

Sudden sensorineural hearing loss

This article highlights the importance of recognizing sudden onset sensorineural hearing loss and summarizes the key diagnostic and management points. There is much to learn about its pathogenesis, and more trials are needed to establish evidence-based management.

Sudden onset sensorineural hearing loss is a medical emergency that continues to be poorly understood despite being recognized in the literature since 1944 (De Kleyn, 1944). A commonly used criterion to qualify for this diagnosis is a sensorineural hearing loss over three contiguous pure-tone frequencies of 30 dB or more that develops within 72 hours. The vast majority of cases are unilateral and the estimated annual incidence is 20 per 100 000 persons (Nosrati-Zarenou et al, 2007). A cause for the hearing loss is only identified in up to 10% of cases but 50% of patients will improve spontaneously (Penido et al, 2009).

Aetiology

The vast majority of cases are considered to be idiopathic, however, a range of possible aetiologies exist, including infectious, traumatic, neoplastic, autoimmune, toxic, vascular, neurological and metabolic (*Table 1*). Sensorineural hearing loss may also be a symptom in a number of disease processes. Published work on this topic is confusing, and approaches to diagnosis and management vary widely (Loughran, 2000). This article presents a practical framework for assessment and management of this disorder.

Viral

The herpes virus is considered the commonest infectious cause of sudden onset sensorineural hearing loss, be it as a result of acute infection of the cochlea or reactivation of the latent virus within the spiral ganglion (Van Dishoeck and Biermann, 1957; Rowson and Hinchcliffe, 1975; Wilson, 1986; Jaffe, 1987). Patients with sudden onset sensorineural hearing loss have statistically significantly raised viral titres for seroconversion for both single and multiple viral infections. These include cytomegalovirus, influenza B, mumps, rubella and varicella zoster (Wilson et al, 1983). Furthermore, post-mortem examination of temporal bones in patients

with sudden onset sensorineural hearing loss has shown histopathological evidence of viral-induced damage in the cochlea. Loss of hair cells, atrophy of the tectorial membrane, atrophy of the stria vascularis and neuronal loss were observed (Schuknecht and Donovan, 1986; Okamoto et al, 1994). Penetration of viruses into the inner ear has been demonstrated in animal studies, which provides further evidence for the viral aetiology (Woolf et al, 1985).

Vascular

The cochlea is exceptionally sensitive to ischaemia and as an end organ has no collateral vasculature. Altered blood flow as a result of thromboembolic disease or vasospasm is the most convincing aetiology for sudden onset sensorineural hearing loss. Thromboembolic events secondary to hyperviscosity disorders have been suggested as a reasonable cause for sudden onset sensorineural hearing loss, although there is a lack of evidence from experimental trials. Sudden onset sensorineural hearing loss is not statistically more commonly seen in patients with hyperviscosity disorders than the general population. It has been shown, however, that patients with sickle cell anaemia and Waldenstrom's macroglobulinaemia are at higher risk of sudden onset sensorineural hearing loss (Wells et al, 1977; O'Keefe and Maw, 1991).

Cerebrovascular accidents can be associated with auditory and vestibular symptoms if the anterior inferior cerebellar artery is involved. Interestingly, there have been a number of reports of sudden onset sensorineural hearing loss in patients post cardiopulmonary bypass (Plasse et al, 1981). Evidence of cochlea damage following labyrinthine vessel occlusion has been demonstrated histologically with early intracochlear haemorrhage leading to fibrosis and ossification (Rudack et al, 2006).

Viral infections have been theorized to cause vascular obstruction through the precipitation of haemagglutination, inflammatory oedema or induction of a hypercoagulable state (Jaffe, 1973). Thus, viral- or vascular-induced cochlea ischaemia could be a common final pathway in the development of sudden onset sensorineural hearing loss. However, cochlea ischaemia typically results in cochlea fibrosis and these ossification changes are not seen on magnetic resonance imaging in patients with sudden onset sensorineural hearing loss (Stokroos and Albers, 1996).

Mr BT Stew is Ear, Nose and Throat ST4 Specialty Trainee in the Department of Ear, Nose and Throat Surgery, Royal Glamorgan Hospital, Llantrisant, Pontyclun CF72 8XR, **Mr SJC Fishpool** is Ear, Nose and Throat ST6 Specialty Trainee in the Department of Ear, Nose and Throat Surgery, Princess of Wales Hospital, Bridgend, and **Mr H Williams** is Ear, Nose and Throat Consultant in the Department of Ear, Nose and Throat Surgery, Royal Glamorgan Hospital, Llantrisant

Correspondence to: Mr BT Stew (stewb@doctors.org.uk)

Intracochlear membrane rupture

A number of thin membranes exist within the ear, first to separate the inner ear from the middle ear and second to separate the perilymph from the endolymph within the cochlea. Although intracochlear membrane rupture has been proposed and favoured in the past as a cause of sudden onset sensorineural hearing loss, there remains a lack of robust evidence with only coincidental case reports in the literature (Schuknecht and Donovan, 1986).

Autoimmune

The inner ear is regarded by many as being immunoprivileged, separated by the blood labyrinthine barrier. However, immunoglobulins, specifically immunoglobulin G, have been isolated in perilymph at a fraction of their serum concentrations (Mogi et al, 1984). The presence of lymphocytes and perisaccular tissues in the endolymphatic sac would suggest that this is the likely site for immune processing. Sudden onset sensorineural hearing loss has been reported in Wegener's granulomatosis, rheumatoid arthritis, polyarteritis nodosum, Sjögren's syndrome, Cogan's syndrome, systemic lupus erythematosus, ulcerative colitis and relapsing polychondritis.

Ménière's disease

Sudden onset sensorineural hearing loss may be the first presentation of Ménière's disease and therefore the classic triad of hearing loss, episodic vertigo and tinnitus should be considered (Nosrati-Zarenou et al, 2007). Typically the hearing loss in Ménière's disease is low frequency. Ménière's disease has an unknown aetiology despite autoimmune and vascular theories.

History

Sudden onset sensorineural hearing loss is considered an otological emergency that requires prompt evaluation and referral to the ear, nose and throat department. The assessment of a patient with sudden onset sensorineural hearing loss should be focussed on identifying any reversible or treatable causes. The key points in the history are onset, time course, pre-morbid hearing level, symmetry, associated symptoms and recent activities such as trauma, straining, flying and exposure to loud noise. The most common presentation is unilateral hearing loss upon waking or when attempting to use the affected ear.

Tinnitus occurs in about 80% of patients and vertigo in about 30% (Nosrati-Zarenou et al, 2007). Up to 80% of patients will report a feeling of ear fullness (Sakata and Kato, 2006).

Past medical history may reveal further risk factors for hearing loss and sudden onset sensorineural hearing loss should be considered as a possible first presentation of underlying systemic disease. A full drug history will identify any ototoxic agents.

Examination

A full and complete head and neck examination should be carried out. Attention paid to otoscopy will help rule out middle ear effusions, infections, wax

Table 1. Causes of sudden sensorineural hearing loss

Infectious	Herpes virus (simplex, varicella zoster, cytomegalovirus)
	Measles
	Mumps
	Rubella
	Meningococcal encephalitis
	Encephalitis
	Syphilis
	Toxoplasmosis
Traumatic	Barotrauma
	Temporal bone fracture
	Perilymph fistula
	Ear surgery (stapedectomy)
	Intense noise exposure
Neoplastic	Cerebellopontine angle tumour, e.g. acoustic neuroma, meningioma
	Leukaemia
	Myeloma
Autoimmune	Wegener's granulomatosis
	Rheumatoid arthritis
	Sjögren's syndrome
	Polyarteritis nodosa
	Relapsing polychondritis
	Lupus erythematosus
	Ulcerative colitis
	Cogan's syndrome
	Antiphospholipid syndrome
	Sarcoid
Toxic	Aminoglycoside antibiotics
	Loop diuretics
	Non-steroidal anti-inflammatory drugs
	Salicylates
	Platinum-based chemotherapeutic agents
Circulatory	Hypercoagulable states, e.g. Waldenström's macroglobulinaemia
	Vertebrobasilar insufficiency
	Cardiopulmonary bypass
Neurological	Multiple sclerosis
	Migraine
Metabolic	Diabetes
	Hyperlipidaemia
	Thyrotoxicosis

impaction and cholesteatoma. A thorough neurological examination, including cranial nerves and cerebellar signs, should be documented in all cases (Chau et al, 2010).

Investigations

A pure tone audiogram will quantitatively assess the degree and pattern of sensorineural loss. This can be particularly helpful in predicting the prognosis as an upward-sloping or cupeloid audiogram has been shown to have a better outcome. A magnetic resonance imaging scan with contrast is considered the imaging modality of choice for evaluation of the cochlea and auditory pathway in sudden onset sensorineural hearing loss and should be requested in all cases. Magnetic resonance imaging is superior to computed tomography and is most sensitive at excluding cerebellopontine angle tumours, such as acoustic neuroma, and inner ear anomalies. It can also identify the underlying aetiology in a significant number of cases such as multiple sclerosis or cerebrovascular accident.

Investigation for systemic disorders should be considered in all patients. However, routine laboratory screening is not warranted in the absence of other clinical evidence of a systemic process. Screening for certain infectious agents that could be causative and can be treated, for example syphilis, should be performed when a risk factor for infection is identified. Routine viral screening is not warranted (Chau et al, 2010). Blood tests may include full blood count, erythrocyte sedimentation rate, C-reactive protein, urea and electrolytes, lipid profile, glucose, thyroid function tests, clotting screen, venereal disease research laboratory (VDRL) and autoantibodies.

Treatment

Idiopathic sudden onset sensorineural hearing loss remains a diagnosis of exclusion and therapy follows a polypragmatic approach based on the different theories of its origin. It is accepted that approximately 50% of cases will resolve spontaneously and therefore proof of effectiveness of all types of therapy is still uncertain. Treatment of sudden onset sensorineural hearing loss is widely variable and often regionally specific. Treatment typically includes anti-inflammatory, antiviral and vasoactive drugs (Table 2).

Steroid therapy is regarded as the standard treatment for sudden onset sensorineural hearing loss, despite a systematic review and meta-analysis revealing no evidence of benefit of standard dose therapy over placebo (Conlin and Parnes, 2007a,b). High-dose steroid therapy has shown improved outcomes when compared with standard dose therapy but caution is advised (Aoki et al, 2006). There have been mixed reviews on the use of intratympanic steroids in refractory cases and in those patients unable to take systemic steroids. A literature review demonstrated that intratympanic steroid treat-

ment as primary treatment for sudden sensorineural hearing loss appears equivalent to treatment with high-dose oral prednisone therapy. As salvage therapy, intratympanic steroids offer the potential for some degree of additional hearing recovery, although it remains uncertain if this improvement is clinically significant (Spear and Schwartz, 2011).

Aciclovir and other antiviral drugs have been trialled in the treatment of sudden onset sensorineural hearing loss with no proven benefit. Vasodilators such as papaverine, histamine, nifedipine and procaine have been used in an attempt to improve cochlear blood flow without documented success. A systematic review of vasodilators and vasoactive substances in the treatment of sudden onset sensorineural hearing loss concluded that the effectiveness of vasodilators in the treatment of idiopathic sudden onset sensorineural hearing loss remains unproven (Agarwal and Pothier, 2009). Altering blood viscosity with rheological agents such as low molecular weight dextrans or anticoagulants such as warfarin has not been proven to have any benefit. There is limited evidence of hearing improvement with hyperbaric oxygen, but this therapy is not widely recommended. There are no valid treatment protocols based on well-constructed randomized controlled trials to date.

Prognosis

The prognosis for hearing recovery in idiopathic sudden onset sensorineural hearing loss is influenced by a number of variables, improvement within the first 2 weeks helps to predict long-term outcome (Xenellis et al, 2006). Evidence from case series suggests recovery is influenced by:

1. Age – those over 60 years of age have a worse prognosis
2. Vertigo – is associated with poorer outcomes

Table 2. Treatments

Anti-inflammatory or immunosuppression	Steroids
	Prostacyclin
Antivirals	Aciclovir
	Valaciclovir
Vasodilators	Carbogen
	Papaverine
	Pentoxifylline
Volume expanders or haemodiluters	Hydroxyethyl starch
	Dextran
Calcium antagonists	Nifedipine
Other agents	Iron
	Vitamins
	Procaine
	Hyperbaric oxygen

3. Audiogram – a profound hearing loss and/or downward sloping audiogram is less favourable
4. Presentation – the earlier the presentation the better the outcome.

Recommendations

- Directed history and examination to identify any treatable or reversible causes
- Pure tone audiometry
- Urgent referral to ear, nose and throat in all cases, regardless of presentation time
- A magnetic resonance imaging scan of the internal auditory canal and cerebellopontine angle is warranted in all patients suffering sudden onset sensorineural hearing loss in whom a cause is not identified
- A short course of empirical high-dose oral prednisolone should be considered within 2 weeks of onset assuming there are no contraindications (Schreiber et al, 2010).

Conclusions

Our understanding of sudden onset sensorineural hearing loss is inadequate and further research is required. Current theories regarding the aetiology are contentious, with the establishment of a direct causal link remaining elusive. The most common suspected aetiologies are idiopathic (71%), infectious (12.8%), otological disease (4.7%), vascular (2.8%), neoplastic (2.3%) and other (6.4%) (Chau et al, 2010). There is a clear lack of consensus regarding optimum treatment protocols and owing to the lack of substantial evidence for any single treatment modality, the risks and benefits of each treatment must be discussed with the patient. **BJHM**

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KEY POINTS

- Sudden onset sensorineural hearing loss is an otological emergency.
- The underlying aetiology remains contentious.
- There are no standardized treatment protocols to date.