Sudden Sensorineural Hearing Loss (SSNHL)

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Learning Objectives

- Understand the basic anatomy and physiology of the peripheral auditory system
- Define Sudden Sensorineural Hearing Loss (SSNHL)
- Describe the epidemiology, diagnostic evaluation, and treatment options for patients with SSNHL
- Distinguish between idiopathic SSNHL and autoimmune hearing loss
- Understand the basics regarding aural rehabilitation
• The middle ear contains 3 ossicles: the malleus, incus, and stapes.

• The inner ear consists of the cochlea, the utricle, the saccule, the endolymphatic duct/sac, and the semicircular canals.

• The cochlea has 2.5-2.75 turns around a bony core (modiolus).
Definition

- Hearing loss can be categorized as conductive (loss of vibratory energy) or sensorineural (related to the inner ear or auditory nerve)

- The most common definition of SSNHL is sensorineural hearing loss > 30 dB in 3 contiguous pure tone frequencies occurring in < 3 days
**Epidemiology**

- SSNHL was first described in 1944
- Estimated incidence is 10-20/100,000
- Highest incidence in patients between 40 and 60 years of age
  - Can affect patients of all ages
- > 95% are unilateral
- No gender preference

**Differential Diagnosis**

<table>
<thead>
<tr>
<th>Infectious</th>
<th>Neurologic</th>
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</thead>
<tbody>
<tr>
<td>Viral: Viral idiopathic</td>
<td>Multiple sclerosis; Focal pontine ischemia; Migraine</td>
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<tr>
<td>Other causes - Meningococcal meningitis; Herpesvirus (simplex, zoster, varicella, CMV); Mumps; Human immunodeficiency virus; Lassa fever; Mycoplasma; Cryptococcal meningitis; Toxoplasmosis; Syphilis; Rubella; Human immunodeficiency virus</td>
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</tr>
<tr>
<td>Vascular</td>
<td>Autoimmune</td>
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<tr>
<td>Alteration of cochlear microcirculation</td>
<td>Autoimmune Inner Ear Disease</td>
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<tr>
<td>Vertebral insufficiency; Red blood cell deformability; Sickled cell disease</td>
<td>Other causes - Ulcerative colitis; Relapsing polychondritis; Lupus erythematosus; Polyarteritis nodosa; Cogan’s syndrome; Wegener’s granulomatosis</td>
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<tr>
<td>Cardiopulmonary bypass</td>
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<td>Traumatic: Intracochlear membrane rupture and Perilymph fistula</td>
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<tr>
<td>Temporal bone fracture; Inner ear concussion; Otologic surgery; Surgical complication of non-otologic surgery</td>
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**For idiopathic SSNHL, treatments have focused on:**

- Minimizing inflammation
- Improving inner ear blood flow/oxygenation
- Re-establishing the endocochlear potential.

- Majority of cases are idiopathic
- No single cause accounts for all cases
- Treatments are directed towards the cause if the cause is known

**Viral Cochleitis**

- **The dominant cause of idiopathic SSNHL**
  - 17-33% of patients recall a viral illness within a month of SSNHL
  - Compared with controls, the rates of herpes virus sero-conversion are higher in patients with SSNHL
**Viral Cochleitis**

- The most compelling evidence comes from temporal bone histopathology consistent with viral injury to the inner ear
  - Loss of inner ear hair cells
  - Atrophy of the stria vascularis
  - Atrophy of the tectorial membrane
  - Neuronal loss
- Diagnosis of exclusion

**Intra-cochlear Membrane Rupture**

- Allows mixing of endolymph and perilymph
- Disrupts the endocochlear potential
  - The stria vascularis generates an endocochlear potential of +80 mV within the scala media.
  - Ion pumps within hair cells create an intracellular potential of -70mV.

**Vascular Compromise**

- The cochlea is an end organ supplied by the labyrinthine artery
  - Vertebrobasilar system
  - No collateral circulation
- Thrombosis, embolic phenomena, vasospasm, and hyper-coagulable states can compromise inner ear oxygenation.

**Perilymph Fistula**

- Leakage of perilymph thru the oval or round windows
- Inciting events:
  - Physical trauma to the head
  - Sneezing
  - Bending/Lifting heavy objects
  - Sudden barometric pressure changes (flying or diving)
  - Acoustic trauma exposure to a loud noise.
### Acoustic Neuroma (Vestibular Schwannoma)

- Nerve sheath tumors originating from Schwann cells (8th cranial nerve).
- Significant patient morbidity due to their critical location
- 1% of patients with SSNHL have a vestibular schwannoma
- 3-12% of patients with VS presented with SSNHL

### Autoimmune Disease

- Introduced in 1979 by McCabe and colleagues as rapidly progressive bilateral SNHL that responds to immunosuppressive medications.
  - Bilateral but onset and progression may be asymmetric
  - Timeline is weeks to months
  - 50% have vestibular (balance) symptoms
  - Unknown epidemiology but much rarer than SSNHL

- May be part of systemic autoimmune diseases
  - Cogan’s syndrome
  - Wegener’s granulomatosis
  - Polyarteritis nodosa
  - Temporal arteritis
  - Buerger’s disease (thromboangitis obliterans)
  - Systemic lupus erythematosus
- May be isolated to the inner ear without systemic involvement.

- Diagnosis: Response to steroids
  - CBC with diff
  - ESR
  - RF
  - ANA
  - Anti-double stranded DNA antibodies
Autoimmune Disease

- Anti SSA/B antibodies
- Anti-phospholipid antibodies
- C3 and C4 complement levels
- Western blot for 68 kD protein
- Raji cell assay for circulating immune complexes

- Rule out syphilis and central pathologies

Autoimmune Disease

- Continue steroids until monthly audiograms demonstrate that hearing has stabilized
- Slowly taper steroids over 8 weeks to a maintenance dose of 10-20 mg every other day
- Most patients require > 6 months of steroid therapy

- Rheumatology consult!

Autoimmune Disease

- Treatment with immunosuppressive medications
  - Prolonged course of high dose oral steroids
    - 60 mg oral prednisone per day for 4 weeks for adults
    - 1 mg/kg oral prednisone per day for pediatric patients
  - Many do not improve until week 4

Evaluation and Management
History

- The immediate goal is discovering a treatable or defined cause of the sudden hearing loss.
- Ask about the onset, time course, and characteristics of the hearing loss
  ✓ Is it constant or intermittent? Associated with position changes?
  ✓ Is it mild, moderate, severe, or profound?
  ✓ Can the patient use a telephone?

Physical Examination

- Otoscopy
  ✓ Believe your physical exam!
  ✓ If there is no ear canal obstruction, ear drum pathology, or fluid in the middle ear... be worried
- Cranial nerve examination
  ✓ Especially cranial nerves 5,7,10,11,12
- Balance Examination
  ✓ Gait testing, Romberg, cerebellar tests, look for nystagmus

History

- Always ask about balance function, tinnitus, prior ear surgery, a history of noise exposure, recent sick contacts, travel history, and inquire about facial nerve dysfunction
- Past medical history may reveal risk factors for hearing loss.
- All medications, including over-the-counter products, must be described.

Tuning Fork Tests

- Weber: Lateralizes to the ear with a conductive HL
- Rinne: A positive Rinne indicates that air conduction is greater than bone conduction. A negative Rinne indicates that bone conduction is greater than air conduction
Audiograms

• Audiograms are graphic representations of auditory sensitivity and are used clinically to test hearing.
• Most audiologists test the 250 Hz – 8000 Hz range (x-axis)
• The y-axis of an audiogram is plotted in decibels (dB), a logarithmic scale.

Lab Tests

• International normalized ratio (INR), activated partial thromboplastin time (aPTT), and clotting time for coagulopathy
• CBC and differential for infection
• Thyroid-stimulating hormone (TSH) for thyroid disease
• Fasting blood glucose for diabetes mellitus
• Cholesterol and triglycerides for hyperlipidemia

Lab Tests

• A shot-gun approach to laboratory examinations is not cost-effective for SSNHL
• Laboratory studies should be directed by the history and physical examination
  ✓ Fluorescent treponemal antibody-absorption (FTA-Abs) for syphilis
  ✓ Antinuclear antibodies (ANA), rheumatoid factor, erythrocyte sedimentation rate (ESR), and 68kD protein Western blot for autoimmune diseases

Imaging

• MRI of the head with and without contrast is the imaging modality of choice
**Imaging**

- CT scans are useful for evaluating the bony anatomy of the temporal bone

![CT scans of temporal bone](www.homepage.mac.com/tigershark/temporalbone)

**Treatment Options**

- Directed towards the cause if one is identified based on work-up
  - Acoustic Neuroma: observation versus stereotactic radiation versus surgical resection
  - Perilymph fistula: middle ear surgical exploration and closure

**Treatment Options**

- The gold standard for the treatment of idiopathic SSNHL is oral steroids
  - Ideal steroid window is within 7 days
  - Steroid therapy within 3 weeks has a reasonable chance at success
About 1/3 of patients recover without therapy, but about 2/3 recover if given oral steroids.

My regimen: 30 mg oral Prednisone twice daily for 2 weeks followed by 30 mg oral prednisone once daily for a week then off.

- An audiogram is obtained 1 week after completion of steroids.

Four variables have been shown to affect recovery from ISSNHL: (1) time since onset, (2) audiogram type, (3) vertigo, and (4) age.

- Steroids within 7 days is optimal.
- Patients less than 15 years of age and over 65 years of age have a poorer prognosis.
- The presence of vertigo portends a poor prognosis.
- Patients with > 90 dB (profound) hearing loss have a poor prognosis.

**Treatment Options**

- Trans-tympanic steroids for patients with a contraindication to oral steroids or patients who fail oral steroids.
  - Type of steroids and optimal frequency for administration are being investigated.
  - My regimen: Dexamethasone 24 mg/ml.
    - 2 injections spaced 2 weeks apart then an audiogram 2 weeks later. If there is benefit, I offer patients another injection.
    - Rare complication is a perforation at the injection site.

**Prognosis**

**Aural Rehabilitation**
Hearing Aids
- Standard hearing aids
- CROS (contralateral routing of signal) hearing aids

The Cochlear Implant

Osteo-Integrated Implant
Bone Conduction Hearing Aid

Summary
- The majority of sudden, unilateral sensorineural hearing loss is idiopathic
- Believe your physical exam. Be worried if the ear drum and middle ear appear intact.
- Start an adequate dose of oral steroids as soon as possible
- Order an MRI to rule out intracranial pathologies such as an acoustic neuroma
Summary

- Direct other evaluations based on history and physical exam
- Refer the patient to an Otologist for formal audiometry and further management
- Several modalities of aural rehabilitation are available to patients to improve their quality of life

Topics to be Discussed:

1) Treatment of Acute and Chronic Sinusitis
2) Options for Therapy of Primary Snoring

Management of Common Ear, Nose and Throat Conditions

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Anatomy of Sinuses

- Paranasal sinuses drain through ostia, whose size and patency greatly influence the health of the sinus
- Biphasic Mucus Blanket
- Mucus moves in spiral pattern to and out the sinus ostia
- Complete clearing every 10 minutes
- Ostiomeatal Complex (OMC): confluence of sinus drainage
### Sinusitis: Definition

- **Recurrent sinusitis:**
  >3 episodes/6mos, >4/1 yr. Sxs clear between episodes.

- **Chronic sinusitis:**
  Sxs >12wks

### Sinusitis: Diagnosis

#### Diagnosis

- **Acute sinusitis:**
  - Diagnosis
    - Major Factors: Facial pain, facial pressure, nasal obstruction, purulent discharge, hyposmia, fever
    - Minor: Headache, fatigue, halitosis, dental pain
  - 2 or more major, 1 major and 2 minor, or 3 minor signs and symptoms, sinusitis strongly suspected

*American Academy of Otolaryngology Task Force Guidelines, 2005*

#### Procedures

- Anterior rhinoscopy
- Nasal endoscopy
- Imaging
Sinusitis: Diagnosis

- Nasal Endoscopy
- Helpful in identifying inflammation not visible anteriorly
- Helpful in identifying anatomic factors causing recurrent disease

Sinusitis: Pathogenesis

- URI: edema, ciliary dysfunction
- Environmental: allergy, cigarette smoke exposure
- Anatomic obstruction: septal deviation, turbinate hypertrophy, nasal polyps
- Immunodeficiency
- Systemic diseases

Sinusitis: Diagnosis

Order CT scan if:

- Diagnostic criteria of chronic sinusitis met
- Complications of sinusitis suspected
- Anatomic abnormalities suspected

Sinusitis: Pathogenesis

- Mucosal edema at OMC leads to obstruction of anterior ethmoid and maxillary sinuses, mucus stasis and then infection.
- Posterior ethmoids affected later
**Sinusitis: Microbiology**

- **Acute sinusitis:** *Pneumococcus* (most common), non-typeable *H. Flu*, *M. catarrhalis*, viruses

- **Chronic sinusitis:** Same as acute plus: *Staph (Coag plus and minus)*, anaerobes, *Pseudomonas*

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**Medical Treatment of Acute Sinusitis**

**GOALS**

- Control infection
- Reduce tissue edema
- Facilitate drainage
- Maintain patency of sinus ostia
- Break the pathologic cycle leading to chronic sinusitis

**Antibiotic Therapy:**

- Excellent Pneumococcal activity
- Good gram negative activity (*H. flu*, *Morax cat*)
- **Antibiotic:** 14 days of High Dose Amoxil or Bactrim DS is in most cases curative. As number of recurrent episodes increase or if symptoms are poorly controlled, more potent agents are required.

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**Medical Treatment of Acute Sinusitis**

- **If Amoxicillin or Bactrim DS are not effective?**
  - Consider culture of mucopurulent secretions
  - **Empiric Therapy:**
    - Beta lactamase PCNs
    - 2nd generation Cephalosporins
    - Macrolides
      - *Clarithromycin* >>> *Erythromycin* >>>> *Azithro*
      - More potent agents in a given class result in less resistance
### Medical Treatment of Acute Sinusitis

- **Adjunctive tx:**
  - Mucolytics (guaifenesin 1200mg BID)
  - Nasal Saline Irrigations (200 ml BID)
  - Oral and topical decongestants
  - Nasal and oral steroids
  - Treatment of Environmental Allergies
- **Re-evaluation after completing therapy**

### Medical Treatment of Chronic Sinusitis

#### Empiric Antibiotic Therapy:
- Excellent Pneumococcal activity
- Good gram negative activity
- Adequate staphylococcal coverage
- Adequate anaerobic coverage
- Culture directed therapy preferred in chronic sinusitis

### Medical Treatment of Acute Sinusitis

- Polyp reduction with 5 days of oral steroids

### Medical Treatment of Chronic Sinusitis

- **Antibiotic:** Broad spectrum Abx X 4 wks.
- **Adjunctive tx:**
  - Mucolytics (guaifenesin 1200mg BID)
  - Nasal Saline Irrigations (200 ml BID)
  - Oral and topical decongestants
  - Treatment of Environmental Allergies
  - Nasal and oral steroids
- **Re-evaluation after completing therapy**
### Danger signs in the patient with sinusitis:

- Failure to improve
  - Resistant infection?, Fungal disease?, Misdiagnosis?
    - Change antibiotic, CT, Nasal Endoscopy
  - Proptosis, orbital cellulites, cranial nerve deficit, high fever, or altered mental state suggest extension
  - Urgent evaluation and management necessary

### Surgical Treatment of Chronic & Recurrent Sinusitis:

- Children
  - Adenoidectomy usually curative in pediatric sinusitis
  - Endoscopic Sinus Surgery indicated in recurrent disease

### Surgical Treatment of Chronic & Recurrent Sinusitis:

- Adults
  - Endoscopic Sinus Surgery is usually curative if predisposing factors are controlled
  - Intent of Endoscopic Sinus Surgery is to restore function to the sinuses by allowing drainage from the natural ostia
### Surgical Treatment of Chronic & Recurrent Sinusitis:

**Image Guided Sinus Surgery**

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### Snoring

- **Snoring**: Audible airway turbulence during sleep
  - Primary Snoring is defined by the absence of obstruction, significant oxygen desaturation, or ventilation deficiencies

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### Current Strategies in the Care of the Snoring Patient

#### Snoring

- **Population studies on snoring**:
  - 60% of American Males
  - 30% of American Females

- **Of which**:
  - 80% of bed partners of snorers relate that this effects their relationship in a negative manner.
  - 20% of bed partners of snorers choose to sleep in another room because of the snoring.
**Can we reliably clinically distinguish O.S.A. from Primary Snoring?**

<table>
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<tr>
<td>Studies have shown that the use of historical and physical examination findings does not reliably predict the degree or presence of Obstructive Sleep Apnea nor distinguish it from Primary Snoring.</td>
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**Obstructive Sleep Apnea versus Primary Snoring**

- The standard for confirming the presence of O.S.A. remains the polysomnogram.
- If O.S.A. is confirmed, the standard of care remains positive pressure therapy.

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**Why can’t we predict the presence or absence of Obstructive Sleep Apnea?**

**Disease Continuum**

No obstruction

- Snoring
- Airway Resistance
- Hypopnea
- Sleep Apnea

The interplay of multiple sites of obstruction, coupled with effects of dynamic anatomy and individual factors (Age, Alcohol intake, Allergies, Reflux, etc) result in great individual variability along this continuum.

**Primary Snoring**

- Is treatment of Primary Snoring necessary?
- Individuals who snore without obstruction when compared to non-snoring controls have:
  - Increased levels of daytime sleepiness
  - Worse general health scores
  - Derioration in exercise capacity
  - Higher rates of divorce/separation
Primary Snoring

- In the patient with Primary Snoring selecting the right therapeutic course is often more challenging than in clear cut severe OSA.

- Behavioral and multiple anatomic factors contribute to this condition and must be addressed for successful therapy to occur.

Treatment of Primary Snoring

- Factor most affecting treatment choice and ultimate success is the correct determination of the site or sites of airway turbulence.

Treatment of Primary Snoring

- Weight Reduction:
  - ✓ Significant association between weight loss and reduction in bed partners’ snoring intensity scores

- Behavioral Modification:
  - ✓ Clear association between alcohol intake and the presence of primary snoring.

Nasal Obstruction

- Increases intrapharyngeal pressure gradient.

- Leads to open mouth posture with resultant posterior tongue base displacement.
Nasal Obstruction

- Inflammatory Cause
  - Allergic
  - Non-Allergic
- Fixed Cause
  - Inferior Turbinate Hypertrophy
  - Nasal Septal Deformity

Treatment of Nasal Inflammation

1) Nasal Steroid Sprays
2) Oral Decongestants
3) Nasal Saline Irrigation
4) Allergy Evaluation

Treatment of Fixed Nasal Obstruction

- Inferior Turbinate Hypertrophy
  - Treated by Turbinate Reduction

- Nasal Septal Deformity
  - Treated by Septoplasty
Tongue
Oral Appliances:

**Hypertrophic Tonsils:**
- While uncommon in adults, reduction of significantly hypertrophic tonsils can have a dramatic effect on the degree of airway turbulence.

Soft Palate and Uvula
- Probably most common site of primary snoring.
- Redundancy creates turbulence and vibration

Soft Palate and Uvula
- In the past surgical therapies to address the soft palate and uvula were associated with significant discomfort and long recuperation.

Uvulopalatopharyngoplasty (UPPP)
Pillar Implantation

- Insertion under local anesthesia of three woven implants into soft palate to increase structural support and reduce palatal vibration and collapse.
- Performed in office in 30 minutes with the majority of patients returning to work the same day and not requiring pain medication.

Summary

- Snoring represents a continuum between Primary Snoring and Obstructive Sleep Apnea.
  ✓ Significant marital and health consequences can be related to Primary Snoring.
  ✓ Polysomnogram required to distinguish where in the continuum an individual patient exists.

Pillar Implantation

Publications on Pillar Implantation:

- Mean bed partner satisfaction score increase 92%
- Return to work day of procedure 85%

Summary

- Behavioral modifications and weight lose can lead to improvements in snoring intensity scores.
- Determining the site of airway turbulence is the key to therapy of primary snoring.