Figure 1. Contralateral routing of sound hearing aid.

Figure 2. Soft band bone conduction hearing aid.

Figure 3. Google Glass, incorporating a bone conduction transducer.

Figure 4. Bone conduction hearing implant – percutaneous.

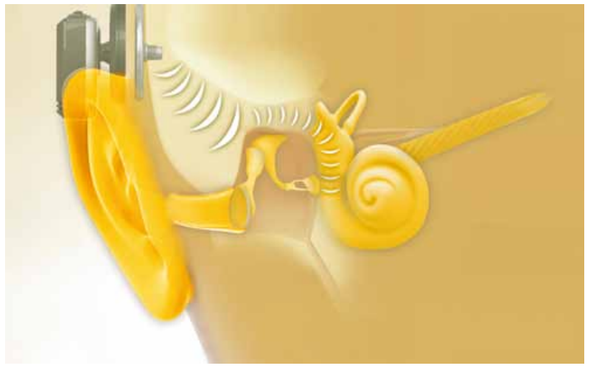

Figure 5. Bone conduction hearing implant – transcutaneous.

The sound of silence

ENT

Single-sided deafness has the potential to dramatically impact patients' quality of life.

DR NICHOLAS JUFAS AND DR NIRMAL PATEL

SINGLE-sided deafness refers to significant or total sensorineural hearing loss unilaterally. There are about 200 new cases of single-sided deafness per million inhabitants in the world and approximately 4000 new cases in Australia annually.[1]

Initial assessment should include full history and examination. Examination should assess the tympanic membrane and include tuning fork testing for sensorineural hearing loss. Referral to an ENT surgeon with an audiogram is required. The more acute the deafness, the more urgent the referral, as the quality of life impact is greater.

The most common causes of single-sided deafness are trauma (head injury, surgery), infections (viral or bacterial), tumours in or around the ear, and sudden hearing loss.

Sudden hearing loss

Around 10% of new cases of single-sided deafness per year will be sudden. Sudden hearing loss occurs rapidly, over a 72-hour period.

The usual audiometric criterion is a decrease in hearing of 30dB or more, affecting at least three consecutive frequencies.[2]

The cause of sensorineural hearing loss is unknown or uncertain in more than 90% of cases. Despite thorough evaluation, treatment decisions are generally made without knowledge of aetiology.[2] The two most common theories of causation are circulatory disturbance and viral infection.[3]

A clinical practice guideline on sensorineural hearing loss was released by the American Academy of Otolaryngology Head and Neck Surgery.[2]

Some of the key points addressed were:

- The importance of clinical and audiometric testing, which should be early and ongoing.
- The need to avoid delay in evaluation and treatment.
- The need to avoid unnecessary and non-targeted laboratory blood tests.
- CT of the head/brain should not be ordered in the initial evaluation as MRI is preferred. This is because up to 10% of those with sudden hearing loss will have a cerebellopontine angle tumour.
- Systemic corticosteroids should be administered orally (1mg/kg/day prednisone for 14 days) if no contraindications or risk factors exist, ideally within the first two weeks and no later than six weeks.
- Intratympanic "salvage" corticosteroid therapy should be commenced for patients who do not recover hearing with systemic steroids. This is generally administered via repeated steroid injection through an intact tympanic membrane or insertion of a grommet for instilling steroids.
- Hyperbaric oxygen therapy may be offered within three months.
- Antivirals, thrombolytics, vasodilators, vasoactive substances or antioxidants should not routinely be prescribed.

Binaural advantages

The Biblical proverb "two are better than one" has a place in otology too. Studies show the benefit of binaural hearing in those with normal hearing, as well as those needing bilateral amplification. Unilateral amplification in the presence of bilateral loss diminishes auditory function.[4]

Binaural hearing provides a number of key advantages, including:

- Squelch effect — a neurological effect whereby brain stem integration of differences in timing, intensity and spectrum of information received binaurally allows for improved perception of sound. Derived from this is the "Cocktail Party" effect, the ability to tune out competing stimuli and tune into a particular one of interest.[5] This is of relevance for those with more significant low frequency hearing loss.[6]
- Summation — a neurological effect whereby the loudness at the same level is greater if heard in two ears rather than in one. The actual effect of this depends on a number of variables, the most important being the loudness level.[6] Given that decibel scale is logarithmic, small increases are important.
- Head shadow — a physical effect most pronounced when speech and noise come from different directions. One ear will have a better signal-to-noise ratio than the other due to the physical acoustic shadow of the head. The effect is more relevant in higher frequencies of speech perception (greater than 1000Hz), as the acoustic head shadow causes greater attenuation.[6]
- Sound localisation — a physical effect following on from head shadow, entailing the ability to localise the direction and distance of the origin of detected sound. This is predominantly achieved through analysis of interaural differences in both time and intensity of perceived sound in each ear. The interaural difference in time is dominant for low frequency sounds (less than 800Hz) and the interaural difference in intensity is dominant for high frequency sounds (greater than 1600Hz).[7]

Rehabilitation options

Contralateral routing of sound and bilateral contralateral routing of sound

Contralateral routing of sound (CROS) hearing aids (figure 1) have been available for decades. Sound directed to the worse ear is recorded by a microphone and transmitted to a speaker in the better ear. Occlusion of the better ear canal and overall poor hearing enhancement is problematic. Despite this, many patients are satisfied with these aids. They remain a first recommendation before patients use other devices.[8]

Bilateral contralateral routing of sound (BiCROS) hearing aids are a variation used when the better hearing ear also has some hearing loss present. Even with minimal hearing loss in the better ear, the benefit obtained appears markedly better than when using CROS aids.[9]

Recent advances that have improved the acceptance rates and overall benefit for CROS/BiCROS aids include digital-signal-processing, more efficient and reliable processors and wireless technologies.

Bone conduction hearing aids and bone conducting hearing implants rely on vibrations transmitted through the skull bones to reach the cochlear.

Bone conduction hearing aids rely on elastic materials to provide tension for the bone oscillator. The most common device carriers commercially available include the head band and soft band (figure 2) models. Glass frame bone conduction hearing aids have also been available for many years and recently this technology has been extended for use in those without hearing impairment, with Google Glass (figure 3) incorporating a bone conduction transducer.

Bone conducting hearing implants involve implantation directly into bone, percutaneously or, more recently, transcutaneously. They rely on the process of osseointegration, a direct functional and structural connection between bone and implant. Feasibility studies have shown equivalence between the two groups of bone conducting hearing implants, but clinical data is too small to show a difference and patient experiences are varied.[10]

The percutaneous models have an abutment that protrudes from the skin, to which receiver/transducer connects directly (figure 4). The transcutaneous models are abutment-free and have the receiver/transducer connect indirectly via magnetic attraction to the implant under the skin (figure 5).

An updated meta-analysis of the application of bone conducting hearing implants in single-sided deafness showed that in the domains of "aversiveness of sound", "reverberation" and "hearing in background noise", bone conducting hearing implants offered significant benefit over CROS. Interestingly, "ease of communication" did not reflect the same benefit.[11]

The studies reviewed were mostly small and not randomised, and only looked at patients who either sought bone conducting hearing implants or failed CROS.[12]

Cochlear implants

Cochlear implants are implanted completely under the skin with up to 22 electrodes inserted into the cochlea (figure 6). Although initially limited, there has been a progressive expansion of the age-range and hearing-loss criteria for these devices.

Single-sided deafness was not initially a recognised indication for cochlear implants, but is now part of the expanded selection criteria.[13,14]

Despite the hearing disadvantages of single-sided deafness, many patients feel the more debilitating aspect of severe, unilateral tinnitus often associated with peripheral deafferentation of neurons. Cochlear implants were serendipitously found to provide partial or total relief of tinnitus initially. This led [**I changed this from 'leads' to 'led' — is that correct???**] to further study on whether these implants could be used as a treatment for severe tinnitus in patients with single-sided deafness. The results so far have shown good efficacy.[15]

Cochlear implants have the advantage of being able to provide real binaural hearing, unlike the pseudo-binaural hearing given by CROS/BiCROS and bone conduction hearing aids/bone conducting hearing implants. Cochlear implants also have the advantage of not interfering with speech understanding and sound perception in the normal hearing ear.[13]

A recent systematic review of the evidence for cochlear implants in single-sided deafness found they provided improved sound localisation and speech perception.[14]

Future directions

In Australia, options for treatment of single-sided deafness are widely available, but not funded by government. Cost-utility analyses show clear benefits for aiding patients totally without hearing, but the quality of life and cost benefits in aiding patients with single-sided deafness are less clear.

Further data affirming these benefits would need to be obtained, or costs would need to decrease, to make these options viable here. In the meantime, for motivated patients who are in a position to pursue treatment options for single-sided deafness, many options exist and can be trialled in a logical, stepwise manner with very favourable results. ●

Dr Jufas is otology/neurotology fellow at the Kolling Deafness Research Centre, University of Sydney and Macquarie University, NSW. Dr Patel is associate professor of surgery, Macquarie University, clinical senior lecturer at the University of Sydney, and director of the Kolling Deafness Research Centre.

Figure 6. Cochlear implant.

SINGLE-SIDED DEAFNESS – KEY POINTS

- Diagnosis — normal tympanic membrane, tuning fork testing and audiogram.
- MRI required to exclude central causes of hearing loss.
- Sudden deafness — quick referral, start oral steroids (if no contraindication).
- Intratympanic steroids may have benefit.
- Rehabilitation options include versions of hearing aids, bone aids, cochlear implantation.

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