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

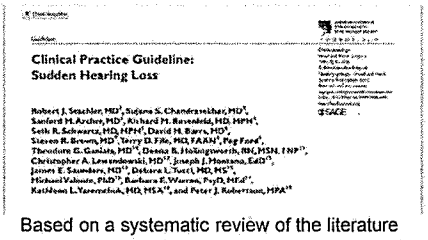
Post mortem histopathologic studies in 12 patients with history of SSNHL

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



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

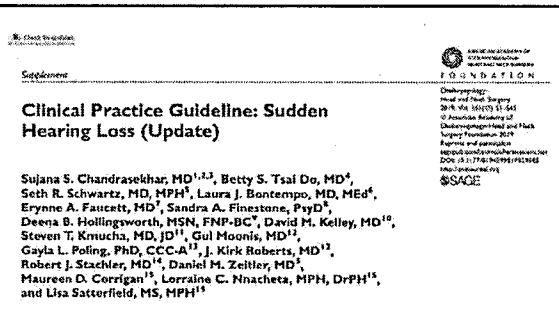
Guideline Update Group

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



Clinical Practice Guideline: Sudden Hearing Loss (Update)

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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

Idiopathic Sudden Sensorineural Hearing Loss

- 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, ginko biloba
- Vasoactive substances – prostaglandin E1
- Rheologic agents – pentoxifylline (Trental), dextran

Frequently asked questions

What is causing the problem?

How is sudden hearing loss diagnosed?

Will my hearing come back?

What additional testing is needed?

How is sudden hearing loss treated?

What are the side effect of each treatment?

What else can I expect?

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 that 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

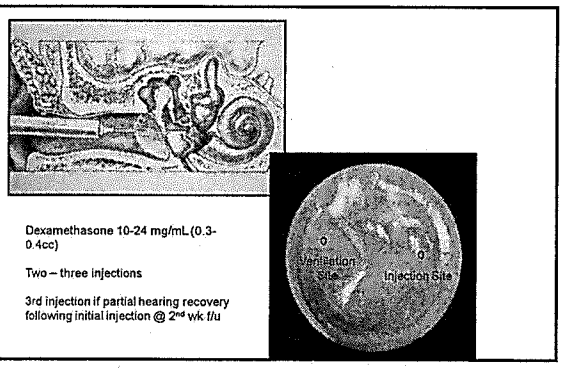
Idiopathic Sudden Sensorineural Hearing Loss

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

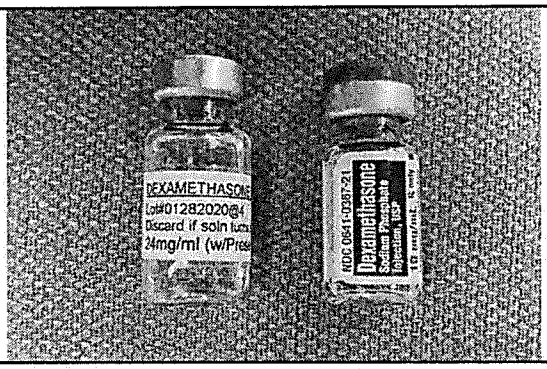
- Patient supine, involved side up, 15-30 minutes, not to swallow
- 3-4 injections over 2-4 weeks, can terminate early if no improvement



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

Two – three injections

3rd injection if partial hearing recovery following initial injection @ 2nd wk f/u



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

Idiopathic Sudden Sensorineural Hearing Loss

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

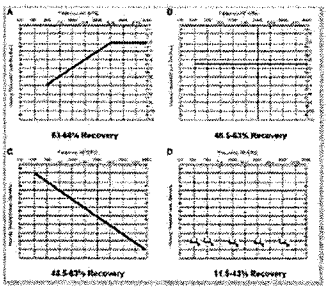
Outcomes

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