Hearing Loss and Osteogenesis Imperfecta

Significant hearing loss has been reported in approximately 50% of people with Osteogenesis Imperfecta (OI). While not everyone who has OI develops hearing loss, the incidence is much higher than in the general population. Sometimes abnormalities involving the ossicular chair in the middle and inner ear can lead to hearing loss. Sometimes the cause of the hearing loss may not be obvious. 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 managed with hearing aids and/or surgery depending on the type and severity of the loss.

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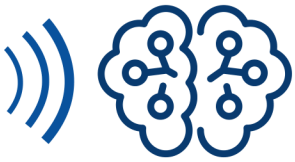
How Does Hearing Work?

Our ability to hear depends on two factors—the transformation 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 (ossicles) to the fluid in the inner ear. The movement of the fluid makes small hair cells in the inner ear 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. Type of hearing loss 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
- Mid
- High
- Or a combination of above

Diagnosis

In all UK NHS hospitals, parents are offered a screening hearing test for their child soon after birth. Most children are also screened at school entry and regular hearing checks are recommended in those with OI. Hearing screeners can carry out hearing screening at birth, but a registered audiologist working with trained paediatrician, Paediatric ENT or Audiovestibular Physician (AVP) should be able to perform the hearing assessment. The tests are carried out as a part of NHS service. Hearing tests should be done in a soundproof room with earphones or headphones.

Hearing tests carried out in open rooms are usually inaccurate because background noise interferes with the results. A special test called Evoked Response Audiometry (ERA) combined with tympanometry and Evoked Otoacoustic Emissions (OAEs), can be used if the child cannot cooperate with the testing process.

Recommendations

It is recommended that children with OI have a formal evaluation of hearing before they start school. Routine testing should begin at age 10 years 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 registered 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.

Hearing Loss in Childhood

Management of hearing loss in children is dependent on the cause similar to other hearing losses. If it occurs early in life, speech therapy usually is necessary. Amplification i.e. hearing aids, and FM devices when of the hearing levels are greater than the normal hearing range (i.e. hearing loss). FM devices can be used if the person has hearing difficulty in noise in site of normal audiometric thresholds.

Preferential seating up front in the classroom is always a good move. If sound muffling can be done (carpeting, acoustic tiles) to soften the overall background noise in the classroom, understanding is easier. Teachers can wear a microphone that sends sound directly to the hearing device the child is wearing or specifically set up classroom FM speaker system. Those with hearing loss may be able to claim disability living allowance. Although most schools readily accommodate special needs, sometimes an advocate is necessary and can be accessed through the school action plus or a statement of educational needs depending on the severity of the disability.

Ear Protection

Exposure to loud noises can lead to sensorineural hearing loss by damaging the hair cells (sound sensing cells) in the inner ear. The loudness and duration of the noise are the important factors determining damage, not the source of the noise. Power tools, chain saws, and loud music are but a few of the noises that can damage our hearing. Use of protection when working around loud noise or avoidance of the loud noise exposure can prevent this problem. Many different kinds of ear plugs and ear muffs are available to block out sound.

The devices carry a rating of effectiveness on their packaging that tells how much sound is blocked out. When one must be around loud noise, preventive measures can help avoid permanent loss. Another important way to protect hearing is to keep the volume turned down whenever using a device that has personal headphones. Those with a conductive hearing loss may have some natural protection (muffling effect of sounds because of the hearing loss) from excessively loud sounds. More information on this should be sought from the audiologists.

Hearing Aids

While hearing aids can't "cure" hearing loss or restore 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. 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. The severity and range of the hearing loss influences which type of aids are best suited to your needs. Thoroughly discuss the styles of hearing aids and the features that suit your needs with your audiologist.

Most individuals with bilateral hearing loss will benefit from binaural aids (one in each ear) that provide a stereo effect.

Aids equipped with a "T-Switch" make it easier to connect with other assistive listening devices. For children hearing aids offer connection to a FM device (e.g. microlink radioaids) so that they can hear the teacher well. People who have sensitive skin or contact allergies may want to get information about non-allergenic earmoulds. Getting a good fit and understanding how a particular model hearing aid is designed to work will help the individual get maximum benefit.

For example, when a hearing aid is turned up too loud, it can give feedback, or the highpitched squeal that some people complain about. If feedback is a problem, then the individual may need a different kind of hearing aid, or a better fitting mould. It takes time and patience to adjust to a hearing aid. Therefore it is important to work with a reputable dealer who can provide the necessary follow-up care.

Hearing aids are provided free to all individuals with hearing loss in the UK through the NHS. However, FM devices are usually provided by the local educational authority. It is good practice to be under one audiologist in your the local audiology department.

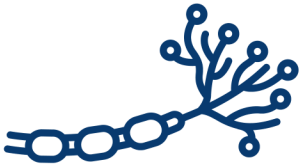
Surgery

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, a copy of the bone, that allows better transmission of the bone movements to the inner ear. Using a laser to do the surgery improves accuracy and visibility during the surgery.

Success of the surgery is highly dependent on the surgeon's experience and on the person's ear anatomy. The procedure itself takes less than an hour and the patient may be discharged from the hospital in less than 24 hours. 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 (stirrup), the attachment of the prosthesis to the adjacent small bone called the incus (anvil) 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 profound sensorineural hearing loss and cannot hear enough with the best possible hearing aids. This surgery has successfully been performed on people with OI. Having a cochlear implant is significant ear surgery that may require a brief period in hospital. 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 anaesthesia, cardiac and respiratory issues. As a general rule, patients should seek treatment centres 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.

Notes

Scan to donate to the BBS

If you found our factsheet helpful and would like to support the BBS in continuing to provide resources, you can donate by scanning our QR Code

Compiled by the Brittle Bone Society in collaboration with BBS Medical Advisory Board and POINT (Paediatric Osteogenesis Imperfecta National Team). The information in this leaflet is correct as at 31st July 2021 but we cannot guarantee that it will be accurate and current at any given time. This leaflet is not intended in any way to replace the advice of your doctor or other medical professional. Leaflets are available online at www.brittlebone.org. This information is available in accessible formats on request.

The Brittle Bone Society (BBS) is a registered charity in Scotland (SCO50854) and company limited by guarantee (SC677346), supporting the OI community throughout the United Kingdom and in Ireland.