

2010 ANS Abstracts Selected for Presentation

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Correlation between Music Perception and Speech Recognition in Cochlear Implant Users

David R. Friedland MD, PhD; John J. Nash, MD
Christina L. Runge-Samuelson, PhD

Objective: To assess the ability of cochlear implant patients to perceive distinct characteristics of music and their relationship to speech performance.

Study Design: Prospective analysis of music and speech perception.

Setting: Tertiary referral center.

Patients: Adult cochlear implant users with at least 1 year post-implant experience.

Intervention: Testing for the ability to distinguish melody patterns and chords and to identify a closed-set of musical instruments (timbre test).

Main Outcome Measure: Comparisons were made across subjects as well as to normal hearing controls. Regression analyses were used to correlate music perception with speech recognition scores.

Results: Instrument identification (i.e., timbre test) was the most difficult task for cochlear implant patients (avg. correct: 53 \square 15.8%). In contrast, normal hearing subjects had an average score of 99.2%. Distinguishing chords 74 \square 10.2% and melodies 67 \square 10.4% were easier. Normal hearing controls, as a group, performed better than cochlear implant patients on the latter tasks but there was some overlap in individual performance. Bimodal hearing provided a modest benefit in most cochlear implant subjects. Regression analyses showed a strong and significant correlation between speech recognition and chord and timbre scores. Timbre, in particular, correlated strongly with CNC ($r=.77$, $p<.001$), HINT/Q ($r=.76$, $p<.001$), and HINT/N ($r=.69$, $p<.001$) scores.

Conclusions: Cochlear implant patients appreciate some aspects of music at levels similar to normal hearing listeners but complex sound perception (i.e., timbre) remains a significant challenge. There is a strong association between speech understanding and timbre which may thus represent a non-verbal substitute for speech recognition testing.

Authors were required to include the following with their abstract submission.

Define Professional Practice Gap & Educational Need: There currently exists a lack of understanding regarding factors that improve music perception among cochlear implant users; factors which not only correspond to music enjoyment but may also be critical to improving speech recognition.

Learning Objective: The learning objective of this talk is to define individual components of music and how they are processed by implant users as well as to relate those musical concepts to speech recognition.

Desired Result: Based on more complete recognition of sound processing with cochlear implants, physicians and paraprofessionals can change their treatment of implant patients to maximize music enjoyment and potentially speech recognition.

IRB # - PRO00010547

Computerized Objective Measurement of Facial Motion: Normal Variation and Test-Retest Reliability

John Gail Neely, MD; Katie X. Wang, BA; Chelsea A. Shapland
Ali Sehizadeh; Amy Wang

Objective: Objective quantitative measurements of facial motion for the assessment of outcomes in patients with facial paralysis have been elusive. This paper will demonstrate a novel computerized program for objective measurement of facial motion and present data on symmetry in normal subjects and test-retest reliability in patients with facial paralysis.

Study design: Prospective analysis of archived images.

Setting: Tertiary referral center.

Patients: Good quality recordings of forty-two normal subjects and 30 facial paralysis subjects with a wide range of paralysis that had been tested twice were selected.

Intervention: Using image subtraction techniques of digital video recordings, computer generated strength-duration curves of prescribed facial movements were automatically constructed. Main outcome measures: The areas under the curve for specific regions of each side of the face were compared as a proportion described as a percentage, left over right, in which 100% would be perfect symmetry.

Results: Forty-two normal subjects had left/right symmetry of 97.88% (95%CI 96.02 – 99.74). Thirty patients with varying degrees of facial paralysis, tested twice, were evaluated to determine the degree of agreement between trials as measured by the intraclass correlation coefficient (ICC). The results showed the ICC (95%CI) for brow was 0.972 (0.943 – 0.987), eye 0.950 (0.898 – 0.976), mouth smiling 0.951 (0.901 – 0.976).

Conclusions: These results demonstrate that a computerized system can be objectively and reliably used to evaluate facial motion. This system can be made available at no cost to interested investigators for their own investigations.

Define Professional Practice Gap & Educational Need: Objective quantitative measurements of facial motion have been elusive. This is especially lacking and needed in comparative outcome assessments for patients recovering from facial paralysis and for planning and evaluating reanimation surgery and rehabilitative methods.

Learning Objective: The learning objective of this talk is to demonstrate a novel computerized system for the quantitative assessment of facial motion and to present data on symmetry in side comparisons in normal subjects and test-retest reliability in facial paralysis subjects.

Desired Result: The attendees will become aware of a valid objective method for facial motion assessment and will be able to obtain the program without cost, upon request, for their own investigations.

IRB # - HRPO #03-0218

Diabetes, Vestibular Dysfunction, and Falls: Analyses from the National Health and Nutrition Examination Survey, 2001-2004

Yuri Agrawal, MD; Lloyd B. Minor, MD

Objective: Diabetics have been shown to be at increased risk both for falls and for vestibular dysfunction, a known risk factor for falls. Our aims were: 1) to evaluate whether the influence of diabetes on vestibular function can be explained by the concomitant peripheral neuropathy and retinopathy associated with diabetes; and 2) to assess the relative contributions of vestibular dysfunction, peripheral neuropathy and retinopathy in mediating the association between diabetes and falls.

Study design: National cross-sectional survey.

Setting: Ambulatory examination centers.

Patients: US adults aged 40 and above who participated in the 2001-2004 National Health and Nutrition Examination Survey (N=5086).

Interventions: Diagnosis of diabetes, peripheral neuropathy and retinopathy.

Main Outcome Measures: Vestibular function measured by the modified Romberg Test of Standing Balance on Firm and Compliant Support Surfaces, and history of falling in the previous 12 months.

Results: We observed that diabetes significantly increased the odds of vestibular dysfunction by 50%, even after adjustment for peripheral neuropathy and retinopathy. Moreover, we found that vestibular dysfunction and peripheral neuropathy were each significant independent mediators of the association between diabetes and fall risk: vestibular dysfunction and severe peripheral neuropathy increased the odds of falling 2.5-fold and 3.8-fold respectively in multivariate analyses.

Conclusions: Diabetes seems to exert direct pathologic effects on the vestibular system. Furthermore, the association between diabetes and falls appears to be mediated in part by dysfunction of vestibular structures. Better diabetes management and control may provide an opportunity to reduce the risk of vestibular dysfunction and falls.

Define Professional Practice Gap & Educational Need: There is currently a lack of awareness about the nature of the associations between diabetes, vestibular dysfunction and falls; it is unclear whether diabetes is an independent risk factor for vestibular dysfunction and falls or whether the effects of diabetes can be explained by the concomitant peripheral neuropathy and retinopathy associated with diabetes.

Learning Objective: To determine if diabetes directly leads to vestibular dysfunction, and whether diabetes increases the risk of falls due to pathologic effects on the vestibular system.

Desired Result: The desired result is that based on a more complete understanding of the associations between diabetes, vestibular dysfunction and falls, clinicians may incorporate better diabetes management and control as part of their fall risk reduction strategy.

IRB # - n/a

Predictive Capability of Historical Data for Diagnosis of Dizziness

Jeff G. Zhao, BA; Jay F. Piccirillo, MD; Edward L. Spitznagel Jr., PhD
Dorina Kallogjeri, MD MPH; Joel A. Goebel, MD

Objective: This study examines categorical responses to questions on a comprehensive dizziness questionnaire, to find its overall predictive power, and identify which question(s) are most predictive of each diagnosis.

Study design: Retrospective chart review.

Setting: Specialized Dizziness and Balance Center at tertiary care hospital.

Patients: 619 patients (19 – 89 yo, 61% F and 39% M), diagnosed with one of 24 types of dizziness or postural instability. Excluded were patients with more than one primary diagnosis.

Intervention: Standardized 163 item dizziness questionnaire (including 77 review of systems items).

Primary outcome measure: Predicted diagnoses from the questionnaire, determined by binary and multinomial logistic regressions, are compared to an ultimate clinical diagnosis made by an expert neurotologist based on full interview, examination, and clinical tests.

Results: Significant question groupings exist for each of the main diagnoses. A subset of 47 questions under multinomial logistic regression gave high predictive accuracies for migraine (92%), BPPV (90%) and Meniere's disease (86%), and fair predictive power for vestibular neuritis (63%), contributing to an overall predictive accuracy of 84%. A smaller subset of 32 questions gave an overall predictive accuracy of 77%.

Conclusions: The capability of historical data to accurately predict the ultimate diagnosis for dizziness emphasizes the importance of a structured questionnaire in the evaluation of such patients. Future developments include the formulation of a computer-based program to generate a differential diagnosis for the practitioner to consider.

Define Professional Practice Gap & Educational Need: 1. Inefficient use of historical patient data among physicians treating dizzy patients. 2. Need to train physicians on the appropriate and efficient use of historical information for developing a differential diagnosis of dizziness.

Learning Objective: To identify and disseminate a concise questionnaire tool to facilitate evaluation of dizzy patients.

Desired Result: Improved efficiency and accuracy of the use of historical data in aiding diagnosis of dizziness.

IRB # - Supported by: NIH TL1RR024995
IRB or IACUC Approval: 07-1085

Cost Effective Evaluation of Dizziness: Brain MRI and ENG

Mohamed Hamid, MD, PhD

Objective: Study the cost effectiveness of brain MRI and ENG in the course of evaluation of dizzy patients.

Study design: Prospective one-year analysis of MRI and ENG test results of dizzy patients.

Setting: Tertiary referral Neurotology center

Patients: New patients (N=237) undergoing neurotologic evaluation.

Intervention(s): Comprehensive neurotologic history and office examination, ENG testing and brain MRI.

Main outcome measure(s): Sensitivity analysis of MRI and ENG studies. Comparative analysis of office vestibular examination findings and ENG results.

Results: The number of brain MRI studies was 153. All studies were normal except two (Chiari malformation and a meningioma) rendering an MRI sensitivity of 1.31%. Given that the average cost of an MRI is about \$4000, it costs \$400,000 to find a single abnormal MRI. The number of ENG studies was 168. Of these, 124 showed abnormalities rendering an ENG sensitivity of 74%. The most common ENG abnormalities were peripheral. Comparative analysis of office vestibular examination and ENG abnormalities showed 95% concordance.

Conclusions: There is a significant number of costly brain MRI studies with very low yield (1.3%) in the course of evaluating dizziness. While ENG sensitivity is 74%, 95% of ENG abnormalities were evident during office vestibular examination. Physicians can rely on vestibular exam when ENG is not available and lower their thresholds in ordering MRI and ENG studies in the course evaluating dizzy patients.

Define Professional Practice Gap & Educational Need: Lack of awareness of MRI and ENG yield in evaluating dizziness

Learning Objective:

1. Cost effective use of brain MRI and ENG in the evaluation of dizziness.
2. Low yield of MRI in dizziness evaluation.
3. Reliance on vestibular examination when ENG is not available.

Desired Result:

1. Low yield of MRI in dizziness evaluation
2. Rely on vestibular exam when ENG is not available

Sudden and Rapidly Progressive Hearing Loss in Bariatric Surgery Patients

Mark Brandt Lorenz, MD; Waleed A. Abuzeid, MD
Teresa A. Zwolan, PhD; Hussam K. El-Kashlan, MD

Objective: To investigate shared risk factors among patients who undergo weight-reduction surgery and subsequently develop sudden or rapidly progressive sensorineural hearing loss

Study Design: Retrospective clinical study from 2003 to present

Setting: Tertiary academic referral center

Patients: 8 adults (>18 y/o) with sudden or rapidly progressive bilateral sensorineural hearing loss after undergoing bariatric surgery. In each case, the hearing loss occurred within 6 months of surgery, and was preceded by marked shifts in weight and nutritional status.

Intervention: Diagnostic

Main Outcome Measures: Audiometric changes, imaging abnormalities, rate and total amount of weight loss, vitamin deficiencies and metabolic abnormalities, and response to conservative and operative hearing therapies.

Results: Patients who experience a rapid decrease in body-mass index, protracted dehydration, or have specific vitamin deficiencies are at risk for permanent bilateral sensorineural hearing loss. Several of these patients required cochlear implantation, and had excellent post-operative outcomes as determined by threshold and speech recognition testing.

Conclusions: Sudden and rapidly progressive hearing loss are not reported complications of bariatric surgery. Numerous patients have suffered permanent bilateral sensorineural hearing loss after weight-reduction surgery in the context of nutritional deficiencies. This project underscores the importance of aggressive nutritional supplementation after bariatric surgery, and sheds light on metabolic influences involved in sudden and rapidly progressive hearing loss.

Define Professional Practice Gap & Educational Need: Sudden and rapidly progressive sensorineural hearing loss are devastating clinical problems, which have not been defined or described as complications of bariatric surgery.

Learning Objective: This project highlights the metabolic influences of sudden hearing loss, and calls attention to a postoperative complication which warrants further research and attention in the otolaryngology and general surgery literature.

Desired Result: This project may increase understanding of the metabolic influences involved in hearing loss, and rehabilitative options available for this population.

IRB # - HUM00033763

The Relationship Between the Air-bone Gap and the Size of Superior Semicircular Canal Dehiscence

Marcus D. Atlas, MBBS; Heng-Wai Yuen, MBBS
Rudolf Boeddinghaus, MBChB; Robert H. Eikelboom, MappIsc, PhD

Objective: To examine the relationship between the ABG and size of superior semicircular canal dehiscence (SSCD) as measured on computer tomography (CT).

Study design: Case series with chart review

Setting: Tertiary referral center

Patients: Twenty-three patients (28 ears) diagnosed with SSCD.

Main Outcome measures: The size of dehiscence on CT scans and the air-bone gap (ABG) on pure-tone audiometry are recorded.

Results: The size of the dehiscence ranged from 1.0 to 6.0 mm (mean 3.5 ± 1.6 mm). Six ears with dehiscence measuring less than 3.0 mm did not have an ABG (0 dB). The remaining 18 ears demonstrated an average ABG at 500, 1000 and 2000 Hz ($AvABG_{500-2000}$) ranging from 3.3 to 27.0 dB (mean 11.6 ± 5.7 dB). Analysis of the relationship between the dehiscence size and $AvABG_{500-2000}$ revealed a correlation of $R^2=0.828$, $p<0.001$ (quadratic fit) and $R^2=0.780$, $p<0.001$ (linear fit). Therefore the larger the dehiscence, the larger the air-bone gap at lower frequencies on pure-tone audiometry.

Conclusions: In SSCD patients, an ABG is consistently demonstrated at the low frequency when the dehiscence is larger than 3mm. The size of the average ABG correlates with the size of the dehiscence. These findings highlight the effect of dehiscence size on conductive hearing loss in SSCD, and contribute to a better understanding of the symptomatology of patients with SSCD.

Define Professional Practice Gap & Educational Need: There is a current lack of understanding of the exact underlying mechanism of the pathophysiology and manifestations of superior semicircular canal dehiscence.

Learning Objective: The learning objective would be to describe the relationship between the size of dehiscence of the superior semicircular canal and the air-bone gap and the role this would play in the pathophysiology of the manifestations of this condition.

Desired Result: The desired result is a better understanding of the pathophysiology and the manifestations of superior canal dehiscence. Clinicians would also be able to explain to patients the reason(s) behind the different presentation of this condition.

IRB # - IRB approval

**Long Term Follow-up of Cochlear Implant Performance
and Residual Hearing Preservation after
Round Window Electrode Insertion**

Ana H. Kim, MD; Simon Parisier, MD; Matthew Jung, MS
Christopher Linstrom, MD; George Alexiades, MD
Lisa Goldin, MA; Sabrina Vitulano, AuD

Objective: To evaluate the long-term hearing preservation rate after round window (RW) electrode insertion, and to compare their cochlear performance to those who underwent traditional cochleostomy surgery.

Study design: Prospective study

Setting: Academic cochlear implant center

Patients: Inclusion criteria used: (1) measurable hearing on unaided or aided testing pre-op, (2) favorable RW anatomy intra-op. Patients who underwent traditional cochleostomy surgery were matched for age, duration of deafness, and pre-operative hearing to serve as controls.

Intervention(s): RW versus traditional cochleostomy

Main outcome measure(s): (1) Unaided/aided test with hearing aid. (2) Speech awareness/reception threshold (SAT/SRT), consonant-nucleus-consonant (CNC), hearing in noise test in quiet (HINT) for adults, and phonetically balanced-kindergarten test (PBK) and Northwestern University children's perception of speech (NUCHIPS) for children, between 3 to 24 months after cochlear implant activation.

Results: 21 patients underwent RW electrode insertion between 2007-2009. Residual hearing was preserved in 8 out of 12 (67%) to within 0-25 dB. Remaining 9 did not have unaided/aided testing. No significant differences in post-op implant speech perception testing were noted between the RW hearing preserved, RW non-hearing preserved, and control groups. In addition, no significant mapping differences (C and T levels) were noted among the three groups. 2 patients in the RW series were bilateral recipients who previously underwent traditional cochleostomy in the contralateral ear prior to sequential RW insertion. Implant performance showed comparable speech performance.

Conclusions: Our study suggests that in select patients, residual preoperative hearing can be preserved using RW insertion. These patients demonstrate comparable speech benefit compared to the traditional cochleostomy group.

Define Professional Practice Gap & Educational Need: Currently, there is great debate regarding the utility and feasibility of round window electrode insertion in cochlear implant surgery.

Learning Objective: Successful audiologic outcome after round window electrode insertion in cochlear implant surgery will be presented.

Desired Result: This surgical technique has important implications in Electrical Auditory Stimulation (EAS) technology.

IRB # - yes

An Animal Model for a Near Real-Time Monitoring System to Assess Intracochlear Electrode Insertion Trauma

Oliver F. Adunka, MD; Adam P. Campbell, BA; Thomas A. Suberman, BA
Craig A. Buchman, MD; Douglas C. Fitzpatrick, PhD

Hypothesis: Monitoring of the functional correlates of intracochlear trauma is possible using a modified cochlear implant electrode system in an animal model.

Objective: To establish an animal model of cochlear implantation that allows for near real-time functional assessments of intracochlear electrode insertion trauma and positioning.

Methods: Experiments were conducted in the normal hearing gerbil. A flexible micro-endoscope (diameter 0.3 mm) was placed on the round window membrane and a platinum-iridium wire electrode was advanced through a basal turn cochleostomy. Electrophysiologic responses (cochlear microphonic and compound action potentials) to acoustic stimuli were obtained at various insertion depths. The endoscope was used to assess the intracochlear electrode position at each recording step. These data were correlated with histologic evaluations of intracochlear trauma.

Results: Results demonstrated the feasibility of this approach. Specifically, endoscopic images correlated well with histologic results and functional assessments. The cochlear microphonic potential provided a near real-time measure of intracochlear trauma. Subtle amplitude changes in the cochlear microphonic and compound action potentials indicated imminent intracochlear damage and subsequent reversibility was possible following electrode withdrawal.

Conclusions: Near real-time functional assessments can predict both reversible and irreversible electrode induced trauma. This system might have utility in human cochlear implantation.

Define Professional Practice Gap & Educational Need: Electrode insertion during cochlear implantation is often associated with various degrees of hearing loss presumably resulting from intracochlear damage. This damage and the positioning of the stimulating electrode might affect postoperative speech perception performance with the implant. Therefore, a functionally based monitoring system that provides real time information about intracochlear electrode positioning and trauma could allow for improved electrode insertions.

Learning Objective: To learn about the animal model work that supports the establishment of a real time, electrophysiological monitoring system for cochlear implant electrode insertion.

Desired Result: To learn more about monitoring the functional status of the cochlea during electrode implantation in an animal model and future human applications.

IRB # - UNC IACUC 08-135

**Endolymphatic Hydrops in Meniere's Disease Detected
by MRI after Intratympanic Administration of
Gadolinium: in Comparison with Sudden Deafness**

Arata Horii, MD, PhD; Yoshihiro Osaki, MD, PhD; Tadashi Kitahara, MD, PhD
Takao Imai MD, PhD; Suetaka Nishiike, MD, PhD
Norihiro Fujita, MD, PhD; Hidenori Inohara, MD, PhD

Objective: Three tesla MRI with intratympanic administration of gadolinium has been reported to successfully visualize the endolymphatic hydrops. The purpose of this study is to investigate the detection rate of endolymphatic hydrops in patients Meniere's disease and sudden deafness.

Study design: A prospective study.

Setting: Tertiary referral university hospital.

Patients: Each of eight patients with intractable unilateral Meniere's disease or sudden deafness who were subjected to intratympanic gentamicin or steroid therapy, respectively.

Intervention: Gadodiamide hydrate (Gd-DTPA-BMA, Omniscan) diluted 8-fold with saline was injected into the ipsilateral tympanic cavity 24 hours before the MRI. Axial MR images by 2D-FLAIR sequences were taken by 3 tesla MR unit.

Main outcome measures: Two doctors independently judged the existence of endolymphatic hydrops in MR images without knowledge of diagnosis.

Results: Endolymphatic space can be detected as a low or no signal intensity area, while perilymphatic space showed high intensity due to enhancement by gadolinium. There was no disagreement of the judge of endolymphatic hydrops between the two doctors: Seven of eight patients with Meniere's disease and two of eight patients with sudden deafness were diagnosed as having endolymphatic hydrops. This detection rate was statistically different between the diseases (Fisher's exact test, $p=0.0406$).

Conclusions: MR images with intratympanic gadolinium injection can be a powerful tool to detect endolymphatic hydrops in Meniere's patients. Secondary hydrops after inner ear insults including sudden deafness may be found in future by this technique that should be strictly separated from the idiopathic primary endolymphatic hydrops, Meniere's disease.

Define Professional Practice Gap & Educational Need: Lack of contemporary knowledge. Endolymphatic hydrops were only diagnosed by physiological test such as ECoG and glycerol test. Radiological diagnosis of endolymphatic hydrops is a new and challenging issue.

Learning Objective: If endolymphatic hydrops can be diagnosed by radiological methods, this can be applied to those who have profound hearing loss in which physiological tests cannot be performed.

Desired Result: MRI in combination with intratympanic gadolinium does not require any special technique or modalities. It can be expected that many attendees can perform this examination in their own hospitals so that endolymphatic hydrops can be easily and surely diagnosed by this technique.

IRB # - 08223

**The Role of Facial Paralysis in Staging of Temporal Bone
and External Auditory Canal Squamous Cell Carcinoma:
A Pooled-data Comparative Survival Analysis**

Thomas S. Higgins, MD; Stephanie A. Moody-Antonio, MD

Objectives: The appropriate designation for facial paralysis in the Pittsburgh staging system for temporal bone and external auditory canal squamous cell carcinoma (TB/EAC SCC) is unclear. The objective of this study was to conduct a survival analysis of published data to compare the survival outcomes of patients presenting with facial paralysis to T-staging in TB/EAC SCC.

Data Sources: A search of MEDLINE (1966 to 2009), EMBASE (1980 to 2009), CENTRAL, and Cochrane databases was supplemented by references in retrieved articles.

Study Selection: The authors independently screened articles for final inclusion.

Data Extraction: Data extracted included study criteria appraisal, demographics, type and stage of cancer, survival data, and presenting facial function.

Data Synthesis: Of the 243 citations reviewed, the search process yielded 21 case series (evaluating 341 patients with TB/EAC SCC) meeting criteria for final inclusion in the analysis. Cox regression survival analyses were performed to compare survival of patients with facial paralysis and T-staging. Using all staging systems, the overall survival for patients with facial paralysis on presentation was significantly worse than stage T3-designated cancers [OR=0.369, 95% CI 0.197-0.689, P=0.002] and demonstrated no difference from stage T4-designated cancers [OR=0.917, 95% CI 0.537-1.567, P=0.752]. Stratified analyses using each staging system demonstrate facial paralysis predictive of survival equivalent to T4 designation.

Conclusions: This pooled-data analysis demonstrates that the survival outcomes of patients with TB/EAC SCC presenting with facial paralysis more closely parallel the survival curves of stage T4 and that facial paralysis would better be classified as stage T4, instead of T3.

Define Professional Practice Gap & Educational Need: The published reports on temporal bone cancer have varied in the staging system used. Depending on the staging system, facial paralysis may be considered stage T2, T3, or T4.

Learning Objective: This study aims to establish the role of facial paralysis in staging temporal bone cancer by evaluating the relative survival outcomes.

Desired Result: The desired application of the results of this study is for researchers to use the modified Pittsburgh staging system with facial paralysis as a T4 designation in temporal bone cancer. This will allow better consistency in the literature and allow more accurate comparison of results.

Migraine-Associated Dizziness: Neuro-Optometric Findings and Treatment

Ian M. Sambur, MD; Sujana S. Chandrasekhar, MD
Neera Kapoor, OD, MS

Study Design: This is a follow-up study of a retrospective review of 19 patients with complex dizziness exacerbated by visual stimuli presented at the ANS meeting in 2008. It was performed to further delineate criteria for assessing visual-vestibular disorder as a subset of migraine-associated dizziness (MAD) and to evaluate the utility of vision rehabilitation therapy in ameliorating the patient's symptoms.

Methods: Nineteen patients were identified by a single neurotologist over a 10 month period as meeting the criteria for visually related vestibulopathy. Thorough neuro-otologic and neuro-optometric evaluations were performed. Most of these patients met the criteria for MAD; a subset was also seen by a neurologist. Interventions included spectacle correction, vision therapy, vestibular rehabilitation, and migraine medications. Follow up ranged from 16 to 28 months. Symptomatic improvement was measured as a percentage of the pre-morbid functionality and objective improvement in sensorimotor vision function.

Results: 78% of patients with visual-vestibular disorder as a subset of MAD, who were treated with visual rehabilitation, being previously incapacitated by their dizziness, achieved significant improvement in symptoms, quality of life and optometric measurements. 67% of patients receiving spectacle change reported significant symptomatic improvement.

Conclusions: Neuro-optometric evaluation and rehabilitation is a useful addition to the treatment portfolio for the complex dizzy patient with definite or probable MAD. Collaboration with a trained neuro-optometrist is beneficial in optimizing treatment outcomes.

Define Professional Practice Gap & Educational Need: There currently exists a lack of awareness regarding the recognition of the vertiginous patient with a prominent visual component in the exacerbation of their symptoms. Such a patient usually falls in the category of Migraine-Associated Dizziness (MAD), and frequently will benefit from vision rehabilitation therapy, performed under the supervision of a trained neuro-optometrist.

Learning Objective: At the conclusion of this presentation, the participants should be able to recognize the vertiginous patient with a prominent visual component in the exacerbation of their symptoms. Such a patient usually falls in the category of Migraine-Associated Dizziness (MAD), and frequently will benefit from vision rehabilitation therapy, performed under the supervision of a trained neuro-optometrist.

Desired Result: To review the presence of visual vertigo in MAD, and the utility of applying neuro-optometric vision therapy to existing neuro-otological treatment protocols.

IRB # - Approval provided by Mount Sinai School of Medicine

Late Failure Rate of Hearing Preservation After Middle Fossa Approach for Resection of Vestibular Schwannoma

Christopher W. Hilton, MD; Stephen J. Haines, MD
Samuel C. Levine, MD

Objective: To explore the relationship between long term hearing results after a middle fossa approach for resection of Vestibular Schwannoma (VS) and recurrence of tumor.

Study design: Retrospective case review

Setting: Tertiary referral center

Patients: All patients undergoing a middle fossa approach for resection of VS at a single institution with intent to preserve hearing between August 1989 and February 2009 were included in the study population.

Interventions: Standard middle fossa approach for resection of VS, Magnetic resonance imaging (MRI), audiogram.

Main outcome measures: Recurrence of tumor as evaluated by MRI, and hearing results as measured by serial yearly audiogram.

Results: 84 patients were identified who met study criteria. 51/84 (61%) had usable hearing (AAO-HNS class A or B) post-operatively. 3/51 (6%) showed late degradation in hearing result (progression to class D) over a mean follow up of 34 months. Mean time to loss of hearing was 42 months. Of the three patients with late degradation in hearing, two were found to have recurrences of their original tumor on MRI.

Conclusions: Late degradation of hearing was a rare occurrence after initially successful hearing preservation over the studied time period. When hearing degradation did occur, there appeared to be a correlation with tumor recurrence.

Define Professional Practice Gap & Educational Need: There is a lack of understanding as to how hearing results fare over time after initially successful hearing preservation with a middle fossa approach for resection of Vestibular Schwannoma.

Learning Objective: The objective of this talk is to explore long term hearing results after initially successful hearing preservation surgery, and to explore the way that tumor recurrence influences those results.

Desired Result: The desired result is that with an understanding of how hearing preservation results fare over time clinicians will be able to better counsel their patients regarding treatment options for this disease.

IRB # - University of Minnesota IRB#: 0811M54102

Cochlear Implantation in the Octo- and Nonagenarian

Matthew L. Carlson, MD; Joseph T. Breen, MD; René H. Gifford, PhD
Annamary Peterson, AuD, MA; Brian A. Neff, MD
Charles W. Beatty, MD; Colin L. W. Driscoll, MD

Objective: Previous studies have shown that cochlear implant (CI) outcomes with respect to surgical morbidity and speech perception may be poorer in elderly patients as compared to younger adults. However, recent anecdotal reports suggest that elderly CI recipients are achieving increasingly higher speech perception performance and fewer surgical complications than previously noted. Our objective is to review CI outcomes using newer generation implants and soft surgical cochleostomy techniques in patients 80 years and older compared to younger adult recipients.

Study design: Retrospective chart review

Setting: Tertiary referral center

Patients: All adult patients (n=264) who underwent implantation with a Nucleus 24RE, Advanced Bionics HR90k, or Med El Sonata device at a tertiary academic institution.

Intervention(s): Postoperative speech perception scores and clinical data extraction using the electronic medical record.

Main outcome measure(s): Surgical time, perioperative and late complication rates, length of hospital stay, and postoperative speech perception scores were collected on patients > 79 years of age (n=50) and those between 18 and 79 years (n=214) for comparison.

Results: Patients >79 years were more likely to have perioperative cardiac arrhythmias and cerebrovascular events, postoperative delirium and require hospital admission (p <0.05). There was no statistical difference between groups with respect to perioperative or long-term dizziness/imbalance and flap complications. Preliminary speech perception analysis revealed similar outcomes for older and younger patients.

Conclusions: Given the high levels of speech perception achieved by older patients, routine implantation of octo- and nonagenarians seems warranted. These results also stress the need for thorough preoperative evaluation of elderly patients given the increased risk for cardiac and cerebrovascular complications.

Define Professional Practice Gap & Educational Need: There currently exists a lack of contemporary knowledge concerning the safety profile and efficacy of cochlear implantation using newer generation implant technologies and surgical techniques in patients eighty years or greater. This understanding is becoming more important with increasing life expectancies and ever broadening implant criteria.

Learning Objective: The learning objective of this talk is to describe cochlear implant outcomes with respect to surgical morbidity and speech perception using newer generation implants and soft surgical cochleostomy techniques in patients 80 years and older compared to younger adult recipients.

Desired Result: The desired result is that based on a more complete understanding of the efficacy and safety profile of cochlear implantation using newer technologies in patients of advanced years, clinicians will apply this information to make informed decisions when determining cochlear implant candidacy and be able to convey accurate expectations and risks of surgery during preoperative discussions with elderly patients undergoing implantation.

IRB # - 09-006785

Temporal Bone Resection for External Auditory Canal Malignancies, Prognostic Factors and Clinical Outcome

Adrien A. Eshraghi MD; Robert Guerring BS; Fred F. Telischi MD
Don Weed MD; Franc Civantos; Craig Buchman MD
Zouka Sargi MD; Morteza Gadiali MD

Objective: To evaluate the outcome of tumors of the external auditory canal (EAC) treated with temporal bone resection (TBR).

Setting: Retrospective chart review of all patients who had TBR for EAC tumors.

Patients: 40 patients with carcinoma of the EAC who underwent TBR were identified.

Main outcome measures: Outcomes were analyzed by means of survival analysis, with respect to Pittsburgh staging system, the status of frozen and final surgical margins, and adjuvant XRT.

Results: At 24 months, 33% developed a recurrence: 0% with Stage I tumor, 29% with Stage II, 40% with Stage III, and 50% with Stage IV. For SCC, mean DFS for Stages I and II combined is 90 +/- 11 months and for Stages 3 and 4 combined is 72 +/- 25 months. The mean OS and DFS are 141 +/- 7 and 102 +/- 12 months respectively. The mean DFS for patients with negative and positive final surgical margins are 104 +/- 9 and 64 +/- 20 months respectively. At 24 months, 64% of patients with positive final surgical margins who underwent radiotherapy have recurrence. However, only 18% of patients with negative final surgical margins who received postoperative radiotherapy recurred.

Conclusions: The outcome of treatment for EAC carcinoma is correlated with positive final surgical margins, however positive frozen sections margins do not affect recurrence or survival. Recurrence is correlated with Pittsburgh stage for SCC of the EAC. For patients with advanced disease (stage III and IV or positive margins), radiotherapy does not ensure a positive prognosis.

Define Professional Practice Gap & Educational Need: Management of tumors of the external auditory canal may require extensive surgery including temporal bone resection and parotidectomy. The prognostic factors and clinical outcome after this type of surgery need to be defined.

Learning Objective: To identify prognostic factors and survival rate in patients undergoing temporal bone resection for external auditory canal malignancies.

Desired Result: Clinicians will apply this information to make informed decisions involving other team member such as otologist or head and neck surgeon when needed. They were be able to discuss the prognosis of patients with external auditory canal carcinoma after temporal bone resection.

Comprehensive Diagnostic Battery For Evaluating Sensorineural Hearing Loss In Children

Jerry W. Lin, MD, PhD; Naweed Chowdhury, BA
Avni Mody; Ross Tonini, AuD; Claudia Emery, AuD
Joanne Haymond, AuD; John S. Oghalai, MD

Objective: Selection of diagnostic tests for children with sensorineural hearing loss (SNHL) is influenced by clinical suspicion. Results reported in the literature are similarly biased. We evaluate the utility of a comprehensive diagnostic battery ordered for each child.

Study Design: Retrospective review.

Setting: Tertiary-care university hospital.

Patients: 268 children referred for cochlear implant evaluation between 1/2005 and 6/2009.

Interventions: none

Main outcome measures: Diagnostic testing: magnetic resonance imaging (MRI), computed tomography (CT), renal ultrasound (US), electrocardiogram (ECG), fluorescent treponemal antibody absorption test (FTA-ABS), and connexin 26 sequencing. Consultation reports: genetic and ophthalmologic evaluations

Results: Barring specialty consultations, which were added to the test battery later, 90% of patients underwent both MRI and CT, and 80% of patients underwent all diagnostic testing. MRI revealed abnormalities explaining SNHL in 23% of patients. CT showed inner ear anomalies in 17% of patients. Renal US found anomalies in 10% of patients, but none led to diagnoses of syndromes associated with SNHL. ECG found 2% of patients with prolonged QT intervals; two patients had Jervell-Lange Nielsen syndrome. FTS-ABS was positive in 0.5%. Biallelic connexin 26 mutations were found in 15%. Genetic consultation diagnosed a hearing loss syndrome in 25%. Ophthalmologic consultation found abnormalities associated with hearing loss in 8%.

Conclusion: As a single diagnostic test, imaging continues to provide the highest yield for evaluating children with SNHL. Genetic consultation with genetic testing is also useful. Other diagnostic tests, although of low diagnostic yield for SNHL, can identify important diseases that significantly impact patient health.

Define Professional Practice Gap & Educational Need: There is no consensus regarding the battery of diagnostic tests that should be ordered for a child presenting with severe to profound hearing loss. Several studies exist in the literature regarding recommendations, but not every patient underwent the full battery of tests to allow for direct comparisons of diagnostic yields between tests.

Learning Objective: This study describes a large group of children who underwent all or most of the diagnostic tests in a comprehensive battery and direct compares the diagnostic yields of each of these tests.

Desired Result: Attendees will have a better understanding of which diagnostic tests will be most useful in diagnosing the cause of severe to profound hearing loss in their pediatric patients.

IRB or IACUC Approval: Yes

Curve-Fitting Vestibular Schwannoma Volume Data to Predict Future Growth

Charles A Mangham, Jr., MD; Uresh Patel, MD

Objective: To test the hypothesis that tumor volume growth follows an S-shaped Gompertz function and that future growth can be predicted using a curve-fitting strategy.

Study Design: Retrospective chart review.

Patients: 70 consecutive patients with the MRI diagnosis of untreated vestibular schwannoma between 2002 and 2007 who had three or more MRI images available to evaluate.

Setting: Subspecialty private practice.

Intervention: MRI volume measurement of 1) patient tumors and 2) targets of known volume, using a manual segmentation technique and curve-fitting using Microsoft Excel.

Main outcome measures: Linear and exponential curves were fit to the first two tumor volume data points and projected in time to the third data point. The process was repeated if four or more MRIs were available. The curve with the best fit was used to calculate volume doubling time.

Results: 70% of tumors displayed positive growth. A linear curve was the best fit in 90% of cases. Curve fitting predicted future tumor volume with an error range of 0.03% to 93% with a median of 14%. Gadolinium enhancement and thick MRI slices systematically overestimated true volume.

Conclusions: An accurate initial measurement of tumor volume was the most important factor in accurately predicting future growth, important enough to justify rescanning when needed. Fitting the first two MRI volume results with a linear curve satisfactorily predicted short-term future growth. This information will help clinicians incorporate patient preferences when discussing treatment alternatives as well as monitor the results of treatment (or observation).

Define Professional Practice Gap & Educational Need:

- 1) Lack of awareness of how basic tumor biological principles can be applied to monitoring growth of vestibular schwannomas
- 2) Lack of awareness of how various MRI techniques affect measurement of tumor size

Learning Objective:

- 1) Learn how to use commonly available computer software to predict short-term tumor growth
- 2) Learn appropriate MRI techniques for following tumor growth

Desired Result:

- 1) Attendees will assess tumor growth, then include patient preferences when making treatment decisions.
- 2) Attendees will learn how interaction with neuroradiologists can improve the assessment of tumor growth.

IRB # - Approved by Chairman of the IRB

Meta-Analysis of Facial Nerve Function after Microsurgery vs. Gamma Knife Radiosurgery for Vestibular Schwannomas

Majestic Tam, MD; Kestutis P. Boyev, MD; Kiran K. Turaga, MD

Objective: There is significant controversy between microsurgery and Gamma Knife radiosurgery (GKR) for the treatment of vestibular schwannomas with regards to facial nerve function (FNF). We pooled published studies to study outcomes of FNF between the two modalities.

Data Sources: We searched the Medline database from 1990-2009 for the following MESH headings and keywords: Vestibular Schwannoma, Acoustic Neuroma, surgery and Gamma Knife.

Study Selection: Studies which compared microsurgery and GKR that utilized the House Brackmann scale (HBS) for FNF were included.

Data Extraction: Of 2166 abstracts identified, seven studies were initially abstracted. One study was excluded because it did not utilize the HBS. A quality assessment scale was applied to each study. We classified HBS I and II as good outcome and HB III – VI as poor outcome. Random effects modeling was applied to the extracted data. Sensitivity analysis after excluding studies with significant heterogeneity was performed.

Data Synthesis: We included 740 patients, 423 undergoing GKR and 317 undergoing microsurgery from 1995-2009. Of patients undergoing GKR, 94% had good FNF, while in patients undergoing surgery 75% had good FNF ($p=0.001$). Pooled Odds Ratio (OR) for Good FNF favoring GKR were 7.2 (95% CI 4.2-12.4, $p<0.001$) but had significant heterogeneity ($p=0.07$). After excluding a heterogeneous study, the pooled OR was 4.1 (95% CI 2.2-7.4 $p<0.001$) still favoring GKR.

Conclusions: Currently there exists no level 1 evidence comparing microsurgery to GKR with regards to FNF and there is significant heterogeneity in current studies. Based on our pooled analysis, GKR was favored over microsurgery in preservation of FNF.

Define Professional Practice Gap & Educational Need: 1) Inconsistencies in selection of microsurgery vs. gamma knife radiosurgery in treatment of vestibular schwannomas
2) Lack of consensus standard treatment

Learning Objective: The learning objective of this talk is to formulate an objective, outcomes based standard of treatment for vestibular schwannomas.

Desired Result: 1) Improve knowledge of treatment outcomes of microsurgery and stereotactic radiosurgery in vestibular schwannomas
2) Evidence based selection of optimal and standard treatment for vestibular schwannomas

IRB# - The proposed research project did not require oversight from an Institutional Review Board because, the project does not meet the definition of human subjects research as defined by the Federal Regulations at 45 CFR Part 46.102. Although the project was conducted for research purposes, all data gathered was collected from publicly available sources and does not contain any identifiable private information.

Post Blunt Head Trauma Hearing Loss in the Context of an Adult Cochlear Implant Program

Simon Greenberg, MBBS; David Shipp, MA; Vincent Lin, MD
Joseph Chen, MD; Julian M. Nedzelski, MD

Objective: To highlight issues that impact on cochlear implant candidacy and performance in individuals who have severe bilateral sensorineural hearing loss following major blunt head trauma.

Study design: Retrospective case series

Setting: Tertiary level neurotology centre

Patients: Seventeen patients treated at our institution for severe hearing loss following blunt head trauma

Intervention: Assessment of cochlear implant candidacy criteria and post implant outcomes in patients presenting with severe hearing loss following major blunt head trauma

Main outcome measures: Etiology of injury, pure tone audiology, open set speech perception (pre and post implant), promontory stimulation and imaging results.

Results: Of seventeen patients with severe hearing loss, eleven ultimately underwent cochlear implantation. Fall from a height was the most common etiology of injury. Six patients suffered bilateral temporal bone fractures. Sixteen of the patients had no open set speech recognition on pre implant testing. For those patients who did undergo cochlear implantation, the mean post implant open set speech recognition score was 69. Issues related to cochlear implant candidacy and outcomes for implantation will be discussed.

Conclusions: Severe hearing loss following major blunt head trauma presents a number of unique clinical challenges. A significant number of these patients are not suitable for cochlear implantation for a variety of reasons. Clinicians working in the area of cochlear implantation should be aware of the various problems that can impact on effective cochlear implantation in such patients.

Define Professional Practice Gap & Educational Need: Lack of awareness of specific issues that impact on cochlear implant candidacy when patients suffer severe hearing loss as a result of significant head trauma.

Learning Objective: To present a large case series of our experience in this area, helping to demonstrate many of the issues that result from head trauma precipitated severe sensorineural hearing loss.

Desired Result: Better understanding of the approach to patients with severe hearing loss requiring cochlear implantation as a result of blunt head trauma.

IRB # - n/a

Estrogen Receptor Expression in Vestibular Schwannomas

Carrie R. Brown, MD; Zana Ahmad, BS; Andrew K. Patel, MD
Weg Ongkeko, MD, PhD; Allen F. Ryan, PhD
Joni K. Doherty, MD, PhD

Hypothesis: Estrogen receptor (ER) is upregulated in some sporadic and Neurofibromatosis 2 (NF2)-associated vestibular schwannomas (VS), and represents one of the cellular pathways responsible for tumorigenesis. VS that exhibit upregulation of ER will have decreased cellular proliferation when treated with the selective estrogen receptor modulator, Tamoxifen.

Background: Sporadic and NF2-related VS arise from mutations in the NF2 gene, encoding merlin. VS have a female predominance of 2:1. Larger and more vascular tumors in women, as well as increased growth rates during pregnancy, have been reported in the literature. However, prior studies on ER and progesterone receptor (PR) expression have been widely varied and conflicting. Most studies have low numbers, and utilize immunohistochemistry, a notoriously difficult and qualitative method. We have previously investigated ER and PR expression in VS at the mRNA level, and found upregulation of ER in approximately 50% of sporadic and 25% of NF2-associated VS.

Methods: We investigated protein expression levels of ER and PR in 20 VS specimens with Western blot technique. Primary cultures of VS exhibiting ER upregulation were then treated with Tamoxifen, alone and in combination with the dual EGFR and ErbB2 inhibitor, Lapatinib. Changes in cellular proliferation were assessed with the MTS assay.

Results: We found upregulation and activation of ER in some VS, and no upregulation of PR.

Conclusion: ER is upregulated in a minority of sporadic VS, is involved in tumorigenesis in these specimens, and may be an effective therapeutic target in combination therapy.

Define Professional Practice Gap & Educational Need: Conflicting results on estrogen receptor and progesterone receptor expression levels in vestibular schwannomas exists in the literature.

Learning Objective: The learning objective of this talk is to convey estrogen receptor and progesterone receptor expression levels in primary vestibular schwannoma samples.

Desired Result: The desired result is for clinicians to have a better understanding of the cellular pathways responsible for vestibular schwannoma tumorigenesis, and to further elucidate which cellular pathways are pharmacologic targets.

IRB # - This research was supported by NIH/NIDCD 5 K08 008523 and NIH/NIDCD T32 DC000028.

Assessment of Electrode Placement and Audiological Outcomes in Bilateral Cochlear Implantation

Georges B. Wanna, MD; Jack H. Noble; Mary S. Dietrich, PhD
Alejandro Rivas, MD; Benoit Dawant, PhD; Robert F. Labadie, MD, PhD

Hypothesis: The goal of this study is to use non-rigid registration to locate the position of cochlear implant (CI) electrodes in relationship to the basilar membrane in bilaterally implanted adults and correlate such with audiological performance.

Background: Several groups have reported correlations between intracochlear electrode position—specifically depth and scala location—and audiological outcomes in CI. We have developed semi-automated, non-rigid registration algorithms and successfully validated its predictive accuracy on cadaver models.

Methods: After obtaining IRB approval, 19 bilateral CI patients who had pre and post-intervention CT scans were recruited. Using an intracochlear atlas built from 7 cadaveric specimens which were microCT scanned, semi-automated non-rigid registration was used to determine the position of the basilar membrane in relationship to the CI electrode in-vivo. Audiological performance was assessed using HINT and CNC testing, and length of auditory deprivation was recorded.

Results: Controlling for the known contributing variable of length of auditory deprivation and using a linear modeling approach to account for the impact of type of surgery (sequential versus simultaneous), initial results from analysis of 3 of the 19 patients show that location of CI electrode in relationship to the scala is not predictive of audiological performance.

Conclusions: Recent excitement regarding the correlation of CI electrode location and audiological performance will need to be confirmed over hundreds of patients. We present a semi-automated, highly-accurate technique that can accomplish this. Our preliminary data contradicts prior reports showing that scala tympani insertion portends better audiological outcome.

Define Professional Practice Gap & Educational Need: Location of the Cochlear Implant in relation to scala tympani and vestibuli might be a major factor in determining the audiologic outcome in these patients. Previous reports have correlated preferential scala tympani electrode insertion with improved audiological outcomes in CI recipients. These labor-intensive studies used rigid registration to identify intracochlear electrode position, which may be less precise than non-rigid algorithms. We sought to apply non-rigid registration via an automated process.

Learning Objective: The goal of this study is to use non-rigid registration to locate the position of cochlear implant (CI) electrodes in relationship to the basilar membrane in bilaterally implanted adults and correlate such with audiological performance.

Desired Result: We present a semi-automated, highly-accurate technique that can accomplish this. Our preliminary data contradicts prior reports showing that scala tympani insertion portends better audiological outcome.

Stereotactic Radiosurgery for Vestibular Schwannomas: A Survey of Current Practice Patterns of Neurotologists

Michael A. German, MD; Mehdi Sina-Khadiv, BA
Hamid R. Djalilian, MD

Objective: To evaluate the use of stereotactic radiation by neurotologists practicing in the United States as a treatment modality for vestibular schwannomas (VS).

Data sources: We sent an anonymous, 17 question survey to 302 members of the American Neurotology Society to assess the number of VS seen, the frequency with which radiation is used as a treatment modality, radiation training and experience, the impact of tumor size and patient age on treatment decisions, and radiation dose ultimately used by neurotologists.

Study selection: Neurotologists were selected based on current membership in the American Neurotology Society.

Data extraction: Data were extracted anonymously from the surveys and tabulated in a spreadsheet.

Data synthesis: Responses were received from 43% (129/302) of surveyed neurotologists; of the respondents, 42% (54/129) perform stereotactic radiation. 56% (30/54) use GammaKnife, while 30% (16/54) use Cyberknife. Respondents indicated that over 35% of VS patients received radiation (median 30%, SD 26%). The upper limit of tumor size showed variation with 23% (22/96) setting it at 2 cm, 32% (31/96) at 2.5 cm, and 31% (30/96) at 3 cm. Neurotologists indicated that they do not perform radiation on patients below 18 years of age.

Conclusions: Conclusion: Neurotologists are increasingly choosing to treat VS with stereotactic radiosurgery (SRS) or fractionated stereotactic radiation. There appears to be discordance in tumor size limits, age limits, and other considerations with regard to treatment decision-making. From the results of our survey, we conclude that neurotologists do not act according to clear consensus when treating VS with radiation.

Define Professional Practice Gap & Educational Need: The practice patterns of neurotologists using stereotactic radiosurgery (SRS) to treat vestibular schwannomas is not well defined. There exists controversy in patient selection, radiation dose used, and tumor size limits, among others.

Learning Objective: The learning objective of this talk is to report on the demographics of neurotologists using stereotactic radiosurgery to treat vestibular schwannomas, what their level of experience and treatment preferences are, and associations between these factors.

Desired Result: The desired result is to create a better awareness of what attendees' colleagues are doing in their practice and hopefully further the dialogue surrounding treatment controversy as it pertains to vestibular schwannomas and stereotactic radiosurgery.

IRB # - HS# 2008-6516 approved by UC Irvine Office of Research Administration

Neuroendocrine Adenoma of Middle Ear: Staging, Treatment and Outcomes

Ilka C. Naumann, MD; Eric W. Sargent, MD
Jack M. Kartush, MD; Noah A. Stern, DO
Michael J. LaRouere, MD; Dennis I. Bojrab, MD

Objective: Neuroendocrine adenoma of the middle ear (NAME)/carcinoid is rare, difficult to diagnose preoperatively, and distinct from papillary tumors of endolymphatic sac origin. Single case reports and small series predominate in the literature. While recurrences are common and metastasis to local lymph nodes have been reported in a few cases, the entity is typically treated with gross total resection with the expectation of a benign outcome. A series of 9 patients treated over the last decade is reviewed to make recommendations for staging, treatment and post-operative monitoring.

Study Design: Retrospective case review

Setting: Tertiary referral center

Patients: 9 patients with NAME/carcinoid tumors (age 31-63, 3 male; 6 female) treated from 2000 - 2009 with surgical resection.

Main Outcome Measures: Clinical presentation, pre- and post-operative hearing and facial nerve function, extent of tumor on CT/MRI, histopathology, and characteristics of recurrence.

Results: In this large, single-institution review of this entity, 7 of 9 patients had a generally unremarkable course following resection. While none have developed metastases, 2 had ipsilateral transient episodes of facial nerve paralysis and inflammatory symptoms (pain) before diagnosis and had a pattern of multiple recurrences eventually resulting in petrosectomy and facial nerve grafting; both had focal or diffuse enhancement of the facial nerve and middle fossa dura on MRI.

Conclusions: 1) long-term serial imaging is required to monitor these patients; 2) Pre-operative facial nerve (paresis/transient paralysis) and/or inflammatory symptoms portend a poor hearing and facial nerve outcome. We suggest that the rare patient presenting with these symptoms may be candidate for either conventional external beam or Gamma Knife irradiation.

Define Professional Practice Gap & Educational Need: Due to the rarity of middle ear adenoma no treatment guidelines exist. Drawing conclusions from mostly single case reports that predominate the literature are difficult.

Learning Objective: Common clinical features are presented which include presenting symptoms, radiological data, pathologic patterns, treatment plans and outcome data are described.

Desired Result: Goal of the presentation is to give a comprehensive review of this rare tumor and to give guidelines for practicing physicians in managing patients with carcinoid tumors of the middle ear.

IRB # - IRB approval from Providence/St John Hospital

Intraoperative Neural Response Telemetry (tNrt) as Predictor of Performance

Maura K. Cosetti, MD; Stacey H. Gunn, MS; William H. Shapiro, AuD
Janet E. Green, AuD; Benjamin R. Roman, MD; Anil K. Lalwani, MD
J. Thomas Roland, Jr, MD; Susan B. Waltzman, PhD

Objective: To determine whether intra-operative neural response telemetry (tNRT) is predictive of post-operative speech perception.

Study design: Retrospective review

Setting: Tertiary referral center

Patients: Children (n=42) between the ages of 5-17 years and adults 18 years and older (n=143) with severe-to-profound hearing loss and implanted with the Nucleus Freedom device between 2005 and 2008 and followed at least 1 year were included.

Intervention: Intra-operative neural response telemetry following insertion of the electrode array.

Main outcome measure(s): Measures included: 1) intra-operative NRT measurements; 2) pre- and 1 year post-operative open set word recognition scores using age appropriate open-set tests for children and adults. tNRT levels for electrodes E20, 15, 10 and 5 in each subject were correlated to performance at the one-year evaluation interval.

Results: No correlation existed between tNRT responses and open set speech performance at the one year evaluation. Several subjects had absent tNRT in the OR but developed speech recognition abilities while the remaining subjects had intraoperative responses with levels of postoperative performance ranging from 0-100%.

Conclusions: This study suggests that there is no significant correlation between intra-operative tNRT and speech perception performance at one year. At the time of surgery, tNRT provides valuable information regarding the electrical output of the implant, the response of the auditory system to electrical stimulation and preliminary device programming data; however, it is not a valuable predictor of post-operative performance. Furthermore, the absence of tNRT does not necessarily indicate a lack of stimulation.

Define Professional Practice Gap & Educational Need: Although widely used to obtain intraoperative measurements of device function during cochlear implantation, it is unknown if tNRT is a useful predictor post-operative speech performance. Further, the implication of absence of tNRT is unknown.

Learning Objective: To determine whether intra-operative neural response telemetry (tNRT) is predictive of post-operative speech perception.

Desired Result: Surgeons and audiologists will incorporate this information into their clinical practice recognizing the role of tNRT intra-operatively and for preliminary post-operative device programming. Lack of intra-operative tNRT should not unduly worry the surgeon, patient or family and should not be used as a reliable predictor of post-operative performance.

IRB # - NYU #11281

A New Software Tool to Optimize Frequency Map Selection for Cochlear Implants

Daniel Jethanamest, MD; Chin-Tuan Tan, PhD
Matthew B. Fitzgerald, PhD; Mario A. Svirsky, PhD

Hypothesis: When cochlear implant (CI) users are allowed to self-select the "most intelligible" frequency-to-electrode table, some of them choose one that differs from the default frequency table that is normally used in clinical practice.

Background: Cochlear implants (CI) reproduce the tonotopicity of normal cochleas using frequency-to-electrode tables that assign stimulation of more basal electrodes to higher frequencies and more apical electrodes to lower frequency sounds. Current audiological practice uses a default frequency-to-electrode table for most patients. However, individual differences in cochlear size, neural survival and electrode positioning may result in different tables sounding most intelligible to different patients. No clinical tools currently exist to facilitate this fitting.

Methods: A software tool was designed that enables CI users to self-select a "most intelligible" frequency table. Users explore a two dimensional space that represents a range of different frequency maps. Unlike existing tools, this software enables users to interactively audition speech processed by different maps, and quickly identify a preferred map. Pilot testing was performed in eight long-term, postlingually deaf CI users.

Results: The software tool was designed, developed, tested and debugged. Patients successfully utilized the tool to sample frequency tables and to self-select tables deemed most intelligible, which for over half of the users differed from the clinical default.

Conclusion: A software tool allowing CI users to self-select frequency-to-electrode maps may help in fitting postlingually deaf users. This novel approach may transform current audiological practices and methods of CI fitting.

Define Professional Practice Gap & Educational Need: Current cochlear implant users receive a standard frequency-to-electrode map and no clinical tools currently exist to facilitate fitting of customized frequency maps. There is a lack of awareness of how frequency mismatches due to variable cochlear size, neural survival and electrode positioning may be partially addressed by individualized frequency maps.

Learning Objective: The learning objective of this talk is to address the potential benefits of a customized frequency-to-electrode map in cochlear implant users, and describe a newly developed software tool for fitting these maps. A review of pilot testing in CI users will also be presented.

Desired Result: The desired result is to provide greater recognition to the potential benefits of customized frequency maps in CI users and demonstrate a new clinical tool that facilitates the fitting process for these patients. Clinicians and researchers will be informed about the possible advantages of this approach that may change the standard of audiological fitting after CI.

IRB # - Acknowledgement: NIH, grant R01-DC03937; PI: M. Svirsky
IRB or IACUC Approval: IRB 05-265

Fundal Fluid as a Predictor of Hearing Preservation in Middle Fossa Craniotomy for Vestibular Schwannoma

John C. Goddard, MD; Rick A. Friedman, MD, PhD

Objectives: To determine whether the presence and specific characteristics of cerebrospinal fluid in the fundal region of the internal auditory canal on pre-operative MRI scans is predictive of hearing preservation following middle fossa craniotomy for vestibular schwannoma.

Study Design: Retrospective review.

Setting: Private practice tertiary referral center.

Patients: All patients undergoing middle fossa craniotomy for unilateral vestibular schwannoma between 2006 and 2009 with pre-operative MRI and pre- and post-operative audiologic data.

Interventions: Middle fossa craniotomy for removal of vestibular schwannoma.

Main outcome Measures: Presence or absence, location, and quantity of fundal fluid as seen on T2-weighted pre-operative MRI will be assessed. These fundal fluid characteristics will be examined in relation to hearing outcomes based on pre- and post-operative pure tone averages (0.5, 1, 2, and 3 kHz) and speech discrimination scores as well as the AAO-HNS guidelines for the evaluation of hearing preservation in vestibular schwannoma.

Results: Hearing was preserved (AAO-HNS Class A, B or C) in 66.7% of patients demonstrating the presence of fundal fluid on pre-operative MRI (90% with Class A or B). Fundal fluid was present in the lower half of the internal auditory canal in 88% of patients who achieved hearing preservation while 61% of patients with hearing preservation had >2mm (transverse dimension) of fluid between the tumor surface and the fundus. Among patients without evidence of fundal fluid on pre-operative MRI, 44.7% had hearing preservation (all Class A or B).

Conclusions: The presence of fundal fluid on pre-operative MRI is predictive of higher hearing preservation rates in patients undergoing middle fossa craniotomy for vestibular schwannoma.

Define Professional Practice Gap & Educational Need: There is a lack of knowledge regarding an objective method of pre-operatively predicting hearing preservation rates among patients undergoing middle fossa craniotomy for vestibular schwannoma surgery.

Learning Objective: To determine if fundal fluid characteristics as seen on pre-operative MRI scans can be predictive of hearing outcomes following middle fossa craniotomy for vestibular schwannoma.

Desired Result: Examination of fundal fluid characteristics on pre-operative MRI scans will aid in counseling patients regarding hearing preservation rates prior to middle fossa craniotomy for vestibular schwannoma.

IRB#: Approval: 09-020