

HEARING DISORDERS

A disorder of hearing may be defined as a problem with any part of the hearing mechanism, which prevents it from functioning normally. Hearing disorders are commonly classified into major categories: *conductive, sensorineural, mixed, nonorganic* hearing losses, and *central auditory processing*.

A. CONDUCTIVE HEARING LOSS

A conductive hearing loss is defined as a malfunction of the external and/or middle ear while the inner ear and nerve are normal. The effect of a conductive hearing loss is reduction in the level of sound being conveyed or "conducted" to the inner ear. The symptoms associated with this type of impairment may vary with the degree of the hearing loss but, in general a person with a conductive hearing loss:

- demonstrates either a hearing loss predominately in the **low frequencies** or a hearing loss **extending equally across all frequencies**;
- understands speech well when the **loudness of the speaker is increased** sufficiently to overcome the amount of the conductive hearing loss;
- is usually **not annoyed** by the presence of **loud sounds** in his/her environment;
- appears to **understand speech in a noisy environment better** than a person who has normal hearing;
- **may speak** in a relatively **soft voice**.

The causes of a conductive hearing loss are described according to the part of the hearing mechanism, which is affected: the external ear or the middle ear.

1. The External Ear

- a) The absence or malformation of the pinna and the external auditory canal. This condition is most often a birth defect commonly referred to as atresia. The treatment may involve the surgical construction of the pinna and the external canal.
- b) Obstruction of the external auditory canal. The presence of a foreign object or a build up of cerumen in the external canal may cause a reduction in the level of sound reaching the eardrum. The treatment is the removal of the obstruction.

2. The Middle Ear

- a) Inflammation or infection of the lining of the middle ear cavity. This inflammatory process is commonly referred to as otitis media and may result from an upper respiratory infection which has gained access to the middle ear through the Eustachian tube. The infection often results in a build up of fluid in the middle ear space, which, because of its presence, reduces the normal transmission of sound across the ossicular chain. Also, a similar problem occurs when the opening of the Eustachian tube is blocked because of enlarged adenoids. In both cases, if drainage from the middle ear space does not occur and the volume of fluid is increased within the cavity, the eardrum begins to bulge from the force of the fluid pressure. The fluid build up is referred to as middle ear effusion or "serous otitis media". A further increase in fluid build up may result in severe pain and possibly a ruptured eardrum. A short-term problem of otitis media is referred to as acute otitis media, whereas, a long-term involvement is referred to as chronic otitis media. Another cause of hearing loss among children is related to allergies. The result of this condition is very similar to those described above.

The treatment for middle ear infections may depend on the severity of the problem or, on the specific cause. Therefore, the treatment may vary from the administration of antibiotics, to the insertion of ventilation tubes through the eardrum, the removal of the tonsils and adenoids, or allergy management. If the presence of a long-term infection has destroyed or damaged the ossicles, or has entered the air-filled spaces of the mastoid, it may be necessary for the physician to remove the infected area and surgically rebuild the damaged structures at a later time.

- (b) Perforation of the eardrum. Damage to the eardrum may be the result of punctures produced by foreign objects, extremely loud sudden sounds such as an explosion or gun fire, increased external pressure, or by fluid pressure build up within the middle ear space. If the eardrum does not spontaneously repair itself, the physician may find it necessary to patch the perforation.
- (c) Otosclerosis. This conductive hearing loss is not thought to be a disease process found in children, but is restricted usually to young adult and older age groups. However, there are isolated reports of this condition occurring among children. Otosclerosis is caused by deposits of a bony substance around the footplate of the stapes. Upon the hardening of the bony substance, the stapes cannot move and the level of the sound reaching the inner ear is reduced.

The treatment for otosclerosis consists of the surgical removal of the stapes or other affected members of the ossicular chain. Following the removal of these bones, the surgeon may insert a synthetic device to bridge the gap between the working portion of the ossicular chain and the oval window, which housed the footplate of the stapes.

- (d) Malformation. The ossicles of the middle ear may be malformed at birth causing the sound reaching the inner ear to be reduced. The treatment for this is removal of affected area and insertion of a synthetic device to bridge the gap between the tympanic membrane and the inner ear.

B. SENSORINEURAL HEARING LOSS

A sensorineural hearing loss is defined as a malfunction of the inner ear (cochlea) and/or the auditory nerve, in the presence of a normal external and middle ear. A sensorineural hearing loss may result in both a reduction in the loudness level of sound, and a loss of the ability to discriminate speech sounds. The symptoms associated with a sensorineural hearing loss may vary with the severity of the problem or the location of the problem. A person with a sensorineural hearing loss:

- demonstrates a hearing loss, which may range from mild to profound in one or both ears which may be greater for the higher frequency sounds;
- may demonstrate a reduced ability to understand speech with the common complaint of "I can hear, but I can't understand". The limitations imposed upon the understanding of speech may vary with the amount of the hearing loss. The high frequency sounds represent majority of the consonant sounds which give most of the information to understand what is being said. Refer to figure 4 and 5 in identifying the speech range and visual clues of consonant sounds.
- may display an inability to tolerate loud sounds;
- may have poor speech because of the inability to hear others as well as to monitor oneself;
- may speak in a relatively loud voice;
- will often complain of a ringing or buzzing sound in his ears. This problem, although not totally understood, is referred to as tinnitus.

The cause of sensorineural hearing loss can best be described according to the time in life when the hearing loss begins. If the hearing problem develops before birth, it is termed congenital; if it occurs after birth, it is termed acquired.

1. Congenital Hearing Loss

A congenital hearing loss is one that is present at the time of birth. The causes of congenital hearing problems are listed in the Joint Committee on Infant Hearing (JCIH), 2000 Position Statement which can be found online at: <http://www.jcih.org/posstatemts.htm>

2. Acquired Hearing Loss

An acquired hearing loss is one that occurs after birth. This loss can be caused by ototoxic medications, infections, exposure to loud noise and others factors. Review the causes of acquired hearing loss listed in the JICH, 2000 Position Statement (<http://www.jcih.org/posstatemts.htm>).

Since the sensorineural hearing loss is rarely corrected through the use of medication or surgery as is a conductive hearing loss, the first and most practical approach to the reduction of sensorineural hearing loss is through a program of prevention. This program should include the identification of drugs that damage the inner ear (ototoxic drugs), inoculation against childhood diseases, control of noise levels, and counseling with regard to hereditary and Rh blood incompatibility problems.

For those individuals who have been born with or have acquired a sensorineural hearing loss, there are rehabilitation programs designed to reduce the effects of these problems. These may include selection of a suitable hearing aid, a cochlear implant (which is a electronic device surgically implanted into the cochlea with external transmitter and microphone) for those with a profound hearing loss, development of auditory speech reading skills (lip reading), speech therapy, sign language, special educational placement, and psychological counseling.

C. MIXED HEARING LOSS

A mixed hearing loss is a combination of a conductive and a sensorineural loss in the same ear. An example of this is the individual who has a sensorineural congenital hearing loss, while at the same time, is experiencing a conductive hearing loss because of the presence of a middle ear infection.

The symptoms associated with a mixed hearing loss may be either characteristic of a sensorineural or conductive problem, or may be a combination of the symptoms of both disorders. Causes and treatments for mixed hearing losses

are the same as those discussed previously in the sections entitled "Conductive Hearing Loss" and "Sensorineural Hearing Loss".

D. NONORGANIC HEARING LOSS

A nonorganic hearing loss may be defined as a hearing loss for which there is no known physical basis but is thought to be a result of the psychological state of the individual. Some of these children may have a positive history of ear infections, a factor which can be misleading to the audiometrist. The following are symptoms which may be associated with this hearing disorder:

- The hearing test usually reflects mild to moderate, flat, bilateral "hearing loss".
- A child who displays a nonorganic hearing loss usually has good speech and no difficulty in communicating in normal conversation. When the hearing of the person is audiometrically tested, however, the results are significantly poorer than would be predicted.
- The results of repeated hearing tests are often inconsistent for an individual manifesting a nonorganic hearing loss.

In these cases it is essential to determine if a hearing loss is indeed present and to find out if there are factors that may have led to the child falsely elevating their thresholds. In children these factors are more commonly a desire for increased attention and may be accompanied with disruptive social behaviors. Dealing with these behaviors will be the necessary treatment.

E. CENTRAL AUDITORY PROCESSING DISORDER

Central auditory processing refers to the way the brain uses the auditory information it receives that originated from the outer, middle and inner ear. A central auditory processing disorder (CAPD) is an auditory communication disorder. CAPD is not a dysfunction of the mechanisms of hearing. People with this disorder have difficulty understanding, interpreting, and using the information they hear. Children may exhibit poor language and/or listening skills and, although, they may have adequate to high intelligence, their academic performance falls below their estimated potential. The major complaint of parents and teachers is that the child is performing below expected levels at school and doing so for reasons that are not clear. While the cause of CAPD is not known, the resulting communication problems are well documented. Those affected have trouble following verbal directions and may seem distracted and inattentive. CAPD can exist alone or with other problems such as attention deficit disorder, learning disabilities, and language disorders. There is a higher incidence in children with middle ear pathologies.

Children with CAPD often give the impression that they are not listening. Listening, one of the basic tools of learning is a skill used to develop speech, language, and psychosocial behavior. In the first two years of school, children are exposed to verbal information between 75 and 95 percent of the time. The child with CAPD has difficulty listening and following rapid verbal information, this difficulty leads to frustration and to an ever widening learning gap. As academic and social demands increase, children frequently become discouraged, and may lose self-esteem. Children with auditory processing difficulties need to experience success and build self-esteem, even more than they need to increase memory and listening skills. When a central auditory problem is suspected or identified, immediate measures must be taken to improve the child's listening environment, especially in the classroom.

The child with normal hearing on traditional audiologic tests, but with a case history that leads one to suspect the child's auditory skills, is a prime candidate for central auditory evaluation and can be confirmed by central auditory testing.

Children with central auditory processing disorder will show some or all of the following behaviors:

- Inconsistent responses to sound. Parents/teachers may suspect a hearing loss, though audiometric test results are normal.
- Academic performances below their estimated potential, especially in reading, spelling, and language arts.
- Speech problems such as omissions, distortions, and substitutions of sounds in words. Poor vocal monitoring of loudness of his/her voice.
- Poor auditory attention and comprehension, especially in the presence of background noises. Does not pay attention or listen carefully to instructions. Short attention span.
- Difficulty following verbal instructions. Inability or confusion with carrying out verbal instructions.
- Difficulty distinguishing between similar words. Difficulty recalling the oral spelling of a simple word.
- Good performance in a one-to-one situation, but poor performance in the classroom.