

A DISCUSSION OF HEARING PROBLEMS IN CHILDREN

Five thousand children are born profoundly deaf each year in the United States alone. Another 10 to 15 percent of newborns have a partial hearing handicap.

FUNCTION OF THE NORMAL EAR

The ear is divided into three parts: an external ear, a middle ear and an inner ear. Each part performs an important function in the process of hearing.

The external ear consists of an auricle and ear canal. These structures gather sound and direct it toward the eardrum. The middle ear chamber lies between the external and inner ear. This chamber is connected to the back of the throat by the eustachian tube, which serves as a pressure equalizing valve. The middle ear consists of the eardrum and three small ear bones (ossicles): malleus (hammer), incus (anvil) and stapes (stirrup). These structures transmit sound vibrations to the inner ear. In so doing, they act as a transformer, converting sound vibrations in the external ear canal into fluid waves in the inner ear.

The inner ear chamber contains the microscopic hearing nerve endings bathed in fluid. Fluid waves stimulate the delicate nerve endings which in turn transmit sound energy to the brain, where it is interpreted.

TYPES OF HEARING IMPAIRMENT

The outer and middle ears conduct and transform sound; the inner ear receives it. When there is some difficulty in the outer or middle ear, a conductive hearing impairment occurs. When there is trouble in the inner ear, a sensorineural or hair cell impairment is the result. Difficulty in both the middle and inner ear results in a mixed hearing impairment.

CONDUCTIVE IMPAIRMENT

A conductive type of hearing impairment occurs when sound is not conducted efficiently through the ear canal, eardrum, or tiny bones of the middle ear. Conductive losses reduce the loudness of sound that is heard.

A conductive impairment may occur from blockage of the outer ear canal, from a perforation (hole) in the eardrum, from middle ear infection or fluid due to blockage of the eustachian tube, or from a congenital defect or disease of any of the three middle ear bones. This type of impairment is usually correctable through surgery.

The child with a conductive hearing loss will never go deaf. He will always be able to hear, either through ear surgery or by use of properly fitted hearing aid.

SENSORINEURAL IMPAIRMENT

A sensorineural hearing loss is used to describe hearing impairments which result from disturbances or defects in the inner ear and transmission of electrical signals from the hair cells. These impairments may be congenital (i.e. present at birth), hereditary, developmental, or a combination of these. In addition, these impairments may result from infections, injuries, ototoxic drug therapy, or lack of oxygen.

Hearing loss may be divided further due to the cause of the hearing handicap.

A. Congenital hearing loss

1. Genetic - In the genetic type there is an actual defect in your child's genes which results in an abnormal development of the ear.

2. Non-genetic - This is a hearing loss which is due to some problem which occurred during the fetal development or the immediate birth period.

B. Acquired hearing loss - This is a hearing impairment which occurs sometime after birth and is not transmitted to future children.

CONGENITAL FACTORS

Several viral infections, including CMV and German measles contracted by the mother during the first three months of pregnancy may interfere with inner ear development in the fetus. Occasionally, other viral diseases are at fault. The viruses of measles and mumps may cause a sensorineural hearing loss after birth, but this happens infrequently. Immunizations are now available for both of these diseases.

PROBLEMS AT BIRTH

A very difficult and complicated labor or premature birth may also result in an inner ear hearing impairment on occasion. This may be due to lack of oxygen. These are many syndromes which can also result in a hearing impairment at birth. One can have a hearing loss at birth without any hereditary relationship.

Jaundice occurring at or shortly after birth is capable of damaging the inner ear. This is most often due to Rh incompatibility between the mother's and the child's blood. Fortunately, this is not a common occurrence.

HEREDITARY IMPAIRMENT

The development and function of the ear is dependent upon hundreds or even thousands of genes interacting with each other and with the inter- and extrauterine environment. A major cause of late-onset hearing loss is genetic. There are several patterns of inheritance. In autosomal dominant disorders, one parent expresses the trait, which he transmits to 50 percent of his children. In autosomal recessive inheritance, the parents of the children are clinically

normal, but carry the recessive gene to 25 percent of their children. X-link inheritance traits are transmitted from a carrier mother to 50 percent of her sons.

Most cases of hereditary childhood deafness are sensorineural rather than conductive in nature. Most examples of hereditary hearing loss are recessive. Recessive deafness characteristically is associated with retention of hearing of low frequency sounds since most of these cases are associated with abnormalities primarily affecting the first turn or the cochlea (i.e. the Scheibe inner ear abnormality). In dominant inherited deafness, the audiogram generally is flat. However, there are other dominant types of mid-frequency sensorineural hearing loss. In X-link recessive deafness, some retention of hearing is usually seen in all frequencies.

Hereditary sensorineural hearing loss may be present at birth, or may develop later in life. This may be due to inner ear malformations or to other associated syndromes which have an associated inner ear hearing loss. One may see a genetic sensorineural hearing loss with or without associated abnormalities.

INFECTIONS

The most common type of acquired sensorineural loss is meningitis. Frequently this may affect both ears, but can involve one ear. Other types of infections would include viral diseases, such as mumps, rubella and otitis media.

HEARING IMPAIRMENT IN ONE EAR

A hearing impairment that is confined to one ear deprives a person of the ability to distinguish the direction of sound. He will also have difficulty hearing from the involved side in a noisy background. These are minor problems to a young child. When this hearing impairment in one ear is conductive, surgery will usually be able to restore the hearing, giving a better balance of hearing. This is usually done in a child who is in his teens. When the impairment is sensorineural, it is often possible when the child grows older to restore some of this balance of hearing through the use of a special hearing aid (i.e. CROS hearing aid).

TREATMENT

There is no known medical or surgical treatment that will restore normal hearing in patients with sensorineural hearing impairments. We, therefore, rely on rehabilitation through the use of a hearing aid and special training. Fortunately, many children with this type of hearing impairment will not show progression of the impairment as they get older.

THE HARD-OF-HEARING CHILD

If your child's hearing impairment is in the range of 35-70 dB HL, he or she should do well with a properly fitted hearing aid. He or she will probably be able to attend school with normal hearing children. He or she will need preschool speech therapy and auditory training in order that communication abilities will be at the optimal level when regular school starts.

HEARING AID EVALUATION

Evaluation of the hearing in a young child may require several visits with the audiologist. It is important to determine an accurate measurement of both the type and the degree of hearing impairment in order to select the proper hearing aid. An aid that is too powerful for a young child may be uncomfortable and cause the child to reject it. On the other hand, if the aid is not strong enough, a child may receive little or no benefit from it and therefore object to wearing it.

SPEECH READING (LIP READING)

Speech reading is very important whatever the type of degree of impairment. This skill enables a person with impaired hearing to understand conversation by attentively observing the speaker. All of us, whether we have a hearing loss or not, employ the sense of sight as well as the sense of hearing in ordinary conversation. We find it easier to comprehend if we can watch the speaker's facial expressions, lip movements and gestures. Just as the visually handicapped learns to use his sense of hearing to compensate for his impaired sight, the person with defective hearing must learn to use his eyes to assist him in hearing. A study of the fundamentals of lip reading or of speech reading, as it is called, will make communication less of an effort and therefore more pleasant for both the speaker and the listener.

Speech reading has its limitations. For example, when the distance between the speaker is great or when there is inadequate lighting or defective vision, one may not always be able to see the speaker's lips clearly enough to speech read adequately. Some persons do not open their mouths very far when they speak and, consequently, their lip movements are very limited. Others have beards, hold their hands over their mouths, or smoke as they talk, making speech reading difficult if not impossible.

It is important to tell other family members and friends to get the child's attention before speaking. The child with a hearing impairment must recognize characteristics of the English language. Many sounds and many words look the same on the lips. The hearing impaired child will find it impossible to see certain words on the lips and therefore needs to continuously fill in the "gaps" of words and sentences. Two thirds of all sounds in the English language are not visible on the lips. Because of the difficulties presented by sounds, the speech reader is encouraged to follow the context or thought of what is being said rather than to try to lip-read each word.

The child, who is learning to speech read, learning to use a hearing aid, or both, should have help from a professional person trained to teach these skills. There are many books on the subject of speech reading. Help is also available at various universities.

Cued speech is a phonemically-based hand supplement to speech reading (speech is made up of sounds called phonemes). The 26 letters in our English language, either singly or in combinations, produce 43 phonemes. Spelling does not illustrate the pronunciation differences whereas cued speech can show the child how something is pronounced while it is being spoken. It is comprised of eight hand shapes used to represent groups of consonant sounds and four positions about the face to represent groups of vowel sounds. Combinations of these hand shapes and placements are used to illustrate exact pronunciations in words in connected speech. Although cueing helps recognize pronunciation, the child will still need speech lessons with a speech therapist. Some deaf educators find some limitations with cued speech, but in certain specific instances this has been used with some success.

PROFOUND SENSORINEURAL HEARING IMPAIRMENT

For those children who are not able to achieve any benefit with a hearing aid and an oral educational program, they will require training in manual forms of communication such as finger spelling or American Sign Language. The type of school depends upon the child's hearing level, progress, and communication skills. If your child's level is greater than 70 dB, he or she will not, in all likelihood, be able to attend classes with normal hearing children, at least in the beginning. It will be difficult for him or her, but with the help of a hearing aid, training in speech reading, and attention to speech correction, he or she may be able to progress through schools for the hearing handicapped to normal schools, to college, and to take his or her place in society with normal hearing people.

For those children who are not able to achieve understanding for speech, special schools are available to train them in the manual form of communication. The type of school a child attends depends upon his progress in communication.

REHABILITATIVE MEASURES

There are two very important factors to be determined upon examining the child with a suspected hearing impairment. First, determination should be made regarding the presence of a hearing loss and the type (i.e., conductive or sensorineural). Secondly, once a hearing loss is found to be present, it should be determined if this loss is progressive or stable. Therefore, your child may require periodic audiograms to be sure that the hearing loss is going to remain stable.

A complete otologic/audiologic examination by a competent ear specialist and audiologist are necessary to determine what type of hearing impairment is present, its probable cause, and its treatment. At times it may be necessary to obtain special x-rays of the inner ear, a balance test or other laboratory tests to make this decision.

A well-rounded program of rehabilitation for children with hearing loss may include speech reading, auditory training, speech therapy and instruction in the use of a hearing aid. One may also consider other adjuvants to assist with their communication skills such as cued speech or other manual techniques. All aspects of the program do not necessarily apply to each child with an impairment, but each individual may be helped through some of these methods.

THE COCHLEAR IMPLANT

The cochlear implant is an electronic device that is implanted into the inner ear of a severe to profoundly hearing impaired child. This device is only utilized in the child who cannot benefit with a hearing aid. It is a device which is used to bypass the disease or nonfunctional hair cells and converts the sounds we hear to electrical impulses which directly stimulate the cochlear nerve. The implant consists of an external portion comprised of a microphone, sound processor, and external coil and an internal portion that must be surgically implanted. The surgical procedure involves the placement of an internal coil beneath the skin behind the ear and a stimulating electrode which is inserted into the cochlea or inner ear.

To determine suitability for this device in the severe to profoundly hearing impaired child, a careful examination is required. The evaluation is performed to determine whether or not the child can receive adequate information from a powerful hearing aid, or whether or not the procedure can be performed and give the expected improvement.

Currently there are several multiple channel devices which are utilized. This is related to the

number of stimulating electrodes within the cochlea.