Onset of sudden sensorineural hearing loss: An update

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Background
Sudden sensorineural hearing loss (SSNHL) can be very confronting for patients.

Objective
This article provides general practitioners, emergency department physicians and other relevant primary care providers with an overview of causes, diagnosis and primary management of patients with SSNHL.

Discussion
The most common cause of SSNHL is idiopathic. SSNHL should be treated as an 'ear emergency' and urgent referral to ear, nose and throat surgery is warranted.

ONE IN SIX AUSTRALIANS have some degree of hearing impairment and this is expected to increase to one in four by 2050 due to an ageing population.[1] New literature has emerged in recent years on the primary management of sudden sensorineural hearing loss (SSNHL), along with a more up-to-date clinical guideline.[2,3] Prompt diagnosis and initiation of primary management for SSNHL is important given the narrow therapeutic window for treatment and has shown a significant correlation with improved patient hearing and quality of life.[4]

Hearing loss can be divided into conductive, sensorineural and mixed hearing loss. Conductive hearing loss occurs due to external or middle ear pathology. Sensorineural hearing loss occurs due to inner ear or retrocochlear pathology; this can be seen in Figure 1.

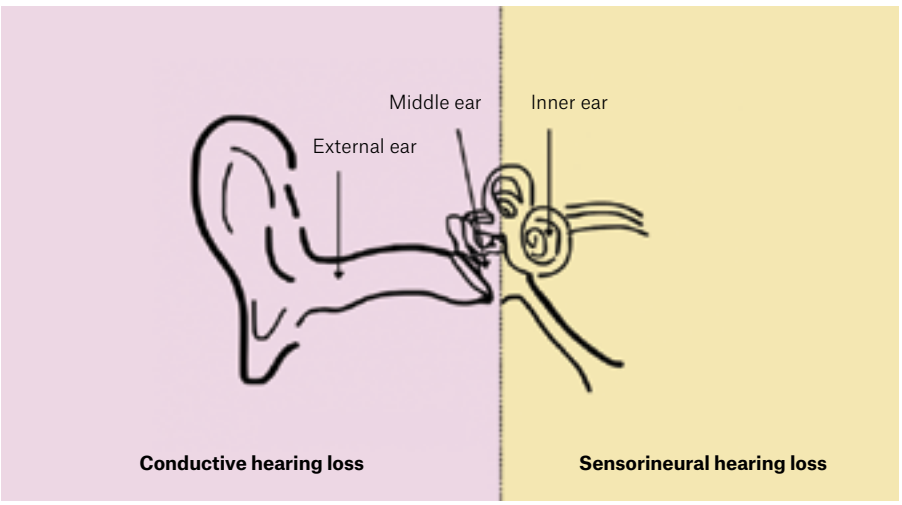

Figure 1. The areas of the ear involved in conductive versus sensorineural hearing loss.

Causes of conductive hearing loss include wax, otitis media with effusion, perforation in the tympanic membrane, foreign body in the external ear canal, ossicular discontinuity or fixation and cholesteatoma.

SSNHL is defined as a decrease in hearing of ≥30 decibels (dB) in three or more consecutive frequencies over 72 hours.[5] This affects approximately 5–20 per 100,000 people per year.[5] The majority of SSNHL presents as unilateral hearing loss, with bilateral hearing loss accounting for 2%.[6]

SSNHL is considered an ear, nose and throat (ENT) emergency. Treatment options for SSNHL are limited and time sensitive. New literature has emerged in recent years on the primary management of SSNHL, along with a more up-to-date clinical guideline.[2,3] These updates include an emphasis on the narrow therapeutic window of treatment for SSNHL,[7,8] the non-utility of routine blood tests,[9] non-utility of CT scans[10] and the importance of patient education on prognosis and multidisciplinary team support and education on the limitations of existing literature.[11]

This article aims to provide general practitioners, emergency department physicians and other relevant primary care providers with an updated overview of primary management of patients with SSNHL.

Aetiology

Overall, 90% of SSNHL is idiopathic.[2] Other causes of SSNHL are shown in Table 1.

Diagnosis

A diagnosis of SSNHL involves history taking, clinical examination and relevant investigations to elicit if the patient has presented with true sudden onset hearing loss and to differentiate between conductive and sensorineural types of hearing loss.

History

There are a number of key questions that should be asked during a history taking session, as noted in Table 2.

Examination

A thorough examination of the ear should be undertaken. This includes examination of the outer ear and otoscopy. On otoscopy, the appearance of the external acoustic canal, middle ear and tympanic membrane should be noted. The presence of copious wax in the external acoustic canal, middle ear effusion and/or abnormalities in the appearance of the tympanic membrane could indicate a conductive aetiology for hearing loss; however, a middle ear effusion can be difficult to correctly diagnose.

The Rinne and Weber tests (Figure 2, Tables 3 and 4) should be conducted with a 512 Hz tuning fork. The 512 Hz tuning fork provides a good balance between sound

Table 1. Causes of sudden sensorineural hearing loss

Idiopathic
Infectious: • HIV • Syphilis • Mumps • Rubella • Cytomegalovirus
Acoustic neuroma
Autoimmune: • Systemic lupus erythematosus • Wegener's granulomatosis • Sjogren's syndrome
Hypothyroidism
Stroke
Ototoxic medications: • Aminoglycosides • Nonsteroidal anti-inflammatories • Loop diuretics • Chemotherapy drugs
Migraine
Diabetes
Physical trauma

Table 2. Key questions to ask during history taking

Which ear is affected?	• SSNHL is most commonly unilateral • Bilateral SSNHL is rare and associated with ototoxic medications, autoimmune and vascular conditions[12]
Duration since onset of symptoms?	Sudden onset versus longstanding hearing loss
What was the patient doing at the time of symptom onset?	• A recent cough or cold could indicate hearing loss secondary to otitis media with effusion • Trauma can result in conductive hearing loss from ossicular chain disruption or sensorineural hearing loss from labyrinthine haemorrhage or perilymphatic fistulae[3,13]
Associated symptoms: • Tinnitus • Vertigo • Otalgia • Otorrhoea • Aural fullness	• Tinnitus and vertigo commonly accompany SSNHL • Otalgia and otorrhoea can help differentiate SSNHL and conductive hearing loss. These might indicate the presence of external or middle ear pathology such as otitis externa or otitis media
Family history	For example, family history of autoimmune conditions (eg otosclerosis)
Past medical history	Diabetes, hypothyroidism, autoimmune conditions, migraine and hyperlipidaemia might be relevant in directing further investigations for SSNHL
Medication history	Ototoxic medications

SSNHL, sudden sensorineural hearing loss.

Table 3. A guide to interpreting Rinne and Weber tests

	Rinne	Weber
Normal hearing	Air conduction > Bone conduction	Sound lateralises to midline
Sensorineural hearing loss	Air conduction > Bone conduction	Sound lateralises to contralateral ear (better ear)
Conductive hearing loss	Bone conduction > Air conduction	Sound lateralises to ipsilateral ear (worse ear)

>, greater than.

duration (decay) and minimising unwanted vibrations that could be felt rather than heard.

Focal neurological examination should be conducted to elicit any deficits indicating central nervous system involvement. These clinical examination findings are an extremely important way to establish a diagnosis in a timely manner and are especially important when audiological testing is not available on the same day.

Initial investigation

Pure tone audiometry should be arranged as soon as possible to elicit the degree of hearing loss and distinguish conductive hearing loss from SSNHL.[10] This should not, however, delay treatment for a SSNHL in the first instance.

Management

Prompt recognition of SSNHL is essential as treatments are time critical, with delays linked to poorer patient outcomes and permanent hearing loss.[2,4,14] A 2023 study by Chen et al found that initiating treatment within 14 days of symptom onset was associated with a 20% improvement in hearing.[4] Furthermore, the efficacy of interventions decreased significantly after 14 days of symptom onset.[4]

Oral corticosteroids (prednisolone) are widely regarded as first-line treatment and should be commenced as soon as possible (preferably on the same day) for suspected SSNHL diagnosed within two weeks of symptom onset. This should be prescribed at a dose of 1 mg/kg/day to a maximum of 70 mg daily for seven days. After seven days, this should be weaned to cease over seven days.[2,15] Evidence on the efficacy of oral corticosteroids is mixed;[16–18] however, it remains one of the few treatments with any data supporting its efficacy for SSNHL. The use of oral corticosteroids largely originated from a 1980 study that found that 61% of patients treated with oral corticosteroids had a significant hearing improvement relative to 32% of patients treated with a placebo.[19] Much literature has emerged on the urgency of commencing treatment for SSNHL as soon as possible given the limited therapeutic window.[7,8] Recent literature by Klein et al found a correlation between earlier initiation of oral prednisolone (within three days of symptom onset) and better patient outcomes.[7]

Oral corticosteroids might be contraindicated in patients with poorly controlled diabetes, peptic ulcer disease, tuberculosis or poorly controlled hypertension. These patients should be referred urgently to an ENT specialist for consideration of a trial of intra-tympanic dexamethasone. [2,20]

Patients with confirmed SSNHL on pure tone audiometry should be urgently referred to ENT. The following could be considered: hyperbaric oxygen therapy and/or intratympanic dexamethasone injections. Much like the use of oral corticosteroids, these treatments are time critical. Hyperbaric oxygen therapy can be used in conjunction with oral corticosteroids within the first two weeks – one month of symptom onset. In the absence of a satisfactory return in hearing after the use of oral corticosteroids, intratympanic dexamethasone can be trialled within 2–6 weeks of the onset of SSNHL.[2,21] Intratympanic dexamethasone is also considered a first line in patients with contraindications to oral steroids. Although these two treatments can be considered,

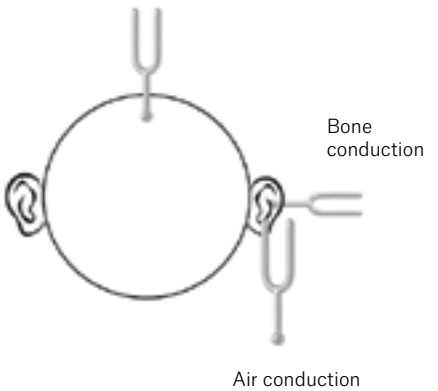

Figure 2. A diagram of Rinne and Weber tests.

Table 4. Example tapering regimen for oral prednisolone

Day 1–7	70 mg oral prednisolone (based on a dosage of 1 mg/kg oral prednisolone (up to a maximum of 70 mg)
Day 8–9	50 mg
Day 10–11	30 mg
Day 12–13	10 mg
Day 14	5 mg
Day 15	Cease all dosages

patients should also be informed that research on their efficacy is still ongoing, and the potential risks and benefits must be carefully evaluated. Additionally, hyperbaric oxygen therapy is not widely available, and accessibility might pose an issue for patients in regional areas. Thus, this might not be recommended for patients in regional areas who would need to travel long distances to access treatment at a tertiary hospital. Antivirals, thrombolytics, vasodilators or vasoactive substances should not be routinely prescribed to patients with SSNHL.[2]

Further investigations in confirmed SSNHL cases

Magnetic resonance imaging internal acoustic meatus (MRI IAM) should be requested to rule out any retrocochlear pathology. New studies have emerged on the use of early MRI IAM in SSNHL.[22] A 2020 study by Kim et al found that performing an MRI early after the onset of SSNHL was associated with higher detection of inner ear abnormalities.[23] CT scans are not indicated in SSNHL and should only be performed where patients cannot tolerate an MRI.[2,10]

Blood tests for further work-up can include full blood count (FBC), erythrocyte sedimentation rate (ESR), autoimmune bloods (anti-neutrophil cytoplasmic antibodies [ANCA], rheumatoid factor [RF], anti-cyclic citrullinated peptide [Anti-CCP], anti-phospholipid antibodies), fasting blood glucose, thyroid function tests (TFT), lipid profile and viral titres.[3] Although these were previously recommended in the work-up of SSNHL,[3] new evidence suggests that these have limited benefit in the diagnosis and management[2,9,15,24] of SSNHL. A study by Heman-Ackah et al found that blood tests routinely ordered in the diagnostic work-up of SSNHL had low diagnostic impacts and high costs.[9] Clinicians should be guided by the clinical scenario of the patient at hand when requesting such tests.

Repeat pure-tone audiometry should be conducted when the course of oral prednisolone is complete to elicit if any improvement to patient hearing has occurred.[10,15]

These have been summarised in Figure 3.

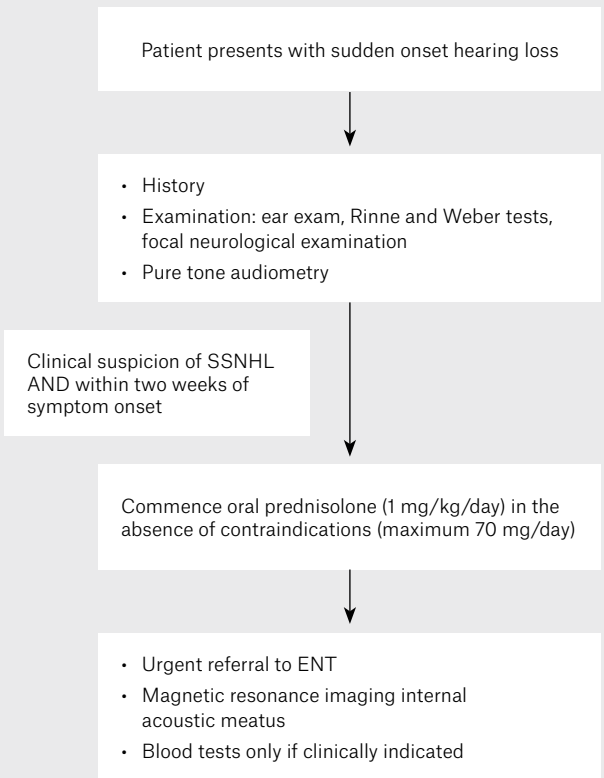

Figure 3. Flow chart on the diagnosis and management of sudden sensorineural hearing loss (SSNHL).

ENT, ear, nose and throat.

Prognosis

Overall, 32–65% of patients with SSNHL recover without treatment within two weeks from symptom onset.[25] Long-term hearing loss, tinnitus and vertigo might persist and have been associated with a poorer quality of life.[11,26] It is important to have an honest discussion on the prognosis of SSNHL with the patient. A study by Chen et al found a correlation between tinnitus and higher depressive thoughts in patients with SSNHL.[26] A multidisciplinary team approach is important in the long-term management of these patients.

In patients who have no or a partial recovery in their hearing, hearing aids might prove beneficial. Patients with persisting vertigo should be referred for vestibular physiotherapy. Patients with tinnitus should be referred to audiology for tinnitus management and advice.

Conclusion

SSNHL requires prompt treatment and diagnosis given the time-critical therapeutic window. If there is clinical suspicion of SSNHL, an urgent audiogram should be performed, and oral prednisolone therapy should begin promptly once SSNHL is confirmed. Patients diagnosed with SSNHL via pure-tone audiometry should receive an urgent referral to an ENT specialist to exclude inner ear or retrocochlear pathology and discuss additional treatment options such as hyperbaric oxygen therapy or intratympanic dexamethasone. MRI IAM should also be ordered to exclude retrocochlear pathology.

Key points

- SSNHL should be treated as an 'ear emergency' and urgent referral to an ENT specialist is warranted.
- 90% of SSNHL is idiopathic.
- SSNHL has a narrow therapeutic window for treatment.
- Prompt diagnosis and initiation of primary management for SSNHL is important.
- High-dose oral prednisolone at a dose of 1 mg/kg (maximum dose of 70 mg) should be commenced for all patients with SSNHL unless contraindicated.

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References

1. Parliament of Australia. Chapter 2: The extent and causes of hearing impairment in Australia. Parliament of Australia, 2010. Available at www.aph.gov.au/parliamentary_business/committees/senate/community_affairs/completed_inquiries/2008-10/hearing_health/report/c02 [Accessed 2 March 2024].
2. Chandrasekhar SS, Tsai Do BS, Schwartz SR, et al. Clinical practice guideline: Sudden hearing loss (Update). Otolaryngol Head Neck Surg 2019;161 Suppl 1:S1–45. doi: 10.1177/0194599819859885.
3. Foden N, Mehta N, Joseph T. Sudden onset hearing loss - causes, investigations and management. Aust Fam Physician 2013;42(9):641–44.
4. Chen I, Eligal S, Menahem O, et al. Time from sudden sensory neural hearing loss to treatment as a prognostic factor. Front Neurol 2023;14:1158955. doi: 10.3389/fneur.2023.1158955.
5. National Guideline Centre (UK). Hearing loss in adults: Assessment and management (NICE Guideline, No. 98.) 11, Sudden sensorineural hearing loss (SSNHL). National Institute for Health and Care Excellence (NICE), 2018. Available at www.ncbi.nlm.nih.gov/books/NBK536521/ [Accessed 4 March 2024].
6. Kuhn M, Heman-Ackah SE, Shaikh JA, Roehm PC. Sudden sensorineural hearing loss: A review of diagnosis, treatment, and prognosis. Trends Amplif 2011;15(3):91–105. doi: 10.1177/1084713811408349.
7. Klein L, Handzel O, Shilo S, et al. Is sudden sensorineural hearing loss an otologic emergency? Evidence-based cutoff for optimal treatment initiation for sudden unilateral sensorineural hearing loss: A case series and meta-analyses. Otol Neurotol 2023;44(3):216–22. doi: 10.1097/MAO.0000000000003808.
8. Battaglia A, Lualhati A, Lin H, Burchette R, Cueva R. A prospective, multi-centered study of the treatment of idiopathic sudden sensorineural hearing loss with combination therapy versus high-dose prednisone alone: A 139 patient follow-up. Otol Neurotol 2014;35(6):1091–98. doi: 10.1097/MAO.0000000000000450.
9. Heman-Ackah SE, Jabbour N, Huang TC. Asymmetric sudden sensorineural hearing loss: Is all this testing necessary? J Otolaryngol Head Neck Surg 2010;39(5):486–90.
10. Herrera M, García Berrocal JR, García Arumí A, Lavilla MJ, Plaza G; Grupo de Trabajo de la Comisión de Audiología de la SEORL. Update on consensus on diagnosis and treatment of idiopathic sudden sensorineural hearing loss. Acta Otorrinolaringol Esp (Engl Ed) 2019;70(5):290–300. doi: 10.1016/j.otoeng.2018.04.007.
11. Carlsson PI, Hall M, Lind KJ, Danermark B. Quality of life, psychosocial consequences, and audiological rehabilitation after sudden sensorineural hearing loss. Int J Audiol 2011;50(2):139–44. doi: 10.3109/14992027.2010.533705.
12. Sara SA, Teh BM, Friedland P. Bilateral sudden sensorineural hearing loss: Review. J Laryngol Otol;128 Suppl 1:S8–15. doi: 10.1017/S002221511300306X.
13. Maillot O, Attyé A, Boyer E, et al. Post traumatic deafness: A pictorial review of CT and MRI findings. Insights Imaging 2016;7(3):341–50. doi: 10.1007/s13244-016-0490-9.
14. Penido Nde O, Ramos HV, Barros FA, Cruz OL, Toledo RN. Clinical, etiological and progression factors of hearing in sudden deafness. Braz J Otorhinolaryngol 2005;71(5):633–38. doi: 10.1016/s1808-8694(15)31268-4.
15. Twigg V, Lawrence R, Fergie TR, Daniel M. Management of suspected unilateral idiopathic sudden sensorineural hearing loss in adults. National Institute on Deafness and Other Communication Disorders, 2018. Available at www.nidcd.nih.gov/health/sudden-deafness [Accessed 4 March 2024].
16. Skarżyńska MB, Kołodziejak A, Gos E, Sanfis MD, Skarżyński PH. Effectiveness of various treatments for sudden sensorineural hearing loss - A retrospective study. Life (Basel) 2022;12(1):96. doi: 10.3390/life12010096.
17. Conlin AE, Parnes LS. Treatment of sudden sensorineural hearing loss: I. A systematic review. Arch Otolaryngol Head Neck Surg 2007;133(6):573–81. doi: 10.1001/archotol.133.6.573.
18. Wei BPC, Stathopoulos D, O'Leary S. Steroids for idiopathic sudden sensorineural hearing loss. Cochrane Database Syst Rev 2013;2013(7):CD003998. doi: 10.1002/14651858.CD003998.pub3.
19. Wilson WR, Byl FM, Laird N. The efficacy of steroids in the treatment of idiopathic sudden hearing loss. A double-blind clinical study. Arch Otolaryngol 1980;106(12):772–76. doi: 10.1001/archotol.1980.00790360050013.
20. Zhao D, Tong B, Wang Q, Hellstrom S, Duan M. A comparison of effects of systemic and intratympanic steroid therapies for sudden sensorineural hearing loss: A meta-analysis. J Otol 2016;11(1):18–23. doi: 10.1016/j.joto.2016.02.002.
21. Wu HP, Chou YF, Yu SH, Wang CP, Hsu CJ, Chen PR. Intratympanic steroid injections as a salvage treatment for sudden sensorineural hearing loss: A randomized, double-blind, placebo-controlled study. Otol Neurotol 2011;32(5):774–79. doi: 10.1097/MAO.0b013e31821fbdd1.
22. Conte G, Di Berardino F, Zanetti D, et al. Early magnetic resonance imaging for patients with idiopathic sudden sensorineural hearing loss in an emergency setting. Otol Neurotol 2019;40(9):1139–47. doi: 10.1097/MAO.0000000000002389.
23. Kim MB, Lim J, Moon IJ. Anatomical and pathological findings of magnetic resonance imaging in idiopathic sudden sensorineural hearing loss. J Audiol Otol 2020;24(4):198–203. doi: 10.7874/jao.2020.00157.
24. Singh A, Kumar Irugu DV. Sudden sensorineural hearing loss - A contemporary review of management issues. J Otol 2020;15(2):67–73. doi: 10.1016/j.joto.2019.07.001.
25. Mattox DE, Simmons FB. Natural history of sudden sensorineural hearing loss. Ann Otol Rhinol Laryngol 1977;86(4 Pt 1):463–80. doi: 10.1177/000348947708600406.
26. Chen J, Liang J, Ou J, Cai W. Mental health in adults with sudden sensorineural hearing loss: An assessment of depressive symptoms and its correlates. J Psychosom Res 2013;75(1):72–74. doi: 10.1016/j.jpsychores.2013.03.006.

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