Sudden Sensorineural Hearing Loss

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Grand Rounds Presentation
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Introduction-SSNHL

- Devastating to patients
- Frustrating for physicians
- Definitive diagnosis and treatment still unknown
- First described by De Klevn in 1944
Introduction-SSNHL

Definition: 30 dB or greater SNHL over at least three contiguous audiometric frequencies occurring within 3 days or less.
Statistics

- 15,000 reported cases per year worldwide
- 4,000 cases per year in the U.S.
- 1/10,000-15,000 will be afflicted
Statistics

- Highest incidence in 50-60 years olds
- Lowest incidence in 20-30 years olds
- M=W
- 2% bilateral
- 90% of cases are idiopathic
Introduction

- Suggested causes of Idiopathic SNHL (ISNHL)
  - Viral infections
  - Autoimmune
  - Vascular compromise
Etiology

1) Infectious
2) Autoimmune
3) Traumatic
4) Vascular
5) Neoplastic
History

- Time course
- Associated symptoms
  - Vertigo/dizziness
  - Aural fullness
  - Tinnitus
- Ototoxic drug use
- Symptoms of URTIs
- H/O head trauma, straining, sneezing, nose blowing, intense noise exposure
- H/O flying or SCUBA diving
History

- PMH:
  - Autoimmune disorders
  - Vascular disease
  - Malignancies
  - Neurologic conditions
  - Hypercoagulable states
    - Sickle cell disease (African Americans)
- PSH: stapedectomy or other otologic surgeries
Physical Exam

🔹 Complete H&N exam in everyone
   ❣ Ears: r/o effusions, cholesteatoma, cerumen impaction
   ❣ Weber/Rinne
   ❣ Neurologic exam – cerebellar findings
      ● Tandem gait
      ● Romberg
      ● Nose to finger, heal to shin
   ❣ Vestibular – Dix-Hallpike test
Diagnostic Testing

- Audiogram
  - Pure tone
  - Speech discrimination
  - Tympanometry
  - Stapedial reflex

- Laboratory testing
  - CBC
  - ESR
  - RPR, VDRL
  - Lymphocyte transformation test
  - Western blot for antibodies to 68 KD protein
Diagnostic Testing

- **MRI:**
  - Rule out cerebellopontine angle tumors
  - Multiple sclerosis
  - Ischemic changes
- 13% of patients with acoustic tumors present with SHL
- 23% may recover hearing
Known Treatable Causes of SSNHL
Autoimmune SHL

- Cogan’s syndrome
- Wegener’s granulomatosis
- Polyarteritis nodosa
- Temporal arteritis
- Buerger’s disease (Thromboangitis Obliterans)
- Systemic Lupus Erythematosus
- Primary
Autoimmune SHL

- Pathogenesis – theories
  - Vasculitis of vessels of the inner ear
  - Autoantibodies (antigenic epitopes)
  - Cross-reacting antibodies
Autoimmune SHL

- **Cogan’s syndrome**
  - Autoimmune disease of the cornea and inner ear
  - Age of onset 22-29 years
  - Presentation – interstitial keratitis and Meniere’s like episodes
  - Associated systemic diseases
    - Aortitis – 10%
Cogan’s Syndrome
Autoimmune SHL

- Cogan’s Syndrome
  - Hearing fluctuates with disease exacerbations and remissions
  - Majority develop bilateral deafness (67%)
  - Etiology is unknown
    - ? Microbial etiology
Autoimmune SHL

- **Cogan’s Syndrome**
  - **Diagnosis** –
    - Requires both eye and inner ear manifestations of inflammation
    - CBC, ESR, RPR, FTAbs
    - MRI/CT
  - **Therapy** –
    - Corticosteroids – prednisone 1mg/kg X 2-4 wks
    - Cochlear implantation
Traumatic SHL

- Breaks in the membranous labyrinth
  - Intracochlear – Meniere’s
  - Oval and/or round window – perilymph fistula
- History – inciting event
  - Blow to the head
  - Sneezing
  - Bending over
  - Lifting a heavy object
  - Exposure to sudden changes in barometric pressure
    - Flying, SCUBA diving
Traumatic SHL

- High risk population
  - Post stapedectomy
  - Inner ear anomalies
    - Mondini malformation
    - Large vestibular aqueduct
Traumatic SHL

- **Diagnosis**
  - **Definitive** – intraoperative
  - **Usually clinical**
    - Audio - Sudden or rapid progressive hearing loss
    - Inciting event
    - R/o inflammatory process, neoplasia
      - (MRI, ESR, syphilis test)
    - Exam – Hennebert’s sign (fistula test)
    - Tullio’s phenomenon
**Traumatic SHL**

- **Treatment**
  - Strict bed rest
  - HOB elevated 30 degrees
  - Avoid lifting > 10 lbs.
  - Avoid straining or hard nose blowing
  - +/- stool softeners
  - Some suggest daily audio
Traumatic SHL

- After 5 days
  - If improvement – 6 weeks of light activity
  - If no improvement – surgery
    - Middle ear exploration
    - Patching of perilymph fistula
Neoplasia

- Acoustic tumors
  - Usually present with gradually progressive SNHL
  - 10%-19% may present with SHL
  - 1% of patients with asymmetric SNHL have acoustic tumors
Idiopathic Sudden Sensorineural Hearing Loss (ISSNHL)
ISSNHL

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

- 1999 – Albers and Schirm
  - Guinea pig model of viral labyrinthitis
    - Similar pathologic findings to patients with ISSNHI
  - Mechanism of viral induced pathology
    - Likely auto immune
- 1990 – Harris
  - Showed immunosuppressed guinea pigs with CMV labyrinthitis had less hearing loss
  - Steroids improve hearing outcomes
Viral

- Direct identification of virus in perilymph
- **Davis and Johnson** – demonstrated ability of rubeola and mumps to infect the inner ears of animal models with immunofluorescent antigen studies
- **Westmore** - cultured mumps form perilymph of SHL patient
- **Davis** cultured CMV from perilymph of infected infant
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
👩‍⚕️ BPRSNHL
👩‍⚕️ Absence of systemic immune disease
👩‍⚕️ 50% with dizziness
👩‍⚕️ Light-headedness and ataxia more common than vertigo
👩‍⚕️ Episodes – multiple, daily
👩‍⚕️ Hearing loss may be:
  👩‍⚕️ sudden, rapidly progressive, or protracted
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%
AIED

1990 – Harris and Colleagues
- Used Western blot to discover anti 68KD autoantibody in sera of patients with ISSNHL
- 22%-58% will have +test
- 94% specificity
AIED

◆ Further studies
  ◆ Billings and Harris
    ◆ Linkage of 68KD protein to heat shock protein 70 (hsp 70)

◆ Theories
  ◆ 1) Cross reactivity
  ◆ 2) Over expression leads to autoimmunity
AIED

- Prednisone 1mg/Kg/day for 4 weeks
- Slow taper
- Relapse during taper – restart
- Slow taper
- If relapse during taper – Cytotoxic agent
  - Methotrexate
  - Cyclophosphamide
- Monitor electrolytes, LFTs, blood counts
McCabe favors starting with cyclophosphamide and prednisone from the start
Vascular

- 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

1980 – Belal

- Examined two temporal bones of patients with SHL
  - Histopathology was similar to animal models of vascular occlusion
  - Extensive fibrosis and ossification
Vascular-histopathology

Fig. 3.—A. Changes in the cochlear duct one day after venous obstruction showing fragmentation of the stria and hemorrhage in the scala tympani with early changes in the end organ and spiral ligament.

B. Changes in the cochlear duct 17 hours after arterial obstruction showing dissolution of the organ of Corti and stria vascularis with marked cellular changes in the limbus, spiral ligament and Reissner’s membrane.
Vascular Anatomy
Vascular

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

- 90% of cases will be Idiopathic
- Treat known causes by addressing the underlying condition
Treatment

- Therapy for ISSNHL is controversial
- Difficult to study
  - High spontaneous recovery rate
  - Low incidence
  - Makes validation of empiric treatment modalities difficult
Treatment

- Proposed treatment modalities
  - Anti-inflammatory – steroids, cytotoxic agents
  - Diuretics
  - Antiviral agents
  - Vasodilators
  - Volume expanders/hemodilutors
  - Defibrinogenators
Treatment

- 1987 – Wilkins and associates
  - “shotgun” regimen – dextran, histamine, Hypaque, diuretics, steroids, vasodilators, carbogen
  - No difference between treated and non-treated patients
  - No control group
  - Treatment for only three days
Treatment

- No benefits found in prospective, randomized, double-blind studies looking at
  - dextran 40,
  - pentoxifylline,
  - low-molecular-weight dextran, and
  - IV procaine
Treatment

- **Steroids**
  
  - 1980 – Wilson and colleagues
    - Double-blind studies with oral steroids in patients with ISSNHL
    - Decadron given over 10-12 days
    - Patients stratified based on audiogram
    - Results: steroids work in patients with hearing loss between 40 and 90 db
    - No effect for patients with >90 db
    - Midfrequency loss – patients excluded from study
  
  - 1984 - Findings confirmed by Moskowitz
Fig 2.—Categories of sudden hearing loss as determined by spontaneous recovery rate.
Treatment

1996 - Review by Hughes

- Recommendations for treatment
  - Low salt (2g/day diet) and Maxide once daily
  - Prednisone 1mg/kg/day
  - Acyclovir 1-2 g orally daily in five divided doses for 10 days
Treatment

- **Carbogen**
  - 95% oxygen and 5% carbon dioxide
  - Shown to increase perilymph $O_2$ saturation
  - $CO_2$ – potent cochleovestibular vasodilator
  - No studies have show benefit over spontaneous recovery

- Hughes recommends use in patients who have one only hearing ear
Treatment

- **Carbogen**
  - Requires in-hospital administration to monitor for raises in BP
  - Insurance does not cover
  - Considered experimental
Treatment

- **Acyclovir**
  - 1999 - Stokroos and Albers
    - Showed therapeutic efficacy of combined steroid and acyclovir in experimental HSV-1 viral labyrinthitis
      - Earlier hearing recovery
      - Less extensive cochlear destruction
  - 1996 – Adour *et al.*
    - Combination therapy shown to be beneficial for tx of Bell’s palsy
  - Benefit of combined therapy has been shown in patients with Ramsay Hunt syndrome
Treatment

- 2000 survey of 100 ENTs (43% otologists) in the United Kingdom
  - 78% - CBC, ESR, Syphilis serology
  - 38% - MRI on initial visit
  - 98.5% - steroids
  - 41% - Carbogen
  - 31% - acyclovir
Prognosis

- 47%-63% spontaneously resolve
  - Combined patients with all audiogram types
- Four prognostic variables
  - Time since onset
  - Audiogram type
  - Vertigo
  - Age
1984 – Byl

- 8 year prospective study of 225 patients with ISSNHL
- Looked at factors for prognosis
  - Age
  - Vertigo
  - Tinnitus
  - Audiogram pattern
  - Time elapsed on presentation
  - ESR level
Prognosis

Age
Prognosis

* Vertigo – 29% affected vs. 55% not affected
Prognosis

🌟 Audiogram type

![Audiogram graph showing SRT, 30 dB Discrim 52%, SRT, 85 dB Discrim 24%, Best, Worst]
Prognosis
## Prognosis

<table>
<thead>
<tr>
<th>Severity of hearing loss</th>
<th>Prognosis for normal or complete recovery</th>
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<tbody>
<tr>
<td></td>
<td>Patients seen within 7 days</td>
<td>Patients seen after 7 days</td>
</tr>
<tr>
<td></td>
<td>No vertigo</td>
<td>Mild</td>
</tr>
<tr>
<td>Mild (24 - 34 dB)</td>
<td>90%</td>
<td>90%</td>
</tr>
<tr>
<td>Intermediate (35 - 54 db)</td>
<td>80%</td>
<td>75%</td>
</tr>
<tr>
<td>Severe (55 - 74 dB)</td>
<td>60%</td>
<td>50%</td>
</tr>
<tr>
<td>Profound (≥ 75 db)</td>
<td>70%</td>
<td>30%</td>
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Conclusion

- SHL is devastating to patients
- Frustrating for physicians to dx and tx
- Thorough H&P
- Rule out treatable cause
- Directed labs, Audiogram MRI
- Discuss risks, benefits, and alternatives of treatment with the patient
- Treat the disorder aggressively
- Rehabilitate those whose hearing does not improve
- Follow patients for development of associated diseases and for contralateral ear disease