



Microtia Repair

TERI JUNGE, CST, CSFA, MED, FAST

The term microtia actually means small ear and the term anotia means lack of the ear. Microtia has become a “catch-all” term that includes any type of deformity of the external ear. Microtia occurs in approximately 1:6,000 births, is more common in males (approximately 63% of the patients are male and 37% are female), and is more likely to occur on the right side (approximately 58% of deformities occur on the right side) however the defect may be bilateral (approximately 9% of patients are affected bilaterally). Microtia can occur as an isolated deformity or can occur in conjunction with other birth defects (related or not) which can include middle and inner ear problems resulting in reduced or absence of hearing on the affected side.

POSSIBLE CAUSES

Microtia is thought to be a random event; although it has been linked to increased maternal age. Other possible causes are intrauterine tissue ischemia (possibly from an artery that was compressed in the womb), maternal medication use (such as the drug thalidomide/Thalomid® which is an immunomodulatory agent used to treat multiple myeloma and erythema nodosum leprosum and/or the drug isotretinoin/Accutane® which is a retinoid that is used to treat acne), evidence of a first trimester of pregnancy rubella (German measles) episode, and genetic factors. Microtia has been associated with several syndromes such as Goldenhar Syndrome and Treacher-Collins Syndrome. A syndrome is a cluster of deformities that often occur together.

Goldenhar Syndrome involves deformities of the face and usually affects one side only. Characteristics of Goldenhar Syndrome include varying degrees of microtia or anotia, the chin on the affected side is closer to the ear, the corner of the mouth on the affected side may be

LEARNING OBJECTIVES

- ▲ Examine the possible causes of microtia
- ▲ Identify the grades of severity related to this condition
- ▲ Review the anatomy of the external ear, middle ear and inner ear
- ▲ Evaluate the steps taken during an autologous rib graft procedure
- ▲ Compare and contrast the conservative approach with the surgical operation

Word Element	Definition
1. acous/o	hearing
2. audi/o	hearing
3. labyrinth/o	portion of inner ear
4. myring/o	eardrum
5. ot/o	ear
6. tympan/o	eardrum

higher than the unaffected side, the eye on the affected side may be missing, deformed, or contain benign growths (such as dermoid cysts). Goldenhar Syndrome is a form of oculoauricular dysplasia.

Treacher-Collins Syndrome is an inherited craniofacial defect that is caused by a defective protein called treacle (a protein involved in transcription of ribosomal DNA) and the significance ranges from mild to severe. Characteristics of Treacher-Collins Syndrome include varying degrees of microtia or anotia, micrognathia (small jaw), a large mouth, coloboma (a full-thickness defect of the eyelid in which there is a hole or a gap), scalp hair growth that extends to the cheeks, and chiloschisis/palatoschisis (cleft lip/cleft palate).

SEVERITY

Microtia is described as typically occurring in 4 grades. Atypical anomalies that do not fit into the grading scale can also occur.

Grade 1 – The pinna is smaller than normal and may have a slightly noticeable malformation. The normal characteristics of the external ear are present and well defined. The acronym FLK which represents the term “funny looking kid” may be seen on the chart of a newborn or child when a dysmorphic feature, such as a slightly malformed ear, is suspected, but has not yet been fully researched and identified.

Grade 2 – The pinna is less developed than in grade 1 and the structures such as the helix and antihelix show less definition.

Grade 3 – The pinna is basically absent however a vertical skin remnant often remains. The remnant consists of cartilage that is not organized and the patient usually has a well-formed ear lobe.

Grade 4 – Complete absence of the auricle (anotia).

ANATOMY REVIEW

There are three structural divisions of the ear.

External Ear

The visible portion of the external ear is also called the pinna and the auricle. The external ear continues through the meatus to the tympanic membrane. The important structures of the external ear that are involved when performing a reconstructive procedure are the:

- Helix
- Triangular fossa
- Antihelix
- Concha
- Scaphae
- Tragus
- Lobule
- External auditory meatus
- External auditory canal which contains the ceruminous glands

The function of the external ear is to collect and funnel sound waves into the ear canal.

Middle Ear

The middle ear begins at the tympanic membrane and ends at the oval window. The three ossicles; the malleus, incus, and stapes are enclosed within the middle ear, as is the opening to the eustachian tube. Additionally, access to the tympanic antrum, which in turn communicates with the mastoid air cells, is provided through the attic (also known as the epitympanic recess). Several physiologic activities occur within the middle ear.

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The ossicles are attached to one another via small ligaments and conduct vibrations from the tympanic membrane to the oval window. Several tiny muscles control/constrict the movements of the ossicles and help to obtund certain noises (such as from chewing crunch food). The two main muscles of the middle ear are the tensor tympani and the stapedius.

The eustachian tube, also called the pharyngotympanic tube, connects the middle ear cavity with the nasopharynx. The eustachian tube functions to drain mucus from the middle ear and provide aeration of the cavity. The eustachian tube opens to equalize atmospheric pressure in the middle ear and closes to protect the middle ear from loud noises and retrograde infection that may result from contaminants ascending from the nasopharynx.

The air spaces of the tympanic antrum function to provide acoustic resonance, dissipate sound, and reduce the weight of the cranium.

Inner Ear

Vibrational movement of the stapes against the oval window, also known as the fenestra ovalis, sets in motion the fluids within the labyrinth. The labyrinth consists of the vestibule, the semicircular canals and the cochlea. The vestibule contains receptors for equilibrium and movement of the fluid within the two bony chambers of the vestibule affects balance. Fluid movement within the three semicircular canals, influences proprioception (sense of position) and balance during movement (dynamic equilibrium) and when still (static equilibrium). Fluid within the bony labyrinth is called perilymph; inside of the bony labyrinth is the membranous labyrinth that contains a fluid called endolymph. The vestibulocochlear nerve (cranial nerve VIII) is responsible for transmission of sound and balance information from the inner ear to the temporal lobe of the brain for interpretation. The vestibular branch of the VIIIth cranial nerve serves the vestibule and the labyrinth and the cochlear branch serves the cochlea.

The organ of Corti, also known as the organ of hearing, is located within the cochlea. The organ of Corti is comprised of ciliated receptor cells that move against the tectorial membrane as the fluid in the middle ear moves. Movement of the tectorial membrane initiates the impulses that move along the cochlear branch of the VIIIth cranial nerve for interpretation in the temporal lobe of the brain. The process of hearing is outlined in Table 1.

Table 1 Process of Hearing	
Step 1	Sound waves enter the external ear
Step 2	Tympanic membrane vibrates
Step 3	Vibrations are transferred through the middle ear by the ossicles
Step 4	Stapes moves the membrane covering the oval window setting in motion the fluids within the bony labyrinth
Step 5	Movement of the fluid within the cochlea sets in motion the hair cells of the organ of Corti
Step 6	Movement of the hair cells against the tectorial membrane initiates a nerve impulse
Step 7	Nerve impulse is transmitted via the VIIIth cranial nerve
Step 8	Impulse is interpreted in the temporal lobe of the brain

CONSERVATIVE/NONSURGICAL APPROACH

A prosthetic device can be constructed and applied to the side of the head at the location of the ear. If an earlobe is present, the prosthesis can be constructed around it. The main reason for using a prosthesis is to avoid undergoing a surgical procedure. Negative qualities of prosthetic ears are that it must be removed and cleaned daily, it may become detached unexpectedly (especially during sporting activities), it will not change coloration according to the amount of sun exposure, will not grow with a child, and will show signs of wear that will require replacement.

HISTORY OF SURGICAL INTERVENTIONS

The first reported microtia repairs occurred in the late 1500s and involved the use of cheek or scalp flaps. Homologous rib graft (particularly from the mother) procedures began to emerge in the late 1920s; however, there was a problem with tissue resorption and the technique has been abandoned. The autologous rib graft procedure, which is still in use today, originated in the 1940s. A major advantage of the

autologous rib graft procedure is that the graft will grow with the child. In an attempt to avoid the thoracic portion of the autologous rib graft procedure, a variety of inorganic implants have been tried. Unfortunately, migration and extrusion of the implants are problematic. Examples of implanted materials that have been tried include Vitallium, polyethylene and Teflon.

AUTOLOGOUS RIB GRAFT PROCEDURE OVERVIEW

Microtia repair is typically described in three to four stages. In preparation for stage one, a template is drawn from the opposite (intact) ear and then reversed to be used of the



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affected side. If the defect is bilateral, a sibling or other model may be used to draw the template. Measurements, or a second template, are used to determine the location of the new auricle. The lateral canthus, alar nasal cartilage, and the corner of the mouth are used as landmarks to estimate the position the graft.

Stage one of the reconstruction is performed under general anesthesia and involves securing the cartilaginous graft from the donor site on the contralateral chest wall. Great

care is exercised to remain outside of the pleura while securing the graft. The cartilage that will represent the main framework of the graft is taken from a section of the false ribs and the cartilage that will become the helical rim is taken from the floating rib cartilage and the chest wound is closed. The surgeon will remove the perichondrium from the cartilage using a periosteal elevator (such as a freer), a scalpel, and a chisel, and then sculpt the main framework of the graft to closely resemble the template. The helical rim is fashioned in much the same way as the main framework and the rim is attached to the main framework with non-absorbable suture (such as silk) or stainless steel wire.

Once the cartilaginous framework (graft) is complete, it is set aside while the incision is made and the pocket is developed at the recipient site. The graft is inserted, the wound is closed, and a wound vacuum/drainage device is employed. A bulky wound dressing is applied and left in place for five to seven days. Additionally, a temporary protective device may be applied to keep very young children from disrupting the wound and to protect the wound while the patient is sleeping. The patient is restricted from using a hair dryer, swimming, and participating in contact sporting activities for four to six weeks.

Stage two of the reconstruction involves transposition of the earlobe (if one is present) and can be performed in conjunction with stage one. Unfortunately, the risk of vascular compromise increases and the esthetic outcome may not be as precise, if performed at the same time as the first stage. Ideally, the earlobe transposition is performed when healing from the first stage is complete. According to the age of the patient, the transposition may be performed under local anesthesia. Any existing lobular tissue is moved into position as a flap to maintain the integrity of

the vascular structures, any unusable tissue is removed, the wound is sutured, and a light dressing is applied.

Stage three of the reconstruction allows for elevation (sometimes referred to as lifting) of the posterior aspect of the newly created framework of the ear. Because skin grafting is often necessary, stage three is performed under general anesthesia. The donor site for the skin graft may be the lower abdomen, the buttocks, or another location determined to be appropriate by the surgeon.

Any of the stages of the reconstruction may be combined, as appropriate. If a fourth stage is needed, it may include laser treatments to remove any scalp hair remaining on the newly created ear and minor additional procedures such as creation of a tragus and/or a false conical shaped entry to simulate the auditory meatus and external ear canal.



ABOUT THE AUTHOR

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