

2012 Abstracts Selected for Presentation

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Meniere's Disease, Migraine, & Chronic Subjective Dizziness

Brian A. Neff, MD; Jeffrey P. Staab, MD; Scott D. Eggers, MD
Matthew L. Carlson, MD; William R. Schmitt, MD; Neil T. Shepard, PhD

Objective: To evaluate similarities and differences in clinical presentations of Ménière's Disease (MD), vestibular migraine (VM), and Ménière's Disease plus vestibular migraine (MDVM), including chronic subjective dizziness (CSD).

Study Design: Retrospective review with consensus conference of investigators to confirm diagnoses

Setting: Ambulatory, tertiary referral, dizziness clinic

Patients: 147 consecutive patients with consensus diagnoses of MD, VM, or MDVM, with/without comorbid CSD. AAO-HNS MD criteria, Neuhauser VM criteria, and newly validated CSD criteria were used for diagnoses.

Interventions: Diagnostic consultation

Main outcome measures: Similarities and differences between diagnostic groups in symptoms and results of neurotology, audiometric, and vestibular laboratory assessments.

Results: 76 patients had MD, 71 had VM. Considerable comorbidity existed between these groups as 21/76 MD patients (28%) had MDVM. Criteria thought to differentiate VM from MD overlapped greatly. 27/71 VM patients (38%) had ear complaints (subjective hearing loss, aural pressure, tinnitus) during periods of vestibular symptoms and headache; most (23/27, 85%) were unilateral. Conversely, 27/55 patients with MD alone (49%) had strong indicators of migraine including past or present migraine (without vertigo), migrainous symptoms (photophobia, headache with vomiting), or 1st degree relative with migraine. Including MDVM patients, 64% (49/76) of all MD patients had migrainous features. Most CSD patients (29/32, 91%) were in the VM group.

Conclusion: Large overlaps exist in symptoms of MD and VM. More specific diagnostic criteria are needed to differentiate them and address their coexistence. These conditions may share common pathophysiologic processes. CSD is not part of MD+VM comorbidity, but is for VM.

Define Professional Practice Gap & Educational Need: Address large variability in diagnostic approach and rationale

Learning Objective: To evaluate similarities and differences in clinical presentations of Ménière's Disease (MD), vestibular migraine (VM), and Ménière's Disease plus vestibular migraine (MDVM), including chronic subjective dizziness (CSD).

Desired Result: Provide better understanding of categorization of Meniere's disease, vestibular migraine, and chronic subjective dizziness

IRB or IACUC Approval: IRB: 09-004579

Cochlear Implant Round Window Versus Cochleostomy Insertion Techniques: Histological Findings

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Hypothesis: Cochleostomies and round window enlargements for cochlear implant electrode insertion result in more abnormal tissue formation in the basal cochlea and are more apt to produce endolymphatic hydrops than round window electrode insertion.

Methods: Twelve temporal bones from patients with a cochlear implant were examined under light microscopy and reconstructed with AMIRA 4.1 (3D reconstruction software) to determine cochlear damage and volume of neo-ossification and fibrosis following electrode insertion. For 3D reconstruction, the cochlea was divided into four segments; segment one was the proximal descending basal turn. The probable role of the electrode was evaluated in each case of hydrops. Amount of new tissue was compared between three groups of bones: insertion through the round window (RW), insertion after enlarging the round window (RWE) and cochleostomy (Cochl).

Results: The volume of fibrous tissue and new bone varied among temporal bones, but the RWE group had more total new tissue (new bone and fibrous tissue) in the two first segments than the RW group and greater initial damage ($p < 0.05$) in the first segment than RW insertions. Hydrops was seen in 5 cases, all belonging to the cochleostomy and RWE groups. Blockage of the duct was due to new tissue formation in 4 of the 5 hydrops cases.

Conclusion: Inserting the cochlear implant electrode through the round window created minimal initial intracochlear damage or new tissue formation as opposed to cochleostomies and round window enlargements. Minimizing extraneous tissue formation is an important consideration when hearing preservation surgery is the goal.

Keywords: Round window, cochleostomy, cochlear implant, fibrosis, new bone.

Define Professional Practice Gap & Educational Need: Amount of trauma induced by cochlear implant electrode insertion varies with type of surgical technique. Technique of insertion is surgeon dependent, is still debated, and is adapted to amount of residual hearing.

Learning Objective: Different techniques of insertion may produce different amounts of intracochlear trauma. Round window insertion is the least traumatic.

Desired Result: In order to preserve residual hearing, minimal trauma to the cochlea is the goal after electrode insertion.

IRB or IACUC Approval: IRB approval for this specific study was not required as approval had already been obtained generically for the temporal bone bank (IRB#06-030) and studies that might ensue from that data.

Cognitive Outcomes and Stress after Cochlear Implantation in Deaf Children with and without Developmental Delays

John S. Oghalai, MD; Susan E. Caudle, PhD; Jerry Lin, MD, PhD
Heather Bortfeld, PhD; Barbara Bentley, PsyD, MS Ed
Homer Abaya, BS

Objective: The benefits of cochlear implantation for children with developmental delays (DD) are often unclear. We compared cognition, adaptive behavior, familial stress, and communication in children with and without DD.

Study Design: Retrospective review

Setting: Two tertiary care pediatric hospitals

Patients: 204 children who underwent cochlear implantation assessed before and >1 year after implantation

Main Outcome Measures: The Mullen Scales of Early Learning (MSEL), Vineland Adaptive Behavior Scales (VABS), Parental Stress Index (PSI), and Preschool Language Scale (PLS).

Results: We developed a specific definition of DD for hearing-impaired children based upon DSM-IV criteria for mental retardation; 60 children met the criteria for DD and 144 children did not. Prior to implantation, multiple linear regression demonstrated that children with DD had lower scores in every domain of the MSEL and VABS tests ($p < 0.05$) but no differences in any domains of the PSI and PLS tests ($p > 0.1$) compared to children without DD. After implantation, children with DD demonstrated improvement in only the MSEL fine motor domain ($p < 0.05$). However, their VABS scores demonstrated a normal developmental trajectory and their PSI demonstrated elevated stress ($P < 0.05$). In contrast, children without DD demonstrated improvements in every MSEL domain ($p < 0.05$), age-appropriate improvements in the VABS, and their stress levels were not increased.

Conclusions: These data indicate that our definition of DD is a reliable method of stratifying deaf children. Improvements in development are noted in children with DD after cochlear implantation, but they are not as large as those found in children without DD.

Define Professional Practice Gap & Educational Need: Lack of awareness of the benefits of cochlear implantation of children with severe developmental delays.

Learning Objective: To report on our data demonstrating the benefits of cochlear implantation in children with developmental delays

Desired Result: The attendees can use this information when counseling parents of children considering cochlear implantation

IRB or IACUC Approval: Yes

Residual Hearing following Cochlear Implantation with a Long Electrode: Implications for Auditory Performance and Speech Perception

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Susan B. Waltzman, PhD

Objectives: To describe the effects of preserved residual hearing in traditional CI patients implanted with a long electrode. Detailed frequency- and hearing level-specific analysis of pre-operative auditory function, preserved residual hearing and post-operative speech performance will be presented.

Study Design: Retrospective review.

Methods: Patients over 18 years of age implanted by a single surgeon at a tertiary care facility between June 2005 and July 2010 with measurable pre-operative pure tone thresholds at any frequency were included. Conservation of hearing was defined as complete for postoperative thresholds within 10 dB of pre-implant values and partial if greater than 11dB. Speech perception was evaluated using monosyllabic word scores in quiet. Pure tone audiometry and speech perception testing were performed preoperatively and at regular intervals postoperatively with the 1-year evaluation being the final outcome period.

Results: Hearing of various degrees was preserved in 40% of patients. While preservation at any frequency or level was not a factor in speech perception outcome, preservation was more common in low frequencies and a correlation was found between level of preoperative hearing and postoperative preservation.

Conclusions: Hearing can be preserved in traditional CI patients implanted with a long electrode. While preservation of hearing may have implications for future technology, it is not currently correlated with performance with a cochlear implant.

Define Professional Practice Gap & Educational Need: 1) incomplete understanding of the ability to preserve residual hearing in traditional CI candidates implanted with a long electrode;
2) Lack of data regarding the effects of preserved residual hearing on postoperative speech performance.

Learning Objective: At the conclusion of this presentation, the participants should be able to (1) recognize the ability to preserve residual hearing in traditional CI candidates implanted with a long electrode; 2) describe the effects of preserved residual hearing on postoperative speech performance.

Desired Result: Attendees will be informed of the ability to preserve residual hearing in traditional cochlear implant candidates implanted with a long-electrode and have a greater understanding of the post-operative implications of preserved residual hearing.

IRB or IACUC Approval: Yes

ANS NEUROTOLOGY FELLOW AWARD WINNER

Understanding the Uses of Computed Tomography for Otosclerosis through Histopathologic Examination of Imaged Temporal Bones

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Objective: Until now, the use of computed tomography (CT) in the diagnosis and evaluation of otosclerosis has been based on correlation of radiologic findings to patient histories, intraoperative examinations, and audiologic data. The purpose of this study was to compare CT findings in otosclerosis to histopathology.

Study design: Prospective blinded

Setting: Radiology by tertiary referral hospital and otopathology laboratory

Patients: Temporal bones from patients with otosclerosis and other otologic diseases (used as controls)

Interventions: Blinded review of CT scans and comparison to histopathology of the same bones

Main outcome measures: The ability of CT to diagnose otosclerosis, identify otosclerotic foci in defined zones of the otic capsule, determine endosteal layer involvement, oval window (OW) obliteration and round window (RW) obliteration

Results: In a randomized blinded evaluation, radiologists identified 8/10 bones with otosclerosis and made 3 false positive diagnoses from 36 control bones. False positive and negative diagnoses are explained by direct comparison of CT images to the corresponding pathology slide. Subsequently, radiologists were asked to identify the location and extent of otosclerotic lesions in bones with known otosclerosis. Radiologists correctly identified otosclerosis in the area anterior to the oval window, the pericochlear area, and the round window niche in 17/17, 9/11, and 3/6 bones, respectively. CT correctly determined involvement of the endosteal layer, OW obliteration, and RW obliteration in 5/8, 2/2, and 2/2 temporal bones.

Conclusions: CT generally correlates well with histopathology in otosclerosis, but this study demonstrates some pathologic findings in otosclerosis that are not easily identified on CT.

Define Professional Practice Gap & Educational Need: Lack of contemporary knowledge: Currently, the accuracy of computed tomography (CT) in diagnosis of otosclerosis is based only on studies comparing CT findings to patient history and physical exam, intraoperative findings, pathology of footplate remnants, and audiometric results. There has been no prior comparison to the true gold standard: histopathology. The ability of CT to define the extent of individual otosclerotic lesions is also unknown, and has clinical implications for management of otosclerosis.

Learning Objective: The objective is to provide a better understanding of the uses and limits of CT in the management and diagnosis of otosclerosis based on comparison of CT findings to histopathology. Answers to the questions stated above will be provided based on review of temporal bones with otosclerosis and comparison to radiologists' evaluation by CT.

Desired Result: Attendees will be able to apply the knowledge learned to the management of their patients with otosclerosis. Understanding the ability of CT to diagnose otosclerosis, and what pathologic changes may lead to false positive or false negative CT results, will help the clinician properly utilize CT for this disease. Attendees may apply information about the ability of CT to diagnose endosteal layer involvement in their decisions about management of sensorineural hearing loss in otosclerosis (and application of any medical treatments).

IRB or IACUC Approval: Yes

Gender and Age Effects on Vestibular Schwannoma Characteristics at Presentation

Aisha Harun, BS; Yuri Agrawal, MD; Marietta Tan, MD
John K. Niparko, MD; Howard W. Francis, MD

Objective: To assess the influence of gender and age on vestibular schwannoma size and symptoms at presentation.

Study Design: Case series with chart review.

Setting: Academic tertiary referral center.

Patients: Subjects diagnosed with unilateral vestibular schwannoma evaluated or treated in either the department of Otolaryngology-Head and Neck Surgery or Neurosurgery between 1997 and 2010.

Intervention: Demographic information, tumor characteristics, presenting symptoms, and treatment management strategy were recorded for each subject.

Main Outcome Measure: Tumor size, presence of hearing loss or dizziness at presentation.

Results: 1269 patients were included in this study. Overall males had significantly larger tumors than females (18.23mm vs. 16.81mm, $p=0.031$); this difference was particularly pronounced in subjects 47 years and younger. The presence of clinical symptoms at baseline also differed by gender: the prevalence of hearing loss was 95.1% in males vs. 90.3% in females ($p=0.001$) and the frequency of dizziness was 74.3% in females vs. 59.0% in males ($p<0.0001$). In multivariate analyses, male gender continued to have a borderline significant positive association with tumor size ($p=0.066$) and males were two-fold more likely to have hearing loss (OR 2.082, 95% CI 1.300-3.336), but half as likely to have dizziness (OR 0.501, 95% CI 0.387-0.649) at presentation.

Conclusion: We observed significant gender-differences in the presentation of unilateral vestibular schwannomas: males had significantly larger tumors at younger ages, and had more hearing loss and less dizziness relative to females at presentation. The implications of these differences for risk stratification, prognosis and treatment merit further study.

Define Professional Practice Gap & Educational Need: Gender differences have been reported in the presentation of sporadic vestibular schwannomas, particularly with regard to tumor size. Age has also been shown to be an independent predictor of tumor size. The predictive value of both age and gender has not been rigorously evaluated yet and may provide useful insights regarding pathophysiology and the patterns of disease progression.

Learning Objective: The learning objective of this presentation will be to describe the influence of gender and age on vestibular schwannoma characteristics at presentation, including tumor size and presenting symptoms.

Desired Result: The desired result will be that clinicians will have increased awareness of patterns of disease presentation and progression for vestibular schwannoma, which may improve risk stratification for patients.

IRB or IACUC Approval: NA_00017143

Neoplastic Causes for Non-Acute Facial Paralysis

John P. Leonetti, MD; Sam J. Marzo, MD
Douglas A. Anderson, MD; Josh M. Sappington, MD

Objectives:

1. To assess the clinical presentation of patients with tumor-related, non-acute facial paralysis.
2. To review our series of 221 such patients.
3. To provide an algorithm for the evaluation and management of these cases.

Study Design: A retrospective chart review of patients with tumor-related non-acute facial paralysis treated at our tertiary care, academic medical center.

Methods: Patients were included with neoplastic related facial paralysis treated at our institution between July of 1988 and July of 2011. The clinical signs and symptoms along with the radiographic findings were assessed.

Results: Gradual-onset facial weakness or facial twitching occurred in all 221 patients. The tumor location according to MRI and/or CT scanning was extratemporal in 128 patients (58%), intratemporal in 55 patients (25%), and intradural in 38 individuals (17%). The majority of the extratemporal tumors were malignant (94%), while (91%) of the remaining patients had benign lesions. Associated clinical manifestations such as hearing loss, tinnitus, other cranial nerve deficits, and the presence of a mass helped identify the tumor site. A transtemporal approach was utilized for the intratemporal and intradural tumor resections while the extratemporal lesions required parotidectomy with partial temporal bone dissection. Facial nerve grafting was performed in 110 of 221 patients (50%).

Conclusion: Gradual onset facial paralysis or twitching may be due to neoplastic invasion of the facial nerve in its course from the cerebellopontine angle to the parotid gland. Beware of the diagnosis of “atypical Bell’s Palsy”.

Define Professional Practice Gap & Educational Need: Delay in diagnosis of facial paralysis due to a tumor.

Learning Objective: This presentation will provide a clinicoradiographic summary of neoplastic causes for non-acute facial paralysis

Desired Result: To provide an algorithm for the evaluation and management of these cases.

IRB or IACUC Approval: Yes, IRB approval # 203938

Hearing Preservation in Management of Epidermoids of the Cerebellopontine Angle

Mia E. Miller MD; Bill Mastrodimos, MD; Roberto A. Cueva, MD

Objectives: Previous reports have advocated the translabyrinthine approach to epidermoid cysts of the cerebellopontine angle (CPA) to achieve complete surgical excision in a single operation. The neurotologic management of epidermoids of the CPA is discussed with an emphasis on hearing preservation.

Study Design: Retrospective chart review.

Methods: A search of archived surgical cases at a single institution between January 1, 1997 and December 31, 2011 revealed 18 cases of epidermoid cysts involving the CPA.

Results: 18 patients with a mean age 40.9 years underwent surgery. Average tumor size was 4.47 cm, and presenting symptoms included headache, vertigo, cranial neuropathies and seizures. Thirteen patients underwent a retrosigmoid approach, 2 translabyrinthine, 2 pterional and 1 retrolabyrinthine/presigmoid. Complications included 1 CSF leak, 1 pseudomeningocele, cranial nerve dysfunction, and persistent imbalance. All but 4 patients had follow-up imaging to chronicle tumor residua/recurrence, varying from 2 to 121 months postoperatively. Two patients had undergone previous surgery for epidermoid excision elsewhere, and 2 patients required reoperation for epidermoid regrowth. The times between surgeries were 3, 7, 18 and 31 years. Of the patients who underwent a retrosigmoid approach, 9 had postoperative audiograms. All of these patients maintained hearing at or near their preoperative level except for 2 patients whose hearing declined and one whose hearing significantly improved.

Conclusions: Hearing preservation approaches for epidermoids of the CPA is a feasible option for long-term control of these tumors. Resection from a retrosigmoid approach can provide years of useful hearing, and the majority of patients do not require reoperation.

Define Professional Practice Gap & Educational Need: 1) Inconsistencies in how epidermoid cysts of the cerebellopontine angle are managed.

Learning Objective: To explain how hearing preservation approaches can be used to effectively manage epidermoid cysts of the cerebellopontine angle.

Desired Result: Attendees will consider retrosigmoid and other hearing preservation approaches in patients with useful hearing over the translabyrinthine approach to epidermoid cysts of the cerebellopontine angle.

IRB or IACUC Approval: n/a

**Direct Round Window stimulation with the
Vibrant Sound Bridge (Med-El):
5-years Experience Using Technique without Fascia**

Henryk Skarzynski, MD, PhD; Lukasz Olszewski MSc
Lorens Artur Eng, PhD; Piotr H. Skarzynski MD, MSc
Marek Porowski, MD

Objective: Different types of congenital malformations of middle ear and external auditory canal are treated with good outcome by Med-El Vibrant Soundbridge system. In the congenital aural atresia with mixed or conductive hearing impairment the classical approach of FMT fixed on the long incus process is used. In special cases where this solution is not appropriate, FMT placement directly on the round window (without fascia) can be used; in our 5-years experience it provides good stimulation and results comparable with other approaches, which we present in this study.

Study Design: Prospective study.

Setting: Tertiary referral center

Patients: 26 patients (14 women, 12 men), ages 11 – 62 (mean 46,8 y.o.), with mixed or conductive, bilateral or unilateral hearing loss.

Intervention: Direct round window implantation with Med-El VSB in selected ear.

Main outcome measures: Pure tone audiogram, free-field speech test and functional gain measurement.

Results: In the 5-year follow up of the study group we observed stable hearing in the implanted ear and significant improvements of free field pure-tone audiometry and speech understanding during the observation period (12 to 62 months). No complications or device extrusion was observed except for three reoperations due to FMT reposition.

Conclusions: Direct round window stimulation is an alternative for patients with hearing impairment caused by chronic otitis media and/or lack of the ossicles, especially after radical modified operations. It allows achieving good results in selected group of patients, although further observation on a larger population is needed to confirm long-term validity and effectiveness of this method.

Define Professional Practice Gap & Educational Need: There is a group of patients with hearing loss and congenital middle ear and/or ear canal malformations for whom the classical application of the Med-El VSB is not possible due to the chronic otitis media and/or lack of the ossicles (e.g. after radical modified operations). For these patients an alternative treatment solution is needed.

Learning Objective: To determine long-term safety, validity and effectiveness of the direct round window stimulation with the FMT.

Desired Result: Safe and effective treatment option for patients for whom the classical VSB (Med-El) application is not applicable.

IRB or IACUC Approval: n/a

**Application of functional Magnetic Resonance Imaging (fMRI)
for Assessment of the Primary Auditory Cortex
Function in Partial Deafened Patients**

Piotr H. Skarzynski, MD; Tomasz Wolak, PhD; Henryk Skarzynski MD, PhD
Artur Lorens Eng, PhD; Lech Sliwa Eng, PhD; Mateusz Rusiniak, MSc

Objective: Assessment of Primary Auditory Cortex (PAC) activations in response to different frequency sounds in hearing impaired children before implantation.

Study design: 3T MAGNETOM TRIO scanner fMRI study using SPARSE paradigm,

Stimuli: tones of two different modulated frequencies: 500Hz (FM 250Hz) and 4000Hz (FM 1000Hz) presented at 90dB SPL via MRI-compatible earphones. Scanner generates noise of 99 dB SPL during 3-second acquisition of whole brain image. T1-weighted structural images acquired with isotropic voxels 0.9x0.9x0.9mm. Two functional 'runs', repetition time TR=18000ms, TE=30ms. Single volume acquired during each TR period, about 3000ms for 48 slices from the start of TR period; until the end of TR period no gradient-generated noise.

Functional data obtained with gradient-echo EPI sequence; 50 volumes per series (each volume - 48 slices, signal acquired with physical matrix 64x64). Functional images' voxel size: 3x3x3mm.

Whole brain morphological images with standard T2-weighted sequences to exclude neurological pathology.

Setting: Tertiary referral center

Patients: 14 right-handed partially deafened children (8-16 y.o.).

Interventions: Diagnostic

Main outcome measure(s): fMRI can serve as additional sub-group classification method for patients with partial deafness.

Results: Most participants displayed activations in PAC corresponding to their hearing loss. Despite better preservation of low frequency hearing in study group, differences from normally hearing subjects were observed. MRI data shows tonotopic structure of PAC and reflects patients' audiogram-based classification (PDT-EC, PDT-EAS, PDT-ES).

Conclusion: fMRI may be a useful additional technique for auditory system function assessment. Based on the results we conclude that our paradigm provides stable and reproducible auditory cortex responses.

Define Professional Practice Gap & Educational Need: Functioning of the auditory cortex in patients qualified for implantation is essential for rehabilitation in the postoperative period. The study was aimed to assess Primary Auditory Cortex (PAC) response to sounds of different frequencies in group of hearing impaired children prior to implantation.

Learning Objective: To develop methods for testing the function of auditory cortex in patients with the partial deafness.

Desired Result: A useful additional method of auditory system function assessment leading to more efficient rehabilitation.

IRB or IACUC Approval: yes

Simultaneous oVEMP and cVEMP Responses Elicited by Novel Head Tap and Recording Device

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Stuart K. Gardiner, PhD; Steven Johnson, PhD; Jon D. Birck, PhD

Objective: To develop a reliable, easy to use bedside, office, or field system that allows the simultaneous measurement of ocular and cervical vestibular evoked myogenic potentials (oVEMP and cVEMP) using a non-auditory stimulus.

Study design: Prospective bioengineering design of the test system with utricular and saccular otolith measurement studies in human subjects.

Setting: Private practice, tertiary neurotology referral center.

Patients: Ten normal adults without history of vestibular dysfunction, five subjects with radiographically confirmed superior canal dehiscence (SCD), and one subject with posterior canal dehiscence (PCD) were studied.

Interventions: The subjects underwent traditional auditory stimuli-based cVEMP studies with a commercially available system and then testing with the oVEMP and cVEMP system.

Main outcome measures: Duration of each diagnostic study, patient comfort, concurrence of auditory and non-auditory induced cVEMPs, and the reproducibility of the oVEMP responses recorded.

Results: For the normal subjects, the mean age was 46.8, with a range of 30 to 61 years; 4 females and 6 males. For the SCD subjects, the mean age was 50.2, with a range of 46 to 54 years; 4 females and 1 male. The PCD subject was a 35 year-old man. While the cVEMP responses were similar using either the auditory or non-auditory stimuli, the subjects preferred the latter. The oVEMP data showed an age-dependent reduction in function.

Conclusions: The combined oVEMP and cVEMP system designed was rapid, more comfortable for the subjects and also allowed recording of both utricular and saccular responses, which cannot be recorded using contemporary auditory stimulus cVEMP protocols.

Define Professional Practice Gap & Educational Need: Clinical use of a vestibular test device that measures utricular function is currently not available.

Learning Objective: Reproducible non-auditory stimulation can be used to generate and record oVEMP and cVEMP responses.

Desired Result: Attendees will understand that non-auditory stimuli (head taps) can be used clinically to study saccular and utricular responses.

IRB or IACUC Approval: LRI-1079

Cerebrospinal Fluid Pressure Abnormalities as a Cause of Chronic Subjective Vertigo with Migraine or Head Pressure

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Linda Gray, MD; Peter Kranz, MD

Objective: The normal range of CSF pressure is poorly defined. Abnormal pressure for some may be normal for others. The objective is to determine if altering the CSF pressure in patients within “normal” range can relieve symptoms of chronic subjective vertigo with migraine headache or head pressure, with or without other otologic complaints

Study Design: Prospective, IRB approved study

Setting: Tertiary center

Patients: Patients with chronic subjective vertigo, migraine or head pressure, with or without other otologic complaints and clouded mentation

Intervention: Diagnostic lumbar puncture with opening pressure measurement and addition or removal of CSF

Outcome measure: Self-reported symptom score immediately after lumbar puncture

Results: 46 patients met entrance criteria. Average opening pressure was 270 mmH₂O ranging from 110 to 265mmH₂O. 27 patients had immediate symptom improvement with removal of CSF - opening pressure for this group averaged 217mmH₂O. Eight had complete resolution of symptoms, 14 had significant improvement and 5 had mild improvement. Eleven patients had improvement with addition of CSF, 3 with complete resolution of symptoms, 4 with significant improvement and 3 with mild improvement with CSF pressure averaging 157mmH₂O. The opening pressure was significantly higher in group that improved with CSF removal. Eight patients had no improvement with addition or removal of CSF. 7 reported no change, one reported significant worsening.

Conclusion: Chronic subjective vertigo with migraine or head pressure, clouded mentation with or without other otologic complaints is often caused by abnormal CSF pressure. Removing or adding CSF in many of these patients provided immediate relief of their symptoms. Lumbar puncture should be considered in these patients.

Define Professional Practice Gap & Educational Need: Lack of contemporary knowledge of treatment for chronic subjective vertigo and migraine headache leads to many patients receiving suboptimal care

Learning Objective: To learn about potential causes of chronic subjective vertigo with migraine and how to treat it

Desired Result: Attendees will be able to apply the knowledge they gain in the treatment of patients with chronic subjective vertigo

IRB or IACUC Approval: yes

Ocular vs. Cervical VEMPs in the Diagnosis of Superior Semicircular Canal Dehiscence Syndrome

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John P. Carey, MD

Objective: To determine whether cervical or ocular VEMPs are better for the diagnosis of superior semicircular canal dehiscence syndrome (SCDS)

Study design: Prospective comparison of tests

Setting: Tertiary referral center

Participants: 34 SCDS patients (mean age 47y; range 20-66y) and 25 age-matched controls (mean age 47y; range 27-66y).

Intervention(s): CVEMPs evoked by clicks; oVEMPs evoked by 500 Hz tone-bursts. All patients underwent surgery for repair of SCDS.

Main outcome measure(s): CVEMP thresholds; oVEMP peak-to-peak amplitudes.

Results: 30 SCDS patients had obvious dehiscences while 4 had thin bone over the superior semicircular canal. In the 34 affected ears, CVEMP thresholds were low (≤ 75 dB nHL) in 17 (50%), normal in 13 (38%) and absent in 4 (12%), while oVEMP amplitudes were high (≥ 24 microV) in 29 (85%), borderline (19-22 microV) in 3 (9%) and absent in 2 (6%) SCDS ears. Of the 2 SCDS ears with absent oVEMPs, one had absent cVEMPs and the other had a normal (95 dB nHL) threshold.

Conclusions: oVEMP amplitudes are superior to cVEMP thresholds in the diagnosis of SCDS. For patient comfort and diagnostic yield, it is recommended to first perform oVEMP testing and subsequently follow-up with cVEMP testing if results are negative but other findings suggest SCDS.

Define Professional Practice Gap & Educational Need: (1) Lack of implementation of oVEMP testing for evaluation of superior semicircular canal dehiscence syndrome (SCDS). (2) Lack of awareness on the advantages of performing oVEMP testing vs. cVEMP testing. (3) Lack of awareness of the weaknesses of cVEMP testing.

Learning Objective: The clinician will learn about the advantages of oVEMP vs cVEMP testing in the evaluation of patients with SCDS.

Desired Result: The clinician will identify which VEMP test is more accurate in the evaluation of patients with SCDS. The clinician will learn about the reliability of cVEMP testing for SCDS and understand that oVEMP may be a better option to evaluate these patients.

IRB or IACUC Approval: NA_00035749

Dural Involvement in Chronic Ear Surgery

Sanjay Prasad, MD

Objective: Does dural exposure occur during chronic ear surgery? Is it iatrogenic or disease-related? What are the clinical implications?

Study design: A retrospective case review of patients who underwent mastoidectomy surgery for chronic otomastoiditis with cholesteatoma or with granulation was performed. Iatrogenic dural exposure and disease-related dural exposure was reported. The clinical implications of untreated areas of dural exposure especially in regards to revision surgery and meningocele/ encephalocele formation are explored.

Setting: Tertiary Otology/ Neurotology Private Practice Referral Center

Patients: 1,817 mastoidectomy procedures were performed on 891 patients for chronic otomastoiditis with cholesteatoma/granulation or with granulation alone by the primary author from 1993 to 2011.

Results: 43.5 % (790/1817) of the procedures had iatrogenic dural exposure and 47.2 % (857/1817) of the procedures had evidence of dural exposure from cholesteatoma or granulation tissue. Granulation tissue and cholesteatoma was safely removed from the dura in all cases. No patient suffered brain injury. The areas of dural exposure were left untreated. Dural exposure occurred more frequently in revision mastoidectomy surgery than primary surgery. On revision surgery, 5 patients developed a meningoencephalocele related to dural exposure from a previous cholesteatoma.

Conclusions: Dural exposure can occur in chronic ear surgery from the disease process and also from the operative process. Granulation tissue and cholesteatoma can be safely removed from the dura. These small areas of dural exposure can be safely left untreated. In our series the risk of meningoencephalocele formation is very low.

Define Professional Practice Gap & Educational Need:

1. Lack of awareness that dura is often exposed in chronic ear surgery.
2. Is it safe to remove granulation/ cholesteatoma from the dura?
3. Is it safe to leave these small areas of exposed dura untreated?

Learning Objective:

1. Increase awareness that dural exposure occurs in chronic ear surgery.
2. Granulation/cholesteatoma can be safely removed from the dura.
3. Small areas of exposed dura do not require repair as the risk of meningocele/ encephalocele formation is very low.

Desired Result:

1. Increase awareness that dural exposure does occur in chronic ear surgery.
2. Caution is advised when performing revision mastoidectomy surgery because dural exposure is more common.
3. Improve documentation of iatrogenic or disease-related dural exposure.
4. It is important to notify the patient and the family that dural exposure has occurred in case of future treatment.

IRB or IACUC Approval: n/a

Evolving Treatment of Large Acoustic Neuromas

Ryan G. Porter, Sr., MD; Michael J. LaRouere, MD
Jack M. Kartush, MD; Dennis I. Bojrab, MD
Daniel R. Pieper, MD

Objective: To describe a successful paradigm for treatment of large acoustic neuromas.

Study design: Retrospective case review.

Setting: Tertiary referral center.

Patients: One hundred fifty-three patients who underwent surgical resection for large acoustic neuromas (greater than or equal to 3cm) between 2000 and 2009.

Intervention(s): Staged surgical resection or single stage surgery with or without adjuvant stereotactic radiosurgery.

Main outcome measure(s): Postoperative facial nerve outcomes are reported using the House-Brackmann (HB) facial nerve grading scale and compared with historical controls from a literature review. Rates of adverse outcomes are also reported.

Results: Seventy patients underwent staged surgical resection of their tumors while 78 patients underwent either single stage surgery or surgery with subsequent stereotactic radiosurgery. Eighty-one percent of patients in the staged surgical resection group had a postoperative HB grade I or II facial nerve function compared with 75% in the single stage surgical group. Overall, 79% of patients in the current study had HB grade I or II following treatment compared with a mean of 49% in the literature for similar sized tumors. Our methods including the decision to use staged surgery when necessary, dissection of tumor with simultaneous intraoperative monitoring and stimulus dissectors, and utilization of adjuvant stereotactic radiosurgery are described.

Conclusions: Using the described paradigm, large acoustic neuromas can be successfully treated with either staged or single stage surgical resection with or without adjuvant radiosurgery to obtain more favorable facial nerve outcomes than historically reported controls while minimizing morbidity for the patient.

Define Professional Practice Gap & Educational Need: Lack of contemporary knowledge of treatment outcomes and methods for large acoustic neuromas.

Learning Objective: To describe a successful paradigm for treatment of large acoustic neuromas.

Desired Result: The attendees should be able to use the presented information to formulate appropriate treatment plans for patients with large acoustic neuromas utilizing available treatment methods and technology.

IRB or IACUC Approval: Providence IRB Approval: 274062-1

Influence of an Intraoperative Cochleostomy Gusher on Cochlear Implant Performance in Children with Labyrinthine Malformations

Oliver F. Adunka, MD; Holly F.B. Teagle, AuD; Lisa Park, AuD
Jennifer Woodard, AuD; Deborah Hatch, AuD; Craig A. Buchman, MD

Objectives: To assess the effect of an intraoperative perilymph fluid gusher during cochlear implantation on speech perception abilities in pediatric patients with labyrinthine anomalies.

Setting: Tertiary care academic referral center.

Methods: 70 subjects with labyrinthine malformations who received a cochlear implant were identified in our pediatric cochlear implant database. In 30 cases, an intraoperative perilymph fluid gusher was encountered during surgery. Fifteen children with GJB2 positive hereditary hearing loss served as controls. A speech perception hierarchy was used to assess performance and displayed as a speech reception index in quiet (SRI-Q). This index was compared among groups and malformation types.

Results: All gushers were controlled intraoperatively without the need for lumbar drainage and there were no postoperative cerebrospinal fluid leaks. Despite considerable variation among individuals, the SRI-Q demonstrated overall good performance for children with incomplete partitioning/enlarged vestibular aqueduct (IP-EVA) type malformations and lesser outcomes for children with hypoplastic malformations when compared to controls. The presence or absence of a perilymph gusher did not significantly influence results after cochlear implantation although some children with gushers did experience fluctuations in performance.

Conclusions: This report documents the variable outcomes for cochlear implantation among children with inner ear malformations. Anatomic parameters such as the type of malformation and cochlear nerve status appear to influence performance with a cochlear implant more so than the presence or absence of a cochleostomy gusher. Patients that experience an intraoperative gusher should be made aware of the possibility of fluctuating auditory performance with the use of their implant.

Define Professional Practice Gap & Educational Need: 1) lack of knowledge of the functional consequences of an intraoperative perilymph gusher during cochlear implantation on speech perception performance.

Learning Objective: The audience will learn about cochlear implant speech perception results in children with inner ear malformations who did or did not have an intraoperative perilymph gusher during surgery.

Desired Result: The desired result is that the audience can apply the knowledge gained in this presentation for their clinical practice. The knowledge gained should help in the clinical management of this patient population.

IRB or IACUC Approval: Yes

Radiographic Evaluation of the Tegmen in Patients with Superior Semicircular Canal Dehiscence

Garani S. Nadaraja, MD; Richard K. Gurgel, MD; Nancy J. Fischbein, MD
Andrew Anglemyer, PhD; Ashkan Monfared, MD
Robert K. Jackler, MD; Nikolas H. Blevins, MD

Objective: To determine a radiographic association between superior semicircular canal dehiscence (SSCD) and tegmen dehiscence (TD).

Study Design: Retrospective Case-Control Series

Setting: Tertiary referral center

Patients: Patients seen between 2003 to 2010 with radiographic SSCD were compared to cochlear implant recipient controls

Intervention: The tegmen and superior semicircular canal were evaluated on CT temporal bone scans.

Main outcome measure: If detected, the widest point of the SSCD was measured. The tegmen was graded on a 5-point scale. Following analysis, a radiographic TD was defined as any single area of absent tegmen greater than 5mm, multiple areas of absent tegmen, or evidence of meningocele.

Results: 38 patients with SSCD and 41 cochlear implant controls were identified. 76% of patients (29/38) with unilateral or bilateral SSCD had a radiographic TD on at least one side compared to 22% (9/41) of controls. Patients with SSCD had a 10.6 times (p-value < 0.001) higher odds of having radiographic TD in either ear compared to controls. Among patients with any SSCD, for every millimeter increase in the width of dehiscence, the relative risk for any TD increased more than two fold (OR 2.5; p=0.019). Age, gender and a body mass index >30 did not confound the association.

Conclusions: There is a strong radiologic association between SSCD and TD, suggesting a similar etiologic process. The tegmen should be carefully evaluated in patients with SSCD. We have also proposed a new system for radiographically grading the integrity of the tegmen.

Define Professional Practice Gap & Educational Need: While there have been case reports of simultaneous tegmen dehiscence with superior semicircular canal dehiscence, there is a lack of literature objectively evaluating the correlation of these conditions.

Learning Objective: There is a strong radiographic correlation between patients with superior semicircular canal dehiscence and tegmen dehiscence. With every increase in millimeter dehiscence of the canal, the risk of tegmen dehiscence increases more than 2 fold.

Desired Result: In patients presenting to clinic with superior semicircular canal dehiscence, the tegmen should be evaluated on imaging. In symptomatic superior semicircular canal patients in which surgical correction is considered, the surgeon may also need to address concomitant tegmen deficiencies. Perhaps a middle fossa craniotomy approach is more suitable than a transmastoid approach.

IRB or IACUC Approval: This study has been approved by the Stanford University IRB (protocol #16879.)

Test–retest Reliability and Construct Validity of MusicEAR Administered to Cochlear Implant Users

Hosam A. Amodi, MD; David Clinkard; Lee Bartel
Lendra M. Friesen; Vincent Y. Lin; Joseph M. Chen

Objective: The main goal of this study was to determine the test–retest reliability of the MusicEAR software from a group of cochlear implant users, over two test sessions. Additionally, we examined the relationship between the MusicEAR test results and selective, alternating and divided attention using the Test of Everyday Attention (TEA).

Study Design: Prospective study.

Setting: Tertiary referral center.

Patients: Forty pre/ peri-lingual and post-lingual deafened cochlear implant users(18-65 years) were tested.

Interventions: Subjects participated in a test and retest session (within 8 weeks) using the MusicEAR software.. The TEA was administered during the first session.

Main Outcome Measure: Test-retest reliability for all the MusicEAR modules, (music enjoyment tool, aural acuity test and perceptual ability) was assessed using two tailed paired t-tests and Pearson correlation coefficients. Correlations were also performed in order to examine the relationship between performance on different MusicEAR tasks and the TEA subtests.

Results: Mean changes in performance on retest on the MusicEAR tasks were not statistically significant compared to the first test ($p < 0.01$). In addition, the responses obtained from the retest correlated significantly to first test responses. Performance on the 5 tasks of the TEA showed no correlation with performance on all MusicEAR tasks except the pitch acuity, which correlated significantly to visual selective attention and attentional switching level ($r = 0.416$ and 0.497 respectively, P value < 0.01).

Conclusion: Overall, MusicEAR software was found to be a reliable measurement tool, although the relationship to attention using the TEA was not established.

Define Professional Practice Gap & Educational Need: Presently, one of the most challenging areas in CI functionality is the lack of consistent performance in music listening. Most previous studies focused only on perceptual skills in the basic elements of music (pitch, timbre and rhythm). In MusicEAR, a more holistic approach is adopted by adding tasks with increasing levels of complexity, involving pattern and melody recognition.

Learning Objective:

This study has two objectives

1. Validate the test-retest reliability of the “Music EAR” software.
2. Correlate “attention” with perceptual performance.

Desired Result:

The desired result is to show that the MusicEAR software is a reliable instrument, and able to discriminate a wide range of music perceptual ability. Another anticipated outcome is that attention will be correlated with perceptual performance. These results will allow this software to be used as a general self-administrable tool for assessing music perception abilities in CI users. Also, these results will provide the basis for developing the MusicEAR retraining module, which can be used to investigate the potential benefits of musical rehabilitation on pitch and speech perception

IRB or IACUC Approval: Yes

Facial and Lower Cranial Neuropathies after Preoperative Embolization of Jugular Foramen Tumors with Ethylene Vinyl Alcohol

Brian C. Gartrell, MD; Marlan H. Hansen, MD; Bruce J. Gantz, MD
Michael B. Gluth, MD; Mustafa K. Baskaya, MD; Samuel P. Gubbels, MD

Objective: Superselective embolization (SSE) is a commonly employed method to reduce the vascularity of skull base tumors prior to surgical extirpation. Cranial nerve palsy is a potential complication associated with SSE. Onyx-18, an ethylene vinyl alcohol (EvOH) copolymer FDA-approved for embolization of dural arteriovenous malformations, has become increasingly utilized in SSE of jugular foramen neoplasms. Although there have been four cases of facial weakness reported, none have been associated with the use of EvOH. In this article, we describe 4 cases of cranial nerve palsy after preoperative SSE with EvOH. The vascular supply of the facial nerve is characterized, and existing literature on SSE is reviewed. Finally, we propose a strategy that might aid in avoidance of this rare but significant complication.

Study design, Setting, Intervention, Patients: Retrospective case review of four individuals at three tertiary care hospitals who experienced cranial nerve palsies after preoperative SSE of a jugular foramen neoplasm using EvOH.

Main outcome measure: Cranial nerve deficits.

Results: Three patients developed facial weakness immediately after SSE, all of whom were complete. One of these patients also had co-incident involvement of multiple other lower cranial nerves. One patient with facial paralysis documented prior to embolization developed cranial nerve X and XI palsies within 24 hours of embolization.

Conclusions: Cranial nerve paralysis, most commonly involving the facial nerve, can occur after SSE with EvOH. Further studies are needed to determine whether the use of EvOH may confer an increased risk of cranial nerve paralysis after SSE for jugular foramen tumors.

Define Professional Practice Gap & Educational Need: There currently exists a lack of awareness of the incidence, risk factors, and prevention strategies for cranial neuropathies after superselective embolization of jugular foramen masses. These factors are important not only for identifying patients who may be at increased risk of such a complication but also in developing methods to protect patients from this occurrence. Furthermore, given the relatively recent introduction of ethylene vinyl alcohol for use in superselective embolization procedures, there is a lack of knowledge regarding the risk of cranial neuropathy associated with this copolymer.

Learning Objective: The learning objective of this talk is to describe four cases of cranial neuropathy after superselective embolization of jugular foramen masses, describe the vascular supply to the facial nerve including normal variants that place patients at risk of facial nerve paresis after embolization, to define the risk of cranial neuropathy associated with superselective embolization, and to propose strategies to assist the clinician in minimizing the risk of cranial neuropathy after preoperative superselective embolization of jugular foramen neoplasms.

Desired Result: The desired result is that based on a more complete understanding of the vascular supply to the cranial nerves, the incidence of cranial neuropathy after superselective embolization, the specific properties of various embolization agents, and strategies to minimize the risk of cranial neuropathy, clinicians who are involved in providing care to patients with jugular foramen neoplasms will apply this information when delivering care to improve patient outcomes.

IRB or IACUC Approval: Permission to proceed with the study was obtained through each participating institution's institutional review board.

Comparison of Speech Performance in Bimodal versus Bilateral Cochlear Implant Users

Ana H. Kim, MD; Derek R. Petti; Lauren Kohan, MS
Lorie Singer, MA; George Alexiades, MD; Ronald Hoffman, MD
Christopher Linstrom, MD; Simon Parisier, MD

Objective: Examine the advantage of bilateral cochlear implantation (CI) over bimodal listening.

Study Design: Retrospective chart review

Setting: Academic cochlear implant center

Patients: Bilateral CI recipients implanted from 1998 to 2010. Bimodal listening was compared to bilateral CI speech perception performance at 1 year post 2nd implant. Of 108 bilateral CI recipients, 19 pediatric patients (mean age of 1st implant = 5.28 ± 3.93 yrs) and 26 adult patients (mean age of first implant = 49.5 ± 14.2) met the inclusion criteria.

Intervention: Cochlear implantation and speech perception testing.

Main Outcome Measure: Speech perception at 50 dB HL, 35 dB HL, and 50 dB HL+5 SNR. Improvement from bimodal to bilateral performance was compared to a control group of bimodal users matched for age of 1st implantation who never received a second implant.

Results: Mean speech perception for the pediatric group using CI and hearing aid were 66.79 ± 21.82 , 40.54 ± 28.92 , and 43.54 ± 27.34 at 50 dB HL, 30 dB HL, and 50dB HL+5 SNR, respectively. One year after receiving the second implant, scores increased significantly to 82.11 ± 16.88 , 66.38 ± 23.33 , and 62.85 ± 29.13 at 50 dB HL, 30 dB HL, and 50dB HL+5 SNR, respectively ($p < 0.05$). Pediatric bimodal controls failed to demonstrate the same significant improvement. In the adult population, speech perception score improved from 45.6 ± 23.5 to 54.4 ± 25.2 ($p < 0.05$) in the bimodal control group, and from 53.5 ± 22.3 to 68.8 ± 19.5 ($p < 0.01$) in the bilateral CI group at 50 dB HL only.

Conclusion: This study supports bilateral cochlear implantation for improved speech perception performance relative to bimodal performance in the pediatric population. Improvement in the adult population was not as dramatic.

Define Professional Practice Gap & Educational Need: Need to understand the advantage of bilateral cochlear implantation in the pediatric and adult population.

Learning Objective: Speech performance advantages with bilateral cochlear implantation over bimodal amplification

Desired Result: Understanding the speech perception outcomes of bilateral cochlear implantation

IRB or IACUC Approval: Yes

Cochlear Implant-Evoked Cortical Activation in Children with Cochlear Nerve Deficiency

Craig A. Buchman, MD; Shuman He, PhD

Objective: Elucidate the value of cochlear implant-evoked cortical potentials in children with cochlear nerve deficiency (CND).

Study design: Comparative cohort study.

Setting: Tertiary referral center.

Patients: Five children with cochlear implants in the setting of CND were compared to a similar group of children without CND.

Intervention(s): Cochlear implantation

Main outcome measure(s): Cortical potentials and speech perception measures. Stimuli for cortical potentials were presented at the maximum comfortable level for each patient. Monopolar stimulation between an active, intra-cochlear electrode and the extra-cochlear ground electrode located in the temporalis muscle (MP1) was used. Responses were elicited from several electrodes along the intra-cochlear array. Raw EEG signals were differentially recorded between surface electrodes placed at the high forehead (Fz) and the contralateral mastoid. The averaged responses were analyzed offline using custom-designed MATLAB software. Speech perception ability was assessed using a hierarchy of measures.

Results: Despite evidence for sound awareness, children with CND demonstrate poor speech perception abilities and morphologically very different patterns of cortical activation using cochlear implants when compared to children with normal inner ear and cochlear nerve anatomy.

Conclusions: Taken together, these findings provide electrophysiological evidence for disordered cortical activation in response to cochlear implant stimulation in children with CND, implying either severely impaired neural transduction along the auditory system or non-auditory stimulation. Development of technologies for gathering this type of information prior to implantation might be clinically useful in choosing intervention strategies.

Define Professional Practice Gap & Educational Need: Lack of knowledge of outcomes of children with cochlear nerve deficiency using cochlear implants and the mechanisms underlying this performance.

Learning Objective: Understand the outcomes and mechanisms underlying this performance of children with cochlear nerve deficiency that have received cochlear implants.

Desired Result: Participants will recognize the need for better assessment tools in the evaluation of children with evidence of cochlear nerve deficiency prior to considering cochlear implantation.

IRB or IACUC Approval: UNC IRB# 09-1202

Cochlear Implantation in Neurofibromatosis II

Mark Brandt Lorenz, MD; Steve R. Otto, MD
Eric P. Wilkinson, MD; Derald E. Brackmann, MD

Objective: Assessment of the function of cochlear implants in patients with neurofibromatosis type II, who have a contralateral auditory brainstem implant

Study Design: Retrospective case series

Setting: Tertiary referral center

Patients: Patients diagnosed with NF2 who have both an ABI and CI

Intervention: Cochlear implantation

Main outcome measure: Audiometric performance testing with NU-CHIPS, CUNY, and Iowa Vowel and Medial Consonant metrics

Results: Eight of 266 NF2 ABI recipients received cochlear implants in an ear contralateral to an ABI. Four males and four females (ages 24-73) were identified. At least 3 months of testing data was collected for six of these patients. Two patients had better CI performance than their contralateral ABI, 2 patients had better function with their ABI. Several patients had reduction in CI performance scores with disease progression. The duration of cochlear implant usage was limited to 3-10 years in four patients. Three patients had radiosurgery to the side implanted with a CI. For ABIs, average NU-CHIPS scores were 59%, Consonants 22%, Vowels 26%, and CUNY Sentences 5%. For CIs, average NU-CHIPS scores were 77%, Consonants 50%, Vowels 60%, and CUNY Sentences were 53%. One patient had no benefit from their CI, one had CI performance deteriorate over 3 years, another over 6 years, and two over 10 years. One patient had his cochlear implant removed for a second ABI. Tumor sizes ranged from 1.75-5cm before CI failure.

Conclusion: CI outcomes are unpredictable in NF2. CI function tends to deteriorate with disease progression, and with time from therapeutic intervention. Further investigation into factors predictive of CI success in NF2 is warranted.

Define Professional Practice Gap & Educational Need: As management of patients with Neurofibromatosis II has changed with the adaptation of radiosurgery, cochlear implantation may have a role in treatment. There is limited research investigating this treatment.

Learning Objective: This work helps describes functional outcomes in patients with Neurofibromatosis II who received a cochlear implant.

Desired Result: This work suggests modification of the treatment algorithm for patients with Neurofibromatosis II.

IRB or IACUC Approval: IRB approved

Factors Associated with Recurrence in Squamous Cell Carcinoma Involving the Temporal Bone

Theodore R. McRackan, MD; Te-Yung Fang, MD; Alejandro Rivas, MD
George B. Wanna, MD; David S. Haynes, MD; Marc L. Bennett, MD

Objective: To better understand characteristics and outcomes in squamous cell carcinoma (SCC) involving the temporal bone.

Study design: Retrospective study

Setting: Subspecialty practice at a tertiary hospital

Patients: Fifty-nine patients diagnosed over a ten year period

Intervention: Patients with SCC involving the temporal bone underwent surgical resection of the malignancy.

Main outcome measures: Intraoperative and pathology tumor data, Recurrence data

Results: Decreased rates of recurrence were seen in those patients with SCC involving the temporal bone through the skin of the external auditory canal (OR=0.01; 95%CI 0.08-0.74; p=0.013) as well as those with a primary site on the auricle (OR=0.17; 95%CI 0.05-0.65; p=0.009). Nodal staging statistically correlated with rates of recurrence. Those staged N1 (OR=3.73; 95%CI 1.04-13.4; p=0.044) and N2 (OR=6.44; 95%CI 1.69-24.47; p=0.006) had increased association with malignancy recurrence. As opposed to perifacial and parotid lymph nodes (p=0.131 and p=0.368, respectively), cervical node involvement was statistically associated with recurrence (OR=4.50; 95%CI 1.40-14.45; p=0.012).

Conclusion: We have identified multiple factors that are associated with increased recurrence in SCC involving the temporal bone.

Define Professional Practice Gap & Educational Need: 1) Lack of awareness of factors associated with recurrence of squamous cell carcinoma (SCC) involving the temporal bone 2) Lack of contemporary knowledge of outcomes in patients undergoing surgical resection of SCC involving the temporal bone

Learning Objective: 1) Better understand which factors (both pre-operative and intra-operative) are associated with increased rates of recurrence 2) Better understand patient outcomes after surgical resection of SCC involving the temporal bone

Desired Result: Attendees should be able to apply the knowledge gained from this presentation to hopefully improve outcomes in patients undergoing surgery for SCC involving the temporal bone. They will also be better equipped to discuss likely prognosis with this patient population.

IRB or IACUC Approval: 110695

ANS TRAINEE AWARD WINNER

Physiology of Vestibular Dysfunction and Aging

Yuri Agrawal, MD; M. Geraldine Zuniga, MD
Marcela Davalos-Bichara, MD; Michael C. Schubert, PhD
Jeremy D. Walston, MD; John P. Carey, MD

Objective: To characterize the physiologic nature of the vestibular dysfunction that occurs with the normative aging process.

Study design: Cross-sectional study.

Setting: Tertiary care academic medical center.

Patients: Fifty individuals age 70 and above.

Interventions: Head thrust dynamic visual acuity testing (htDVA) and cervical and ocular vestibular-evoked myogenic potential (VEMP) testing.

Main Outcome Measures: Semicircular canal function measured by htDVA in each of the three semicircular canal planes, and saccular and utricular function measured by cVEMP and oVEMP testing, respectively.

Results: We observed significant declines in semicircular canal function in each of the canal planes as well as otolith function associated with aging. We found that individuals with impaired horizontal and superior semicircular canal function were likely to also have concomitant deficits in utricular but not saccular function. Overall, we noted that the prevalence of semicircular canal dysfunction was highest followed by saccular then utricular impairment, although we did observe individuals with isolated otolith deficits.

Conclusions: These data suggest an overall decline in semicircular canal as well as otolith function associated with aging, although the magnitude of impairment was greater for the semicircular canals than the otoliths in this elderly population. A better understanding of the specific vestibular deficits that occur with aging can inform the development of rational screening, vestibular rehabilitation and fall risk reduction strategies in older individuals.

Define Professional Practice Gap & Educational Need: Vestibular function has been shown to decline with aging, however it is unknown whether age-related changes occur in the semicircular canals or the otoliths or both.

Learning Objective: The learning objective is to understand the specific physiologic nature of age-related decline in vestibular function.

Desired Result: These results will be used to inform the development of rational vestibular rehabilitation and fall risk reduction strategies in older individuals.

IRB or IACUC Approval: IRB Approval: NA_00035749

Cochlear Implantation in Children with Cochlear Nerve Hypoplasia

Joseph T. Breen, MD; Matthew L. Carlson, MD
Melissa D. De Jong, AuD; Charles W. Beatty, MD
Colin L.W. Driscoll, MD; Brian A. Neff, MD

Objective: To review outcomes after cochlear implantation (CI) in children with known cochlear nerve hypoplasia (CNH) on the implanted side.

Study design: Retrospective case series with matched controls.

Setting: Tertiary referral center.

Patients: All implanted patients with radiographic evidence of ipsilateral CNH. A control patient with a radiographically normal cochlear nerve matched for age at implantation, length of deafness, and preoperative hearing loss was identified for each study patient.

Intervention: Implantation in an ear affected by CNH.

Main outcome measures: Implant usage, primary form of communication, postoperative pure tone audiometry, speech awareness and reception thresholds, speech perception scores, parent questionnaires.

Results: Six patients met study criteria (3 males, 3 females). Patients ranged in age from 22 months to 4 years at time of implantation. All but one had bilateral cochlear nerve anomalies. No patient received an implant in an ear with a radiographically absent cochlear nerve. Two patients had an associated cochlear or vestibular malformation on the implanted side. Mean length of follow up was 3.5 years. While all continue to use their implants, no patients with CNH were able to achieve open set word recognition abilities. In contrast, 5 of the 6 controls (CNC 84-90%, WIPI 72%) attained open set discrimination ($p=0.0152$). Five of the 6 subjects with CNH were dependent on manual communication.

Conclusions: Cochlear implantation in the setting of CNH continues to be controversial. We present a series of 6 implanted children with CNH with generally poor audiometric outcomes when compared to matched controls.

Define Professional Practice Gap & Educational Need: Inconsistent and sparse data regarding outcomes after cochlear implantation for patients with cochlear nerve anomalies.

Learning Objectives: 1) Review outcomes for a series of 6 patients with cochlear nerve hypoplasia who underwent cochlear implantation. 2) Review the relevant anatomy, pathophysiology, and current literature regarding cochlear implantation in patients with cochlear nerve anomalies

Desired Result: Develop appropriate expectations for outcomes after cochlear implantation for patients with cochlear nerve anomalies in the implanted ear.

IRB or IACUC Approval: IRB Approval: #11-003814

Thresholds for Human Perception of Roll Tilt Motion Depend on Visual-Vestibular Integration

Vartan Mardirossian, MD; Faisal Karmali, PhD
Daniel Merfeld, PhD

Hypothesis: The brain's estimate of spatial orientation is improved by integrating visual and vestibular inputs.

Background: A previous study we performed showed that visual and vestibular cues are fused when the brain judges the roll-tilt direction. However it was unclear whether the brain was picking the more reliable of the two cues (gating) or was able to do better than with either individual cue (integration).

Methods: A novel method of vestibular evaluation - the measurement of vestibular thresholds, was used. We used a Moog mobile platform with dedicated software. 4 subjects were tested at 1, 2, 3, 4 and 5 Hz with adaptively decreasing amplitude. Subjects were asked to indicate the direction of motion in 3 conditions: vestibular only – subjects roll tilted in the dark, visual only – a visual scene was tilted in front of subjects, and combined visual + vestibular – subjects rotated while watching a stationary visual scene. For each subject, we calculated the percent difference between the threshold for combined visual/vestibular stimuli and the best of either the vestibular or visual threshold.

Results: Thresholds with combined visual/vestibular stimuli were significantly lower than those predicted by the gating hypothesis (2-way, 1-sided ANOVA, $p=0.0365$).

Conclusions: The brain determines the body motion in space during roll tilt motion by integration of vestibular and visual inputs: the combined estimate of motion is better than with either cue individually. This research may be useful in work-up of vertiginous disorders with impaired integration of vestibular and visual cues, (motion-sickness and migraine dizziness).

Define Professional Practice Gap & Educational Need: Despite advances in the study of the human vestibular system and the diagnostic methods for the measurement of vestibular function, little is known about vestibular perception and vestibular thresholds or the mechanisms that the human brain uses to integrate visual and vestibular input about the body position in space

Learning Objective: Learn that the measurement of vestibular thresholds provides a novel method to evaluate vestibular function, as well as having significant potential in studying the pathways of vestibular and visual input integration in the brain

Desired Result: Understanding the modalities of visual-vestibular integration in the brain represents a fundamental finding for further clinical investigations in mal deDebarquement and migraine patients

IRB or IACUC Approval: Human Studies Committee Protocol Number: 98-09-027

Mechanisms of Hearing Loss Associated with Sporadic Vestibular Schwannomas: Clues from Tumor Secretions

Sonam Dilwali, Andrew Lysaght; Daniel Roberts, MD, PhD
Konstantina M. Stankovic, MD, PhD

Hypothesis: We hypothesize that the severity of hearing loss (HL) associated with sporadic vestibular schwannomas (VSs) is correlated with tumor secretion of proteins with ototoxic potential.

Background: Since it has been recognized that HL associated with VSs is not solely due to auditory nerve compression, elucidating the tumor pathophysiology that causes hearing loss has been an important and intriguing task. We have previously shown that VSs stratified by hearing have differential gene expression. We have now focused on identifying differentially expressed proteins in tumor secretions.

Methods: Fresh surgical specimens of VSs were incubated in sterile PBS for 1 hour at 37°C to collect secretions. The specimens were divided into a group associated with good hearing (word recognition >70% and pure-tone average ≤30 dB, N=2) or poor hearing (N=5). The two groups were compared using a customized RayBiotech L-series cytokine array.

Results: Of the 22 cytokines that we studied, only 3 were expressed in VSs secretions: IL-1 Receptor Antagonist (IL-1ra), platelet derived growth factor PDGFaa and PDGFbb. All 3 proteins were expressed at substantially higher levels in tumors associated with poor re good hearing. Interestingly, we also detected merlin, a cytoskeletal protein, in secretions of tumors associated with poor, but not good hearing.

Conclusions: This study highlights several proteins that may mediate hearing loss associated with VSs. Experiments are underway to validate these preliminary findings using more tumor samples for each hearing group.

Define Professional Practice Gap & Educational Need: There is a lack of effective treatments for the progressive sensorineural hearing loss associated with vestibular schwannomas. This hearing loss substantially decreases patients' quality of life and leaves them with inadequate treatment options for prevention of further hearing loss. Our superficial understanding of the pathophysiology of vestibular schwannomas hinders our goals to design potent therapies.

Learning Objective:

- (1) Understand the possible mechanisms through which hearing loss could be correlated with tumor secretion of proteins that have an ototoxic potential
- (2) Be informed of potential therapeutic targets that could decrease or prevent hearing loss in patients with sporadic vestibular schwannomas

Desired Result: If we could understand the mechanisms that lead to hearing loss associated with vestibular schwannomas, we will be able to develop effective options for treatment and/or prevention. Further, it can also provide us insight into the general pathophysiology of vestibular schwannomas.

IRB or IACUC Approval: IRB Protocol Number 10-04-028

Cochlear Implantation in Patients with Neurofibromatosis Type II

Matthew L. Carlson, MD; Joseph T. Breen, MD
Colin L. Driscoll, MD; Michael J. Link, MD
Brian A. Neff, MD; Charles W. Beatty, MD

Objective: To investigate cochlear implant (CI) performance outcomes among patients with Neurofibromatosis Type II (NF2)

Study design: Retrospective case series

Setting: Tertiary academic referral center

Patients: All patients with NF2 having an anatomically intact ipsilateral cochlear nerve who underwent CI

Intervention(s): Cochlear implantation

Main outcome measure(s): Postoperative audiometric performance in the electric-only condition including speech reception thresholds, Consonant-Nucleus-Consonant word scores, and Hearing in Noise Test performance.

Results: Nine patients (4 females, mean age at implantation 49 yrs) met study criteria. The mean duration of follow-up was 32 months (median 31 mo, range 2-72). Three patients underwent previous stereotactic radiosurgery, 5 received prior microsurgical resection, and a single patient was observed without intervention. The mean pre-implant duration of deafness was 6.0 years. All subjects achieved sound awareness with an average speech reception threshold of 31 dB; 5 attained open-set speech recognition (CNC 22-46%; HINT 63-98%). Patients with a duration of deafness less than 4 years were more likely to attain open-set discrimination capacity compared to those with longer durations of deafness ($p=0.048$).

Conclusions: Cochlear implantation remains a viable method of hearing rehabilitation in patients with NF2 who have an anatomically intact cochlear nerve. Open-set speech discrimination is possible in more than half of subjects. Similar to conventional CI recipients, a prolonged duration of auditory deprivation may negatively influence subject performance.

Define Professional Practice Gap & Educational Need: Currently, there is a lack of knowledge regarding the best method of hearing rehabilitation for patients with Neurofibromatosis Type II and severe-to-profound sensorineural hearing loss. Much of the literature within the last decade has focused on auditory brainstem implantation while cochlear implantation among patients with an intact ipsilateral cochlear nerve has received much less attention.

Learning Objective: The learning objective of this talk is to describe cochlear implant performance outcomes among patients with Neurofibromatosis Type II and an intact ipsilateral cochlear nerve.

Desired Result: The desired result is that based on a more complete understanding of the expected audiometric outcomes and variables that may influence individual subject performance, clinicians will apply this information to make informed decisions on recommended treatment strategies to improve patient outcomes.

IRB or IACUC Approval: IRB Approval: 11-007118

Efficacy of Hearing Aid Trial in Determining Pediatric Cochlear Implantation Candidacy

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Objective: To determine whether hearing aid (HA) trials affect the decision to perform cochlear implantation (CI) in children with severe to profound sensorineural hearing loss (SNHL).

Study Design: Retrospective review.

Setting: Tertiary-care university hospital.

Patients: 383 children with severe to profound SNHL referred for CI evaluation between 1/2002 and 6/2009.

Interventions: Hearing aid use and weekly speech therapy.

Main outcome measures: Aided sound field testing, Meaningful Auditory Integration Scale (MAIS) evaluation or infant-toddler MAIS (ITMAIS) evaluation.

Results: Of the 383 children, 238 were approved and underwent CI, 54 children were in the review process, and 91 were not implanted. The rationale for not implanting these 91 children were reviewed: 32 lost to follow up, 19 not audiometric candidates, 19 due to family decision, 8 due to poor social situation, 7 relocated, 6 medically contraindicated. Notably, no child was not implanted due to positive benefit from amplification. Of the original 383 children, we examined the benefit of the 3-6 month hearing aid trial on 95 children whose hearing loss was identified before 18 months of age and who received amplification before 24 months of age. Linear regression analysis demonstrated poor correlation between MAIS/ITMAIS performance and time of hearing aid use. Ultimately, 86 of these 95 children underwent CI. The slopes of MAIS/ITMAIS performance versus time of hearing aid use for the 86 implanted children versus the 9 non-implanted children did not differ significantly.

Conclusion: HA trials do not change the decision to perform CI in children with severe to profound SNHL.

Define Professional Practice Gap & Educational Need: Generally, children with severe to profound sensorineural hearing loss who are undergoing evaluation for cochlear implantation need to undergo a hearing aid trial of at least 3 months. There is little evidence for the length of the hearing aid trial or for the need for the hearing aid trial at all.

Learning Objective: To determine in our cochlear implant program whether the result of the hearing aid trial affected our decision whether or not to perform a pediatric cochlear implantation.

Desired Result: We find that the hearing aid trial did not ever ultimately affect the decision whether or not to perform a pediatric cochlear implantation. Attendees may decide to proceed with cochlear implantation earlier if all other aspects of candidacy have been satisfied and only completion of the hearing aid trial is pending.

IRB or IACUC Approval: Yes, Baylor College of Medicine protocol# H-24980

Intraoperative Round Window Recordings of Acoustically Evoked Cochlear Potentials in Cochlear Implant Recipients

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Hypothesis: Acoustically evoked cochlear potentials can be measured from the round window (RW) intraoperatively even in profoundly deaf patients undergoing cochlear implants.

Background: Patients with a substantial amount of functional hearing but poor speech recognition scores are receiving cochlear implants. Recent trends are towards preservation of residual hearing. However, even patients who are profoundly deaf may have some residual function of hair cells, likely to be in low frequency regions. If true, improved methods to assess cochlear functioning and preserve hearing even in these cases may serve to enhance patient outcomes postimplantation.

Methods: Patients (6 months – adult) undergoing cochlear implantation were included. The Navigator Pro auditory diagnostic system (Biologic Systems Corp.) was used to measure potentials by placing a facial nerve monitor probe at the RW after surgical access to the RW was obtained. The cochlear microphonic (CM), compound action potential (CAP), and neurophonic potential were recorded to tone bursts at frequencies of 0.25-2 kHz at various levels administered through a sound tube.

Results: All patients tested so far (n=9) had measurable potentials to acoustic stimuli, including those who were audiometrically determined to be profoundly deaf at all frequencies. In general a larger magnitude of response was noted for patients with higher audiometric thresholds but this was not true in all cases.

Conclusions: Acoustically evoked cochlear potentials are detectable in candidates for cochlear implantation even if detectable hearing is limited. These findings indicate that a more sensitive measure of cochlear status could be useful in tailoring interventions for hearing loss.

Define Professional Practice Gap & Educational Need: Lack of awareness of possible residual cochlear function in profoundly deaf individuals.

Learning Objective: To become aware of the amount of residual cochlear function that may be present in a profoundly deaf patient.

Desired Result: Attendees will keep in mind hearing preservation techniques even in profoundly deaf individuals to possibly improve overall hearing outcomes.

IRB or IACUC Approval: IRB Approval: Study # 05-2616

Association of Benign Intracranial Hypertension and Spontaneous Encephalocele with Cerebrospinal Fluid Leak

Laura Brainard, MD; Douglas A. Chen, MD; Todd A. Hillman, MD

Objective: To determine the incidence of intracranial hypertension in patients with spontaneous encephalocele with cerebrospinal fluid (CSF) leak.

Study design: Retrospective case review

Setting: Tertiary care neuro-otologic practice

Patients: Patients presenting since 2008 with spontaneous encephalocele and CSF leak in the temporal bone.

Intervention(s): Lumbar puncture with opening pressure measurement following encephalocele repair.

Main outcome measure(s): Patient age, sex, post-operative course, Body Mass Index and intracranial pressure as measured by post-operative lumbar puncture opening pressure.

Results: Of the 26 patients identified with spontaneous encephalocele with CSF leak, 9 patients had post-operative lumbar puncture data. Of those 9, 89% were female, 11% male, with a mean age of 57 and a mean BMI of 41 kg/m² (morbidly obese). The mean opening pressure was 24.5 cm H₂O. 33% had normal intracranial pressure (mean 15cmH₂O, range 10 to 17 cmH₂O). 67% had elevated intracranial pressure (mean 29 cmH₂O, range 23.5 to 40 cmH₂O). The incidence of benign intracranial hypertension in the general population is 0.001%. Of the 6 with intracranial hypertension, 3 (50%) were placed on acetazolamide, for fundoscopic findings, postoperative headache, and/or visual changes. Mean time to LP after repair of encephalocele was 13 months (range 4 days to 75 months).

Conclusions: This study shows that BIH is prevalent in a significant number of patients presenting with spontaneous encephalocele with CSF otorrhea at a rate much higher than is found in the general population. This suggests that all patients with spontaneous encephalocele with CSF leak warrant evaluation for BIH.

Define Professional Practice Gap & Educational Need: Although the association of benign intracranial hypertension and spontaneous encephalocele with cerebrospinal fluid leak has been hypothesized, it has not been previously demonstrated, and there is a lack of awareness about this association.

Learning Objective: The learning objective is to describe the association of benign intracranial hypertension and spontaneous encephalocele with cerebrospinal fluid leak.

Desired Result: The desired result is that clinicians will understand that there is an association of benign intracranial hypertension and spontaneous encephalocele with cerebrospinal fluid leak and that these patients have a much higher incidence of benign intracranial hypertension (BIH) than the general population. It is also desired that, based on this understanding, clinicians will evaluate these patients for BIH, so that they may be treated for it, if indicated.

IRB or IACUC Approval: n/a

Temporal Binding Window in Vestibular Migraine

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Hypothesis: Patients with vestibular migraine have widened temporal binding windows.

Background: Maintenance of balance requires accurate integration of multiple sensory cues. Physical and physiologic limitations prevent cues deriving from a single event to be perceived exactly simultaneously. Related cues must be integrated despite slight asynchronies separating their perception. If cues are not temporally integrated accurately, confusion can ensue, much like a movie with the audio and video tracks out of synch. Temporal integration is characterized by the temporal binding window, which is the length of time over which two sensory cues may be presented and still perceived as simultaneous. The temporal binding window is widened in dyslexia, autism, and the elderly.

Methods: Subjects with vestibular migraine were rotated sinusoidally at 0.5 Hz, 20 deg/sec peak velocity about an earth-vertical axis. A metronome-like click at the same frequency was presented dichotically via headphones, alternating between the ears. Subjects estimated the alignment of the alternating clicks and vestibular stimulus using a method of constant stimuli.

Results: The temporal binding window was defined as the width of time over which subjects felt the clicks and vestibular stimulus were aligned. Subjects with vestibular migraine demonstrated temporal binding windows approximately twice as wide as control subjects.

Conclusions: The temporal binding window of subjects with vestibular migraine is widened. Inaccurate temporal integration of vestibular inputs with other stimuli may underlie motion intolerance and imbalance in these subjects. A widened temporal binding window offers a plausible explanation of why migraineurs are particularly sensitive to sensory overstimulation.

Define Professional Practice Gap & Educational Need: Lack of awareness of cause of vestibular migraine; lack of diagnostic tool for vestibular migraine

Learning Objective: Recognition of multisensory integration as a factor in causation of vestibular migraine

Desired Result: Will consider timing relationships as a possible diagnostic tool for vestibular migraine

IRB or IACUC Approval: 08-1227

Prognosis for Contralateral Hearing in Children Presenting with Unilateral Sensorineural Hearing Loss

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Study design: Retrospective review

Setting: Tertiary-care university hospital

Patients: 115 children with USNHL referred between 1/2004-6/2011

Intervention(s): none

Main outcome measure(s): Audiometry, diagnostic imaging

Results: 115 children with USNHL were stratified based on inner ear findings on diagnostic imaging: A) normal bilaterally, (60.9%, n=70); B) ipsilateral abnormality, (30.4%, n=35); C) bilateral abnormality, (8.7%, n=10). No patients had contralateral abnormality only. The incidence of contralateral SNHL at presentation was 11% (8) in group A, 17% (6) in group B, and 40% (4) in group C. Of patients with follow-up audiometry, 10% (5/51) of group A, 11% (2/18) of group B, 44% (4/9) of group C developed CSNHL at an average follow-up time of (A) 1.9, (B) 1.2, and (C) 1.8 years. Of the patients developing progressive CSNHL, 55% (6/11) had inner ear imaging abnormalities including: 50% (3/6) hypoplastic cochlear nerve/aperture stenosis, 50% (3/6) dysmorphic/absent semicircular canals, 33% (2/5) cochlear malformation, 33% (2/5) enlarged vestibular aqueduct.

Conclusions: Imaging provides an important role in the diagnostic as well as prognostic evaluation of unilateral SNHL. Even with normal diagnostic imaging, however, serial audiometry remains essential for detecting onset of CSNHL as well as progression of USNHL.

Define Professional Practice Gap & Educational Need: Lack of medical knowledge regarding management of unilateral sensorineural hearing loss

Learning Objective: Determine prevalence and progression of contralateral sensorineural hearing loss in children presenting with unilateral sensorineural hearing loss

Desired Result: Increased awareness and utilization of audimetry and imaging studies in diagnostic as well as prognostic evaluation of unilateral sensorineural hearing loss

IRB or IACUC Approval: Pending