

New Ears Transforming Congenital Deformities

BY DR. JOSEPH ROBERSON, M.D.

The human ear is a complex body organ that begins development early in a mother's pregnancy. Those who have been pregnant know the ear works by the last third of pregnancy and that babies respond to sounds in their environment by startling, by kicking or by stopping movement.

For the ear to form correctly, structures grow from the skull base and must join with growth from the outside of the head which progresses in toward the inner ear. The two processes meet at the eardrum and middle ear bones. A part of the process involves formation of the outer ear (called the pinna) as several small folds of skin merge to give our ears their normal appearance.

These processes can become disordered to produce abnormalities at birth – a congenital deformity. The deformity may be mild,

such as a middle ear bone that has not become entirely separated from the surrounding bone, causing a restriction in its ability to vibrate and thereby to transmit sound energy to the inner ear. More severe abnormalities may result in a normal inner ear but no ear canal (called Atresia) and no outer ear (called Microtia). Other problems with formation of the ear may cause the structure of the ear to be normal but without the correct receptive nerves in the inner ear, which produces deafness.

Audiologists can perform specialized tests to determine if the cause of a hearing loss is due to a lack of normal nerve function (sensorineural hearing loss) or if the problem is due to a lack of function in the ear canal, eardrum or middle ear bones (conductive hearing loss). Hearing loss that is sensorineural may be treated with hearing aids or cochlear implants and responds very well to

There's No Disguising It Prosthetic Ear is a Big Success

BY DEVORAH FOX

ALL PHOTOS COURTESY OF CANDACE SHOCK

Madison Shock was born with microtia, a congenital malformation of the external and middle ear. Her right ear is fine, though her hearing is limited. On the left side, Madison's middle ear is underdeveloped, so she is deaf on the left side.

Madison's parents, Brian and Candace Shock, considered a surgical solution. A piece of her rib bone and skin grafts from her legs would be used to shape a new ear. The Shocks decided against the extensive surgery.

Candace then learned about Robert Barron who designs custom prosthetics. A former CIA disguise specialist, Barron retired from the CIA in 1993. Barron now owns Custom Prosthetic Designs in Ashburn, Va. Madison was too young initially to work with Robert Barron, who wouldn't see her until she was 5 years old.



these treatments when started in the first few weeks or months of life. Conductive hearing loss can be repaired, allowing reestablishment of sound transmission to the inner ear (the inner ear and nerve function in conductive hearing loss is usually normal). These situations are rare and may be very challenging surgically.

One such congenital deformity in which hearing loss can be restored is Atresia – Microtia. In some children, the anatomy of the deformity allows creation of an ear canal and restoration of hearing. Experienced surgeons can “grade” a type of x-ray called a CT scan and predict the chance of success for surgery. In similar fashion, the outer ear can be reconstructed to resemble a normal outer ear around the reconstructed ear canal.

Let me describe how that happens surgically. The ear canal is fashioned with a diamond drill that allows the removal of bone and the sculpting of a canal of normal size and contour. This canal must line up exactly with the middle ear bones. Once this channel to the outside world is opened, it must be lined with skin, taken as a skin graft and transplanted from another area of the body. More importantly, an eardrum must be created to connect to functioning middle ear bones. Very specialized equipment and techniques, like lasers, are needed to free the ear bones from their fixation point with the bone of the middle ear. The eardrum receives the sound vibrations that proceed down the skin-lined ear canal, transmitting their energy to the vibrating middle ear bones and to the inner ear, where the normal hearing nerves are waiting to receive them. From there, the hearing nerve takes the signal to the brain. In this way, a deaf ear can be turned into a hearing ear!

For the outer ear reconstruction, three options exist. The first is to build a prosthetic ear that either glues on in the proper position or is affixed to implanted magnets in the skull that match up with

similar magnets in the prosthesis. Both options are removable. Two other options include creation of an outer ear covered with skin that stays in place for the rest of a person’s life. In most patients, the technique uses the patient’s own rib cartilage. After harvesting a block of cartilage from the chest, an ear is carved and inserted under the skin. Several surgeries are required to carve, place, lift and position the ear to make it as good as it can be. A newer technique uses a manmade material called Medpor® that is also customized to be the right size and shape. The prosthetic ear is inserted under a skin graft and a tissue layer brought down from the scalp. One or two surgeries are normally required for this technique to achieve the best form and function.

Ear canal surgery and outer ear surgery have been performed separately for many years. Recently, we have been performing both outer ear and ear canal surgery in one day. While it takes eight hours or longer to do the work, many patients and parents prefer to get it all done at one time.

Of particular note to parents and patients are laws in more than 30 states that require insurance coverage for congenital defects. While not everyone with a congenital defect is a candidate for surgical reconstruction, it’s nice to know it may be a covered expense in many situations. Further help achieving insurance coverage can be gained free of charge through a foundation my lovely wife of 25 years, Julia, and I started in 2002 called the Let Them Hear Foundation (www.letthemhear.org).

In other patients with severe middle ear, ear canal or outer ear malformations surgery is not an option because of the severity of the malformation. Fortunately, other options allow hearing to be restored. One strategy is to pick up the tiny sound vibrations we hear, amplify them and apply them to the bone of the skull.

Robert Barron uses liquid silicone to fashion Madison’s new ear.



The ear goes through several shaping, refining and coloring processes.



Robert Barron attaches Madison’s new ear with adhesive. The ear isn’t permanently affixed to Madison’s head. The adhesive can stand up to swimming and play. When Madison isn’t wearing the ear, she keeps it in a small plastic container. With regular wear and tear, the ear will last about three to five years. Then Madison will need a new one.

Amazingly, the inner ear will pick up these vibrations and allow them to be heard. The vibrational energy may be applied to the skull through a hearing aid that presses on the skin and vibrates the bone of the skull behind the ear, or it may be attached to a surgically implanted titanium post which comes through the skin and allows the specialized device to be attached. Another new technology picks up the vibration with an external microphone, transmits it through the skin to a surgically implanted device that releases energy very close to the inner ear via a specialized vibrating transducer supplying sound to the inner ear. To learn more about these options, read "Hear in Stereo Again" in the Winter 2009 issue of Hearing Health (available in the archives at www.drff.org).

Hearing in one ear is good but inferior to hearing with two. For example, children with congenital deformities causing hearing impairment in one ear are over 10 times more likely to repeat a grade and end up with salaries nearly 25 percent lower than their peers with two ears! It's wonderful that options exist for those with congenital deformities. ■

Joseph Roberson, M.D., is CEO of California Ear Institute and president and founder of Let Them Hear Foundation.

Further information about how to treat a congenital deformity may be gained at www.atresiarepair.com and www.letthemhear.org. Let Them Hear also offers several conferences around the world to interested patients and parents. Free conferences in 2009 will be in Korea, India, Mexico and Texas.

Support Groups for People and Parents of Children with Ear Deformities

Yahoo® Online Support Groups

<http://health.groups.yahoo.com/group/AtresiaMicrotia/>

For parents who have or are expecting to adopt a child with Atresia-Microtia

<http://groups.yahoo.com/group/Atresia-Microtia-Adoption/>

Children's Craniofacial Association, 800.535.3643, www.ccakids.org

Offers physician listings, educational resources, a newsletter, annual family retreat and family networking opportunities. ■

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Log on to www.hearinghealthmag.com



Candace, Brian and Madison Shock celebrate the completion of the prosthetic ear.



Madison shows off her new ear, and her appreciation.