

Seminar Summary Report:

“Early MRI Protocols to Understand Pathophysiology of Sudden Sensorineural Hearing Loss (SSNHL)”

Date: Saturday, 24th May 2025

Time: 09:00–11:00 GMT-07:00

Speakers:

- John A. Butman, MD, PhD, NIH (ret.)
- Bryan Ward, MD, Johns Hopkins Medicine
- Akira Ishiyama, MD, UCLA
- John L. Go, MD, UCLA
- Amy Juliano, MD, Mass Eye and Ear
- William Slattery, MD, House Ear Clinic, House Institute Foundation

Attendees:

- Seilesh Babu
- Parveen Bazard
- Derald Brackmann
- Jane Cabison
- Keiko Hirose
- John House
- Claudia Kirsch
- Hossein Mahboubi
- Sudhir Manickavel
- Shemms Najjar
- Madelynn Poe
- Stefan Plontke
- Ali Andalibi
- Akil Turner
- Andrea Vambutas
- Adam Vesole
- Mary Ying
- 3 unknown names

Introduction: HSA and SSNHL

The House Institute Foundation Hearing Science Accelerator (HSA) is a strategic initiative designed to advance biomedical research in hearing disorders that have historically lacked effective treatment options. By bringing together leading experts in hearing science and thought leaders from other disciplines to an in-person workshop, the HSA fosters innovative, cross-disciplinary collaboration. For each targeted disorder, the HSA identifies critical research questions with the aim of shifting from progress to transformative innovation.

Sudden Sensorineural Hearing Loss was defined during the seminar as a rapid loss of hearing, typically in one ear, amounting to more than 30 decibels over at least three consecutive frequencies and occurring within a 72-hour window—often instantaneously. Despite decades of investigation, its etiology remains unclear. Experts discussed various theories, including vascular, viral, autoimmune, and inflammatory causes, but no definitive conclusion has been reached. William Slaterry emphasized that much of the existing research remains stagnant and lacks translational impact, urging for more purposeful collaboration and innovation, which is a goal of the Hearing Science Accelerator initiative.

Q2: Can acute MRI predict response to treatment?

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

Q3: How can we use functional and imaging studies to identify the site of lesion, better predict prognosis or treatment protocol?

- a. Would earlier testing within (72 hours of onset) help?
- b. Would a natural history study of SSNHL help?
- c. Does the pattern of SSNHL on audiometric testing predict treatment response?
- d. 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

Radiology Seminar

The seminar titled “*Early MRI Protocols to Understand Pathophysiology of Sensorineural Hearing Loss*” brought together a multidisciplinary group of clinicians, radiologists, neuroscientists, and researchers to address the persistent challenges in diagnosing Sudden Sensorineural Hearing Loss (SSNHL). The meeting aimed to evaluate current imaging approaches, explore emerging MRI techniques, and develop consensus on the implementation of more effective and timely imaging protocols to understand the pathophysiology behind SSNHL.

The primary objective of this session is to evaluate the current landscape of MRI imaging in the context of SSNHL and to establish a standardized early-stage MRI protocol. By reviewing existing imaging techniques and exploring opportunities for enhancement, the session aims to advance the understanding of SSNHL’s underlying causes. Special emphasis will be placed on how MRI advancements can facilitate earlier diagnosis and more effective treatment strategies, ultimately improving patient outcomes.

The meeting was hosted by the HSA chair, William Slattery, MD, and featured focused presentations, followed by open discussions on future research directions and ethical considerations.

Speaker Presentations

John A. Butman, MD, PhD: “FLAIR imaging to monitor inner ear inflammation”

Dr. John Butman presented the challenges and potential of using FLAIR imaging in monitoring inner ear conditions. He expressed concern over the limited publication and integration of FLAIR imaging into current protocols, pointing to a significant gap in both knowledge and clinical practice. While high-resolution water-weighted imaging (T2, CISS, SPACE etc.) is routinely used to detect structural abnormalities, it falls short in identifying the mechanisms underlying sudden sensorineural hearing loss (SSNHL). T1-weighted imaging, both with and without contrast, remains standard for detecting vestibular schwannomas and some inflammatory masses, but it rarely reveals inflammation within the inner ear due to its limited sensitivity.

Dr. Butman highlighted that certain inflammatory inner ear pathologies, such as those seen in Kogan syndrome and granulomatosis with polyangiitis, underscore the diagnostic value of FLAIR imaging—especially in identifying inflammation not visible on T1-weighted scans. He referenced that despite the resolution limitations of standard FLAIR sequences, they are still sensitive to detect inner ear inflammation on immediate/early post contrast imaging. He also discussed how post-contrast FLAIR imaging has proven effective not only for detecting inner ear inflammation in conditions like NOMID, a rare neonatal autoinflammatory disease, but also monitoring the effectiveness of anti-inflammatory therapies and suggested that similar imaging strategies could be applied to inner ear disorders.

In terms of clinical application, Dr. Butman advocated for the development of a streamlined MRI protocol for acute cases, incorporating high-resolution T2 imaging and pre and post-contrast

FLAIR sequences. This protocol, which could be completed in under 15 minutes, would help detect early inflammatory changes and guide timely treatment to prevent progressive hearing loss.

Bryan Ward, MD: “1.5 vs. 3.0 vs 7t imaging”

Dr. Bryan Ward emphasized the vital role of MRI in diagnosing sudden sensorineural hearing loss (SSNHL), particularly in ruling out retrocochlear pathologies such as vestibular schwannomas. However, he acknowledged that many subtle inner ear conditions remain undetectable with current imaging techniques. This underscores a critical gap in diagnostic capability, as many theoretical vestibular disorders are still inferred rather than directly observed through imaging.

Dr. Ward discussed the technical limitations contributing to this issue, including partial volume averaging and large voxel sizes, which can obscure small but clinically significant structures like membrane ruptures or otoconial debris. He highlighted the importance of ultra-high field MRI systems, noting that 3 tesla MRI offers double the signal of 1.5 tesla, and 7 tesla provides even greater improvements in resolution. With more than 130 7T scanners globally—over 50 in North America—access to advanced imaging tools is growing.

Despite these advances, Dr. Ward noted that even 7T imaging may not reveal all underlying mechanisms of SSNHL. He pointed to emerging technologies like 11.7 tesla MRI, which are beginning to resolve finer inner ear structures such as the cupula and crista, showing promise for the future. Nevertheless, he expressed frustration with the current inability to visualize the causes of hearing loss in many patients.

As a path forward, Dr. Ward recommended further research into finer spatial resolution, minimizing partial volume effects, and developing new contrast techniques to enhance the detection of inner ear pathologies. These advancements are necessary to bridge the diagnostic gap and improve outcomes for patients with SSNHL.

Akira Ishiyama, MD: “Delayed imaging protocol and its effect on inner ear”

Dr. Akira Ishiyama’s presentation on delayed MRI contrast infusion for inner ear imaging focused on its application in diagnosing endolymphatic hydrops, particularly in patients with Meniere's disease. He highlighted over 15 years of successful use of this protocol, demonstrating its effectiveness in identifying regional changes in the cochlear duct, saccule, and utricle. In cases of isolated cochlear hydrops, patients primarily experienced cochlear symptoms without vascular involvement, reinforcing the diagnostic value of the imaging approach.

Notably, these localized inner ear changes can mimic the full spectrum of Meniere's disease symptoms, such as hearing fluctuations and vertigo. Imaging findings, such as FLAIR signal changes, were also linked to early detection of Meniere’s disease, as shown in a 2012 study by Yamazaki and later in a 2016 study that found increased FLAIR signals in asymptomatic

contralateral ears of symptomatic patients. Additionally, diuretics were found to improve hearing in some patients, suggesting the condition may be dynamic and responsive to treatment.

Dr. Ishiyama supported these imaging observations with histological evidence, showing significant edema and thickening of the basement membrane in Meniere's patients, providing a physiological explanation for the MRI findings. Further data, including results from pupillometry measurements in nearly 600 patients, revealed a correlation between sympathetic nervous system activity and the onset of Meniere's attacks.

In conclusion, high-resolution MRI, particularly with delayed contrast infusion, is a critical tool for diagnosing audiovisceral dysfunction and tracking physiological changes associated with inner ear disorders like Meniere's disease.

John L. Go, MD: "Imaging Endolymphatic Hydrops"

Dr. John L. Go presented on advanced imaging techniques used to evaluate the inner ear, highlighting key challenges and developments in diagnosing conditions such as Meniere's disease. He noted that while high-resolution T2-weighted MRI is commonly used, it has limitations in distinguishing between the membranous and bony labyrinths, necessitating more specialized imaging methods.

One such technique involves intratympanic gadolinium injections to enhance MRI contrast, enabling clearer visualization of the membranous labyrinth by highlighting the perilymph. Additionally, Dr. Go emphasized the utility of 3D FLAIR imaging, which is widely available and particularly helpful when subtraction imaging is not feasible.

For endolymph visualization, the protocol includes scanning with sub-millimeter slice thickness and isotropic voxels, using modified FLAIR sequences with specific inversion times (2,350 ms and 2,050 ms). This method allows clinicians to detect filling defects—a key sign of endolymphatic hydrops in Meniere's disease. Imaging abnormalities are considered significant when volume uptake exceeds 3%, with 50% being strongly indicative of pathology.

Dr. Go also discussed cochlear duct imaging, explaining that if the cochlear duct fills more than 50% of the apical turn, it is considered a positive finding for Meniere's. However, he raised concerns about the practicality of delayed imaging protocols, particularly the challenge of scheduling four-hour follow-up scans, which may limit widespread clinical adoption.

Finally, he warned of the risk of image corruption due to patient movement between imaging sequences, stressing the need for careful acquisition protocols to ensure diagnostic accuracy.

Amy Juliano, MD: "Timing of MRI with onset of disease"

Dr. Amy Juliano provided an in-depth discussion on the etiologies and imaging considerations for sudden sensorineural hearing loss (SSNHL), emphasizing the importance of timing and the

choice of imaging protocols. She outlined that SSNHL may arise from both central (brain-related) and peripheral (inner ear) causes, including ischemia, infections, and neurologic conditions.

A major concern raised was the timing of MRI imaging, especially for conditions like ischemia or inflammation, which require early detection to avoid missing transient or evolving abnormalities. Imaging delays can lead to false negatives or misdiagnoses, particularly after steroid treatment, which may reduce inflammation before imaging findings related to it can be captured by MRI. A standard temporal bone MR imaging protocol at Massachusetts Eye and Ear includes thin-slice high-resolution T1-weighted and heavily T2-weighted sequences through the internal auditory canal and temporal bone region for detailed assessment of the labyrinthine structures, internal auditory canal, and cerebellopontine angle cistern, as well as DWI, SWI, T2-weighted, and T1-weighted sequences of the whole brain to rule out central pathology. Dr. Juliano noted that heavily-T2-weighted imaging (e.g. fast/turbo spin echo with driven equilibrium, balanced steady-state free precession), while useful for delineating fluid-filled structures, cannot distinguish between types of fluid such as endolymph from perilymph. As such, in select cases, a four-hour delayed 3D FLAIR sequence is used for inner ear/Meniere's disease imaging, noting that this type of 3D FLAIR sequence employs a different inversion time than conventional CSF-null FLAIR imaging.

Dr. Juliano highlighted recent trends in the utilization of AI in radiology, noting the recent development of some AI tools designed to accentuate certain imaging features such as contrast enhancement. A thoughtful and judicious approach to integrating and adopting these new AI software tools would be important to ensure accurate and robust imaging diagnostics.

Citing a 1998 study¹ and a 2017 study,² Dr. Juliano suggested that MRI for idiopathic SSNHL should likely be performed as early as possible, ideally before the initiation of any treatment, to allow the capture of imaging findings that reflect the initial pathologic process with greatest integrity and maximize diagnostic sensitivity and specificity. She also mentioned a paper that examined the utility of the labyrinthine 3D FLAIR sequence in the setting of idiopathic SSNHL, noting that the study found abnormalities on this sequence in 27% of patients in their cohort, and that in those with initial severe and profound hearing loss, the presence of 3D FLAIR abnormalities indicated a poor prognosis.³

1. Stokroos RJ, Albers FW, Krikke AP, Casselman JW. Magnetic resonance imaging of the inner ear in patients with idiopathic sudden sensorineural hearing loss. *Eur Arch Otorhinolaryngol*. 1998;255(9):433-6. doi: 10.1007/s004050050093. PMID: 9833208.
2. Conte G, Di Berardino F, Sina C, Zanetti D, Scola E, Gavagna C, Gaini L, Palumbo G, Capaccio P, Triulzi F. MR Imaging in Sudden Sensorineural Hearing Loss. Time to Talk. *AJNR Am J Neuroradiol*. 2017 Aug;38(8):1475-1479. doi: 10.3174/ajnr.A5230. Epub 2017 May 25. PMID: 28546251; PMCID: PMC7960407.
3. Lee JI, Yoon RG, Lee JH, Park JW, Yoo MH, Ahn JH, Chung JW, Park HJ. Prognostic Value of Labyrinthine 3D-FLAIR Abnormalities in Idiopathic Sudden Sensorineural Hearing Loss. *AJNR Am J*

Summary:

The seminar emphasized that while MRI is a cornerstone tool in ruling out retrocochlear pathology such as vestibular schwannomas, current imaging methods frequently fail to identify more subtle, inner ear-specific abnormalities. Many speakers noted the limitations in resolution, timing, and contrast sensitivity that hinder current diagnostic efforts. Timing of imaging was a repeated concern—steroids, often administered early in treatment, may obscure inflammation and other markers on scans. Therefore, participants agreed that imaging should be performed before steroid administration to maximize diagnostic yield.

Several imaging modalities were explored in-depth, including T1- and T2-weighted sequences, post-contrast FLAIR imaging, and emerging 3D high-resolution techniques.

Challenges in the healthcare system's ability to perform immediate imaging were acknowledged, including logistical and scheduling hurdles. Participants discussed the importance of aligning clinical and research protocols to ensure findings could translate into practice. The group agreed that further collaboration is essential, particularly through initiatives like the Hearing Science Accelerator, which can foster protocol sharing, preliminary data generation, and cross-disciplinary dialogue.

In terms of clinical implications, the seminar highlighted the complexity of diagnosing inner ear diseases. Advances in imaging may help distinguish between autoimmune, infectious, ischemic, and inflammatory pathologies that present similarly. The role of autoimmune markers and studies on perilymph fluid were noted as promising directions for future research.

In conclusion, the seminar underscored that current MRI practices are insufficient for detecting many cases of SSNHL. There is a need for early, targeted, high-resolution imaging protocols to improve diagnosis and treatment outcomes. Participants emphasized collaboration, technology adoption, and clinical integration as key to advancing the field. The next steps include developing formalized imaging protocols, leveraging high-field MRI systems, sharing research across institutions, and integrating imaging data with clinical findings to drive precision diagnosis and therapy for sensorineural hearing loss.

Next Steps:

The seminar concluded with a set of actionable next steps.

1. Optimal MRI timing is upon ER admission or on the same day as clinical evaluation, before steroids are administered.
2. At this time, there is no need for serial MRI imaging until we can identify abnormalities with an initial protocol.

3. Development of a standardized imaging protocol for:
 - a. 7 tesla
 - b. Quick scanning protocol
 - c. Comprehensive MRI scanning protocol
4. This will likely have to be done clinically opposed to a funded research endeavor, unless associated with another clinical trial
5. Put together groups of individuals who want to work together to develop protocols and implement them in their institutions, in a prospective collection manner that can be published.