

**TITLE: Microtia: With An Emphasis On Reconstructive Options**  
**SOURCE: Grand Rounds Presentation, UTMB, Dept. of Otolaryngology**  
**DATE: January 26, 2010**  
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## **INTRODUCTION**

Microtia, the abnormal development of the external ear, is a condition that affects up to two individuals per 10,000 cases although some sources cite a higher incidence of up to 1-2 cases per 7,000-8,000 births. The incidence is higher in Hispanics, Asians, and Native Americans—especially among the Navajo and Eskimos. In addition, there appears to be a higher risk among children born to multiparous mothers although there is only a positive family history in 15% of cases. Microtia tends to affect males more than females with a predilection more for the right ear than the left.

Not unexpectedly, conductive hearing loss (CHL) may be associated in 80-90% of microtia cases although sensorineural hearing loss may be present in up to 15% of affected individuals. Most of the etiology for the CHL is from some degree of congenital atresia of the external auditory canal, but there may be middle ear abnormalities. However, there has been no association between the severity in dysmorphic features of the microtic ear and the degree of hearing impairment. Preserving normal hearing is crucial in proper speech development, and it is generally accepted that additional auditory testing may be delayed for 6-7 months if an affected child passes the newborn hearing screen in the nonmicrotic ear.

Other abnormalities affiliated with microtia include cleft lip or palate, microphthalmia or anophthalmia, cardiac defects, limb development, renal malformation, and holoprosencephaly. Microtia may also be found in congenital birth conditions such as Goldenhar Syndrome or Treacher Collins Syndrome. While often overlooked amidst all these other potential defects to consider, facial nerve dysfunction may also be present.

Marx is credited as the first to attempt classifying the various degrees of microtia around 1926. A grade I microtia referred to an ear that was slightly smaller than a normal ear. The

auricle exhibits a mild deformity but each structure can still be identified. Ears with grade II microtia are distinguished from grade I microtic ears by a size 50-60% smaller than normal ears. The symbolic “peanut ear” is classified under grade III microtia while grade IV refers to anotia. While there have been a number of refinements and amendments since then, the current microtia classification system generally accepted by most is the one developed by Aguilar in 1996. Under this new system, a normal ear is denoted as grade I while grade II encompasses grades I and II from the Marx classification scheme. Grade III still refers to a severely malformed ear but also includes anotia.

## **RECONSTRUCTION**

The first reports of the first attempt at surgical reconstruction of microtia date back to the 1500’s albeit to minimal success. A folded mastoid flap was described by Dieffenbach in the mid-1800’s regarding the repair of a traumatic defect to a monk’s ear. Pierce first introduced the concept of autogenous cartilage in the 1930’s, but it was Tanzer’s work with the subcutaneous placement of an autogenous cartilage graft framework in 1959 that laid the foundation for modern-day auricular reconstruction.

Microtia repair, especially for unilateral cases, is typically delayed until a child reaches six years of age. The size of the ear is approximately 85-95% of its adult dimensions at this age, and performing surgery at this time allows for sufficient costal cartilage development for autogenous reconstruction. Intervention at this age also occurs during a period when most children begin attending school and may be subjected to ridicule and ostracization from their peers if they possessed a deformed ear. If congenital aural atresia is also present, drill-out procedures are usually postponed until age seven at the earliest although some surgeons prefer not to consider such surgeries until after reconstruction is completed so as to minimize any potential disruption to the vascular supply to the repaired auricle.

Currently, autogenous cartilage grafts are considered the gold standard for surgical repair of microtia. There are some inherent advantages and disadvantages regardless of the technique undertaken. Autogeneous cartilage frameworks are favored because they are native tissue and are more resistant to infection. In addition, they possess the potential to demonstrate a 5% growth with time. The cartilage, however, may warp or resorb with time and collecting them from the rib cage subjects patients to complications such as pneumothorax, atelectasis, or chest wall deformity or scarring. In addition, there is a risk for extrusion of the grafts with necrosis of the overlying skin flaps. One subtle downside to this technique is most noticeable in individuals with a low hairline. A low hairline may affect the final cosmetic result if it is involved in the skin flaps overlying the cartilage framework. Also, skin possessing hair follicles tend to be thicker and demonstrate less contouring typically needed to cover the frame, and it can be predisposed to inflammation and infection beginning with just folliculitis.

## **BRENT TECHNIQUE**

While Tanzer is considered to be the “father of modern auricular reconstruction,” Brent is viewed as the leading authority in the use of autogenous cartilage graft frameworks. He adapted Tanzer’s six-stage approach into a four-stage one: (1) developing the auricular framework, (2) transposition of the lobule, (3) elevating the framework, and (4) forming the tragus. As alluded

to earlier, a child undergoes surgery when he or she reaches six years of age, and any otologic surgery follows the reconstructive effort.

The first stage begins with designing a template for the auricular framework. This is done by tracing the patient's unaffected ear on a piece of x-ray film. Tracing a parent's ear is generally acceptable in cases of bilateral microtia. Once drawn, the size of the template is decreased by a few millimeters to account for the thickness of the skin that will cover the cartilage graft. The costal cartilage is then harvested from ribs 6-8 contralateral to the side of the microtic ear to form the two major components of the framework. The based is composed of the synchondrosis from ribs six and seven while the helical rim is created by the "floating" piece of cartilage from the eighth rib, and both of these pieces are affixed to each other with clear nylon suture. This construct is then nestled in a subcutaneous pocket along the inferior-posterior aspect of the microtic vestige with two suction drains placed nearby and left for five days. There is a strong preference to avoid using pressure or bolster dressings. Extra cartilage is banked either with the cartilage graft or within the chest incision.

Performed several months after the completion of the first stage, the lobule is transposed with an inferiorly based rotational flap in the second stage of the procedure. The lobule is mobilized as to receive the end of the framework while any extraneous unused tissue is excised. The framework is then elevated in the third stage. This is typically done with making an incision a few millimeters from the helical rim followed by dissection along the posterior aspect of the capsule until the desired amount of projection is achieved. The ear position is secured with the extra cartilage that was banked earlier in the first stage by placing it posteriorly beneath the frame in a fascial pocket. The retroauricular scalp is then advanced forward as much as possible while a split-thickness skin graft is utilized to cover the remaining postauricular space.

A tragus is formed in the final stage of the procedure. First, a composite skin and cartilage graft is obtained from the contralateral (i.e. normal) conchal vault. A J-shaped incision is then made along the posterior aspect of the tragal margin of the framework. The composite graft is then inserted through the incision and used to project the neotragus and cavitate the retrotragal hollow. The shadow of the neotragus imitates an external auditory canal. Subcutaneous tissue is excavated as needed to deepen the conchal bowl while the reconstructed ear is adjusted for frontal symmetry with its normal counterpart as necessary.

There have been a number of alterations and adjustments to this technique since its inception in 1971. One modification has entailed the creation of the tragus with forming the cartilage framework. The eighth rib creates the helix while a second strut is arched around to form the tragus, antitragus, and intertragic notch. The tip of this strut is then affixed to the helical crus of the main frame with a clear nylon horizontal mattress suture. More recent surgeries have utilized laser hair removal on the scalp flaps for cosmetic purposes.

Although the Brent technique has served as the basis for essentially all of the other modern-day reconstructive techniques, there has been some criticism. Opponents argue that the high number of stages carries an inherently higher operative morbidity and associated financial burden. Others have pointed out a less than ideal aesthetic result of the reconstructed tragus and the lack of definition to the conchal bowl, intertragic notch, and antitragal contour. There are some reports of aesthetic consequences from hyperpigmentation of the skin grafts to the conchal

bowl, while others have discussed the decreased projection of the reconstructed ear secondary to the effacement of the postauricular sulcus with skin graft contraction. Brent counters this latter critique by stating that this decreased projection can be minimized with the use of thicker skin grafts, preferably full-thickness ones, or by advancing the postauricular skin to the depth of the sulcus and grafting only the posterior aspect of the ear.

## **NAGATA TECHNIQUE**

Almost seemingly like a response to such criticisms, Nagata championed a two-stage technique in 1993. While there were technical refinements dependent on the type of microtia present such as lobular, small concha, conchal, and anotia, the first stage essentially grouped the first three stages of Brent's technique into one. The second stage involved wedging a piece of cartilage to elevate the auricular framework and serve as the posterior conchal wall. One striking distinguishing characteristic to this procedure, however, was that surgery did not begin until a child was ten years old and possessed a chest circumference of at least 60 centimeters around.

Significantly more costal cartilage is collected than with the Brent technique as cartilage from ribs 6-9 are harvested. In addition, one important difference is that the harvested ribs are on the ipsilateral side as the microtic ear. This construct possesses three "floors" that correspond to three different elevations of the frame: the base houses the cyma, cavum, and conchae; the crus helix, fossa triangularis, and scapha are located along the second level; and the top level contains the helix, antihelix, tragus, and antitragus. Once collected, the cartilage framework is assembled together with fine-gauge wire sutures where the base is composed of the sixth and seventh ribs, the eighth rib contributes to the helix and crus helix, and the superior and inferior crus and antihelix are derived from the ninth rib. The remaining structures are carved from the leftover residual cartilage.

The first stage with this technique also involves creating a W-shaped incision along the posterior lobule that extends toward the anterior lobule and where the tragal incision would be made. This type of incision increases the surface area available to cover the cartilage construct. The resulting flaps are undermined and reapproximated to form the cup of the intertragal notch, and the auricular framework is then inserted and positioned beneath the flaps. Lobular transposition is performed by reassembling the flaps in a Z-plasty fashion. A 2mm circular portion of skin is removed at the most anterior aspect of this incision and intentionally inverted to create the incisura intertragica. Finally, bolsters are secured in place with mattress sutures and left there for two weeks.

The second stage of the reconstruction process occurs six months after the first stage is completed. A crescent-shaped piece of costal cartilage is harvested from the fifth rib and wedged into position to elevate the framework and serve as the posterior conchal wall. Afterward, a temporoparietal fascial (TPF) flap is then raised through a new scalp incision and tunneled subcutaneously to cover the reconstructed auricle, the posterior aspect of the cartilage graft, and the mastoid surface. The retroauricular skin is advanced forward as much as possible, and an ultra-delicate split-thickness skin graft from the occipital scalp is placed over the posterior aspect of the cartilage framework.

Proponents for this technique emphasize that the conchal bowl appears deeper and more natural partly because there is no need to excavate any subcutaneous tissue as described with the Brent technique. However, critics cite high rates of vascular compromise leading to peri-lobular flap necrosis (up to 14%) and extrusion rates of the cartilage framework from the wire sutures (up to 8%). Others remark on the substantially larger amount of costal cartilage needed which can result in a significant anterior chest wall deformity and a thicker appearance to the reconstructed auricle. Nagata counters these assessments claiming that there is a lower incidence of necrosis and framework extrusion if a subcutaneous vascular pedicle along the posterior flap is preserved. In addition, the wire loops exhibit less extrusion if they are embedded into the substance of the anterior surface of the frame. Nagata also emphasizes the importance in preserving the perichondrium when obtaining the cartilage during the first stage, alleging that there is less chest wall deformity with eventual cartilage regrowth so long as the perichondrium is intact.

## **SINGLE-STAGE RECONSTRUCTION**

The indication for single-stage reconstruction efforts has traditionally revolved around partial auricular defects such as with the superior helix or lobule. A prospective single-stage microtia repair would be favorable with its relatively lower operative risk and cost, but would still necessitate the use of skin and fascial flaps in addition to external stents. Unfortunately, the results from the procedures that have been performed generally have not been shown to be comparable to that obtained with the various multi-staged procedures. For example, Park's two-flap single-stage procedure was eventually converted to a three-stage one with a marked improvement in the aesthetic results (1997, 2000).

## **TISSUE EXPANDERS**

Some have explored the role of tissue expanders in microtia repair. Supporters of this modality have suggested that the auricle could be created in a single stage without using a skin graft. Furthermore, the expanded skin possesses a good texture that matches the type of skin that is normally found on the auricle. In addition, skin innervation to this area is reportedly preserved.

The use of tissue expanders essentially entails two major phases. The skin and fascial layers are expanded in the first stage. A cartilage framework is sandwiched between both layers around this same time frame. It may still be necessary to place a skin graft over the fascial layer. Cartilage is removed and structures are shaped from the framework in the second phase. Despite the provision of extra tissue from use of the expander, skin grafts may still be required to cover the posterior surface of the frame.

Criticism of this modality includes pain with the expansion process itself, and consequently, this makes it less tolerable in younger children. Another important complication to consider is that a fibrous capsule may form around the expander which can marginalize the contouring results and diminished the aesthetic quality. Others argue that the insertion of the expander is a procedure in itself, and hence, there is not a true reduction in the overall number of surgeries undertaken.

## **OSSEO-INTEGRATED PROSTHESIS**

A bone-anchored prosthesis is a viable alternative when constructing autogenous cartilage grafts may not be a feasible option. Good candidates for this consideration include individuals who demonstrate severe soft-tissue or skeletal hypoplasia or have poor native tissue secondary to neoplasm or prior history of radiation exposure. Individuals who have had a failed autogenous reconstruction previously and those with high operative risk factors are other acceptable candidates. The most important aspect in the preoperative evaluation is to ensure that the patient has at least 3mm of bone by which to secure the implant.

Osseo-integrated prostheses are notable for providing the best aesthetic outcome although this is highly dependent on whoever actually designs and makes the prosthesis itself. This option is also favored in that it requires only a single, less-involved procedure as opposed to multiple stages inherent with other surgical alternatives. Typically, there may be some mild inflammation around the anchoring pins or even tissue overgrowth, but there are few complications related to infection. One distinguishing characteristic is that the prosthesis does not resorb because it is positioned outside of the body.

On the other hand, these prostheses are notable for degrading with ultraviolet light exposure and will eventually necessitate replacement every 2-5 years. In addition, multiple prostheses are needed to account for seasonal skin tones. Consequently, one drawback to this reconstructive modality is the substantial financial burden it imposes as each prosthesis may cost between \$2,000-7,000. Another important risk is the “Mr. Potato Head” social stigma that can accompany a child in case the ear unintentionally falls off or if it is forcibly removed by others. This would have a devastating negative effect from the potential ridicule that he or she may be subjected to should those situations occur. Finally, placement of an osseo-integrated prosthesis precludes any prospective autogenous reconstructive efforts.

## **TISSUE ENGINEERING**

The concept of tissue engineering engenders much interest due to its potential to offer autogenous cartilage without the morbidity associated from the rib harvest. In conjunction with actually growing it, the cartilage needs to be assimilated onto a prefabricated framework. The challenge comes in that taut pressure from the overlying skin can deform the frame. Firmer alloplastic frames may be able to withstand the pressure, but they are associated with a risk of extrusion. In addition to replicating numerous human chondrocytes to create an adequate amount of cartilage, it is difficult to match it to the size and shape of a normal human ear.

Cao *et al* sparked interest in tissue engineering after successfully transplanting bovine chondrocytes grown *in vitro* onto a synthetic ear-shaped biodegradable scaffold and implanting this scaffold into an immunocompetent mouse (1997). New cartilage formation in a human ear shape was noted after twelve weeks. Kamil (2004) expanded on this by demonstrating the creation of a human-sized auricle using a hydrogel scaffold in pigs. Kamil was also able to grow chondrocytes on a perforated ear mold made of pure gold and has reported the harvest of human chondrocytes from microtic ears in a mouse model.

Despite such promising results, the durability and compatibility of this neocartilage has yet to be defined. Consequently, this treatment modality is currently not a practical option in microtia repair although future research and developments may improve its feasibility.

## **ALLOPLASTIC IMPLANTS**

In line with the motivation to avoid extracting costal cartilage, alloplastic materials have been investigated as a reconstructive option. Silicone implants were the first to be studied in the 1960's and 1970's with good results initially. Such promise was replaced with disappointing long-term outcomes notable for a high incidence of implant extrusion or resorption due to skin flap erosion or necrosis. Implant failure was also observed after just minor trauma or abrasions. Consequently, the use of silicone alloplastic implants has since been abandoned.

A prefabricated porous polyethylene auricular implant, known to many as the Medpor implant, was introduced in the late 1990's and provided much improved biocompatibility, stability, tissue integration, and resistance to infection. The Medpor implant may be inserted alone or concurrently with a bone-anchored hearing aid. Although the Medpor implant appears to exhibit inherently less extrusion rates compared to its silicone counterpart, the incidence of extrusion decreases significantly if the framework is covered with a water-tight TPF flap. Soft tissue flaps may be employed to treat early implant exposure from flap ischemia if caught early for exposures less than 1cm assuming that tissue integration has already initiated. Furthermore, supporters of this treatment modality allege that the cosmetic results are superior to that obtained with autogenous cartilage grafts. Similar to silicone, Medpor implants still carry a risk for infection with even a small laceration or abrasion, and there is some degree of sensitivity to direct contact or minor trauma. As a whole, they have demonstrated good short-term (two-year) results, but long-term results are still pending.

Proponents cite five major advantages: (1) good projection and definition to the reconstructed auricle, (2) decreased reconstruction time, (3) no chest wall scarring or deformity since harvesting costal cartilage is not necessary with this technique, (4) can perform in children as young as 3-4 years old, and (5) the learning curve to this procedure is felt to be shorter than with performing an autogenous cartilage graft.

While different surgeons may alter certain details, microtia repair with the Medpor implant typically involves two major stages. The first stage begins with identifying the superficial temporal arteries utilizing a Doppler signal. Once these vessels are located, scalp incisions are marked to avoid junctions over the arteries but still remain perpendicular to the direction of hair growth. The scalp flaps are then lifted to provide access to a long and wide TPF flap. Similar to the Nagata technique, a small subcutaneous pedicle is preserved to provide a blood supply to what will eventually become the conchal bowl of the reconstructed ear. The lobule is mobilized, typically from a vestige of the microtic ear. Attention is then directed toward the contralateral unaffected ear and the postauricular skin is harvested. An abdominal skin graft is then placed over the postauricular harvest site, and an antibiotic-covered bolster is used to secure the skin graft in place.

The Medpor implant is constructed of two components similar in appearance to the autogenous cartilage base and helical rim as made in the Brent technique. After soaking in

betadine, the pieces are attached to each other with either clear non-absorbable suture or affixed with electrocautery. Of note, the manufacturers of the Medpor implant only endorse the use of suture to fasten the components together although anecdotally other physicians have had similar success with cautery.

Once constructed, the Medpor implant is fitted into position and the TPF flap is raised and draped over the implant. A drain is situated underneath the implant while another is placed behind it and put to suction to ensure a watertight enclosure. If positioned properly, the implant should fit inside a subcutaneous pocket encompassing its lower two-thirds. The remaining superior portion is covered with another abdominal skin graft posteriorly and the anterior portion is covered by the previously harvested postauricular skin from the contralateral ear.

The second major stage takes place a few months after completing the first one and is generally less complex in nature. Excess skin along the reconstructed auricle is excised, and an anteriorly-based flap is made near the location of the tragus. Part of the parotid gland is removed to allow room for the conchal bowl, and finally, the anterior flap is sutured over itself to make a tragus.

## **CONCLUSION**

Whether occurring as a single entity or in conjunction with other abnormalities, microtia is a condition that can significantly impact a child's self-image. Surgical repair should be delayed until the child is older regardless of the reconstruction option chosen, but close coordination with other services, such as audiology and otology, is absolutely warranted to ensure proper speech development and preserved hearing. While reconstruction with autogenous cartilage grafts is currently considered the gold standard, repair with the Medpor alloplastic implant may soon become a preferred treatment modality for the improved cosmetic results it offers with less staged procedures.

## **DISCUSSION by HAROLD S. PINE, MD, FACULTY**

**Having a child with microtia, especially if it's of the more severe type can cause a fair amount of distress for parents. The otolaryngologist is often consulted early on, sometimes while children are in the newborn nursery. These kinds of consults are often "dismissed" by ENT residents as there is "nothing to do" until the child is older. I would strongly argue that there is plenty to do and early involvement from ENT can help alleviate stress in families and also ensure appropriate work up and follow up.**

**While microtia may be an isolated finding in up to 65% of cases, a thorough exam may point to other subtle anomalies or suggest an underlying syndrome, ie Treacher Collins, Goldenhar. Chromosomal abnormalities occur in roughly 15% of the cases and so involvement from a genetics team can prove helpful.**

**Over 80% of the microtia cases are going to be unilateral and of those there is a slight preponderance for the right side. Where the ENT surgeon can be helpful here is trying to document whether there is also canal stenosis or atresia as well. In cases with combined microtia and atresia it is a certainty that there is going to be a component of at least conductive hearing loss. I support quick follow up within 1 month after birth to an audiologist.**

Early follow up is necessary for all of these children and most will end up getting ABR testing done to confirm the hearing loss and to initiate amplification. While speech and language development will usually be normal when there is one normal hearing ear, our audiologists are anxious to see even the unilateral cases and they do suggest that there is benefit from early fitting of bone conducting hearing aids.

When one ear is normal, it is vital to keep this ear healthy. For young patients who go on to have recurrent otitis media or present with evidence of OME, I tend to have a low threshold for placing a PE tube in the “normal” ear. I would advise for at least yearly ENT visits until which time the formal reconstructive process has begun.

If the audiological testing supports a normal inner ear, I generally will not recommend early radiographic evaluation. I think it makes more sense to wait until just prior to reconstructive efforts especially if there are plans for middle ear and canal reconstruction along with the microtia repair. There are a host of reconstructive options as reviewed by Dr. Pham. It pays to make some calls in your own area to find out who has the interest and expertise in providing this service.

Finally, as the ear expert who is called to the nursery to consult with families who have children with microtia, it is helpful to have resources available. There are a host of web-based support groups like the Yahoo health group for AtresiaMicrotia and Baby Center Community. There are even sites designed by parents of children with microtia to help the community at large. [www.microtiasupport.com](http://www.microtiasupport.com)

## DISCUSSION by TOMOKO MAKISHIMA, MD

Just a clarification on unilateral hearing loss, as I think I may have confused some people today:

1. The bone conductance attenuation from one ear to another is 5-10dB.
2. The attenuation from air conductance in one ear to bone conduction to the other ear is 45-50dB.
3. The maximum conductive hearing loss you can have is 50-60dB.

These are known observations in humans.

Based on these facts, here are some examples:

**Case 1:** a child with unilateral conductive hearing loss from left aural atresia. Right ear bone threshold 0dB, air threshold 0dB, left ear air threshold 50dB, bone threshold 0dB. This child will be able to hear out of right ear normally, and when the sound is greater than 50dB, the child will have input into left ear which is attenuation from right ear and from direct hearing from left ear. There is usually no developmental issues regarding speech in children as long as one ear has normal hearing. As a reference 30dB is when you can hear conversation in the next room or when you speak to them from behind, and 50dB is roughly the level of being able to use a regular phone to communicate, which is not all that bad. So, for this child, if the better ear has worse hearing than 30dB, it will most likely cause problems with speech development, and will need some intervention to bring the hearing level in the good ear up to about 0dB. If the child has very good hearing in one ear, the child doesn't necessarily need hearing aids. The purpose of using bone anchored hearing aids (not BAHA), is to increase the chance of stimulating the child as well as developing sense of direction.

**Case 2:** an adult with sensorineural hearing loss in the left ear, with bone conduction 70dB and air conduction 70dB, and normal hearing in the right ear with bone conduction 0dB and air conduction 0dB. This person has sensorineural hearing loss of 70 dB air bone gap in the left ear, and normal hearing in the right ear. First of all, if there is a sensorineural hearing loss which has more than a 50dB gap between the right and left ear, it is almost impossible to obtain a correct

**audiogram. In unilateral hearing loss, you will need to mask the better ear with white noise. However, when the white noise needs to be larger than 50-60dB, it will start to interfere with the bone threshold in the other ear. Therefore, it is very tricky to get an accurate audiogram from such patients. Many times on an audiogram, this will present as a person with 70dB "conductive hearing loss" and not a sensorineural hearing loss, in the left ear, with normal hearing in the right ear. When you see someone with a sensorineural hearing loss in one ear with more than a 50dB difference bilaterally, or a conductive hearing loss with more than a 50dB air bone gap, be suspicious about the validity of the audiogram itself. Audiograms can trick you in many ways!**

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