Sudden onset hearing loss

Causes, investigations and management

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Background
Sudden onset hearing loss (SOHL) has a number of causes, ranging from the simple and reversible to the profound and permanent. The sequelae of a sudden loss of hearing can be significant.

Objective
This article seeks to address the various aetiologies of SOHL, how they can be diagnosed at the earliest opportunity, and outlines the methods of investigation and management.

Discussion
SOHL causes great concern for the patient. It is when there is a 30 dB or greater hearing loss over less than 72 hours. History and examination, with discerning use of investigations, can identify whether the hearing loss is of conductive or sensorineural origin; and those individuals who have a potentially reversible hearing loss that can be addressed quickly and in an appropriate fashion. However, in the majority of cases of sudden sensorineural hearing loss (SSNHL), no cause is identified and it is considered idiopathic SSNHL. In these patients, high dose oral prednisolone may improve hearing outcome, particularly if started early.

Keywords
hearing loss, sudden; hearing loss, sensorineural

Sudden onset hearing loss (SOHL) is a subjective symptom in one or both ears, as perceived by the patient. To objectively determine whether there is a hearing loss, a pure tone audiogram (PTA) must be performed. A clinical assessment can differentiate between conductive hearing loss (CHL) and sensorineural hearing loss (SNHL).

SOHL is an alarming symptom, and potentially a medical emergency, depending on the cause. It is of rapid onset, typically occurring within a few days. There are a number of causes of SOHL, both conductive and sensorineural (*Table 1*). All patients presenting with SOHL require urgent assessment. 'Red flags' associated with SOHL are listed in *Table 2*. The most common type of sudden hearing loss is CHL; a thorough clinical evaluation will help to determine this.

Sudden sensorineural hearing loss (SSNHL) is defined as a hearing loss of 30 dB or more over at least three contiguous frequencies, over a period of 72 hours or less.[1] Hearing loss can range from mild hearing impairment to a total loss of hearing, and may be temporary or permanent. Regardless of the cause and degree of hearing loss, rapid assessment and early treatment is vital.

Assessment

A methodical approach to the assessment of SOHL can help determine the cause.

Conditions affecting the external and middle ear are causes of CHL, while those affecting the inner ear are usually SNHL, but they may not be mutually exclusive. A combination of the two results in mixed hearing loss.

Although there are a number of causes of SSHNL, most cases are idiopathic, and no cause can be identified in 85–90% of cases.[2]

History

Initially ascertain which ear is affected, or if the loss is bilateral. Bilateral SSNHL is extremely rare, but can be caused by autoimmune disease, syphilis, trauma, neoplasia and vascular causes. Clarify that it was a truly sudden hearing loss – a gradual loss may be associated with a pre-existing disorder, such as Ménière's disease. Pre-existing disease in the affected ear may provide clues to the current diagnosis.

Any activity being undertaken at the time of the hearing loss may provide relevant information. Water may precipitate wax impaction. Trauma (physical or acoustic) can result in ossicular discontinuity (CHL), or SNHL in cases of extreme noise exposure or brain damage. Excessive straining might cause a perilymph fistula, however, tinnitus and vertigo would also usually be present. Tinnitus and dizziness are non-specific and may not help to differentiate between CHL and SNHL. Coryzal symptoms, fever, discharge, otalgia

or recurrent ear infections with foul smelling discharge all provide clues to the aetiology of the hearing loss.

The patient's past medical history may include potential causes, such as an autoimmune disease, diabetes mellitus, sarcoidosis or vascular disease. Excess noise exposure or acoustic trauma may be important, especially if the resultant hearing loss occurred at the time of exposure. Previous otological surgery may point to disease recurrence or failure of outcome.

A medication history, paying attention to known ototoxic drugs, such as the aminoglycosides, frusemide, non-steroidal anti-inflammatory drugs (NSAIDs), chemotherapeutic agents (cisplatinum),[3] quinine and salicylates at high doses, is essential. Any relevant family history should also be sought, such as for otosclerosis.

Examination

Examination of the head and neck and regional lymph nodes is indicated. Lymphadenopathy may indicate malignancy or a middle ear infection affecting the facial nerve. Cranial nerve abnormalities may suggest intracranial lesions (such as acoustic neuromas or malignancy) or multiple sclerosis. Otitis externa may be indicated by discharge at the external meatus or pain on moving the pinna. Mastoiditis may cause mastoid tenderness or fluctuance.

Otoscopy should be performed using a systematic approach, starting from the external meatus medially. Foreign bodies, wax, discharge or masses can cause CHL. The drum may have retraction or evidence of a middle ear effusion, or a tympanic membrane perforation (especially if there is a history of trauma or infection).

Tuning fork tests can help to differentiate the type of hearing loss, and can be especially helpful in determining SNHL. A 512 Hz tuning fork provides the most reliable response.[4] The Rinne test is positive when air conduction is better than bone conduction in that ear (normal test), and negative when bone conduction is louder. It has a low sensitivity.[5]

Free field testing is a simple method of testing a patient's hearing, with a reported sensitivity of 90–100% and specificity of 70–87% in adults.[6] The examiner tests the patient's hearing with whispered, conversational and loud voice (indicating higher sound thresholds) while standing 60 cm behind the seated patient. If responses are poor, then the test can be repeated at 15 cm from the patient. Free field testing does not differentiate the type of hearing loss, but can be useful in the consulting room where audiology is not available.

Table 1. Aetiology of SOHL

Outer ear (conductive hearing loss)
Foreign body
Wax
Otitis externa
Other ear canal pathology (eg. exostoses)
Trauma (syringing)
Middle ear (conductive hearing loss)
Otitis media with effusion
Haemotympanum
Ossicular chain discontinuity
Trauma
Barotrauma
Iatrogenic (post-operative)
Tympanic membrane perforation
Cholesteatoma
Inner ear (sensorineural hearing loss)
Idiopathic
Infective: viral/bacterial (human immunodeficiency virus [HIV], cytomegalovirus [CMV], herpes simplex [HSV], mumps, rubella, syphilis)
Noise induced
Trauma (temporal bone fracture)
Ototoxic drugs
Autoimmune (systemic lupus erythematosis [SLE], granulomatosis with polyangiitis [formerly Wegener], Cogan syndrome, relapsing polychondritis, ulcerative colitis)
Tumour (vestibular schwannoma, leukaemia, myeloma)
Vascular (cerebrovascular disease, sickle cell disease)
Perilymphatic fistula
Barotrauma
Neurological (multiple sclerosis, cerebrovascular accident, migraine)
Other (diabetes mellitus, sarcoidosis)
Non-organic hearing loss

Investigations

Investigations may be arranged by the general practitioner or an ear, nose and throat (ENT) specialist, depending on local resources and access. Investigations are especially important if there is a poor initial response to treatment.

Pure tone audiogram is the initial audiological test used to distinguish CHL from SNHL by assessing both air and bone conduction thresholds. A PTA will determine whether there is any hearing loss, the degree and the type of loss. Serial PTAs can be used to determine a response to treatment.

Tympanometry assesses tympanic membrane mobility and middle ear function. Fluid in the middle ear is represented as a 'flat' trace (type B tympanogram), and is consistent with CHL.

Flexible nasoendoscopy (or alternatively mirror examination of the nasopharynx) visualises the postnasal space for possible masses.

Blood tests can establish the cause of SSNHL in some cases, and therefore direct treatment. Blood tests may not need to be done urgently and can be guided by clinical findings (*Table 3*).

Table 2. Red flags associated with SOHL

- Concurrent head trauma
- Neurological signs or symptoms
- Unilateral middle ear effusion (post-nasal space must be examined)

Imaging

Magnetic resonance imaging (MRI) with gadolinium contrast of the internal acoustic meatus and brain is essential in unilateral or asymmetrical SNHL (>15 dB) to exclude a vestibular schwannoma. It may also identify other diagnoses relevant to the SNHL, such as demyelination, typically seen in multiple sclerosis, and small vessel ischaemic changes.

Computed tomography (CT) of the temporal bones can be used in patients with contraindications to MRI. A CT can exclude large acoustic neuromas in SNHL, and can also evaluate the middle ear/ossicular chain in conductive hearing loss. A chest X-ray may be required if there is suspicion of sarcoidosis with mediastinal involvement.

Management

Initial management in the general practice setting

Conductive hearing loss

Many of the causes of CHL, such as wax, foreign bodies or otitis externa, can usually be managed in the general practice setting, with only complicated cases requiring referral.

Acute otitis media with effusion can result in impaired hearing. Referral is indicated for evaluation of the postnasal space (usually via flexible nasendoscopy) if the effusion is unilateral in all cases.

Tympanic membrane perforations (from infection or trauma) usually heal spontaneously within a few days to weeks. Topical antibiotics are not routinely required unless there is an associated infection. Referral to an ENT surgeon for consideration of repair (tympanoplasty) may be required should the perforation not heal.

Idiopathic sensorineural hearing loss

Idiopathic SSNHL management generates much debate. It must be remembered that this is a diagnosis of exclusion. Idiopathic SSNHL affects approximately 20 per 100 000 people per year and accounts for up to 90% of SSNHL. Tuning fork tests are essential in determining SNHL. A history of the patient describing a clear and sudden change in hearing or awakening with a new hearing loss may help the diagnosis, and treatment can be commenced on a clinical basis.

Table 3. Blood tests to consider in the investigation of SSNHL

- Full blood count
- Erythrocyte sedimentation rate (ESR)
- Urea and electrolytes
- Fasting blood glucose
- Fasting cholesterol/triglycerides
- Viral titres (HIV, CMV, HSV, mumps, rubella)
- FTA-abs for syphilis (or VDRL)
- Lyme titres (this is rare and could be considered in patients returning from endemic areas)
- Thyroid function tests/anti-thyroid antibodies
- Angiotensin-converting enzyme (ACE)
- Anti-neutrophil cytoplasmic antibodies (ANCA)
- Antinuclear antibodies (ANA)
- Rheumatoid factor (RF)
- Anti-cyclic citrullinated peptide (anti-CPP)
- Anti-phospholipid antibodies

Indicators of a better prognosis include low frequency hearing loss,[7] a less severe hearing loss at presentation, and early commencement of treatment.[8,9] Interestingly, patients with a more severe hearing loss appear to respond better to steroid therapy.[1] A worse prognosis has been found in patients aged less than 15 years, in patients aged more than 60 years and in the presence of vertigo.[8] Between one- and two-thirds of patients will recover some hearing within 2 weeks of onset.[10] It is also possible that improvements may reflect a degree of spontaneous improvement rather than a true response to therapy. Treatment should be started as early as possible, however, as this most likely improves the chances of recovery.[11] Completing a full diagnostic work-up should not delay treatment.

The mainstay of treatment is early initiation of oral steroids (prednisolone at a dose of 1 mg/kg/day to a maximum of 60 mg/day) unless contraindicated. The use of steroids should be tailored to the individual patient, and any comorbidities or risk factors taken into account and discussed with the patient. Even in diabetic patients, the individual risk-benefit analysis usually favours steroid use, with appropriate monitoring and management of diabetes.

Treatment with oral steroids is usually between 7–14 days, and tapering is not required with shorter courses. Studies have not shown a conclusive benefit,[2,12] however, oral steroids have been recommended in recent guidelines as the standard treatment for idiopathic SSNHL.[11] The guidelines suggest the use of intratympanic steroid injections should this initial therapy fail, as it may offer some hope as a salvage treatment if the response to systemic steroids is poor.[13] Patients who do not respond to oral steroids should be referred early for consideration of intratympanic steroids. There may be a role for hyperbaric oxygen therapy, but the clinical significance of this remains to be seen.[14] There is no evidence for the use of other therapies, such as carbogen, vasodilators or thrombolytics.[15,16]

There may be concern about whether or not to treat SSNHL in the first instance, especially before PTA, as there is conflicting evidence in the literature as to whether or not treatment methods work. It is essential to empower the patient to make an informed choice and to provide reassurance.[17]

We propose a basic management strategy for use in primary care:

- Discuss the likely diagnosis of SSNHL and that the cause may not be found
- Explain that there is some evidence for the use of oral steroids, but their effectiveness is difficult to predict and hearing may return spontaneously. (The authors would, on balance, advocate the use of oral steroids)
- Discuss the risks and potential side effects of steroid use
- Help the patient to come to an informed choice.

Treatment with oral steroids can be commenced early if the clinical evaluation is convincing for SSNHL, with PTA and ENT review after this. We would always recommend discussion with the ENT specialist or department you are referring to, as there is a degree of variability within ENT specialists on how to treat idiopathic SSNHL.[18] However, a United Kingdom survey study found that 98.5% of otolaryngologists would use steroids as part of their initial management.[19] There is no data to support the routine use of antivirals.[11]

The importance of audiological rehabilitation, often with a multidisciplinary team, should not be underestimated in patients who suffer a non-reversible SSNHL.[20]

Further management

The management of SOHL depends on an accurate diagnosis and the expediency of management. Aside from trauma, most cases of SOHL due to a conductive deficit do not need to be treated as an emergency. A foreign body in the external ear canal can be removed within a reasonable time period unless the offending item is toxic, corrosive (batteries), or if the patient is in pain. Interventions aimed at restoring or improving a conductive deficit can be performed following adequate planning.

There are a number of other causes of SSNHL that must be considered, usually following referral to an ENT specialist.

Vestibular schwannoma

Vestibular schwannoma, also known as acoustic neuroma, is a benign lesion that arises from the Schwann cells of the vestibular nerves. It comprises 8% of all intracranial tumours. Neurofibromatosis type 2 comprises 5%, while the other 95% are sporadic. While 95% of patients with vestibular schwannoma suffer hearing loss, SSNHL can be found in 7–20%.[21,22] The reason for imaging patients with SOHL is largely to exclude vestibular schwannoma.[22]

Vascular

There is debate as to the role of known cardiovascular risk factors and their relevance in the pathogenesis of SNHL. Studies have not been able to determine a definite causal effect.[23,24] Patients with diabetes mellitus and coronary heart disease are at a greater risk of developing SSNHL.[25] This risk seems to increase according to the underlying disease severity, and improved control of risk factors may benefit.

Other causes

Sarcoidosis is a rare cause of SSNHL occurring in less than 0.5% of patients with sarcoidosis, who have eighth cranial nerve involvement.[26]

Infections can cause SSNHL. Syphilis is an established cause and early treatment may lead to a better outcome.[27] Generalised systemic viral illnesses have been mooted as a possible risk factor.

SSNHL has been associated with systemic lupus erythematosis and other autoimmune diseases. It has been suggested that screening for antiphospholipid antibodies should be carried out on all patients presenting with SSNHL.[28]

Key points

- SOHL is a worrying symptom for the patient and should be addressed immediately.
- History and examination can guide which of the many possible investigations are relevant for an individual patient.
- Oral prednisolone should usually be started early for idiopathic sudden sensorineural hearing loss.
- Urgent ENT referral is essential for the patient with sudden sensorineural hearing loss, but less so for patients with conductive hearing loss.

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