SUDDEN HEARING LOSS

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HEARING LOSS FACTS

- Men are more likely to experience hearing loss than women.
- Approximately 17 percent (36 million) of American adults report some form hearing loss.
- About 2 to 3 out of every 1,000 children in the United States are born deaf or hard-of-hearing.
  - Nine out of every 10 children who are born deaf are born to parents who can hear.
HEARING LOSS FACTS

- The NIDCD estimates that ~15 percent (26 million) of Americans between the ages of 20 and 69 have **high frequency** hearing loss due to exposure to loud sounds or noise at work or in leisure activities.

- Only **1 out of 5** people who could benefit from a hearing aid actually wears one.

- Three out of 4 children experience ear infection (otitis media) by the time they are 3 years old.
Hearing Loss Facts

- Strong relationship between age and reported hearing loss:
  - 18 percent of American adults 45-64 years old, 30 percent of adults 65-74 years old, and 47 percent of adults 75 years old or older have a hearing impairment.

- Roughly 25 million Americans have experienced tinnitus.

- Approximately 4,000 new cases of sudden deafness occur each year in the United States.
HEARING LOSS FACTS

- Approximately 615,000 individuals have been diagnosed with Ménière's disease in the United States. Another 45,500 are newly diagnosed each year.

- One out of every 100,000 individuals per year develops an acoustic neuroma (vestibular schwannoma).
AUDIODEGRAM

Red Line = Right Ear
Blue Line = Left Ear
TYPES OF HEARING LOSS

1. **Sensorineural** Hearing Loss (nerve loss)
2. **Conductive** Hearing Loss
3. **Mixed** Hearing Loss (both nerve and conductive loss)
CONDUCTIVE HEARING LOSS

External Ear Canal
Cerumen impaction, foreign body

Tympanic Membrane
Perforation, tympanosclerosis, Hematoma

Middle Ear
Otitis Media with Effusion, Cholesteatoma

Ossicles
Otosclerosis
SENSORINEURAL HEARING LOSS
ETIOLOGIES

1. Infectious:
   ▪ Meningitis, Herpes virus, HIV, Mumps, Rubella, Rubeola, Mycoplasma, Toxoplasmosis, Syphilis, Lyme disease

2. Autoimmune
   ▪ Lupus erythematosus, Cogan’s syndrome, Wegener’s granulomatosis

3. Traumatic
   ▪ Perilymph fistula, T-bone fracture, Acute blast injury
SNHL ETIOLOGIES

4. Vascular
   - Vertebrobasilar insufficiency (VBI), Sickle cell disease, Hyperviscosity syndromes, Waldenstrom’s macroglobulinemia, Polycythemia vera, thrombocytopenia

5. Neurologic
   - Multiple sclerosis, Migraine

6. Neoplastic
   - Acoustic neuroma, Meningioma, Metastasis, Leukemia, Myeloma
SNHL ETIOLOGIES

7. Iatrogenic
   ♦ Ototoxic Meds, Otologic surgery

8. Congenital
   ♦ Hereditary, Toxic, Infectious, Spontaneous

9. Toxic
   ♦ Chronic noise

10. Idiopathic Sudden SNHL
SNHL: HISTORY

- Onset (sudden vs. progressive)
- Duration
- Fluctuations in hearing
- Associated symptoms:
  - tinnitus, vertigo, imbalance, aural fullness
- h/o otologic surgery or recurrent AOM, head trauma, vascular disease, autoimmune disease
- Family history of hearing loss
SNHL: PHYSICAL EXAM

- Full head and neck exam
- Otoscopic exam
  - Pneumatic otoscopy / Tympanometry
- Tuning forks (Weber & Rinne)
- Cranial nerve exam
- Cerebellar exam/Balance testing as appropriate
SNHL: PHYSICAL EXAM

WEBER TEST: typically with a 512 Hz tuning fork

- Normal = sound heard centrally or in both ears
- unilateral SNHL should lateralize to better hearing ear,
- unilateral CHL should lateralize to diseased ear
**SNHL: PHYSICAL EXAM**

**Rinne Test:** compare air conduction (AC) and bone conduction (BC); place tuning fork ≤ 1 cm of the EAC (AC) and then place on the mastoid (BC);

- **Positive** = SNHL (GOOD)
  - tuning fork heard better by AC = normal or most SNHL

- **Negative** – CHL
  - BC is perceived louder than AC = CHL > 15–30 dB HL or severe to profound SNHL with cross-over

**Normal:** AC > BC


**Table 4. Recommended Technique for Weber and Rinne Testing**

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<tr>
<th>Weber Test</th>
<th>Rinne Test</th>
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<tr>
<td>1.  Place vibrating tuning fork (256 or 512 Hz) at midline of forehead or on maxillary teeth (not false teeth)</td>
<td>1.  Place vibrating tuning fork (256 or 512 Hz) over the mastoid bone of one ear, then move the tuning fork to the entrance of the ear canal (not touching the ear)</td>
</tr>
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<td>2.  Ask where the sound is heard; it is normal to hear at the midline or “everywhere”</td>
<td>2.  The sound should be heard better via air conduction (at the entrance to the ear canal).</td>
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</table>
| 3.  If the sound lateralizes to one ear then:  
   a.  There is a CHL in that ear, OR  
   b.  There is SNHL in the opposite ear | 3.  If the sound is heard better by bone conduction, then there is a CHL in that ear.  
Repeat for the other ear:                                                                                                                                 |

Abbreviations: CHL, conductive hearing loss; SNHL, sensorineural hearing loss.
SNHL: WORKUP

- Full audiogram with pure tones, speech recognition, and word recognition
- For sudden sensorineural hearing loss or asymmetric sensorineural hearing loss: MRI + gadolinium
PRESBYACUSIS

- Age-related hearing loss
- 40% U.S. population >75 y/o affected
- Often familial (>50%)
- Bilateral and symmetric
Presbyacusis: Treatment

- Treatment
  - Hearing aids
  - Assistive listening devices
  - Cochlear Implantation
NOISE INDUCED HEARING LOSS (NIHL)

- Most common cause of preventable SNHL
- Most frequently occurs from exposure through years (> 90dB)
- Can result from single exposure to very loud noise (>120-130 dB)
- Typically bilateral and symmetric
3 types noise:
- Continuous
- Impulse (e.g., gun)
- Impact

2 types exposure:
- Chronic
- Acute

2 types damage:
- TTS (temporary)
- PTS (permanent)
CHRONIC NIHL: AUDIOGRAM

Frequency (hertz)

Years Exposure

dB HL

- 1--2
- 5--9
- 15--19
- 25--29
- 35--39
**Ototoxic Medications**

- **Macrolides**
  - High frequency SNHL, tinnitus, vertigo
  - Usually reversible within 2 weeks
  - Unknown mechanism

- **Vancomycin**
  - High freq SNHL progresses to bilateral profound SNHL

- **ASA**
  - Doses > 2700 mg/day
  - Affects stria vascularis, reversible
OTOTOXIC MEDICATIONS

- **Antineoplastics/cisplatinum**
  - Begins with high freq HL, progresses as total dose accumulates
  - Irreversible when profound deafness occurs
  - Can be vestibulotoxic
  - Affects OHC

- **Loop diuretics/ethacrynic acid**
  - Affects stria vascularis, rarely permanent

- **Phosphodiesterase type 5 inhibitors (Viagra > Levitra, Cialis)**
  - Unknown mechanism; question of nitric oxide effects on ear
PERILYMPHATIC FISTULA (PLF)

- **Definition**: Communication between perilymph space and middle ear/mastoid

- **Etiology**
  - Increased pressure/trauma → communication → Decreased perilymph volume → 2\textsuperscript{nd}ary endolymphatic hydrops → symptoms

- **Potential causes (rare)**:
  - Otologic surgery (stapedectomy)
  - Head trauma
  - SCUBA diving
  - Congenital ear malformation
  - Forced *valsalva* / suppressed sneezing
Acoustic tumors:

- Most common: Acoustic Neuroma (misnomer) = Vestibular Schwanomma
- Usually present with gradually progressive SNHL
- 1% of patients with asymmetric SNHL have acoustic tumors
Clinical Practice Guideline: Sudden Hearing Loss

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Sponsorships or competing interests that may be relevant to content are disclosed at the end of this article.

Abstract

Objective. Sudden hearing loss (SHL) is a frightening symptom that often prompts an urgent or emergent visit to a physician. This guideline provides evidence-based recommendations for the diagnosis, management, and follow-up of patients who present with medical interventions, and the limitations of existing evidence regarding efficacy; and (3) counsel patients with incomplete recovery of hearing about the possible benefits of amplification and hearing-assistive technology and other supportive measures. The panel made recommendations that clinicians should (1) assess patients with presumptive SSNHL for bilateral SHL, recurrent episodes of SHL, or focal neurologic findings; (2) diagnose presumptive ISSNHL if audiometry confirms a 30-dB hearing loss at 3 consecutive frequencies and an underlying condition cannot...
### CLINICAL PRACTICE GUIDELINE SUMMARY

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IDIOPATHIC SUDDEN SENSORINEURAL HEARING LOSS (ISSNHL)

- Theories:
  - Viral
  - Autoimmune (autoimmune inner ear disease – AIED)
  - Vascular
  - Intracochlear membrane breaks
ISSNHL: VIRAL

- Current belief – viral cochleitis causes the majority of cases of ISSNHL
- 1983 – Wilson and colleagues
  - Viral seroconversion rates greater in patients with ISSNHL (63%) compared to control (40%)
    - Influenza B
    - Mumps
    - Rubeola
    - VZV
ISSNHL: VIRAL

- 1981 - Veltri et al.
  - 65% seroconversion

- 1986 – Schuknecht and Donovan
  - Temporal bone studies (n. 12)
    - ISSNHL vs. cases of known viral labyrinthitis
    - Similar pathologic findings
      - Atrophy of the organ of Corti, tectorial membrane, stria vascularis, cochlear nerve, and vestibular organ
ISSNHL: VIRAL

FIG. 2. Light microscopy of the organ of Corti in experimental herpes simplex virus type 1 labyrinthitis. Extensive destruction of the organ of Corti and supporting cells. The tectorial membrane is disrupted from the sensory cells but has a normal appearance (arrow) (bar = 100 µm).
New - **Sleep apnea** linked to sudden hearing loss

- No causality proven
- Sleep apnea causes major inflammation in the bloodstream/brain promoting vascular complications
- Need for prospective studies, and causality studies with treatment trials (i.e., improved sleep apnea, improved hearing).

ISSNHL: TREATMENT

- 90% of cases will be Idiopathic
- Treat known causes by addressing the underlying condition
Therapy for ISSNHL is **controversial**

- Difficult to study
  - High spontaneous recovery rate
  - Low incidence
  - Makes validation of empiric treatment modalities difficult
ISSNHL: TREATMENT

- Proposed treatment modalities
  - Anti-inflammatory – steroids, cytotoxic agents
    - e.g., *Prednisone* 1mg/kg/day (80mg) PO QD taper over 2 weeks.
  - Diuretics
  - Antiviral agents
General Guidelines for Corticosteroid Therapy for Idiopathic Sudden Sensorineural Hearing Loss (ISSNHL)

| Table 9. General Guidelines for Corticosteroid Therapy for Idiopathic Sudden Sensorineural Hearing Loss (ISSNHL)³ |
|---|---|---|
| **Oral Corticosteroids** | **Intratympanic Corticosteroids** |
| **Timing of treatment** | Immediate, ideally within first 14 days. Benefit has been reported up to 6 weeks following onset of sudden sensorineural hearing loss (SSNHL) | Immediate |
| **Dose** | Prednisone 1 mg/kg/d (usual maximal dose is 60 mg/d) or Methylprednisolone 48 mg/d or Dexamethasone 10 mg/d | Dexamethasone 24 mg/mL or 16 mg/mL (compounded), or 10 mg/mL (stock)Methylprednisolone 40 mg/mL or 30 mg/mL |
| **Duration/frequency** | Full dose for 7 to 14 days, then taper over similar time period | Inject 0.4 to 0.8 mL into middle ear space every 3 to 7 days for a total of 3 to 4 sessions |
| **Technique** | Do not divide doses | Anterosuperior myringotomy after topical anesthetic |
| | | Inject solution into the posterior inferior quadrant via narrow-gauge spinal needle to fill middle ear space |
| | | Keep head in otologic position (one side down, affected ear up) for 15 to 30 minutes |
| **Monitoring** | Audiogram at completion of treatment course and at delayed intervals | Audiogram before each subsequent injection, at completion of treatment course, and at delayed intervals |
| | | Inspect tympanic membrane (TM) to ensure healing at completion of treatment course and at a delayed interval |
| **Modifications** | Medically treat significant adverse drug reactions, such as insomnia Monitor for hyperglycemia, hypertension in susceptible patients | May insert pressure-equalizing tube if planning multiple injections, but this increases risk of TM perforation |
| | | May consider adding round window transport facilitator |

³This table is designed to provide guidance for systemic and intratympanic steroid treatment for SSNHL. Treatment is routinely individualized by provider and per patient. The most important principles pertain to early institution of high enough dosages of treatment. Prednisone 1 mg/kg/d or its equivalent and/or adequate concentration of intratympanic dexamethasone or solumedrol should be administered.

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TREATMENT

- **BAHA (Cochlear / Oticon)**
  - Bone Anchored Hearing “Aid”
  - Surgically implanted pin in the skull, using a vibrating digital hearing amplification device.

- **CROS/BICROS**
  - Contralateral Routing Of Signal
## Table 6. Selected Conditions That May Be Associated with Bilateral Sudden Hearing Loss

<table>
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<tr>
<th>Cause</th>
<th>Other Features</th>
</tr>
</thead>
<tbody>
<tr>
<td>Meningitis (infectious, inflammatory, neoplastic)</td>
<td>Headache, fever, abnormal cerebrospinal fluid (CSF) studies, possibly other cranial nerve palsies(^{211})</td>
</tr>
<tr>
<td>Autoimmune inner ear disease</td>
<td>Fluctuation of hearing may sometimes occur; vertigo may occur in some cases(^{41})</td>
</tr>
<tr>
<td>Lyme disease</td>
<td>Erythema chronicum migrans, abnormal CSF, fluctuating bilateral audiovestibular symptoms(^{212})</td>
</tr>
<tr>
<td>Syphilis</td>
<td>Abnormal fluorescent treponemal antibody absorption (FTA-abs) test, bilateral fluctuating hearing loss, tabes dorsalis, multiorgan involvement(^{213})</td>
</tr>
<tr>
<td>Ototoxic medications</td>
<td>Vestibular loss, oscillopsia(^{214,215})</td>
</tr>
<tr>
<td>Trauma</td>
<td>Significant head trauma, barotrauma, temporal bone fractures(^{214})</td>
</tr>
<tr>
<td>Herpes zoster oticus (Ramsay-Hunt syndrome)</td>
<td>Otalgia, pinna and/or ear canal vesicles, facial nerve paresis, positive viral titers, positive viral cultures(^{216})</td>
</tr>
<tr>
<td>Human immunodeficiency virus (HIV) otitis</td>
<td>Positive HIV titers, altered T cell counts, and often other cranial neuropathies may be associated with mastoiditis out of proportion to clinical complaints(^{217,218})</td>
</tr>
<tr>
<td>Lead poisoning</td>
<td>Learning disabilities, other stigmata of lead poisoning(^{219})</td>
</tr>
<tr>
<td>Genetic disorders</td>
<td>May be syndromic or nonsyndromic(^{220,221})</td>
</tr>
<tr>
<td>MELAS (metabolic encephalopathy, lactic acidosis and stroke-like episodes)</td>
<td>Periods of confusion, elevated serum lactic acid levels around times of attacks, stroke-like spells, magnetic resonance imaging (MRI) white matter signal changes, migraine-like headaches, seizures, diabetes, mitochondrial gene mutation (Mt-RNR1, Mt-TS1, POLG genes)(^{222,223})</td>
</tr>
<tr>
<td>Other mitochondrial disorders</td>
<td>Variable phenotypes(^{224})</td>
</tr>
<tr>
<td>Bilateral synchronous internal auditory artery occlusion associated with vertebrobasilar vascular disease</td>
<td>Vertigo, dysarthria, facial weakness, ataxia, nystagmus, unilateral numbness, abnormal computed tomography or magnetic resonance angiogram of the vertebrobasilar vasculature(^{48,50,225,227})</td>
</tr>
<tr>
<td>Cogan syndrome</td>
<td>Nonsyphilitic interstitial keratitis of the cornea, hearing loss, vertigo(^{49})</td>
</tr>
<tr>
<td>Neoplastic (neurofibromatosis II, bilateral vestibular schwannomas, intravascular lymphomatosis, others)</td>
<td>Abnormal brain MRI or cerebrovascular imaging study(^{228-230})</td>
</tr>
<tr>
<td>Sarcoïdosis</td>
<td>Pulmonary symptoms, bilateral vestibular loss, elevated serum angiotensin-converting enzyme level or abnormal Gallium scan(^{231,232})</td>
</tr>
<tr>
<td>Hyperviscosity syndrome</td>
<td>Mucous membrane bleeding, neurologic and pulmonary symptoms, associated retinopathy(^{233})</td>
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AUTOIMMUNE INNER EAR DISEASE (AIED)

- 1979 – McCabe
  - Described patients with bilateral rapidly-progressive SNHL (BRPSNHL)
  - Proposed the term – **autoimmune inner ear disease (AIED)**
  - Evidence of **autoimmunity**
    - Lymphocyte inhibition test
    - Substantial hearing improvement with steroids
AIED

- Clinical characteristics
  - Middle-aged **females**
  - **BRPSNHL**
  - Absence of systemic immune disease
  - 50% with **dizziness**
  - Light-headedness and ataxia more common than vertigo
  - Episodic – multiple, daily
  - Hearing loss sudden, rapidly progressive, or protracted
“RUSH LIMBAUGH’S severe-to-profound, bilateral, rapidly progressive hearing loss generated considerable public interest in sudden deafness. In his case, its cause was reportedly an autoimmune disease of the cochlea.”

- CNN.com

“FOXY BROWN, real name Inga Marchand, has revealed that she is slowly losing her hearing. She first noticed a problem when her label boss, Jay-Z told her the sound levels on her new record were way too high when she had thought they were perfect.”

- Hip Hop News
**AIED**

**Diagnosis**
- Based on Hearing loss and response to treatment
- Hughes –
  - Lymphocyte transformation test
    - Sensitivity – 50-80%
    - Specificity – 93%
    - Positive predictive value 56-73%
- Western blot
  - Sensitivity – 88%
  - Specificity – 80%
  - Positive predictive value – 92%
1. **Prednisone** 1mg/Kg/day for 4 weeks
2. Slow taper
3. Relapse during taper – restart
4. Slow taper
5. If relapse during taper – **Cytotoxic** agent
   - Methotrexate
   - Cyclophosphamide
   - Monitor electrolytes, LFTs, blood counts
VASCULAR

- **Etiologies** - Embolism, vasospasm, hypercoagulable states/sludging
- **Pathophysiology** – anoxia to vestibulocochlear apparatus
- Cochlea is intolerant to disruption of blood supply
  - 1957 Kimura and Perlman
    - Clamped the labyrinthine artery in guinea pigs
    - Demonstrated irreversible loss of cochlear function after 30 minutes of disruption
VASCULAR ANATOMY

Anterior Inferior Cerebellar Artery
Basilar Artery
Labyrinthine Artery
Common Cochlear Artery
Main Cochlear Artery
Cochlear Ramus
Vestibulocochlear Artery
Anterior Vestibular Artery
Posterior Vestibular Artery
Arteries of the Canals
VASCULAR

- Abnormal **circulatory** states
  - Sickle-cell disease
  - Waldenstrom’s macroglobulinemia
  - Hearing loss is usually reversible with tx
  - AICA strokes
  - Cardiopulmonary bypass
PROGNOSIS

- 47%-63% spontaneously resolve
  - Combined patients with all audiogram types

- **Prognostic** variables:
  1. Time since onset
  2. Audiogram type (severity of hearing loss)
  3. Vertigo
  4. Age
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