SELECTED ABSTRACTS

IN ORDER OF PRESENTATION

POSTER PRESENTATIONS

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Predictive Value of EABRs in NF2 Adults and Non-NF2 Children Receiving ABIs

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Objective: To compare the predictive value of intraoperative electrically evoked auditory brainstem response (EABR) between NF2 adult ABI recipients and non-NF2 pediatric ABI recipients.

Study design: Retrospective case series.

Setting: Single tertiary academic referral center.

Patients: All ABI recipients from 1994 to 2015, which included 34 NF2 adults and 10 non-NF2 children.

Intervention(s): EABR recordings during ABI placement.

Main outcome measure(s): The morphologies of the intraoperative EABRs were evaluated for the number of waveforms showing a response, the number of positive peaks in those responses, and the latencies of each of these peaks.
Results: 27/34 adult NF2 patients and 9/10 children had EABR waveforms. 20/27 (74.0%) of the adult patients and all of the children had ABI devices that stimulated post-operatively. When comparing the waveforms between adults who stimulated and those who did not stimulate, the proportion of total number of intraoperative EABR peaks to total possible peaks was significantly higher for the adults who stimulated than for those who did not ($p<0.05$). Children had a significantly higher proportion of total number of peaks to total possible peaks when compared to adults who stimulated ($p<0.02$). Additionally, there were more likely to be EABR responses at the initial stimulation than intraoperatively in the pediatric ABI population ($p=0.065$)

Conclusions: This study shows that a higher number of total peaks seen on intraoperative EABRs may indicate a higher likelihood of eventual device stimulation, although the predictive value of intraoperative EABR for speech perception outcome requires further study.

Define Professional Practice Gap & Educational Need: 1. Unclear utility of intraoperative EABRs during ABI placement and whether they may be predictive of clinical outcome. 2. Lack of contemporary knowledge about EABRs in children receiving ABIs

Learning Objective: To compare the predictive value of intraoperative electrically evoked auditory brainstem response (EABR) between NF2 adult ABI recipients and non-NF2 pediatric ABI recipients.

Desired Result: Attendees will try to obtain the most number of peaks on intraoperative EABRs when placing the ABI device. They will further investigate whether EABRs are predictive of clinical outcome.

Indicate IRB or IACUC Approval: Approved

The Oncomir Mir-21 Facilitates AKT Pathway Activation in Vestibular Schwannomas and Meningiomas

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Abraham Jacob, MD

Hypothesis: Overexpression of oncomir miR-21 in vestibular schwannomas (VS) represses tumor suppressor genes involved in the inhibition of the PI3K/AKT pathway.

Background: MicroRNAs regulate gene expression at the post-transcriptional level by binding to the 3â€™-UTR of their targets. The oncomir (oncogene) miR-21 is up-regulated in different types of cancers during tumor growth inducing cell proliferation and inhibiting cell death. VS are caused by mutations in the neurofibromatosis type 2 (NF2) gene and activation of the PI3K/AKT signaling pathway drives VS tumor growth. PTEN, BTG2, and TIMP1 are targets of miR-21 and function as inhibitors of the PI3K/AKT pathway. Therefore, miR-21 may represent a putative target for VS therapy.

Methods: Expression levels of miRNAs were analyzed by stem-loop quantitative Real Time PCR (qRT-PCR) based on SYBR-Green I. MirVana kit was used to isolate miRNA and RNA fractions. Expression of miRNA targets was analyzed by qRT-PCR. Protein expression of the miR21 targets was analyzed by western blots. MiRNA expression was normalized to the small RNA U6 while expression of miRNA targets was normalized to GAPDH.

Results: High expression levels of miR-21 were found in VS and meningioma cells as well as in VS tumors compared to Schwann primary cell lines. Additionally, miR21 targets and tumor suppressors, PDCD4, BTG2, PTEN and TIMP1, were found in very low expression levels.

Conclusion: MiR-21 over-expression supports VS tumor growth by inducing the AKT signaling pathway via the repression of PTEN, BTG2, TIMP1 and PDCD4. MiR-21 is a promising molecular target for the development of VS therapy.

Define Professional Practice Gap & Educational Need: There are no drugs currently FDA approved for the treatment of vestibular schwannomas and meningiomas; therefore, the discovery of new molecular targets represents an urgent clinical need for the development of novel therapeutics.

Learning Objective: (1) To study molecular mechanisms of microRNA regulation in Vestibular Schwannomas growth. (2) To reveal microRNA targets for putative VS therapy.

Desired Result: To develop microRNA inhibitors as a viable treatment strategy for vestibular schwannomas and meningiomas.

Indicate IRB or IACUC Approval: Approved

Initial Operative Experience and Hearing Preservation
Results with a Mid-Scala Cochlear Implant Electrode Array

Maja Svrakic, MD; J. Thomas Roland, Jr., MD
Sean O. McMenomey, MD; Mario A. Svirsky, MD

Objective: To describe our initial operative experience and hearing preservation results with the Advanced Bionics (AB) HiFocus Mid Scala Electrode (MSE)

Study Design: Retrospective review.

Setting: Tertiary referral center.

Patients: Sixty-three MSE implants in pediatric and adult patients were compared to age- and gender-matched 1j electrode implants from the same manufacturer.

Intervention: Cochlear implantation with either the AB 1j electrode or the AB MSE.

Main Outcome Measures: The MSE and 1j electrode were compared in their angular depth of insertion (aDOI) and ability to preserve residual hearing. Hearing preservation was analyzed as a function of aDOI. Secondary outcome measures included operative time, incidence of abnormal intraoperative impedance and telemetry values, and incidence of postsurgical complications.

Results: The aDOI was slightly shallower for the MSE electrode (391° vs. 418° for the 1j, $p<0.01$). Patients with MSE electrodes had better hearing preservation. Thresholds shifts at four audiometric frequencies ranging from 250 to 2,000 Hz were 10 dB, 7 dB, 2 dB and 6 dB smaller for the MSE electrode ($p<0.05$). Hearing preservation at low frequencies was worse with deeper insertion. Operative time was similar and complication rate was similarly low for both electrodes. The incidence of abnormal intraoperative impedances and neural response telemetry was slightly higher for the MSE.

Conclusions: The MSE electrode resulted in slightly shallower insertions and significantly better hearing preservation than the 1j electrode. Differences in other outcome measures were small or unlikely to have a meaningful effect.

Define Professional Practice Gap & Educational Need: Lack of surgical experience and lack of knowledge in depth of insertion and hearing preservation with a new mid scala electrode compared to the older electrode from the same device company

Learning Objective: To characterize the surgical aspects, depth of insertion and hearing preservation outcomes with a new mid scala electrode compared to the older electrode from the same device company

Desired Result: Will know what to expect with respect to depth of insertion and hearing preservation as well as operative time, incidence of abnormal intraoperative impedance and telemetry values, and incidence of postsurgical complications for the new mid scala electrode compared to the older electrode from the same device company

Indicate IRB or IACUC Approval: Approved

Depression, Self-Esteem, and Quality of Life in Vestibular Schwannoma Treatment Decision-Making

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Seth E. Pross, MD; Lisa Ishii, MD, MSH
Masaru Ishii, MD, PhD; Howard W. Francis, MD, MBA

Objective: To identify psychological factors associated with treatment modality selection in vestibular schwannoma.

Study Design: Prospective observational study

Setting: Tertiary care neurotology clinic

Patients: Data was prospectively collected from patients initially presenting to a tertiary care neurotology clinic between 2013-2015. Patients who did not have MRI, demographic, psychometric, or audiometric data were excluded from analysis.

Intervention: Demographic information, tumor size, Beck depression inventory, Rosenberg self-esteem scores, headache severity, and clinical symptoms were collected to determine factors associated with undergoing acoustic neuroma surgical resection using univariate and multiple logistic regression analysis.

Results: A total of 143 acoustic neuroma patients (mean age 54.2 years, 50.4% female) were included. 62 patients (43.3%) pursued surgical resection, 81 patients (56.6%) pursued active surveillance. Surgical treatment was significantly associated with larger tumors, lower word discrimination scores, and higher headache severity scores ($p<0.05$). Depression, self-esteem, quality of life, hearing thresholds, and clinical symptoms were not significantly associated with treatment modality. On multiple logistic regression analysis, the likelihood of undergoing surgical resection significantly increased in patients with moderate (OR 115.75; 9.21-1454.34) and large grade tumors (OR 72.75; 95% CI 4.33-1221.28), and higher headache severity scores (OR 1.03;95% CI 1.01-1.06). The likelihood of undergoing surgical resection was not significant for small and medium grade tumors.

Conclusions: Patients with larger acoustic neuromas and worse headaches are more likely to undergo surgery rather than active surveillance. However, psychometric factors such as depression, quality of life, and self-esteem do not seem to impact patient decision-making.

Define Professional Practice Gap & Educational Need: Lack of understanding regarding the impact of psychological factors on treatment decision-making by acoustic neuroma patients.

Learning Objective: To identify psychological factors associated with treatment modality selection in vestibular schwannoma patients.

Desired Result: With a better understanding of the factors impacting treatment decision-making, clinicians may provide better patient-centered discussions regarding management of acoustic neuromas.

Indicate IRB or IACUC Approval: Approved

Defining the Limits of Endoscopic Access to Internal Auditory Canal: Anatomical and Computed Tomographic Analysis of an Exclusively Endoscopic Approach

Adam N. Master, MD; L. Gale Gardner, MD
Maura K. Cosetti, MD

Hypothesis: It is possible to quantify surgical access to internal auditory canal (IAC) via a transcanal endoscopic technique in reference to neurovascular and osseous surgical landmarks of the temporal bone

Background: Transcanal endoscopic ear surgery is a reliable, effective and minimally invasive technique for middle ear and mastoid pathology, however its application to the IAC has not been widely investigated.

Methods: Anatomic dissection of 2 paired and 15 unpaired cadaveric temporal bones was performed through an exclusively endoscopic approach to the IAC. Dissection proceeded until access to the cerebellopontine angle (CPA) was achieved. Following dissection, all specimens underwent computed tomography (CT) scans. Anatomage InVivo5 software was used to analyze the CT scans and record measurements.

Results: Access to the CPA and visualization of the labyrinthine segment of the facial nerve was achieved in all specimens. The mean distance from the carotid artery, jugular bulb, and middle fossa from our fundostomy of the IAC was 4, 5, and 5 mm, respectively. Mean surface area of the fundostomy and tympanic ring were 42 and 81mm2. The mean distance from the osteocartilaginous junction and tympanic ring to the porus acousticus was 29 and 21mm.

Conclusion: Transcanal access to the entire IAC can be achieved safely achieved via an endoscopic approach. Generous removal of the cochlear promontory can be accomplished while maintaining a safe distance from key neurovascular structures. The endoscopic transcanal approach to the IAC offers a minimally invasive alternative in patients without serviceable hearing for intra-meatal and medial IAC tumors.

Define Professional Practice Gap & Educational Need: 1. Need for better understanding of the emerging applications of transcanal endoscopic ear surgery to the lateral skull base 2. Need for greater understanding of the anatomic limitations and potential application of exclusively endoscopic technique to the internal auditory canal

Learning Objective: to quantify surgical access to internal auditory canal (IAC) via a transcanal endoscopic technique in reference to neurovascular and osseous surgical landmarks of the temporal bone

Desired Result: Attendees will understand the anatomic limitations and potential applications of the exclusively endoscopic approach to the internal auditory canal

Indicate IRB or IACUC Approval: Exempt

Dosimetric Analysis of Adjacent Neurovascular Structures in Treatment of Skull Base Tumors with CyberKnife Radiation Therapy

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Objective: To examine the relationship between the prescribed target dose and the dose to healthy neurovascular structures in patients with skull base tumor treated with CyberKnife radiation therapy.

Study Design: Retrospective analysis

Setting: Academic Tertiary Care Center

Patients/Interventions: Twenty patients with vestibular schwannomas who were treated with fused CT/MRI-guided CyberKnife radiation therapy.

Main outcome measures: Average radiation dose delivered to healthy neurovascular structures (e.g. carotid artery, basilar artery, and facial nerve, trigeminal nerve, and cochlea) was analyzed.

Results: The prescribed dose ranged from 18-23.35Gy over 1-5 fractions to cover 95% of the target tumor volume. The mean dose to the carotid artery was 5.71Gy (range 0.36-17Gy); basilar artery was 2.14Gy (range 0.20-5.32Gy); facial nerve was 2.95Gy (range 1.74-7.54Gy); trigeminal nerve was 5.21Gy (range 0.39-17.09Gy); and the cochlea was 8.06Gy (range 2.45-12.61Gy).

Conclusions: Stereotactic radiosurgery for some vestibular schwannomas can expose the basilar artery and carotid artery to radiation doses that can initiate atherosclerotic processes. The dose delivered to other structures such as the cochlea and facial nerve appears to be lower and much less likely to cause immediate issues. The relationship between the size and location of the tumor with the radiation doses will be discussed.

Define Professional Practice Gap & Educational Need: Lack of knowledge on the dosing to adjacent healthy neurovascular structures when using CyberKnife radiation therapy in the treatment of skull base tumors.

Learning Objective: To examine the relationship between the prescribed target dose and the dose to healthy neurovascular structures in patients with skull base tumor treated with CyberKnife radiation therapy.

Desired Result: Participants will be able to discuss toxicity of adjacent healthy neurovascular structures as a result of CyberKnife radiation therapy when treating skull base tumors.

Indicate IRB or IACUC Approval: Exempt

Treatment of Lateral Skull Base and Posterior Cranial Fossa Lesions Utilizing the Extended Middle Cranial Fossa Approach

Joseph P. Roche, MD; Andrew J. Goates, BS
Marlan R. Hansen, MD; Bruce J. Gantz, MD

Objective: Define the indications and outcomes for patients undergoing treatment utilizing the extended middle cranial fossa approach (EMCF).

Study design: Retrospective records review.

Setting: University-based tertiary referral center.

Patients: Patients undergoing treatment of PCF lesions.

Intervention(s): EMCF exposure and treatment of the indicating PCF lesion.
Main outcome measure(s): Demographic, indication for surgery, audiometric, and cranial nerve functioning variables were assessed.

Results: Thirty-five subjects who underwent an EMCF exposure were identified over a 12-year period. The most common indication was meningioma (18, 51%) followed by various schwannomas (6, 17%) and vascular lesions (5, 14%). Preoperative cranial nerve complaints were common (31, 91%) as were objective cranial nerve abnormalities on physical exam (21, 60%). Available pre-operative audiometric data were demonstrated good functioning including three-tone pure tone averages (23 +/- 15 dB HL and word understanding scores (92 +/- 20 %). Most (34, 97%) subjects intact facial nerve function. There were no intraoperative complications and no perioperative deaths attributable to the surgical intervention. The average length of stay was 11.6 days (median = 9). Cranial neuropathies were common postoperatively with 24 (68%) subjects demonstrating some objective cranial nerve dysfunction, the most common of which was trigeminal nerve hypesthesia (20, 57%). Subjects with identifiable pre- and post-operative audiometric data demonstrated small declines in the three-tone average (<3dB HL) and word recognition scores (~9%). Four subjects had complete hearing loss postoperatively. No subject had a change in facial nerve function postoperatively.

Conclusions: The EMCF approach can provide safe and effective exposure of the anterior PCF.

Define Professional Practice Gap & Educational Need: The extended middle cranial fossa (EMCF) approach to the cerebellopontine angle, parapontine and prepontine cisterns can be utilized for a variety of skull base and posterior cranial fossa (PCF) lesions. Currently, few reports exist documenting the variety of lesions that can be approached and outcomes that can be expected. This report adds large series of patients undergoing EMCF approachs to the petrous apex and PCF and the outcomes that can be expected using this approach.

Learning Objective: To recognize the breadth of indications and possible outcomes that can be expected when utilizing the EMCF approach.

Desired Result: To aid in the decision making process when choosing how to best approach PCF lesions.

Indicate IRB or IACUC Approval: Approved

Treatment Paradigms in the Management of Late Stage Neurofibromatosis Type II Patients

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Sean O. McMenomey, MD; Matthias A. Karajannis, MD
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Objective: Available treatments for neurofibromatosis type II patients at a comprehensive multi-disciplinary center are presented along with algorithms for decision-making to maximize patients' quality of life.

Study design: Retrospective case series

Setting: Tertiary academic medical center.

Patients: Neurofibromatosis type II patients managed at a comprehensive center.

Intervention(s): Medical and surgical treatments for tumor control and to rehabilitate cranial neuropathies including resection of symptomatic tumors, treatments of hearing loss, dynamic and static facial nerve reanimation and systemic therapy and drug trials where eligible.

Main outcome measure(s): Restoration of function or delay in debilitating dysfunction.

Results: Eleven patients with a broad spectrum of disease progression were included. Age ranged from 12-54 years. Patients were managed with serial volumetric imaging, surgical resection, and systemic therapeutic drug trials, such as bevacizumab, trametinib, everolimus, and lapatinib. Co-morbidities from disease progression include facial paresis, hearing loss, and poor nutritional status. Patients were offered facial nerve grafts, cochlear or auditory brainstem implants and procedures for the treatment of dysphagia. Through a well-coordinated algorithmic approach, these patients are able to enjoy improved quality of life.

Conclusions: Management of late-stage neurofibromatosis type II patients, especially at a young age, is a complex issue requiring extensive counseling and discussion about the goals of care. There are multiple medical and surgical treatment options that can be utilized to maximize quality of life for these patients. Some of these address the disease process while others seek to rehabilitate functions lost from disease progression or surgery. Management requires a multi-disciplinary team well versed in these options in order to create individualized treatment plans.

Define Professional Practice Gap & Educational Need: Neurofibromatosis type II is a complicated potentially life threatening disease with various considerations that direct the course of management. Certain approaches in the management of aggressive phenotypes may maximize quality of life.

Learning Objective: After this presentation, participants should have an understanding of: 1.) the clinical presentation of late-stage neurofibromatosis type II, 2.) the importance of a multi-disciplinary approach to management, and 3.) current management options to maximize the quality of life for neurofibromatosis type II patients as it relates to hearing and facial nerve outcomes.

Desired Result: Attendees will have a better understanding of the approach to management in advanced neurofibromatosis type II. Use of the discussed algorithms will allow them to provide more thoughtful, patient-centered care.

Indicate IRB or IACUC Approval: Approved

Rare Metastatic Lesions of the Internal Auditory Canal

Richard J. Wiet, MD, Robert A. Battista, MD
R. Mark Wiet, MD; Jenna K. Little, BS
Richard J. Wiet, MD

Objective: The goal of this study is to elucidate the key differences between the more common vestibular schwannoma of the internal auditory canal (IAC), and the rare metastatic lesion to the IAC.

Study design: Retrospective case series

Setting: Tertiary referral center

Patients: A review of 1200 known IAC/cerebellopontine angle (CP angle) tumor cases was conducted. Six patients (0.5% of the known cases) were found to have tumors metastatic to the IAC. A study of patterns that set the metastatic cases apart from vestibular schwannoma was conducted.

Outcome measures: History, histology, audiogram, MRI scans, cerebrospinal fluid (CSF) cytology.

Results: All six patients presented with sudden hearing loss; two were bilateral. Leptomeningeal carcinomatosis was found in two of three CSF samples. Three cases presented with facial paralysis. Except for one, all five died within months of the diagnosis. Histology was available for four cases. Two cases had metastatic breast cancer; one case had adenocarcinoma, mammary type. Histology in the remaining two cases was squamous cell carcinoma and gastrointestinal adenocarcinoma. The diagnosis of breast carcinoma preceded the IAC lesion by two years in one case. The remaining patients demonstrated variability in time between the primary carcinoma diagnosis and the metastatic IAC lesion finding.

Conclusion: Metastatic disease to the IAC/CP angle should be suspected in cases with sudden hearing loss, age exceeding 55 years, facial nerve neuropathy, and/or a history of prior malignancy. CSF cytology should be considered as a diagnostic tool in cases suspected of metastatic disease.

Define Professional Practice Gap & Educational Need: Few case studies present more than one to two cases on metastatic lesions of the internal auditory canal. Our efforts are to broaden the scope of these rare cases by offering more evidence and identifying key factors that distinguish suspicion for metastatic lesions of the IAC from vestibular schwannoma.

Learning Objective: To learn the range of symptoms that may require additional evaluation for possible metastatic lesions of the internal auditory canal.

Desired Result: To obtain a greater knowledge on the differentiation between the diagnostic investigation of rare metastatic lesions and vestibular schwannoma. In addition, the study aims to classify instances where metastatic disease should be suspected.

Indicate IRB or IACUC Approval: Exempt

Vestibular Schwannoma Volume to Posterior Fossa Volume Ratio as a Predictor of Clinical Outcomes following Resection

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Oliver F. Adunka, MD

Hypothesis: Radiographically-determined vestibular schwannoma volume relative to posterior fossa volume is a better predictor of postoperative outcomes than tumor volume alone.

Background: Larger tumor volume is known to portend a poorer prognosis with respect to post-operative outcomes such as hearing preservation and facial nerve function. Current clinical practice, however, does not take into account the size of the posterior fossa relative to the tumor dimensions.

Methods: A retrospective review of patients undergoing retrosigmoid or translabyrinthine vestibular schwannoma resection at the study institution was conducted. Individual volumetric posterior fossa dimensions and preoperative tumor volumes were determined. Clinical outcomes considered included post-operative House-Brackmann Score, resection status, complications, and surgical duration. One-way and two-way ANOVAs were performed.

Results: A total of 95 patients were identified. A one-way ANOVA demonstrated that surgical time ($p < 0.001$) and whether a Good Outcome (House-Brackmann score of 1 or 2, no complications, and a complete resection) was achieved ($p = 0.009$) correlated very well with preoperative tumor volume, but not with posterior fossa volume ($p = 0.412$ and $p = 0.345$, respectively). However, in medium sized tumors, facial function was significantly correlated with posterior fossa volume ($p = 0.032$).

Conclusions: Our results suggest that the ratio of vestibular schwannoma volume to posterior fossa volume may hold clinically-useful potential in planning surgery and prognosticating outcomes for patient with medium sized tumors.

Define Professional Practice Gap & Educational Need: This submission targets lack of awareness and lack of contemporary knowledge.

Learning Objective: The primary learning objective is to make the audience aware of the potential utility of considering the ratio of vestibular schwannoma volume to posterior fossa volume in pre-operative planning and predicting post-operative outcomes.

Desired Result: The desired result of this knowledge is that the audience could use vestibular schwannoma volume to posterior fossa volume ratio as an additional variable to guide surgical approach selection and planning, as well as pre-operative patient counseling.

Indicate IRB or IACUC Approval: Exempt

Facial Nerve Schwannomas Mimicking as Vestibular Schwannomas

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Jason R. Bell, MD; Dennis I. Bojrab, MD
Michael J. LaRouere, MD; Matthew L. Kircher, MD
Seilesh C. Babu, MD

Objective: To describe our series of presumed vestibular schwannomas, found to be facial schwannomas, and to determine methods to distinguish their differences preoperatively.

Study Design: Retrospective chart review.

Setting: Tertiary Referral Center.

Patients: Eighteen cases with a presumed diagnosis of vestibular schwannoma found to have a facial schwannoma intraoperatively, from October 2002 to July 2015, were reviewed.

Intervention: Thirteen patients underwent tumor resection: 9 incomplete, 4 complete. Five patients had decompression of the tumor and two of those required no further treatment.

Main Outcome Measures: Demographics, surgical approach, intra-operative findings, hearing and facial nerve status, and adjunctive treatment were documented.

Results: Pre-operative hearing loss and imbalance were seen in 72% and 61%, respectively. Pre-operative electroneuronography (ENOG) revealed a mean weakness of 19%. Pre-operative imaging showed a mean tumor size of 1.6 x 1.6 cm. Suspicious intraoperative findings included: facial nerve incorporated intimately with the tumor capsule in 12 cases; spontaneous action potentials noted while drilling the bony IAC in 3 cases; and action potentials noted on stimulation of the entire tumor capsule in 10 cases. The mean long-term facial function was House-Brackman grade 2 and the mean length of follow-up was five years.

Conclusions: Facial neuromas are rare and may be difficult to distinguish from a vestibular schwannoma pre-operatively. Surgical findings that should raise concern include: spontaneous action potentials during drilling the bony IAC, absence of a plane of dissection between the facial nerve and tumor, or stimulation of the tumor capsule.

Define Professional Practice Gap & Educational Need: Facial Nerve Schwannomas are rare tumors of the temporal bone but may be difficult to distinguish from a vestibular schwannoma, as classic clinical and radiological findings may be absent.

Learning Objective: To describe our series of presumed vestibular schwannomas, found to be facial schwannomas, and to determine methods to distinguish their differences preoperatively.

Desired Result: That attendees gain knowledge in the diagnosis and treatment of facial nerve schwannomas and are equipped to provide better care for these patients.

Indicate IRB or IACUC Approval: Approved

An Easy and Reliable Method to Locate the Dehiscence during Middle Fossa Superior Canal Dehiscence Surgery: It's a (C) inch

Neil S. Patel, MD; Jacob B. Hunter, MD
Brendan P. O'Connell, MD; George B. Wanna, MD
Matthew L. Carlson, MD

Objective: The middle fossa floor lacks reliable surface landmarks. Furthermore, in cases of superior semicircular canal dehiscence (SSCD), multiple skull base defects may be present further confounding the location of the labyrinth. Misidentification of the SSCD during surgery may lead to treatment failure or sensorineural hearing loss. Anecdotally, the authors have observed the distance from the lateral edge of the craniotomy to the SSCD to be consistently one inch. Herein we present radiologic evidence of this practical and clinically useful relationship.

Study Design: Retrospective radiological analysis and chart review.

Setting: Tertiary center.

Patients: Consecutive patients with radiological evidence of SSCD.

Interventions: Analysis of CT imaging.

Main outcome measures: The horizontal distance from the outer cortex of the squama temporalis immediately superior to the bony external auditory canal (approximating lateral edge of craniotomy) to the SSCD was measured in the coronal plane by two independent reviewers.

Results: A total of 151 adult ears with SSCD were analyzed. A Shapiro-Wilk goodness-of-fit test confirmed that measurements were normally distributed. Pearson inter-rater correlation was 0.95, confirming very strong agreement between observers. The mean distance between the outer cortex of the squama temporalis and SSCD was 25.9 mm, or 1.02 inches. Sixty-eight percent of the SSCD population would fall between 0.92 and 1.12 inches and 95% would lie between 0.83 and 1.21 inches.

Conclusions: The horizontal distance from the outer cortex of the squama temporalis to the SSCD consistently approximates one inch. This easily-remembered distance can aid surgeons in locating or confirming the SSCD during middle fossa surgery.

Define Professional Practice Gap & Educational Need: 1. Lack of anatomic landmarks to guide middle fossa superior semicircular canal dehiscence surgery.

Learning Objective: 1. To learn that the average distance from the lateral edge of a middle fossa craniotomy defect to a dehiscent superior semicircular canal is approximately one inch.

Desired Result: 1. To apply the one-inch distance estimate to superior semicircular canal dehiscence surgery to promote safe surgery and improve outcomes.

Indicate IRB or IACUC Approval: Approved

Semicircular Canal Dehiscence in Patients with Cadherin 23 Related Hearing Loss

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Jack E. Russo, MD; Clifford J. Eskey, MD
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Objective: To investigate the prevalence and relative risk of semicircular canal dehiscence in pediatric patients with CDH23 mutations compared to an aged-matched control group.

Study Design: Retrospective cohort study

Setting: Multi-institutional study

Patients: Sixteen pediatric patients (ages 0-5 years) were compared based on the presence of a CDH23 mutation with a control population.

Interventions: Retrospective review of high resolution CT temporal bone scans

Main outcome measures: Superior and posterior semicircular canals were evaluated by a neuroradiologist for presence of semicircular canal dehiscence in the CDH23 variant and control groups

Results: Sixteen CT scans were reviewed for semicircular canal dehiscence. Five of six children with CDH23 variant had dehiscence in at least one canal compared to only one child out of ten in an age-matched control population. Out of six patients (12 ears) in the CHD23 variant group there were five dehiscencent superior semicircular canals (RR=8.3) and three dehiscencent posterior canals (RR= 5.9). Three children had bilateral dehiscence of the canals. However no children had dehiscence in both the superior and posterior canals. Relative risk of semicircular canal dehiscence in children with CDH23 mutation is 8.33 compared to the pediatric control population. 95% confidence interval (1.25, 55.35).

Conclusions: Children with CDH23 mutations are at significantly increased risk of having semicircular canal dehiscence in both the superior and posterior semicircular canals and may be a contributing factor to the vestibular dysfunction in USH1D patient population.

Define Professional Practice Gap & Educational Need: Lack of awareness/contemporary knowledge: There are several competing theories about the etiology of semicircular canal dehiscence and the ossification of the otic capsule. It has been theorized that CDH23 mutations may be associated with delayed ossification. This study sheds new light on the topic by showing a correlation between canal dehiscence and mutations of the cadherin 23 protein.

Learning Objective: To describe the correlation between CHD23, a gene linked to Usher Syndrome 1D and non-syndromic hearing loss, and semicircular canal dehiscence in children.

Desired Result: Increased awareness of the correlation between this genetic mutation and semicircular canal dehiscence will allow physicians to better evaluate and treat children with CDH23 associated hearing loss

Indicate IRB or IACUC Approval: Approved

Pediatric Superior Semicircular Canal Dehiscence and Inner Ear Anomalies

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Objectives: To determine the incidence and association of superior semicircular canal dehiscence (SSCD) with inner ear (IE) anomalies in the pediatric population.

Study Design: Retrospective chart review.

Setting: Three tertiary referral centers in ambulatory and hospital settings.

Patients: Children less than 18 years who received a 0.5 mm or less collimated CT including the temporal bones between 2010 to 2013 for reasons including, but not limited to, hearing loss, trauma, and infection.
Interventions: Radiologic software was used to reformat images into Pöschl and Stenver planes. Five hundred three CT images were reviewed by experienced neuroradiologists.

Main Outcome Measures: Incidence of SSCD and IE anomalies, noting laterality. Patient age, sex, and diagnosis were recorded. Statistical analysis was performed to compare CT findings across outcome measures and patient demographics.

Results: Pediatric incidence of SSCD was 6.2% (31/503) and an IE anomaly was 15.1% (76/503). Within SSCD patients, incidence of an IE anomaly was 22.6% (7/31); and within IE anomaly patients, incidence of SSCD was 9.2% (7/76). Incidence of SSCD with an IE anomaly together was not significantly correlated (1.4%, 7/503; P=0.23). The Incidence of an IE anomaly with bilateral (0.8%, 4/503) vs. unilateral (0.6%, 3/503) SSCD was similar.

Conclusions: SSCD incidence is higher in the pediatric population compared to adults. SSCD and inner ear anomalies rarely occur together and are unlikely related. Similar incidences of having an IE anomaly with unilateral vs. bilateral SSCD further supports this.

Define Professional Practice Gap & Educational Needs: Lack of contemporary knowledge regarding pediatric superior semicircular canal dehiscence and its relationship with inner ear anomalies.

Learning Objective: To understand the increased incidence of pediatric superior semicircular canal dehiscence and its rare occurrence with inner ear anomalies, being unlikely related.

Desired Result: Changes in physician knowledge of superior semicircular canal dehiscence, specifically in pediatric populations as it may contribute to the understanding of its pathogenesis.

Indicate IRB or IACUC Approval: Approved

Cochlear Patency is Maintained after Transmastoid Labyrinthectomy

Eric W. Sargent, MD; Eric Liao, MD; Roger L. Gonda, Jr., MD

Objective: Labyrinthectomy is considered the 'gold standard' in the treatment of intractable vertigo attacks due to Ménière's Disease (MD) but sacrifices all residual hearing. Interest in auditory rehabilitation has lead to cochlear implantation in some patients. Concern remains that the cochlear lumen may fill with tissue or bone after surgery. This study sought to determine the incidence of obliteration of the cochlea after transmastoid labyrinthectomy.

Study Design: Retrospective observational study.

Setting: Tertiary referral center.

Patients: 18 Patients with intractable vertigo from MD who underwent surgery.

Interventions: Transmastoid labyrinthectomy between 2008 and 2013. Cochleas were imaged with unenhanced, heavily T2-weighted MRI.

Main Outcome Measure: Presence of symmetrical cochlear fluid signals on MRI.

Results: There was no loss of fluid signal in the cochleas of operated ear compared to the contralateral, unoperated ear in any subject an average of 3 years (SD: 1.2) after surgery. 3/18 patients had the vestibule blocked with bone wax at the time of surgery. Blocking the vestibule with bone wax did not change the cochlear fluid signal.

Conclusion: The risk of cochlear obstruction after labyrinthectomy for MD is very low. The significance of this finding is that patients with MD who undergo labyrinthectomy will likely remain candidates for cochlear implantation in the labyrinthectomized ear long after surgery if this becomes needed. Immediate cochlear implantation or placement of a cochlear lumen keeper during labyrinthectomy for MD is probably not necessary.

Define Professional Practice Gap: The persistence of cochlear patency after labyrinthectomy is unknown. There is concern that labyrinthectomy may lead to later cochlear obstruction in some labyrinthectomized patients, making them less able to benefit from cochlear implantation should it become required. This has lead some surgeons to advocate implanting labyrinthectomized ears immediately, perhaps unnecessarily.

Learning objective: To show that obliteration of the cochlea by tissue or bone after labyrinthectomy for Ménière's Disease is probably rare.

Desired Results: Patients undergoing labyrinthectomy for Ménière's Disease do not require immediate cochlear implantation or placement of a cochlear lumen keeper to remain eligible for later cochlear implantation. This result varies from findings after translabyrinthine vestibular schwanomma surgery in which cochlear obliteration is common.

Indicate IRB or IACUC Approval: Approved

Evaluation of Cochlear Anatomy Models for Determining Intra-cochlear Electrode Position

Ahmet Cakir, MRes; Robert Labadie, MD, PhD
Benoit Dawant, PhD; Jack Noble, PhD

Hypothesis: Use of high-resolution non-rigid models of intra-cochlear anatomy permits more accurate determination of cochlear implant (CI) electrode position than rigid models.

Background: Quantifying intra-cochlear position of CI electrodes is important for studying the relationship between electrode position and hearing outcomes and when using electrode position-based CI programming strategies. Evaluating electrode position within the cochlea requires identifying the basilar membrane, which is not visible in CT images. Two commonly used approaches for estimating basilar membrane position in CT are: (1) Register scans with a high resolution image of a single cochlear specimen where the membrane is visible (rigid model) and (2) register scans with a non-rigid model created based on the anatomy of 10 individual cochleae. There were no studies quantifying the utility of the non-rigid versus the rigid model.

Methods: The automatic non-rigid method was used to localize cochlear anatomy of 79 ears. Rigid anatomical models of 7 cochlear ÂµCT specimens were registered to each ear. Electrode position was quantified relative to each model.

Results: Standard deviation across rigid models in measures of electrode depth, distance to modiolus, and distance to basilar membrane were, 4.92. 0.53, and 0.67 mm. Mean difference in those measures between the non-rigid model and the average result of the rigid models was 0.64, 0.49, and 0.35 mm.

Conclusion: Variance of inter-rigid-models is high. In contrast, differences between non-rigid and the average of rigid model estimations are relatively low, demonstrating that non-rigid models are necessary to achieve the most accurate estimate of intra-cochlear electrode position.

Define Professional Practice Gap & Educational Need: Multiple methods exist for identifying the intra-cochlear position of cochlear implant electrodes, but no studies have been performed to contrast the performance of the two methods.

Learning Objective: To inform the community of the reliability and limitations of different methods that have been proposed to assess electrode position using CT images.

Desired Result: Attendees will be aware of the limitations of existing techniques for assessing cochlear implant electrode position.

Indicate IRB or IACUC Approval: Approved

The Central Auditory System and Cochlear Implantation: Using Olfactory Testing to Evaluate a Potential Central Component in Cochlear Implant Performance

Hinrich Staecker, MD, PhD; Thomas Muelleman, MD
Valerie Wood, MD; Elizabeth Ripley, AuD

Hypothesis: Olfactory testing may identify patients that are at risk for poor cochlear implant performance.

Background: Cochlear implantation is a highly successful intervention that despite remarkable improvements in hardware and software continues to show a high degree of variability in outcomes. Performance in adult patients can potentially be affected by the integrity of spiral ganglion neurons or by the performance of the central auditory system. Prolonged deafening and dementia are conditions that affect the central auditory system and can negatively impact cochlear implant outcomes. Central auditory test batteries can evaluate the central component of hearing in patients that significant residual hearing but can not be effectively used in most cochlear implant patients. A wide variety of recent studies have shown that decline in olfaction predates and often predicts a variety of central nervous system degenerative disorders. We set out to evaluate if olfaction testing could identify patients with poorer implant outcomes than age matched controls.

Methods: Adult cochlear implant candidates were recruited and olfaction measured with the University of Pennsylvania smell identification test (UPSIT). At 6 months post implant activation composite scores of the UPSIT were compared to the patients CNC and AzBIO scores. Poor performance on the UPSIT was correlated with poorer speech scores suggesting that olfactory testing may be useful in preoperative evaluation of cochlear implant patients.

Conclusions: Identification of patients at risk for central auditory system dysfunction may be possible by evaluation of patients' olfactory function.

Define Professional Practice Gap & Educational Need: Awareness of the role of central auditory function in cochlear implantation

Learning Objective: Understand the relationship between olfactory function and neurodegeneration

Desired Result: Screening patients for central auditory degeneration prior to cochlear implantation may alter rehab strategies

Indicate IRB or IACUC Approval: Approved

Cochlear Histopathology as Seen in Two Patients with a Clarion® Cochlear Implant Electrode with Positioner

Takefumi Kamakura, MD, PhD
Joseph B. Nadol Jr., MD

Hypothesis: This study reports the cochlear histopathology of two cases who during life underwent cochlear implantation with a positioner.

Background: A silastic positioner introduced by the Advanced Bionics Corporation in 1999 was designed to position the electrode of the ClarionÂ® cochlear implant close to the modiolus. The positioner was recalled in the United States in July 2002 because of an apparent higher incidence of bacterial meningitis in patients in whom the positioner had been placed.

Methods: Four celloidin-embedded temporal bones from two patients with cochlear implants with a positioner were included in the study. In a previous study, we reported histopathologic findings in Case 1, and in this report, we present the findings in a second case in a 94-year-old woman (Case 2), and the similarities and differences between the two cases. All four specimens were prepared for histologic study by conventional techniques and 2-D reconstruction.

Results: Evidence of insertion trauma was seen in all three implanted specimens. More significant trauma was found in Case 2 than Case 1 including disruption of the osseous spiral lamina and the basilar membrane. In addition, there was more new fibrous tissue and bone in Case 2 than Case 1. There was a large fluid space in all three implanted temporal bones around the electrode and positioner.

Conclusion: The findings seen in the two cases may help to explain the increased risk of meningitis in patients implanted with a positioner. Significant cochlear trauma was seen in all implanted specimens.

Define Professional Practice Gap & Educational Need: Lack of awareness of cause of bacterial meningitis after cochlear implantation with a positioner.

Learning Objective: To discuss possible causes of bacterial meningitis after cochlear implantation with a positioner as revealed by histopathologic study.

Desired Result: Evidence of insertion trauma, new fibrous tissue and bone, and a large fluid space around the electrode and positioner were seen in all three implanted specimens, which may help to explain the increased risk of meningitis.

Indicate IRB or IACUC Approval: Approved

Interactive iPad-Based Education for Cochlear Implant Candidates

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Afsheen Moshtaghi, BS; Harrison W. Lin, MD
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Objective: One challenge cochlear implant (CI) teams face is the tremendous counseling required prior to the procedure. This study seeks to evaluate the efficacy of an iPad-based interactive educational module in patient education on CI, the various devices and their unique features, expectations/outcomes, and the surgery.

Methods: CI candidates were randomized into two groups. One group received the iPad-based interactive iBook while the other didnt. Both groups received standardized verbal information from the surgeon. The iBook provides a step-by-step interactive description of all necessary information regarding CI. Complete with animations and illustrations, the patient learns all the risks and benefits associated with the surgical procedure in addition to what to expect post operatively. Pre and post-education surveys were performed.

Results: Data is still being collected, however, preliminary results indicate statistically significant (p=.026) increases in patient understanding of: CI, expectations post-CI, differences between CI devices, long-term expectations of CI, CI risks, and recovery post-CI surgery when given the iBook.

Conclusion: By using the iBook to increase patient knowledge, the patient can have a more effective and productive conversation with the physician and audiologist. In this setting, the physician and audiologist can use his/her expert knowledge to address difficult question specifically pertaining to the patient to achieve a superior level of understanding.

Define Professional Practice Gap & Educational Need: There might be a lack of knowledge about the cochlear implanta among the patients or their families and several sessions for consultations may be required. The proper and ideal method for giving information prior the surgery still remains a challenging problem.

Learning Objective: To observe the effectiveness of the iPad-based education for cochlear implant in improvement of the knowledge in patients and their families prior to consultation with physician and surgery.

Desired Result: To evaluate the efficacy of an iPad-based interactive educational module in patient education on Cochlear Implant, the various devices and their unique features, expectations/outcomes, and the surgery.

Indicate IRB or IACUC Approval: Exempt

Preservation of Low Frequency Hearing in Children with Enlarged Vestibular Aqueduct

Kevin D. Brown MD, PhD; Lisa Park, AuD
Erika Gagnon, AuD; Jennifer Woodson, AuD
Holly Teagle, AuD

Objective: To determine the rates of preservation of functional acoustic hearing in children undergoing cochlear implantation with enlarged vestibular aqueduct and the benefits of hybrid (electric and acoustic) stimulation in this population Study Design: Case series

Setting: Tertiary children's hospital

Patients: Pediatric subjects 12 months to 18 years with preoperative low frequency hearing < 80 dB at 250 and 500Hz.

Intervention: Cochlear implantation with 20mm electrode by soft round window technique Main Outcome Measure: Preservation of functional acoustic hearing following cochlear implantation defined as < 80 dB at 250 and 500Hz.

Results: Pediatric patients with enlarged vestibular aqueduct consistently achieve high levels of hearing preservation and have substantial benefits of hybrid stimulation of the implanted ear similar to other hearing preservation candidates.

Conclusions: High levels of hearing preservation are possible in pediatric patients with enlarged vestibular aqueduct and substantial benefits are realized with hybrid stimulation.

Define Professional Practice Gap & Educational Need: Lack of contemporary knowledge of high likelihood of hearing preservation in children with enlarged vestibule aqueduct and benefits of hybrid hearing in children with enlarged vestibule aqueduct.

Learning Objective: The learner will understand that children with enlarged vestibular aqueduct can have high levels of low frequency hearing preservation and substantially benefit from hybrid stimulation of their cochleas.

Desired Result: The learner will be able to apply this information to educate parents of children with enlarged vestibular aqueduct of high likelihood of hearing preservation after cochlear implantation and the benefits of hybrid sound stimulation.

Indicate IRB or IACUC Approval: Approved

Hearing Loss after Round Window Surgery in Mice is due to Middle Ear Effusion

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Kevin T. Isgrig, BS; Lisa L. Cunningham, PhD
Wade W. Chien, MD

Hypothesis: Hearing loss associated with round window (RW) injection in mice is caused by middle ear effusion (MEE).

Background: Delivery of therapeutic agents directly through the RW offers promise for treating sensorineural hearing loss. However, hearing loss can result from the surgical approach itself, and the reasons for this are poorly understood. We examined the hearing loss following the three major steps involved with the RW approach to access the mouse cochlea: bullostomy, RW puncture, and RW injection.

Methods: 21 adult CBA/J mice underwent bullostomy alone; 10 underwent RW puncture, and 8 underwent RW injection with PBS with 5% glycerol. Auditory brainstem responses and otoscopy were performed preoperatively and up to six weeks postoperatively. Hair cells were stained and survival was assessed using immunofluorescence.

Results: One week postoperatively, mice in all groups showed significant threshold shifts. Otoscopy revealed approximately half of all mice had MEE, with a higher incidence of effusion in the RW puncture and RW injection groups. Those with MEE had significant ABR threshold shifts, whereas those without MEE had minimal hearing loss. MEE persisted through six weeks in a majority of cases, but in those mice with MEE resolution, there was at least partial improvement in hearing. Immunohistochemistry showed minimal loss of hair cells in all animals.

Conclusion: MEE is highly correlated with hearing loss in mice undergoing bullostomy, RW puncture, and RW injection. Otoscopy is an important adjunct to consider after ear surgery in mice, as MEE may contribute to post-surgical hearing loss.

Define Professional Practice Gap & Educational Need: Lack of knowledge regarding the causes of hearing loss after round window surgery in mice.

Learning Objective: Middle ear effusion is a major cause of hearing loss after round window surgery in mice.

Desired Result: Otoscopy should be performed after ear surgery in mice, and otoscopic findings should be taken into account when assessing hearing outcomes.

Indicate IRB or IACUC Approval: Approved

Incidence of Sigmoid Sinus Wall Anomalies in Patients with Idiopathic Intracranial Hypertension

Kristen Angster, MD; Erica Archer, MD
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Robert Morales, MD; David Eisenman, MD

Objective: Determine the incidence of sigmoid sinus wall anomalies (SSWA) in patients with idiopathic intracranial hypertension (IIH).

Study Design: Prospective diagnostic interventional study

Setting: Tertiary center

Patients: Adults with papilledema referred to neuro-ophthalmology and subsequently diagnosed with IIH

Intervention(s): Diagnostic Computed tomography (CT) Main outcome measure(s): Subjective pulse synchronous tinnitus (PST) and diagnosis of SSWA by CT scan

Results: 20 subjects were enrolled. The incidence of SSWA in patients with IIH and PST was 71%. No subjects without PST had a SSWA, though 2 had PST without a SSWA. All identified anomalies were sigmoid plate dehiscences without diverticulum. This incidence of SSWA in subjects with IIH and PST is significantly higher than than the 23% reported for patients who underwent CT for PST ($p<0.001$) and far exceeds the 1.2% reported for patients undergoing CT for complaints other than PST. The mean BMI was lower in subjects with SSWA (32.46 kg/m2) compared to subjects with no SSWA (43.9 kg/m2)(44.47) ($p=0.02$).

Conclusions: There is a high incidence of sigmoid sinus dehiscence in subjects with IIH and PST. These results further support an association between sigmoid sinus dehiscence and IIH, and implicate SSWA as necessary, though possibly not sufficient, cause of PST in patients with IIH. Patients with IIH and PST should undergo high-resolution diagnostic CT. Transmastoid sinus wall reconstruction should be considered if PST persists despite medical management. Patients with SSWA and PST should be screened for IIH.

Define Professional Practice Gap & Educational Need: The incidence of SSWA in patients with Idiopathic Intracranial Hypertension has yet to be defined, and the relationship between these two pathologies is poorly understood.

Learning Objective: Know the incidence of sigmoid sinus wall anomalies (SSWA) in patients with idiopathic intracranial hypertension (IIH), and explain the need to screen for IIH in all subjects with sigmoid sinus dehiscence.

Desired Result: Following the presentation, attendees will recognize the importance of screening patients with sigmoid sinus dehiscence for IIH. Attendees will also be able to counsel patients with IIH about potential medical and surgical treatment options for associated pulse synchronous tinnitus.

Indicate IRB or IACUC Approval: Approved

Outcomes of the Suture "Pull-through" Technique after Repair of Lateral Skull Base CSF Fistula and Encephaloceles

Brendan P. O'Connell, MD; Jacob B. Hunter, MD
Alex D. Sweeney, MD; Reid C. Thompson, MD
Lola B. Chambless, MD; George B. Wanna, MD
Alejandro Rivas, MD

Objective: 1) Report recurrences after repair of lateral skull base CSF fistula and encephalocele utilizing suture "pull-through" technique; 2) Examine post-operative audiometric outcomes using this method.

Study Design: Retrospective

Setting: Tertiary care hospital

Patients: Patients undergoing surgery for CSF fistula and/or encephalocele

Intervention: Combined transmastoid and middle fossa approach using suture pull-through technique

Main Outcome Measures: The primary outcome measures of interest were recurrence of CSF fistula or encephalocele, and post-operative ABG.

Results: 21 patients were included; mean age at surgery was 59Â±14 years and 66%(14/21) of patients were female. The majority of defects involved both the tegmen mastoideum and tympani(62%,13/21); multiple defects were present in 9 cases. Small craniotomy(2X3 cm) was performed and defects were repaired using composite grafts constructed with fascia, bone and/or cartilage, and dural substitute affixed with suture. The suture tail was left long and passed from the middle fossa through the defect into the mastoid. This allows centered and accurate reconstruction of the defect. At average follow-up of 9Â±8 months, no cases of recurrent CSF leak or encephalocele were noted. After excluding patients with ossicular chain discontinuity related to chronic ear pathology, a trend towards improved post-operative ABG at last follow-up(15Â±7 dB) was noted when compared to pre-operative ABG(24Â±12 dB)(p=0.06).

Conclusion: A combined transmastoid and middle fossa approach for repair of lateral skull base CSF fistula and encephaloceles using the aforementioned technique is highly efficacious. This method facilitates reliable placement of a composite graft in the center of skull base defects, even though small craniotomies.

Define Professional Practice Gap & Educational Need: This is a novel technique, there is lack of awareness of this surgical technique.

Learning Objective: Describe the combined transmastoid and middle fossa approach with suture pull through and report clinical outcomes.

Desired Result: Attendees will have an understanding of our technique.

Indicate IRB or IACUC Approval: Approved

Transverse Sinus Considerations in Idiopathic Intracranial Hypertension and Visual Impairment Intracranial Pressure Syndrome

Glenn W. Knox, MD

Objective: Visual impairment intracranial pressure syndrome (VIIP) can occur in astronauts exposed to microgravity. VIIP is characterized by visual decrements, cotton-wool spot formation, choroidal fold development, papilledema, optic nerve sheath distention and/or posterior globe flattening, along with elevated ICP. All of these manifestations can occur in idiopathic intracranial hypertension (IIH). The objective of the present study is to compare IIH to VIIP with respect to transverse sinus stenosis and pulsatile tinnitus.

Study Design: Retrospective case review.

Setting: Primary care and occupational medicine ambulatory center.

Patients: The first 15 astronauts identified as having developed VIIP syndrome during long-duration (six months) missions on the International Space Station. Six patients had been analyzed in detail sufficient for comparative purposes.
Interventions: Diagnostic. The VIIP patients were analyzed for the presence or absence of pulsatile tinnitus and transverse sinus stenosis. This series of patients was compared to a recently published meta-analysis of 19 studies with a total of 207 patients which examined outcomes of transverse sinus stenting for treatment of IIH.
Main Outcome Measures: Presence or absence of pulsatile tinnitus; presence or absence of transverse sinus stenosis on MRV.

Results: In the group of patients with VIIP syndrome, none of them had pulsatile tinnitus. Of the six cases, three of them had post-flight MRVs, all of which were negative for transverse sinus stenosis. In the meta-analysis of a total of 207 patients which looked at the overall clinical outcomes of transverse sinus stenting for the treatment of IIH, the authors reported an improvement rate of 81% with regard to headaches, 87% for papilledema, and 95% for pulsatile tinnitus.

Conclusions: Stenting of the transverse sinus does appear to be of therapeutic benefit in IIH. The small number of patients with VIIP syndrome suggests that the temporary elevation of ICP seen in microgravity does not result in transverse sinus stenosis. Transverse sinus stenosis may be a causative factor in IIH. In addition, transverse sinus stenosis may be a causative factor in the pulsatile tinnitus seen in IIH, since pulsatile tinnitus is absent in VIIP.

Define Professional Practice Gap & Educational Need: Lack of contemporary knowledge of the pathophysiology and treatment of IIH.

Learning Objective: To characterize pathophysiologic factors in IIH by comparison with an analogous disorder, VIIP.

Desired Result: Attendees can use this information to counsel IIH patients regarding pulsatile tinnitus and visual disturbances, and provide information to the patients regarding transverse sinus stenting as a possible treatment modality.

Indicate IRB or IACUC Approval: Approved

Reconstruction Outcomes Following Lateral Skull Base Resection

Nicholas J. Thompson, BS; Joseph P. Roche, MD
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Marlan R. Hansen, MD

Objective: Compare reconstruction outcomes for various lateral skull base closure techniques.

Study design: Retrospective medical records review.

Setting: University-based tertiary referral center.

Patients: Patients who underwent resections of tumors involving the lateral skull base requiring reconstruction beyond primary closure.

Intervention(s): Reconstructive techniques, from rotational flaps to free tissue transfer.

Main outcome measure(s): Outcome data including wound complications, CSF leakage, and need for surgical revision were tabulated.

Results: Eighty-six patients underwent lateral skull base tumor resection and reconstruction. Procedures were primarily lateral temporal bone resections but also included subtotal temporal bone, total temporal bone, and infratemporal fossa resections. Cutaneous malignancy was the most common resection indication (83%) and the temporalis rotational flap was the most commonly employed reconstructive option (30%). When free tissue transfer techniques were utilized, the radial forearm, anterolateral thigh, and latissimus dorsi were the most frequent donor sites. Patients with T2 disease were more likely to undergo temporalis flaps, whereas patients with T4 disease were more likely to undergo free flap reconstruction. Major complications were uncommon (~8%), the most frequent being stroke (~3%). The postoperative wound complication rate was approximately 45%. The majority involved minor dehiscence and were managed conservatively. Patients with T4 disease were more likely to have wound complications ($p<0.05$). Radial forearm free flaps were less likely to have wound complications when compared to other reconstruction techniques ($p<0.05$).

Conclusions: Many factors go into planning lateral skull base reconstruction. Free flaps were more often utilized for T4 disease. Radial forearm free flaps tended to have lower wound complication rates when compared to other techniques.

Define Professional Practice Gap & Educational Need: There are currently no discrete guidelines regarding optimal management of reconstructing lateral skull base defects.

Learning Objective: To recognize the different reconstruction options following skull base surgery and compare complication rates for each.

Desired Result: To aid in the decision making process of reconstructing lateral skull base defects with a focus on complication rates.

Indicate IRB or IACUC Approval: Approved

Cost Analysis of Cerebrospinal Fluid Leaks and Cerebrospinal Fluid Leak Prevention in Patients Undergoing Cerebellopontine Angle Surgery

Alexander Chern, BS; Jacob B. Hunter, MD
Marc L. Bennett MD

Objective: To determine if cranioplasty techniques following translabyrinthine approaches to the cerebellopontine angle (CPA) are cost-effective

Study Design: Retrospective case series

Patients: One hundred and eighty patients with available financial data who underwent translabyrinthine approaches at a single academic referral center between 2005 and 2015

Intervention: Cranioplasty with a dural substitute, layered fat graft and a resorbable mesh plate secured with screws

Main Outcome Measures: Billing data was obtained for each patient's hospital course for translabyrinthine approaches and postoperative cerebrospinal fluid (CSF) leaks

Results: One hundred and nineteen patients underwent translabyrinthine approaches with an abdominal fat graft (AFG) closure, with a median cost of $25,759.89 (range $15,885.65-$136,433.07). Sixty-one patients underwent translabyrinthine approaches with a dural substitute, AFG, and a resorbable mesh for closure, with a median cost of $29,314.97 (range $17,674.28-$111,404.55). The median cost of a CSF leak was $50,401.25 (range $0-$384,761.71). The additional cost of a CSF leak when shared by all patients who underwent translabyrinthine approaches is $6,048.15. The addition of a dural substitute and a resorbable mesh plate following translabyrinthine approaches reduced the CSF leak from 12% to 1.9%, an 84.2% reduction, and a median savings per patient of $2,932.23. Applying our cohort's billing data to previously published cranioplasty techniques, costs, and leak rate improvements following translabyrinthine approaches, all techniques were found to be cost-effective.

Conclusions: Resorbable mesh cranioplasty is cost-effective at reducing CSF leaks following translabyrinthine approaches. Per our billing data and achieving the same CSF leak reduction, cranioplasty costs exceeding $5,080.32 are not cost-effective.

Define Professional Practice Gap & Educational Need: Minimal literature has discussed costs associated with post-operative CSF leaks. It is imperative for physicians to appreciate such costs as the American healthcare system shifts towards a bundled payment system.

Learning Objective: To describe costs associated with CSF leaks and cranioplasty techniques used to reduce postoperative CSF leak formation.

Desired Result: Attendees will appreciate costs of postoperative CSF leaks and cranioplasty techniques that can drastically reduce both the incidence and costs of postoperative CSF leaks.

Indicate IRB or IACUC Approval: Approved

Corneal Complications after Lateral Skull Base Surgery

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Matthew Ehrenburg; Heather Weinreich, MD
Howard W. Francis, MD

Objective: To analyze the rate of corneal complications after lateral skull base surgery, and the relative risk of each potential contributing factor.

Study Design: Retrospective cohort study

Setting: Tertiary care center

Patients: Adult patients who had undergone lateral skull base surgery involving an otolaryngologist at our institution from 2007 to 2015

Intervention: none

Main outcome measure: Relative risk (RR) for each potential contributing factor to corneal complications

Results: 469 patients met inclusion criteria. Of those, 35 developed mild exposure keratopathy, 13 developed moderate exposure keratopathy, and 5 developed severe exposure keratopathy. Age, gender, prior eye surgery, tumor side, and pathology were not significant predictors of keratopathy. Tumor size greater than 30 mm (RR 4.75), post-operative trigeminal palsy (RR 3.42), post-operative abducens palsy (RR 9.08), House-Brackman score 5-6 (RR 4.77), lagophthalmos (RR11.85), ectropion (RR 4.29), prior eye disease (RR 1.83), and an anatomically non-intact facial nerve (RR 3.95) were all significantly associated with the development of corneal complications. On multivariate analysis, lagophthalmos and the presence of an abducens palsy were independent predictors of keratopathy.

Conclusions: There are several important risk factors for exposure keratopathy after lateral skull base surgery, and knowledge of these risk factors can help identify high risk patients in whom early, aggressive preventative therapy is warranted.

Indicate IRB or IACUC Approval: Approved

Endoscopic-Assisted Repair of CSF Otorrhea and Temporal Encephaloceles via Keyhole Craniotomy

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Norman Chan, MD; Vishad Sukul, MD
Kadir Erkmen, MD

Objective: Temporal lobe encephaloceles and cerebrospinal fluid otorrhea from temporal bone defects that involve the tegmen tympani and mastoideum are general repaired using middle fossa, mastoidectomy, or combined approaches. Standard middle fossa craniotomy exposes patients to dural retraction which may lead to postoperative neurologic complications. Here we describe novel approach using endoscope visualization through a keyhole middle fossa craniotomy to repair tegmen defects.

Study design: Retrospective case review of a series of patients treated for temporal encephaloceles or cerebrospinal fluid originating from defects in the tegmen utilizing an endoscopic-assisted middle fossa keyhole craniotomy approach.

Setting: Tertiary referral center.

Patients: Patients included in the study underwent endoscopic-assisted or fully endoscopic repairs of temporal encephalocele and/ or cerebrospinal fluid otorrhea originating from a defect in the tegmen. Only adult patients were included. Patients of multiple ethnicities and body mass indices were included.

Intervention: Endoscopic-assisted or fully endoscopic middle fossa repair of tegmen dehiscence through a keyhole craniotomy approach.

Main outcome measure: Recurrence of cerebrospinal fluid otorrhea or temporal encephalocele.

Results: All cases were performed successfully using a keyhole craniotomy with endoscopic visualization and minimal retraction. There were no recurrences of encephaloceles or cerebrospinal fluid otorrhea in these patients. Additionally, surgical times did not increase. There were no major postoperative complications within this series.

Conclusions: Endoscopic visualization allows for smaller incisions and craniotomies and less risk of brain retraction injury without compromising repair integrity during temporal lobe encephalocele and tegmen repairs.

Define Professional Practice Gap & Educational Need: lack of knowledge of added advantage of use of endoscopy for visualization for care of temporal encephaloceles and cerebrospinal otorrhea

Learning Objective: Introduction of the technique of endoscopic visualization of the tegmen via a middle fossa keyhole craniotomy approach in the surgical treatment of temporal lobe encephaloceles and cerebrospinal fluid otorrhea

Desired Result: Attendees will understand the technique and advantages of the use of endoscopic visualization of the tegmen in the surgical treatment of temporal lobe encephaloceles and cerebrospinal fluid otorrhea

Indicate IRB or IACUC Approval: Approved

Middle Cranial Fossa (MCF) Approach for the Management of Spontaneous Cerebral Spinal Fluid (CSF) Leaks

Rick F. Nelson, MD PhD; Joseph P. Roche, MD
Bruce J. Gantz, MD; Marlan R. Hansen, MD

Objective: To determine the efficacy and morbidity of repairing spontaneous CSF leaks with the MCF approach without the use of a lumbar drain (LD), as perioperative use of LD remains controversial.

Study Design: Retrospective review from 2008-2015

Setting: Two university academic centers

Patients: Those with lateral skull base spontaneous CSF leaks and/or encephaloceles.

Intervention: MCF approach for repair of spontaneous CSF leak and/or encephalocele

Main Outcome Measure: Spontaneous CSF leak patient characteristics (age, sex, BMI, obstructive sleep apnea) were collected. Length of stay (LOS), post-operative complications, CSF leak rate, and need for LD were calculated.

Results: 25 patients underwent MCF repair. CSF diversion with LD was not used in most of the patients (21 of 25). The average LOS was significantly shorter without the use of a lumbar drain (3.9 vs. 9.5 days). Post-operative complications included transient mental status change (2), meningitis (1) and seizure (1). No patients experienced long-term neurologic sequelae or long-term CSF leak recurrence with an average length of follow up of 21.9 months. All patients with spontaneous CSF leaks were overweight (BMI >25 kg/m2) with an average BMI of 38.8 +/- 8.6 kg/m2. The average age was 58.1 +/- 9.1 years and 56 % were female. Most patients had obstructive sleep apnea (15 of 25).

Conclusions: The morbidity of the MCF craniotomy for repair of spontaneous CSF leaks is low and the efficacy of repair is high without the use of lumbar drain. Obesity and obstructive sleep apnea are highly associated with spontaneous CSF leaks.

Define Professional Practice Gap & Educational Need: The use of CSF diversion with lumbar drain remains controversial in the repair of spontaneous CSF leaks. There is a need to understand the morbidity and efficacy of surgical management of spontaneous CSF leaks

Learning Objective: To demonstrate the low morbidity and high efficacy of MCF repair for spontaneous CSF leaks without the use of a lumbar drain.

Desired Result: Physicians who manage lateral spontaneous CSF leaks should consider the middle fossa craniotomy as a safe and effective. CSF diversion is often not required for successful repair of spontaneous CSF leaks.

Indicate IRB or IACUC Approval: Approved

Visualization of Vestibular Structures using Optical Coherence Tomography in Mouse Models

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Akiko Taura, MD, PhD; Shin-ichiro Kitajiri, MD, PhD
Takayuki Nakagawa, MD, PhD; Juichi Ito, MD, PhD
Koichi Omori, MD, PhD

Hypothesis: Optical Coherence Tomography (OCT) is an effective tool to visualize the internal structures of the vestibular systems in mouse models

Background: OCT is an imaging modality that utilizes near infrared light as an imaging medium. The strength of OCT is in the extremely high resolution. OCT has been utilized to visualize normal and abnormal internal structures of rodent cochleae through the cochlear bony capsules in vivo. However the feasibility of OCT for vestibular systems has not been elucidated yet.

Methods: ICR and Slc26a4 knock-out mice at the age of postnatal day 1 and 105 were used. After the removal of the inner ear and fixation with 4% paraformaldehyde, near infrared light was applied from the cranial side of the vestibule to image the internal structures. The inner ears were then prepared for paraffin sections, and stained with hematoxylin and eosin.

Results: OCT effectively demonstrated granules of otoconia, as strong signals in the vestibule of P1 and adult ICR mice. After decalcification, signals of otoconia showed attenuation, and deeper structures such as the stapedial footplate could be visualized. Giant otoconia in the utricle and saccule were observed in Slc26a4 knock-out mice.

Conclusion: OCT is a useful modality for inspection of the internal morphology of normal and abnormal vestibular structures in mouse models.

Define Professional Practice Gap & Educational Need: Lack of contemporary knowledge for the effectiveness of optical coherence tomography applied to the mouse vestibule.

Learning Objective: To reveal the ability and limitation of optical coherence tomography for use in the mouse vestibular structures, including otoconia and membranous labyrinth.

Desired Result: Audience will recognize the effectiveness of optical coherence tomography for the vestibular systems and hopefully it will be applied further for the basic research of vestibules and development of the imaging device for clinical application.

Indicate IRB or IACUC Approval: Approved

The Efficacy of Color Mapped Fusion Imaging in the Postoperative Follow-up Evaluation for Residual Cholesteatomas

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Toshinori Kubota, MD, PhD; Masafumi Kanoto, MD, PhD
Seiji Kakehata, MD, PhD

Objective: To assess the efficacy of color mapped fusion imaging (CMFI) in evaluating shadows found on CT scans during postoperative follow-up evaluations for residual cholesteatomas in patients whom had undergone surgery for removal of primary acquired cholesteatomas.

Study Design: Prospective case study

Setting: Single university hospital

Patients: Ninety patients who had undergone surgery for removal of primary acquired cholesteatomas and were undergoing postoperative follow-up evaluations for residual cholesteatomas at 6-month intervals.

Intervention: Patients underwent a CT scan. If shadows were found suggesting the presence of a residual cholesteatoma, CMFI was performed to determine if the shadows were a cholesteatoma.

Main Outcome Measure(s): Shadows were found on the CT scan in 68 of 90 patients. The presence of a residual cholesteatoma was strongly suggested in 5 of the 68 patients based on CMFI and these 5 patients all underwent additional surgery. The CMFI evaluations for these patients were compared to intraoperative findings.

Results: All 5 patients were found to have a residual cholesteatoma in the same anatomical location as indicated by CMFI and these cholesteatomas were all successfully removed. CMFI facilitated accurate and immediate detection of the cholesteatoma anatomical location in contrast to a CT scan. A CT scan alone requires waiting until the next 6-month follow-up evaluation to determine whether the shadow is a cholesteatoma based on its growth. Thus all residual cholesteatomas were removed at the earliest possible stage in the postoperative follow-up evaluation process.

Conclusions: CMFI is a reliable diagnostic modality for postoperatively identifying early-stage residual cholesteatomas.

Define Professional Practice Gap & Educational Need: To spread awareness of the newly developed CMFI which facilitates accurate identification of early-stage residual cholesteatomas.

Learning Objective: To demonstrate that CMFI is a reliable diagnostic modality for not only preoperatively identifying cholesteatomas but also postoperatively identifying early-stage residual cholesteatomas.

Desired Result: CMFI will be widely applied to all types of cholesteatomas to more precisely determine the anatomical location of cholesteatomas at an earlier stage than previously possible. These improvements will also facilitate determination of whether a patient is indicated for transcanal endoscopic surgery (TEES) in the treatment of such cholesteatomas.

Indicate IRB or IACUC Approval: Approved

Effects of Large-dose Steroid Administration in Bell's Palsy

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Objective: Recent large-scale investigations have not been conducted on the efficacy of large-dose steroid administration of prednisone (PSL) for Bell's palsy. We compared treatment results between normal dose steroid (PSL 1 mg/kg/day) and large-dose steroid (PSL 200 mg/day) administration.

Study Design: Retrospective case review

Setting: Tertiary referral center

Patients: A total of 923 patients with Bell's palsy were treated in our department between 1995 and 2014. These patients could be divided into a normal dose group of 355 and large-dose group of 568 patients.

Methods: We separately assessed treatment outcomes for the three groups of H-B grade V patients, H-B grade IV patients and all patients. Logistic regression analysis was also performed to investigate factors which can impact treatment outcomes, i.e. gender, age, days to start of treatment, PSL dosage and antiviral drug administration.

Results: Recovery rates were significantly better in the PSL 200 mg/day group in comparison with the PSL 1 mg/day for H-B grade V (83.5% vs. 96.2%) H-B grade IV (77.7% vs. 100%) and all patients (68.2% vs. 92.5%). Recovery rates were also superior in the PSL 200 mg/day group when an antiviral agent was also administered. Significant factors for treatment outcomes were PSL 200 mg/day administration and early initiation of treatment for grade IV and PSL 200 mg administration for all patients. Insignificant factors were gender, age and the antiviral agent.

Conclusion: We showed the PSL 200 mg/day administration produced significantly better outcomes than PSL 1 mg/kg/day administration in the treatment of patients with Bell's palsy.

Define Professional Practice Gap & Educational Need: Lack of large-scale investigation about efficacy of large-dose steroid (PSL) administration for Bell's palsy.

Learning Objective: We showed large-scale investigation about efficacy of large-dose steroid (PSL) administration for Bell's palsy.

Desired Result: To demonstrate that PSL 200 mg administration and early initiation of treatment are superior in terms of treatment outcomes.

Indicate IRB or IACUC Approval: Approved

Investigation of Piezoelectric Sensors for Totally Implantable Otologic Microphones

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Hypothesis: To determine if piezoelectric sensors can measure umbo motion or intracochlear pressure change to function as totally implantable otologic microphones.

Background: Current implantable microphones have functional and design limitations. We have previously reported on the idea of a microphone to be imbedded in a cochlear implant electrode array. Additionally, we have been exploring feasibility of sensing umbo motion. Here we report resolution, sensitivity and limitations of our piezoelectric microphone designs.

Methods: An extended facial recess was created in fresh human cadaveric temporal bones, and sound pressure was applied to the ear canal. Intracochlear pressures were measured with a piezoelectric sensor in scala tympani. Umbo motions were measured with a piezoelectric device above the cochlear promontory. Both piezoelectric sensors were constructed of polyvinylidene fluoride (PVDF) material. Ear canal pressure, stapes velocity and umbo velocity were concurrently measured.

Results: Both intracochlear pressure and umbo motion were successfully recorded with our sensors, with minimal effect on ossicular motion. These sensor measurements were similar to the ear-canal pressures across a broad frequency range.

Conclusions: Sensing umbo velocity and intracochlear pressures are feasible for implantable microphones. Future studies will focus on refining these designs to increase sensitivity, improve signal to noise ratio, improve the design for surgical ease, and ultimately incorporate the design for use with active middle ear devices and cochlear implants.

Define Professional Practice Gap & Educational Need: Lack of contemporary knowledge of methods to create fully implantable otologic microphones

Learning Objective: To understand the current possibilities for the use of piezoelectric sensors as fully implantable otologic microphones

Desired Result: Attendees will be able to apply knowledge of different technologies for fully implantable microphones to future research directions in active middle ear implants and fully implantable cochlear implants

Indicate IRB or IACUC Approval: Approved

Modification of Osseointegrated Device Parameters to Improve Speech in Noise and Localization Ability: Clinical Recommendations

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Objective: To determine how best to modify osseointegrated (OI) device or environmental settings to maximize hearing ability.

Study Design: Prospective cohort study
Setting: Tertiary referral center

Patients: 13 adults with single-sided deafness (SSD), normal contralateral hearing, and a minimum of 6 months OI usage

Interventions: Speech in noise (SIN) and localization were assessed in a multi speaker array (R-Space) with patients repeating sentences embedded in competing noise and verbally indicating the source speaker, respectively.

Main Outcome Measures: SIN and localization were assessed with multiple OI microphone settings (adaptive, fixed-directional and omnidirectional) as well as an unaided (i.e., OI off) condition. Participants completed the Abbreviated Profile of Hearing aid Benefit.

Results: Localization performance remains compromised for OI users with a high number of front-back confusions, but fixed-directional microphone settings improve side angle localization ($p<0.01$). SIN performance is greatly enhanced with speech presented to the contra hearing ear (+6dBSNR; $p<0.001$). Subjective report of hearing ability is highly predictive of objective localization measures ($p<001$).

Conclusions: Clinicians should consider implementing a fixed-directional microphone setting for improved localization performance. For better hearing in noise, clinicians should counsel OI recipients to orient the speech signal to their normal hearing ear rather than their OI device.

Define Professional Practice Gap & Educational Need: 1. Lack of clinician awareness for maximizing osseointegrated device performance with device or environmental settings. 2. Variation in practice patterns for maximizing hearing in noise and sound localization ability for osseointegrated device users.

Learning Objective: 1. To determine how to maximize osseointegrated device performance with device and environmental settings. 2. To determine how to maximize hearing in noise and sound localization ability for osseointegrated device users.

Desired Result: Clinicians will know how to maximize hearing in noise and sound localization ability for osseointegrated device users by appropriately adjusting device and environmental settings.

Indicate IRB or IACUC Approval: Approved

Management of Mal de Debarquement Syndrome as Vestibular Migraines

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Objective: Mal de debarquement syndrome (MdDS) is a balance disorder which typically starts after an extended exposure to passive motion such as boat and plane rides. Management is typically supportive (e.g. physical therapy), and symptoms that persist beyond six months have been described as unlikely to remit. This study was conducted to evaluate the response of MdDS to management with migraine prophylaxis, including lifestyle changes and medical therapy.

Study Design: Prospective review.

Setting: Outpatient clinic, tertiary medical center

Methods: Clinical history, detailed questionnaires and audiograms were used to diagnose patients with MdDS. Those patients with the diagnosis of the MdDs were placed on our institutional vestibular migraine management protocol.

Results: Fifteen patients were diagnosed with MdDS with a predominance of females (11 (73%) female), with a mean age of 50 Â± 13 years. Concurrent diagnosis of vestibular migraine, Meniereâ€™s disease and sinus headaches was present in 11 (73%), 9 (60%) and 13 (87%) of these patients, respectively. Nine patients (60%) responded well to management with a vestibular migraine protocol, which included lifestyle changes, as well as pharmacotherapy with verapamil, nortriptyline or topiramate.

Conclusions: Management of MdDS as vestibular migraine yields successful results in improving patientsâ€™ symptoms and increasing the quality of life. Nearly all the patients suffering from MdDS had a personal or family history of migraine headaches or had signs or symptoms suggestive of atypical migraine.

Define Professional Practice Gap & Educational Need: Sometimes there might be a lack of knowledge about Mal de Debarquement Syndrome (MdDS) and its diagnosis. Also the proper and ideal treatment for these patients still remains a challenging issue.

Learning Objective: To observe response of the patients with MdDS to the medications used for (vestibular) migraine and changes in the quality of life in these patients.

Desired Result: To find out the prevalence of patients with MdDS and also to improve the quality of life in patients with MdDS.

Indicate IRB or IACUC Approval: Approved

Novel Device for Measuring Ossicular Chain Compliance before and after Surgical Repair

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Hypothesis: We have developed a device that is unique in its process of measuring ossicular chain compliance, which we hypothesize can be used intraoperatively both before and after surgical repair to measure the stiffness of the ossicular chain.

Background: Conductive hearing loss is a very common surgically treated entity. However, no commonly accepted objective measure exists to assess stiffness and motion of the ossicular chain intraoperatively. This is a problem both for diagnostic purposes when considering surgical repair, and for assessing the integrity of surgical repair.
Ossicular chains of human cadaver heads with well preserved auditory anatomy were treated with poly(methyl methacrylate) to simulate ossicular fixation. Ossicular chain reconstructions were then performed to restore normal function. Our device was used to measure ossicular chain compliance at each step. Laser Doppler Vibrometry was utilized to confirm normal middle ear function prior to the experiment, to confirm ossicular fixation, and finally to test the integrity of the surgically repaired ossicular chain.

Results: Stapes and lateral chain fixation significantly increased ossicular chain stiffness at all locations tested. After ossicular chain reconstruction, chain compliance tested with our device returned to the "pre-fixation" level.

Conclusion: Early results demonstrate feasibility for assessing changes in stiffness between normal and disease-simulated ears, as well as the ability to measure the improvement of ossicular chain compliance following surgical repair. Further testing is needed to determine its true diagnostic utility in different forms of conductive hearing loss, as well as its predictive capacity following surgical repair of conductive hearing loss.

Define Professional Practice Gap & Educational Need: There is a lack of objective measures for determining ossicular chain stiffness during middle ear surgery. Be it a stapedectomy for otosclerosis, suspected lateral chain fixation, or just a hearing loss that is out of proportion to a small perforation; our only way to approximate ossicular chain compliance is by palpating the chain and relying on the surgeon's hand to sense extremely small forces.

Learning Objective: 1) Understand the basic physiology of the ossicular chain, including the forces on and displacement of the ossicles. 2) Understand how best to measure ossicular chain compliance, and the potential importance in surgical decision making

Desired Result: Attendees may begin to think about the overall subjective nature of a large portion of middle ear surgery, and how their surgical decision making can be made more objective.

Indicate IRB or IACUC Approval: Exempt

Cochlin-tomoprotein (CTP) Detection Test Revealed Idiopathic Perilymphatic Fistula in Patients with Idiopathic Sudden Sensorineural Hearing Loss

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Tomoo Watanabe, MD, PhD; Takatoshi Furukawa, MD, PhD
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Objective: To demonstrate that the presence of the previously ignored idiopathic perilymphatic fistulas (PLF) in patients with idiopathic sudden sensorineural hearing loss (ISSNHL) using a cochlin-tomoprotein (CTP) detection test.

Study design: A prospective case series

Setting: Tertiary referral center

Patients: Twenty-one patients with ISSNHL received intratympanic steroid therapy using dexamethasone (IT DEX) between December 2013 and July 2015.

Intervention: Dexamethasone was injected through a perforation made by laser-assisted myringotomy. IT DEX administration was performed on 8 consecutive days. A lavage was performed on the middle ear through the perforation on the first or second day of treatment to collect samples for the CTP detection test. CTP is defined as present at a value of 0.8 ng/ ml or higher.

Results: The CTP detection test revealed that 5 out of the 21 patients (23.8%) with ISSNHL were CTP positive (0.81 to 3.44 ng/ml, mean 1.5 ng/ml). This result suggests that idiopathic PLF can be a cause of ISSNHL.

Conclusions: Five of the 21 patients with ISSNHL were CTP positive, suggesting that idiopathic PLF may be a causative factor in some cases of ISSNHL. These findings underscore the need for physicians to consider idiopathic PLF in their diagnosis of ISSNHL.

Define Professional Practice Gap & Educational Need: To spread awareness that idiopathic perilympatic fistula can be important cause of idiopathic sudden sensorineural hearing loss.

Learning Objective: To show that idiopathic perilympatic fistula is not a rare but can be an important cause of idiopathic sudden sensorineural hearing loss.

Desired Result: More than 20% of patients with idiopathic sudden sensorineural hearing loss were idiopathic perilympatic fistula.

Indicate IRB or IACUC Approval: Approved

Vestibular Functions in Otitis Media with Antineutrophil Cytoplasmic Antibody (ANCA)-Associated Vasculitis (OMAAV) Patients

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Objective: Otitis media with antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis (OMAAV) is a recently reported novel ear disease, which is characterized by a rapidly progressing, mixed or sensorineural hearing loss with presence of serum ANCA. OMAAV is sometimes accompanied by systemic lesions in the lungs and kidneys, for which steroid plus cyclophosphamide therapy is provided. OMAAV causes profound hearing loss, sometimes bilaterally, and in the absence of immediate treatment, cochlear implants may be needed. Although characteristics of auditory disturbance in patients with OMAAV have been briefly described, there is no report on vestibular dysfunction in these patients. In this study, we investigated vestibular function and the clinical features of dizziness in patients with OMAAV.

Study Design: Retrospective case series.

Setting: University hospital.

Patients: Twenty-eight patients diagnosed with OMAAV.

Main outcome measures: Clinical findings and caloric response.

Results: Nine of the 28 patients (32.1%) complained of dizziness: 1 patient showed acute vertigo with sudden hearing loss and the other 8 patients slowly developed dizziness with the progression of hearing loss. Among these 9 patients, 3 (33.3%) still had dizziness after the treatment. All 9 patients showed absence of vestibular response on caloric testing. Eight patients with OMAAV who had no vestibular symptoms underwent equilibrium function test. Of these, 5 patients showed caloric weakness.

Conclusions: These results suggest that vestibular function is disturbed in almost all patients with OMAAV, but may not be noticed because of gradual progression. However, early treatment such as vestibular rehabilitation is important for symptomatic patients.

Define Professional Practice Gap & Educational Need: Lack of contemporary knowledge of otitis media with antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis(OMAAV)

Learning Objective & Desired Result: To understand of the inner ear function of OMAAV patients OMAAV causes vestibular dysfunction as well as profound hearing loss.

Indicate IRB or IACUC Approval: Approved

A Retrospective Review of Temporal Bone Imaging with Respect to Bone-Anchored Hearing Aid Placement

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Objective: Current bone-anchored hearing aid (BAHA) guidelines recommend placement of the titanium implant 5-7cm posterior to the ear canal. Previous studies show that bone conducted hearing is maximized the closer the transducer is to the cochlea. We aim to demonstrate that BAHA implants may be safely placed closer to the ear canal in patients with chronic ear disease, enhancing the amplification available to the patient.

Study Design: We performed a retrospective review of high-resolution temporal bone CTs, comparing multiple measurements between ears with chronic ear disease and normal controls.

Setting: Images were obtained at a single academic medical center.

Patients: 80 patients (160 ears) with temporal bone CTs performed between 2006 and 2009 were measured. Patients with chronic ear disease were identified by ICD-9 code and confirmation by review of the imaging.

Main Outcome Measures: Measurements were made on axial CT slices from a point 1cm posterior to the sigmoid sinus to the posterior margin of the external canal. The squamous temporal bone thickness was also measured a this point.

Results: 47 patients (55 ears) had chronic ear disease. Distance from the posterior canal was significantly different between normal and diseased ears (36.3mm vs. 33.5mm, $p<0.001$). Squamous temporal bone thickness varied widely, and was similar between groups (6.9mm vs. 6.8mm, $p=.76$).

Conclusions: According to our data, titanium implants for bone-anchored hearing aids may be safely placed closer to the external canal than the current recommendations. This could allow for better transduction as well as sound localization in BAHA patients.

Define Professional Practice Gap & Educational Need: 1. Current guidelines for BAHA placement are based on safety concerns 2. Lack of contemporary data on the safety of alternative locations for placement of BAHA implants, and comparison between normal ears and ears with chronic disease.

Learning Objective: 1. We will show the learner that placement of BAHA implants closer to the ear is safe while avoiding the sigmoid sinus, especially in patients with known chronic ear disease.

Desired Result: The learner will be able to consider placement of BAHA implants closer to the ear, particularly in patients who have known chronic ear disease.

Indicate IRB or IACUC Approval: Approved

Content Validity of Temporal Bone Models Printed Via Inexpensive Methods and Materials

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Hypothesis: The 3D printed models will be within 15% accuracy of the scans of the cadaveric temporal bones.

Background: Previous studies have evaluated the face validity of 3D-printed temporal bone models designed to train otolaryngology residents. The purpose of the study was to determine the content validity of temporal bone models printed using inexpensive printers and materials.

Methods: Four cadaveric temporal bones were randomly selected and clinical temporal bone CT scans were obtained. Models were generated using previously described methods in acrylonitrile butadiene styrene (ABS) plastic using Makerbot Replicator 2x and Hyrel printers. Models were radiographically scanned using the same protocol as the cadaveric bones. Four images from each cadaveric CT series and four corresponding images from the model CT series were selected, and voxel values were normalized to black or white. Scan slices were compared using PixelDiff software. Gross anatomic structures were evaluated in the model scans by 4 board certified otolaryngologists on a 4 point scale.

Results: Mean pixel percent difference between the cadaver and model scans was 16.89% ± 6.8%. Mean cortical bone width difference and mean external auditory canal width difference were 0.58 ± 0.66 mm and 0.55 ± 0.46 mm respectively. Expert raters felt the mastoid air cells were well represented (2.5 ± 0.5), while middle ear and otic capsule structures were not accurately rendered (all averaged <1.8)..

Conclusion: These results suggest that these models would be sufficient adjuncts to cadaver temporal bones for training residents in cortical mastoidectomies, but less effective for middle ear procedures.

Define Professional Practice Gap & Educational Need: Lack of knowledge about the internal structures of printed temporal bone models

Learning Objective: Identify why printed temporal bone models are useful. Describe a novel method to assess the accuracy of the printed temporal bone. Explain why portions of the model are poorly rendered by the printing process.

Desired Result: Attendees will be able to assess the accuracy of models regardless of the method used to create them.

Indicate IRB or IACUC Approval: Exempt