INTRODUCTION

Microtia means “little ear” and is derived from the Latin words, micro + otia. It can be associated with absence of the ear canal known as aural atresia. Microtia can range from a small loss or displacement of ear structures to complete anotia, or entire absence of the external ear. Hearing and middle ear function is often affected. In most cases, there is a small remnant of cartilage and earlobe. These remnants are used in later stages of reconstructions and should not be removed.

There is a range of types which have been classified as follows:

<table>
<thead>
<tr>
<th>Type</th>
<th>Description</th>
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<tbody>
<tr>
<td>Type I</td>
<td>Small pinna&lt;br&gt; EAC present&lt;br&gt; earlobe present&lt;br&gt; e.g. cup ear, lop ear</td>
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<tr>
<td>Type II</td>
<td>Absent pinna&lt;br&gt; with EAC&lt;br&gt; auricular remnant anteriorly earlobe</td>
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<tr>
<td>Type III*</td>
<td>Absent pinna&lt;br&gt; No EAC&lt;br&gt; auricular remnant anterior earlobe</td>
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<tr>
<td>Type IV</td>
<td>Anotia&lt;br&gt; no earlobe no EAC&lt;br&gt; no pinna&lt;br&gt; no remnant</td>
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Subclassification:

- subtype A – no associated syndrome or other craniofacial anomaly
- subtype B – with associated syndrome or other craniofacial anomaly

*The most common type is III-A

EXAMPLES OF MICROTIA
FIRST ASSESSMENT BY PEDIATRICIAN OR PRIMARY CARE PRACTITIONER
The diagnosis of microtia is usually not made until birth. It is rarely detected by antenatal ultrasound. When first discovered it may cause distress, worry and fears for the parents. Pediatricians may want to thoroughly evaluate the baby for any other associated conditions, such as kidney problems, which are rare but an ultrasound is sometimes done. An MRI or CT scan is generally not necessary until the child is much older, unless there is concern about any central nervous system abnormality. Consultation with an otologist and plastic surgeon is done as soon as possible after birth. A genetic evaluation may be helpful. At the first visit I like to review the condition, offer support, show before and after pictures to the family. I then follow the child yearly until it is time to perform the reconstruction, usually after the child turns 7 or 8 years old.

EPIDEMIOLOGY and CAUSATION
Microtia is a very rare condition occurring once in every 6,000 births. The incidence is much higher in Native Americans. It is estimated that in New Mexico, there are approximately 4 to 5 new cases per year. Microtia is more common in boys (63 per cent) and on the right side (58 per cent). It occurs bilaterally (on both sides) in 10 percent of cases. Five percent of cases have a positive family history. Microtia occurs usually as a random, sporadic event, although it may be associated with certain syndromes. It is important to understand that there is no known causative relation with environmental factors, medications or drugs. Despite many superstitions, it is important to emphasize that the deformity is not caused by anything the mother did before or during the pregnancy. The risk of recurrence within the family is about five percent. During embryological development the parts of the fetus that are destined to form the ear fail to grow for reasons that are unknown. I often compare this to a flower bud which fails to open.

ASSOCIATED CONDITIONS
In some cases, microtia may be associated with other congenital abnormalities of the face and jaw. The most commonly associated anomaly is underdevelopment of the mandible or lower jaw known as Hemifacial Microsomia. This is due to the fact that the mandible and ear develop from the same embryonic structures. In mild cases, no treatment is necessary, but if the mandibular deformity is severe, then corrective orthognathic surgery may be required. Aural atresia refers to the absence an external ear canal. When someone has aural atresia, there is a high incidence of malformation of the external ear, known as Microtia. Atresia/Microtia is frequently isolated, meaning, the patient with atresia and/or microtia has no other health problems. However, approximately 40% of the time, Atresia/Microtia is one symptom of a larger syndrome.
List of associated syndromes:
- Treacher-Collins
- Hemifacial Microsomia
- Goldenhar’s syndrome (oculo-auricalar-vertebral syndrome)
- Crouzon’s syndrome
- Pfeiffer syndrome
- Apert’s syndrome
- Nager / Miller syndrome
- Klippel-Feil
- BOR (Branchio-Oto-Renal) also known as Melnick-Fraser
- Pierre Robin sequence
- CHARGE syndrome
- Chromosome 18q- syndrome (This is only known source of atresia without microtia that can be confirmed by blood test)

ALTERNATIVES FOR RECONSTRUCTION
There are several alternatives for treatment of microtia. The parents and child must weigh these options carefully before making such an important decision.

- **First option:** The first option is simply to wait and do nothing. Although the benefits of ear reconstruction are well known, absence of the external ear poses no long term health risk to the child, therefore, reconstruction is entirely elective. Reconstruction can be done at any time in their life, even as an adult.

- **Second option:** Reconstruction using the patient’s own rib cartilage is by far the best overall option for the child and family for many reasons. Since it is the patient’s own tissues, there is little chance for rejection. The tissue used is alive and grows with the patient. After full healing it is resistant to injury and infection. Note: it is not possible to use the parent’s ribs since this will result in tissue rejection.

- **Third option:** This involves the use of a synthetic framework implanted under the skin or under a flap to avoid harvesting a rib graft. Use of materials such as Teflon or Silicone have been used in the past but often result in extrusion requiring removal. Sometimes these materials are used to supplement a cartilage graft, but in general, are a poor choice for primary or first-time reconstruction. A technique has been described where tissue from the scalp is placed over the plastic material and covered with a skin graft. The results are not as good as those obtained using conventional rib graft techniques.
• **Fourth option:** This involves the use of an external ear prosthesis or artificial ear. In order to accomplish this, the ear remnant is removed and fixation screws are placed on the skull. This makes it virtually impossible to perform conventional reconstruction in the future. Although excellent cosmetic results can be obtained with an artificial ear there is a very high complication rate such as screw site infections. They are typically made of silicone which is colored to match your individual skin and can be attached using 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 Appliance) screws. The skin match is not always perfect and the appearance depends on the skill of the artist who makes the prosthesis. Moreover, the prosthesis can be lost or damaged. Replacement may be difficult or impossible after a certain age. Most insurance plans will not cover the cost of replacement. The maker of the prosthesis does offer monthly insurance payments which must be made for the rest of the patient’s life. If for any reason, the prosthesis cannot be replaced, then there will be exposed metal clips and scarring. If a prosthesis is to be considered, it is best to wait until the child is at least 12-14 years. A prosthesis may be a valuable alternative in cases where the rib graft has failed, or in certain cases where conventional reconstruction is not possible. It is useful for adults who have suffered from cancer or traumatic amputation of the ear. Ordinarily a prosthesis should not be used in children.

**HEARING ISSUES**
The side with microtia generally has no or little hearing due to the lack of an ear canal and ear drum. The middle ear may be functional but will only pick up low tones and vibrations. The inner ear is usually intact. MRI or CT scan are not routinely required when the hearing on the other side is normal. It is imperative to have the other side checked by and ENT doctor to make sure hearing is normal. In cases of bilateral microtia, conductive hearing aids must be worn from early infancy so that normal speech can develop.

If hearing is normal on the opposite side, then ordinarily nothing is done to restore hearing in the affected microtic ear because of the adverse effect on the results of reconstruction. Depending on the severity of the middle ear pathology, surgery may be attempted although surgical attempts to restore hearing may actually interfere with the hearing of the normal ear. On occasion in unilateral cases, a hearing aid may be worn, in which case, the reconstructed ear is essential for fitting the device. When both sides are affected, then restoration of hearing and middle ear function is indicated. This is done after reconstruction working closely with an otologist, or ENT surgeon hearing specialist such as Dr. Karl Horn.
BAHA IMPLANTATION (BONE ANCHORED HEARING APPLIANCE)
For patients who are not good atresia repair candidates, or where a skilled atresia repair surgeon is not available, a BAHA device can be used to correct the hearing loss on the atretic side. Current advances can result in normal hearing for both unilateral or bilateral cases. Surgically implanted BAHA devices use an abutment embedded in the skull to transmit sound by direct conduction through bone to the inner ear, bypassing the external auditory canal and middle ear. A sound processor is attached to the titanium abutment. The implant vibrates the skull and inner ear, which stimulate the nerve fibers of the inner ear, allowing hearing while bypassing all the sources of the conductive hearing loss caused by the atresia. The minimum age in the United States for Baha implantation under FDA guidelines is 5 years old.

INSURANCE COVERAGE AND PAYMENT
Currently, all Medicaid and Salud programs cover all aspects of reconstructive ear surgery. Insurance companies will also cover surgery but they require prior authorization for all stages. For children not eligible for Medicaid, Salud, or insurance, coverage is available for those who qualify through Children’s Medical Services. Dr. Cuadros and Dr. Horn work with the following plans: Presbyterian Health Plan, Cigna, Blue Cross Blue Shield, United Health Care, Tricare, Medicaid, Pres Salud, Molina Salud, Lovelace, Lovelace Salud, and Children’s Medical Services. If your child is not on this list, contact us and we will most likely be able to make arrangements for full coverage of reconstructive surgery.

AGE TO BEGIN RECONSTRUCTION
I have found that the ideal age to begin is when the child is a little older, between 7 and 9 years old because by this time the ear is nearly full grown. Also, the child is more mature and cooperative with the process of reconstruction and can actively participate in the decision to proceed with surgery. Many children are actually enthusiastic about the prospect of a “big” ear. Most parents choose to begin the first stage of reconstruction in the early summer, soon after school is out. Subsequent stages are then scheduled at 3 to 4 month intervals.

The normal ear reaches 85% of the adult size by age 6-7 years old. Reconstructed ears can be expected to grow proportionately as the child matures. Normally, four stages are required to finish the ear, but more may be required for further refinements. Although it is impossible to create an exact duplicate of a normal ear, the goal of surgery is to create a natural replica.
BENEFITS AND MEDICAL NECESSITY OF RECONSTRUCTION
Microtia is a congenital birth defect and monitored by the Birth Registry of the US department of Health and listed in the National Birth Defects Prevention Network. All surgery for birth defects is considered reconstructive in nature. The functional improvements are many, including the ability to wear hearing aids, should they be required. The reconstructed ear allows the wearing of prescription eye wear for vision, and sunglasses for UV protection – especially important in New Mexico. Since one of the most important functions of the face is communication, by restoring a more normal appearance, there is improved social function. It is therefore vital to understand that this type of surgery is *not* cosmetic as defined by the American Medical Association.

ANATOMY AND PROPORTIONS OF THE NORMAL EAR
The ear is made of skin, cartilage and connective tissue. The outer upper portion of the ear is known as the auricle or pinna. The helix and helical rim form the outer edge. The antihelix forms the inner rim dividing superiorly into anterior and posterior helical crura. This is space is the fossa triangularis. The space between the helix and antihelix is the scapha or scaphoid fossa. The concha, or large central depression is divided into the upper cymba and lower cavum by the crus of the helix. The tragus covers the front of the opening of the external auditory canal, or meatus. The lower inner portion of the concha has a prominence known as the anti-tragus, divided by the intertragal notch. The lobule is the lower fleshy part and contains no cartilage.

PREPARATION AND PLANNING FOR THE SURGERY
In the office, before surgery, a template using exposed x-ray film is made of the contralateral normal ear. This is then reversed and used in the operating room for sculpting the framework. Photographs are taken.
STAGES OF RECONSTRUCTION
The technique followed is based precisely on the techniques of Dr. Burt Brent who pioneered modern microtia reconstruction. The surgery is done in four to five separate stages to achieve the goals of surgery. All surgery is done at Presbyterian Hospital. The first stage requires a 3-4 night inpatient admission. Subsequent procedures can be done as an outpatient.

FIRST SURGICAL STAGE - Harvesting, sculpting and implantation of rib graft

This is the longest and most complicated stage of reconstruction, it takes 4 to 5 hours requiring hospitalization for 3 to 4 nights. This and all stages require general anesthesia.

- Harvesting of rib cartilage graft - The 6th, 7th, and 8th ribs are harvested from the opposite chest wall. Great care is taken to protect the delicate lining of the lung.
- Sculpting of framework using custom carving tools. The free floating 8th rib is used to form the helix, or rim of the ear. A base for the ear is made from the conjoined cartilage of the 6th and 7th ribs. They are sewn together to create the framework.
- Creation of subcutaneous pocket. Skin under the proposed new ear site is undermined. The framework is placed into subcutaneous pocket.

For the last several years, we have been using a special implantable pain pump at the rib site in addition to a PCA machine to control the pain.
SECOND SURGICAL STAGE - Transposition of ear lobule

This is done 3 months after the first stage and is not as complicated. It involves transposition of the ear lobule into normal position and removal of any excess remnant.

THIRD SURGICAL STAGE - Creation of concha

Creation of the conchal depression and tragus using a graft using the vestigial remnant.

FOURTH SURGICAL STAGE - Elevation and skin graft and contralateral otoplasty

Elevation or “lifting” of the framework from behind to separate the structure from the head, giving projection. A split thickness skin graft taken from the buttock region, or a full thickness skin graft taken from the opposite ear. Contra-lateral otoplasty - the other ear is pinned back for symmetry. An ear piercing for girls can be done at this time. Additional procedures or stages may be necessary.
RESULTS OF SURGERY

Alysia M, 10 years old underwent 4 stages of reconstruction.

Marco R., 9 years old from Silver City, New Mexico underwent 5 stages of reconstruction including contralateral otoplasty. He also had placement of a BAHA device to improve hearing.
PAIN AND RECOVERY FROM ANESTHESIA AND SURGERY
After the first stage, recovery time is 2-3 weeks. Subsequent stages require a 1-2 week recovery time. Pain is controlled after the first stage with a pain medication infusion and a special anesthetic pain pump. At home the children are given Tylenol with codeine and antibiotics. Parents are instructed on wound care.

POTENTIAL RISKS, SIDE EFFECTS AND COMPLICATIONS
Complications are rare and can arise at both the donor and ear reconstruction sites. They can occur at any stage:

- Inaccurate positioning or poorly designed cartilage framework resulting in a poor result.
- Infection resulting in loss of reconstructed ear.
- Bleeding and hematoma.
- Pain or tenderness after surgery.
- Pneumothorax (collapse of the lung) and atelectasis (incomplete expansion of the lung) at the time of obtaining rib grafts. If severe, may require thoracostomy or chest tube. This has never happened in my patients due to the extreme care taken during surgery.
- Pneumonia
- Partial or total lung collapse due to poor breathing.
- Ischemia (loss of blood supply) of the skin overlying the graft. In extreme cases this can result in skin necrosis of the covering flap and exposure or loss of the graft. Additional surgery in the form a temporo-parietal flap may be necessary.
- Symptomatic protrusion of sutures.
- Need for multiple additional surgeries. Possible need for additional rib graft or implantation of synthetic plastic parts.
- Pressure necrosis due to the patient sleeping on the reconstructed ear.
- Permanent scars including hypertrophic scars and keloids at the ear and at the donor sites, such as buttock where skin graft is taken or scarring of the chest wall incision site.
- Partial or total resorption of cartilage leading to sub-optimal result.
- Anesthesia risks – extremely rare in these health kids.
RESOURCES and CONTACTS

Dr. Karl Horn - An otolaryngologist or ENT surgeon specializing in Otology. Dr. Horn is a leading specialist and performs a variety of procedures to improve hearing including PE tubes, myringoplasties, middle ear reconstruction, cranial base surgery, cochlear implants, and bone-anchored hearing aids (BAHA).

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tel (505) 224-7610

Dr. Luis Cuadros, Pediatric Plastic Surgery
Dr. Cuadros has been in practice in New Mexico since 1988. He is a graduate of Columbia University and trained at Harvard Medical School. He is a member of the American Society of Plastic Surgeons and American Cleft Palate Association. He is a consultant for Healing the Children and a member of Operation Smile. He has been a provider for Children’s Medical Services for 20 years and has extensive experience in surgical management of congenital facial deformities.

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Oral and Maxillofacial Surgery of New Mexico
Drs. Candelaria, Mitchell and Urquhart are experts in oral surgery and mandibular reconstruction
6800A Montgomery Blvd. N.E.
Albuquerque, New Mexico 87109-1425
(505) 881-1130
info@omsanewmexico.com

Other resources:
- New Mexico School for the Deaf, 1060 Cerrillos Road, Santa Fe, NM 87505
  (505) 476-6300 (v/tty/video-phone)
- Children’s Medical Services Statewide Office – Toll Free 1-877-890-4692
- http://www.earsurgery.com
- http://www.earreconstruction.com
- http://www.microtia.net