

Sudden Sensorineural Hearing Loss in Adults

Collaborative Multi-Institutional Otolaryngology
Residency Education Program

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Sudden Hearing Loss

- Rapid onset of subjective sensation of hearing impairment in one or both ears
- Conductive hearing loss (CHL) – primary care or ER physician/APP
- Sensorineural hearing loss (SNHL)
- Mixed hearing loss (CHL and SNHL) in the same ear

Sudden Sensorineural Hearing loss

Idiopathic Sudden Sensorineural Hearing Loss
(ISSNHL)

- Incidence in the US:
- 5-20 cases per 100,000 people annually
 - Likely underestimated due to spontaneous recovery, people not seen
- 66,000 new cases per year
- Often associated with tinnitus
- Seen initially in Urgicare, ER, or primary care clinics

Sudden Sensorineural Hearing Loss

- Occurs within a 72-hour time window
- Decrease in ≥ 30 dB
- Affecting at least 3 consecutive frequencies
 - Often defined in relation to the opposite ear
- These definitions may not be stringently adhered to
- Can be associated with tinnitus and/or vertigo

Idiopathic Sudden Sensorineural Hearing Loss

- Defined as Sudden Sensorineural Hearing Loss (SSNHL) with no identifiable cause
- 90% of people with SSNHL have ISSNHL
- Look for underlying disease for non-idiopathic cause of SSNHL include vestibular schwannoma, stroke, malignancy, noise, head trauma and ototoxic medications
- History of fluctuating HL, bilateral onset, autoimmune diseases, infections - viral (herpes, HIV); bacterial; mycoplasma, Lyme

Idiopathic Sudden Sensorineural Hearing Loss

Proposed Etiology

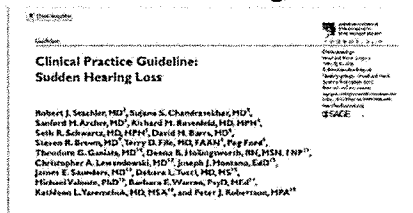
- Direct viral invasion into the cochlea or 8th nerve
- Reactivation of latent virus in the spiral ganglia
- Immune mediated mechanisms from systemic infection
- Cochlea ischemia – hemorrhage, embolism, vasospasm
- Cochlear membrane rupture

Sudden Sensorineural Hearing Loss

Support for viral etiology
 Post mortem histopathologic studies in 12 patients with history of SSNHL
 Atrophy of organ of Corti and tectorial membrane similar to known cases of viral labyrinthitis (mumps, rubella)
 No cochlear membrane rupture or vascular occlusion

Schuknecht, Donovan 1986

Clinical Practice Guideline: Sudden Hearing Loss



Based on a systematic review of the literature
 Otolaryngology-Head and Neck Surgery 2012, Vol 146 (IS) S1-S35

Clinical Practice Guideline: Sudden Hearing Loss Update

Guideline Update Group

Disciplines:

Otolaryngology-head and neck surgery, otology, neurotology, family medicine, audiology, emergency medicine, neurology, radiology, advanced practicing nursing, and consumer advocacy. Previous 2012

Based on a systematic review of the literature
 Otolaryngology-Head and Neck Surgery 2019, Vol 16(IS) S1-S45

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Guidelines

Clinical Practice Guideline: Sudden Hearing Loss (Update)

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 SAGE

Clinical Practice Guideline: Sudden Hearing Loss Update

Review generates Key Action Statements (KAS)
 Strength of Action Terms

Strong Recommendation – can be positive or negative
 Recommendation - can be positive or negative
 Option – quality of evidence is suspect or good evidence does not favor one approach versus another

Idiopathic Sudden Sensorineural Hearing Loss

Factors affecting the prognosis for recovery

Patient age
 Vertigo at onset
 Degree of hearing loss
 Configuration pattern of the audiogram
 Time between onset of hearing loss and intervention

Idiopathic Sudden Sensorineural Hearing Loss

Poor prognosis for recovery

Profound hearing loss or down sloping, with vertigo, and prolonged time since onset

Idiopathic Sudden Sensorineural Hearing Loss

Potential Treatment Options

Systemic and intratympanic Steroid
Hyperbaric Oxygen Therapy (HBOT)
Antiviral agents
Rheologic agents
Diuretics
Herbal and other complementary and alternative treatments
Middle ear surgery (fistula repair)
Observation

Idiopathic Sudden Sensorineural Hearing Loss

Recommendation FOR

- Retrocochlear testing
- ABR - patient preference, will miss 20% (8-42%) IAC lesions, severe hearing loss 4KHz
- MRI - gold standard; T1 with contrast
 - Patients with SSNHL have a CPA tumor in 2.7 - 10.2%
 - 10-20% patients with VS can present with some degree of SSNHL
 - Can potentially avoid contrast by obtaining heavily weighted T2 images
 - FIESTA or CISS
 - Other central pathology - labyrinthine hemorrhage, cochlear inflammation, arachnoid cyst, demyelination,

Idiopathic Sudden Sensorineural Hearing Loss

• Strong recommendation FOR

- Obtaining an audiogram - ASAP but within 2 weeks of symptom onset
 - Confirms the type of HL (CHL; MHL: SNHL)
 - OAEs and ABR are useful in patients with suspected non-organic (functional) hearing loss
- Patient Education - shared decision making
 - Diagnosis - possible causes
 - Evaluation and treatment options

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• Strong Recommendation AGAINST

- Obtaining a CT scan - unless there are focal neurologic signs, trauma
 - Potential use in patients with retained metal or pacemaker, severe claustrophobia, or having bone disease like Paget's or possible metastasis
- Routine "shotgun" Laboratory Testing
 - Can have false positive or negative results
 - Unless specific concerns like Lyme in endemic areas - positive serology patients had better recovery - antibiotic treatment, steroids, natural recovery?
 - Autoimmune screen-negative for antithrombin III, protein C, fibrinogen
 - Association between elevated cholesterol and poorer hearing recovery

Idiopathic Sudden Sensorineural Hearing Loss

• Strong Recommendation AGAINST

Use of:

- Antivirals
- Thrombolytics - aspirin
- Vasodilators - calcium antagonists, ginkgo biloba
- Vasoactive substances - prostaglandin E1
- Rheologic agents - pentoxifylline (Trental), dextran

Frequently asked questions	Table 8. Hearing Loss: Causes and Treatment
What is causing the problem?	Question What is causing the problem? Answer The loss of hearing is usually due to damage to the cochlea, the part of the inner ear that contains the hair cells. The hair cells are responsible for converting sound waves into electrical signals that the brain can understand. If the hair cells are damaged, the brain will not receive the signals and hearing will be lost. The most common cause of hearing loss is age-related hearing loss, which is caused by the natural wear and tear of the hair cells over time. Other causes include noise-induced hearing loss, otitis media with effusion, and congenital hearing loss.
How is sudden hearing loss diagnosed?	Question How is sudden hearing loss diagnosed? Answer Sudden hearing loss is diagnosed by a hearing test. The hearing test will show a sudden drop in hearing in one or both ears. The hearing test will also show that the hearing loss is not due to a conductive hearing loss, such as ear wax or a middle ear infection. The hearing test will also show that the hearing loss is not due to a mixed hearing loss, which is a combination of conductive and sensorineural hearing loss.
Will my hearing come back?	Question Will my hearing come back? Answer The hearing may or may not come back. The hearing may come back if the hearing loss is due to a conductive hearing loss, such as ear wax or a middle ear infection. The hearing may also come back if the hearing loss is due to a mixed hearing loss. However, if the hearing loss is due to a sensorineural hearing loss, the hearing will not come back. The hearing loss is permanent.
What additional testing is needed?	Question What additional testing is needed? Answer The hearing test is the first step in diagnosing sudden hearing loss. If the hearing test shows a sudden drop in hearing, the doctor will order additional tests. These tests include an audiogram, an otoscopy, and a tympanometry. The audiogram will show the hearing loss in more detail. The otoscopy will look for any physical changes in the ear. The tympanometry will measure the pressure in the middle ear.
How is sudden hearing loss treated?	Question How is sudden hearing loss treated? Answer The treatment for sudden hearing loss depends on the cause of the hearing loss. If the hearing loss is due to a conductive hearing loss, the treatment will be to remove the cause of the hearing loss. For example, if the hearing loss is due to ear wax, the wax will be removed. If the hearing loss is due to a middle ear infection, the infection will be treated with antibiotics. If the hearing loss is due to a mixed hearing loss, the treatment will be to treat both the conductive and sensorineural components. If the hearing loss is due to a sensorineural hearing loss, the treatment will be to use hearing aids or a cochlear implant.
What are the side effect of each treatment?	Question What are the side effect of each treatment? Answer The side effects of treatment for sudden hearing loss depend on the treatment. The side effects of hearing aids include tinnitus, dizziness, and discomfort. The side effects of a cochlear implant include infection, dizziness, and tinnitus. The side effects of antibiotics include diarrhea, nausea, and allergic reactions. The side effects of steroids include weight gain, mood changes, and increased blood sugar.
What else can I expect?	Question What else can I expect? Answer The prognosis for sudden hearing loss depends on the cause of the hearing loss. If the hearing loss is due to a conductive hearing loss, the hearing will usually come back. If the hearing loss is due to a mixed hearing loss, the hearing may come back. However, if the hearing loss is due to a sensorineural hearing loss, the hearing will not come back. The hearing loss is permanent.

Idiopathic Sudden Sensorineural Hearing Loss

Option for treatment

- Suggest instituting within 2 weeks
 - Systemic glucocorticoids (oral, IV, IM) and/or Intratympanic
 - Inner ear sites of action affect viral, vascular, syphilitic, autoimmune, and hydrops
- 3 Trials of RCT of oral vs. placebo – 2 showed no benefit, 1 had improvement; inconclusive
- 1 RCT IT steroids vs. placebo – patients with moderate HL
 - better early recovery with early steroids

Idiopathic Sudden Sensorineural Hearing Loss

Option for treatment

- Studies comparing IT to oral steroids
 - No benefit – Crane (2015)
 - IT steroids alone or in combination with oral steroids did much better than oral steroids alone (Battaglia 2008)
 - Multicenter RCT (16 centers) – oral compared to IT steroids has similar outcome at 2 months (Rauch 2011)
 - IT and oral steroids started within 7 days of onset had 20dB PTA and 30% WRS improvement compared to those treated after 7 days (Battaglia 2014)

Idiopathic Sudden Sensorineural Hearing Loss

Protocols for Treatment

Oral Steroids

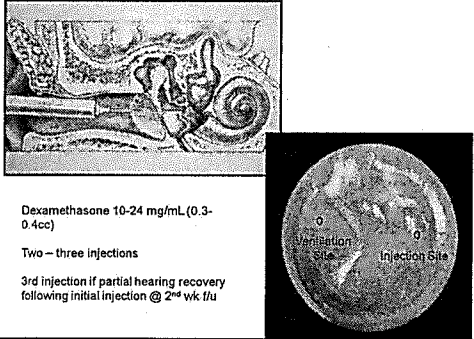
- Prednisone 60mg/day for 4-10 days with taper beginning at day 4, 5, 7, or 10 for total of 10-17 days
- Equivalent 48 mg methylprednisolone
- Equivalent 10 mg dexamethasone
- Caution: diabetes, labile hypertension, glaucoma, TB, stomach ulcers, psych problems

Idiopathic Sudden Sensorineural Hearing Loss

Protocols for Treatment

IT Steroids

- Good option for those who cannot take oral steroids
- Dexamethasone (4 to 24 mg/ml)
- Methylprednisolone >30mg/ml
- Patients receiving 24mg/ml had 53% get >30dB improvement compared to 17% getting 10 mg/ml (Alexander 2015)
- Possible harm – transient dizziness, TM perforation, syncope (vasovagal), infection
- Patient supine, involved side up, 15-30 minutes, not to swallow
- 3-4 injections over 2-4 weeks, can terminate early if no improvement



Dexamethasone 10-24 mg/mL (0.3-0.4cc)

Two – three injections

3rd injection if partial hearing recovery following initial injection @ 2nd wk. 1fu



Idiopathic Sudden Sensorineural Hearing Loss

Hyperbaric Oxygen

- Vascular compromise cochlear ischemia, not FDA approved 100% oxygen at 2.4 ATA (atmospheres absolute), 90-120 minutes each, 10-20 sessions
- Underseas and Hyperbaric Medical Society in 2011 approved HBO with steroids within 2 weeks onset SSNHL
- Hardships – limited centers, ETD, claustrophobia, sporadic insurance coverage (cost)

Idiopathic Sudden Sensorineural Hearing Loss

Hyperbaric Oxygen

Oral steroids	19%	HBOT alone	17.5%
IT Steroids	16%	HBOT and oral steroids	42.6%
Full hearing recovery			

(Alimoglu 2011)

Idiopathic Sudden Sensorineural Hearing Loss

Hyperbaric Oxygen

Option

- Initial Therapy - combined with steroids within 2 weeks of onset SSNHL
- Salvage Therapy – between 2- 4 weeks of onset SSNHL
- No benefits noted beyond 3 months

Idiopathic Sudden Sensorineural Hearing Loss

• Recommendation FOR

- IT Steroids for Salvage Therapy 2-6 weeks after onset SSNHL
- Meta-analysis 11 trials – IT steroids had better outcome when used as salvage treatment (Garavello 2012)

Idiopathic Sudden Sensorineural Hearing Loss

- CPG indicates oral or IT steroids are an *option* in management of acute SSNHL
- CPG indicates IT steroids are *recommended* in salvage treatment after oral steroids, HBO or observation provided incomplete hearing recovery from SSNHL 2-6 weeks after onset.
- Most frequent means of delivery is with transtympanic needle injection or via myringotomy with tube
- Many studies show the benefit of IT salvage treatment

Idiopathic Sudden Sensorineural Hearing Loss

Siegel's Recovery

Pre-treatment Hearing Groups

1. <25dB
2. 26 – 45dB
3. 46 – 70dB
4. 71 – 90dB
5. >90dB

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Siegel Hearing Recovery Criteria

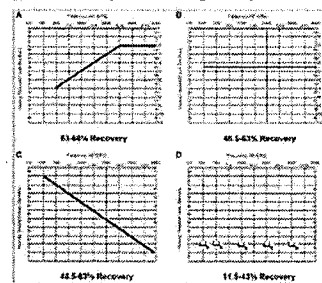
Type	Hearing recovery
I. Complete recovery	Final hearing better than 25 dB
II. Partial recovery	More than 15 dB gain, final hearing 25–45 dB
III. Slight improvement	More than 15 dB gain, final hearing poorer than 45 dB
IV. No Improvement	Less than 15 dB gain, final hearing poorer than 75 dB

Idiopathic Sudden Sensorineural Hearing Loss

Outcomes

Natural recovery rate 32-65%
 Overall response rate 50-70% including partial return
 Hearing recovery occurs within 1 month in 90% and with 3 months in 98% (Yeo 2007)

Hearing Recovery following idiopathic SSNHL



Kuhn, Maggie. Sudden Hearing Loss Trends in Hearing 2011

Idiopathic Sudden Sensorineural Hearing Loss

Meta-analysis review; 7 randomized trials, 710 patients
 IT steroids – complete recovery 24%
 Systemic steroids – complete recovery 25%
 Combined systemic and IT – complete recovery 30%
 Conclusion: No difference in recovery comparing IT, systemic or combination therapy

Mirian, C JAMA March 2020

Idiopathic Sudden Sensorineural Hearing Loss

Conclusions

The earlier the presentation better chance of recovery
 Confirm it is SNHL, no other suggestive history
 No laboratory studies
 Obtain imaging – MRI IAC w/ (exceptions - CT, ABR)
 Institute therapy – IT and/or systemic steroids, HBO?
 Salvage therapy – IT steroids (2-6 weeks) or HBO?
 I usually set the window at 4-6 weeks post onset