

Hearing Loss and Osteogenesis Imperfecta

Introduction

Significant hearing loss has been reported in approximately 50% of people with osteogenesis imperfecta (OI) beginning any time from childhood into middle age. While not everyone who has OI develops hearing loss, the incidence is much higher than in the general population. Sometimes visible deformities in the ossicles and inner ear can lead to hearing loss. Sometimes the cause of the loss is not visible. Environmental factors can contribute to hearing loss in a person with OI in the same way as for anyone else. These include loud noises, head injuries and infection. Hearing loss can start at any time but among people who have OI the loss often starts at an earlier age and the likelihood increases with age. Hearing loss can be treated with hearing aids and/or surgery depending on the type and severity of the loss.

How Does Hearing Work?

Our ability to hear depends on two factors—the mechanical translation of the sound waves into movement within the middle ear and then translation of that motion into a nerve transmission to the brain. In the first part, the sound waves make the tympanic membrane vibrate. That movement is transmitted through three small bones in the middle ear to the fluid in the inner ear. The movement of the fluid makes small hair cells move and that movement alters the signal along the hair cell nerve to the brain.

Types of Hearing Loss

There are two main types of hearing loss: conductive and sensorineural. Both types of hearing loss can occur with all types of OI. Hearing loss type can vary within a family just as much as between unrelated people. Sensorineural hearing loss tends to be more frequent as a person ages. Conductive loss generally occurs around age 20 or 30, but there are many exceptions. OI related hearing loss can occur at any age, including childhood.

Conductive Hearing Loss: Usually results from a physical problem in the external or middle ear. It may occur as a result of ear infection, blockage of the middle ear, or from fixation or fracture of the stapes, one of three tiny bones in the middle ear that transmits the movement of the eardrum through the middle ear to the fluid in the inner ear.

Sensorineural Hearing Loss: This type of hearing loss occurs when the inner ear is not transmitting the nerve signals normally to the brain.

In addition, hearing losses are classified according to the degree of severity:

- Mild,
- Moderate,
- Severe,
- Profound.

Hearing losses are also classified according to the sound frequency that cannot be heard: low, high, all frequencies.

Symptoms

Some of the early signs of hearing loss are:

- Difficulty understanding certain words or parts of words.
- Frequently asking others to repeat themselves.
- Difficulty understanding others on the telephone.
- Turning up the sound on the television or radio to a level that is too loud for others in the room.
- Difficulty hearing in noisy surroundings.

As it progresses, hearing loss can interfere with communication, performance at work or school, social activities, and personal relationships. Left untreated, hearing loss may even result in isolation and depression.

Diagnosis

In many states, newborns are screened for hearing loss. Most children are screened every few years at school or as part of a routine physical. Regular hearing checks are recommended. A pediatrician can do the general screening, but a registered audiologist, or a physician who is an ear nose and throat (ENT) specialist (otolaryngologist) with pediatric experience should perform the hearing assessment. Hearing tests should be done in a soundproof booth with headphones. Hearing tests done in open rooms are inaccurate because background noise interferes with the results. A special test called Evoked Response Audiometry can be used if the person cannot cooperate with the testing process.

Recommendations

It is recommended that children with OI have a formal hearing evaluation before they start school and be repeated every 3 years. Any child with OI who demonstrates articulation problems, speech delays, recurrent ear infections, or whose parents suspect a hearing loss should have a formal audiologic assessment regardless of age. If borderline hearing is discovered, then yearly testing with a certified audiologist is recommended.

Adults with borderline hearing should have yearly testing and follow up appointments similar to the schedule for children. Adults experiencing tinnitus (ringing in the ear) or symptoms of hearing loss should also have an audiologic assessment that includes a hearing test with air and bone conduction and speech reception threshold. This type of test can determine if the hearing loss is conductive or sensorineural. Adults who have an identified hearing loss should continue to see their audiologist or ENT on a regularly scheduled basis and whenever they feel their hearing has changed.

Ear Protection

Since OI increases the risk for hearing loss. It is important for people to protect and preserve their hearing to defer or minimize any loss.

1. **Noise is a common cause of hearing loss.** Damage from noise actually kills the tiny hair cells in the inner ear. This damage is permanent, painless and adds up over time.
2. **Use ear protection** -- ear plugs or ear muffs -- when you attend noisy events such as rock concerts, car races, or indoor sporting events.
3. Control the volume on iPods and other devices that send sound directly into your ear. Don't go above the halfway setting on the volume control, or level 4 on a scale of 1-10. If the person sitting next to you can hear your music, the volume is too loud.

Hearing Aids

While hearing aids can't "cure" hearing loss or duplicate natural hearing, they provide amplification that can help all age groups. When hearing loss is identified, hearing aids are usually tried first because, unlike surgery, they carry little risk. Hearing aids come in a wide variety of shapes, sizes and ability to amplify. Hearing aid technology has been rapidly improving. No single model has been identified as appropriate for all people who have OI. The organizations listed at the end of this fact sheet offer literature to help people learn about hearing aids and become educated consumers.

Surgery

Surgical procedures such as **stapedectomy** or **cochlear implant** help some people with OI related hearing loss that is severe, conductive and progressively getting worse. In a stapedectomy, the fixed footplate of the stapes is replaced by a prosthesis that allows the normal transmission of signals to the inner ear. Using a laser to do the surgery improves outcomes. Success of the surgery is highly dependent on the surgeon's experience and on the person's ear anatomy. Complications from stapedectomy surgery can include dizziness, change in taste to part of the tongue and worsening of the hearing loss rather than an improvement. Initially successful stapedectomies can fail for a number of reasons. There may be more problems in the ear than just with the stapes, the attachment of the prosthesis to the adjacent small bone called the incus may be faulty, the incus may be too thin to hold the prosthesis long term, or damage can occur to the inner ear leading to nerve loss or malfunction.

A cochlear implant is an electronic device that can provide partial restoration of hearing that was lost because the very first part of the nerve transmission pathway is defective. A portion of the device is implanted behind the

ear and electrodes are threaded into the cochlea inside the ear. This is an option for someone who has a sensorineural hearing loss. This surgery has successfully been performed on people with OI. Having a cochlear implant is significant ear surgery that may require a brief hospitalization. Complications can include dizziness and facial nerve injury.

Since OI bone is not as solid or dense as bone in other people, the electrical signals from the device seem to travel a little more easily through the bone. This means that the facial nerve, which runs near the inner ear, may be stimulated from the implant. Stimulation of the facial nerve can be corrected by turning off some of the electrodes. Hearing with a cochlear implant is variable. It requires a period of training and adjustment after the surgery. Hearing does not return to “normal,” but many people adapt well and can even return to talking on the telephone.

A number of important issues need to be assessed, discussed, and clarified before any person with OI can be considered a “good candidate” for surgery. These include anesthesia, cardiac and respiratory issues. As a general rule, patients should seek treatment centers where the otologists (physicians who specialize in ear disorders) have considerable experience with stapes and/or cochlear implant surgery.

Don't Be Afraid to Seek Help

There are many devices available on the market to help people cope with hearing loss. These can include special doorbells, telephones, smoke alarms that light up and alarm clocks that vibrate. Since hearing loss is an issue for the general population, a number of local and national organizations provide information and have services available to those with hearing loss.

Additional Resources

American Speech-Language-Hearing Association

(800) 638-8255

www.asha.org

Alexander Graham Bell Association

(202) 337-5220, TTY (202) 337-5221, Fax (202) 337-8314

Email: info@agbell.org

www.agbell.org

Hearing Loss Association of America

(301) 657-2248; (301) 657-2249 TTY

www.hearingloss.org