

# **ACUTE HEARING LOSS**

## **Background**

1. Definition: Categorized as conductive, sensorineural, or mixed<sup>1,5</sup>
  - Conductive hearing loss occurs when sound cannot travel through the external ear, middle ear, or both
  - Sensorineural hearing loss results from pathology of cochlea, eighth cranial nerve, or central auditory pathways
  - Mixed hearing loss is combination of both conductive and sensorineural
2. General Information: Sudden sensorineural hearing loss (SNHL) is rapid loss of hearing that occurs all at once or over a period of up to 3 days diagnosed as at least a loss of 30 decibels in three connected frequencies<sup>2</sup>

## **Pathophysiology**

1. Pathology of Disease:<sup>3</sup>
  - Infections (viral, acute otitis media, external otitis, syphilis, Lyme disease) causing SNHL from toxins affecting the inner ear through direct viral invasion or latent virus reactivation<sup>13</sup>
  - Immunologic hypothesis based on theory that circulating antibodies cross-react with inner ear antigens or activated T cells and damage the inner ear<sup>13</sup>
  - Hydrops and perilymphatic fistula cause abnormalities of labyrinthine fluid
  - Thrombosis, embolic occlusion, and hyperviscosity state (e.g. polycythemia vera) may result in cochlear ischemia
  - Diabetes mellitus, atherosclerosis, and sickle cell anemia obstruct small vessels
2. Incidence, Prevalence<sup>4</sup>
  - Estimated incidence between 5 and 20 per 100,000 persons per year
  - Typically occurs between 43 years and 53 years of age
  - Equal gender distribution according to several large case series of 7500 cases in United States, Europe and Japan
  - Approximately 1% of cases related to vestibular schwannoma, demyelinating disease or stroke
  - 10 to 15% due to other causes such as Meniere's disease, trauma, autoimmune disease, syphilis, Lyme disease, or perilymphatic fistula
  - Majority are idiopathic
3. Risk Factors<sup>3</sup>
  - Head trauma, noise, and barotrauma
  - Retrocochlear neoplasm in cerebellopontine angle
  - May be presenting symptoms of multiple sclerosis
  - Associated ophthalmologic disease should be investigated for Susac's syndrome (rare immunological disorder characterized by encephalopathy, branched retinal arterial occlusion [BRAO], and hearing loss)
  - Ototoxic medications (aminoglycosides, diuretics, chemotherapy)
  - Conductive (otosclerosis, chronic otitis media with effusion, malleus head fixation, tympanosclerosis, myringosclerosis, tympanic membrane perforation, cholesteatoma, ossicular chain disruption)
  - Autoimmune disease (polyarteritis nodosa, relapsing polychondritis, sarcoid, ulcerative colitis, systemic lupus erythematosus, Wegener's disease, Churg-Strauss syndrome, Behcet's disease, Cogan's syndrome)

4. Morbidity / Mortality
- 15% with Sudden SNHL will have hearing loss that gets worse over time<sup>2</sup>

## **Diagnostics**

1. History
  - Current symptoms/PHx:<sup>3</sup>
    - Unilateral or bilateral
    - Fluctuating or constant
    - Chronology
    - Current and past treatments with oral and intravenous medications, nonprescription drugs
    - Screen for systemic disease
    - Prior ear surgery, cardiac bypass, surgery, and lumbar puncture
    - Family history of hearing loss, neoplasms, renal disease, and balance disorders
    - Previous sharp or blunt head trauma, noise trauma, barotrauma
  - Sudden SNHL symptoms:<sup>4</sup>
    - Immediate rapid hearing loss, or hearing loss upon waking
    - Majority are unilateral
    - Tinnitus and ear fullness
    - Vertigo
    - Ear pain or paresthesia
2. Physical Exam<sup>1,4</sup>
  - Visualize and palpate the auricle
  - Examine external auditory canal and tympanic membrane
  - Evaluate drum mobility and middle ear effusion with pneumatic (air inflation) otoscopy or tympanogram
  - Perform Weber and Rinne tests (512 Hz tuning fork)
  - Softly whisper simple words or numbers and ask patient to repeat
  - Instruct patient to hum and report hearing asymmetry (sound lateralizes to affected ear in conductive and unaffected ear in SNHL)
  - Evaluate cranial nerves
3. Diagnostic Testing
  - Audiometry<sup>1,4</sup>
    - Sensorineural hearing loss: sensitivity to bone-conducted and air-conducted sound stimuli are equally reduced in affected ear (thresholds elevated)
    - Conductive hearing loss: bone conduction normal bilaterally; air-conducted thresholds elevated in affected ear (sensitivity reduced)
  - Speech testing<sup>1,5</sup>
    - Subject given list of words and asked to repeat them
    - Speech reception threshold is sound level at which 50% of spoken words are understood
    - Speech recognition score is percentage of spoken words understood at 40 dB above speech reception threshold
  - Tympanometry<sup>5</sup>
    - Assesses mobility of tympanic membrane and pressure status of middle ear
4. Diagnostic Imaging<sup>4</sup>

- MRI
  - Rule out retrocochlear abnormality (e.g. neoplasm, stroke, or demyelination)
- CT or auditory brain-stem response audiometry
  - Alternatives for patients who cannot have MRI
  - Less sensitive than MRI for detecting retrocochlear abnormality

### **Differential Diagnosis**

#### 1. Key Differential Diagnoses:<sup>3</sup>

- Acute
  - Sudden idiopathic SNHL
  - Infection (acute otitis media, external otitis, syphilis, Lyme disease, viral)
  - Perilymphatic fistula
  - Ischemia of retrocochlear structures
  - Multiple sclerosis
  - Autoimmune disease
  - Traumatic causes
  - Metabolic (chronic renal failure)
  - Hematologic (sickle cell anemia)
- Rapidly progressive
  - Autoimmune inner ear disease
  - Meningeal carcinomatosis
  - Vasculitis secondary to infection (Rocky Mountain spotted fever)
  - Lyme disease
  - Otoxic exposure (aminoglycosides, diuretics, chemotherapy)

#### 2. Extensive differential diagnoses:

- Fluctuating
  - Perilymphatic fistula
  - Meniere's disease
  - Multiple sclerosis
  - Migraine-associated hearing loss
  - Infection (syphilis)
  - Autoimmune (Cogan's syndrome, systemic lupus, polyarteritis nodosa, Wegener's syndrome, temporal arteritis, scleroderma)
  - Sarcoidosis
- Gradual
  - Presbycusis
  - Noise-induced
  - Familial
  - Retrocochlear neoplasm
  - Chronic otitis media, cholesteatoma
  - Otosclerosis
  - Endocrine (hypothyroidism, diabetes mellitus)
  - Paget's disease
  - Metabolic (chronic renal failure, hyperlipoproteinemia)
  - Mucopolysaccharidosis

## Therapeutics

### 1. Acute Treatment:

- Oral corticosteroids (prednisone or methylprednisolone) tapered over 10 to 14 days
  - Immediate treatment for unilateral idiopathic sudden hearing loss and additional symptoms (dizziness or tinnitus) is 14-day course of 60 mg prednisone (with taper). EBM rating: (SOR:C)<sup>7</sup>
  - Spontaneous recovery occurs within the first 2 weeks after onset of Sudden SNHL
  - Greatest recovery of hearing when corticosteroids is initiated within 2 weeks; minimal benefits if greater than 4 weeks from symptom onset
  - Systemic steroids cannot be considered gold standard for Sudden SNHL; benefits remains unclear<sup>8,9,10</sup>
  - Audiogram should be done within 24 to 48 hours after treatment for documenting extent of hearing loss

### 2. Further Management:

- Intratympanic corticosteroids as primary or salvage therapy<sup>4,11,12,16,18</sup>
  - Rationale: delivers high concentration to specific tissue with less systemic effects
  - As primary treatment, appears equivalent to treatment with high-dose oral prednisone therapy
  - As salvage therapy for patients who do not improve with oral treatment, may result in hearing improvement
  - May cause discomfort, less convenient, more costly
- Randomized trials comparing corticosteroids alone to corticosteroids plus antiviral agents failed to show added benefit from antiviral agents<sup>4,6,10,13</sup>
- Vasodilators and vasoactive substances showed no evidence of benefit; studies were poor quality and number of patients were small<sup>4,6,9,14</sup>
- No evidence of benefits from hyperbaric oxygen; its use is not recommended<sup>4,6,13,15</sup>
- Superiority of fibrinogen-LDL-apheresis over standard first line treatments not established<sup>17</sup>

### 3. Long Term Care:<sup>4</sup>

- Scuba diving contraindicated due to risk of tympanic membrane rupture, hearing loss, tinnitus, and balance problems
- Ear plugs or earmuffs should be used to protect against loud noises or music
- Unaffected ear should have immediate otolaryngologic evaluation to assess for any signs/symptoms of pathology

## Follow Up

1. Return to Office: follow up preferably face-to-face to
  - provide support,
  - assess efficacy of therapy and stage of recovery,
  - address any concerns or side effects
2. Refer to Specialist: immediate referral to otolaryngologist
3. Admit to Hospital: not specified unless other serious diagnoses (e.g. stroke) considered

## Prognosis

1. Not well documented
2. Patients with Sudden SNHL may have a 1.64 times greater risk of stroke during 5 years follow up compared to patients undergoing appendectomy<sup>4,6</sup>
3. Patients with Sudden SNHL should have audiometric monitoring repeated over the course of a year to monitor recovery, direct rehabilitation (hearing aids), and monitor signs of relapse in affected ear or development of hearing loss in contralateral ear<sup>4</sup>

## Prevention

3. Not well documented

## Patient Information

1. Information for patients:  
<http://www.nidcd.nih.gov/health/hearing/pages/sudden.aspx>

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