

## Key points

- Hearing loss is very common in the Australian adult community. When the loss is severe, it can have a major impact on employment and quality of life.
- The surgical implantation of an electronic prosthesis into the inner ear – cochlear implantation – is the intervention of choice for severe-to-profound levels of hearing loss.
- The fundamental indication for cochlear implantation is bilateral severe-to-profound hearing loss for which well-adjusted hearing aids have given little or no benefit.
- Well-selected candidates with reasonable expectations can achieve highly satisfying outcomes from cochlear implantation, including benefits to auditory communication within their work and social environments.
- GPs have an important role in recognising potential candidates for cochlear implantation and facilitating their preliminary investigations, as well as in the early recognition and treatment of implant infections.

# Severe hearing loss in adults

## Is cochlear implantation an option?

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Severe hearing loss can have a major impact on employment and quality of life. The surgical implantation of an electronic prosthesis into the inner ear, commonly known as cochlear implantation, is now the intervention of choice for adults with severe-to-profound levels of hearing loss. GPs play an important role in identifying potential candidates for cochlear implantation as well as in the early recognition of infections associated with the implant.

**H**earing loss is common in the community.<sup>1</sup> When mild, it responds well to a hearing aid and the disability associated with it can be minimised. However, disability increases with the extent of the hearing loss and when the loss is severe, it can have a major impact on employment and quality of life. Safety in dangerous environments also becomes a significant issue.

The surgical implantation of an electronic prosthesis into the inner ear, commonly known as cochlear implantation, is now the intervention of choice for severe-to-profound levels of hearing loss. For the severely deafened patient for whom the most powerful hearing aids have failed, the outcomes of implantation are often a dramatic and beneficial improve-

ment in hearing and communication ability. These benefits are shared with the recipient's family, social associates and work colleagues.

### HOW WE HEAR

Hearing requires sound to be transmitted mechanically through the external and middle ear components (the conductive components) to the inner ear (Figure 1). Within the inner ear, the mechanical sound energy is converted to electrical signals by the hair cells (the sensory components) in the organ of Corti (the end organ of hearing within the cochlea), which in turn leads to stimulation of the auditory nerve and higher neural pathways, ultimately reaching the auditory cortex in the temporal lobe.

Disruptions to the conductive hearing

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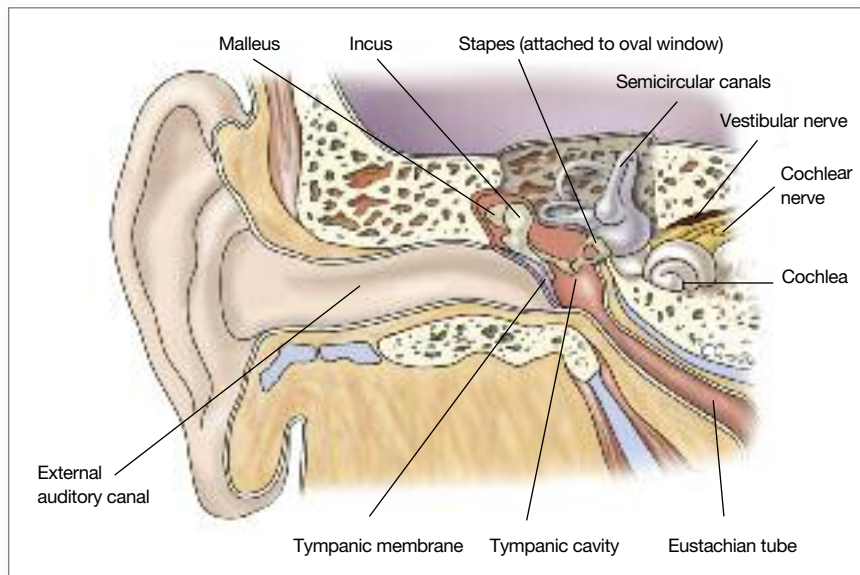


Figure 1. The anatomy of the ear.

mechanisms (e.g. caused by otitis media, perforations or otosclerosis) produces hearing losses that are mild-to-moderate in degree. Such hearing loss responds well to treatment with surgery or hearing aids. In contrast, problems affecting the sensory mechanisms in the inner ear (e.g. caused by ageing, genetics, Meniere's disease,

trauma, meningitis or ototoxicity) may produce severe-to-profound hearing losses. Severe-to-profound hearing loss responds poorly to hearing aids and is associated with a significant reduction in quality of life. It is for patients with this latter type of hearing loss that the cochlear implant has been designed and developed.

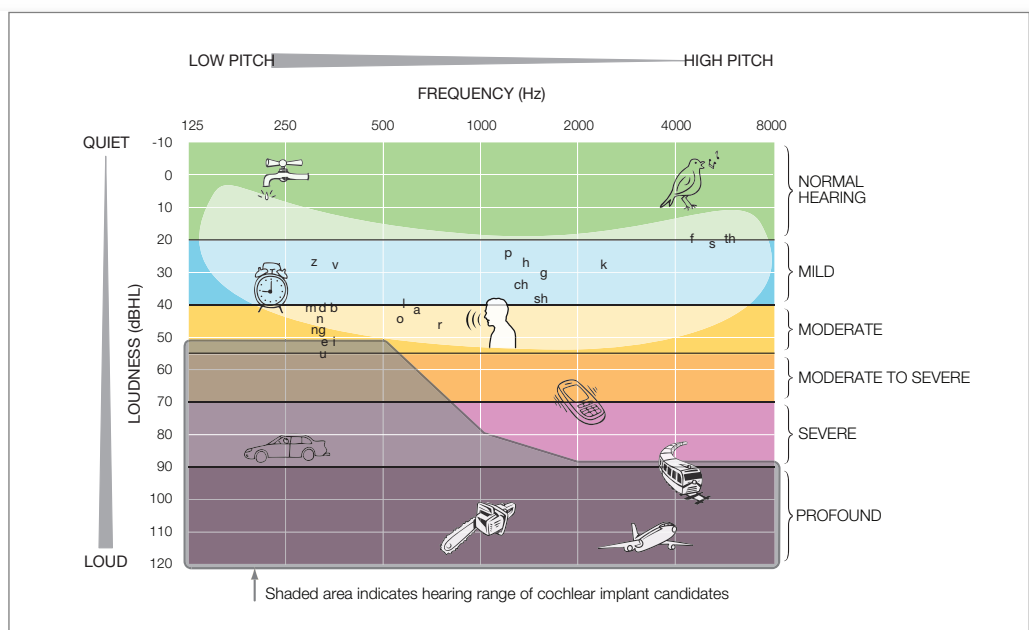
## HOW WE GO DEAF

Most causes of acquired hearing loss involving the sensorineural parts of the hearing pathway are due to hair cell lesions of the inner ear. The hair cells within the organ of Corti are vulnerable to a variety of toxicities. In some cases of profound hearing loss there may be a single identifiable causative agent that leads to hair cell loss and subsequent deafness, such as exposure to ototoxic agents (e.g. gentamicin, cisplatin) or following inflammation caused by meningitis. More often there are a variety of factors that add together to produce hair cell loss and profound hearing loss or, alternatively, the deafening aetiology is unknown. One common pattern of hearing loss is the effect of ageing (presbycusis) added to a pre-existing, nonageing factor of hearing loss, such as noise exposure with a background genetic susceptibility leading to early hair cell dysfunction.

Sustained stimulation of the higher neural pathways linking the inner ear and the auditory cortex is necessary for development of normal sound and speech perception. Most causes of deafness involve the hair cells within the cochlea,

Figure 2. A schematic diagram of our hearing environment. Patients with extreme hearing losses (in the severe-to-profound range) generally fulfill the indications for cochlear implantation.

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with the higher pathways remaining intact. However, hearing losses that occur very early in life are associated with poor maturation of these pathways, particularly at a cortical level. Hearing loss acquired before the acquisition of cortical speech perception (prelingual) is usually associated with disordered higher pathways and characterised by abnormal speech quality, which in extreme cases can be completely unintelligible. Hearing loss acquired later in life, after the acquisition of speech (post-lingual), is usually associated with well-formed higher pathways and characterised clinically by normal speech quality.

The separation of these two broad categories of hearing loss (prelingual versus postlingual) is an important factor in the selection and counselling of prospective cochlear implantation candidates. In general, candidates with postlingual deafness have predictable and satisfactory outcomes. With prelingual hearing losses, outcomes are more difficult to predict and can vary since they are dependent on many factors, including the age of the patient at implantation. Although the outcomes with regard to speech perception may be limited because cortical perception of speech sounds is poorly developed, the individual's appreciation of environmental sounds allows many adults with prelingual deafness to derive considerable auditory benefit from cochlear implantation. However, the prospective candidate should receive careful counselling to ensure their expectations are realistic.

### HOW COMMON IS HEARING LOSS?

Hearing loss is very common in the Australian adult community. Its prevalence in Australia is 17% of 50-year-olds, 48% of 60-year-olds and 64% of 70-year-olds.<sup>2</sup> It is more common in males at all ages. The majority of the affected population has a mild-to-moderate degree of hearing loss that is highly suitable for assistance with a hearing aid. Severe hearing losses (greater than 70 dB; Figure 2) affect a

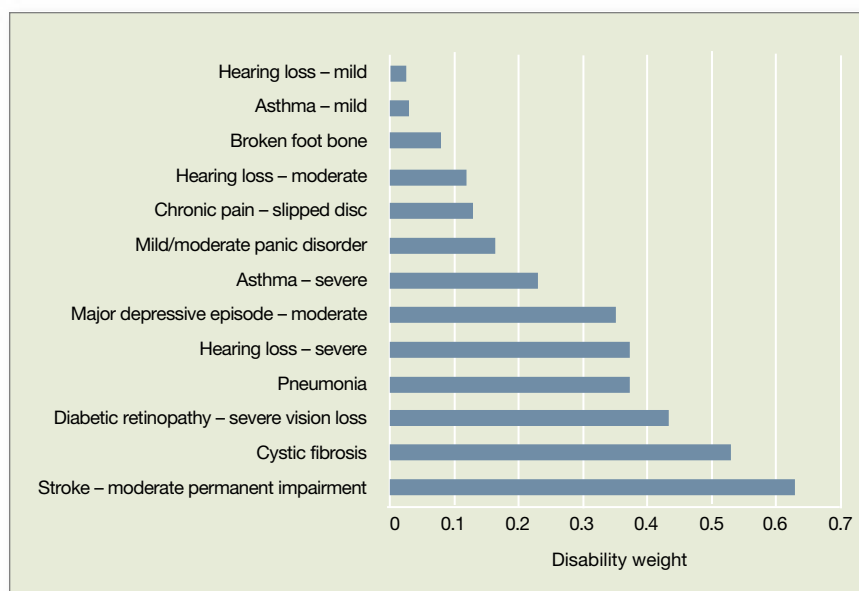


Figure 3. The burden of disease and injury in Australia.

SOURCE: MATHERS ET AL. THE BURDEN OF DISEASE AND INJURY IN AUSTRALIA (1996). CAT. NO. PHE 17. CANBERRA: AUSTRALIAN INSTITUTE OF HEALTH AND WELFARE; 1999.<sup>2</sup>

smaller proportion of the community but carry a greater disability burden. In the elderly, hearing loss is often combined with visual failure and cognitive decline. The impact of this combination of sensory losses on quality of life and independent living can be profound.

In adults of employable age, the impact of hearing loss on potential employment, quality of life and general function is much higher than has been commonly realised, and is equivalent to the disability burden of many health conditions recognised as national health priorities.<sup>3</sup> For example, in terms of disability burden, mild hearing loss is comparable with mild asthma, moderate hearing loss is comparable with severe pain related to degenerative spinal disease (such as a slipped disc), and severe hearing loss is comparable with pneumonia or severe diabetes associated with visual failure (Figure 3).

### HOW A COCHLEAR IMPLANT WORKS

A cochlear implant is an electronic prosthesis that works by bypassing the residual damaged hearing elements within the

organ of Corti and directly stimulating the intact nerve endings in the auditory nerve. The device has two components:

- a fully implanted intracochlear electrode attached to a receiver stimulator package (this is inserted during cochlear implant surgery; Figure 4)
- an external speech processor unit that sits behind the ear and looks like a sophisticated hearing aid (Figure 5).

Both components are required for the cochlear implant system to function properly.

Speech and environmental sounds are captured by the microphone within the speech processor unit. The sounds are then processed within the electronic package to a series of electrical pulses and transmitted to the internal receiver–stimulator unit wirelessly. The internal unit then further processes the signals and sends them down the multiple channels of the intracochlear electrode array, which in turn stimulates the intact auditory nerve endings. These intact auditory nerves carry electrical signals through the higher pathways and eventually the auditory cortex





Figure 4. Diagram of the implanted multichannel intra-cochlear electrode and receiver-stimulator unit of a cochlear implant.

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where they are perceived as speech and sound sensations.

### WHEN TO CONSIDER A COCHLEAR IMPLANT

Current management of hearing loss involves a combination of strategies. Patient education regarding the nature, magnitude and impact of hearing loss is important. There is a focus on useful devices such as hearing aids of various designs used alone and in combination with other assistive technologies, for

example, phone amplification and frequency modulation systems.

A common complaint of those using hearing aids is that although the volume at which sounds and speech can be heard is improved, the clarity of word recognition is still poor. The clarity of speech declines dramatically in noisy listening conditions and higher listening functions, such as music perception, are greatly impaired. When the hearing loss is severe-to-profound, the efficacy of hearing aids fails, telephone usage is severely limited and, unless lip reading skills are developed and maintained, general day-to-day aural communication becomes difficult. It is at this point that a cochlear implant should be considered. A good indication that hearing has declined to the point where a cochlear implant may be useful is when the clarity of speech over the telephone with a maximally adjusted hearing aid and familiar voices is poor.

### INDICATIONS FOR COCHLEAR IMPLANTATION

The fundamental indication for cochlear implantation is bilateral severe-to-profound hearing loss for which well-adjusted hearing aids have given little or no benefit. Establishing these criteria requires a detailed and rigorous assessment

of aidable residual testing. Specially designed tests of speech hearing presented to each aided ear in isolation and both ears together, under a variety of controlled quiet and noisy conditions, provide quantification of residual hearing capacity. This preimplantation data can help predict the hearing outcomes following implantation.

All patients are carefully evaluated with CT and MRI before undergoing cochlear implantation to demonstrate normal cochlear anatomy and identify any factors that may preclude accurate placement of the stimulation electrodes. Potential sources of infection should also be evaluated. Any health factors that might interfere with the anaesthetic or surgery should be identified and managed beforehand. Prospective candidates for cochlear implantation should be well motivated and supported. Candidates need to understand the commitment required for rehabilitation and auditory training once the implant is placed. Advanced age is rarely a limiting factor in cochlear implantation.

### COCHLEAR IMPLANT SURGERY

Surgery to place a cochlear implant has evolved considerably since the first operations were designed 30 years ago. The procedure takes about two hours to perform and recovery usually requires an overnight stay in hospital. It can take four to five days before most patients can return to their usual work and social activities, including driving and work.

The operation involves performing a limited mastoidectomy to access the middle ear cleft and cochlear structures. A small opening measuring about 1 mm in diameter (cochleostomy) is then drilled into the cochlea to allow placement of the intracochlear electrode close to the auditory nerve endings (Figure 6). The implant is tested to confirm its function and correct placement within the cochlea (Figure 7).

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Figure 5. The external speech processor component of a cochlear implant.

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Figure 6. The multichannel intracochlear electrode placed within the cochlear spiral at the time of surgery.

COURTESY OF DR J. THOMAS ROLAND JR, NEW YORK UNIVERSITY COCHLEAR IMPLANT PROGRAM.



Figure 7. Postoperative x-ray of the multichannel electrode placed within the cochlear spiral and part of the attached receiver-stimulator unit placed in a subcutaneous pocket behind the ear.

are fully healed, usually within two to three weeks, the implant device can be switched on and programmed (a process called mapping) to optimise the patient's hearing outcome. Although useful sound and speech perception often occurs at the time of switching on the device, it usually takes many months to reach the best possible hearing outcomes. Persistence with the rehabilitation program is required.

## HEARING OUTCOMES

Many factors determine the hearing outcomes of cochlear implantation. These include the following:

- patient factors – including age, level of cognition and auditory stimulation and persistence with the rehabilitation program
- the nature of the hearing loss – including age of onset (prelingual or postlingual), duration of deafness and degree of residual hearing
- the experience of the cochlear implantation team – including completeness of electrode insertion, type of implant and persistence with the rehabilitation program.

Well-selected candidates with reasonable expectations of what is involved in the cochlear implantation process can achieve highly satisfying outcomes, including benefits to auditory communication within

their work and social environments. Previously employed and socially active adults often return to their usual work and social roles. Elderly patients and their families report a lessened sense of social isolation and the ability to continue living independently and safely as a result of the cochlear implant. Many, but not all, patients can return to fluent listening over the telephone and some to music appreciation. Quality of life, mood and sense of vitality and energy can also be enhanced following cochlear implantation. In general terms, people with cochlear implants that are performing well can hear in quiet environments as effectively as matched patients with aided moderate hearing losses.<sup>3</sup>

## LIMITATIONS

There are limitations to cochlear implantation that are important for patients to understand and accept before embarking on the surgery. Although outcomes are clustered at the higher range of the hearing spectrum, some patients derive only modest benefit for perception of environmental sounds and very limited speech understanding. Occasionally, such patients become nonusers of their implant.

Several of the limiting factors will be known preimplantation. Longstanding deafness (of several decades) and prelingual hearing loss in which the patient's

speech quality is unintelligible are two conditions in which the hearing outcomes are variable. Preimplant counselling is important in these patient groups, with the aims of implantation being awareness of environmental sounds and as an aid to lip reading, rather than speech perception.

Patient support associations such as Cicada ([www.cicada.org.au](http://www.cicada.org.au)) play an important role in informing patients and families of the range of possible outcomes following cochlear implantation. Information about the assessment process, surgery to place the devices and the rehabilitation process following the switching on of the cochlear implant is conveyed by means of a series of informal social events and information sessions. The future candidates have the opportunity to meet previous cochlear implantation recipients and view their outcomes and experiences from both the patient and the family points of view.

## NEW INDICATIONS FOR COCHLEAR IMPLANTS

Given the increasing predictability of the cochlear implant selection and speech perception outcomes, there has been interest in expanding the selection criteria to include candidates with aidable residual hearing, single-sided deafness (with normal hearing on the contralateral



side) and even tinnitus occurring in association with severe hearing loss.

Newer devices that incorporate a fusion of a cochlear implant with a hearing aid have been developed to assist patients with residual audible hearing. As most of these patients have residual hearing in the lower frequencies with severe high-tone hearing losses, the aim is for the cochlear implant to rehabilitate the higher (speech) frequencies while preserving the lower tones. When successful, this combination of 'electro-acoustic' hearing can produce a hearing result with near normal listening experiences; however, the risks of losing functional residual hearing during surgery to place the implant should be accepted.

### THE ROLE OF THE GP

The GP has an important role in recognising potential candidates for cochlear implantation and facilitating their preliminary investigations – particularly basic audiometry and temporal bone imaging. GPs also have a role in the early recognition of implant infections and in the initiation of early treatment and prompt review with the implant surgeon or clinic.

### Recognising potential candidates

Adult patients with severe hearing loss should be considered for a cochlear implant when their hearing aids are optimally adjusted but still give poor hearing experiences in quiet listening environments. In common terms, this level of hearing impairment is reached when the deaf patient is having difficulty understanding conversation over the telephone with familiar voices and subjects. If readjustment of the hearing aids by the hearing aid provider fails to improve the situation then a referral to an ENT surgeon with an interest in cochlear implantation should be considered. A recent audiogram as well as the results of previous hearing tests will be useful in determining the rate of progression of the hearing loss.

### Recognising implant infections

Implant infections are signalled by pain, swelling and discharge over the incision line or implant site. They can occur many months or years following the initial surgery. Fortunately, such infections are becoming infrequent but when they do occur they require prompt recognition and treatment, as an established infection often requires removal of the infected device to achieve treatment resolution. **MT**

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COMPETING INTERESTS: None.

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