Sudden Hearing Loss

Treating this rare but potentially devastating condition

by Dr Annabelle Leong

Sudden hearing loss is a rare but potentially devastating condition which involves an acute unexplained hearing loss of at least 30 decibels (dB) over three audiogram frequencies occurring at a 72-hour period. Many patients present with a sensation of a blocked ear and do not recognise that they have lost hearing. In actuality, the phenomenon of sudden hearing loss is sensorineural in nature. The true incidence of Sudden Sensorineural Hearing Loss (SSNHL) is uncertain with the incidence estimated at 2 to 20 per 100,000 people per year. SSNHL can occur at any age, but most commonly affects patients in their 40s and 50s, with equal gender distribution. Most patients have unilateral hearing loss, but up to 3% of the cases may be bilateral.

The aetiology of most cases of SSNHL is unknown. The possible causes of SSNHL are listed in **table 1.** The prognosis of SSNHL depend on several factors. Young age, male gender, mild severity of hearing loss, lack of associated vertigo, and U-shaped audiogram curve are associated with better prognosis and recovery. In one cohort study, 54.5% of the patients who recovered at least partially showed improvement within 10 days, and only one patient showed any recovery beyond six months.

Impact on Quality of Life

SSNHL has a profound impact on the quality of life, which is further impaired if there is dizziness and/or tinnitus. For patients with unilateral hearing loss, difficulty in localising sound may increase the risk of accidents. These patients also experience difficulty communicating with others in a noisy environment and in group conversations. This can affect their performance at work. Approximately, 90% of patients with unilateral SSNHL experience tinnitus. SSNHL is difficult to treat, thus increasing its psychological burden.

In a study conducted on patients hospitalised in Taiwan for the treatment of SSNHL, the risk of stroke over a five-year follow-up period was found to be higher compared to patients of similar age and demographics. However,

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Table 1.* Main Causes of SSNHL

Infections
Viral cochleitis associated with e.g. herpes viruses, parainfluenza virus, mumps, measles, rubella, or HIV Bacterial meningitis Mycoplasma pneumoniae infection Lyme disease Syphilis infection
Ototoxic Drugs
Aminoglycosides, vancomycin, loop diuretics, antimalarials, cisplatin, sildenafil (Viagra), cocaine
Neoplasms
Vestibular schwannoma Meningeal carcinomatosis Lymphoma Leukaemia
Trauma
Autoimmune Disease
Autoimmune inner ear disease e.g. Cogan's syndrome, systemic lupus erythematosus, antiphospholipid antibody syndrome, rheumatoid arthritis Vasculitides e.g. Wegener's granulomatosis, temporal arteritis, or primary central nervous system vasculititis
Vascular Disorder
Vertebrobasilar cerebrovascular accident; cerebellar infarction; inner ear hemorrhage
Other Causes
Meniere's disease; Paget's disease; multiple sclerosis; sarcoidosis

**This list is not exhaustive. Most cases of sudden sensorineural hearing loss are idiopathic.*

in a literature review, the committee developing 2012 guidelines for the American Academy of Otolaryngology-Head and Neck Surgery (AAO-HNS) concluded that the relationship between SSNHL and risk of stroke did not meet their threshold for significance.

Diagnosis

Patients who complain of sudden hearing loss should be evaluated to determine if the hearing deficit is due to a conductive or sensorineural problem. In the clinic, the Weber and Rinne tuning fork tests can help distinguish between sensorineural and conductive loss. Patients should be asked about a history of trauma, otalgia, ear discharge, fever, focal neurologic symptoms, headache, diplopia, recent eye pain, or erythema. A history of fluctuating hearing loss may point to Meniere's disease or auto-immune inner ear disease.

Physical examination should not reveal signs of otitis media or obstruction in the ear canal. Otoscopic examination is performed to exclude otitis media, foreign bodies, tympanic membrane perforations, otitis externa, or cholesteatoma. Any impacted cerumen should be removed and the hearing loss evaluated afterwards.

A neurological examination should be carried out to exclude stroke in the territory of the Anterior Inferior Cerebellar Artery (AICA), which feeds the internal auditory artery, as a cause of the sudden hearing loss. AICA occlusion may manifest as ipsilateral Horner syndrome, diplopia, nystagmus, facial weakness, limb clumsiness, ataxia, and contralateral loss of pain or temperature sensation.

All patients with sudden SSNHL should undergo audiometric evaluation and Magnetic Resonance Imaging (MRI) scanning with gadolinium, even if it spontaneously recovers. Accurate audiometric evaluation is necessary to support the initial diagnosis and to follow up changes over time. Laboratory studies should be tailored according to the history and physical examination findings that suggest a specific aetiology that warrants diagnostic studies. For example, Lyme titres may be indicated in patients with a history suggesting exposure in an endemic area.

Many patients with SSNHL are likely to have a viral, auto-immune, or microvascular aetiology, which will not be identifiable by MRI. However, other causes of unilateral hearing loss, such as vestibular schwannoma, perilymphatic fistula, vascular insufficiency, multiple sclerosis, or other conditions involving the central nervous system may only be discovered on MRI scan. In a series of 556 patients, 17 patients

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(3%) had a retrocochlear tumour (schwannoma, neurofibroma, or undifferentiated carcinoma). Other studies report the rate of vestibular schwannomas to range from 2.7% to 10.2% in patients presenting with sensorineural hearing loss.

Treatment

There is currently no strong evidence to support the efficacy of any treatment modality for patients with SSNHL. Steroids are considered the first-line of therapy for SSNHL and may be administered systemically (usually orally) or locally via intratympanic injection into the middle ear. Intratympanic glucocorticoids are often reserved for use as salvage therapy, i.e. when hearing fails to improve after a trial of systemic therapy. There is also a role for intratympanic steroids when systemic steroid therapy is best avoided, for example, in patients with diabetes or peptic ulceration or immunocompromised states.

Management strategies vary, reflecting the uncertainty of treatment efficacy and the significant rate of spontaneous recovery. One survey found that steroids were employed more often by otolaryngologists than general practitioners who treated patients with SSNHL (98% and 73%, respectively) and used in higher doses by specialists (at least 60mg of prednisone daily).

Oral Steroids

While oral steroids are considered first-line therapy for SSNHL, their benefit remains unclear. Two systematic reviews and a meta-analysis identified only two randomised studies of sufficient quality for analysis, with both studies differing in their conclusions. Wilson *et al* in 1980 is credited with the first double-blind clinical study thus far to evaluate the role of oral steroids in treating SSNHL. This study assessed 67 patients with SSNHL within 10 days of hearing loss at two centres and demonstrated some benefit from glucocorticoid treatment. More patients treated with oral steroids recovered hearing (61% vs. 32%). Patients with mild or moderate hearing loss recovered hearing, regardless of treatment group. Steroid therapy did not appear to improve rates of recovery compared with placebo in 23 patients with profound hearing loss (18% vs. 1%).

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The second study by Cinamon *et al* in 2001 evaluated the efficacy of steroids vs. carbogen inhalation therapy in 41 patients with SSNHL and found no difference in hearing level at six different frequencies between the steroid and placebo-treated patients. Improvement was found in 60% and 63%, respectively, at early follow-up and 80% and 81% at late follow-up. The systematic reviews conclude that the effectiveness of steroids remains unproven. Results of the meta-analysis that pooled data from the two randomised controlled studies showed no difference between treatment groups.

The latest guidelines from AAO-HNS in 2012 suggest that patients with SSNHL be offered treatment with oral steroids promptly. Patients should be counselled about the risks and benefits of steroid treatment. The recommended dose for prednisone is 1mg/kg/day (maximum 60mg) given as a single dose for 10 days to 14 days.

Intratympanic Steroids

Intratympanic (IT) steroids are primarily used as salvage therapy for patient's refractory to initial oral steroids. Dosing regimens for intratympanic steroids include dexamethasone 10mg/mL to 24mg/mL or methylprednisolone 30mg/mL to 40mg/mL, while dosing frequency ranges from continuous infusion to a few times a day through a ventilation tube to several days consecutively or once weekly. Some side effects associated with IT dexamethasone include transient otalgia, dizziness, ear fullness, and tympanic membrane perforation.

A systematic review and meta-analysis of several small, prospective cohort studies and randomised trials found that intratympanic glucocorticoid therapy following failed oral glucocorticoids was associated with a modest, statistically-significant improvement in hearing (13.3 dB). Almost all studies found some benefit, but whether this represents a clinically significant improvement in hearing remains to be seen. A randomised trial compared IT methylprednisolone with oral prednisone in 250 patients and found that within 14 days of onset the IT steroids were just as effective as oral steroids when hearing loss was assessed at two months. More robust evidence from future studies is certainly needed.

In another randomised trial of 88 patients with SSNHL, there was no difference in hearing recovery between those assigned to simultaneous IT and oral dexamethasone vs. oral dexamethasone followed by IT dexamethasone.

Antiviral Agents

Randomised trials have found no superiority of treatment with antiviral medication plus steroids compared with steroids alone. Guidelines from the AAO-HNS suggest not treating with antivirals, citing the lack of evidence for efficacy and some risk of medication side effects.

Other Treatment Options

A myriad of therapeutic modalities have been evaluated, with difficulty comparing results of studies due to varying definitions of recovery and small sample sizes. In a synergistic effect, some effectiveness has been suggested for oral magnesium and zinc. A systematic review of Chinese herbal medicines for SSNHL found significant flaws in all reported trials and no credible evidence to support their use. Agents to improve cochlear blood flow and oxygen delivery have been tried, including carbogen, dextran, mannitol, pentoxifylline, gingko biloba, nifedipine, heparin, carbogen, and prostaglandin E1 but have not proven to be of benefit.

If the hearing does not return, even after treatment, then it should be managed with assistive devices.

Hearing Amplification

The options for hearing amplification in SSNHL, i.e. single-sided deafness, include a standard hearing aid, a bone conduction hearing (CROS hearing aid), and surgically implantable options such as Bone Anchored Hearing Aids (BAHA) and more controversially, cochlear implantation.

A standard hearing aid may be beneficial if large asymmetry in hearing thresholds between both ears is not present. Often, however, there is severe to profound hearing loss in the affected ear with normal hearing in the unaffected ear. In this scenario, a CROS hearing aid may be more useful as the patient wears a transmitter microphone in the affected ear to detect sound input on that side, and this is then routed via a steel wire headband to the receiver in the ear with good hearing. Latest developments for CROS aid devices include Bluetooth wireless technology which is far more comfortable than the traditional steel headband connector.

The commonest surgically implantable option is BAHA which involves the surgical insertion of a percutaneous titanium implant which becomes osseo-integrated and is then able to transmit sound through the patient's skull to the cochlea on the good hearing side. The quality of sound obtained with a BAHA is far superior to that of a CROS hearing aid.

A newer device, the Bonebridge, is transcutaneous so that the actual implant, which includes a floating mass transducer and a magnet fixed to the mastoid bone, is hidden entirely beneath the skin. MG

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