

Hearing Concerns

Infants born with cleft palates and other craniofacial anomalies are likely to have more hearing problems than other infants, and the condition of their middle ear is likely to be of concern for the rest of their lives. Since hearing loss is a major cause of language learning problems, it is important to be on the alert for any signs of hearing impairment in your child. Your child should undergo regular examinations to detect and prevent chronic ear disease and deafness.

The Ear

While hearing is a key factor in communication, possibly even more important is that it gives people a feeling of life participation and security. A sense of hearing is vital to well-being. Anytime there are problems involving the ear, a physician should be consulted.

The normal human ear can distinguish among some four hundred thousand different sounds, some

weak enough to cause the eardrum to move as little as one-tenth the diameter of a hydrogen molecule. When a telephone rings, it produces a series of disturbances in the surrounding air, and these disturbances, or sound waves, travel out and away from the source. Your hearing mechanism perceives these and transforms and transmits these sound waves as a message to your brain.

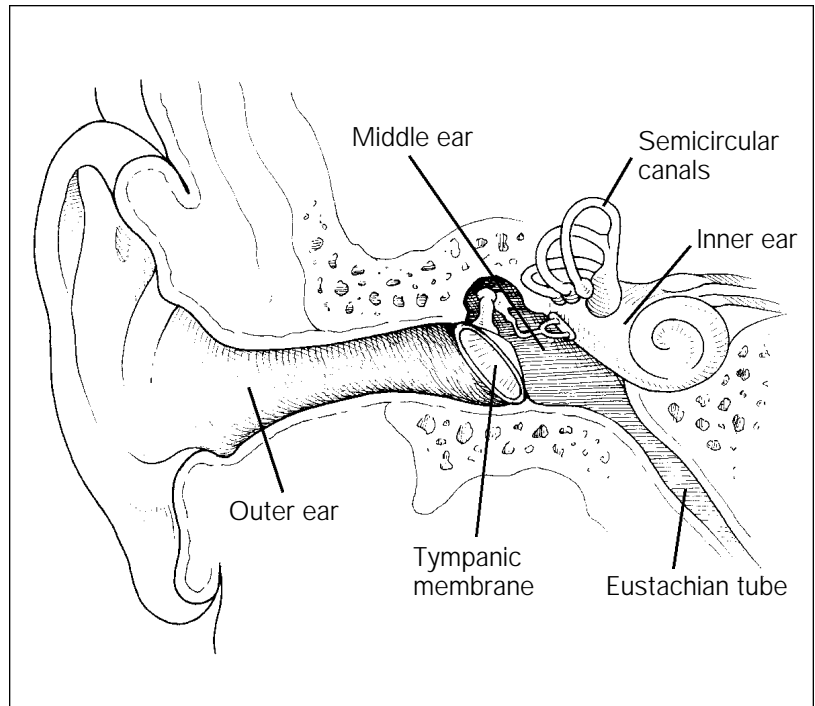
Before the message can get to your brain, however, it has to pass through three well-defined sections of the ear; the outer ear, the middle ear, and the inner ear. The outer ear includes the *pinna*, the part of the ear we can see, plus the ear canal. The pinna is designed to help gather sound waves and funnel them down the ear canal to the eardrum.

The sound waves then strike the eardrum, or *tympanic membrane*, which is about as thin as tissue paper but very strong. The eardrum separates the outer and middle ear. The eardrum vibrates when sound waves strike it. Attached to the eardrum is a chain of three small bones called the *ossicular chain*. The bones are in the pea-sized middle-ear cavity.

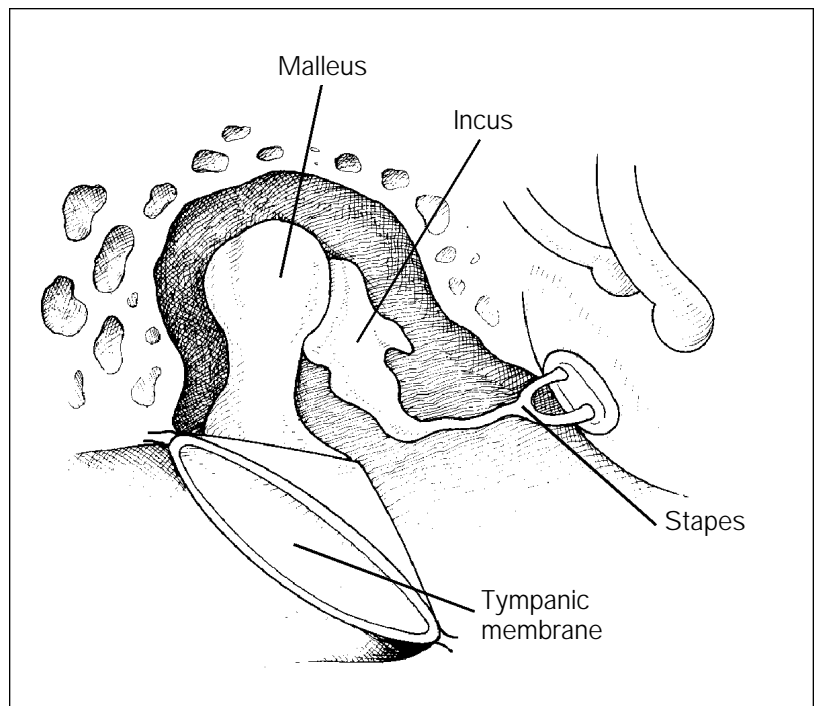
The ossicles, the smallest bones in the human body, are full-size when we are born. These individual bones are smaller than grains of rice, and they are named after objects they resemble. The bone attached to the eardrum is the *malleus* (hammer), the second bone is the *incus* (anvil), and the third is the *stapes* (stirrup). As sound waves move the eardrum, they move the ossicles. The three bones actually form a lever system that transfers the energy of the sound waves from the outer ear through the middle ear and into the inner ear.

The *eustachian tube* is the part of the middle ear that connects the middle ear cavity with the back of the throat. The upper end is normally open because it is surrounded by bone. The lower end is normally closed, or collapsed, because it is surrounded by soft tissue. The eustachian tube helps maintain a

The ear has three chambers: the outer, middle, and inner ear. The two connecting structures are the *semicircular canals* responsible for maintaining balance and the *eustachian tube* for the equalization of air pressure to the middle ear and drainage of fluids to the nose.



The middle ear contains the *ossicular chain*, which transmits vibrations from the tympanic membrane to the oval window. It is composed of the *malleus*, *incus*, and the *stapes*. Cleft palates can cause problems with the opening and closing mechanism of the eustachian tube. Malfunction of the eustachian tube can lead to otitis media, inflammation of the middle ear. (Adapted from "Physiology of the Ear," Starkey Laboratories, Inc.)



balance between the air pressure on both sides of the eardrum. The tube opens about every third time we swallow or yawn, allowing air pressure in the middle ear to equalize with the outside air pressure. The normal ear-popping sensation sometimes experienced in an airplane or with other sharp changes in altitude is caused by this equalization process.

The last bone in the ossicular chain, the stapes, is attached to a tiny membrane called the *oval window*. The oval window is an entrance to the inner ear that contains the organ of hearing, or cochlea. When the stapes bone moves, the oval window moves with it. The cochlea is a fluid-filled channel. The fluid is disturbed by movements of the oval window. Inside the cochlea are thousands of microscopic hair-like cells that are set in motion whenever the fluid is disturbed. Stimulation of these cells, in turn, causes electrical impulses to be sent to the brain.

Our inner ears also contain a very important organ that is actually connected to the cochlea but does not contribute to our sense of hearing. Instead, these three small loops, called semicircular canals, help us maintain balance. Problems within the *semicircular canals* may result in symptoms such as dizziness.

Hearing Problems

Individuals with craniofacial anomalies may have congenital abnormalities of the ear structures, and they are thus subject to ear disease. Such children are at high risk for hearing disorders varying from mild to severe. The 1990 Joint Committee on Infant Hearing in the United States recommended that “at risk” newborns have their hearing screened at the age of three months and no later than six months.

Infection of the middle ear is one of the most common problems for all children, and it can be one of the most serious complaints during childhood. An untreated or repeated ear infection can inhibit learning at critical stages of language development because it can reduce a child's ability to hear.

More children with cleft palates suffer decreases in hearing than do similar groups of children without cleft palate. Since it is very difficult to see the eardrum in small infants, the physician and parents may not be initially aware of the presence of ear disease, and changes in the tympanic membrane and bones of the middle ear may occur that might have been prevented by early treatment.

The part of the ear of most concern in the child with a cleft palate is the middle ear. The eustachian tube normally drains fluid that collects during a cold or respiratory infection. The muscles that help the eustachian tube function properly are connected to the soft palate. If there is a cleft palate, these muscles may not operate efficiently, thus interfering with the ability of the eustachian tube to drain fluid from the middle ear to the back of the nose.

A child's eustachian tube is shorter, narrower, and more horizontal than an adult's; for these reasons it is more likely to swell during a cold and become blocked. Poor drainage of the middle ear may lead to the accumulation of fluid (effusion). The effusion may become infected with bacteria or viruses and cause pressure and/or fever. The fluid hampers the movement of the eardrum and associated small bones and may lead to hearing loss. This loss is about the same level of decrease you would get by putting a finger in your ear canal. It is, however, unusual for a child with a cleft palate to experience total hearing loss.

Since these changes occur early in life, the child may not show obvious hearing loss problems but may simply learn to live with the disease. Moderate

hearing loss may not be noted until the child is older, when it becomes obvious that he or she does not hear properly. Occasionally, an infection will be severe enough to cause a hole in the eardrum, resulting in a draining ear. While it is difficult to evaluate hearing in infants, it is not impossible. A specialist in the testing of hearing (audiologist) may work with the ear specialist (otolaryngologist or otologist) to evaluate your child's hearing as he or she grows. In almost all children, this team can evaluate the hearing level.

Otitis media

In the later stages of infancy and throughout their childhood, there is a higher incidence of *otitis media*, inflammation of the middle ear, in children with clefts. This suggests a disturbance of some kind around the eustachian orifice. Whether this is caused by an anatomic or functional variation associated with the cleft, or by an increased susceptibility to irritation and infection, remains to be resolved.

Those who suffer from respiratory allergies, as well as infants who usually drink from a bottle while lying down, are at risk for otitis media. Because this disease of the middle ear can cause deafness, prevention and detection are essential.

The basic instrument for examining the ear for otitis media is the otoscope. Another screening technique is tympanometry, which measures the ability of the tympanic membrane to vibrate. Parents should be aware that school screening tests are not always sufficient to identify otitis media.

Management of middle ear disease, elimination of middle ear fluid, and aeration (which balances air pressure between the middle ear and the nasopharynx) of the middle ear are the responsibility of the otologist and can be accomplished by draining (by

myringotomy), aspiration (removing fluid by means of suction), or the insertion of tympanotomy or pressure equalization tubes. If the tubes fall out, a myringotomy and reinsertion of the tubes may be necessary. The pressure equalization tubes should be reinserted as many times as necessary until the eustachian tube is functioning adequately. Aeration of the middle ear will prevent serious hearing problems.

Eustachian tube function

The eustachian tube serves three physiologic functions for the middle ear:

1. Ventilation (balancing air pressure between the middle ear and the nasopharynx)
2. Protection (from secretions and pressure)
3. Clearance (draining middle-ear secretions into the nasopharynx)

The normal eustachian tube at rest is closed, with a slight negative pressure existing in the middle ear. There is no muscle tissue in the eustachian tube itself to open or close it. Intermittent contractions during swallowing of the tensor veli palatine muscle, which is attached to the soft palate, cause the eustachian tube to open, equalizing pressure in the middle ear. (This is why repeated swallowing can often relieve painful pressure disequilibrium in the ears during a sharp airplane descent; when the ears “pop,” it is a sign that more equalized pressure has been restored.) Yawning and sneezing also open the eustachian tube. Conversely, relaxation of the tensor veli palatine muscle causes the eustachian tube to close.

Obstruction of the eustachian tube may result in a negative pressure in the middle ear. If this persists, atelectasis, or collapse of the tympanic membrane, may occur and lead to the accumulation of fluid in

the resulting space. This negative pressure also creates a situation in which bacteria from the nose may be aspirated into the middle ear, resulting in acute bacterial otitis media with effusion.

When a persistent hearing loss is identified, hearing aids and auditory training systems should be considered. When hearing loss occurs in the presence of *atresia* (blockage), whether in one or both ears, bone conduction amplification may be considered. Depending upon degree of loss, an implantable bone conduction aid may be an option in treatment. These treatment programs require continual monitoring. If a child has a hearing loss, the appropriate school official should be notified. If no hearing loss is detected, the child should still have a yearly checkup.

Mechanical obstructions

Mechanical obstructions of the ear, which can adversely affect persons with or without cleft palates, are classified into two types: extrinsic and intrinsic. The extrinsic variety includes obstructions such as cotton swabs, and anatomic abnormalities such as nasal-pharyngeal tumors and enlarged adenoids. Intrinsic obstructions include blockage by secretions resulting from inflammation caused by infection or possibly allergy.

It must be stressed that in children with cleft palate the condition of the middle ear, even with corrective treatment, will remain a concern throughout their lifetime.

How to Prevent Ear Problems

Ear disease can exist with minimal symptoms in all infants. The ears should be examined regularly, and the wax that interferes with seeing the eardrum removed. Hearing tests should be scheduled as early as possible.

When the child with a cleft palate has a cold in the nose, throat, or chest, the ears should be examined to see whether there is an associated ear infection. If there is, it is usually treated with the appropriate antibiotic. When indicated, the eardrum may have to be lanced to allow fluid to escape from the middle ear. Opening the eardrum will not result in bad scars or permanent holes. It may be necessary for your child's surgeon to perform a *myringotomy*, a procedure in which a small slit is made in the eardrum to permit the drainage of trapped fluid. Ventilating tubes may or may not be placed.

If permanent changes in the ears caused by disease can be prevented during infancy, the need for corrective surgery later in life may be avoided. If disease-caused changes do occur, techniques are available to allow the surgeon to try to reconstruct the middle ear in hopes of restoring proper function.

As the child gets older, examining hearing function becomes easier. Periodic hearing tests are valuable throughout childhood and adolescence to determine whether treatment is necessary.

Hearing Tests

Pure-Tone Audiometer. This device provides information about the integrity of the peripheral auditory system, that is, the outer, middle, and inner ear.

Whatever sound-transmitting defect exists in the ear will be revealed on an audiogram as a loss in hearing sensitivity.

Bone-Conduction Audiometry. This procedure basically measures defects of the sensorineural (pertaining to the sensory nerve) mechanism of the ear. The bone-conducted signal bypasses the external and middle ear and goes directly to the inner ear through vibrations of the cranial bones.

The classic method of distinguishing middle ear defects from cochlear or sensorineural impairment is to find the difference between air-conduction and bone-conduction hearing levels (known as the air-bone gap).

Impedance Audiometry. This is an objective hearing test that does not require the child's active participation. This test includes tympanometry, static compliance or acoustic impedance, and acoustic reflex threshold measurements. An instrument is used to detect the possible presence of fluid in the middle ear. The entire battery of three tests can be administered by an experienced person in sixty to ninety seconds per ear. The child can be sedated if necessary, and the tests can be used successfully with persons of all ages, including newborn children.

Tympanometry cannot detect sensorineural hearing impairment and thus cannot be substituted for pure-tone audiometry as a screening technique. However, tympanometry is more sensitive and reliable than air-conduction audiometry in identifying a pathologic condition of the middle ear, and probably is equal or superior in reliability to the otoscopic examination. It is recommended that tympanometry, in combination with air-conduction audiometry, offers the best method for detecting middle ear disease and hearing impairment.

Otoscope. An otoscope is used to visualize the tympanic membrane that separates the middle ear chamber from the outer ear canal. Otologic assessment of the middle ear during the first year of life is important to determine whether or not otitis media is present. Regular examination (every three months) allows prompt treatment if the disease is present.

Audiologic Evaluation Instruments. These hearing evaluation instruments aid in the detection of hearing disabilities. An audiometer, for example, is used to evaluate any degree of hearing loss. The detection of a disability by the physician marks the beginning of a diagnostic process that varies in duration and complexity with the chronological and mental age of the child and the nature of the auditory problem.