microtic side. Surgical placement of an aesthetically pleasing ear will not achieve the goals of the patient or surgeon if it is hidden from view because it is placed on a portion of the skull that is depressed inward; thus, temporal hypoplasia may require adjustment of the location of framework implantation (i.e., more anterior or posterior), or fabrication of a thicker construct with an underlying cartilage "wedge," in order to improve projection when viewed from the front.
4. In the child with microtia, maturity may bring a progressive dental/skeletal canting, with asymmetric maxillary growth and dental eruption occurring in response to the mandibular hypoplasia on the side where the microtia is present. A simple clinical test used to demonstrate this is to have the patient

bite down on a wooden tongue depressor that is placed horizontally into the mouth, as far posteriorly as is comfortable. Relative to the sagittal plane of the patient's face, the tongue blade will tend to slant upwards towards the involved side. An orthodontist should participate in the routine evaluation of the patient with microtia.

5. The hypoplastic hemimandible reveals itself in a noticeable ipsilateral "chin point" in repose, with further lateral deviation of the jaw towards the microtic side evident when the patient opens his or her mouth. It is helpful to perform the mandibular lengthening procedure used to surgically correct the hypoplastic mandible prior to embarking on the ear reconstruction so that proper placement of the ear framework can be achieved.
6. The presence of hairline abnormalities may influence the placement of the ear framework and the possible need for management of hair-bearing skin overlying the reconstructed ear (e.g., by means of electrolysis or laser treatments).

Looking for and documenting possible associated craniofacial syndromes, to ensure that they are addressed and to distinguish between pre-existing and iatrogenic conditions, is also part of any proper examination of a child with microtia. Evaluate seventh cranial nerve function, looking for asymmetry of facial motor activity which is not uncommon in patients with microtia. Look for the presence of macrostomia (the congenital form of which is often referred to as a Tessier #7 facial cleft), which is associated not uncommonly with severe external ear deformities. Rule out any unusual neck mass or sinus that may represent a branchial cleft cyst. Inspect the ocular region for features that may be associated with Treacher-Collins syndrome, Nager syndrome, or Goldenhar syndrome (epibulbar dermoid cysts on the conjunctival surface of the lower lids, microphthalmia, or colobomas of the lid, iris, or retina; partial absence of the medial lower eyelid lashes, paucity of lower lid skin; and down-sloping of the palpebral fissures).

Evaluation by a geneticist may be indicated to identify any associated malformations (e.g., renal anomalies, defects within the oculoauricular vertebral spectrum, mandibulofacial dysostosis syndromes, etc.).

Treatment Options

Inner ear. Bone-conductive hearing aids are indicated for severe bilateral hearing loss (i.e., for bilateral microtia or when there is abnormal hearing in the contralateral ear) within weeks of birth. These devices are somewhat awkward and stigmatizing for an older child to wear, and so the implementation of a bone-anchored hearing apparatus is an appealing option for many patients when they get older.

Surgical exploration of the middle ear, involving drilling of the temporal bone to create a neo-canal, and fabrication of a tympanic membrane using graft material, is an approach offered by some otologists with particular expertise in this area. Consideration of this challenging procedure is more common in cases of bilateral microtia, but not exclusively so [5]. In cases of autogenous ear repair, the middle ear

exploration should occur *after* the reconstruction in order to preserve the blood vessels of the overlying skin pocket and allow for strategic placement of the otologist's incision.

Outer ear. Reconstruction of the external framework may utilize either autogenous tissue or an alloplastic implant. In alloplastic reconstruction, the outer ear is constructed using an artificial material (Medpor, a porous polyethylene, is popular; silicone, though largely of historical note, is still used in some places). The construct is wrapped under an inferior flap of temporoparietal fascia and covered with a skin graft. There are well-established pros and cons of the two techniques (autogenous versus alloplastic) [6-8]. The benefits of alloplastic reconstruction include reduced donor site morbidity and the ability to perform reconstruction at a younger age (the patient can be as young as 4-5 years of age, whereas autogenous reconstruction is usually not performed until the patient is 6 to allow for harvest of sufficient size rib cartilage). The disadvantages of alloplasty are increased framework exposure, concern over long-term permanence of the implant, and the necessary use of the temporoparietal fascial flap which sacrifices a valuable salvage procedure in the event of an infection. The majority of reconstructive ear surgeons today employ autogenous material.

The modern technique of autogenous total ear reconstruction was developed by Tanzer and popularized and modified by Brent, Nagata, and others [9-12]. Reconstruction involves a staged approach, which varies depending on the surgeon. The Nagata modification, for example, performs reconstruction in two stages. At the second stage, the concha and tragus are reconstructed. Another common sequence, utilizing the Brent technique, is as follows. (Stages may be combined in the case of bilateral microtia.)

1. Stage one entails the fabrication and insertion of a costochondral auricular framework. The cartilage substrate for the framework is harvested via an oblique incision measuring roughly 6-8 centimeters positioned overlying the sixth and seventh costal cartilages. This region of costal cartilage synchondrosis is used to form the "base-block." The first free-floating costal cartilage rib (eighth) is used for the helical rim. Prior to closing the donor site, the chest should be checked for a collapsed lung. Injury to the parietal pleura is not an uncommon occurrence during costal cartilage harvest, but management is usually straightforward.

   The base-block is trimmed and tailored using a template to guide shape and sizing. The free rib cartilage is thinned and made flexible enough to wrap around the periphery of the base-block, fashioning a natural helical contour. The two components of the framework are spliced together. It is important to exaggerate the contours of the framework since the overlying skin flap is thicker than that of a normal ear and will tend to obscure the sculpted detail. The back wall of the concha, antitragus, scapha, and triangular fossa may be carved with sharp gouges, fine curettes, and a scalpel.

Finally, a pocket is created into which the framework will be placed. The incision for access may be placed anterior or superior to the microtic remnant. Any vestigial cartilage (i.e., the remnant) is removed at this time. The dissection is carried out beyond the immediate outline of the framework to facilitate draping of the skin over the cartilage and into its sculpted grooves and nooks. An adequate seal is imperative to maintain apposition of the skin to the framework. The ear is dressed with a soft Vaseline dressing to maintain contour.

2. Stage two occurs at least 2 months later and involves repositioning of the lobule, which, in microtia, is typically malpositioned and oriented vertically. The lobule is surgically elevated, except for a small vascular pedicle, and rotated around into a more normal transverse position, overlying the caudal portion of the implanted cartilage framework. A postauricular sulcus is concurrently created and the hairline advanced into it. An incision is placed several millimeters outside and all along the framework from the helical root to the lobule. Soft tissue is left on the posterior surface of the framework to allow for skin graft take.

3. At stage three, soft tissue is removed from the planned concha and lined with a skin graft harvested from the posterior lobule or posterior surface of the opposite ear. The tragus is reconstructed by combining a skin flap with a composite chondrocutaneous graft harvested from the opposite ear.

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Mitchell A. Stotland, MD, MS, is an associate professor of surgery, associate professor of pediatrics, and director of the Craniofacial Anomalies Program at Dartmouth-Hitchcock Medical Center in Lebanon, New Hampshire. His interests include shared decision making and facial reconstruction in adolescents, surgical device design, and facial perception and stigma as they pertain to congenital anomalies and aging.

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