

Superior Semicircular Canal Dehiscence Size and Its Relationship with Clinical Symptoms

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Objective: To define the relationship between patients' clinical presentation of superior semicircular canal dehiscence (SCD) and the size of the dehiscence.

Study design: Retrospective review.

Setting: Tertiary care neurotologic clinic.

Patients: Clinical symptoms and signs as well as multiplanar CT scans of the temporal bones were analyzed in 51 patients who were diagnosed with SCD syndrome. The ages of the patients ranged from 13 – 68 years (mean 45.6 years).

Main outcome measures: The SCD length was measured in the plane of the canal using high-resolution temporal bone CT scans. The height of the residual lumen of the superior canal was measured as the length of a perpendicular line drawn from a chord connecting the two ends of the dehiscence to the inner surface of the canal lumen. Clinical signs and symptoms were evaluated as either absent or present. Vestibular-evoked myogenic potentials (VEMPs) were also measured.

Results: SCD length in symptomatic ears ranged from 0.6 to 6.1 mm (mean 3.3 +/- 1.4 mm STDEV). Dehiscence size correlated with the presence of sound-induced vertigo and sound-evoked nystagmus ($p < 0.05$). A smaller residual lumen also correlated with sound-evoked nystagmus ($p < 0.05$). We did not see significant correlation between dehiscence size and the presence of chronic dysequilibrium, autophony, pulsatile tinnitus, or valsalva or pressure-evoked nystagmus. The larger the dehiscence, the lower the VEMP threshold ($\rho = -0.57$, $p < 0.001$).

Conclusions: Precise measurement of SCD can be made on imaging with multiplanar analysis. Larger dehiscences are more likely associated with sound-evoked vertigo and nystagmus as well as lower VEMP thresholds.

Superior Semicircular Canal Dehiscence: A Clinical and Radiographic Survey

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Objective: To determine the incidence of clinical and radiographic superior semicircular canal dehiscence in the general population, as well as in those undergoing temporal bone imaging.

Study Design: Retrospective patient series

Setting: Tertiary referral center

Patients: All patients who underwent sinus or temporal bone imaging using our Xoran MiniCAT scanner.

Interventions: All sinus and temporal bone scans were screened for superior semicircular canal dehiscence. Coronal images were used for the initial screen, followed by isotropic on-axis reconstructions if dehiscence was suspected. The length of dehiscence was measured if present. Charts of patients with radiographic dehiscence were reviewed for audiometric data, history of vertigo, and findings on pneumatic otoscopy. If such data were not available, patients underwent audiometric evaluation and an in-office consultation.

Main Outcome Measures: Length of dehiscence, audiometric data, history of vertigo, pneumatic otoscopy.

Results: In our ongoing screen, 230 sinus and 137 temporal bone scans have been examined. Among the sinus scans, 24 individuals (10.4%) and 27 ears (5.9%) were found to be dehiscent, with 3 individuals having bilateral involvement. Among the temporal bone scans, 19 individuals (13.9%) and 23 ears (8.4%) were found to be dehiscent, with 4 individuals having bilateral involvement. A total of seven ears displayed dehiscence less than 2mm in length. A high percentage (>40%) of patients suspected clinically of having superior canal dehiscence were found to have radiographic dehiscence on temporal bone imaging. Additional clinical data are pending at this time. We anticipate screening over 1000 patients by the conclusion of the study.

Conclusions: Our Xoran sinus CT database is proving useful for determining the incidence of superior semicircular canal dehiscence in the general, non-otologic population. Our temporal bone CT database is likewise useful for determining the incidence in a neurotologic practice, as well as among patients suspected of having dehiscence. We hope to demonstrate a minimum length of dehiscence for hearing loss or vertigo to occur. IRB: Pending None

Vestibular Evoked Myogenic Potential Induced B Stimuli of Bone Conduction in Patients with Unilateral Chronic Otitis Media

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Objective: We previously reported that vestibular evoked myogenic potential (VEMP) induced by stimuli of bone-conduction (B-VEMP) could evaluate vestibular function in patients with normal hearing (Miyamoto, et al. Otol Neurotol, 2006 (1)). The present paper investigated B-VEMP in patients with conductive hearing loss due to perforated chronic otitis media (COM).

Study design: Prospective study.

Setting: University Hospital.

Patients: Subjects were 52 patients with unilateral COM-induced conductive hearing loss. The vertigo group consisted of 26 patients and the non-vertigo group consisted of 26 patients. For the control group, 35 normal healthy volunteers were used.

Intervention: Each subject was examined by B-VEMP according to the procedure reported in (1).

Main outcome measures: Results of B-VEMP were evaluated by inter-aural ratio (IAR) and compared with the presence of vertigo/dizziness in each subject.

Results: There were no significant differences between the mean IAR of the non-vertigo group ($1 \pm 14\%$) and control group ($1 \pm 21\%$) ($p > 0.05$; Mann-Whitney's U test). Abnormal results in B-VEMP were not demonstrated in any of the non-vertigo group but in 9 patients (34.6%) of the vertigo group ($p = 0.0008$; Fisher's Exact Test). The COM-affected ear demonstrated a lower response than the contralateral intact ear in all COM cases.

Conclusions: The B-VEMP findings indicated high specificity in the presence of vertigo in patients with unilateral COM.

Key words: VEMP - Bone conduction - Chronic otitis media - specificity

IRB: This kind of study is not required in Japan

Transmastoid Superior Semicircular Canal Occlusion

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BACKGROUND: The traditional surgical repair for superior semicircular canal dehiscence (SSCD) involves either canal plugging or resurfacing via the middle cranial fossa approach. A novel transmastoid approach for occlusion of the superior semicircular canal is presented.

RESULTS: Two patients with SSCD underwent surgical occlusion of the superior semicircular canal via the transmastoid approach. The first patient was a primary case of SSCD and the second patient had failed a previous middle fossa occlusion using fascia at an outside institution. It was noted in this failure that the fascia plug was mobile and a permanent occlusion had not been achieved. Occlusion was achieved by using bone pâté, formed from a mix of bone dust and fibrin glue, which allowed a permanent bony partition to be achieved between the dehiscence and the remainder of the labyrinth. In both cases, hearing was preserved and the procedure was successful in controlling symptoms. During the presentation, intraoperative videos will be shown to describe the details of the procedure.

CONCLUSION: In the treatment of SSCD, transmastoid superior semicircular canal occlusion is a viable alternative to traditional middle fossa techniques. Otologic surgeons inexperienced in traditional techniques will benefit from the ease and familiarity of the transmastoid exposure. The approach has minimal morbidity and obviates the need for a craniotomy with temporal lobe retraction. Finally, the use of bone pâté to create a bony partition may be more reliable than standard occlusion techniques utilizing fascia.

PI3-kinase/AKT Pathway Activation in Human Vestibular Schwannoma

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Hypothesis: The *Neurofibromatosis 2 (NF2)* gene, which encodes the tumor suppressor protein merlin, is frequently mutated in vestibular schwannomas (VS). Merlin can inhibit phosphatidylinositol 3-kinase (PI3-kinase) by binding to PI3-kinase enhancer long isoform (PIKE-L), a brain-specific GTPase. Therefore, we hypothesize that the PI3-kinase/AKT pathway is activated in VS.

Background: Despite advances in diagnosis and treatment, VS continue to cause significant hearing and balance dysfunction. Better understanding of the signaling pathways deregulated in VS will aid in developing novel medical therapeutics. Activation of the PI3-kinase/AKT pathway, resulting in increased cell survival and proliferation, has been detected in a variety of human cancers. However, no group has demonstrated PI3-kinase/AKT activity in VS.

Methods: cDNA microarrays were performed on cultured Schwann cells, 4 VS specimens, and 2 paired normal vestibular nerves. Immunostaining using antibodies to activated phospho-AKT (p-AKT) on 14 VS tissue sections was performed. Western blots using various antibodies to the PI3-kinase/AKT pathways were also conducted.

Results: Microarray data found upregulation of AKT RNA expression in VS compared to normal vestibular nerves. Immunostaining on 14 VS tissue sections demonstrated positive reactivity to p-AKT at both serine-473 and threonine-308. Western blots found that total AKT, p-AKT, PI3-kinase, p-PTEN, p-PDK1, p-FOXO, p-GSK3 β , and p-mTOR were all upregulated in VS.

Conclusion: The PI3-kinase/AKT pathway is activated in VS. Using the quantifiable VS xenograft model that we recently reported, novel inhibitors of the PI3-kinase/AKT pathway may be tested for VS growth inhibition *in vivo*.

Management of Facial Neuroma: Review of 16 Surgical Cases and Literature Review

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Objective: Primary tumors of the facial nerve are rare, representing 1% of all intrapetrous lesions. We analyzed the management and surgical outcomes of 16 patients with multisegment facial neuromas treated at our institution over a 16 year period. We also reviewed the presenting symptoms based on tumor location, facial nerve function, and post-operative hearing results.

Study Design: A retrospective chart review

Setting: Tertiary referral center

Patients: All patients included in the study had surgical management of their facial neuroma. There were 9 females and 7 males. The mean age was 43 years old with a mean follow-up period of 27 months.

Intervention: Surgical excision (n=15) or decompression (n=1) of facial neuroma.

Main outcome measures: Tumor location, presenting symptoms, hearing outcomes, and facial function.

Results: Unilateral hearing loss was present in eight (50%) patients and thirteen (81%) patients had facial paresis as their presenting symptom. All tumors involved multiple segments of the facial nerve and ranged in size from 1-2.5 cm. Fifteen (94%) patients had the tumor completely excised and 1 (6%) patient underwent a needle decompression of the cystic component of the tumor. Nine (56%) patients had involvement of the geniculate ganglion and the tumor involved the labyrinthine and tympanic segments of the facial nerve in seven (44%) patients. Despite multiple types of reconstructive options used, the best recovery of facial function was a House-Brackmann Grade III in 11 (73%) patients. There were no peri-operative complications and no cases of recurrent tumors.

Conclusion: Treatment of facial neuromas depends on the extent of tumor, degree of facial paresis, and hearing function. We advocate complete resection of tumor when facial palsy exists. Patients with normal facial function and hearing may be advised on a more conservative treatment option, such as radiological observation, drainage of any cystic component of the tumor for histological diagnosis, or stereotactic radiotherapy.

IRB: 109293

The Evolution of Surgical Approaches in the Treatment of Petroclival Meningiomas at a Single Center: A Retrospective Review

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Objective: The treatment of petroclival meningiomas remains controversial. Various surgical approaches have been described and proposed for their management. We describe our surgical experience in treating these lesions and how our management methods have changed. **Setting:** Tertiary Referral Center.

Methods: Study Design: A retrospective review of craniotomies done between September 1994 and July 2005 for treatment of petroclival meningiomas was performed. **Patients:** All patients undergoing surgical treatment for petroclival meningiomas were included in the study. Cases were also compared to previously published series from our institution dating back to 1986.

Results: A total of 63 cases were reviewed. Approaches utilized included combined petrosal approaches (39%), standard retrosigmoid (RS) craniotomy with or without some degree of petrosectomy (69%), orbitozygomatic (OZ) craniotomy with or without anterior petrosectomy (8%), and combined OZ-RS approaches (23%). A majority of the combined petrosal approaches were performed prior to 1994 (72%). The percentage of cases with gross total resection was significantly higher in the combined petrosal group (68% vs. 43%) as was the complication rate (64% vs. 36%). There were no deaths in any of the groups. There was no significant difference in progression-free survival at 4.2 years mean follow up (84% vs. 98%). No patients treated with stereotactic radiosurgery developed progression.

Conclusion: The utilization of combined petrosal approaches in the treatment of petroclival meningiomas leads to greater rates of complete resection while increasing the rates of morbidity. Progression-free survival remains excellent in both groups. The availability of radiosurgery as an adjunct to therapy has led to a reduction in the utilization of aggressive resection and combined approaches in favor of minimizing patient morbidity.

Acknowledgements:

U. Kumar Kakarla
Louis J. Kim
Peter Nakaji
Randall W. Porter

Adenoid Cystic Carcinoma of the Parotid Gland with Temporal Bone Invasion

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Objective: To present our series of 16 patients with adenoid cystic carcinoma of the parotid gland with temporal bone invasion.

Study design: A retrospective case review.

Setting: Tertiary care, academic medical center.

Patients: All patients treated at our institution between July 1988 and July 2006 with parotid gland adenoid cystic carcinoma with temporal bone invasion.

Interventions: Preoperative radiographic assessment with combined surgical and radiation therapy treatment.

Main outcome measures: Postoperative- or radiation-related complications and overall 2-year, 5-year, and 10-year survival.

Results: The most common surgically related complications were new-onset cranial nerve deficits, while osteoradionecrosis of the bony E.A.C. was the most frequently noted complication associated with RT. The overall survival at 2 years, 5 years, and 10 years was 94%, 75%, and 60%, respectively.

Conclusions: Lateral skull base access should be employed in the extirpation of adenoid cystic carcinoma of the parotid gland with temporal bone involvement.

Experience with a Totally Implantable Cochlear Implant: Initial Clinical Outcomes

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Objectives: To evaluate the effectiveness and issues associated with use of a prototype Totally Implantable Cochlear Implant (TIKI)

Study design: Limited patient trial.

Setting: Tertiary referral centre.

Patients: Three adult human subjects with severe to profound sensorineural hearing loss.

Interventions: A prototype TIKI device has been designed and developed by a research team in Cochlear Limited and CRC HEAR. The TIKI has an inbuilt lithium ion rechargeable battery and a package-mounted internal microphone. The TIKI design enables "Invisible Hearing" using the internal microphone, speech processor and battery power, without any externals. The TIKI device also has similar functionality to the Nucleus 3 cochlear implant system and can be used with an external speech processor and transmitting coil in "Conventional mode", which also serves to recharge the battery. After initial safety studies 3 patients were implanted with the prototype TIKI. The standard surgical technique was modified to accommodate the larger device package. Postoperatively, subjects used the TIKI in both Invisible Hearing and Conventional modes.

Main outcome measures: Device use was recorded in both Invisible Hearing and Conventional modes. Performance of the internal battery and microphone was assessed over time, psychophysical MAP data was collected and speech discrimination was measured at 1, 3, 6, and 12 months.

Results: There were no surgical or postoperative complications. All patients could successfully use both the Invisible Hearing and Conventional function modes. The subcutaneous internal microphone provided reasonable high frequency response, enabling subjects to use Invisible Hearing in situations where they could not normally use their cochlear implant. The rechargeable battery functioned well, with a cycle time indicating that the low power implant design is effective and will deliver long battery life. Speech perception outcomes for all patients showed improvement on their preoperative scores. As expected, speech perception results were significantly better in the Conventional mode than the Invisible Hearing mode, while the subjects also reported body noise interference that limited use of the Invisible Hearing mode. All subjects achieved benefit with their cochlear implant system and continue to use the Invisible Hearing mode on a limited daily basis

Discussion: The challenges in developing a safe and effective totally implantable cochlear implant are considerable. In developing the TIKI prototype, a conservative approach was taken with the microphone and battery being included in the stimulator housing. Specific challenges were: to have a safe, small, long life battery; an effective microphone with good high frequency response; low power electronics; and package size suited to surgical implantation.

Conclusion: Three subjects were implanted with a prototype totally implantable cochlear implant. The hearing outcomes using the Invisible Hearing mode, whilst not as good as the Conventional mode, demonstrate that this prototype provides a sound basis for future development of totally implantable cochlear implant technology.

IRB: 05/593 H

Acknowledgements:

This study was funded by Cochlear Limited through contributions to the Co-operative Research Centre for Cochlear Implant and Hearing Aid Innovation (CRC HEAR)

Revision Cochlear Implant Surgery in Children

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Objective: The purpose of this study is to determine the incidence of revision CI surgery in children, the indications for revision surgery, and to examine the pattern of events that lead to revision CI surgery.

Study Design: Retrospective chart review

Setting: Two large tertiary pediatric cochlear implant programs

Patients: Pediatric cochlear implant recipients who underwent revision surgery related to the cochlear implant from 1990 – 2005.

Interventions: Revision surgery for cochlear implant related reasons.

Main Outcome Measures: The main outcomes of the study were the incidence of revision CI surgery in children, the indications for revision surgery, and to examine the pattern of events that lead to revision surgery.

Results: 961 pediatric CI surgeries were performed during the study period. 92 pediatric patients underwent a total of 104 (10.8%) revision surgeries for CI related issues. The reasons for revision surgery include device failure (46%), medical or surgical causes (38%), and suspected device malfunction (15%). Within the device failure and suspected device malfunction groups, the majority of CIs had defects found on device analysis. Performance after cochlear implantation met or exceeded pre-revision performance in most patients; however, this improvement may be delayed in some patients.

Conclusions: Revision CI surgery accounts for ~10% of surgeries at two large pediatric cochlear implant programs. Device failure and medical/surgical issues are the most common reasons for revision surgery. Suspected device malfunction was present in a significant number of patients. This diagnosis requires a careful scrutiny of symptoms in this patient population.

Clinical Assessment of Music Perception in Cochlear Implant Listeners

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Objective: Cochlear implants (CI) have been immensely beneficial for speech recognition in quiet for severely and profoundly hearing-impaired patients, but CI users have great difficulty perceiving music. The purpose of this study was to develop a test to quantify music perception by CI listeners in clinically practical manner that could be standardized for administration at any implant center.

Study Design: Prospective, case-controlled study.

Setting: Hearing research center at an academic hospital.

Patients: Eight CI listeners, including 5 males and 3 females with implant experience ranging from 0.5 to 6 years, participated in this study. They represented a variety of implant devices and strategies.

Intervention: Music test presented via a loud speaker within a standardized, double-walled sound-treated booth.

Main Outcome Measure: Music perception was assessed using a computerized test comprising pitch direction discrimination, melody identification, and timbre identification. The pitch subtest utilizes a 2-alternative forced choice adaptive procedure to determine a threshold interval for discrimination of complex pitch direction change. The melody and timbre subtests assess recognition of 12 isochronous melodies and 8 musical instruments, respectively.

Results: Testing demonstrated a broad range of perceptual accuracy on all three subtests. Test duration averaged less than 45 minutes. Test-retest reliability was good.

Conclusions: The University of Washington Clinical Assessment of Music Perception (UW-CAMP) is able to efficiently differentiate many levels of music perception in CI users.

IRB: 05-7483-V01

Acknowledgements: The authors are grateful for the dedicated efforts of our listeners.

This work was supported by the University of Washington, the VM Bloedel Hearing Research Center, NIH grant R01-DC007525, and a subcontract of NIH grant P50-DC00242.

Simultaneous Bilateral Cochlear Implantation: Prospective Study In Adults

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Objectives: The primary aim of this in-progress multicenter study is to document bilateral vs. unilateral listening benefits in postlinguistically deafened adults implanted simultaneously with two HiRes 90K cochlear implants. A secondary aim is to assess the general clinical utility of a new methodology for testing cochlear implant recipients which eliminates the need for a sound booth or loudspeaker array.

Study Design: A prospective counterbalanced between- and within-subjects design is used to evaluate bilateral vs. unilateral listening benefits. Subjects are evaluated at 3, 6, 8, and 12 months after implantation.

Setting: Nine tertiary referral centers in the United States.

Patients: Subjects are adults with bilateral severe-to-profound sensorineural hearing loss who meet the candidacy criteria for cochlear implantation.

Intervention: Subjects are implanted with two HiRes 90K devices during the same surgery.

Main outcome measures: Speech perception (in quiet and in noise) and localization accuracy are assessed for each ear alone and both ears together. A unique aspect of the study is use of a direct-connect system for postimplant testing. The system was developed by the House Ear Institute and eliminates the need for a sound booth or a speaker array. Left-ear and right-ear head-related transfer functions appropriate to the selected source location are applied to the selected signal and presented via direct connection to the auxiliary input of the Auria/Harmony sound processor.

Results and Conclusions: Clinical results to date from 13 subjects indicate that bilateral cochlear implantation provides improved sound localization and better speech perception in noise compared with unilateral implantation.

Individual IRB approvals obtained at nine study sites

Replacement of the Cochlear Prosthesis: Indications and Results in Adults

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Objective: To assess the results of cochlear reimplantation surgery and to identify the clinical, audiological and electro-prosthetic characteristics that most accurately predict improved and/or restored auditory performance.

Study Design: Retrospective case series.

Setting: Academic tertiary referral center.

Patients: Adults who underwent revision cochlear implantation (n = 43).

Intervention: Cochlear reimplantation surgery.

Main Outcome Measures: Surgical findings, and audiologic performance.

Results: 43 revision cochlear implant surgeries have been performed between 1993 and 2006.

The indication for revision implantation included soft failures (46%), hard failures (23%), electrode extrusion (12%), facial nerve stimulation (9%), infection (7%), and intern receiver- stimulator migration (2%).

To date, 23 cases have been audiological followed for more than 6 months. Of those, speech perception peak performance improved in 52% of cases, 39% achieved the same peak performance, and only 2 cases (9%) had a decrease in peak performance compared to scores prior to reimplantation. All of the patients operated for electrode extrusion, facial nerve stimulation, infection, and intern receiver- stimulator migration showed improvement in peak performance after reimplantation surgery.

Device analysis failed to reveal any obvious abnormality in 34% of the cases. 26% of the failures were confirmed to be due to device defects, of which failure of hermetic seal was the most common.

Conclusion: Following cochlear reimplantation surgery, auditory benefit is restored in at least 90% of the cases. The possibility of worse hearing makes it imperative to establish a hierarchy of predictors that will guide the decisions regarding the appropriateness of revision surgery.

Difluoromethylornithine (DFMO) Lowers Endocochlear Potential and Distortion Product Otoacoustic Emissions (Dpoaes) in Neonatal Gerbils

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Background: Difluoromethylornithine (DFMO) is a cancer chemoprevention drug that inhibits polyamine synthesis and is ototoxic. Previous experiments have shown that daily injections of DFMO (750 mg/kg) from postnatal 3 through 20 in neonatal gerbils causes mild shifts of 10 to 20 dB in auditory brainstem response thresholds for the frequencies 2000 to 32000 Hz.

Hypothesis: The objectives of the study were to measure DPOAEs and endocochlear potential (EP) in this same animal model. DPOAE levels and EPs were compared for DFMO and control groups of 15 gerbils each.

Methods: On postnatal day 21, animals were anesthetized with ketamine/xylazine. DPOAEs were measured for the geometric mean frequencies 4000 to 16000 Hz at 75 dB SPL. EPs were measured with a glass micropipette electrode filled with 150 mM KCl advanced through a small hole in the basal turn of the cochlea, connected to a high-impedance DC amplifier, measuring the potential difference in mV between the scala media and a subcutaneous reference electrode. Left ears were tested and DPOAE levels were compared between groups using ANOVA, while EP thresholds were compared using students t-test.

Results: There was no difference in DPOAE levels for the two groups at geometric mean frequency 4000 Hz but significantly higher DPOAE levels were found at geometric mean frequencies 8000, 12000, and 16000 Hz for control animals compared with DFMO-treated animals. The mean EPs for DFMO-treated and control gerbils were 53 mV and 72.3 mV, respectively ($p < .0001$).

Conclusion: DFMO lowers endocochlear potential and DPOAEs in neonatal gerbils.

Dexamethasone Perfusion vs. Natural History in Meniere's Disease

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OBJECTIVE: Comparison between five year results of intratympanic dexamethasone perfusion vs. natural history in Meniere's disease.

STUDY DESIGN: Prospective cohort study of Meniere's patients followed up for 5 years post intratympanic dexamethasone perfusion.

SETTING: Tertiary-care hearing and balance center.

PATIENTS: The study included 99 patients diagnosed according to the 1995 AAO-HNS guidelines for the diagnosis of Meniere's disease.

INTERVENTION: Intratympanic injection of dexamethasone 24 mg/ml once weekly for a maximum of 3 treatments. Pre- and post-treatment neurotologic examination and tests of auditory and vestibular function. Follow-up every 3 months for the first year, then annually for 5 years.

MAIN OUTCOME MEASURES: Pre- and post-treatment hearing (pure-tone average and word-recognition score), vertigo frequency and functional levels, and subjective changes in tinnitus and aural fullness. Results were compared to the 5 year natural history of Meniere's disease as documented in neurotology literature. Correlated t test and 95% confidence interval were used for statistical analysis.

RESULTS: Significant gain of 30% speech discrimination was evident in 90% of patients. The five year average WRS was 80%. Vertigo frequency and functional levels were significantly improved in 90% of patients. Aural fullness and tinnitus subsided significantly in 90% and 60% of patients, respectively. No significant adverse reactions were reported by patients. Outcome is significantly superior to the reported natural history of Meniere's disease.

CONCLUSION: Dexamethasone 24 mg/ml intratympanic perfusion is an effective treatment for Meniere's disease. The treatment is associated with significant speech discrimination recovery and vertigo control by comparison to the natural history of the disease.

Key Words: Inner ear perfusion, intratympanic perfusion, Meniere's disease, dexamethasone, speech discrimination recovery, natural history of Meniere's disease

Longitudinal Results with Intratympanic Dexamethasone in the Treatment of Meniere's Disease

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Objective: To assess the success of intratympanic dexamethasone in controlling vertigo in patients with Ménière's disease and to determine the rate of failure requiring ablative interventions.

Study design: Retrospective study.

Setting: Tertiary referral center.

Patients: 129 subjects with Ménière's disease.

Intervention(s): Intratympanic dexamethasone injections as needed to control their vertigo attacks.

Main outcome measure(s): A Kaplan-Meier time-to-event method was used to determine the rate of "survival," meaning sufficient satisfaction with vertigo control that the subject did not wish to have subsequent ablative treatment. "Failure" was defined as poor control and the choice to proceed to ablative treatment.

Results: Acceptable vertigo control ("survival") was achieved in 117 of 129 subjects (91%), most of whom required 3-4 injections per year. Of 12 failures (9%), 9 occurred within 6 months of the first intratympanic dexamethasone injection. Follow-up for more than 2 years was available for 87 subjects. Of these, 62 (70%) required no further injections after 2 years, 23 (26%) continued to receive intratympanic dexamethasone injections, and 3 (3%) chose intratympanic gentamicin treatment.

Conclusions: Intratympanic dexamethasone injection therapy on an as-needed outpatient basis can provide satisfactory vertigo control in most patients with Ménière's disease. The traditional method of comparing vertigo rates before and for 2 years after treatment does not adequately address this growing clinical practice. Instead, the Kaplan-Meier method provides a quantitative means of assessing vertigo control using the patient's decision whether or not to proceed with more destructive treatment.

A Novel Controlled Local Drug Delivery System for Inner Ear Disease

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Much interest has been generated in developing a system for local drug delivery to the inner ear for the treatment diseases, such as Meniere's disease and sudden idiopathic or noise-induced hearing loss. Studies in recent years demonstrate the pharmacokinetics of the inner ear is influenced by the route of administration. We are developing a novel biodegradable system we hypothesize can continuously deliver, in a single application to the middle ear, a controlled dose course to the inner ear of drugs such as dexamethasone and gentamycin.

We developed a chitosan hydrogel which is prepared in a liquid state at room temperature. At body temperature, it undergoes a phase transition becoming solid. Drug is released from the hydrogel as the hydrogel degrades. Randomized, controlled in vivo studies were performed in our mouse model. The hydrogel system in combination with dexamethasone or gentamycin was placed at the round window niche of the tympanic cavity. By collecting perilymph at different time points and analyzing the samples with high performance liquid chromatography and mass spectrometry we characterized the drug release of our drug delivery system.

Our novel hydrogel delivery system provides sustained and controlled drug release curves of dexamethasone. Our data reveals this novel delivery system can deliver dexamethasone, continuously, into the inner ear through the round window membrane as evidenced by the detection of dexamethasone in the perilymph fluid.

This novel system provides a unique approach for sustained local drug delivery for inner ear therapy.

IRB: N/A IACUC: 801487

Acknowledgements: University of Pennsylvania School of Medicine Department of Pharmacology

Feasibility of Auditory Cortical Stimulation for the Treatment of Tinnitus

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Christina Runge-Samuelsen, John L. Ulmer, Brian H. Kopell

Objective: To test the safety and feasibility of auditory cortex stimulation for the treatment of tinnitus.

Study Design: Prospective controlled single-blinded study followed by open label stimulation period.

Setting: Tertiary care referral center.

Patients: Adults (n=8) with constant tinnitus for greater than one year scoring greater than 33 on the Tinnitus Reaction Questionnaire. Tinnitus was predominantly unilateral and the frequency of tinnitus was below 8,000Hz.

Interventions: Surgical implantation of an investigational extradural two contact electrode over the auditory cortex using fMRI image guidance. A two week stimulation period alternated with a two week sham stimulation period in random order to which the subjects were blinded. This was followed by continuous stimulation with adjustment of parameters to maximize tinnitus suppression.

Main Outcome Measure: Subjective rating of tinnitus severity and loudness as well as device efficacy (0-100 scale). Subjects also underwent repeat audiometric evaluation and re-application of the Tinnitus Handicap Questionnaire, Tinnitus Reaction Questionnaire and Beck Depression Scale.

Results: Response of tinnitus to auditory cortical stimulation appears to be time dependent. No sudden changes in tinnitus were noted within the 4-week crossover period but significant declines in tinnitus loudness were found for several patients with continuous stimulation. Two subjects experienced periods of total tinnitus suppression lasting from 24-48 hours. Scores for tinnitus and depression questionnaires mirrored improvements in subjective ratings of tinnitus severity.

Conclusions: Electrical stimulation of the auditory cortex for the suppression of tinnitus is feasible and appears safe. Further studies will investigate tinnitus characteristics that may predict greatest response.

IRB: HRRC # 304-05 FMLH# 05-139

Evaluation of Facial Function with a Questionnaire: A Validation Study

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Objectives: To determine the validity and inter-observer reliability of a patient administered questionnaire in the evaluation of long-term facial function after vestibular schwannoma (VS) surgery.

Study Design: Validation and inter-observer reliability study of a patient administered questionnaire of facial nerve function.

Methods: 34 patients had an office evaluation of facial function at least one year after microsurgical resection of a VS. 30 patients had a sporadic VS, 4 patients had NF2. The average tumor size was 2.2cm.

A post-operative questionnaire regarding facial function was completed within two months of the office evaluation. The office evaluation was considered the standard by which to evaluate the validity of the questionnaire. Six physicians independently graded the questionnaires to determine each patient's facial function, using the House-Brackmann (HB) facial grading scale. The physicians were blinded to the patients' identities and clinical evaluations of facial function. Statistical analysis was performed to compare the office evaluation rating with the questionnaire rating.

Results: The correlation between the office evaluation and the questionnaire was highly significant (0.01 level) with a Spearman's rho = 0.915. The inter-observer reliability was high, with coefficients ranging from 0.814-0.996 (0.01 level).

There were a large proportion of patients with normal (HB grade 1) facial function. As this may be an "easy" rating based on the questionnaire, an additional analysis was done with this patient sample excluded. The high levels of correlation and inter-observer reliability persist.

Conclusions: When compared to office evaluation, a patient administered questionnaire provides an accurate and reliable measure of long-term facial function after VS surgery.

Steroid Responsive Auditory Neuropathy/Dys-Synchrony is Suggestive of CNS Pathology

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Objective: To review case series of patients with AN/DS due to CNS disturbance.

Study Design: Retrospective case review

Setting: Tertiary referral center

Patients: Two children presenting with steroid responsive acute sensorineural hearing loss

Interventions: Audiometric studies, Otoacoustic emissions, Auditory brainstem response, MRI imaging

Background: Auditory Neuropathy / Dys-Synchrony (AN/DS) is a disorder defined by sensorineural hearing loss associated with recordable otoacoustic emissions but absent or grossly abnormal auditory brainstem responses. Recent focus on pathophysiology has shifted from abnormality of the auditory nerve to dys-synchrony within the cochlea. This case series highlights the continued need to be vigilant of CNS pathology as the etiology of AN/DS specially if the SNHL is steroid responsive.

Main outcomes measured: Pure tone audiometry, speech recognition testing, OAE, ABR, MRI imaging

Outcome: Two cases of auditory neuropathy produced by disease of the CNS were identified: an intrinsic neoplasm of the middle cerebellar peduncle (juvenile pilocytic astrocytoma) and multiple sclerosis affecting the brainstem. Both patients initially presented with acute sensorineural hearing loss and speech recognition testing worse than expected for the degree of hearing loss. Interestingly, both patients demonstrated steroid responsiveness. Definitive treatment consisted of gross total resection for brainstem mass, and cyclophosphamide immunomodulation for the demyelinating disorder. The complete case histories, including audiologic, radiologic and pathologic findings will be reviewed.

Results: Both cases of steroid responsive SNHL fitting AN / DS criteria were of central etiology

Conclusion: AN/DS responsive to steroid therapy may be due to CNS pathology. Therefore, it is crucial to obtain brain MRI with contrast enhancement in all patients with AN/DS. This is especially critical in patients undergoing cochlear implantation as MRI may be contraindicated postoperatively.

H#12300

Electrode Migration after Cochlear Implantation: Incidence and Solutions

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Objective: To review the occurrence of electrode extrusion following cochlear implantation

Study Design: Retrospective case review

Setting: Tertiary academic referral center, ambulatory

Patients: Retrospective review of patients in the U.S. Food and Drug Administration MAUDE(Manufacturer and User Facility Device Experience) database and all patients whose cochlear implants were performed at our center using the 'Split Bridge' technique between 1996 and 2006

Intervention: Cochlear implantation

Main outcome measure: Incidence of electrode migration

Results: During 2001-2006, 116 reports of electrode migration were filed in the MAUDE database. Of the eight hundred cochlear implant surgeries using the 'Split Bridge' technique performed at the University of Miami, only one case of electrode migration was identified.

Conclusions: Although electrode migration is a known complication of cochlear implantation, three isolated cases have been reported in the literature during the past 10 years. The substantial numbers detailed in the MAUDE database demonstrate that electrode migration continues to be a significant complication of cochlear implant surgery. Electrode immobilization using the 'Split Bridge' technique may be an effective way to reduce the incidence of cochlear implant electrode migration.

IRB Number: 20060869

Value of Computed Tomography in the Evaluation of Children with Deficient Cochlear Nerves

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Objective: To assess the value of high resolution computed tomography (HRCT) in the evaluation of children with absent or small (deficient) cochlear nerves (CNs). Of special emphasis was the diagnostic significance of the bony cochlear nerve canal (BCNC), which forms the bony aperture of the modiolus within the internal auditory canal.

Study Design: Retrospective review of medical records.

Setting: Tertiary referral center.

Methods: Nineteen children with cochlear nerve deficiency, who underwent both MRI and HRCT imaging of the temporal bones, were included. The diagnosis of CN deficiency was made based on a set of exams including MRI and thorough audiometric evaluation. HRCTs were reviewed and several parameters including the presence/size of the BCNC were evaluated.

Results: Twenty-seven ears had absent CNs on MRI and 22 (81.5%) of which had abnormal (absent or small) BCNCs. Nine ears (9 of 27, 33.3%) had absent CNs and present but morphologically small BCNCs. Of those ears, 3 had small IACs and 4 had inner ear malformations. Five ears (5 of 27, 18.5%) had absent CNs in the presence of normal BCNCs – all of which had normal size internal auditory canals (IACs) and 2 of which had normal inner ear morphology. Also, 4 ears had small cochlear nerves, 3 of which had normal BCNCs.

Conclusions: This paper documents the diagnostic significance of the BCNC in the evaluation of children with deficient CNs. In this series, almost 20% of ears with absent CNs had normal BCNCs and normal size IACs evidenced via HRCT – thus absence of the CN would have been missed with HRCT imaging only. We therefore advocate MRI for screening children with sensorineural hearing loss.

Unilateral Inner Ear Malformations in Children

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Objective: Unilateral temporal bone malformations are being increasingly diagnosed with advancements in newborn screening and imaging. It is unknown whether they represent a distinct entity from bilateral disease, a fact which would have significant implications for counseling and workup. We studied a large cohort of unilateral malformations to determine any distinguishing features from bilateral disease.

Study design: Retrospective case series.

Setting: Tertiary pediatric center.

Patients: From a database of children with unilateral sensorineural hearing loss (USNHL) who had imaging data (N=112), those with unilateral inner ear malformation were reviewed. Mean follow-up was 3 years.

Main outcome measure(s): Clinical course, radiographic findings, hearing loss progression.

Results: Thirty-nine children with USNHL (35%) had unilateral malformations. Enlarged vestibular aqueduct (EVA) was exceedingly common (92%). EVA was usually isolated; additional anomalies occurred in only 22%. Cochlear and vestibular dysplasias were generally mild and rarely occurred without EVA. Hearing loss was mild to moderate in most (62%), and progressive in only 21%. No incidents of stepwise progression were observed. Although 3 patients had renal or cardiac anomalies, no syndromes were identified, and all ancillary testing, including genetic evaluation, was normal.

Conclusions: Unilateral inner ear malformations may represent a distinct disease process from bilateral malformations, showing on average a milder phenotype in the affected ear and more homogeneity in anatomic structure. Progression of hearing loss may be somewhat less likely, including stepwise progression with head trauma. Extensive ancillary testing is of limited value. These observations may help in counseling parents and in guiding further work-up.

Auditory Benefits of Bilateral Sequential Cochlear Implantation in Children and Adults

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Objective: To assess the clinical benefits of bilateral sequential cochlear implantation in adults and children with at least 12 months time between implantations

Study design: Retrospective

Setting: Tertiary academic referral center

Patients: 69 patients (46 children, 23 adults) with bilateral profound sensorineural hearing loss underwent sequential bilateral implantation with at least 12 months between surgeries. The pediatric age range at the time of the second implant was 2.58 to 18.42 years (mean 7.7 years) and the time between implants ranged from 1.59 to 15.18 years (mean 5.2 years). In the adults, the ages ranged from 24.75 to 67.3 years (mean 49.9 years) and the time between implantations ranged from 1.33 to 9.75 years (mean 4.29 years).

Intervention: Patients with bilateral severe-to-profound sensorineural hearing loss underwent sequential implantation with 12 months or more between the first and second surgery.

Main outcome measures: Standard age appropriate speech perception tests were performed preoperatively prior to the second implantation and postoperatively. The tests included measures of open-set phoneme, word and sentence recognition and localization tasks. For the purposes of this study, the 12-month evaluation was the final outcome interval.

Results: The results indicated an overall improvement in subjective and objective auditory benefits following second-sided implantation. Several factors contributed to bilateral performance including length of implant usage, time between implantations, overall length of deafness, and other disabilities.

Conclusions: Sequential bilateral cochlear implantation yields better results as compared with unilateral implantation, and the benefits were not dependent on length of time between first- and second-implantations.

IRB: 06-740

Clinical Characterization and Genetic Analysis of Familial Migrainous Vertigo

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Background: Migrainous vertigo, characterized by episodic vertigo associated with migraine, is known to be genetically determined in some cases. To date, no loci or genes have been reported for familial migrainous vertigo.

Objectives: To describe the clinical features and natural history of migrainous vertigo, and to perform genetic analysis in a large, multi-generational family.

Methods: We studied a six generation Caucasian family from Brazil over the course of ten years, and collected data from 146 members. Clinical data included detailed case histories, otolaryngological and neurological examinations, audiometric evaluations and imaging studies. Serial clinical and audiometric evaluations were done. We have also undertaken a genome-wide linkage analysis, with subsequent fine mapping using microsatellite markers.

Results and Conclusions: The pattern of transmission was autosomal dominant with incomplete penetrance and variable expression of the gene. Of the 146 members, 32 suffered from migraine with aura. Of these 32 individuals, 10 also suffered from episodic vertigo. Migraine headaches preceded onset of vertigo by 15-20 years, on average. Overall, migraine symptoms decreased with time, while vertigo had a tendency to get worse. Longitudinal audiometric studies over 10 years generally showed stable, high frequency sensorineural hearing loss, consistent with presbycusis. Low tone or fluctuating hearing loss was generally not observed. Imaging studies were normal. Genetic analysis revealed a novel locus on chromosome 5 (lod score 4.05). Studies are ongoing to investigate candidate genes at this locus. This is the first reported locus for migrainous vertigo.

IRB: 11516 Supported by NIH/NIDCD

The Relationship Between the Modified Somatic Perception Questionnaire and Dynamic Platform Posturography

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Objective: To determine if patients whose vestibular symptoms are associated with non-organic sway patterns show more evidence of somatization and/or malingering than patients whose vestibular symptoms are associated with normal or physiologically abnormal sway patterns seen in persons with documented vestibular pathology.

Design: Prospective study of patients undergoing routine vestibular testing including computerized dynamic posturography.

Setting: Neurotology tertiary referral center.

Participants: One hundred four consecutive patients with complaints of vestibular dysfunction and hearing impairment.

Interventions: Computerized dynamic posturography (CDP) and completion of the Modified Somatic Perception Questionnaire (MSPQ) – a validated test for malingering and somatization.

Main Outcome Measures: CDP results classified into four categories (normal, physiologic abnormal, borderline-aphysiologic, and aphysiologic) were correlated with the results of the MSPQ.

Results: A significant CDP group effect on the MSPQ ($F [3,100] = 7.57, p < 0.0001, \eta^2 = 0.19$) was found. The frequency of high MSPQ scores were examined across CDP groups at various MSPQ cutoff scores. Kruskal-Wallis Test demonstrated a significantly higher proportion of the Aphysiologic group had positive scores regardless of the cutoffs used ($> 10: X^2 [3] = 11.28, p < 0.05; > 11: X^2 [3] = 11.82, p < 0.01; > 14: X^2 [3] = 16.95, p < 0.01$).

Conclusions: Results indicate that patients who have aphysiologic CDP sway patterns are more likely to have higher MSPQ scores. Both aphysiologic CDP results and high MSPQ scores have been associated with intentional exaggeration. Patients in this study would likely be exaggerating if they had both aphysiologic CDP results and high MSPQ scores

Chronic Suppurative Otitis Media (CSOM), Caloric Testing and Rotational Chair Testing

Gerard Gianoli, MD, James Soileau, MD

Objective: To determine the incidence of caloric and rotational chair testing abnormalities in patients with CSOM.

Design: Retrospective study of preoperative vestibular testing in patients to undergo tympanomastoidectomy.

Setting: Neurotology tertiary referral center.

Participants: Twenty-three patients with chronic suppurative otitis media with or without cholesteatoma who were to eventually undergo tympanomastoid surgery.

Interventions: Caloric testing and rotational chair testing (ROT).

Main Outcome Measures: History of dizziness. Vestibular test abnormalities defined by caloric weakness (CW), reduced gain, abnormal phase or asymmetry on ROT.

Results: Among the 23 patients, 12 had bilateral CSOM and 11 had unilateral CSOM – most with longstanding disease and history of prior surgical intervention. Seventeen of the 23 patients (74%) demonstrated either unilateral or bilateral CW. Sixteen (70%) demonstrated abnormalities on ROT. Eleven patients (48%) had complaints of vertigo/dizziness, although two of these patients had both normal caloric testing and ROT.

Conclusions: The incidence of CW among CSOM patients in this study was high and correlated well with abnormalities on ROT. Interestingly, ROT results correlated better with CW than symptoms of dizziness/vertigo. While CW findings could be the results of technical limitations in testing patients with CSOM, ROT corroboration of these results suggest that they are valid findings.

Neuro-Optometric Evaluation and Rehabilitation as a Useful Adjunct in the Management of the Complex Dizzy Patient

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A small but significant subgroup of patients who present with dizziness have visual symptomatology as a major complaint. Visual stimuli that may incite or exacerbate dizziness include reading, transferring attention between a computer monitor and written material, and busy or crowded situations. These patients do not demonstrate consistent abnormalities on standard vestibular testing, nor is posturography evaluation often helpful. They do not respond adequately to medical management with or without vestibular rehabilitation. Neuro-optometric evaluation in these patients is valuable, and may reveal a myriad of problems, including those of convergence-divergence and fusion.

We present our findings in 13 patients with complex dizziness and significant eye complaints who underwent thorough neuro-otologic, rehabilitative, and neuro-optometric evaluation. The patients received a combination of medical treatment, vestibular rehabilitation and neuro-optometric rehabilitation as indicated. Significant improvement in symptoms and quality of life was seen in over 62% of patients, all of whom were incapacitated by their dizziness previously.

Neuro-optometric evaluation and rehabilitation is a key part of the care of certain patients with complex dizziness. We will present our findings as well as a simple set of eye evaluations that can be performed by the neurotologists in the office to screen for these disorders.