### **SELECTED ABSTRACTS**

# POSTER PRESENTATIONS



# 52<sup>nd</sup> Annual Spring Meeting AMERICAN NEUROTOLOGY SOCIETY

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# Cost-Effective MRI Screening for Acoustic Neuromas in Patients with Asymmetric Hearing Loss

Matthew G. Crowson, MD; Daniel J. Rocke, JD, MD David M. Kaylie, MD, MS

**Objective:** To determine if a screening MRI with FIESTA/CISS sequences is cost-effective compared to a traditional full MRI protocol with contrast for the evaluation of acoustic neuromas.

**Study Design:** Cost-effectiveness (CE) analysis from a third-party payer perspective.

**Methods:** A decision tree was constructed to evaluate full MRI and screening MRI strategies for patients with asymmetric sensorineural hearing loss. If a patient were to have a positive screening MRI, they received a full MRI. Acoustic neuroma prevalence, MRI specificity and sensitivity, and gadolinium anaphylaxis incidence were obtained through literature review. Institutional charge data were obtained using representative patient cohorts. One-way and probabilistic sensitivity analyses were completed to determine CE model threshold points for MRI performance characteristics and charges.

**Results:** The mean charge for a full MRI with contrast was significantly higher than a screening MRI (\$4089  $\pm$  1086 vs. \$2872  $\pm$  741; p<0.05). The screening MRI protocol was more cost-effective than a full MRI protocol with a willingness-to-pay from \$0-20,000 USD. Sensitivity analyses determined that the screening protocol dominated when the FIESTA/CISS MRI charge was less than \$4,318., and the specificity exceeded 63.8%. The screening MRI protocol also dominated when acoustic neuroma prevalence was varied between 0 to 1,000 in 10,000 people.

**Conclusions:** A screening MRI protocol is more cost-effective than a full MRI with contrast in the diagnostic evaluation of an acoustic neuroma. A screening MRI also confers benefits of shorter exam time, and no contrast use. Further investigation is needed to confirm the performance of FIESTA/CISS MRI sequences for acoustic neuromas.

**Define Professional Practice Gap & Educational Need:** 1) Lack of awareness of the benefits of utilizing a screening MRI protocol in the assessment of a possible acoustic neuroma in an adult patient presenting with asymmetric hearing loss. 2) Lack of a contemporary economic evaluation of the utility of a screening MRI protocol compared to traditional MRI protocols with contrast.

**Learning Objective:** 1) At the conclusion of this presentation, the participants should be able: 1) To describe a cost-effective strategy using a screening MRI protocol for the evaluation of a patient with history and audiogram findings suggestive of an acoustic neuroma. 2) To compare the reported imaging performance of a screening MRI protocol without contrast versus a full MRI protocol with and without contrast for the identification of an acoustic neuroma.

**Desired Result:** It is the authors' hope that attendees will consider that a screening MRI protocol without contrast – a protocol that exclusively utilizes FIESTA or similar sequences – may be a more cost-effective strategy versus proceeding with a full MRI with and without contrast as the initial imaging modality in the evaluation of asymmetric hearing loss.

### Prevalence of Hearing Impairment and Hearing Care Utilization among Asian Americans

Janet S. Choi MD, MPH; Laurel M. Fisher, PhD Rick A. Friedman, MD, PhD; Elina Kari, MD

**Objective:** To assess the prevalence of hearing impairment and utilization of hearing care among Asian Americans (AA), using the first nationally representative sample of AA adults.

Study design: National cross-sectional survey

**Setting:** Ambulatory examination centers

**Patients:** 3,733 adults (539 AAs) aged 20-70 in the 2011-2012 National Health and Examination Survey whose hearing were assessed by pure-tone audiometry

**Intervention(s):** Hearing impairment defined as a speech frequency pure-tone average (PTA)  $\geq$ 25dB in better hearing ear.

Main outcome measure(s): Rates of hearing impairment, recent hearing test, and hearing aid use. Analyses incorporated sampling weights to account for complex sampling design.

**Results:** The prevalence of hearing impairment was 5.8% [95% CI: 3.0-8.6%] among AAs and increased substantially with age, which was comparable to other races/ethnicities including Whites, Blacks, and Mexican-Americans. After adjusting for age and PTA, AAs with hearing impairment were less likely to have received a recent hearing test compared to Whites (OR: 0.21 [95% CI: 0.08-0.53, p=0.003] and to Blacks (OR: 0.24 [95% CI: 0.08-0.70, p=0.012]), less likely to have used hearing aids compared to Whites (OR: 0.07 [95% CI: 0.01-0.65], p=0.022), and less likely to report trouble hearing compared to Whites (OR: 0.30 [95% CI: 0.10-0.89], p=0.032). Among AAs, private insurance (OR: 2.4 [95% CI: 1.10-5.04]) and Medicare (OR: 7.6 [95% CI: 1.9-30.3]) was associated with higher levels of receiving a recent hearing test.

**Conclusions:** This first examination of a nationally representative sample of AA and hearing loss suggests that the prevalence is similar to other races. However, widespread adoption of a hearing assistive device may be a more difficult challenge.

**Define Professional Practice Gap & Educational Need:** 1. Lack of knowledge in prevalence of hearing loss and patterns of hearing care utilization among Asian Americans despite substantial growth of the population group in the US 2. Lack of awareness in racial disparities and barriers to hearing care among racial and ethnic minorities especially among Asian Americans

**Learning Objective:** 1. To understand the prevalence of hearing loss and hearing care utilization patterns among Asian Americans in comparison to Whites, Blacks, and Mexican-Americans using the nationally representative sample. 2.To investigate the extent of racial/ethnic disparities in hearing health including hearing exams, assistive device use, and differential factors associated with the hearing care utilization among Asian Americans.

**Desired Result:**1. Attendees will learn that the racial/ethnic disparities exist in hearing care utilization among Asian Americans based on the first nationally representative sample 2. Attendees will understand the need for further research to identify barriers to hearing care and ways to promote hearing health to address the racial/ethnic disparities.

#### Influence of Co-morbidity and Surgical Strategy on the Clinical Outcome in the Treatment of SCDS

Prof Arne Ernst, MD

**Objective:** SCDS is a disorder with varying symptoms. The management is complicated by the need to offer the patient a realistic outcome with either conservative or surgical measures.

**Patient:** In our prospective, non-randomized series, 52 patients (31 female, 21 male, average age 48.8 yr) with radiologically confirmed SCDS were counselled, 41 agreed to undergo a surgical therapy. The surgical therapy consisted of canal occlusion via a transmastoid approach. In addition, all patients were given a hearing implant (CI or VSB) for auditory rehab. The patients were followed up for 14 months (6 - 24 months) on average (DHI, neurotological testing- c/oVEMPs, excentric rotation, HIT and PTA).

**Intervention:** A positive outcome (n = 28, i.e. significant improvement in DHI, normalisation of cVEMP thresholds) was correlated to the severity of vertigo preoperatively, to the parallel implantation of a CI, to the age at implantation (younger than 45 yrs).

**Outcome Measure:** A negative outcome (n = 13; i.e., non-significant improvement in DHI, no normalization of cVEMP thresholds) was correlated to the co-morbidity of vestibular migraine and/or Menière's disease, the bilateral occurrence of SCDS, SCDS defect size (larger than 4 mm), old age (55 yrs). In those 13 patients, 5 were revised which significantly improved the DHI outcome and led to a normalization of cVEMP amplitudes.

**Results:** The 10 patients who declined surgery were treated by antivertiginous drugs, which lead to a reduction of vertigo, but no significant improvement in DHI.

**Conclusions:** In essence, canal occlusion in SCDS can be highly effective when considering co-morbidity and adding hearing implants for auditory rehab.

**Define Professional Practice Gap & Educational Need:** The treatment strategies in Superior Canal Dehiscence Syndrome (SCDS) vary form none to surgery and focus on either/or the rehabilitation of hearing and/or balance. It is the aim of the present paper to better define diagnostic hints which will lead to resolving inconsistencies of present-day knowledge in choosing treatment strategies in SCDS and to produce awareness for the high interindividual variability of the clinical picture.

**Learning Objective:** The learning objective should be to realistically judge on the appropriate tratement strategy before starting any therapy and to learn how to counsel the patient adequately that he/she has a realistic expectation as well how to decide.

**Desired Result:** The take-home-message which is applicable in daily practice should be that the individual need of the patients (resolving their hearing and/or balance problem) must be the primary guideline for choosing any treatment option and this presentation should lead the way.

### Exclusive Transcanal Endoscopic Ear Surgery for Excision of a Facial Nerve Hemangioma with Interposition Nerve Grafting: A Case Report

Cameron C. Wick, MD; Mark Sakai, BS Brandon Isaacson. MD

**Objective:** To illustrate a novel approach for surgical management of a facial nerve hemangioma, including interposition nerve grafting, via an exclusively transcanal endoscopic ear surgery (TEES) approach.

**Patient:** 39-year-old female with a preoperative House-Brackmann (HB) grade IV facial paresis secondary to a facial nerve hemangioma.

**Intervention(s):** Surgical excision and interposition nerve graft via a transcanal endoscopic approach.

Main Outcome Measure(s): Completeness of resection, approach morbidities, and facial nerve outcome.

**Results:** The TEES approach provided wide exposure of the facial nerve from the geniculate ganglion through the vertical segment. This visualization facilitated complete tumor resection, incus interposition ossicular reconstruction, and placement of an interposition nerve graft. The nerve graft was positioned in the fallopian canal and was secured at both ends with Surgical. The patient had no postoperative complications. At 11-month follow-up her facial function had returned to HB grade IV.

**Conclusions:** To our knowledge, this is the first report of a facial nerve hemangioma resection and interposition nerve graft via an exclusively endoscopic approach. This report adds to the growing body of evidence for an endoscopic role in the management of diverse middle ear and lateral skull base pathology. Additional studies are needed to fully elucidate the risk-benefit profile of this technique.

**Define Professional Practice Gap & Educational Need:** This is novel approach to resect and reconstruct a facial nerve tumor in the middle ear. To our knowledge it is the first description of an entirely endoscopic approach for a facial nerve hemangioma resection and interposition nerve graft.

**Learning Objective:** Review differential diagnosis of middle ear neoplasms. Discuss when intervention is recommend for facial nerve tumors. Discuss different approaches and considerations for each approach.

**Desired Result:** Introduce a novel approach and reconstruction option.

# Dizziness in Vestibular Schwannoma Patients Due to Underlying Comorbidities

Yarah M. Haidar, MD; Ronald Sahyouni, BA Omid Moshtaghi, BS; Harrison W. Lin, MD Hamid R. Djalilian, MD

**Objectives**: To report findings from a cohort of vestibular schwannoma patients with a secondary comorbid vestibular disorder, and to discuss management strategies for this subset of patients presenting with both episodic vertigo and vestibular schwannoma (VS).

**Study design**: Retrospective case series.

**Methods**: VS patients presenting to a neurotology clinic in a tertiary care academic center with episodic vertigo.

**Results**: Nine VS patients presented with vertigo. Eight (89%) suffered from vestibular migraine (VM) and two (22%) had benign positional vertigo (BPV), one patient had both. All VM patients had at least partial improvement of their dizziness symptoms when treated with migraine lifestyle with migraine prophylactic therapy. One of the two patients with BPV had complete resolution of symptoms following particle repositions maneuvers. In seven of the nine patients (78%), treating the underlying non-VS condition resulted in complete dizziness resolution. Five patients (56%) avoided surgery and are continuing observation, while four patients (44%) had radiosurgery.

Conclusions: Many patients presenting with a VS report a history of recurrent episodic vertigo, and this may be due to underlying conditions and should be thoroughly elucidated prior to attributing it to the VS. All VS patients presenting with vertigo should have the standard vertigo history and examination, including a Dix-Hallpike exam, to determine if there is an alternate cause to their vertigo. Some authors have advocated surgical treatment for VS patients with vertigo. Other vestibular pathologies should be ruled out prior to considering microsurgical excision for VS patients with episodic vertigo.

**Define Professional Practice Gap & Educational Need:** Lack of awareness of alternate causes of dizziness due to underlying comborbidities in patients with vestibular schwannomas.

**Learning Objective:** To understand the workup and treatment of dizziness in patients with vestibular schwannomas.

**Desired Result:** Practitioners will appropriately workup and treat additional comorbidities in vestibular schwannoma patients to treat their underlying dizziness.

### The Effect of Citalopram Versus a Placebo on Central Auditory Processing in the Elderly

Jose Fernando Polanski, MD Alexandra Dezani Soares, MSc Liliane Desgualdo Pereira, PhD Oswaldo Laercio de Mendonça Cruz, PhD

**Objective:** Evaluate the effects of therapy with citalopram on the central auditory processing in the elderly measured by central auditory tests.

**Study Design:** Prospective, randomized, double-blind, placebo-controlled study.

**Setting:** Tertiary referral center.

**Patients:** Forty-nine patients older than 60 years with normal hearing thresholds or symmetrical sensorineural hearing loss up to 70 dBHL, word-recognition score equal to or better than 70%, and diagnosed with central auditory processing disorders. They underwent the mini-mental state examination, as a way to screen those with the possibility of dementia; they also underwent the Beck depression inventory, for screening individuals with depression.

**Intervention:** Citalopram 20mg/day or placebo for 6 months.

**Main outcome measure:** The central auditory tests were applied to the selection of individuals with auditory processing disorders and repeated after 6 months' treatment. The tests were: sound localization, speech in noise, dichotic digits' test, pitch pattern sequence, duration pattern test and gaps-in-noise.

**Results:** Comparisons of central auditory tests pre - and posttreatment in groups showed: sound localization (p=0,735), pitch pattern sequence humming (p=0,102), pitch pattern sequence nomination (p=0,157) duration pattern test humming (p=0,102), duration pattern test nomination (p=0,196) and gaps-in-noise (p=0,683). Dichotic tests in right and left ear respectively: speech in noise (p=0,143; p=0,052), dichotic digits test (p=0,492; p=0,233). Conclusions: There was no statistically significant effect with the use of citalopram in central auditory processing tests of the subjects.

**Define Professional Practice Gap & Educational Need:** A general lack of contemporary knowledge on the use of selective inhibitors of serotonin reuptake in central hearing disorders, despite the pathophysiological evidence of the importance of serotonin in the functioning of central auditory pathways.

**Learning Objective:** To recognize the difficulty in the management of central hearing disorders and to analyze the effect of citalopram in their treatment.

**Desired Result:** Hope to encourage new thinking and research on new therapy approaches in central hearing disorders.

#### Use of Positive Airway Pressure Following Middle Ear Surgery: A Practice Survey of Otologists

Douglas S. Ruhl, MD; Anthony M. Tolisano, MD Bradley W. Kesser, MD; George T. Hashisaki, MD Macario Camacho, MD

**Background:** Positive Airway Pressure (PAP) devices are used as treatment for obstructive sleep apnea (OSA). PAP may increase middle ear pressure which is concerning for otologic surgeons. There is a lack of data to guide how surgeons should manage PAP therapy following middle ear surgery.

**Objective:** To elucidate how otologic surgeons manage PAP in their patients after ear surgery.

**Study design:** A survey e-mailed to practicing members of the American Neurotology Society.

**Results:** Among sixty respondents, the most common advice given to patients was to avoid PAP use for one week (40%), return to normal use immediately (20%), avoid PAP for one month (13.3%) or avoid PAP for one day (13.3%) after surgery. Twenty percent of providers reported that they change their advice based on the PAP pressure settings (i.e. shorter hiatus for higher settings). Among respondents, 40% believe that they have patients with middle ear issues secondary to PAP and 20% attributed surgical failures to PAP use. One-third of providers routinely pack the Eustachian tube during surgery. Providers that attributed a negative surgical outcome to PAP use were more likely to routinely pack/plug the Eustachian tube during otologic surgery (p=0.0013).

**Conclusion:** Postoperative PAP treatment for OSA varies greatly among practicing otologists. Providers who believe that an adverse outcome was attributed to PAP use were more likely to prophylactically plug the Eustachian tube during surgery. Future research will provide additional information which will allow us to better understand the effect of PAP on the middle ear especially following otologic surgery.

**Define Professional Practice Gap & Educational Need:** 1. There is a lack of awareness that positive airway pressure (PAP) may affect middle ear pressure. 2. There is a paucity of data to guide how surgeons should manage PAP therapy following middle ear surgery.

**Learning Objective:** 1. Explain that positive airway pressure (PAP) may increase middle ear pressure - which is of concern for otologists. 2. Elucidate how otologic surgeons manage PAP in their patients after ear surgery.

**Desired Result:** 1. Attendees will better understand the potential relationship that positive airway pressure may have on the middle ear. 2. Attendees will consider the variations in practice patterns when managing PAP after ear surgery.

# **Epidemiology of Dizzy Patient Population in a Neurotology Clinic and Predictors of Peripheral Etiology**

Thomas Muelleman, MD; Matthew Shew, MD Rahul Subbarayan, MD; Axel Shum, MS Kevin Sykes, PhD; Hinrich Staecker, MD James Lin. MD

**Objective:** To compare the proportion of peripheral versus non-peripheral etiologies among all patients, inclusive of those presenting primarily or as referrals, to rank diagnoses in order of frequency, to determine whether or not age and gender predict diagnosis, and to determine which subgroups tended to undergo formal vestibular testing.

Study design: Retrospective cohort.

Setting: Academic neurotology clinic.

Patients: Age >18 neurotology clinic patients with the chief complaint of dizziness.

**Intervention(s)**: None.

Main outcome measure(s): Age, gender, diagnosis, record of vestibular testing.

**Results**: 2079 patients were assigned 2468 diagnoses, of which 57.7% and 42.3% were of peripheral and non-peripheral etiologies, respectively. The most common diagnoses were Meniere's (23.0%), vestibular migraine (19.3%), BPPV (19.1%), and central origin, non-migraine (16.4%). Peripheral diagnoses are more likely to be found in men than in women (odds ratio 1.59). Peripheral diagnoses were most likely to be found in the 60-69 age group (odds ratio 3.82). There was not a significant difference in rate of vestibular testing between women and men. Among patients with two diagnoses, the most common combinations were vestibular migraine and BPPV then vestibular migraine and Meniere's.

**Conclusions:** A large proportion of patients seen for the chief complaint of dizziness in the neurotology clinic were found not to have a peripheral etiology of their symptoms. These data challenge a prevalent dogma that the most common causes of vertigo are peripheral: BPPV, vestibular neuritis, and Meniere's disease. Age and gender are statistically significant predictors of peripheral etiology of dizziness.

**Define Professional Practice Gap & Educational Need:** 1. Lack of awareness of proportion of non-peripheral causes of dizziness in patients in neurotology clinic 2. Lack of collaboration to optimize clinical care for patients with dizziness 3. There are numerous studies of demographics of dizzy patients from various populations: general population, population in primary care clinics, geriatric patients, emergency department patients, and neurology clinics. Interestingly, there have not been reports in the literature of demographics of dizzy patients in a neurotology clinic.

**Learning Objective:** 1. To understand the proportion of peripheral etiologies of dizziness to non-peripheral etiologies 2. To understand the most common diagnoses for dizzy patients in neurotology clinic 3. To understand whether or not age and sex predict peripheral dizziness 4. To understand whether which subgroups of dizzy patients tend to undergo formal vestibular testing 5. To understand which dizzy diagnoses are commonly associated with each other

**Desired Result:** Attendees will be able to contextualize their own neurotology practice relative to this large population of dizzy patients and better appreciate the importance of a close working relationship with other services in the treatment of patients who suffer from dizziness.

#### An Analysis of Safety and Adverse Events Following Cochlear Implantation in Syndromic Children

Vijay A. Patel, MD; Huseyin Isildak, MD Michele M. Carr, DDS, MD, PhD

**Objectives:** To determine the safety profile and perioperative morbidity of syndromic children undergoing cochlear implantation (CI).

**Study Design:** Retrospective study utilizing the American College of Surgeons National Surgical Quality Improvement Program Pediatric Database (ACS-NSQIP).

**Methods:** Pediatric patients who underwent CI were queried using the ACS-NSQIP database from 2012-2014. Univariate analysis and multivariate logistic regression were used to determine group comparisons. Outcomes included prematurity, comorbidities, operative time, length of stay, complications, readmissions, and related reoperations.

**Results:** 395 of the 1,993 cases of pediatric CI were found to have an associated congenital abnormality. The mean age of syndromic children was 4.98 years compared to 5.27 years for non-syndromic children. Syndromic patients were more likely to have an unplanned readmission (4.3% vs. 2.3% of children, p=0.045), unplanned reoperation (2.0% vs. 0.8% of children, p=0.035), and present with significant medical comorbidities (i.e. prematurity p=0.002, cardiac anomalies p<0.001, asthma p=0.015). However, syndromic children were found to have no statistically significant difference in postoperative complications including superficial and deep wound infections. Finally, syndromic children had significantly shorter operative times (145.90 vs. 166.81 minutes, p<0.001) and a shorter length of stay (0.54 vs. 0.90 days, p=0.041).

**Conclusions:** In spite of the medical complexity of syndromic patients, these children have no increase in adverse events in the immediate postoperative period although readmissions and reoperation were found to be more common.

**Define Professional Practice Gap & Educational Need:** Lack of contemporary knowledge with respect to the safety and adverse events associated with cochlear implantation in syndromic children.

**Learning Objective:** To determine the safety profile and adverse events of syndromic children undergoing cochlear implantation (CI).

**Desired Result:** As early CI continues to reveal decreased auditory deprivation, improved speech perception, and early primary language development, considerations may be made for the equivalent safety profile of CI in syndromic children.

#### **Publishing Trends in Otology and Neurotology**

Ryan M. Boerner, MD; Jonathan Hatch, MD Elizabeth Harruff, BS; Shaun A. Nguyen, MD Theodore McRackan, MD; Ted A. Meyer, MD, PhD Paul R. Lambert, MD

**Objectives**: 1) Describe publishing trends for otologic/neurotologic disorders over a 35 year span 2) Compare trends in publishing with disease prevalence.

**Methods**: PubMed searches were performed on 35 otologic/neurotologic disorders using medical subject headings (MeSH) terms from 1980-2015 to determine the number of published articles per year. A Mann-Kendall trend analysis evaluated changes in publication frequency over time as a discrete variable while correcting for total number of articles published per year. Topics were evaluated both as annual count and as a percentage of the total number of articles cited.

**Results**: The total number of publications on the 35 topics increased from 853 in 1980 to a peak of 3068 in 2013. Otitis externa ( $\tau$  = -0.634, p < 0.001), cholesteatoma ( $\tau$  = -0.629, p < 0.001), and Meniere's disease ( $\tau$  = -0.724, p < 0.001) all showed decreasing publication trends with otitis media ( $\tau$  = -0.799, p < 0.001) showing the largest decrease. Topics with positive trends included cochlear implants ( $\tau$  = 0.740, p < 0.001), congenital hearing loss ( $\tau$  = 0.629, p < 0.001), and temporal bone encephaloceles ( $\tau$  = 0.743, p < 0.001). Glomus jugulare showed the least variability ( $\tau$  = -0.0244, p=0.085). Rapid rise in publications on superior canal dehiscence and vestibular migraine illustrate novel diagnoses.

**Conclusion**: This study displays trends in the literature over the past 35 years that are often inconsistent with common disorders seen by otologists/neurotologists. Certain diagnoses that are currently being researched less commonly continue to impact patients with the same regularity.

**Define Professional Practice Gap & Educational Need:** There are currently inconsistencies in publication trends in Otology and Neurotology with disorders encountered more frequently being published with decreasing frequency.

**Learning Objective:** To identify disorders at the forefront of current research efforts while recognizing the disorders being published at decreasing rates but effecting individuals with the same regularity.

**Desired Result:** This study will attempt to identify the current direction of the field of Otology and Neurotology and promote research efforts in disorders that continue to effect patients despite decreasing publication trends.

#### Bilateral Bifurcation of the Vertical Facial Nerve Segments in an Asymptomatic Patient

Spencer E. Lindsey, MD; Melissa Kang, MD Michael Hoa, MD

**Objective:** To describe a clinical encounter with asymptomatic, bilateral vertical segment facial nerve bifurcation and literature review of the anomaly

**Study design:** Case study, literature review

**Setting:** Veterans Administration hospital

**Patient:** 32 year old Asian American male with bilateral conductive hearing loss and Eustachian tube dysfunction, noted to have asymptomatic facial nerve bifurcation on pre-operative imaging for right tympanoplasty and bilateral Eustachian tube balloon dilation. Patient noted to have bilateral bifurcation of the distal vertical segment of the facial nerve, with complete duplication unliaterally and incomplete on the contralateral. He did not exhibit any signs of facial weakness or other temporal bone anatomical variations.

**Intervention:** CT temporal bone, MRI IAC with and without contrast

**Results:** Approximately five prior publications reporting facial nerve bifurcations and anomalies exist in the otolaryngology literature. Facial nerve anomalies are most commonly associated with other dysplasias of the middle and inner ear. Tympanic segment anomalies, typically unilateral, are most commonly identified, though labyrinthine and mastoid anomalies have been reported as well. The presentation of our patient is novel in that it represents a rare bilateral distal mastoid segment facial nerve bifurcation in a non-syndromic patient without any other anatomic abnormalities. The possibility of incomplete extension from the temporal bone of the extratemporal facial nerve is raised.

**Conclusions:** Facial nerve bifurcation is a rare malformation that can occur along any segment of the nerve and can have significant effect on potential complications. While the incidence is low, pre-operative high resolution CT imaging is key to its detection.

**Define Professional Practice Gap & Educational Need:** 1. Lack of awareness of condition 2. Unknown incidence of this variant 3. Poorly defined significance of finding

**Learning Objective:** To increase awareness of a rare anatomical variant and potential effects on surgical planning and potential complications. To encourage identification and further study of this variant.

**Desired Result:** To educate providers regarding a rare anomaly in order to increase pre-operative identification and avoid complications through use of imaging and careful surgical planning. To improve documentation of the variation in the reported literature

#### Decreased Percutaneous Threshold of Facial Nerve Stimulation Predicts Facial Canal Dehiscence

Patricia Johnson, MD; Taha Mur, BS Kamil Amer, BS; Rich Vogel, PhD Pamela Roehm, MD, PhD

**Objective:** Preoperative temporal bone high-resolution computed tomography (HRCT) is used to assess middle ear and mastoid anatomy. However, HRCT is an unreliable method of detecting facial canal dehiscence. We aimed to determine if preoperative transcutaneous facial nerve (FN) stimulation at the stylomastoid foramen could predict middle ear facial canal dehiscence.

**Study design:** Retrospective review

**Setting:** Tertiary center

**Patients:** Records of adult patients who underwent otologic surgery at our institution from January 2015 to October 3, 2016 were reviewed. Those selected for inclusion had preoperative FN stimulation and HRCT available for review.

**Intervention:** Using an FN stimulator placed on the skin over the stylomastoid foramen, the FN was stimulated at amperages ranging from 0.01—30 milliamperes (mA). Thresholds to waveform formation and amplitudes of compound muscle action potential (CMAP) were recorded at all electrodes.

**Main outcome measures:** Threshold to CMAP, average threshold to CMAP, threshold to maximum amplitude of CMAP, and maximum amplitude of CMAP.

**Results:** 34 patients met inclusion criteria. Of the 11 with intraoperatively confirmed dehiscences, 6 were identified by the attending surgeon on HRCT and 2 were identified on official radiology report. Mean lowest threshold to CMAP (0.75mA vs 8.0mA), and mean maximum amplitude of CMAP (1835 $\mu$ V vs 1197 $\mu$ V) of dehiscent versus non-dehiscent nerves were significantly different (p<0.05).

**Conclusions:** Iatrogenic facial nerve injury is one of the most devastating potential complications of otologic surgery. The use of facial nerve stimulation is a simple and cost-effective tool that can give the surgeon presurgical confirmation of facial nerve anatomy.

**Define Professional Practice Gap & Educational Need:** Lack of contemporary knowledge

**Learning Objective:** Understand that preoperative facial nerve stimulation can be used to provide information about facial nerve anatomy to supplement high resolution CT scan.

**Desired Result:** They will use preoperative facial nerve stimulation to supplement knowledge of patient's facial nerve anatomy as described by high resolution CT scan.

#### Serum Levels of Prestin in a Mouse Model of Cisplatin Ototoxicity

Benjamin Liba; Elizabeth Bezyk Charlene Campbell Michael Mei James Naples, MD; Kourosh Parham, MD, PhD

**Hypothesis:** Prestin in circulation may serve as a biomarker after exposure to cisplatin.

**Background:** Biomarkers are commonly used in detection of disease processes and provide an easy method of early diagnosis and monitoring. At present, no serologic biomarkers are available for inner ear diseases. Our group has proposed outer hair cell-specific protein, prestin, as a possible biomarker of inner ear damage in circulation. Here we investigate prestin as a biomarker in a mouse model of cisplatin ototoxicity.

**Methods:** Control and five groups of experimental mice were used. Baseline click-evoked auditory brainstem response (ABR) thresholds were recorded for all mice. Baseline blood was collected from untreated, control mice. The remaining groups were injected with cisplatin (16 mg/kg) and ABR threshold measurements and blood draws were repeated either 12 hr, 1, 3, 7 or 14 days later. Prestin concentrations in the serums were measured using ELISA.

**Results:** ABR thresholds were significantly elevated 24 hrs after cisplatin injection, but declined over days 3-14. There was a trend toward increased prestin concentrations reaching a maximum at 24 hr after cisplatin and then declined over days 3-14. These changes, however, failed to reach statistical significance.

**Conclusions:** The preliminary results presented here suggest that the time course of change in circulatory prestin parallels that of ABR threshold changes. Additional work is needed to further clarify the relationship between serum prestin levels and ABR thresholds as well as histological changes present in the cochlea. Prestin appears to have the potential to serve as a biomarker.

**Define Professional Practice Gap & Educational Need:** 1. Lack of an early detection system for inner ear diseases. 2. Lack of awareness that the outer hair cell-specific protein, prestin, can serve as a biomarker.

**Learning Objective:** To understand that the current methods used to measure inner ear disease lack the ease of use and reliability that a biomarker measured in circulation can provide.

**Desired Result:** They will begin to actively seek ways in which prestin can be used as a biomarker to monitor inner ear disease.

### Mastoid and Inner Ear Measurements in Patients with Ménière's Disease

Eric M. Sugihara DO; Alexander L. Marinica, DO Nathan D. Vandjelovic DO; Benjamin M. Kelley, DO Said S. Sana DO; Seilesh C. Babu MD

**Objective**: To determine the relationship between radiographic temporal bone anatomy of patients with Meniere's disease in medically and surgically managed populations.

**Study Design:** Retrospective chart review.

**Setting**: Two tertiary referral centers.

**Patients**: Adults older than 18 years with Meniere's disease treated with endolymphatic sac decompression (ESD) or medical management (non-ESD) vs controls.

**Interventions:** MRI and CT imaging studies of the temporal bones were reviewed by blinded radiologists.

**Main Outcome Measures**: Radiographic temporal bone dimensions were measured in Meniere's disease and control patients. Age, sex, symptoms, nadir audiogram data, academy classification of Meniere's disease, number of acute medical interventions, further surgery, and follow up were recorded. Statistical analysis was performed to compare outcome measures across groups and demographics.

**Results**: A total of 90 imaging studies were reviewed. Meniere's disease represented 52/90 studies (ESD 22/52; non-ESD 30/52). ESD and non-ESD groups had similar pure tone averages (33.95±20.55 vs 41.63±22.56 dB HL; p=0.21) and frequency of definite Meniere's disease (59.1% vs 53.3%; p=0.69). Mean vestibule width was less in Meniere's disease (2.99±0.46mm) vs controls (3.19±0.39mm; p=0.024). Mean vestibule length significantly increased between the control (5.45±0.54mm), non-ESD (5.80±0.96mm), and ESD (5.94±0.81mm) groups (p=0.018). Mean mastoid height was less in the ESD group (35.3±8.7mm) compared to non-ESD (38.9±4.9mm) and controls (38.9±5.8mm), which approached significance (p=0.11).

**Conclusions**: Medically and surgically managed Meniere's disease patients were clinically similar based on academy classification. Although surgical Meniere's disease patients may have more compact mastoid and elongated vestibular anatomy, these radiographic differences may not be a conclusive clinical predictor.

**Define Professional Practice Gap & Educational Need:** There are very few studies investigating the relationships between mastoid and inner ear anatomy and Meniere's disease. As a result, there is a lack of knowledge and awareness of such a relationship. Furthermore, there has never been a clinical study looking at the temporal bone anatomy and the various degrees of Meniere's disease, such as medically and surgically managed patients.

**Learning Objective:** The learning objective is to understand the different measurements of mastoid and inner ear anatomy in Meniere's disease patients versus controls, especially in the surgical subset of patients.

**Desired Result:** The desired result will be to introduce awareness of radiographic differences in Meniere's disease patients, which may be applied as a possible predictor or guide in otologic surgery for these patients.

# Hearing Preservation after Cochlear Implantation and Administration Different Models Steroid Therapy

Piotr H. Skarzynski, MD, PhD; Magdalena B. Skarzynska, MA Bartlomiej Krol, MD; Magdalena Koziel, MSc Kamila Osinska, MA; Henryk Skarzynski, Prof. MA, PhD

**Objective:** The objective of the study is to assess how different models of steroid therapy influence hearing preservation.

**Data sources:** During the last decade we observed the increase of number of scientific reports on topical application of steroids in treatment of inner ear diseases such as sudden deafness, tinnitus, Meniere's disease, autoimmune inner ear disease and hearing loss caused by the introduction of the cochlea electrode implant.

**Study selection:** To assess influence of different models of steroids administration on hearing preservation after cochlear implantation Pure Tone Audiometry was applied. The hearing preservation was calculated based on special formula developed by the Hearing Group, meaning Hearing preservation = (1-(PTApost-PTApre/PTAmax-PTApre)) x 100.

**Data extraction:** Patients included in the study were divided into 3 groups: intravenous steroid administration, intravenous + oral steroids administration and a control group. Since May to October (2016) 15 patients were included to the study group. By the end of the year 2016 additionally 10 patients will be included in the study. In all of the cases the cochlear implant electrode was inserted via the RW approach.

**Data synthesis:** After the analysis, the first data obtained from 13 out of 15 patients. The complete hearing preservation was observed in 5 cases, partial hearing preservation was observed in 6 cases and minimal hearing preservation was observed in 2 cases.

**Conclusions:** In the most of cases we observed hearing preservation after application of intravenous or combination of intravenous and oral steroids. A larger number of patients is required to assess the effectiveness and compare the two models of steroid therapy.

**Define Professional Practice Gap & Educational Need:** Lack of contemporary knowledge on use of steroids in Partial Deafness Treatment

**Learning Objective:** Information about feasibility of using steroids in this kind of treatment

**Desired Result:** Application of knowledge in everyday clinical practice

#### Forward Masking of the Speech-Evoked Auditory Brainstem Response

Sarah E. Hodge, MD; Denise C. Menezes, PhD Kevin D. Brown, MD, PhD; John H. Grose, PhD

**Hypothesis:** The goal is to determine if susceptibility to forward masking of the speech-evoked Auditory Brainstem Response (sABR) increases with advancing age.

**Background:** Older listeners have difficulty understanding speech in background noise even when their audiograms are normal. This is partly due to deficits in the processing of time-domain speech information, including masking of speech sounds by prior sounds (forward masking). The goal of this study was to determine if forward masking of the sABR can be used as an objective measure of temporal processing ability.

**Methods:** Forward-masked sABRs were measured in normal hearing young adults using a 40-ms synthetic /da/ signal preceded by a 100-ms speech-shaped noise masker. The masker was presented at 75 dB SPL and the signal at 70 dB peSPL. The interval between masker offset and signal onset ( $\Delta t$ ) was 4, 16, 32, or 64 ms. Baseline was the response to the unmasked signal. The dependent variables were the latencies of vertex-positive peaks in the response waveform.

**Results:** Response peak latency varied inversely with  $\Delta t$ . This was most pronounced for the initial peak ( $\approx$  ABR Wave V) which showed a maximal mean shift of > 1 mSec. Later peaks showed only minor systematic latency shifts. The initial peak was generated primarily by the plosive onset of the signal. Preliminary data on older listeners suggests a similar behavior but with latency shifts extending out to later peaks.

**Conclusion:** Forward-masked sABRs provide a viable and objective measure of temporal processing that appears to be sensitive to listener age.

**Define Professional Practice Gap & Educational Need:** Clinical use of the auditory brainstem response (ABR) test has been limited primarily to threshold estimation or site-of-lesion assessment using simple click or tone stimuli. This study demonstrates that the ABR test is capable of providing much more information regarding auditory function such as temporal processing ability.

**Learning Objective:** The learner will understand how speech-evoked ABRs can be used in the assessment of age-related temporal processing deficits

**Desired Result:** The desired result is a rigorous demonstration that the forward-masked sABR can provide a reliable objective gauge of temporal processing that, in turn, will lead to a better assessment of the speech understanding difficulties of older listeners even with normal audiograms.

# Prevalence of Extracochlear Electrodes: CT Scans CI Maps, and Operative Reports

Jourdan T. Holder, AuD; Jack H. Noble, PhD David M. Kessler, BA; René H. Gifford, PhD Robert F. Labadie, MD, PhD

**Objective:** To quantify and compare the number of cochlear implant (CI) electrodes found to be extracochlear on postoperative CT scans, the number of basal electrodes deactivated during standard CI mapping (without knowledge of the post-operative CT scan results), and the degree of electrode insertion noted by the surgeon.

Study Design: Retrospective

**Setting:** Academic Medical Center

**Methods:** 357 patients underwent standard cochlear implantation followed by postoperative temporal bone CT scanning, which was analyzed to determine the number of extracochlear electrodes. Standard CI programming was completed without knowledge of the number of extracochlear electrodes identified on the CT. These standard CI maps were reviewed to record the number of deactivated basal electrodes. Lastly, each operative report was reviewed to record the degree of reported electrode insertion.

**Results:** Twenty-eight percent (n=100) of the CIs were found to have at least one electrode at the entrance of or outside of the cochlea on the CT scan. Review of CI mapping indicated that audiologists had deactivated extracochlear electrodes in 62% (n=62) of these cases. Review of operative reports revealed that surgeons indicated incomplete insertion in only 8% of cases (n=8).

**Conclusions:** Extracochlear electrodes were identified audiologically in 62% of cases and in surgical reports in 8% of cases; however, it is possible that at least a portion of these cases involved postoperative electrode migration. Nevertheless, given these findings, postoperative CT scans can provide information regarding basal electrode location which could help improve programming accuracy, associated frequency allocation, and audibility with appropriate deactivation of extracochlear electrodes.

**Define Professional Practice Gap & Educational Need:** 1. Lack of awareness of the prevalence of extracochlear electrodes 2. Lack of understanding of how extracochlear electrodes can adversely affect audibility for CI patients, if not deactivated appropriately 3. Incomplete information provided in operative reports, which could help guide CI programming, if reported more accurately

**Learning Objective:** 1. Understand prevalence of extracochlear electrodes and how extracochlear electrodes are identified 2. Recognize the importance of postoperative CT scanning to optimize CI mapping 3. Recognize the importance of thorough and descriptive operative reports to help guide CI mapping

**Desired Result:** Increased awareness of the prevalence of extracochlear electrodes will lead to more postoperative CT scans and/or more accurate operative reports to help guide appropriate deactivation of these electrodes during CI mapping, which will in turn improve programming accuracy, associated frequency allocation, and audibility for patients.

# Vestibular Preservation After Cochlear Implantation with Modern Electrode Arrays

Samuel J. Trosman, MD; Erika A. Woodson, MD

**Objective:** To determine the prevalence of vestibular symptoms and functional change after cochlear implantation (CI) with slim or mid-scalar electrode arrays.

**Study design:** Prospective, single-blinded pilot study

**Setting:** Tertiary care academic center

**Patients:** Eleven ears from 10 patients who underwent CI with slim or mid-scalar electrode arrays. Exclusion criteria included history of a vestibular disorder or preoperative vestibular hypofunction (VH).

**Intervention:** Cochlear implantation

Main outcome measure: Subjective and objective vestibular function was assessed using the Dizziness Handicap Inventory (DHI), caloric electronystagmography (ENG) and cervical vestibular evoked myogenic potentials (c-VEMP) before CI and >2 months post-CI. Hearing was assessed by unaided pure tone audiometry.

**Results:** Postoperatively, 2 implanted ears had clinically significant VH (>25% caloric asymmetry), but no ear lost >50% caloric response. Three ears had a total loss of c-VEMP response, including both ears with postoperative VH. The median postoperative low-frequency pure tone average (LF-PTA) shift was 18 dB (range 4-31 dB). The two ears with VH and c-VEMP loss had postoperative LF-PTA shifts of 16 and 18 dB, and the ear with c-VEMP response loss had a LF-PTA shift of 11 dB. The DHI for the subjects with objective findings did not change after CI.

**Conclusions:** All patients who underwent CI with a slim or mid-scalar electrode array had labyrinthine preservation, with only 2 of 11 ears (18.1%) showing clinically significant VH. Neither DHI score nor low-frequency hearing preservation appeared predictive of postoperative VH.

**Define Professional Practice Gap & Educational Need:** 1. Lack of contemporary knowledge of the impact of modern-era cochlear implantation on vestibular function.

2. Inconsistencies in preoperative evaluation of patients considering sequential cochlear implantation.

**Learning Objective:** 1. Understand the risk to vestibular function after modern cochlear implant surgery, 2. Recognize that hearing preservation and vestibular symptoms may not predict vestibular loss after cochlear implantation.

**Desired Result:** Pursue vestibular testing when evaluating candidates for sequential cochlear implantation. Utilize knowledge of vestibular loss after cochlear implant surgery to guide candidacy evaluation, primary and sequential. Counsel patients appropriately on their likelihood of dizziness or vestibular loss after cochlear implantation.

### Everyday Listening Experience Does Not Improve Osseointegrated Device Performance in Single-sided Deafened Individuals

P. Cody Buchanan, DO; Jacky V. Tran, BS Jake Hillyer, BS; Elizabeth Elkins, AuD Stacey D. Watson, AuD; Douglas D. Backous, MD Alexandra Parbery-Clark, AuD, PhD

**Objective:** Determine if 12 months of daily osseointegrated (OI) device usage improves hearing in noise and localization performance in single-sided deafened individuals.

**Study Design:** Prospective longitudinal study

**Setting:** Tertiary referral center

**Patients:** 10 adults with single-sided deafness (SSD) and normal contralateral hearing; minimum of 6 months OI usage prior to initial visit.

Interventions: Speech in noise (SIN) understanding and localization were assessed in a multi-speaker array (R-space) with patients either repeating sentences embedded in competing noise or verbally indicating the source speaker of a sentence stimuli (i.e. Where am I coming from now?), respectively. To gauge the impact of microphone settings on performance, directional, omnidirectional, and adaptive microphone modes were used. Tests were completed at two time points: 1) following 6 months of consistent device use (i.e., 2) itial provides a problem of the points of the po

**Main Outcome Measures:** SIN understanding and localization abilities were assessed longitudinally over a 12-month time period using multiple microphone modes.

**Results:** Localization and SIN understanding performance show no significant improvement for OI users after 12 months of daily use regardless of microphone mode.

**Conclusions:** Daily usage of an OI device in SSD individuals does not result in performance gains over a 12-month period. Developing a targeted auditory training program focused on improving OI hearing in noise and localization performance will be important future directions for improving OI user SIN and localization abilities.

**Define Professional Practice Gap & Educational Need:** 1. Lack of knowledge of ways to improve osseointegrated device performance among single-sided deafened individuals. 2. Lack of the ability to counsel single-sided deafened individuals on their performance using osseointegrated devices over time.

**Learning Objective:** 1. To understand the effect of everyday listening experience on osseointegrated device performance in single-sided deafened individuals. 2. To understand how to properly counsel single-sided deafened patients regarding osseointegrated device performance over time.

**Desired Result:** Attendees will be able to properly counsel single-sided deafend individuals regarding osseointegrated device usage over time.

# Vestibular Rehabilitation Utilizing a Novel Palatal Alternative Sensory Feedback Device

Benjamin F. Erhardt, MD Carmen Casanova Abbott, PT, PhD Arnaldo Luis Rivera, MD

**Objective:** Establish the feasibility of palatal alternative sensory feedback utilizing a novel device (EquiCue V1.0, Innervo Technology LLC) to improve vestibular function in patients with vestibular loss or vestibulopathy.

Study design: Pilot case series

**Setting:** Tertiary Care Medical Center

**Patients:** Four English speaking adults, ages 18-85, with documented vestibulopathy who completed a course of vestibular rehabilitation.

**Intervention:** Patients were custom fit with a novel frequency modulated electrostimulatory palatal device (EquiCue V1.0, Innervo Technology LLC), which was custom calibrated to their sensory sensitivity. Patient underwent 1 to 3 90-minute sessions administered by physical therapists.

**Main outcome measures:** Validated outcome measures (Sensory Organization Test (SOT), Dynamic Gait Index (DGI), and Verbal Analog Dizziness Scale(VADS)) were administered with and without the appliance.

**Results:** SOT: 4/4 patients saw consistent improvement in vision and vestibular integration with use of the device. Their reliance on the somatosensory/visual systems normalized with use of the appliance. All patients showed correction of posterior center of gravity alignment to central. DGI/VADS: 3/4 of patients had immediate relief of symptoms of dizziness/nausea/unsteadiness with the appliance on their first visit, as calculated using the VADS, which persisted on repeat sessions. Total DGI score increased 1-2 points. Increased speed of movement and steadiness was observed. Patients were able to perform pivot turns and stepping over obstacles without staggering.

**Conclusions:** Use of a novel non-invasive frequency modulated electrostimulatory palatal device may afford relief of vestibular symptoms and improvement in vestibular function in patients with documented vestibulopathy. Further study is indicated to confirm these results.

**Define Professional Practice Gap & Educational Need:** Patients with documented vestibulopathies not amenable to medical or surgical therapy are currently offered vestibular rehabilitation with physical therapy. Motivated investigation has taken place to produce alternative sensory feedback devices to assist in augmentation of the patients' residual balance and vestibular function. These have included devices focusing on intraoral alternative sensory feedback, specifically, the tongue. We present our initial experience with a novel palatal alternative sensory feedback device (Equicue V1.0, Innervo Technology LLC) which, if effective, could prove to be a more practical and ergonomic alternative.

**Learning Objective:** Those in attendance will learn about our initial positive subjective and objective results in vestibular rehabilitation utilizing a novel palatal alternative sensory feedback device (Equicue V1.0, Innervo Technology LLC).

**Desired Result:** Attendees will understand the potential application of this novel palatal alternative sensory feedback device (Equicue V1.0, Innervo Technology LLC). With further research, such a product could provide benefit to many of their patients.

### Non-Contrast MRI for Monitoring of Patients with Acoustic Neuroma

Mathieu Forgues, MD; Rahul Mehta, MD Dwayne Anderson, MD; Christian Morel, MD Laura Miller, MD; Alexander Sevy, MD Moises Arriaga, MD

**Objective**: To assess the feasibility of high resolution non-contrast T2 MRI images as compared to T1 post-contrast images for detecting acoustic neuroma growth.

**Study design**: Retrospective clinical study.

**Setting**: Tertiary referral center.

**Patients**: Adults with a diagnosis of acoustic neuroma who underwent an MRI of the internal auditory canals with and without contrast in the past 9 years.

**Intervention**: T1 post-contrast and T2 non-contrast MRI images were separately, randomly, and blindly reviewed by neuroradiologists. Measurement of tumor size on T1 post-contrast MRI was used as the "gold standard."

**Main outcome measure**: Tumor size was measured in one millimeter increments. The accuracy of the measurements on T2 non-contrast images was defined as a difference of less than or equal to two millimeters from the measurement on T1 post-contrast image. The mean of the measurements on T1 and T2 images were compared.

**Results**: 203 MRI images of 88 acoustic neuroma patients were reviewed. Measurements of tumor size on T2 non-contrast MRI were 80% accurate. Measurements of tumor size on T2 images were on average 13.5% smaller than on T1 images.

Conclusions: Our results suggest that acoustic neuroma growth could accurately be detected with non-contrast high resolution T2 MRI images. For patients monitored with serial imaging, this avoids the risks to patient health associated with intravenous gadolinium and saves significant time and hospital resources. We recommend T1 post-contrast images still be used for initial diagnosis. Additional analysis including inter-observer reliability and serial comparisons of each modality will be included in the final presentation.

**Define Professional Practice Gap & Educational Need:** Lack of evidence comparing the efficacy of non-contrast and post-contrast MRI images for detecting growth of acoustic neuromas.

**Learning Objective:** 1. Understand the capability and limits of non-contrast T2 images for detecting acoustic neuroma size and growth. 2. Define the role of non-contrast T2 images and post-contrast T1 images in the diagnosis and management of acoustic neuromas.

**Desired Result:** 1. Appropriate use of non-contrast MRI for monitoring of patients with acoustic neuromas.

- 2. Decrease in patient health risks related to intravenous gadolinium, such as Nephrogenic Systemic Fibrosis.
- 3. Decreased use of hospital resources and time spent in serial imaging of patients with acoustic neuromas.

# Stratification of Presentation and Management of Encephaloceles by Etiology

Maja Svrakic, MD; Spiros Manolidis, MD

**Objective**: To characterize the clinical and surgical findings in patients with temporal bone defects and describe surgical repair methods.

Study Design: Retrospective chart review

**Setting:** Multiple tertiary referral centers

**Patients:** Eighty-nine temporal bone encephaloceles diagnosed in eighty-seven patients

**Intervention:** Tegmen defects were repaired via a transmastoid (TM) approach alone or in combination with a middle cranial fossa (MCF) exposure. Repair methods included reconstruction with cartilage, bone, fat and fascia, including a pedicled superficial temporal fascia flap. Other surgical interventions were tympanoplasty, facial nerve decompression and mastoidectomy.

Main Outcome Measures: Etiology of encephalocele, presence of cholesteatoma, infection and cerebrospinal fluid (CSF) leak, incidence of prior surgeries, size of tegmen dehiscence, facial nerve and labyrinthine involvement, surgical intervention, and approach and tissue materials utilized for repair.

**Results**: Etiology of encephaloceles included chronic otitis media (74.2%), spontaneous (15.7%) and all other causes (9.0%). In chronic otitis media, most bony defects were associated with cholesteatoma (90.9%) and 80.3% cases had prior surgeries. The encephaloceles were repaired via a TM approach only in 26 (29.2%) ears, in combination with a mini-craniotomy in 37 (41.6%) ears, via a MCF approach in 18 (20.2%) ears and with fat obliteration in 8 (9.0%) ears. The approach, associated surgical interventions and reconstructive materials were dependent on the etiology of encephalocele.

**Conclusions**: Encephaloceles resulting from chronic otitis media present differently and are managed differently than those from spontaneous or traumatic etiologies. Associated findings and proposed algorithms for surgical management are described.

**Define Professional Practice Gap & Educational Need:** Inconsistencies within the field on reconstructive methods utilized for encephalocele repairs based on etiologies

**Learning Objective:** Characterize the clinical and surgical findings in patients with temporal bone defects and describe surgical repair methods based on etiology of the defects

**Desired Result:** The attendees can utilize our proposed algorithms for the surgical management of encephaloceles based on their etiology.

# Assessment of Disruptive Behavioral Problems in Children with Hearing Loss

Caitlin E. Fiorillo, MD; Vania Rashidi, BS Philip M. Westgate, PhD; Julie A. Jacobs, MPH Christina R. Studts. PhD: Matthew L. Bush. MD

**Objective:** To compare the prevalence of disruptive behavioral problems between children with hearing loss and normal hearing.

**Study design:** Prospective cross-sectional study

**Setting:** Tertiary academic center

**Patients:** Caregivers of children (2-5 years old) with normal hearing (n=39), hearing loss using hearing aid(s) (n=29), or cochlear implant(s) (n = 21) were recruited.

**Intervention(s):** Demographic information and a childhood mental health history were obtained. Childhood behavior and language development were assessed.

Main outcome measure(s): The Child Behavior Checklist, the Young Child-Diagnostic Interview Schedule and the MacArthur-Bates Communication Development Inventory (MBCDI)

**Results:** Similar distributions of race, socioeconomic and insurance status were observed across all groups. Parents of children with hearing loss were significantly more likely to report disruptive behavior (HA=41%, CI=38%) than parents of normal hearing children (10%) (p=0.002). Children with hearing loss were significantly more likely to meet criteria for oppositional defiance disorder (HA=48%, CI=48%) than normal hearing children (23%) (p=0.02). Few normal hearing children (8%) and no hearing impaired children had accessed mental health services (p=0.08). Normal hearing children were found to have more advanced language development on MBCDI than hearing impaired children (p=0.01), but controlling for MBCDI percentiles, the observed behavioral differences remained.

**Conclusions:** Controlling for language development, children with hearing loss have higher prevalence of disruptive behaviors than their normal hearing peers. These children are less likely to receive appropriate behavioral interventions. Further research is warranted to investigate the impact of disruptive behavioral problems on speech and hearing rehabilitation and to explore methods to improve access to effective behavioral intervention.

**Define Professional Practice Gap & Educational Need:** 1. The prevalence of behavioral problems in children with hearing loss is not known 2. No previous studies have investigated the type and degree of behavior problems using validated structured-diagnostic interviews in children who are deaf and hard of hearing 3. Few previous studies have looked at the family impact, parent sense of competence and parental stress in raising children with hearing loss

**Learning Objective:** 1. To determine the prevalence of behavior disorders in children with hearing loss versus normal hearing 2. To assess the level of parenting sense of competence and parent stress in parents of children with hearing loss versus normal hearing 3. To provide novel information on the impact on families of having children with hearing loss versus normal hearing

**Desired Result:** 1. To provide data obtained with validated structured diagnostic interviews on behavioral problems and related outcomes in children who are deaf and hard of hearing versus normal hearing

# **Development of Bone Density of the Temporal Bone in Healthy Subjects**

Kuniyuki Takahashi, MD; Yuka Morita, MD Shinsuke Ohshima, MD; Yamato Kubota, MD Shuji Izumi, MD; Arata Horii, MD

**Hypothesis:** The maturation of bone density in the temporal bone shows regional differences.

**Background:** Severe cases of acute otitis media can spread laterally and progress to acute mastoiditis in infants. However, they can cause intracranial complications more often in older children than in infants. Bone density maturation may affect the spread pattern of acute otitis media. Although the developing forms of mastoid air cells have been well studied, few research studies have been conducted on bone density maturation.

**Methods:** Eighty subjects aged 3 months to 42 years who had normal hearing participated in this study. Computed tomography (CT) values (Hounsfield unit [HU]) in various regions of the temporal bone, such as the otic capsule (OC), lateral surface of the mastoid cavity (LS), posterior cranial fossa (PCF), and middle cranial fossa (MCF), were measured. Bone density maturation was defined if the CT value exceeded 1000 HU, which is an indicator of compact bone. The age at which bone density maturation occurred was compared between regions of the temporal bone.

**Results:** The bone density of the OC showed maturation even just after birth, while that of the LS, PCF, and MCF matured at 1.7, 3.9, and 10.8 years of age, respectively. Bone density maturation showed significant regional differences.

**Conclusion:** Regional differences in bone density maturation may be a factor influencing the spread pattern of acute otitis media.

**Define Professional Practice Gap & Educational Need:** Lack of awareness of healthy temporal bone development

**Learning Objective:** To learn the development of bone density of various regions in healthy temporal bone

**Desired Result:** The bone density maturation showed significant regional differences. This may affect the spreading pattern of acute mastoiditis. We can learn that we should change management of acute mastoiditis according to age.

### Use of an Endoscope for Resection of Acoustic Neuroma through Middle Fossa Craniotomy

Adam N. Master, MS, MD; Daniel S. Roberts, MD, PhD Gregory P. Lekovic, MD, PhD

**Objective:** Visualization of the fundus of the internal auditory canal, especially below the transverse crest, is limited in conventional surgical exposure. The surgeon's impression of extent of resection may therefore be inaccurate. This study examines the utility of the endoscope to assess complete tumor removal in middle cranial fossa approach for acoustic neuroma

Study design: Prospective case series

**Patients:** Eight consecutive patients from December 2014 to August 2016 (6F,2M; mean age 51, range 22-66) undergoing middle fossa craniotomy for acoustic neuroma resection.

**Intervention:** After completion of microsurgical resection, surgical endoscopes were used to visualize the fundus of internal auditory canal and confirm extent of resection of tumor. Prior to introduction of the endoscope, the surgeon's response to questions including whether tumor residual is expected, et al. were recorded.

**Main outcome measures:** The primary outcome measure was to determine whether residual tumor was present after standard microsurgical resection, and whether the presence of residual tumor was expected or not expected by the surgeon. Additional data included were: 1) whether the endoscope changed management; 2) rate of hearing preservation; 3) facial nerve outcome.

**Results:** In two (25%) patients additional tumor was identified using the endoscope that was not seen during microsurgical resection. In both patients the residual tumor was removed under endoscopic guidance.

**Conclusion:** The endoscope may be a beneficial tool to identify residual tumor in patients undergoing middle fossa craniotomy for removal of acoustic neuroma.

**Define Professional Practice Gap & Educational Need:** Lack of knowledge of whether there is a benefit of using an endoscope in surgical resection of acoustic neuroma during middle fossa craniotomy

**Learning Objective:** Examines the utility of the endoscope to assess complete tumor removal in middle cranial fossa approach for acoustic neuroma

**Desired Result:** attendees will have a better understanding on the applications of endoscopic visualization in middle fossa craniotomy for acoustic neuroma resection

### The Combined Linguistic and Indexical Speech Perception Assessment (CLISPA): A Novel Measure of Speech Perception for Cochlear Implants

Chad V. Ruffin, MD; Taylor Curry, BS Janice Farlow, BS; Cullen Taylor, BS Charles W. Yates, MD

**Background:** Perception of the words in spectrally degraded speech requires high cognitive load that may interfere with simultaneous perception of indexical speech cues.

**Hypothesis:** The number of indexical cues that a listener perceives while simultaneously maintaining maximal levels of linguistic speech perception is significantly less than if assessed as a single outcome measure.

**Subjects:** Three different subgroups (N = 15 each) of normal hearing native speakers of American English.

**Methods:** The AzBio Sentence Lists were re-recorded to include greater indexical variation and more speakers. These stimuli were processed through a noise band vocoder. All three groups listened to the same sentence lists. The Sentence-only subgroup (N = 15) performed sentence discrimination. The Indexical-only subgroup (N = 15) identified indexical cues. The primary task of the Sentences+Indexical subgroup (N = 10) of targeted 15) was sentence perception followed by the secondary task of identifying indexical cues.

**Results:** The Sentences-only and Sentences+Indexical group exhibited high and similar levels of linguistic speech perception (94.7%  $\pm$  2.6 vs. 92.9%  $\pm$  2.7; p = 0.12). Indexical performance was significantly better in the Indexical-only vs. the Sentences+Indexical subgroup on gender discrimination (80.2%  $\pm$  4.7 vs. 72.5%  $\pm$ 7.2; p = 0.005) and emotion discrimination (41.8%  $\pm$  4.9 vs. 33.0%  $\pm$  7.2; p = 0.001). There were no differences in speaker identification between groups (40.9%  $\pm$ 11 vs. 37.3  $\pm$ 9.3; p= 0.42)

**Conclusions:** Under spectrally degraded conditions, listeners are significantly less fluent at integrating linguistic and indexical speech cues. This has important implications in designing outcome measures for CIs.

**Define Professional Practice Gap & Educational Need:** Highlight the insufficiency of traditional methods of assessing speech perception.

**Learning Objective:** The learner will identify alternative methods of assessing speech perception.

**Desired Result:** The learner will identify the gap in

### Environmental Sound Awareness among Cochlear Implant Users and its Relationship to Spectral Resolution and Speech Perception Skills

Michael S. Harris, MD; Lauren Boyce, BA David B. Pisoni, PhD; Aaron C. Moberly, MD

**Hypothesis:** The objective of this study was to evaluate environmental sound awareness (ESA) as a means of studying auditory perception among cochlear implant (CI) users and to determine to what extent ESA is correlated with speech perception and spectral resolution abilities.

**Background:** ESA is an ecologically valid auditory perception skill about which little is known following CI. It is largely a presumed benefit of implantation, often discussed clinically, but a paucity of objective data exists. The degree to which ESA relates to spectral resolution and speech perception skills is incompletely characterized among CI users.

**Methods:** A cohort of post-lingually deaf adult CI users and a cohort of normal hearing peers were assessed using an environmental sound identification task (Familiar Environmental Sound Test), a spectral ripple task, and a battery of speech and word perception tasks including recognition of isolated words (CID-22), phonetically balanced sentences (Harvard Sentences), and challenging sentences involving multiple speakers and dialects (PRESTO). The contributions of patient age and duration of auditory deprivation were considered.

**Results:** Environmental sound identification accuracy among CI users was poorer compared to that of normal hearing listeners. Speech perception skills were strongly and consistently correlated with environmental sound identification accuracy among CI users. This relationship remained significant when controlling for patient age and spectral resolution. No relationship was observed between duration of auditory deprivation and ESA.

**Conclusions:** ESA is a poorly understood aspect of auditory perception among CI users, which represents an opportunity for development of new assessment strategies and potential training targets.

**Define Professional Practice Gap & Educational Need:** 1. Environmental sound awareness is often discussed in clinical settings as an anticipated benefit of cochlear implantation. Little objective research has been performed to provide an evidence basis for this expectation. This study specifically addresses this gap. 2. Since the introduction of cochlear implants as an intervention for hearing loss, a need for non-speech measures of auditory sensitivity has been clear. This study introduces data that supports the use of environmental sound awareness as a new avenue of auditory perception assessment following implantation. 3. This study addresses the incompletely characterized relationship among speech perception, spectral resolution, and environmental sound awareness.

**Learning Objective:** 1. The learner will come away with an evidence base to ground their counseling of patients on environmental sound awareness following cochlear implantation. 2. The learner will be up-to-date on the potential utility of environmental sound awareness as an additional target for auditory perception assessment among patient with hearing impairment and cochlear implant users. 3. The learner will have an appreciation for the connection between speech perception skills and environmental sound awareness: how they are similar, how they are dissimilar, and why the difference matters.

**Desired Result:** 1. Learners will have the opportunity to apply what they learn regarding environmental sound awareness among cochlear implant users in their routine counseling of patients anticipating cochlear implant surgery -- this is a topic that families and patients often raise in clinical encounters. 2. Learners may consider incorporating the concept of non-speech indicators of auditory perception into their evaluation of cochlear implant candidates. 3. Learners may be able to use knowledge that speech perception skills are highly correlated with environmental sound awareness to inform patient expectations and in considering non-speech auditory perception assessments for patient who cannot perform speech perception testing.

#### Risk Factors of Recurrence in Pediatric Congenital Cholesteatoma

Yuka Morita, MD, PhD; Kuniyuki Takahashi, MD, PhD Shuji Izumi, MD, PhD; Yamato Kubota, MD, PhD Shinsuke Ohshima, MD, PhD; Yutaka Yamamoto, MD, PhD Arata Horii, MD, PhD

**Objective:** To examine the risk factors of residual and retraction recurrence in pediatric congenital cholesteatoma.

Study Design: Retrospective chart review.

**Setting:** University hospital.

**Patients:** Sixty-seven patients having tympanic type of congenital cholesteatoma under 15 years-old at surgery.

**Interventions:** Canal wall-up tympanoplasty (n=30) or transcanal atticotomy (n=37) was performed depending on cholesteatoma extension, 16 of which were followed by second-look surgery. Preoperative CT before second-look surgery or follow-up CT was performed to detect residual recurrence one year after the surgery. Cholesteatoma found at the second surgery was also included in the recurrence. All patients had no recurrent cholesteatoma at the last follow-up (Median, 61 months after surgery).

Main outcome measures: Possible predictive factors were compared between the groups.

**Results:** Residual cholesteatoma and retraction cholesteatoma occurred in 21% and 6%, respectively. There was no significant difference in age, gender, and type of cholesteatoma (open or closed) between the groups; however, Potsic stage and status of stapes involvement were more advanced in the residual recurrence group. All residual lesions could be detected by follow-up CT or by second-look surgery. All of four retraction recurrence (+) patients were male and young at the surgery.

**Conclusions:** Recurrence mostly occurred as residual cholesteatoma, suggesting that CT is recommended as a follow-up tool for congenital cholesteatoma. Advanced lesions had the risk of residual recurrence, suggesting that complete removal of epithelium is important. Although rare, young advanced-stage patients had risk of retraction cholesteatoma and therefore normal mucosa should be preserved as much as possible for these patients.

**Define Professional Practice Gap & Educational Need:** Lack of awareness of risk factors of recurrent lesions in pediatric congenital cholesteatoma

**Learning Objective:** To examine the risk factors of residual and retraction recurrence in pediatric congenital cholesteatoma.

**Desired Result:** Potsic stage and status of stapes involvement were more advanced in the residual recurrence group.

### Fluoroscopy-assisted Transnasal Onyx Occlusion of the Eustachian Tube for Lateral Skull Base Cerebrospinal Fluid Fistula Repair

Neil S. Patel, MD; Matthew L. Carlson, MD

**Hypothesis:** Transnasal occlusion of the Eustachian tube (ET) with Onyx liquid embolic solution (LES) is feasible for lateral skull base cerebrospinal fluid (CSF) leaks.

**Background:** The rate of CSF leak following vestibular schwannoma surgery can be as high as 12%, exposing patients to risk of meningitis, pneumocephalus, and the need for lumbar drainage or additional surgery. We sought to develop a minimally-invasive approach to occlusion of the ET when CSF fistulae develop after lateral skull base surgery.

**Methods:** A CSF fistula model was developed by the authors using fresh cadaveric heads. Using a transtympanic needle, regulated pressurized pigmented saline was continuously irrigated into the middle ear cleft and visualized endoscopically in the nasopharynx. An angioembolization catheter and Onyx 18 LES was placed just medial to the bony ET. Under endoscopic and fluoroscopic guidance, the material was deployed into the bony ET segment up to the middle ear space.

**Results:** In two cadavers, a CSF fistula model was developed and endoscopic visualization of irrigant flow into the nasopharynx was confirmed. Fluoroscopy provided adequate anatomic views of the ET and middle ear, in addition to dynamic views of embolization. Cessation of flow after occlusion was achieved with pressures up to 25 mm Hg, mimicking physiological intracranial pressure in patients with meningitis or benign intracranial hypertension.

**Conclusion:** Eustachian tube occlusion with Onyx is feasible in a novel cadaveric CSF leak model. This may be employed as a short, outpatient treatment for intermittent or low-flow CSF fistulae following lateral skull base surgery.

**Define Professional Practice Gap & Educational Need:** 1. Lack of widely accepted technique for occlusion of the Eustachian tube for lateral skull base CSF fistulae 2. Lack of minimally-invasive treatment for lateral skull base CSF fistulae

**Learning Objective:** 1. The learner will be able to describe a minimally-invasive technique for transnasal Eustachian tube occlusion and acknowledge its value in repairing lateral skull base CSF fistulae

**Desired Result:** 1. Attendees will include this option among presently available treatments for lateral skull base CSF fistulae

#### **Heat Shock Proteins in Human Perilymph**

Athanasia Warnecke, MD; Heike Schmitt, MD Ariane Roemer, MD; Carsten Zeilinger, MD Martin Durisin, MD; Hinrich Staecker, MD, PhD Thomas Lenarz, MD, PhD

Objective: Data about the etiology and pathophysiology of inner ear diseases leading to hearing loss and especially changes of the composition of the perilymph fluid are still very limited. This is mainly due to the difficult access to structures and cochlear fluids. Heat shock proteins (HSP) belong to a superfamily of stress proteins and promote refolding of denatured proteins. Interestingly, HSP may either prevent or promote cell injury. The aim of the study was to analyze the presence of HSP in human perilymph derived from cochlear implant and correlate patients to their presence with audiological data. etiologic

**Methods:** Sampling of the perilymph was performed during CI implantations and vestibular schwannoma surgeries with translabyrinthine approach via the round window or the semicircular canal. Individual proteins were identified by a shot-gun proteomics approach and data-dependent analysis using orbitrap mass spectrometry (Thermo Fisher Scientific) and Max Quant software for identification. The residual hearing of patients was determined by prae- and postoperative data and compared with different HSP identified in the perilymph. Also, differences in HSP occurrence of children and adults and vestibular schwannoma patients were analyzed.

**Results:** 10 subgroups of HSP were identified in HP samples. Only 33% of the patients with protected residual hearing showed an expression of HSP90. However, in two of three patients that lost their hearing, HSP90 (alpha and the beta subtype) were identified. In the perilymph of all patients with preserved residual hearing, HSP70 (subtypes 1 and 6) was identified, whereas subtype 4 was identified in only 17%.

**Conclusions:** In-depth proteome analyses of perilymph samples in correlation to patients' audiogram data leads to the hypothesis that HSP70 is associated with preservation of residual hearing after cochlear implantation, whereas HSP90 is associated with loss of residual hearing.

**Define Professional Practice Gap & Educational Need:** 1) Currently there are no prior studies showing sampling of perilymph as a potential diagnostic test for inner ear disease 2) There currently is no way to sample inner ear tissue for diagnostic purposes

**Learning Objective:** 1) Show that perilymph sampling could be used to better understand inner ear disease 2) Perilymph sampling is feasible without damaging the ear

**Desired Result:** 1) The inner ear produces proteins that may alter its response to stress. 2) Different disease states may be defined by changes in the perilymph proteome

#### Multidisciplinary Management of Head and Neck Malignancies Involving the Lateral Skull Base

Nauman Manzoor, MD; Kate Clancy, BA Rod Rezaee MD; Chad Zender, MD Sarah Mowry, MD; Cliff Megerian, MD Maroun Semaan, MD

**Objective:** Analyze loco-regional control after management of advanced stage malignancies involving lateral skull base.

**Study design:** Retrospective chart review.

**Setting:** Tertiary care center.

Patients: Adult patients with malignancies involving lateral skull base.

**Intervention(s):** Lateral temporal bone resection with transpetrous approach to the stylomastoid foramen and jugular foramen  $\pm$  post operative radiotherapy.

#### **Main outcome measure(s):**

Loco-regional control and distant failure rates. Facial nerve sacrifice and post operative radiotherapy was assessed using the using Kaplan-Meier method and compared with log-rank test.

**Results:** Between 2011 and 2016, 29 patients were identified. Mean age was 73 years and 86.2 % were male. Median follow up was 8.3 months (range 0.6-46.8). 65.5 % cases were recurrent disease. Site of origin was external auditory canal (EAC) in 9 (31 %) cutaneous (auricle, lateral facial and scalp) in 17 (58.6%) and parotid in 3 (10.3 %). Of the 9 EAC cases, 5 were T2 while 2 cases each were T3 and T4 (Pittsburg staging system). 8 cutaneous primaries were AJCC stage 3 (temporal bone involvement) and 9 were AJCC stage 4 (facial nerve involvement at skull base). Histologically 72.4 % were squamous cell cancer (SCC), 17.2 % were basal cell cancer (BCC), and 10.3 % were malignant parotid disease (stage T4a). The facial nerve was sacrificed in 10 cases (32.3 %). 20 patients (69.0 %) received post operative radiotherapy. Five (17.2 %) patients had recurrent disease (3 locoregional and 2 distant). Unadjusted recurrence free survival trended toward significance when facial nerve was not sacrificed (p=0.06, log rank). Unadjusted recurrence free survival was significantly improved when post op radiotherapy was used (p=0.04, log rank).

**Conclusions:** Improved loco-regional control can be achieved in advanced stage malignancies involving lateral skull base with multi-modality treatment. Facial nerve sacrifice portends a worse survival.

**Define Professional Practice Gap & Educational Need:** Inconsistencies with the extent of lateral skull base resection and utilization of post operative radiotherapy.

**Learning Objective:** Multi-modality treatment with ablative procedure including facial nerve sacrifice (when indicated) and adjuvant radiation improves loco-regional control in various malignancies involving the lateral skull base.

**Desired Result:** Pre-operative planning to achieve negative margins including facial nerve sacrifice when indicated and the utility of post operative radiation. Extending traditional LTBR to remove disease around stylomastoid foramen and jugular bulb in the presence of peri-neural spread.

### Single Stage Removal of Osseointegrated Implant and Insertion of Ipsilateral Cochlear Implant with Myofacial Temporalis Muscle Flap: A Novel Technique

Michael F. Foster, DO; Douglas D. Backous, MD

**Objective:** To describe a novel technique for conversion from an ipsilateral osseointegrated implant (OI) to a cochlear implant (CI) utilizing a single stage myofascial tissue rearrangement and traditional cochlear implant insertion.

Case: One patient with single-sided deafness (SSD) whom had an OI placed using the dermatome technique had their bone anchored hearing aid abutment removed in office then later underwent a single stage operation including fixture removal then rotation of a temporalis myofascial flap to fill in the soft tissue defect immediately followed by a traditional uncomplicated cochlear implantation.

**Conclusion:** With emerging interest in cochlear implantation for single sided deafness we predict an increase in the number of patients undergoing cochlear implantation whom have previously had an ipsilateral OI. This can be complicated by the significant soft tissue defect in the anticipated location of a CI receiver stimulator after a past OI placement using the dermatome technique. This case presents a safe and efficient technique to accomplish cochlear implantation and traditional placement of the receiver-stimulator in this group of patients. A single staged procedure prevents inconvenience to the patient and the potential complications of multiple trips to the operating room.

**Define Professional Practice Gap & Educational Need:** 1. Lack of contemporary knowledge

**Learning Objective:** 1. To provide a novel technique that will become more prevalent as Cochlear Implantation for Single-Sided Deafness is performed more frequently in patients with poor performance using an Osseointegrated Implant

**Desired Result:** Attendees will use this technique when necessary.

### 3D Reconstruction and Topographical Analysis of Human Cochlear Nucleus: Designing a Better Auditory Brainstem Implant Array

Vivek V. Kanumuri, MD; Osama Tarabichi, MD Maria Duarte, BS; Julian Klug, BS M. Christian Brown, PhD; Daniel J. Lee, MD

**Hypothesis**: Quantification of the cochlear nucleus curvature will inform design of a flexible auditory brainstem implant (ABI) array.

**Background**: The ABI is an auditory neuroprosthesis placed on or near the surface of cochlear nucleus (CN) to restore hearing in children and adults who are not candidates for cochlear implantation. Current ABI arrays are stiff and do not conform to brainstem topography. Recent advancements in flexible electrode arrays allow for anatomy conforming electrical stimulation. Herein, we measure the surface curvature of the cochlear nucleus and adjoining regions to assist in design of next generation flexible ABI arrays.

**Methods**: Data collection and sharing for this project was provided by the MGH-USC Human Connectome Project and was IRB exempt. Images were obtained from the advanced Siemens 3T Connectome imaging system. T1 weighted MRI sequences were imported into segmentation software (Amira; TGS, Berlin, Germany) to create accurate volumetric models and surface rendering. Local curvature analysis was performed on manually-selected regions of interest.

**Results**: 3D reconstruction of the left cochlear nucleus and adjoining brainstem regions was successfully rendered. Mean principal curvature values were calculated along 5 equidistant points along the lateral – medial (range: -0.28 to 0.08) and rostral-caudal (range: -0.10 to 0.11) axes. These values were plotted on kmin-kmax graphs and indicated significant complexity in curvature with broad variation and a unique combination of convex and concave surfaces.

**Conclusions**: This study reveals the complex curved topography of the human cochlear nucleus. This supports and informs the design of soft moldable flexible electrodes for the ABI.

**Define Professional Practice Gap & Educational Need:** Lack of Contemporary Knowledge

**Learning Objective:** Define the complex surface topography of the cochlear nucleus and auditory brainstem. Gain insight into improved Auditory Brainstem Implant (ABI) electrode design.

**Desired Result:** The attendees will gain an improved understanding of the surface anatomy of the cochlear nucleus and adjacent brainstem along with an introduction into recent advancements in Auditory Brainstem Implant technology

### Cartilage or Abscess Found Instead of Cholesteatoma in Five Patients who Underwent Tympanomastoidectomy Following False Positive Non-Echo-Planar Diffusion-Weighted MR Imaging

Alexander Sevy, MD; Joshua Sappington, MD Rahul Mehta, MD; Moises Arriaga, MD, MBA

**Objective:** To evaluate the false positive cholesteatoma diagnoses on MRI in results from patients who underwent tympanomastoidectomy surgery.

Study design: Retrospective case series.

**Setting:** Tertiary referral center. Patients: n=5 adults, single institution with diagnosis of cholesteatoma based on clinical findings and Non-echo-planar diffusion-weighted MR Imaging who underwent tympanomastoidectomy for resection of presumed cholesteatoma. All patients were found to have no evidence of cholesteatoma during surgery and surgical pathology.

**Intervention:** Patients with clinical suspicion for cholesteatoma underwent Non-Echo-Planar Diffusion-Weighted MR Imaging. Half-Fourier acquisition single-shot turbo-spin echo (HASTE) protocols were used for this imaging. All of these patients underwent tympanomastoidectomy and surgical findings and surgical pathology specimens were analyzed.

**Main outcome measure:** Comparison of preoperative HASTE MR imaging with surgical findings and surgical pathology results. Results: 80% (4/5) with falsely positive detection of cholesteatoma on HASTE MRI were found to have cartilage only at the site of presumed cholesteatoma, and in 20% (1/5) there was an abscess at the site of presumed cholesteatoma.

**Conclusions:** Non-echo-planar diffusion-weighted MR Imaging has become the gold standard in the detection of cholesteatoma, particularly in the setting of possible residual or recurrent disease with better sensitivity and specifity than any other current clinical imaging. However, it is important to realize that the specifity of these imaging protocols are not 100%. What may resemble cholesteatoma on MR imaging may indeed be other entities such as cartilage or infection, which should be included in the differential of patients with prior ear surgery.

**Define Professional Practice Gap & Educational Need:** 1. Lack of awareness

**Learning Objective:** Awareness of false positives in Non-echo-planar diffusion-weighted MR Imaging, and discussion on ways to reduce false positive scan results, in addition to more information for counselling of patients

**Desired Result:** Review of images by surgeon prior to surgery with suspicion for potential false positives and additional information for counselling to patients

### Duration of Eligibility Prior to Cochlear Implantation: Have We Made Any Progress?

Eric N. Appelbaum, MD; Shannon S. Yoo, BS Robert A. Perera, PhD; Daniel H. Coelho, MD

**Objective:** To determine if eligibility (as defined as the duration of severe to profound hearing loss prior to cochlear implantation (CI)) has changed over the 30 years since FDA approval.

**Data Sources:** English language, peer-reviewed articles, theses, and trial data available through PubMed and Cochrane Library databases up until and including May 31st, 2016.

**Study Selection:** 1006 unique articles were identified. Prospective studies that reported duration of severe/ profound hearing loss before CI in adult patients were included. Retrospective studies, reviews, meta-analyses, articles reporting pediatric or mixed data, hybrid/electroacoustic CI, and articles from centers outside the United States were excluded. 71 studies met inclusion criteria and were included for analysis.

**Data Extraction:** Contributing authors independently reviewed included studies for data validity and applicability.

**Data Synthesis:** Meta-regression was used to assess the relationship between the year of publication and duration of hearing loss. To account for a possible age effect, a second model was estimated including mean age at time of study as a covariate.

**Conclusions:** A positive association between study year and the duration of hearing loss prior to implantation was found showing a 0.30 year increase in the duration of hearing loss for every increasing study year. Contrary to conventional assumption, duration of eligibility for CI appears to be increasing. Though the reasons for this are not clear, current strategies to increase both awareness and access to CI appear to be falling short.

**Define Professional Practice Gap & Educational Need:** 1. Lack of awareness 2. Lack of contemporary knowledge

**Learning Objective:** 1. To determine if eligibility (as defined as the duration of severe to profound hearing loss prior to cochlear implantation (CI)) has changed over the 30 years since FDA approval.

**Desired Result:** 1. Attendees will attain greater understanding of change in referral patterns of cochlear implantation candidates.

## Getting the Feeling? Salience of Musical Emotion with a Cochlear Implant

David R. Friedmann, MD; Daniel Jethanamest, MD Joshua Horton MD; David Landsberger, PhD J. Thomas Roland Jr., MD; Susan B. Waltzman, PhD

**Objective:** Music like all art seeks to communicate with the audience. Some are motivated by the emotional connection to its content. Studies cochlear implants do not faithfully represent the psychophysical relationships inherent in music. We sought to determine whether targeted musical emotions are conveyed through electric only stimulation.

**Intervention:** Twenty musical pieces from western musical canon for which there was concordance among normal hearing (NH) listeners as to the emotion conveyed: (1)happy, (2)sad, (3)scared or, (4) peaceful were presented. Cochlear implant (CI) subjects(n=20) were presented with stimuli through sound field with a specifically designed IOS application administered on an iPad. Subjects also completed a musical background questionnaire.

**Results:** CI users identified the target emotion in only 52% of cases compared to NH listeners who correctly identified the target emotion in 87% of pieces. "Happy" pieces were recognized in 86% of cases by CI users while the remaining target emotions were much more difficult for CI users sad (44%), scary (38%) and peaceful (41%) compared with NH raters. Length of deafness, CI experience, speech perception scores and prior musical experience did not correlate with performance on this task.

**Conclusions:** Current CI technology do not convey the range of emotions conveyed in music as recognized by NH subjects. Specific cues in musical pieces with a target emotion of happy seem to be better preserved for CI users but not for the others tested. This may explain the lack of interest many patients have in music after CI despite a passion for music before implantation.

**Define Professional Practice Gap & Educational Need:** Studies cochlear implants do not faithfully represent the psychophysical relationships inherent in music though it does not explain musical enjoyment. We believe emotional content may be related to music enjoyment measures.

**Learning Objective:** We sought to determine whether targeted musical emotions are conveyed through electric only stimulation.

**Desired Result:** Current CI technology may not convey the range of emotions conveyed in music as recognized by NH subjects. This may explain the lack of interest many patients have in music after CI despite a passion for music before implantation and help target improvements in processing strategies that preserve such cues

## Hearing Outcomes after Stereotactic Radiosurgery for Glomus Jugulare Tumors

Neil S. Patel, MD; Michael J. Link, MD Colin L. W. Driscoll, MD; Bruce W. Pollock, MD Brian A. Neff, MD; Christine M. Lohse, MS Matthew L. Carlson, MD

**Objective:** Describe audiometric outcomes following stereotactic radiosurgery (SRS) for glomus jugulare tumors (GJT).

**Study Design:** Retrospective review.

**Setting:** Tertiary referral center.

**Patients:** Patients with serviceable hearing (AAOHNS Class A or B) and serial audiometric follow-up who underwent Gamma Knife SRS for GJT between 1990 and 2015.

**Intervention(s):** Gamma Knife SRS.

Main outcome measure(s): Preservation of serviceable hearing; univariate and multivariate associations with time to non-serviceable hearing.

**Results:** Of 81 patients with GJT who underwent SRS during the study period, 34 (65% female, median age 54) had pretreatment class A or B hearing and post-treatment audiometry. Median tumor volume at the time of treatment was 7180 mm<sup>3</sup>, the mean cochlear point dose was 5.6 Gy (range 2-8.8 Gy), and the median marginal and maximum tumor dose were 16 and 32 Gy, respectively. Seven patients (21%) developed non-serviceable hearing (class C or D) at a median of 13.2 months following SRS (IQR 0.4-2.0 years). Among those who maintained serviceable hearing, median audiometric follow up was 35 months (range 5-123 months). The Kaplan-Meier estimated rates of serviceable hearing at 1, 3, and 5 years following SRS were 91%, 79%, and 79%, respectively. Associations between time to non-serviceable hearing and pre-treatment hearing levels, tumor volume, marginal and maximum dose, cochlear dose, and age are reported.

**Conclusions:** The short- and intermediate-term risk of progression to non-serviceable hearing following SRS for GJT is low. In contrast to vestibular schwannoma, the impact of marginal tumor dose and cochlear dose on hearing preservation appears to be less significant.

**Define Professional Practice Gap & Educational Need:** 1. Lack of understanding of the effect of stereotactic radiosurgery on hearing in patients with glomus jugulare tumors. 2. Lack of contemporary knowledge on the etiology of hearing loss after radiosurgery for glomus jugulare tumors.

**Learning Objective:** 1. Describe the risk of progression to non-serviceable hearing after stereotactic radiosurgery for glomus jugulare tumors. 2. Describe factors associated with a higher risk of developing sensorineural hearing loss after radiosurgery for glomus jugulare tumors.

**Desired Result:**1. Attendees should be able to estimate the frequency of progression to non-serviceable hearing among patients with glomus jugulare tumors 2. Attendees should acknowledge the difference between hearing outcomes following stereotactic radiosurgery for vestibular schwannoma and glomus jugulare tumors.

### Endoscopic Infracochlear Approach for Drainage of Petrous Apex Cholesterol Granulomas: A Case Series

Cameron C. Wick, MD; Alexander R. Hansen, BS Joe Walter Kutz Jr., MD; Brandon Isaacson, MD

**Objective**: To describe the feasibility and technical nuances of a transcanal endoscopic infracochlear approach for drainage of petrous apex cholesterol granulomas.

**Study Design:** Retrospective case review.

**Setting:** Tertiary care university hospital.

**Patients**: A 32-year-old male with bilateral petrous apex cholesterol granulomas and a 54-year-old male with a left-sided petrous apex granuloma each with symptoms necessitating surgical intervention.

**Interventions**: Transcanal endoscopic infracochlear approach for drainage of the cholesterol granulomas.

Main Outcome Measures: Operation efficacy, corridor size, and perioperative morbidity.

**Results**: All three cholesterol granulomas were successful drained without violating the cochlea, jugular bulb, or carotid artery. The dimensions of the infracochlear surgical corridor measured 5 mm by 6 mm, 3.5 mm by 3.5 mm, and 6 mm by 4 mm, respectively. All corridors facilitated visualization within the cyst and allowed lyses of adhesions for additional cyst content eradication. All patients had resolution of their acute symptoms. Two of the three subjects had serviceable hearing prior to and after their procedures. One patient required revision surgery 2-months after their initial procedure secondary to recurrent symptoms from acute hemorrhage within the cyst cavity. The infracochlear tract in this patient was noted to be patent.

**Conclusions**: A transcanal endoscopic infracochlear approach appears to be a viable and effective technique for the management of cholesterol granuloma. The surgical access was wide enough to introduce the endoscope into the petrous apex cavity in each case. Further studies are needed to compare the efficacy and perioperative morbidity versus the traditional post-auricular transtemporal approaches.

**Define Professional Practice Gap & Educational Need:** The petrous apex remains a challenging anatomical location to reach surgically. This has led to multiple approaches for the surgical management of petrous apex cholesterol granuloma. We report on the feasibility and efficacy of the endoscopic transcanal infracochlear approach for drainage of petrous apex cholesterol granulomas. This route has potential to be less invasive and less morbid than the more traditional transtemporal routes.

**Learning Objective:** To describe the feasibility and technical nuances of a transcanal endoscopic infracochlear approach for drainage of petrous apex cholesterol granulomas.

**Desired Result:** Knowledge of this surgical approach may change the way neurotologists approach petrous apex cholesterol granulomas.

### The Effect of Materials Used for Superior Canal Dehiscence Repair on Inner-ear Pressures

Deepa J. Galaiya, MD; Xiying Guan, PhD Y. Song Cheng, MD; Hideko Heidi Nakajima, MD, PhD

**Introduction:** The effects of repairing superior canal dehiscence (SCD) on the vestibular and auditory system are poorly understood. By measuring inner-ear pressures, we can determine the input drive for the cochlea and the superior canal ampulla in a controlled manner, furthering our knowledge of such repairs. Using this technique, we studied how different materials used to repair the SCD affect inner-ear pressures evoked by air and bone conduction (AC and BC) stimulation.

**Methods:** Intracochlear pressures in the scala vestibuli (Psv) and scala tympani (Pst) were measured in 6 human cadaveric temporal bones using micro-fiberoptic pressure sensors sealed and firmly glued to the otic capsule near the oval and round windows. Psv, Pst, ear-canal pressure (Pec) and stapes velocity were measured while stimulated by AC (with speaker at ear canal) or BC (with a bone anchored hearing aid) in normal intact superior canal, with SCD, and with repaired SCD. Repairs were made with soft materials (e.g. dental impression material, Jeltrate), hard materials (e.g. dental cement), and bone wax.

**Results:** With AC, SCD reduced both Psv and Pst, and reduced the cochlear and ampulla input drives at low frequencies. Patching the dehiscence with various materials reversed this effect to the normal initial pressures, as long as a hermetic seal was achieved. In BC, SCD reduced Psv across a limited low-frequency bandwidth, but did not generally change Pst. Estimates of cochlear and ampulla input drives were reduced at limited low-frequency bandwidths. The type of material used to repair the SCD affected the ability to reverse the effect of SCD during BC. A soft material was not effective in fully reversing the SCD effect during BC. However, a harder material could reverse the SCD effect on BC. Bone wax had a variable impact on SCD reversal.

**Discussion and Conclusion:** Reversing SCD effects on bone-conducted inner-ear pressures were more challenging than reversing air-conducted inner-ear pressure effects. The type of material used for SCD repair affected reversal of the SCD effect on the input drive to the auditory and vestibular systems during BC.

**Define Professional Practice Gap & Educational Need:** 1) Lack of a clear model to explain the effects of superior canal dehiscence on air-conducted and bone-conducted inputs to the cochlea and ampulla. 2) Absence of an understanding of which kind of surgical repair of superior canal dehiscence will result in the most complete reversal of these effects on air-conducted and bone-conducted inputs to the cochlea and ampulla.

**Learning Objective:** 1) Understand the effects of superior canal dehiscence on air-conducted and bone-conducted inputs to the cochlea and ampulla. 2) Understand that the material properties of the substance used to repair the superior canal dehiscence can affect how well the repair can reverse the effects of superior canal dehiscence.

**Desired Result:** 1) Consider which of the myriad materials that are used to surgically repair superior canal dehiscence would be most effective in reversing the symptoms of superior canal dehiscence, based on its physical and material properties.

### A Report of Two Distal AICA Aneurysms Presenting as IAC Masses A Review of the Literature and Characteristic Radiologic Features

Michael F. Foster, DO; Douglas D.Backous, MD Roberto A. Cueva, MD; P. Cody Buchanan, DO

**Objective:** Medial branch of Distal Anterior Inferior Cerebellar Artery (AICA) aneurysms are exceedingly rare, but potentially lethal if ruptured. We report on two cases of a distal AICA aneurysm masquerading as an intracanalicular vestibular schwannoma and review the literature.

Case 1: 47yo Hispanic male experienced right sudden SNHL and was treated with combined high dose oral prednisone taper and intratympanic (IT) Dexamethasone injections with complete recovery. An MRI revealed a right 1cm intracanalicular internal auditory canal lesion. The patient underwent elective craniotomy through a retrosigmoid approach. Intraoperatvely the mass was identified as an aneurysm arising from a loop of the AICA within the IAC. The aneurysm was successfully clipped. 4-year post op angiogram demonstrated patent AICA without aneurysm.

Case 2: 51yo Caucasian male with sudden deafness on the left side. Subsequent MRI showed a 7x7mm left cerbellopontine angle (CPA) mass with unusual calcification. Patient elected to have the mass surgically resected via a translabyrinthine approach. Inraoperatively the mass was identified as a distal AICA aneurysm and was clipped successfully. Postoperative angiogram revealed no residual or recurrent aneurysm and a patent AICA.

**Conclusion:** Vestibular Schwannomas make up the 85% of CPA and intracanalicular masses. Aspects of our cases and review of the literature provide insight into atypical signs and symptoms that can raise suspicion for a distal AICA aneurysm. These characteristics can provide prudent insight when counseling a patient on etiology of their IAC lesion and the plan of intervention.

**Define Professional Practice Gap & Educational Need:** 1. Lack of awareness 2. Lack of contemporary knowledge

**Learning Objective:** 1. To call to attention the prevalence and characteristic clinical and radiologic features of a distal AICA aneurysm that can mimic the presentation of a vestibular schwannoma.

**Desired Result:** Physicians will critically examine an atypical appearing IAC mass and order further angiographic testing when necessary.

#### Safety of Intratympanic Fludrocortisone Injection in a Mouse Model

George Kurien, MD; Vincent Lin, MD

**Background:** Intratympanic glucocorticoid injection is commonly used in the treatment of various otologic conditions. In addition to their anti-inflammatory effect, there is evidence that their mineralocorticoid effects on ionic transport in the inner ear may be partially responsible for their therapeutic effect. Intratympanic mineralocorticoid injection has not been tested for safety.

**Hypothesis:** Fludrocortisone (a mineralocorticoid) is safe for intratympanic injection in mice, with no difference in hearing thresholds from carrier injection in normal hearing mice.

**Methods:** 25 Swiss Webster Mice between 9-12 weeks of age had ABR testing followed by transbullar injection of one of four treatments. The mice had repeat ABR testing at 1 week and 4 weeks post-procedure. Treatment groups were - (A) Fludrocortisone (0.14mg/mL) in Normal Saline (B) Fludrocortisone (0.14mg/mL) in DMSO (0.5%) in Normal Saline (C) DMSO (0.5%) in Normal Saline

**Results:** Six mice were in groups A,B, D, and 7 mice in group C. Five mice had middle ear infections following injection, and resultant hearing loss (A,B,D,D,D). At 1 week, the mean difference from baseline in hearing thresholds at 8, 16, and 32KHz were (A)1dB,-1dB,8dB, (B)14dB,11dB,21dB (C)9dB,2dB,22dB (D)13dB,13dB,52dB. Three mice died between week 1 and 4 post-procedure (A,B,C) for unrelated reasons. At 4 weeks, the mean difference from baseline in hearing thresholds at 8, 16, and 32KHz were (A)3dB,-3dB,14dB, (B)8dB,4dB,20dB (C)3dB,-2dB,18dB (D)12dB,18dB,48dB. The Kruskal-Wallis test showed no statistically significant differences between groups at any frequency at any time point (p>0.05).

**Conclusions:** Fludrocortisone has a similar safety profile in intratympanic injection in normal hearing mice when compared to carrier. Intratympanic mineralocorticoid injection has potential utility in the treatment of disorders of the inner ear.

**Define Professional Practice Gap & Educational Need:** 1. Lack of intratympanic treatments available for inner ear conditions. 2. Uncertainty about the exact mechanism of action of intratympanic injected corticosteroids.

**Learning Objective:** 1. Examine the potential role for injected intratympanic mineralocorticoids in the treatment of inner ear conditions. 2. Understand the safety profile of intratympanic mineralocorticoid in a mouse model.

**Desired Result:** 1. Further investigate the role for intratympanic mineralocorticoids in the treatment of inner ear conditions.

### Ultra Long-term Audiometric Outcomes in the Treatment of Vestibular Schwannoma with the Middle Cranial Fossa Approach

Joseph P. Roche, MD; Erika A. Woodson, MD Marlan R. Hansen, MD; Bruce J. Gantz, MD

**Objective:** Define the long-term audiometric outcomes from vestibular schwannomas treated with the middle cranial fossa (MCF) approach.

**Study design:** Retrospective records review.

**Setting:** University-based tertiary referral center.

**Patients:** Patients undergoing treatment of small vestibular schwannomas with the MCF approach.

**Intervention(s):** MCF exposure and treatment of the small vestibular schwannomas.

Main outcome measure(s): Demographic and audiometric variables were assessed.

**Results:** Thirteen subjects had audiometric data for review. The average time between surgery and the most recent audiometric testing was 14 years (range 10-18 years). The mean baseline pure-tone average (PTA) was 36dB and the most recent PTA was 59dB in the operated ear. The mean baseline PTA was 16dB and the most recent PTA was 37dB in the non-operated ear. The mean change from baseline to most recent follow-up was a threshold elevation of 22dB and 19dB in the operated and non-operated ears, respectively. The mean baseline speech discrimination score (SDS) was 83% and the most recent SDS was 73% in the operated ear. The mean baseline SDS was 98% and the most recent SDS was 94% in the non-operated ear. The mean changes from baseline to most recent follow-up were 10% and 0% SDS depression in the operated and non-operated ears, respectively. The rates of changes in PTA and SDS were not statistically different between the operated and non-operated ears.

**Conclusions:** Surgically preserved hearing is maintained in the majority of patients at more than 10 years from surgery. PTA and SDS changes were similar between the ipsilateral and contralateral ears.

**Define Professional Practice Gap & Educational Need:** The middle cranial fossa approach to the internal acoustic canal allows for the treatment of vestibular schwannomas with hearing preservation in appropriately selected patients. Currently, few reports exist documenting the long-term hearing outcomes at greater than 10 years. This report adds a series of patients with audiometric data at greater than 10 years from the index procedure and compares these outcomes to hearing results from the contralateral ear.

**Learning Objective:** To recognize the long-term hearing outcomes from treatment of vestibular schwannoma via the middle cranial fossa approach.

**Desired Result:** To aid in the decision making process when choosing how to best approach small vestibular schwannomas and understand the long-term hearing outcomes that can be expected with the MCF approach.

#### Validation of a Subjective Visual Vertical Test App

George Kurien, MD; Day Dai, BSc; Leah Smith, MA Euna Hwang, MDCM; Vincent Lin, MD

**Objective:** To validate a subjective visual vertical test app in healthy controls

Study design: Cross-sectional sample

**Setting:** Medical office

**Patients:** Volunteers

**Intervention:** Healthy subjects underwent a subjective visual vertical (SVV) test using the validated bucket with plumbline method and simultaneously had a measurement made with the Visual Vertical iOS app. Each subject underwent 10 iterations of the test.

Main outcome measure(s): Internal reliability, intraclass correlation, comparison of means.

**Results:** 22 healthy subjects underwent testing. Mean (SD) results using the plumbline were 0.330 (1.37), and using the app, were 0.350 (1.34). Internal reliability (Cronbach's) for both tests were high at 0.976 for the plumbline method, and 0.978 for the app. Correlation (Pearson coefficient) between the two testing methods was high at 0.974 (p < 0.0001). Comparison of means showed a mean (SD) difference of 0.02 (0.12), which was not statistically significant (p=0.382, Paired T-test).

**Conclusions:** The Visual Vertical iOS app is a reasonable alternative to the standard SVV plumbline test with a high degree of internal reliability, correlation with the established test, and no statistically significant difference from the established test when tested in healthy patients. Further testing is required for assessment of those with vestibular pathology.

**Define Professional Practice Gap & Educational Need:** 1. Lack of a practical test battery in clinical assessment of vestibular dysfunction

**Learning Objective:** 1. Recognize the utility of an app-based subjective visual vertical test used in the clinical assessment of vestibular dysfunction

**Desired Result:** 1. Utilize the app-based subjective visual vertical test clinically

# Middle Ear Cancer in the American College of Surgeon's National Cancer Database

Jason A. Brant, MD; Michael J. Ruckenstein, MD

**Objective:** Evaluate patterns in carcinoma of the middle ear in a large national database.

**Study Design:** Retrospective review of a nationally collected cancer database.

**Setting:** The American College of Surgeon's National Cancer Database Patients: Patients with tumor location of the middle ear (C30.0)

**Interventions:** Surgery. Main Outcome Measures: Demographics and survival.

**Results:** From 2004 to 2013, 329 cases of primary cancer of the middle ear were included in the database. A majority of patients were male (51.7%) and white (82.7%). Most cases were squamous cell carcinoma or its variants (60.0%). The only other tumor types that represented over five percent of tumors were carcinoid (5.8%) and embryonal rhabdomyosarcoma (5.5%). Seventy-one percent of patients underwent surgery. Overall median survival was 71.3 months, and those who had surgery had a significant survival advantage over those that did not (85.4 vs 45.2 months, p = 0.03).

Conclusions: Carcinoma of the middle ear is a relatively rare condition and understanding of the impact it has on patient's as well as the treatments those patients are receiving can be advanced by evaluation of large national databases. The National Cancer Database includes important variables that can add to the understanding of this disease and help to impact treatment decisions for individual patients. This evaluation found that a majority of patients with cancer of the middle ear underwent surgery and this was correlated with a significant increase in survival.

**Define Professional Practice Gap & Educational Need:** Lack of contemporary knowledge of the full extent of carcinoma of the middle ear across geographic location and practice types.

**Learning Objective:** To define the extent of disease and evaluate treatment and survival patterns in a large national database.

**Desired Result:** Better understanding of the treatment patterns of carcinoma of the middle ear and their impact on patient outcomes.

### Radiosurgery of Glomus Tumors of Temporal Bone: A Meta-Analysis

Omid Moshtaghi, BS; Ronald Sahyouni, BA Hossein Mahboubi, MD, MPH; Yaser Ghavami, MD Harrison W. Lin, MD; Hamid R. Djalilian, MD

**Objectives:** (1) Perform a meta-analysis of the available data on the outcomes of stereotactic radiosurgery (SRS) for treatment of temporal bone glomus tumors (GT), and (2) evaluate the collective outcomes of SRS treatment with respect to tumor control.

**Data Sources:** A thorough literature search of published English-language literature from 2011-2016 was performed in PubMed, Ovid, and Cochrane databases using the keywords ("gamma knife" or "CyberKnife" or "linear accelerator" or "radiosurgery") and ("glomus jugulare" or "jugular paraganglioma.")

**Study Selection:** Studies reporting outcomes of SRS for temporal bone GT were included.

**Data Extraction:** Of 24 articles found, 9 studies met our inclusion and exclusion criteria and were used for qualitative and quantitative analyses containing 395 patients.

**Data Synthesis:** Average margin dose, modality, isodose line, volume decrease, follow-up duration, and tumor control rate data were extracted and analyzed.

Conclusions: The mean follow-up duration ranged from 37-148 months. Margin dose varied from 13.6-50.4 Gy. The collective mean tumor control rate was 95.7% (95% CI: 93.4%-98.0%). Clinical data on outcomes of SRS for treatment of GTs are sparse and primarily limited to single institutional analyses, with considerable variation in tumor volume and follow-up time. This meta-analysis provides an in-depth analysis of available data in the literature and reviews reported outcomes. Combined data with a meta-analysis performed in 2011 will be presented.

**Define Professional Practice Gap & Educational Need:** Lack of contemporary data on radiosurgery of glomus tumors in the temporal bone, and studies are primarily single institutional experiences with different tumor control rates.

**Learning Objective:** The reader may develop a better understanding of tumor control rates and and outcomes following radiosurgery of glomus tumors in the temporal bone.

**Desired Result:** Radiosurgical approaches can be considered in patients presenting with glomus tumors of the temporal bone with a better understanding of tumor control rates.

### Vestibular Schwannoma Resection via Keyhole Retrosigmoid Craniotomy Approach

Spencer Falcon, BS; Andrew K. Conner, MD Joshua D. Burks, BA; Anthony M. Alleman, MD, MPH Michael E. Sughrue, MD; Betty S. Tsai Do, MD

**Objectives:** The purpose of this study is to describe the surgical technique and outcomes of the keyhole retrosigmoid craniotomy for the resection of small to large vestibular schwannomas.

**Study Design:** Retrospective chart review.

**Setting:** Single-institution tertiary referral center.

**Patients:** 15 patients underwent a keyhole retrosigmoid craniotomy between for a primary diagnosis of vestibular schwannoma between 2012 and 2015.

**Intervention:** Keyhole retrosigmoid craniotomy with resection of tumor.

Main Outcome Measures: Outcomes studied include residual tumor volume, facial function as classified by the House-Brackmann (HB) scale, complication rates, and length of hospitalization. Results: Average preoperative tumor volume was 12.9 cm3 (range 1.2 cm3 - 33.8 cm3) with median extent of resection of 79% (range 37%-89%). Preoperative facial dysfunction (House-Brackman II) was present in 2 patients. Postoperative facial function at 1 month was HB I in 80% of patients, HB II in 13% of patients, and HB V in 6% of patients; at 1 year one patient had HB III with remainder all HB I. Postoperatively, disequilibrium was documented in 20, CSF leak was recorded in 13%, while unilateral dysesthesia, dysgeusia, and diplopia were recorded in 6%. Symptomatic dysesthesia and disequilibrium resolved within first year; however dysgeusia, and diplopia persisted. No infectious or hemorrhagic complications were recorded. The average length of hospitalization was 5.8 days (median 4 days, range 1-20 days).

**Conclusions:** Keyhole retrosigmoid craniotomy is a safe and feasible option for the resection of vestibular schwannomas with excellent long term facial function preservation and low rate of complications.

**Define Professional Practice Gap & Educational Need:** 1. Lack of awareness of the use of keyhole craniotomy for resection of acoustic neuroma. The keyhole craniotomy approach is routinely used for many neurosurgical procedures, but little has been described regarding the safety and efficacy in skull base surgery.

**Learning Objective:** 1. Describe the keyhole retrosigmoid craniotomy approach. 2. Evaluate the outcomes in consecutive patients who have undergone the keyhole retrosigmoid craniotomy approach for resection of an acoustic neuroma. 3. Compare the keyhole approach to the standard retrosigmoid craniotomy.

**Desired Result:** Attendees may change their practice in the retrosigmoid approach for resection of acoustic neuromas. This may result in better outcomes as the keyhole craniotomy is a smaller craniotomy and does not require the use of retractors that are commonly used in the standard approach. These retractors increase likelihood of intracranial injury and smaller craniotomies have been shown to result in fewer headaches.

### Ultra-high Resolution MRI Aids in Treatment of Audiovestibular Symptoms in Patients with Acoustic Neuroma and Endolymphatic Hydrops

Roxana Moayer, MD; Gail Ishiyama, MD Ali Sepahdari, MD; Akira Ishiyama, MD

**Objective:** To use high-resolution MRI to detect co-morbidity of endolymphatic hydrops (EH) and acoustic neuroma (AN) in patients presenting with audiovestibular complaints and correlate the MRI findings with clinical symptoms and audiovestibular testing.

**Setting:** Academic tertiary care referral center

**Patients:** Nine patients imaged with high resolution MRI had AN. Materials and Methods: Magnetic resonance EH imaging sequences on 3-Tesla included "cisternographic" 3D T2, and delayed intravenous enhanced three-dimensional fluid-attenuation-inversion-recovery (DIVE 3D FLAIR).

**Results:** Of the nine patients, three (33.3%) had coexisting AN and EH, on side ipsilateral to AN. All of the patients with comorbidity of AN and EH had a presentation indistinguishable from Meniere's disease: recurrent spells of vertigo associated with ipsilateral aural fullness, tinnitus, and hearing loss. One patient with both AN and EH had resection of AN at outside hospital, but continued to suffer imbalance likely secondary to central vestibular complications. The other two opted for surveillance of AN and are well-managed with low-salt diet and diuretics. The six patients with AN but not EH had varied presentations: one patient meets criteria for MD, and three had hours-long vertigo spells, but unaccompanied by otological symptoms.

**Conclusion:** Patients with AN presenting with episodic vertigo may have coexisting EH. We recommend that patients with AN and recurrent vertigo undergo high-resolution MRI with EH protocol, and if EH is present, then medical management should be considered prior to making surgical decisions.

**Define Professional Practice Gap & Educational Need:** The purpose of this study is to highlight a lack of awareness in detection which has only become possible with recent technology advances. New ultra-high resolution MRI can now detect patients with endolymphatic hydrops. Currently, standard protocol MRI is used to detect and perform surveillance for acoustic neuroma. We would like to share that ultra-high resolution MRI can be used to detect co-morbid endolymphatic hydrops in patients with acoustic neuroma who present with audiovestibular systems that are indistinguishable from Meniere's disease.

**Learning Objective:** The primary learning to objective is to recognize that patients with audiovestibular systems, which are indistinguishable from Meniere's disease, may in fact have two co-existing disease processes e.g. acoustic neuroma and endolymphatic hydrops. The second learning objective is to understand that ultra-high resolution MRI findings are critical in the work-up and treatment of patients with episodic vertigo.

**Desired Result:** It is our goal to propose that patients with acoustic neuroma who present with episodic vertigo should first undergo ultra-high resolution MRI to rule out endolymphatic hydrops prior to proceeding with surgery, as patients with co-morbid endolymphatic hydrops may be successfully treated medically.

### Quantifying the Hair Cell and Neural Sources of ECochG Signals Recorded in Patients with ANSD Receiving a Cochlear Implant

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**Hypothesis:** To characterize the contribution of the auditory nerve neurophonic (ANN) to electrocochleography (ECochG) of pediatric CI recipients with and without auditory nerve spectrum disorder (ANSD).

**Background:** ECochG is an emerging technique for predicting outcomes in cochlear implant (CI) recipients. Its utility may be increased using a computational model to independently quantify cochlear microphonic (CM), produced by hair cells, and the ANN, which are mixed in the ongoing portion of the response to low frequency tones.

**Methods:** Study conducted with UNC prior IRB approval . ANSD was diagnosed by a present CM and abnormal or absent Wave V on auditory brainstem response testing. The CM was modeled using a sinusoid modified by rectification and saturation. The ANN was modeled using a convolution of the unit potential and cycle. CM and ANN waveforms were extracted from ECochG signals recorded from the round window of pediatric CI recipients using 250-1000 Hz tones (90 dB nHL).

**Results:** The model matched 112 raw signals from ANSD and 314 from non-ANSD patients with mean R2>0.95. Signals of ANSD patients (n=112) had larger overall magnitudes (mean±std 3.04±4.32 uV) than non-ANSD patients (n=314, 0.73±1.23uV). The model identified similar ranges of ANN magnitudes in both groups (0-5.75 uV for ANSD compared to 0-3.54 uV).

**Conclusions:** The neural contribution to the typically large ECochG signals of ANSD patients is highly variable, often larger than is typical for non-ANSD subjects. The presence of a neural contribution would be expected to improve their clinical outcomes with the CI.

**Define Professional Practice Gap & Educational Need:** 1. Lack of awareness of the role of ECochG in predicting speech perception outcomes of cochlear implant recipients with auditory neuropathy spectrum disorder (ANSD). 2. Lack of knowledge of the newly developed signal analysis methods used with ECochG to separate neural and hair cell potentials.

**Learning Objective:** 1. Characterize the patterns of neural contribution to ECochG signal in patients with ANSD. 2. Demonstrate the advances in ECochG signal analysis methods which improve our ability to quantify the functional status of the neural components of the peripheral auditory system.

**Desired Result:** Improved understanding and confidence in the information conveyed by an ECochG recording as it relates to counselling the patient or their family regarding the potential range of outcomes for a patient with ANSD receiving cochlear implant.

# The Pattern of Hearing Outcome Following Surgeries of the Semicircular Canals

Amit Wolfovitz, MD; Thomas A. Babcock, MD Simon I. Angeli, MD

**Objective:** to analyse demographic, clinical and surgical factors that predict hearing outcome following surgeries of the semicircular canals (SCC) for various vestibular indications

**Study design:** retrospective case review

**Setting:** Tertiary referral center

Patients: adults who underwent surgeries for superior SCC dehiscence, Meniere's disease (MD), or BPPV

**Intervention:** therapeutic.

**Main outcome measure:** post surgical hearing outcome and its association with preoperative demographic, clinical features as well as surgical features.

**Results:** 11 cases underwent surgery for superior SCC dehiscence (5 middle fossa and 6 transmastoid approach), 4 cases for the lateral SCC (intractable MD) and 2 for the posterior SCC (BPPV). The mean age in the cases with postoperative similar (or better) bone conduction thresholds (n=15; 88.2%, 6 males) was 47.9±13.3 (21-66) while in cases with deteriorated thresholds (n=2, 1 male) was 50.5 ±2.1 (49-52). age, side, gender, indication for procedure, presenting symptoms and preoperative air and bone pure tone averages as well as surgical approach and technique were not found to be associated with worse hearing outcome. Moreover, in cases of deteriorated bone conduction threshold (a case of superior SCC dehiscence treated via middle fossa approach and another case of lateral SCC plugging for intractable MD), there was no unified audiometric pattern of hearing loss

**Conclusions:** hearing outcome was found to be favorable in 88.2% of SCC surgeries. non of the assessed demographic, clinical and surgical parameters was found to be associated with worse audiologic outcome.

**Define Professional Practice Gap & Educational Need:** 1. There is uncertainty regarding how many of the surgeries for the semicircular canals (SCC) for various vestibular indications, are ending with worse bone conduction threshold. 2. In cases of post surgical worse bone conduction threshold, it is unknown what preoperative demographic and clinical, as well as intra-operative approach and technique might played a key factor in the final outcome 3. additionally it is unknown whether there is a stereotypic pattern of hearing loss post surgeries of the semicircular canals

**Learning Objective:** 1. Assess the percentage of cases post surgeries for the SCC that are ending with worse bone conduction 2. Analyse demographic, clinical and surgical factors that predict postoperative hearing outcome 3. Understand the nature and pattern of hearing loss in cases of worse postoperative audiologic outcome

**Desired Result:** Clinicians aiming to perform surgery for the SCC for their patients will have additional tool in their armamentarium for prediction of post surgical outcome and for better counselling their patients about possible outcome and complications.

### Recurrent Vestibular Migraine Vertigo Attacks Associated with the Development of Profound Bilateral Vestibulopathy: A Case Series

Jacob L. Wester, MD; Akira Ishiyama, MD Gail Ishiyama, MD

**Background:** Idiopathic bilateral vestibulopathy is a debilitating condition characterized by gait ataxia, oscillopsia, and imbalance.

**Objective:** Case series of patients with profound bilateral vestibulopathy with migraine-linked vertigo spells.

**Patient 1:** 69 yo male with recurrent severe vertigo spells lasting up to 3 days in duration associated with prostrating migraine headaches starting at age 60. Hearing was normal. Misdiagnosed for 9 years as anxiety syndrome. At age 68, an ENG revealed absent caloric responses, and profound vestibulopathy.

**Patient 2:** 51 yo male presents with a history of brief "earthquake-like" vertigo, sharp head pains and phonophobia, occurring a handful of times in his lifetime over 7 years. At age 43, ENG was normal; however, ENG at age 48 revealed profound bilateral vestibulopathy. Subjectively, he noted improved balance with acetazolamide, although ENG was unchanged.

**Patient 3:** 49 yo female with a history of recurrent migraines with visual aura associated with vertigo lasting one hour. ENG at age 50 revealed profound bilateral vestibulopathy. Subjectively, she noted improved balance with acetazolamide, and ENG demonstrated mild improvement.

**Patient 4:** 43 yo male with a 5-year history of optical migraines and recurrent vertigo spells lasting 30 seconds presented with a 10 year history of oscillopsia. Misdiagnosed as BPPV. ENG at age 61 revealed profound bilateral vestibulopathy.

**Conclusion:** In these cases, migraine was linked with vertigo spells that eventually led to complete bilateral vestibular loss. Potential pathophysiological mechanisms are discussed.

**Define Professional Practice Gap & Educational Need:** Lack of contemporary knowledge of migraine-linked vertigo and its temporal relation to complete bilateral vestibular loss

**Learning Objective:** Relation of migraine-linked vertigo to complete bilateral vestibular loss

**Desired Result:** Attendees will have a better understanding of migraine-linked vertigo spells leading to complete bilateral vestibular loss and its underlying pathophysiology