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# an Ear

*When an ear fails to develop in an infant it is known as aural atresia. Doctors can build an ear using the patient's own tissue and special surgical techniques.*

One baby in every 20,000 is born with aural atresia, a condition that is characterized by an abnormally small or absent bony ear canal. This is frequently accompanied by aicrotia, or a vestigial outer ear (pinna), and by abnormalities of the middle ear. Though the cause is not known, atresia is a congenital condition that occurs by the sixth week of fetal development, and can affect one or both ears. Parents of a child born with atresia should seek a hearing evaluation, and a hearing aid evaluation, during the baby's first few weeks of life.

Typically, two of the three parts that comprise the middle ear are also affected by aural atresia. The malleus (hammer) is deformed and is fused to the incus (anvil), although the stapes (stirrup) is usually normal. The resulting lack of sound conduction through the middle ear causes a 35%-60% hearing loss. Though the outer ear, middle ear, and ear canal are affected and require surgical correction, the delicate inner ear structure usually exists intact. In most cases of aural atresia significant improvement of hearing levels can be attained through surgery.



**Question:** What is aural atresia?

**Answer:** An abnormally small or absent ear canal, which is frequently accompanied by a vestigial outer ear and abnormalities of the middle ear that affect hearing.

Surgery is performed at age five or six to widen a narrow ear canal or create one where none exists. The bones of the middle ear are then inspected and mobilized. The surgeon next creates an eardrum using a graft from the patient's own fascia (muscle covering). Keeping the new ear canal open can present a challenge, since the body's natural healing process is to close openings that remain following surgery. Therefore, in an attempt to avoid closure, the new ear canal is lined with a skin graft that is carefully held in place with special packing for 6-8 weeks as it heals.

In less than 10 percent of cases, a second surgery is required to correct problems caused by scar tissue during the healing process. Scarring can sometimes re-close the ear canal, and dislodge the new eardrum, immobilizing or reducing contact with the bones of the middle ear that are necessary for sound conduction, and diminishing hearing quality that was initially improved. Scar tissue may also form around the bones of the middle ear, immobilizing them.

“Atresia occurs more frequently in males than females, and affects the right ear more often than the left.”

Reconstructive surgery of a malformed pinna (outer ear), does more than offer the patient a normal-looking ear. The “new” pinna contributes approximately six decibels (dB) of sound to the ear from the environment, augmenting the average 60 percent hearing improvement achieved through surgical construction of the ear canal and middle ear.

Some atresia patients are inoperable due to lack of development of the mastoid, absent oval window that normally contains the stapes, or an abnormally-placed facial nerve. These patients can receive bone-conducting hearing aids, such as the bone-anchored hearing appliance, or BAHA. Children in the U.S. may receive implantable hearing aids at age five or six, while in Europe, children as young as two or three years can receive implantable bone conduction hearing aids. ❖

## Professional Education at HEI

The physicians of the House Clinic volunteer their time to teach courses at HEI through its **Visiting Physicians Program**. Since its inception, the program has attracted thousands of doctors from 75 countries. It welcomes otologists, otolaryngologists, neurosurgeons, residents in training and hearing health specialists from all over the world to participate in special ear surgery training courses. Under the direction of Antonio De la Cruz, M.D., participants follow programs for periods ranging from three days to three months. They can attend a Temporal Bone Surgical Dissection Course, observe physician-patient consultations in the House Clinic, special procedures performed by Clinic surgeons at St. Vincent Medical Center, professional presentations and lectures. Visiting physicians may also utilize the Institute’s otology library and collection of surgical videotapes.



*Medical expertise and background information for this article were provided by Antonio De la Cruz, M.D., who practices neurotology and skull base surgery at the House Clinic in Los Angeles. Dr. De la Cruz is also director of education at the House Ear Institute.*