

Hearing Science Accelerator (HSA)

Sudden Sensorineural Hearing Loss (SSNHL)

Executive Summary & Research Progress

June 2025

What is the Hearing Science Accelerator (HSA)?

The HSA aims to advance the science of hearing disorders for which there has long been a lack of new treatment options. Through this initiative, we aim to foster collaboration and change the approach to hearing research from iterative to innovative.

HSA Process Overview

1. Disease Prioritization

- recruitment of experts
- literature search/reviews

2. Disease Workshop

- in-person workshop with experts
- begin writing white paper

3. Disease Initiative

- disseminate research questions (i.e. white paper, conferences)
- facilitate workgroups to answer research questions

Goals

- Improve diagnostic treatment options for hearing disorders
- Identify priority research questions
- Involve disciplines outside of Otolaryngology
- Identify barriers to research
- Identify short, intermediate, and long-term goals for advancing clinical care and research

Who is Involved?



Why Sudden Sensorineural Hearing Loss (SSNHL)?

SSNHL is the rapid-onset subjective sensation of hearing impairment in one or both ears. It occurs within a 72-hour window and consists of a decrease in hearing of >30 dB affecting at least three consecutive audiometric frequencies.



SSNHL Subtopics

- **Diagnosis**
 - clinical diagnosis of exclusion; no test specific for disease
- **Etiology**
 - unknown
- **Treatment**
 - 80 years without new successful therapies

Why SSNHL?

SSNHL was selected given its prevalence, disability, and the lack of advances that have been made towards our understanding and treatment of the disease.

Disease Prioritization:

Literature search and review

Findings

Etiology

- There are likely multiple etiologies that lead to sudden hearing loss
- There is evidence to support several, but it is not definitive

Diagnosis

- Comprehensive audiometric testing can provide useful information
- MRI provides the most useful diagnostic information
- Serologic testing is low yield without positive history

Treatment

- Primary steroid therapy:
 - No definitive evidence for steroids over placebo
 - No conclusive evidence on dose, timing, delivery route, or formulation
- Salvage steroid therapy:
 - Multiple meta-analyses support intratympanic steroids
- Hyperbaric oxygen treatment:
 - Cochrane review supports it when the outcome is limited to 25% improvement
 - Numerous lower-level study suggest benefit
- All other therapies:
 - Limited to non-existent

Disease Workshop: HSA SSNHL Workshop

In-Person Workshop

April 21-23, 2023
Washington D.C.

Participants

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Andrea Vambutas, MD
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Disease Initiative: Questions

Question 1

- Do steroids really work? Are steroids effective therapy for sudden sensorineural hearing loss? What studies are required?
 - Placebo-Controlled Trial
 - Placebo vs oral steroid? Low dose vs Standard dose (10mg vs 60mg prednisone)
 - Does a higher concentration of IT dexamethasone (24mg/ml) improve hearing outcomes? (i.e. 24 mg/ml vs 10mg/ml)

Question 3

- How can we use functional and imaging studies to identify the site of lesion, better predict prognosis or treatment protocol.
 - Would earlier testing within (72 hours of onset) help?
 - Would a natural history study of SSNHL help?
 - Does the pattern of SSNHL on audiometric testing predict treatment response?
 - A standardized testing protocol would allow comparison of standard data and potential a central registry for SSNHL. The following test should be included in such a testing protocol.
 - 1. Behavioral audiogram: pure tones averages (PTA) 250 Hz to 8 kHz, air and bone conduction, word recognition scores, tympanometry, acoustic reflexes
 - 2. Extended high-frequency audiogram if possible – up to 18 kHz
 - 3. Distortion product otoacoustic emissions (DPOAE)
 - 4. Electrocochleography (ECoG)
 - 5. Vestibular evoked myogenic potentials (VEMPs) at 250 and 500 Hz

Question 2

- Can acute MRI predict response to treatment?
 - Is the MRI noise safe in the setting of acute inner ear hearing loss?
 - Is it possible to get a quiet inner-ear protocol?
 - Can early MRI help us determine etiology of SSNHL? Diagnostic tests protocols for intralabyrinthine, inflammation, infection, ischemia, hydrops, bleeding or schwannoma would be required.
 - Can MRI determine the role of cochlear hydrops in SSNHL?

Question 4

- Can a biobank and patient registry be developed in order to determine the etiology of SSNHL? Possible specimens would include genetic, serum, and/or perilymph testing.
 - Can Genetic testing determine susceptibility or predict prognosis of SSNHL?
 - Are there biomarkers in the serum predictive of etiology, diagnosis or treatment?
 - What test of perilymph would aid in the treatment of SSNHL?

Question 5

- Can perilymph obtained during cochlear implantation or during the acute presentation aid in diagnosis and/or treatment of SSNHL?
 - Would biologic markers elucidate perilymph antigens helpful in etiology or diagnosis?
 - Could the etiology of hearing loss be determined by analysis of perilymph content in the future?
 - Would variation in perilymph composition be predictive of different SSNHL etiologies?
 - A standard procedure for perilymph collection should be developed for maximum yield and minimal risk to hearing and balance.

Question 7

- How can we use existing temporal bone specimens to better understand the etiology of SSNHL?
 - What are the sites of injury within the cochlea in patients with SSNHL?
 - Is SSNHL associated with cochlear hydrops? To what extent (i.e. severity and distribution along the cochlea from base to apex)? With what frequency? Is cochlear hydrops associated with specific patterns of cellular injury in SSNHL?
 - Is there an immune and inflammatory response in the cochlea of patients who suffered SSNHL? Does this occur only acutely, or does it persist even years after the SSNHL?
 - Is there evidence of viral infection in the human cochleae of patients who suffered SSNHL? Or, is there evidence of viral reactivation or viral induced immune response?

Question 6

- Can we standardize outcome measures in SSNHL?
 - Can we create a standard for outcomes reporting?
 - Can we define what constitutes an acceptable functional outcome?
 - Is there a way to standardize word recognition scores across languages?
 - At what time intervals should we measure outcome?

Question 8

- How does the timing of treatment affect outcome? Most studies suggest that earlier treatment is better, but the natural history is still not well understood.
 - Does Immediate vs Intermediate vs Late Intervention make a difference and how do we define and measure this difference?
 - How can we obtain patients quickly after onset of symptoms and institute immediate (<24 hours of loss) treatment?
 - Can we measure and correlate outcome of treatment with timing of start of treatment; Timing of HBO2 +/- steroids?

Disease Initiative: Research Progress

Key Initiatives and Collaborative Efforts

- **HSA proved to be successful**
 - Completed HSA disease prioritization and workshop phases.
 - Published findings to accelerate research translation.
 - Fostered high-level collaboration among researchers.
- **NIH Clinical Database**
 - Developing standardized SSNHL patient database.
 - Aims to enhance data analysis, trend identification, and treatment evaluation.
- **Steroid Treatment Study**
 - Pending study on effectiveness of steroids in SSNHL.
- **Genetic Biobank**
 - Will support research into genetic causes and treatment outcomes of SSNHL.
- **Early MRI Protocol and New Imaging Techniques**
 - Collaborative effort with neuroradiologists in progress.
 - Aims to enhance early diagnosis and understanding of SSNHL.
 - Seminar on new MRI techniques scheduled for May 24, 2025.
- **Perilymph Sampling Protocol**
 - Collaborative protocol development for perilymph sampling and testing in progress.
 - Seminar on perilymph protocol scheduled for May 10, 2025.
- **Blood Biobank**
 - Creating a cooperative biobank to study serum biomarkers.
 - Investigates autoimmune and infectious disease links in SSNHL.

Your Support to the HSA

For additional information, questions, or ways to get involved, please email hsacoordinator@hifla.org or visit <https://hifla.org/sudden-sensorineural-hearing-loss-general-info/>

To support our mission of developing a cure for Sudden Sensorineural Hearing Loss, please **click the link** below.



Thank You!