

Sudden hearing loss

Leong, A.C., Fairley, J.W., & Padgham, N.D.

Department of Otolaryngology, William Harvey Hospital, Kennington Road, Willesborough Ashford, Kent, TN24 0LZ

A 38-year-old lady is referred by her GP with a history of unilateral hearing loss for 48 h.

What should you cover in the history?

The definition of sudden hearing loss is widely accepted as a sensorineural hearing loss of 30 dB or more, over at least three contiguous audiometric frequencies, that develops over 3 days or less.¹⁻³ It must either be a new loss occurring in an ear with previously normal hearing or an incremental deterioration in an ear with pre-existing hearing loss. Although conductive hearing loss may also be sudden, with middle ear effusion as the most common cause, the term sudden hearing loss specifically refers to sudden sensorineural hearing loss.^{1,2} Its low incidence (eight per 100 000 persons per year), heterogeneous pathologies and spontaneous recovery (50% of patients) mean that few controlled studies exist.¹⁻³ Comparisons are made difficult by diverse inclusion criteria and outcome measures, leading to a myriad of treatment protocols.¹⁻³ In all, 5–10 % of sudden hearing loss cases are because of identifiable causes, which may be infectious, traumatic, neoplastic, auto-immune, toxic, circulatory, neurologic and metabolic.^{1,3} The overwhelming majority of cases are idiopathic, with labyrinthine viral infection, vascular insult, intracochlear membrane rupture, autoimmune inner ear disease and activation of cochlear nuclear factor *kappa* B (NF κ B) proposed as possible causes.³

The aim of the history is to diagnose potentially treatable causes of sensorineural hearing loss, particularly ototoxicity

- *How severe is the hearing loss?* This varies from mild to total loss, and may occur at a different frequency for each patient.
- *How quickly did the loss occur?* The exact circumstance and characteristic of the onset of hearing loss should be ascertained, as it may have been instantaneous or progressive over several days. Many patients first notice sudden hearing loss upon awakening in the morning, whilst others only realise their hearing loss upon using

the telephone with the affected ear, which may make it difficult to pinpoint the precise instant of onset. Sudden hearing loss may also occur in patients with a pre-existing disorder characterised by sudden fluctuations in hearing, such as Meniere's disease.

- *Was there pre-existing hearing loss?* From acoustic trauma, ageing, or other disease processes.
- *Is the loss unilateral?* The vast majority (96–99%) of sudden hearing loss is unilateral.^{3,4} Causes of bilateral sudden hearing loss include autoimmune disease, syphilis, especially if rapidly progressive, and, as it occurs more commonly in older patients with pre-existing diabetes mellitus and lipid abnormalities, a vascular aetiology.
- *Are there associated aural symptoms?* Vertigo, tinnitus (especially if low-pitched or 'roaring') and aural pressure may occur in idiopathic Meniere's disease and acquired endolymphatic hydrops, which are important differential diagnoses.
- *How relevant is a full medical history?* It is extremely unlikely that sudden hearing loss will be the sole presenting feature of a systemic disease such as diabetes mellitus, hyperlipidaemia and auto-immune disease such as antiphospholipid syndrome, sarcoidosis and multiple sclerosis, and there is no evidence at present to suggest that sudden hearing loss is more common in these conditions than the population at large.³ Anecdotal experience of hypo- and hyperthyroidism, syphilis, renal failure and renal dialysis and disorders of iron metabolism have shown associations with sudden hearing loss.¹ However, sudden hearing loss may also be caused by vertebrobasilar insufficiency, especially in the territory of the anterior inferior cerebellar artery, and a history of nuchal pain, cerebrovascular embolism, or delayed associated neurologic symptoms should be elicited.⁵ A history of malignancy, e.g. leukaemia, or metastases, is significant. Treatment for malignancy can lead to sudden hearing loss, e.g. post-irradiation effects. It has been estimated that \approx 1–2% of sudden hearing loss patients will turn out to have acoustic tumours, and conversely, 3% of acoustic neuromas present with sudden hearing loss.⁶ Contralateral hearing loss has also been noted after acoustic neuroma surgery.

- *Take a drug history.* Enquire about general anaesthesia, aminoglycosides and other antibiotics, platinum-based antineoplastic agents, salicylates and loop diuretics. Ototoxicity is typically associated with bilateral high-frequency sensorineural hearing loss and tinnitus but may be asymmetrical. Hearing loss can be temporary, e.g. that caused by aspirin, but is irreversible with most agents, and may be progressive even after discontinuation. The usual time of onset is often unpredictable, and marked hearing loss can occur after only a single dose.⁷
- *Ask about direct temporal bone trauma, noise trauma or barotrauma.* Perilymphatic fistula may only be considered if sudden hearing loss is closely associated with a well-defined event of trauma, diving, exertion or intense noise exposure.
- *Ask about preceding infection.* Otitis media and upper respiratory tract infections may affect the inner ear, as may meningitis and encephalitis. Herpes zoster, measles, mumps and HIV may also affect the hearing. More uncommon causes include Lyme disease (borreliosis) and a history of tick bites or recent foreign travel may be relevant.

What should you cover on examination?

- *Perform otoscopy* to exclude conductive causes such as impacted wax or otitis media with effusion.
- *Do a fistula test when there is a history of trauma* but its sensitivity and specificity to detect a perilymphatic fistula even with a suggestive history is controversial. Gentle pressure on the tragus or with a pneumatic otoscope may produce dizziness or objective nystagmus in the presence of a perilymphatic fistula.
- *Neurotologic examination* with particular emphasis on cranial nerves and cerebellar signs. Associated cranial nerve palsies or cerebellar abnormalities may indicate the presence of intracranial lesions such as acoustic neuromas and other malignancies, neurosyphilis, multiple sclerosis or vascular insufficiency.

What management should you offer?

Current literature suggests that the sooner a patient is treated, the better the recovery.^{1,2,4} However, the significant rate of spontaneous recovery means that those who received early treatment may well have included the subset who would have recovered spontaneously without treatment.

Good prognosis is associated with youth, male gender and a short history.⁸ Poor prognosis is associated with profound hearing loss, age over 60 years and the presence of vertigo.

At first visit:

- *Do a pure tone audiogram.* Downward sloping audiograms and profound hearing impairment are bad prognostic signs.²
- *Blood tests* to screen for systemic disorders should be directed by clinical history and examination findings. There is a divergence of opinion over the merits of performing a set of 'screening' blood tests in patients with sudden hearing loss.³ Full blood count, acute phase reactants such as ESR, blood chemistry and autoimmune screen may be helpful when autoimmune disease is suspected.¹ Thyroid-stimulating hormone is important to diagnose thyrotoxic hypokalaemia in those who present with associated severe muscle weakness and electrolyte abnormalities.¹ A clotting screen may be indicated in those with a vascular aetiology. Fasting blood glucose and glycosylated haemoglobin (HbA1c) are useful screening tools for diabetes mellitus, particularly in bilateral cases. It may be helpful to measure post-prandial blood glucose in diabetic patients as a prognostic indicator for cochlear dysfunction.⁹ Syphilis serology (VDRL) has a predictive value of 22% in an otologic population with 75% false-positive rate because of such low disease prevalence, hence clinical assessment should dictate the decision to screen for otosyphilis, and perhaps limit it to bilateral sudden hearing loss only.¹ Serologic tests such as for Lyme disease, HIV and various viral titres should be limited to patients with suspect histories or symptomatology.¹⁰
- *Order an MRI of the internal acoustic meati with gadolinium enhancement* to look for vestibular schwannoma. Intralabyrinthine haemorrhage may also be detected. The urgency of obtaining an MRI scan depends on the clinical presentation, with bilateral sudden hearing loss and associated neurological symptoms clearly warranting greater priority.
- *For the minority with an identifiable cause, treatment is specific but not always successful.* Ototoxic medication should be stopped, suspected perilymphatic fistulae should be explored, and zoster may be treated with acyclovir. Vestibular schwannomas may merit surgical removal.

For the majority with idiopathic sudden hearing loss, discuss the risks, benefits and alternatives of treatment with the patient, sharing the uncertainty about the natural history of sudden hearing loss but explaining some of the possible causes. Decide to treat aggressively or not at all. The duration between time of sudden hearing loss onset and presentation to hospital may influence the decision to institute treatment: some clinicians may choose to treat only those who present within 1 week of symptom onset, whilst others will treat up to 30 days of onset.

More than 50 treatment regimens have been tried without good evidence of success, including surgery, cytotoxic agents, ginkgo biloba and magnesium.^{1,3} There is no high quality evidence to support systemic corticosteroid administration due to small studies thus far.¹¹ A randomised controlled trial of oral steroid *versus* placebo showed promising results (73% recovery rate for patients with mid-frequency hearing loss *versus* 38% for the untreated controls) but its methodological quality and subgroup analysis were criticised.¹²

Ultimately, the risk of complications from steroids is very low, and most patients would prefer some form of treatment than none at all, to maximise hearing recovery. Super high-dose steroid therapy of a reducing course over 18 days with an initial dose of 1200 mg hydrocortisone was found to result in better hearing recovery than a similar course starting off at 600 mg, without any serious complications.¹³ Take caution with steroid use if the patient is diabetic, pregnant, elderly, or has a history of peptic ulcer, but no reliable data actually exists about the risks of a short course of oral steroids in these groups.³ However, high-dose corticosteroid therapy is not contra-indicated in diabetic patients with sudden hearing loss, as optimal glycaemic control is still achievable.⁹ Patients should be informed that aseptic necrosis of the femoral head is a very rare, idiosyncratic reaction to the use of steroids that may require total hip replacement.

The type, duration and optimum dose of steroid is unknown, but many clinicians will prescribe steroids at 1 mg/kg/day of prednisolone for 10 days in unilateral cases, and for longer periods up to a month for bilateral cases, or where auto-immune aetiology is suspected.¹ Proton pump inhibitors such as lansoprazole should be prescribed in conjunction with steroids. *Intratympanic steroid administration* has recently been proposed as treatment for refractory sudden hearing loss where systemic steroids have failed^{14,15} but the efficacy and safety of intratympanic steroid therapy is unknown. Common adverse effects include pain, transient vertigo, otitis media and tympanic membrane perforation.

- *Acyclovir does not appear to improve hearing outcome, whether given alone or combined with steroids.*¹⁶
- *'Shotgun' therapy with dextran, histamine, diuretics, steroids, vasodilators, Hypaque and Carbogen* (5% carbon dioxide-95% oxygen) inhalation is certainly no better than spontaneous recovery.¹⁷
- *Hyperbaric oxygen* has been shown to improve hearing in those who present early, but the clinical significance of the level of improvement is unclear, and its routine application is not justified.¹⁸

- *Reassure the patient with unilateral sudden hearing loss that 50% of patients will have complete spontaneous recovery.*³ The true recovery rate may be higher, as many patients with sudden hearing loss who recover spontaneously within a few days probably do not seek medical evaluation. However, in bilateral cases, the recovery rate is reportedly only 37%⁴ and these patients should be counselled accordingly.

- *Arrange to see the patient again.* In our unit, we review patients at intervals of 2 weeks, 3 months, 6 months and 1 year from onset of sudden hearing loss to monitor the degree of recovery, rehabilitate those who do not improve and manage those with delayed symptoms.

At follow-up.

- *Improvement rate at 1–2 weeks after treatment may predict the longterm prognosis for recovery of hearing level in sudden hearing loss.*¹⁹ Pure tone audiometry and speech discrimination scores should be obtained at each follow-up visit to monitor functional hearing.
- *Rehabilitate those patients whose hearing does not improve.* There is poor evidence to support the efficacy of contralateral bone-anchored hearing aids in unilateral acquired sensorineural hearing loss in adults,²⁰ and in most cases no aiding is required. For bilateral cases, hearing aids may be offered when the hearing impairment is mild-to-moderate. Severe-to-profound hearing impairments may be better served with a cochlear implant.
- *Follow up all patients adequately for possible delayed symptoms.* Late otologic complications may occur in nearly 30%, including fluctuating hearing thresholds, Meniere's syndrome, recurrent vertigo without hearing loss and progressive loss after apparent stabilisation of thresholds.²
- *Inform patient that whilst the risk of contralateral sudden hearing loss is low,³ there is a critical need to seek medical help and begin treatment as soon as possible if it occurs.* Recurrence of sudden hearing loss in the same ear after hearing recovery is very rare.²¹ Those with sudden hearing loss in their only hearing ear may be treated the same way as other patients because their outcome appears to be the same, i.e. about 60% will regain functional hearing. This information may be useful to alleviate anxiety and fears.

Information sources

This article is based on evidence from a literature search performed using Entrez Pubmed on 8 March 2007, using search terms 'sudden hearing loss' and 'sensorineural hearing loss'.

Conflict of Interest

None to declare.

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