

SELECTED ABSTRACTS

***ORAL
PRESENTATIONS***

IN ORDER OF PRESENTATION



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AMERICAN NEUROTOLOGY SOCIETY***

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New Orleans, LA***

Evaluating Functional Hearing Outcomes and Associated Predictors after Resection of Cerebellopontine Angle Meningioma

*Khalil Baddour, MD; Vanessa Helou, MD; Parthasarathy D. Thirumala, MD
Philip L. Perez, MD*

Objective: To characterize functional hearing outcomes after resection of cerebellopontine angle (CPA) meningioma, and determine factors associated with preserved or improved postoperative hearing

Study Design: Retrospective cohort

Setting: Tertiary referral center

Patients: Adults with CPA meningioma

Interventions: Surgical resection

Main Outcome Measures: Preservation or improvement of hearing via subjective reporting, pure tone averages (PTA), word recognition scores (WRS), and/or American Academy of Otolaryngology-Head and Neck Surgery (AAO-HNS) hearing classification grade

Results: Twenty-five patients (72% female) with a median (range) age at surgery of 55 (47) years were included. Most tumors (40%) originated from the posterior petrous ridge. The internal auditory canal (IAC) was involved in 72% of cases. The median (range) tumor volume was 5.98 (46.78) cm³. Meningiomas were most commonly resected via a combined approach involving a retrosigmoid craniotomy (53%) with 64% of procedures resulting in gross total resection. The preoperative median (range) PTA and WRS were 33.75 (92.5) dB and 88% (100%), and were preserved postoperatively at PTA 33.13 (108.75) dB and WRS 94% (100%). Eighty percent of patients had reported or measured improvement/preservation in their postoperative hearing. Of those, 11 had improvement/maintenance of functional hearing based on audiometric data. Univariable analysis evaluated the association between functional hearing outcomes and age, tumor laterality, volume, location, IAC involvement, surgical approach, extent of resection, preoperative hearing loss, audiometric data and AAO-HNS class. The final proposed model to predict postoperative hearing outcomes included age (OR 1.19, 95% CI 1.01-1.41, *p*-value 0.04) and preoperative WRS (OR 0.97, 95% CI 0.94-1.00, *p*-value 0.08), although neither remained statistically significant in multivariable analysis.

Conclusions: Postoperative preservation or improvement of hearing can be expected after hearing-preserving resection of CPA meningiomas. Older age and lower preoperative WRS may portend poorer functional hearing outcomes.

Professional Practice Gap & Educational Need: There is scarce literature evaluating functional hearing outcomes after CPA meningioma resection and potential predictive factors that may be associated with worse postoperative hearing.

Learning Objective: To quantify postoperative functional hearing outcomes after CPA meningioma resection and associated factors

Desired Result: To improve preoperative patient counseling regarding expected functional hearing outcomes and any potential patient-specific associated risk factors

Level of Evidence – Level IV

Indicate IRB or IACUC: University of Pittsburgh STUDY18120055

Proteomic Analysis Identifies Novel Plasma Biomarkers in Patients with Vestibular Schwannoma

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Hypothesis: Patients with sporadic vestibular schwannoma (VS) have unique plasma protein biomarkers that can accurately distinguish and classify them from healthy, non-tumor controls.

Background: Conventional radiographic modalities, such as MRI, are resource intensive and offer limited insight into tumor biology. Histological markers such as ki-67, while informative, require invasive craniotomy and are not applicable in non-surgical patients. Identification of plasma-derived biomarkers for VS could enhance disease prognostication and guide treatment decisions.

Methods: High-throughput, multiplexed, DNA aptamer-based proteomic analysis was performed in plasma samples from 12 patients, six with sporadic non-irradiated VS and six age-/gender-matched healthy controls. Dysregulated proteins were identified using a cut-off value of $|\log_2\text{foldchange}| > 1$ and $p_{adj} < 0.05$. Enriched pathways were determined using Metascape and STRING bioinformatic analysis. Candidate biomarker expression was validated in tumor tissue, a schwannoma cell line, and primary VS culture.

Results: A total of 7310 proteins were profiled. Of 1499 differentially expressed proteins, 152 (10%) were upregulated and 273 (18%) were downregulated in VS. There was an enrichment in cancer proliferation, protein catabolism and immune cell activation processes. A panel of 40 proteins distinguished VS from HC, accounting for 84% of the variance on principal component analysis. These included NFKBIA, WNT10A and WNT16, proteins integral to NF- κ B and Wnt signaling. Four were further validated *in vitro*. *IGFBP-1*, *IGFBP-2* and *FCGR3A* mRNA expression were elevated >20-fold in schwannoma cells. Hepcidin (HAMP), a regulator of iron homeostasis and metabolism that influences tumor growth, was enriched in VS tissue and in primary VS culture-derived secretions compared to wild-type Schwann cells (308 vs. 6 pg/mL).

Conclusions: Our study identified several plasma biomarkers that classify VS patients. Identification of hepcidin warrants further investigation into its role in tumor progression.

Professional Practice Gap & Educational Need: Vestibular schwannomas are associated with significant morbidity. No consensus plasma biomarkers for VS exist. There is a critical gap to identify prognostic biomarkers to molecularly stratify VS patients and guide clinical decision making.

Learning Objective: To better understand the role of plasma proteins as a predictive tool to distinguish VS patients from healthy controls and identify potential novel therapeutic targets.

Desired Result: Identification of novel circulating plasma proteins in VS that may promote tumor growth.

Level of Evidence – Level V

Indicate IRB or IACUC: The Ohio State University IRB Protocol #IRB1994H0241

**HERBERT SILVERSTEIN AWARD FOR RESEARCH
EXCELLENCE IN OTOTOLOGY/NEUROTOLOGY**

**Whole Genome Sequencing of Sporadic Vestibular Schwannoma
Identifies Novel Molecular Pathways**

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Hypothesis: Whole genome sequencing of sporadic vestibular schwannoma (VS) specimens will reveal novel genetic mutations and molecular pathways involved in the pathogenesis of the disease.

Background: The optimal treatment for VS remains uncertain due to inability to predict tumor behavior, growth, and symptom progression. While molecular changes and mutational burden may govern tumor behavior, genotype-phenotype correlations are not well established. The objective of this study was to describe the genomic landscape of sporadic VS utilizing whole genome sequencing (WGS), with the aim to uncover novel genes and pathways involved in tumor behavior.

Methods: Tumor and matched peripheral blood specimens were collected from 28 patients with sporadic VS who underwent surgical resection. Demographic and clinical characteristics including pre-operative hearing status, cystic change, tumor size, and tumor growth were obtained. Specimens were processed and DNA extraction was completed. WGS was performed and mutational burden and functional enrichment analysis were completed.

Results: WGS was performed on 23 tumor specimens. 46 genes were mutated in 4 or more samples. Commonly mutated genes included TTN, KMT2C, AHNK2, CCDC168. STRING network analysis demonstrated several network associations. ADGRV1, OTGL, CCFC168, and TRIOBP, genes which have previously been shown to play an essential role in the development of hearing and inherited deafness, were affected. Mucin genes, important in middle ear and endolymphatic sac inflammation, were also altered. Functional enrichment analysis revealed several enrichments, with affected genes involved in histone methylation and extracellular matrix structure. 16 of 23 samples had 32 identified mutations in the NF2 gene. Of these, 24 were intron variants.

Conclusions: WGS of sporadic VS tumors identified several novel genes and molecular pathways which may be important drivers of tumor behavior.

Professional Practice Gap & Educational Need: The optimal treatment for vestibular schwannoma has been debated for decades. Much of this debate derives from an inability to predict tumor behavior, which generates significant uncertainty. An improved understanding of the genetic and molecular drivers of tumor behavior may advance management of the disease and lead to novel therapeutic targets.

Learning Objective: To describe novel genetic mutations and molecular pathways involved in the pathogenesis and behavior of sporadic vestibular schwannoma tumors.

Desired Result: To utilize genomics data to better understand vestibular schwannoma behavior and to inform future research.

Level of Evidence - Level V

Indicate IRB or IACUC: University of California San Diego Institutional Review Board #180556 and #181755

Automated Segmentation of Bilateral Vestibular Schwannoma

*Krish Suresh, MD; Ryan Weiss, MS; Daniel J. Lee, MD
D.'Bradley Welling, MD, PhD; Yin Wu, PhD; Matthew G. Crowson, MD*

Objective: Automated segmentation models for volumetric measurement of vestibular schwannoma (VS) have been developed for sporadic VS but not for bilateral VS. Automated segmentation would be especially valuable in this setting: Patients with neurofibromatosis 2 (NF2) undergo numerous MRI scans, and automated analyses would aid in timely therapeutic decision-making. We aim to develop a computer vision model for the volumetric measurement of bilateral VS.

Study Design: Retrospective study

Setting: Tertiary referral centers

Patients: 87 individuals with VS (59 sporadic, 28 NF2) from our institution; 30 patients with sporadic VS from an open-source dataset.

Interventions: A nnU-Net was trained on our institutional data augmented with the public data to develop an automated segmentation model for VS on T1-post contrast MRI. The model was tested on a holdout set of sporadic and bilateral VS scans.

Main Outcome Measures: Dice score to compare pixel-wise agreement, qualitative review

Results: Median tumor volumes were 0.345 cc for institutional sporadic VS, 2.05 cc and 0.501 cc for the larger and smaller tumors for institutional bilateral VS, and 1.36 cc for the public sporadic VS. There was a high incidence of comorbid intracranial pathology in the NF2 cases, including 39% with meningiomas and 32% with trigeminal schwannomas. The final model achieved a mean Dice score of 0.94 on the holdout set of internal sporadic VS, 0.95 on public sporadic VS, and 0.87 on bilateral VS. On qualitative review of the NF2 cases, the model performed well at distinguishing VS from adjacent non-VS lesions. The model also performed well at detecting small tumors in cases with significant size asymmetry. The model struggled in cases with non-contiguous segments of tumor, often including one segment but not the other.

Conclusions: This study is the first in the literature to report on the automated segmentation of bilateral VS. Further work is necessary to improve model performance, extend it to the postoperative setting, and apply it to other intracranial tumors.

Professional Practice Gap & Educational Need: Despite the well-established benefits of volumetric measurements for VS, adoption is limited.

Learning Objective: To identify challenges with automated segmentation for bilateral VS and understand ongoing efforts to overcome these.

Desired Result: Attendees will consider applying automated segmentation for volumetric measurement of VS in sporadic and simpler NF2 cases.

Level of Evidence - IV

Indicate IRB or IACUC: Exempt

Comparing Cochlear Implantation With vs. Without Resection in Intralabyrinthine Schwannoma: A Systematic Review and Meta-Analysis

Brendon K. Warner, MD; Lawrence Lee, MD; Nauman F. Manzoor, MD

Objective: To analyze the differences in outcomes between cochlear implantation (CI) with and without primary resection of intralabyrinthine schwannomas (ILS).

Data Sources: PubMed, Embase, and CINAHL.

Study Selection: Two-person title/abstract screening followed by full text screening. Of 759 studies screened, 13 met inclusion criteria.

Data Extraction: JBI risk of bias tool for case series was used to assess validity of included studies.

Data Synthesis: Statistical testing included meta-analysis of mean differences and two-sample t-tests.

Results: A total of 97 patients were analyzed, 80 in CI with resection, and 17 in CI without resection. CI with resection showed favorable audiometric outcomes, with a mean difference in PTA of -76.5 ($p < 0.00001$, [-84.0, -69.1]) and in WRS of 47.5 ($p = 0.0002$, [22.8, 72.1]). CI without resection also showed favorable audiometric outcomes, with PTA mean difference: -82.8 ($p < 0.00001$, [-96.0, -69.5]), and in WRS: 41.1 ($p < 0.00001$, [25.7, 56.4]). Comparison of PTA mean difference between CI with and without resection showed no statistically significant difference, $t = -0.3$ ($p = 0.77$, [-17.2, 12.9]). WRS comparison also showed no statistical significance, $t = 0.64$ ($p = 0.53$, [-25.3, 49.0]).

Conclusion: CI placement, both with and without resection of ILS, showed improvement in audiometric outcomes. There does not appear to be a statistically significant difference in audiometric outcomes if the tumor is resected or not when CI is placed. More research should be done regarding CI without resection to give a better understanding of outcomes and when resection may or may not be warranted.

Professional Practice Gaps: Current minimal application of cochlear implantation of ILS patients without tumor resection, with even less research done on outcomes.

Learning Objectives: To understand audiometric outcome differences between cochlear implantation with and without ILS tumor resection, and to weigh the risks and benefits of each approach.

Desired Results: To improve audiometric outcomes for patients with ILS, to improve surgeon ability to decide between best approaches to ILS treatment for optimum patient care, and to increase awareness for the need for further research regarding cochlear implantation in ILS with and without concurrent resection.

Level of Evidence - Level I

IRB: Exempt

NEUROTOLOGY FELLOW AWARD

Radiosurgery for Sporadic Facial Nerve Schwannomas: An International Multi-Institutional Study

*John P. Marinelli, MD; Justin Cottrell, MD; Eric E. Babajanian, MD; Simon K.W. Lloyd, BSc (Hons), MPhil
Jason P. Sheehan, MD; J. Walter Kutz, Jr., MD; Matthew L. Carlson, MD*

Objective: To characterize patient outcomes following primary stereotactic radiosurgery (SRS) for management of sporadic facial nerve schwannomas.

Study Design: Retrospective cohort study.

Setting: Six tertiary referral centers across the United States and United Kingdom.

Patients: Adults undergoing SRS from 2000 to 2023 for sporadic facial nerve schwannomas along any segment of the facial nerve were included. Patients with NF2-related schwannomatosis were excluded.

Interventions: SRS.

Main Outcome Measure: Long-term tumor control, defined as salvage treatment-free survival at 10 years.

Results: Among 59 patients meeting inclusion, the median age at SRS was 52 years (IQR 42-65) with a median tumor size of 19.5 mm (IQR 15.4-22.8). Tumors commonly involved the internal auditory canal (72%), cerebellopontine angle (50%), geniculum/labyrinthine (48%), and tympanic segments (22%). Cochlear fistula was present in 3 (5%). Two patients underwent salvage treatment; salvage-free survival rates (95% CI; number still at risk) at 1-, 3-, 5-, and 10-years following SRS were 100% (100-100; 53), 100% (100-100; 36), 100% (100-100; 18), and 87% (72-100; 9), respectively. Among 31 (53%) patients with House-Brackmann (HB) grade I facial function at presentation, only 5 demonstrated worse facial function at a median of 3.2 years (IQR 1.7-7.1) following SRS. Of 18 patients with serviceable hearing (AAO class A/B) at SRS, 12 maintained serviceable at a median of 1.1 years (IQR 0.5-5.2) of post-SRS audiometric follow-up.

Conclusions: Durable tumor control following primary SRS for sporadic facial nerve schwannomas is achieved in most patients. Among those with HB grade I facial function at presentation, the majority demonstrated preservation of facial nerve function at last follow up.

Professional Practice Gap & Educational Need: Secondary to disease rarity, limited evidence currently exists surrounding the efficacy and expected outcomes of stereotactic radiosurgery when used in the primary management of sporadic facial nerve schwannomas which limits patient counseling and understanding of treatment response.

(1) Understand the common clinical presentations of sporadic facial schwannomas that undergo radiosurgery.

Learning Objectives: (2) Describe long-term tumor control following primary radiosurgery for sporadic facial nerve schwannomas.

(3) Describe evolution in facial function, hearing, and other common related symptoms following treatment.

Desired Result: Providers would be able to describe the long-term efficacy of stereotactic radiosurgery when used as primary management of sporadic facial nerve schwannomas, as well as the expected evolution in patient symptoms, including but not limited to facial function, hearing, facial spasm, xerophthalmia, dysgeusia, trigeminal hypesthesia and pain, and vestibular function.

Level of Evidence: IV

Indicate IRB or IACUC: 24-000217; S22-01574; STU112016-040

Evaluating for Endolymphatic Hydrops in Meniere's Disease Using In-Vivo 7 Tesla Magnetic Resonance Imaging and Advanced Post-Processing Techniques

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John P. Carey, MD; Jun Hua, PhD; Bryan K. Ward, MD*

Objective: To conduct a volumetric analysis of the inner ear's membranous labyrinth in patients with Meniere's Disease (MD) vs. healthy controls using high-resolution 7 tesla (T) MRI.

Study Design: Prospective cohort study.

Setting: Tertiary Medical Center.

Patients: Adult participants with MD (n=12) and healthy controls (n=5).

Interventions: Axial T2-weighted and 3D-Fluid Attenuated Inversion Recovery (FLAIR) sequences were obtained at 7T MRI before and four hours after intravenous gadolinium-based contrast agent administration. Following image co-registration and subtraction with Statistical Parametric Mapping (SPM12) and MATLAB, images were uploaded into 3D Slicer for 3D reconstruction and volumetric analysis.

Main Outcome Measures: Voxels measuring 0.3 x 0.3 x 0.5 mm corresponding to areas of endolymph were manually identified and highlighted to create 3D reconstruction, delineation, and volume quantification of the a) utricle and semicircular canals (SCC), b) saccule, c) cochlea, and d) total endolymph (all three compartments).

Results: The median [interquartile range (IQR)] volume in ears affected by MD (n=15) was larger than healthy control ears (n=10) for all measured structures—utricle+SCC: 86.79 mm³ (IQR 83.35-97.94 mm³) vs. 58.32 mm³ (IQR 53.99-71.04 mm³) (p<0.001), saccule: 5.76 mm³ (IQR 4.70-7.64 mm³) vs. 2.80 mm³ (IQR 2.70-4.29 mm³) (p=0.002), cochlea: 62.01 mm³ (IQR 51.75-75.85 mm³) vs. 27.50 mm³ (IQR 26.83-37.39 mm³) (p<0.001), and total endolymph: 149.36 mm³ (IQR 135.99-179.73 mm³) vs. 86.62 mm³ (IQR 80.84-111.05 mm³) (p<0.001).

Conclusions: Ears affected by MD exhibited increased volume in all compartments of the membranous labyrinth compared to healthy controls, with the largest changes observed in the saccule and cochlea. 7T MRI enables in-vivo volumetric measurement of the membranous labyrinth.

Professional Practice Gap & Educational Need: While endolymphatic hydrops (EH) is known to be associated with MD, the underlying cause of MD remains poorly understood. Additionally, in-vivo analysis has traditionally been limited by poor spatial resolution. Therefore, there is a need for advanced imaging techniques to evaluate EH in live human subjects, which could help bridge the gap in understanding the cause, diagnosis, and treatment of MD.

Learning Objective: To demonstrate that EH can be visualized in live human subjects with MD, compared to healthy controls, using advanced imaging techniques.

Desired Result: To enhance understanding of the cause and treatment of MD, ultimately improving patient outcomes.

Level of Evidence - III

Indicate IRB or IACUC: Johns Hopkins IRB00259196

**Risk Factors Associated with Superior Semicircular Canal Dehiscence:
A National Database Study**

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Craig Buchman, MD; Nedim Durakovic, MD*

Objective: To characterize the demographics of patients with superior semicircular canal dehiscence (SSCD) in the US and understand the association with elevated BMI, concurrent CSF leak, obstructive sleep apnea (OSA) or osteoporosis (OP).

Study Design: Retrospective cohort study

Setting: National database (TriNetX) sourced from 67 HCOs in the USA

Patients: Adults diagnosed with SSCD (ICD-10H81.8X9).

Interventions: Evaluation of age, gender, race, BMI > 25, CSF leak, OSA or OP diagnosis.

Main Outcome Measures: 1) Mean age, gender, race 2) BMI > 25 (overweight) 3) CSF leak diagnosis 4) OSA diagnosis 5) OP diagnosis

Results: 11,589 subjects with SSCD were identified. Mean age at diagnosis of SSCD was 53.0 ± 19.5 (mean \pm SD). The majority were female (63.6%, n = 7,375) and white (73.3%, n = 8,490). Subjects with SSCD had a greater risk of being overweight (40.6% vs 28.4%, $p < 0.0001$), having a diagnosis of CSF leak (0.67% vs 0.065%, $p < 0.0001$), having a diagnosis of OSA (11.5% vs 3.65%, $p < 0.0001$) and OP (7.55% vs 2.21%, $p < 0.0001$) compared to age-matched controls (n = 89,720,908).

Conclusions: Patients with SSCD demonstrated significant associations with BMI > 25, a concurrent diagnosis of CSF leak, OSA, and osteoporosis. Findings have implications for the understanding of the etiopathogenesis of SSCD.

Professional Practice Gap & Educational Need: Prior studies have implicated gender, BMI, OSA and underlying predisposition to bony thinning (development of additional CSF leaks and/or concurrent osteoporosis) to the development of SSCD. Herein we leverage a large national database of healthcare outcomes to better characterize these associations.

Learning Objective: 1) Demonstrate the basic demographics (age, gender, race) of subjects with SSCD in the US 2) Identify key associations including elevated BMI, concurrent CSF leak, OSA or OP with the development of SSCD.

Desired Result: Attendees will appreciate the demographics and common risk factors associated with SSCD.

Level of Evidence - IV

Indicate IRB or IACUC: Exempt

Changes in the Neurovascular Unit in Meniere's Disease

Steven D. Curry, MD; Ivan A. Lopez, PhD; Gail Ishiyama, MD; Akira Ishiyama, MD

Hypothesis: Degenerative changes in the neurovascular unit (NVU) in the human spiral ganglia (SG) in patients with Meniere's disease (MD) compared to normal patients underly the clinical manifestations of MD.

Background: Endolymphatic hydrops (EH) is the most consistent pathological correlate of MD, yet the etiology of MD is poorly understood. EH does not explain the changes in permeability of the cochlear blood-labyrinthine barrier seen with delayed contrast MRI or the fluctuations in hearing and impedances of patients with MD with cochlear implants.

Methods: Hematoxylin and eosin sections of the cochlea were obtained from temporal bones of normal patients (n=8, age 50-90 years, 5 male/3 female) and patients diagnosed with MD (n=8, age 44-92, 4 male/4 female). The number of spiral ganglia neurons (SGNs) in each cochlea was estimated. SGNs and blood vessels in the cochlea from normal and MD patients (archival celloidin sections from the same patients) were reliably identified with antibodies against acetylated-3-tubulin and glucose transporter-1 (GLUT-1) respectively and visualized by immunofluorescence and laser confocal microscopy.

Results: There was a significant decrease (50% loss) of SGNs among patients diagnosed with MD compared with age-matched controls ($p<.05$). Immunofluorescence-stained sections showed a marked decrease of blood vessels parallel to the loss of SGNs in MD specimens as compared to the controls.

Conclusions: The decrease of neurons and blood vessels as well as the clinical outcomes showed a correlation between regional damage of the cochlea and patient presentation. These results suggest that the NVU interaction maybe critical to preserve the SGNs in MD and establishes a framework for understanding the etiology and treatment of MD beyond EH.

Professional Practice Gap & Educational Need: Meniere's disease is poorly understood, with limited treatment options available. A better understanding of Meniere's disease at a cellular level may identify targets for therapeutic interventions.

Learning Objective: Understand the changes in the neurovascular unit seen in the cochlea in patients with Meniere's disease based on evidence from human temporal bone research.

Desired Result: Participants will understand how changes in the neurovascular unit contribute to our understanding of the pathophysiology of Meniere's disease beyond its association with EH.

Level of Evidence: Not applicable

Indicate IRB or IACUC: Approved, IRB #22-001587, UCLA Medical Center

NICHOLAS TOROK VESTIBULAR AWARD

Clinical Vestibular Function in Children with and without Sensorineural Hearing Loss

Graham D. Cochrane, MD, PhD; Emily Buss, PhD; Carlton Zdanski, MD

Objective: The objective of the study was to compare vestibular function between children with and without sensorineural hearing loss (SNHL)

Study Design: Cross-sectional, single visit

Setting: Tertiary hospital outpatient specialty clinic

Patients: Forty children ages 6-12 with (n=20) and without (n=20) SNHL of any severity, aided or unaided, without significant physical limitations that would impair their ability to complete balance and gait tasks.

Interventions: All participants completed a one-hour clinical vestibular protocol consisting of measures of static and dynamic balance, vestibular-ocular reflex function, and subjective visual vertical. Parents completed a self-report survey related to their child's birth, motor development, and current activity levels.

Main Outcome Measures: Participants completed the Modified Clinical Test of Sensory Integration on Balance (mCTSIB), a Dynamic Visual Acuity (DVA) task, a Subjective Visual Vertical (SVV) bucket task, the Functional Gait Assessment (FGA), and the Emory Modified Clinical Vestibular Chair Test (EMCVCT).

Results: Children with SNHL demonstrated significantly worse performance on FGA, DVA, SVV, and EMCVCT compared to children without SNHL. Parents of children with SNHL more commonly reported their child was sensitive to motion, clumsy, and less physically able than their peers. Children with bilateral SNHL and children with greater severities of hearing loss were more likely to demonstrate vestibular deficits across multiple tasks.

Conclusions: Deficiencies in vestibular function can be identified with low-cost tasks in children with SNHL and are associated with severity of hearing loss. Parents of children with SNHL report their children have greater physical limitations and trouble keeping up with peers. Future research should investigate whether these identifiable deficits can be improved with vestibular therapy and whether improvement in these functions may help alleviate those perceived physical limitations.

Professional Practice Gap & Educational Need: Vestibular deficits are identifiable in children with SNHL with simple tools clinics could use to screen and these deficits are associated with perceived physical limitations of the children. More children with SNHL should be screened for vestibular deficits as there are low-cost screening tools available and proper identification may aid in their motor development and physical activity levels with peers. More accessible vestibular screening protocols may help children with SNHL address balance difficulties and engage more actively with peers.

Learning Objective: Children with SNHL have easily identifiable deficits in vestibular function and are more likely to have perceived sensitivity to motion compared to children without SNHL.

Desired Result: These results should encourage providers to screen for vestibular deficits in children with SNHL and refer them appropriately to services such as vestibular therapy. Vestibular screening may not need to be done in a specialized, high-cost clinic to identify children who may benefit from therapy.

Level of Evidence – Level 3

Indicate IRB or IACUC: University of North Carolina at Chapel Hill IRB #23-2458

Otic Capsule Demineralization and Hearing Outcomes of Stapes Surgery for Otosclerosis

*Akira Kimura, MD; Chihiro Yagi, MD, PhD; Yuka Morita, MD, PhD; Tatsuya Yamagishi, MD, PhD
Shinsuke Ohshima, MD, PhD; Shuji Izumi MD, PhD; Arata Horii, MD, PhD*

Objective: To correlate the otic capsule demineralization with pre-/post-operative hearing parameters in otosclerosis.

Study Design: Retrospective study

Setting: University hospital

Patients: 181 consecutive ears with otosclerosis that underwent stapes surgeries from January 2003 to December 2020.

Interventions: Demineralization of the otic capsule was examined by CT.

Main Outcome Measures: Demineralization loci were examined using high resolution computed tomography seen as hypodense area in the otic capsule. Hearing parameters before and 12 months after surgeries were compared between demineralization (+) and (-) ears and were also correlated with number of loci in demineralization (+) group.

Results: There were eighty-five ears in the demineralization (-) group, while one or more hypodense areas were found in 96 ears (53%) (demineralization (+) group). Preoperative air-conduction (AC) threshold was significantly worse due to larger air-bone gap (ABG) in demineralization (+) group than that in demineralization (-) group, while there was no difference in bone-conduction (BC) thresholds between groups. Postoperative AC was significantly worse due to larger ABG in demineralization (+) group, whereas there were no significant differences in postoperative BC and closure of ABG between groups. No significant relationship was found between the number of demineralization loci (single to triple) and pre-/post-operative hearing parameters in demineralization (+) group.

Conclusions: While both pre- and post-operative AC were worse in demineralization (+) group than those of (-) group, significant and similar closure of ABG was obtained in demineralization (+) group as in (-) group. Otic capsule demineralization may attribute to possible third window syndrome and ABG that cannot be fixed by stapes surgeries.

Professional Practice Gap & Educational Need: The role of demineralization for otosclerosis has no consensus.

Learning Objective: The extent of demineralization observed on CT does not correlate with hearing outcomes after stapes surgery.

Desired Result: Learning the significance of the demineralization

Level of Evidence - Level IV

Indicate IRB or IACUC: Niigata University Hospital (No. 2020-0446)

Pseudomembranes and Tissue Plugs in the Round Window Niche: Implications for Inner Ear Drug Delivery

*Nicole Kim, BA; Liliya Benchetrit, MD
Anbuselvan Dharmarajan, MD, MPH; Alicia M. Quesnel, MD*

Hypothesis: We hypothesize that a significant proportion of human temporal bone specimens has anatomical obstructions in the round window niche, including pseudomembranes and tissue plugs, and certain patient factors predict these obstructions.

Background: Therapies for sensorineural hearing loss (SNHL) may utilize intratympanic drug delivery through the round window membrane (RWM) of the cochlea. However, anatomical barriers like pseudomembranes and tissue plugs may impede access if drug delivery relies on diffusion across the RWM or intracochlear access without surgical dissection of the RWM. This study, the largest survey of RWM obstructions to date, aims to assess their prevalence and identify factors predictive of obstructions relevant to inner ear drug delivery.

Methods: Temporal bone cases with SNHL and normal hearing controls were selected from Massachusetts Eye and Ear, University of Pittsburgh, and Hospital Nacional de Niños using the NIDCD National Temporal Bone Registry. Exclusion criteria included prior otologic surgery, middle ear disease, and congenital ear anomalies. Histopathologic analysis was performed.

Results: 279 temporal bones were analyzed, including 215 with SNHL and 64 controls. RWM obstruction prevalence was 37.3% for pseudomembranes and 28.3% for tissue plugs, with no significant differences between SNHL and controls. Multivariate analysis showed age over 18 was significantly associated with pseudomembranes (Odds Ratio [OR]: 8.45), while age 18 or younger was associated with tissue plugs (OR: 2.63). Hearing loss due to Meniere's disease (OR: 0.08) or ototoxicity (OR: 0.13) predicted a lower likelihood of pseudomembranes.

Conclusions: Pseudomembranes and tissue plugs are common in the RWN, with 2/3 of ears demonstrating either obstruction. Adults were more likely to have pseudomembranes, while pediatric patients were more likely to have tissue plugs. These findings may impact inner ear drug delivery strategies.

Professional Practice Gap & Educational Need: This research highlights the prevalence of RWM obstructions, including pseudomembranes and tissue plugs, and begins to elucidate associations of obstruction types with age and specific hearing loss etiologies. It aims to stimulate discussion on potential anatomical assessment protocols for more effective inner ear drug delivery.

Learning Objective: Recognize that the presence of RWM obstructions, including pseudomembranes and tissue plugs, is relatively common. Identify age and certain hearing loss etiologies as factors relevant to the prevalence of obstructions.

Desired Result: Improve awareness of types of RWM obstruction, which may have implications for intratympanic or intracochlear approaches to inner ear drug delivery.

Level of Evidence – III

Indicate IRB or IACUC: Protocol # 2021P001593 (Human Temporal Bone Pathology of Sensorineural Hearing Loss, Otosclerosis, and Post Cochlear Implantation), Mass Eye and Ear U24 grant

Are Jak Inhibitors Contributing to Ototoxicity? Investigating Their Role in Aminoglycoside-Induced Damage

Jonathan Fleegel; Marisa Zallocchi, PhD

Hypothesis: Inhibition of the JAK-STAT signaling pathway, either pharmacologically with JAK or genetically through JAK2 knockout, potentiates the ototoxic effects of aminoglycosides. JAK2 knockout mice will show increased susceptibility to aminoglycoside-induced ototoxicity.

Background: Aminoglycosides are effective antibiotics but are linked to irreversible ototoxicity. Although the mechanisms underlying aminoglycoside-induced ototoxicity are being explored, the role of specific signaling pathways in modulating this ototoxicity remains unclear.

Methods: Auditory function was assessed using auditory brainstem response (ABR) and distortion product otoacoustic emissions (DPOAE) across multiple frequencies. Vestibular function was evaluated with vestibular short-latency evoked potentials (VsEP). C57BL/6J Cdh23-corrected mice were treated with JAK inhibitors (mometinib 20 mg/kg, tofacitinib 20 mg/kg, upadacitinib 10 mg/kg) via oral gavage for 14 days, alone or combined with kanamycin (600 mg/kg, S.C.). In a separate experiment, Pax2-Cre; JAK2^{fl/fl} knockout mice were treated with kanamycin, or tobramycin (200 mg/kg, S.C.). Vestibular testing was conducted for all animals exposed to tobramycin.

Results: All JAK inhibitors significantly potentiated the ototoxic effects of kanamycin, as evidenced by elevated ABR and DPOAE thresholds across multiple frequencies. JAK2 knockout in mice treated with kanamycin or tobramycin exhibited significant hearing loss and vestibular dysfunction compared to controls. RM-ANOVA with Tukey's HSD was used for statistical testing.

Conclusions: Systemic administration of JAK inhibitors enhances the ototoxic effects of kanamycin, emphasizing the role of the JAK-STAT pathway in auditory protection. JAK2 knockout mice were particularly vulnerable to aminoglycoside-induced hearing loss and vestibular dysfunction. These findings highlight a critical role for JAK-STAT signaling in mitigating aminoglycoside ototoxicity, suggesting that patients treated with JAK inhibitors could be at higher risk for hearing loss when exposed to aminoglycosides. Monitoring for ototoxicity may be beneficial in these populations.

Professional Practice Gap & Educational Need: Jak inhibitors are a rapidly growing class of drugs that may increase the susceptibility of patients to ototoxic effects of various drugs. A formal clinical assessment through ototoxicity monitoring programs would be beneficial in identifying the risk associated with the use of these drugs and adverse hearing and vestibular consequences.

Learning Objective: The identification of unknown at-risk clinical cohorts for ototoxicity.

Desired Result: Clinical monitoring and risk assessment in patients exposed to Jak inhibitors.

Level of Evidence – I

IACUC Protocol number(s): Creighton University IACUC: 1148 and 1251.

Computer Vision-Based Extraction of Structured Data from Scanned Audiograms in the Electronic Health Record

Peter R. Dixon, MD, MSc; Ruoyu Yang, PhD; Dana Mae Salvador, BSc; Carl Ehrett, PhD

Objective: A barrier to leveraging real-world data from electronic health records (EHRs) for hearing research is the inability to confirm hearing loss diagnosis and quantify its severity. This study aims to develop a machine learning-based approach to interpret scanned audiogram test sheets from the EHR at scale.

Study Design: A contour-based computer vision (CV) method to extract structured data from scanned audiograms with hand-drawn symbols stored in the EHR

Setting: Tertiary academic health network

Patients: Models were trained on 907 audiograms (Jan 1, 2014 – Dec 31, 2022) selected using stratified random sampling to ensure balanced representation of normal hearing, bilateral sensorineural, asymmetric sensorineural, conductive, and mixed hearing loss. Audiograms were download, de-identified, and pure tone thresholds manually extracted to serve as the ground truth.

Interventions: The CV pipeline accepted audiogram PDF files as inputs and returned frequency (Hz) and threshold (dB HL) estimates for all symbols.

Main Outcome Measures: Mean absolute error (MAE) on 30 test audiograms randomly sampled from the EHR comparing CV-generated and ground truth values

Results: The pipeline has three steps: (1) Image cropping to segment the pure tone threshold plot, (2) Pattern detection using grayscale conversion and contrast enhancement preprocessing, followed by contour identification to calculate symbol center points, and (3) Pattern coordinate calibration to identify axis labels and translate center points from pixel to audiogram coordinates. In the test set, the MAE was 136 Hz for frequency and 1.3 dB HL for threshold across all symbols.

Conclusions: The CV method accurately estimates symbol coordinates for pure tones. Future work will expand the pipeline for automated identification of audiograms within EHR media and integrate deep learning algorithms, such as convolutional neural networks, to enhance pattern classification and scalability for hearing health research.

Professional Practice Gap & Educational Need: Despite the growing availability of EHR data, clinicians and researchers face challenges in identifying hearing loss and quantifying its severity due to the unstructured nature of scanned audiograms stored as images. There is a need for education on how machine learning and computer vision techniques can automate the extraction of structured data from these audiograms, enabling more efficient research and clinical workflows. This session addresses the gap by introducing practical approaches for leveraging automated methods to enhance hearing health services research.

Learning Objective: Understand how computer vision can extract data from scanned audiograms and explore its applications in hearing health research using EHR data.

Desired Result: Attendees will be able to describe the potential of machine learning-based methods for audiogram interpretation, recognize how automated data extraction can improve research workflows, and identify future directions for integrating deep learning in hearing health studies.

Level of Evidence - Level IV

Indicate IRB or IACUC: Exempt

MICHAEL E. GLASSCOCK SCIENTIFIC MERIT AWARD

Conductive Hearing Loss is Associated with Dementia in the All of Us Research Program

*Hannah N. W. Weinstein, BA; Lauren H. Tucker BA; Michael W. Denham, MD, MS, MPhil
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Objective: Sensorineural hearing loss (SNHL) has been associated with cognitive impairment and dementia. However, because SNHL contains a neural component, it is difficult to exclude reverse causation whereby dementia causes worse SNHL. Conductive hearing loss (CHL), a purely peripheral phenomenon, would not have this limitation. We investigate the association between CHL and dementia in a large national cohort.

Study Design: Cross-sectional epidemiologic study

Setting: The NIH All of Us Research Program, which includes aggregated data from the electronic health records of voluntary participants.

Patients: ≥ 18 years old ($n=399,927$).

Main Outcome Measures: Dementia defined by ICD-10 codes (F01, F03, G30-32).

Methods: The exposure was CHL defined by ICD-10 codes (H90.0-90.2). The odds of dementia in subjects with and without CHL were assessed with multivariable regression, controlling for potentially confounding variables (age, sex, education).

Results: The mean (SD, range) age was 56 years (± 17 , 20-124). 242,911 (60.7%) participants identified as female. The cohort included 1,274 (0.3%) individuals with CHL and 398,653 (99.7%) without CHL. Overall, 6,425 (1.6%) participants had dementia. After controlling for covariates, the odds of dementia were 2.7 times (95% CI 2.1-3.4; $p < 0.0001$) higher for those with CHL compared to those without CHL. The odds remained unchanged (OR=2.7, CI 2.1-3.5; $p < 0.0001$) when examining only participants > 60 years old who are in an age range at higher risk for dementia.

Conclusions: In the All of Us dataset, CHL was strongly associated with dementia. This supports CHL as a risk factor for cognitive disorders, and the theory that any sensory deprivation to the brain can be detrimental. As it is implausible for dementia to cause CHL, a purely peripheral process, the possibility of reverse causation as an explanation is eliminated.

Professional Practice Gap & Educational Need: The association between CHL and cognition is emerging, with additional investigation needed to improve understanding, monitoring, diagnosis, and management of the association between these chronic conditions.

Learning Objective: Participants will understand the association between CHL and dementia, while considering the utility and pitfalls of using national databases and EHRs in large epidemiologic investigations.

Desired Result: Participants will better understand the relationship between CHL and cognition.

Level of Evidence: IV

Indicate IRB or IACUC: All study procedures were confirmed as meeting criteria for non-human subjects research by the All of Us Research Program IRB.

Safety of Middle Cranial Fossa Repair of Spontaneous Cerebrospinal Fluid Leaks in Superobese Patients

*Hunter Elms, MD; Douglas Totten, MD; Evan Cumpston, MD
Charles Yates, MD; Rick Nelson, MD, PhD*

Objective: Determine how complication rates vary according to body mass index (BMI) in middle cranial fossa (MCF) repairs of spontaneous cerebrospinal fluid leaks (sCSF-L)

Study Design: Retrospective case series

Setting: Tertiary academic

Patients: Adults with MCF sCSF-L

Interventions: Middle fossa craniotomy skull base repair

Main Outcome Measures: 90-day rates of any perioperative complications according to American Surgical Association severity (grades 1-5) and rates of ipsilateral persistent leak within 90 days of initial surgery

Results: 125 patients (64% female), aged 28-81 (57.7 ± 11.3) years, underwent MCF repairs of 143 spontaneous tegmental leaks (50% right-sided, 36% left-sided, 14% bilateral). Patient BMIs ranged from 23.6-85.4 (39.5 ± 10.1) kg/m². 2.4% of patients were normal weight, 11.2% were overweight, 23.2% class 1 obese, 21.6% class 2, and 41.6% class 3. Successful initial surgical repair was accomplished in 96.8% of cases, with 91.2% of cases complication-free. When comparing BMI classes to each other, as well as comparing class 3 obese patients to all others, there were no significant differences in overall complication rates, severity scores, or rates of persistent CSF leaks.

Conclusions: In a population where 41.6% of patients had class 3 obesity, BMI and class 3 obesity did not significantly affect surgical success, complication rates, or severity of perioperative complications from MCF sCSF-L repairs. MCF repair had a first-case success rate >95%, with complication rates <10%.

Professional Practice Gap & Educational Need: level of surgeon willingness to offer MCF repair of sCSF-L to very obese patients

Learning Objective: Evaluate the effect of BMI on safety profile of MCF sCSF-L repair

Desired Result: Consideration of surgery for very obese patients who meet criteria

Level of Evidence: V

Indicate IRB or IACUC: Exempt

A Systematic Review and Meta-Analysis of Meningitis Risk Reduction after Repair of Spontaneous Lateral Skull Base Cerebrospinal Fluid Leaks

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Estephania Candelo, MD; Zhen Wang, PhD; Tara Brigham, MLIS, AHIP-D; Mallory Raymond, MD*

Objective: To estimate the relative risk of meningitis after repair of spontaneous lateral skull base cerebrospinal fluid (CSF) leaks through meta-analysis of published studies.

Data sources: PubMed, Medline, Embase, Cochrane, SCI-EXPANDED, ESCI, Epistemonikos, and WHO Global Index Medicus were queried for terms including and related to meningitis, CSF leak, surgical repair, and spontaneous from inception to April 02, 2024.

Study selection: Prospective and retrospective studies in any language reporting either preoperative or postoperative rates of meningitis in adult patients with spontaneous lateral skull base CSF leaks were included.

Data extraction: Two reviewers independently screened 2,564 studies identified by the search and extracted data and evaluated risk of bias in 57 studies that met inclusion criteria. Risk of bias was assessed using the JBI Critical Appraisal Checklist for Case Series and the Newcastle-Ottawa Quality Assessment Scale for Cohort Studies.

Data synthesis: Of 1,310 patients who met inclusion criteria, 1,239 underwent CSF leak repair. 52 studies reported rates of preoperative meningitis, and 27 studies reported rates of postoperative meningitis. 22 studies with a total of 511 patients reported both rates. The average follow-up period of 17 studies reporting duration of follow-up was 23.1 months. A random effects generalized linear mixed model was used to pool and compare logit transformed risk of meningitis before and after repair. The pre-operative pooled risk of meningitis was 0.16 (95% CI: 0.13 to 0.25, $I^2=40.57\%$). The post-operative pooled risk was considerably lower at 0.01 (95% CI: 0.01 to 0.14, $I^2=4.16\%$). With substantial heterogeneity ($I^2=62.94\%$), we observed a relative risk of meningitis after repair of 0.02 (95% CI: 0.00 to 0.79, $p=0.04$).

Conclusions: Surgical repair significantly reduces the risk of meningitis in adults with spontaneous lateral skull base CSF leaks.

Professional Practice Gap & Educational Need: The risk reduction of meningitis from undergoing surgical repair of spontaneous CSF leaks is unknown. Without an estimate of both the pre- and postoperative risk of meningitis, counseling patients, especially those who are asymptomatic, on the need for surgical repair can be challenging. Pooled rates of pre- and postoperative meningitis can help to guide decisions on whether to proceed with surgical repair.

Learning Objective: To understand the relative risk of meningitis in adult patients after repair of spontaneous lateral skull base CSF leak.

Desired Result: Clinicians can quote the estimated rate of pre- and postoperative meningitis in patients with spontaneous lateral skull base CSF leak and the relative risk after surgical repair.

Level of Evidence: Level III.

Indicate IRB or IACUC: Exempt.

Intracranial Dural Arteriovenous Fistula Can Mimic Sigmoid Sinus Wall Anomalies Induced Pulsatile Tinnitus: Caution Before Considering It's Venous

Yue-Lin Hsieh, MD, PhD; Xu Liu, MD; Wuqing Wang, MD, PhD

Objective: To highlight that dural arteriovenous fistula (DAVF) can exist in subjects with unilateral vascular pulsatile tinnitus (PT), positive internal jugular vein (IJV) compression tests, and radiologic evidence of sigmoid sinus wall anomalies (SSWA). 2) To introduce the "moth-eaten sigmoid plate" sign and emphasize the importance of retroauricular compression in diagnosing PT.

Study Design: Retrospective data analysis.

Setting: Multi-institutional tertiary university medical centers.

Patients: 81 subjects with PT as sole symptom and intracranial DAVF.

Interventions: High-resolution temporal bone CT and magnetic resonance angiography (MRA) were conducted.

Main Outcome Measures: The moth-eaten sigmoid plate sign and DAVF-induced SSWA were defined, and their correlation with PT duration was studied.

Results: Significant differences were observed between ipsilateral IJV and retroauricular compression outcomes in DAVFs located at the transverse-sigmoid sinus ($p < 0.01$) and hypoglossal canal ($p < 0.01$). Among 71 subjects with CT data, the moth-eaten sign was found in 29 of 37 subjects (78.4%) with DAVFs at the transverse-sigmoid sinus. SSWA and JB anomalies were observed in 40.8% of subjects. PT duration significantly differed between subjects with SSWA and those without SSWA ($p < 0.01$).

Conclusions: The presence of SSWA on CT and a positive IJV compression test should not be considered conclusive for diagnosing venous PT. The "moth-eaten sigmoid plate" sign on non-contrast CT and positive retroauricular compression are strong indicators of DAVF as the primary cause of PT.

Professional Practice Gap & Educational Need: 1) A positive IJV compression test and non-contrast CT presence of SSWA is not sufficient for diagnosis of venous PT. 2) The "moth-eaten sigmoid plate" sign on non-contrast CT and positive retroauricular compression are strong indicators of DAVF as the primary cause of PT.

Learning Objective: Understanding the significance of conducting MRA examinations for patients with vascular PT is crucial to prevent misdiagnosing the vascular causes of PT. This understanding is essential for accurate diagnosis and appropriate treatment planning.

Desired Result: Improved diagnosis rates in treating PT caused by DAVF and decreased incidences of unnecessary and potentially harmful surgical interventions.

Level of Evidence – III

Indicate IRB or IACUC: The present study was approved by the medical ethics committee of the Eye, Ear, Nose, and Throat Hospital of Fudan University (No. 2024125) on January 2024.

Risk Factors and Developmental Patterns of Spontaneous New Encephaloceles Following Initial Repair

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Objective: To identify risk factors and temporal patterns for the spontaneous development of new encephaloceles following initial repair of skull base defects.

Study Design: Retrospective case-control study

Setting: Tertiary academic medical center

Patients: Consecutive adult patients with skull base defects repaired from October 2012 to October 2024.

Interventions: Electronic medical records of qualifying patients were searched for relevant demographics, medical history, disease characteristics, surgical approach, local and non-local encephalocele recurrence, and postoperative outcomes. Analysis was performed using Mann-Whitney U, Kruskal-Wallis, and Fischer's exact tests.

Main Outcome Measures: Risk factors for, median time to, and location of recurrent skull base encephaloceles.

Results: 176 adults met inclusion criteria. Mean age was 56.8 years old (SD 12.4 years), average BMI was 35.2 kg/m² (SD 8.5 kg/m²), and 110 (62.5%) patients were female. 23 (13.1%) patients developed new encephaloceles after initial repair, with a median time to recurrence of 0.86 years (IQR [0.13, 2.63]). Of these newly developed skull base encephaloceles, 3 (13.0%) were contralateral, 7 (30.4%) were anterior, and 13 (56.5%) were ipsilateral. History of idiopathic intracranial hypertension (IIH) (OR 12.1 [4.4, 33.4]; p<0.001) was significantly associated with new encephaloceles; bilaterality (OR 2.6 [0.97, 7.1]; p=0.07), revision surgery (OR 3.0 [0.86, 10.6]; p=0.09), and history of sleep apnea (OR 2.1 [0.86, 5.2]; p=0.10) approached significance. BMI, when calculated both including and excluding patients with IIH, was not significant.

Conclusions: Although new spontaneous encephaloceles are uncommon following initial repair, the management of comorbid medical conditions and routine postoperative surveillance is crucial, especially for higher-risk individuals such as those with IIH. Higher-powered studies are warranted to further explore associations that approached significance in this study, such as sleep apnea, bilaterality, and revision surgery.

Professional Practice Gap & Educational Need: Skull base encephaloceles have become an increasingly common pathology in adults. It is critical to identify patients who are at risk for recurrent or new encephaloceles following an initial repair and elucidate their risk factors, developmental timeframe, and anatomic characteristics.

Learning Objective: 1) Recognize risk factors associated with locally and non-locally recurrent encephaloceles and 2) familiarize oneself with the typical timeframe and pattern in which they recur.

Desired Result: Synthesize pre-existing knowledge about encephalocele symptoms with empiric data about risk factors and temporal patterns to better monitor patients for local and non-local recurrence.

Level of Evidence: IV – Retrospective case-control study

Indicate IRB or IACUC: University of Pennsylvania Institutional Review Board (#856621)

Factors Associated with Cochlear Implant Utilization in Adults with Single-Sided Deafness

*Matthew J. Wu, MD; Amit Walia, MD, MSCI; Shubhanjali Minhas, BA; Jill Firszt, PhD
Jacques A. Herzog, MD; Craig A. Buchman, MD; Matthew A. Shew, MD*

Objective: Determine daily utilization of cochlear implants (CI) in adults with single-sided deafness (SSD)

Study Design: Retrospective, cross-sectional study (7/2018-9/2024)

Setting: Tertiary referral center

Patients: Adult CI recipients implanted for SSD (pure tone average [PTA] ≤ 35 dB hearing loss [HL] in non-implanted ear)

Interventions: CI

Main Outcome Measures: Mean daily CI usage (in hours) determined through datalogging and speech perception measures at pre-op, 3- and 6-month intervals.

Results: One-hundred twenty-four adult patients underwent CI for SSD. The most common HL etiologies included sudden hearing loss (n=52, 41.9%) and unknown (n=42, 33.9%). Median length of audiometric follow-up was 1 year (range 0.1-8.1). Of the 112 patients with available datalogging, 71 (63.4%) were full-time users (≥ 6 hours/day). Analyses of the 41 partial users showed 41 (36.6%), 25 (22.3%), 12 (10.7%) and 2 (1.8%) patients used the device <6 , <4 , <2 , or 0 hours/day, respectively. A logistic regression model was used to identify factors associated with CI utilization (≥ 6 hours/day). Age at implantation ($\beta=1.02$; 95%CI: 0.99–1.05), preoperative PTA ($\beta=1.00$; 95%CI: 0.98–1.03), HL duration ($\beta=1.00$; 95%CI: 0.96–1.05), and preoperative CNC word scores ($\beta=1.01$; 95%CI: 0.97–1.05) were not associated with utilization. A separate logistic regression was used to identify factors associated with use <2 hours/day. While controlling for duration of HL, 6-month CNC word scores ($\beta=0.99$; 95%CI: 0.94–1.04) were not associated with non-use.

Conclusions: Non-utilization of CI in SSD patients appears low although more limited use (<6 hours/day) is common. Patient factors did not explain more limited utilization. Future work is needed to better understand more limited daily utilization of CI in SSD patients.

Professional Practice Gap & Educational Need: Given the relatively recent expansion of indications for adult CI to include SSD, large scale analyses of daily utilization in this population are needed to inform patient and clinician expectations.

Learning Objective: Understand CI utilization rates in adults with SSD (full-time usage, partial usage, and non-usage).

Desired Result: Physicians will understand that traditional CI performance metrics may not explain under-usage in the SSD population. Datalogging in SSD patients may also not capture quality of life associations driving satisfaction. Future work needs to explore factors associated with use to improve patient counseling and selection.

Level of Evidence - IV

Indicate IRB or IACUC: Washington University in St. Louis IRB #202408206 (8/30/24).

NEUROTOLOGY FELLOW AWARD

Factors Associated with Delayed Loss of Residual Acoustic Hearing Following Cochlear Implantation

*Michael W. Canfarotta, MD; Ankita Patro, MD, MS; Natalie Schauwecker, MD; Connie Ma, MD
Aaron C. Moberly, MD; Jourdan T. Holder, AuD, PhD; Elizabeth L. Perkins, MD*

Objective: To identify factors associated with delayed loss of preserved low-frequency acoustic hearing following cochlear implantation.

Study Design: Retrospective cohort.

Setting: Tertiary referral center.

Patients: One hundred fifty-four adult cochlear implant (CI) recipients with initial “functional” hearing preservation (low-frequency pure-tone average [LFPTA; 125, 250, and 500 Hz] ≤ 80 dB HL) at 1 month postoperatively.

Interventions: Cochlear implantation with a straight ($n = 114$), precurved non-styled electrode array ($n = 26$), or precurved styled electrode array ($n = 14$).

Main Outcome Measures: Binary logistic regression was performed to investigate variables associated with delayed loss of functional residual hearing during the first year after implantation.

Results: Among 154 CI recipients, 114 (74%) retained functional residual hearing through the first year postoperatively. Patients with a younger age at surgery (odds ratio [OR], 0.96 [95% CI, 0.93-0.99]; $p = 0.034$), precurved styled electrode array (OR, 5.5 [95% CI, 1.34-22.55]; $p = 0.018$), and higher LFPTA at the 1-month postoperative interval (OR, 1.31 [95% CI, 1.18-1.45]; $p < 0.001$) were more likely to experience delayed loss of functional residual hearing. There were no significant effects of biological sex, diabetes, postoperative steroid use, or initial LFPTA shift due to surgery ($p \geq 0.15$).

Conclusions: For the present cohort of CI recipients with initial low-frequency hearing preservation, younger age at surgery, implantation with a precurved styled electrode array, and a higher LFPTA at 1 month postoperatively were associated with delayed loss of functional hearing. Additional studies are needed to determine the mechanisms by which these factors place patients at risk for delayed hearing loss.

Professional Practice Gap & Educational Need: While variables associated with acute and long-term hearing preservation have been extensively studied, there is limited data on factors contributing to delayed hearing loss in CI recipients that initially preserve low-frequency acoustic hearing. As we aim to reduce the postoperative inflammatory response with the use of robotics and drug-eluting electrode arrays, it will be imperative to understand baseline characteristics that are associated with delayed hearing loss.

Learning Objective: (1) Understand previously identified factors associated with acute and long-term hearing preservation. (2) Describe factors that are associated with delayed hearing loss in CI recipients that initially preserve low-frequency acoustic hearing.

Desired Result: At the conclusion of this presentation, providers should have a better understanding of factors related to delayed hearing loss following cochlear implantation and possible mechanisms to explain these findings.

Level of Evidence – Level IV

Indicate IRB or IACUC: IRB #240876, Vanderbilt University

Perimodiolar Electrode Locations Outperform Lateral Wall Arrays when Controlling for Cochlear Health and Speech Processing Strategy

*Amit Walia, MD, MSCI; Matthew A. Shew, MD; Amanda Ortmann, PhD
Matthew Wu, MD; Shannon Lefler, AuD; Jacques A. Herzog, MD; Craig A. Buchman, MD*

Objective: To assess how variations in scala tympani (ST) electrode position affect speech-perception performance, controlling for cochlear health and stimulation strategy.

Study Design: Retrospective cohort study

Setting: Tertiary referral center

Patients: Ninety-eight postlingual adult cochlear implant (CI) recipients participated—21 received lateral wall electrodes (CI624;20-mm) and 77 received perimodiolar electrodes (CI632;18.4-mm). All patients used the Advanced Combination Encoder (ACE) strategy with ST insertions. Cochlear health was assessed via pre-insertion round window electrocochleography-total response (ECochG-TR). Postoperative 3D CT-reconstructions determined electrode proximity to the modiolus (wrapping factor, WF) and angular insertion depth (AID).

Main Outcome Measures: Speech-perception testing at 6-months post-activation (CNC)

Results: A strong positive correlation existed between ECochG-TR and CNC scores ($r=0.61$; 95% CI:0.42-0.84). WF showed a weak negative correlation with CNC ($r=-0.36$; 95% CI:-0.53 to -0.16; tighter WF with better performance), while AID demonstrated a weak positive correlation ($r=0.29$; 95% CI:0.08-0.47; deeper AID with better performance). A regression model using ECochG-TR underestimated performance for electrodes with tighter WF and deeper AID, and overestimated performance for lateral wall location and shallower insertions. A multivariate regression combining ECochG-TR, AID, and WF improved speech perception estimation significantly ($R^2=0.51$; $P=0.001$).

Conclusions: Electrode location within the ST varies significantly, even within the same electrode type; perimodiolar arrays can reside along the lateral wall if over-inserted, and lateral wall arrays can approach the modiolus. Incorporating electrode location and ECochG-TR into speech-perception models is crucial for understanding differences between electrode types. When controlling for these variables using the same processing strategy, our study shows that perimodiolar electrodes with tighter WFs outperform lateral wall electrodes on 6-month CNC scores. This underscores the importance of controlling these factors to determine optimal electrode location.

Professional Practice Gap & Educational Need: Perimodiolar electrodes are designed to assume a position closer to the modiolus, in closer proximity to the neural elements for more precise stimulation. However, the wide variability in CI performance makes the numerous comparative studies on electrode type and location inconclusive. Controlling for cochlear health using electrocochleography-total response (ECochG-TR) allows for logical segmentation of the recipient population, thereby enabling comparisons of variables such as device type, electrode location, processing strategy, and many others. This study investigates whether controlling for stimulation strategy (i.e., Advanced Combination Encoder strategy) and cochlear health (i.e., ECochG-TR) can reveal the impact of intrascalar electrode location on performance. Notably, not all perimodiolar electrodes maintain close proximity to the modiolus. Without proper pull-back techniques, these arrays can be displaced to an anti-modiolar location, undermining the intent of the precurved design.

Learning Objective: To evaluate the impact of perimodiolar electrode proximity (i.e., WF) and angular insertion depth on speech-perception performance, emphasizing the impact of cochlear health and electrode position in CI studies.

Desired Result: Investigators will recognize the importance of cochlear health and electrode position when designing studies to improve CI outcomes—such as mapping, auditory rehabilitation, electrode selection, steroid use, and more.

Level of Evidence - IV

Indicate IRB or IACUC: Washington University in St. Louis IRB #202007087 (5/16/23).

One or Two Cochlear Implants? A Comparison of Bimodal Hearing Versus Bilateral Cochlear Implantation in a Large Cohort of Traditional Candidates

*Ankita Patro, MD, MS; Michael W. Canfarotta, MD; Natalie Schauwecker, MD; Elizabeth L. Perkins, MD
David S. Haynes, MD; René H. Gifford, PhD; Aaron C. Moberly, MD*

Objective: 1) To identify preoperative factors that impact the pursuit of bimodal hearing versus bilateral implantation among traditional cochlear implant (CI) candidates, and 2) To compare postoperative outcomes in these two groups.

Study Design: Retrospective cohort.

Setting: Tertiary referral center.

Patients: 499 adult CI recipients (386 bimodal, 113 bilateral CI users) with preoperative best-aided AzBio scores in quiet \leq 60% in both ears.

Main Outcome Measures: Demographics; pure-tone average (PTA); AzBio in quiet; Speech, Spatial, and Qualities of Hearing Scale (SSQ); datalogging.

Results: Preoperatively, compared to bimodal recipients, bilateral CI recipients were significantly younger (58.6 vs. 67.7 years, $p < 0.001$) and had worse audiometric thresholds (PTA in worse hearing ear: 95.4 vs. 88.4 dB HL, $p < 0.001$; PTA in better hearing ear: 90.0 vs. 74.3 dB HL, $p < 0.001$). Bilateral CI recipients had worse AzBio in quiet scores at initial CI evaluation: worse hearing ear (8.6% vs. 12.9%, $p = 0.009$); better hearing ear (19.3% vs. 25.7%, $p = 0.003$); and binaural (27.2% vs. 35.6%, $p = 0.002$). Gender, race, duration of deafness, and preoperative SSQ scores were similar between the two groups ($p > 0.05$). Compared to bilateral CI users after their second CI, bimodal patients had equivalent device usage (10.7 vs. 11.8 hours, $p = 0.109$) but lower AzBio in quiet scores in the everyday listening condition at 12 months (60.0% vs. 73.3%, $p = 0.005$). After controlling for datalogging, age, and preoperative scores on multivariable analysis, 12-month speech recognition scores remained higher in the bilateral CI group.

Conclusions: Traditional CI candidates who pursue bilateral implantation over a bimodal hearing configuration are younger and have worse preoperative hearing and speech recognition. Even after controlling for age and baseline scores, long-term speech recognition may be superior with bilateral CIs.

Professional Practice Gap & Educational Need: To our knowledge, the impact of preoperative factors on whether patients pursue one or two CIs, when both ears qualify, has not been reported. While the benefits of bimodal hearing and bilateral cochlear implantation have been explored in the literature in smaller sample groups, this is the first study to compare the benefits of bimodal hearing versus bilateral CIs in a large cohort of patients.

Learning Objective: 1) To understand factors that influence when patients pursue a bimodal hearing configuration versus bilateral cochlear implants, 2) To describe differences in postoperative outcomes between bimodal and bilateral CI users.

Desired Result: Providers will have knowledge about the impact of age and preoperative hearing on the pursuit of bilateral CIs when both ears qualify during initial evaluation. Moreover, they will understand differences in speech recognition performance between bimodal and bilateral CI users. These findings can help counsel patients in both the preoperative and postoperative settings.

Level of Evidence: Level IV – Historical cohort or case-controlled studies.

Indicate IRB or IACUC: IRB Exempt (240876, Vanderbilt University, approved 8/23/24).

Evaluating the Impact of Cochlear Implantation on Cognitive Outcomes in Older Adults: A 5-year Follow-up

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Neil S. Patel, MD; Richard K. Gurgel, MD, MSCI*

Objective: To assess the impact of cochlear implantation (CI) on cognitive outcomes in older adults 5 years post-implantation.

Study Design: Prospective, interventional study.

Setting: Tertiary care center.

Patients: Cochlear implant recipients aged 65 or older.

Interventions: Subjects underwent pre-operative cognitive testing with a novel battery of neuropsychological tests including those assessing global cognition (Mini-Mental Status Exam), verbally-based cognition (Digit span, Stroop, Hopkins Verbal Learning Test – Revised, Hayling Sentence Completion) and comparable visually-based cognition (Spatial span, d2 Test of Attention, Brief Visuospatial Memory Test – Revised [BVM-T-R], Trails A and B). Testing was repeated 5 years post-operatively.

Main Outcome Measures: Cognitive outcomes assessed with cognitive testing battery.

Results:

After 5.71 ± 1.14 years following cochlear implantation, 16 subjects (mean age 83 ± 7.22 years, 93.75% male, 87.5% with normal pre-operative cognitive status) repeated the cognitive battery. In comparison to pre-operative testing, subjects showed a significant improvement on the Hayling Sentence Completion test, which evaluates executive functioning (Wilcoxon signed rank: $Z=-2.275$, $p=0.023$). Conversely, there was a significant decrease in their scores on a verbal test of executive functioning (Stroop Color-Word: $Z=-2.557$, $p=0.011$) and visually-based tests of attention (d2 standardized score: $Z=-2.24$, $p=0.025$; Spatial span total score: $Z=-2.388$, $p=0.017$), memory (BVM-T-R: Total T score $Z=-2.500$, $p=0.012$, delayed T score $Z=-2.703$, $p=0.007$), and executive functioning (Trails B seconds: $Z=-2.158$, $p=0.031$). Other test scores did not show a significant change on follow-up.

Conclusions: In a 5-year follow-up, participants demonstrated a significant improvement on a verbally based test of executive functioning from baseline, suggesting a beneficial role of cochlear implantation in enhancing this cognitive domain in older recipients. Other tests of cognition showed stable scores or a decline, highlighting how cognitive domains may be affected differentially with aging following CI.

Professional Practice Gap & Educational Need: The long-term effect of CI on cognitive status of older adults who are at risk of dementia associated with hearing loss.

Learning Objective: Learners will better understand the long-term impact of cochlear implants on cognition in older adults.

Desired Result: To demonstrate that cochlear implants in older adults can improve cognition in certain domains, while other domains are unaffected or continue to decline with age.

Level of Evidence: Level III

Indicate IRB or IACUC: IRB 00083983, The University of Utah.

Tone Adaptation among Cochlear Implant Candidates to Inform Hearing Preservation during Awake Cochlear Implantation

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Objective: Cochlear implantation (CI) under local anesthesia facilitates intraoperative behavioral patient feedback to potentially improve acoustic hearing preservation. The degree to which tone adaptation to a sustained pure tone threshold during electrode insertion impacts the accuracy of this feedback is a critical factor to understand and remains undefined.

Study Design: Prospective study.

Setting: Tertiary academic medical center.

Patients: Prospective cochlear implant candidates.

Interventions: Standard behavioral audiogram, audiogram obtained in ambient operating room (OR) background noise, and tone adaptation to sustained suprathreshold (+10 and +20dB) pure tones at 500, 1000, 2000, and 4000Hz in both quiet and in OR noise.

Main Outcome Measures: Pure tone average (PTA) in quiet and in ambient OR noise, timing of tone adaptation at different suprathreshold levels and frequencies, and reliability of perceived loudness changes.

Results: In total, 20 ears were included for study. The median PTA was similar between testing in quiet (68dB, IQR 9) and in ambient OR noise (68dB, IQR 11). For pure tone adaptation, 10 patients reported decreased perception of tone at +10dB, at a median time of 55 seconds (IQR 58), most commonly at 250 Hz. For perception of loudness change, all patients were able to report a difference of 10dB across the frequency range at both +10 and +20dB when testing in quiet and in ambient OR background noise.

Conclusions: Audiometry demonstrated similar testing in quiet and ambient OR background noise, with 50% of patients reporting neural adaptation at +10dB, and all patients perceiving a 10dB change in stimulus amplitude both in quiet and in ambient OR noise.

Professional Practice Gap & Educational Need: Pure tone testing in noise, rates of neural adaptation, and ability to perceive changes to loudness amongst patients with hearing loss who qualify for CI are unknown.

Learning Objective: To describe audiometric testing features, frequency of pure tone adaptation, and patient perception of loudness changes in patients who qualify for CI.

Desired Result: To report similarity in pure tone average testing, the relative frequency of pure tone adaptation, and the accurate perception 10dB changes in both quiet and testing in ambient OR background noise.

Level of Evidence – Level III

Indicate IRB or IACUC: IRB #24-002837, “Systematic Assessment of Patients’ Perception of Amplitude (Loudness) Changes During Audiological Testing: Data to Inform Methods for Hearing Preservation Cochlear Implant Surgery Under Local Anesthesia”, Approval date 3/28/2024