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QUICK REFERENCES AND FACT SHEETS

Hearing Loss

Hearing loss is one of the most common birth defects; about 3-4 in 1,000 newborns have significant hearing impairment. Hearing loss that is present at birth is called congenital hearing loss. Hearing loss also can develop later in childhood or during adulthood.

Hearing loss can have a major impact on the life of a child and his family. Because language and communication develop so rapidly during the first 3 years of life, an undetected hearing loss is likely to interfere with a child's speech, language and communication with others. Hearing loss also can result in learning problems that affect a child's performance at school. The goal of early screening, diagnosis and treatment is to help children with hearing loss to develop language and academic skills equal to their hearing peers.

Because hearing loss in infancy is hard to recognize, most hospitals screen all newborns before they are discharged. Most states have an Early Hearing Detection and Intervention program to help ensure that infants who don't pass the screening receive follow-up care. The March of Dimes, the American Academy of Pediatrics, the Maternal and Child Health Bureau, the Centers for Disease Control and Prevention (CDC) and others strongly support these programs.

What causes hearing loss in babies and children?

Hearing loss can be inherited (genetic) or can be caused by illness or injury. In some cases, the cause of hearing loss is not known. About 90 percent of babies with congenital hearing loss are born to hearing parents.

Genetic factors are believed to cause about 50 percent of cases of congenital hearing loss. About 25 genes that play a role in hearing loss have been identified.

About 30 percent of children with hearing loss also have other birth defects. In such cases, hearing loss is part of a syndrome (group of birth defects that occur together).

Illnesses that can cause congenital hearing loss include infections during pregnancy, such as rubella (German measles), cytomegalovirus, toxoplasmosis, herpes or syphilis. Babies born preterm also are at increased risk.

After birth, head injuries or childhood infections, such as meningitis, measles or chickenpox, can cause permanent hearing loss. Certain medications, such as the antibiotic streptomycin and related drugs, also can cause hearing loss. Ear infections (otitis media) may cause temporary hearing loss.

Are there different types of hearing loss?

Hearing loss is the decreased ability to hear sounds. When sound enters the outer ear (auricle or pinna), it moves through the ear canal to the eardrum (tympanic membrane). Incoming sound causes the eardrum to vibrate which moves three small bones (ossicles) in the middle ear. In this way, the ear canal, the eardrum and the middle ear transmit sound from the outside world to the inner ear (cochlea). Within the inner ear, thousands of tiny hair cells detect the incoming vibrations and convert them into signals that are relayed to the auditory nerves, which send neural impulses to the hearing center in the brain.

Hearing loss is often discussed in terms of where the loss occurs in the hearing pathway.

- Conductive hearing loss occurs when something interferes with sound passing through the outer or middle ear. A blockage in the ear canal, damage to the eardrum, or fluid or an infection in the middle ear (called otitis media) are examples of conditions that can cause a conductive hearing loss. This type of hearing loss is usually temporary and can often be corrected with medication or surgery.
- Sensorineural hearing loss usually occurs when the hair cells in the inner ear cannot detect all incoming vibrations or when neural impulses are not transmitted to the brain. Prenatal infections, lack of oxygen at birth, or genetic factors can cause this type of hearing loss, which is generally permanent. However, many children can be aided with devices that amplify sound. Sensorineural hearing loss also can result from damage to the brain's auditory center.
- Mixed hearing loss occurs when a child who has a sensorineural hearing loss also has a conductive loss (such as fluid in the middle ear). It is very important that children with permanent hearing loss be monitored and treated for middle ear problems so hearing is not further reduced.

How are newborns screened for hearing loss?
Newborns are screened with one of two tests, both of which measure how a baby responds to sound. Both tests take 5 to 10 minutes, are painless, and can be done when the baby is resting.

In the otoacoustic emissions (OAE) test, a small microphone is placed in the baby's ear. The microphone, connected to a computer, sends soft clicking sounds into the ear and records the inner ear's response to sound.

In the automated auditory brainstem response (AABR) test, soft clicking sounds are presented to the ear through small earphones. Sensors placed on the head and connected to a computer measure brain wave activity in response to sound.

What happens if a baby doesn't pass the hearing screening?
If a baby does not pass the OAE or the AABR, the test should be repeated or the baby should be referred to a hearing specialist (audiologist) or an ear, nose and throat specialist (ENT or otolaryngologist) for more extensive tests to determine if the baby has a hearing loss. It is important for babies to be assessed by specialists who have experience testing very young children. Diagnostic testing should be completed by 3 months of age.

Parents must keep in mind that the screening tests cannot diagnose hearing loss. Up to 5 percent of babies will have abnormal results on their hearing screening test. However, additional tests show that only about 1 in 10 of these babies actually have hearing loss.

How are babies and children tested for hearing loss?
The most common hearing test for infants under 6 months of age is the diagnostic auditory brainstem response test. It is similar to the automated screening test, but it provides more information and must be administered by a specialist.

Children between 6 months and 2 years of age often are tested with visual reinforcement audiometry (VRA).

During VRA testing, a series of sounds is presented to the child through earphones or speakers. The child is trained to turn toward any sound, and is then rewarded with an entertaining visual image for responding.

Children between 2 and 4 years of age are tested with conditioned play audiometry (CPA). They are asked to perform a simple play activity (like placing a ring on a peg) when they hear a sound. This is similar to the test for older children and adults, who are asked to press a button or raise their hand when they hear a sound.

These tests also may be recommended if a child was not screened as a newborn; if he has had persistent ear infections, meningitis or other illness that can cause hearing loss; has been diagnosed with a syndrome that can include hearing loss; or if a parent suspects the child is not responding normally to sounds.

What are some signs of hearing loss in infants and young children?
Parents should be alert to any signs of hearing loss and discuss them with their child's pediatrician. Some signs include: failure to startle at loud sounds; not turning toward the sound of a voice or imitating sounds after about 6 months of age; lack of babbling at 9

months; not using single words by 18 months; or using gestures instead of words to express needs. Parents should be concerned about hearing loss in older children if they develop vocabulary more slowly than their peers; have speech that is difficult to understand or that is too loud or too soft; often ask you to repeat what was said; turn the TV too loud. At school age, children with hearing loss often appear inattentive and have difficulties learning to read or perform simple mathematics, and fall behind at school.

How is hearing loss treated?
A child with a congenital hearing loss should begin receiving treatment before 6 months of age. Studies suggest that children treated this early are usually able to develop communication skills (using spoken or sign language) that are as good as those of hearing peers. Because of a federal law (the Individuals with Disabilities Education Act), children with a hearing loss between birth and 3 years of age have the right to receive interdisciplinary assessment and early intervention services at little or no cost. After age 3, early intervention and special education programs are provided through the public school system.

There are a number of treatment options available, and parents will need to decide which are most appropriate for their child. They will need to consider the child's age, developmental level and personality, the severity of the hearing loss, as well as their own preferences. Ideally a team of experts including the child's primary care provider, an otolaryngologist, a speech-language pathologist, audiologist and an educator will work closely with the parents to create an Individualized Family Service Plan. Treatment plans can be changed as the child gets older.

Children as young as 4 weeks of age can benefit from a hearing aid. These devices amplify sound, making it possible for many children to hear spoken words and develop language. However, some children with hearing loss are helped more than others by hearing aids. Some children with severe to profound hearing loss may not be able to hear enough sound, even with a hearing aid, to make speech audible. A behind-the-ear hearing aid is often recommended for young children because it is safer and more easily fitted and adjusted as the child grows as compared to one that fits within the ear.

Parents also will need to decide how their family and child are going to communicate. If the child is going to communicate orally (speech), he may need assistance learning listening skills and lip reading skills to help him understand what others are saying. Many children with hearing loss also need speech or language therapy.

A child also can learn to communicate using a form of sign language. The type preferred by most deaf adults is American Sign Language (ASL), which has rules and grammar that is distinct from English. There are also several variations of sign language that can be used along with spoken English.

Surgery may be recommended if a child has a permanent conductive hearing loss caused by malformations of the outer or middle ear, or by repeated ear infections. Although fluid in the middle ear usually results in only temporary hearing loss, chronic ear infection can cause a child to fall behind in language skills. In some cases, a doctor may suggest inserting a tube through the eardrum to allow the middle ear to drain. This procedure generally does not require an overnight hospital stay.

Surgery also may be an option for some children with severe to profound sensorineural hearing loss. A device called a cochlear implant can be surgically inserted in the inner ear of children as young as 12 months of age to stimulate hearing. The surgery requires a hospital stay of one to several days. With additional language and speech therapy, children with cochlear implants may learn to understand speech and speak reasonably well, but the amount of improvement is variable.

Does the March of Dimes support research on hearing loss?
Several March of Dimes grantees are exploring the role that specific genes play in causing hearing loss, with the goal of developing treatments for hereditary hearing loss. Others are seeking to prevent hearing loss by preventing infections that can cause it and to improve treatment of individuals with hearing loss. One is developing improved hearing aids that amplify speech more clearly.

For More Information
The National Center for Hearing Assessment and Management provides resources for both health care providers and families. The center has produced a 6-minute educational video for parents "Giving Your Baby a Sound Beginning," which is available in English and Spanish. The video may be viewed online at no charge, or a VHS copy may be purchased for $15.

The Early Hearing Detection and Intervention Program of the Centers for Disease Control and Prevention provides educational materials for both families and health care providers. Visit the program's Web site for more information.

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