About 50% of people who have OI experience OI-related hearing loss during their lifetime. In some people it can progress to a profound loss or almost total deafness. Hearing aids are the first treatment. Stapedectomy surgery will benefit some people with OI. In recent years, because of improvements in technology and surgical techniques, cochlear implants have become available to people with OI-related hearing loss.

What is a cochlear implant?
The cochlea is an important part of your hearing equipment. It is the major organ of the inner ear. It is often described as looking like a snail. A cochlear implant is a sophisticated device that combines digital technology and information processing. The system consists of two parts: a tiny electrode that is surgically implanted inside the ear and threaded into the cochlea, plus an external part that combines a microphone, speech processor, and power source. A cochlear implant won’t cure deafness, but it can offer a significant improvement.

How does it help a person hear?
Our ability to hear depends on two factors:
• the mechanical translation of sound waves into movement within the middle ear, and
• the translation of that motion into a nerve transmission to the brain.

In normal hearing, sound is collected by the outer ear and sent into the middle ear. Here the sound waves make the eardrum vibrate. The vibrations go through three tiny bones including the stapes to reach the inner ear. Next the sound waves are transported to the cochlea by the fluid of the inner ear. Inside the cochlea, sound wave vibrations are converted to electrical impulses by tiny hair like cells. The electrical impulses are sent to the brain through the auditory nerve. The brain interprets this information and recognizes specific sounds. All of this happens much quicker than it takes to explain.

The cochlear implant bypasses the middle ear, and substitutes for the defective inner ear. Sound is captured by the speech processor and coded so it can be sent into the inner ear as electrical signals. These signals are picked up by the auditory nerve and sent to the brain for interpretation.

Who is a candidate for this type of surgery?
Adults with severe to profound sensorineural hearing loss in both ears are candidates for a cochlear implant. Other requirements include:
• A working auditory nerve
• Intact cochlea
• No longer benefiting from hearing aids
• Score of less than 50% of words in sentences on a speech recognition test such as the Hearing in Noise Test
• Generally healthy enough to be able to handle a 2-3 hour surgery. Adults with cardiac or respiratory problems may not be able to tolerate the surgery.

Continued on next page
How can I find out if this treatment would work for me?

The process for getting a cochlear implant involves a consultation with an otolaryngologist, a number of tests including a CT scan, a psychological evaluation, and a series of informational and counseling appointments.

Are there risks that a person with OI should know about?

There are several possible side effects, but risks are relatively minimal. One reported side effect is facial nerve injury. This occurs when the facial nerve, which runs near the inner ear, is stimulated by the implant and causes twitching. This does not damage the nerve and long term problems do not usually result. This injury occurs in about 1 percent of patients, and facial nerve monitoring is now done during the surgery to minimize this risk. It is possible that people with OI might be more at risk than others for facial nerve injury because OI bone is not as solid or dense as bone in other people. This may allow the electrical signals from the device to travel a little more easily through the bone. Stimulation of the facial nerve can be corrected by adjusting the electrodes.

Dizziness or a sense of imbalance can occur after cochlear implant surgery, but it is usually temporary.

A small risk of meningitis has been reported. This can be prevented with a vaccination before surgery.

It has been discovered that the cochlea is misshapen in some people with OI. This makes implantation of the electrodes more difficult and may lengthen time in surgery.

Will my hearing return to normal after an implant?

Hearing with a cochlear implant is variable. It requires a period of training and adjustment after the surgery. It takes time, training and commitment to learn how to listen through the device. After the surgery there is a required series of follow-up care appointments so the implant can be activated. Hearing does not return to “normal,” but many people adapt well and can even return to talking on the telephone.

Is a cochlear implant expensive?

Newspaper reports indicate that total cost is around $40,000 (this includes all evaluation tests, out patient surgery, the device itself, and follow-up appointments.) Many insurance companies cover all or most of the cost. Medicare also covers cochlear implants in some situations.

Where can I get more information?

- American Academy of Otolaryngology
  www.entnet.org
- Hearing Loss Association of America
  (formerly Self-Help for Hard of Hearing or SHHH)
  www.hearingloss.org
- The Listening Center at Johns Hopkins Medical Center
  601 North Caroline Street
  Baltimore, MD 21287-6214
  410-955-6554

This information was prepared with the assistance of David Vernick, M.D., Chief – Otolaryngology, Beth Israel Deaconess Hospital, Boston, MA, a member of the OI Foundation Medical Advisory Council.