



PROGRAM and ABSTRACTS

of the

***One Hundred Forty - Seventh
Annual Meeting***

**AMERICAN OTOLOGICAL
SOCIETY, INC.**

May 16 - 17, 2014

***Roman Ballroom
I/III***

**Caesars Palace
Las Vegas, NV**

OFFICERS
JULY 1, 2013—JUNE 30, 2014

PRESIDENT

John W. House, M.D.
House Ear Clinic
Los Angeles, CA

PRESIDENT - ELECT

D. Bradley Welling, M.D., Ph.D.
Harvard Medical School
Massachusetts Eye and Ear Infirmary
Massachusetts General Hospital
Boston, MA

SECRETARY - TREASURER

Steven A. Telian, M.D.
University of Michigan Medical Center
Ann Arbor, MI

EDUCATION DIRECTOR

Debara L. Tucci, M.D.
Duke University Medical Center
Durham, NC

COUNCIL

The above officers and
Herman A. Jenkins, M.D.
Paul R. Lambert, M.D.
Samuel H. Selesnick, M.D.
Roberto A. Cueva, M.D.

Accreditation Statement

This activity has been planned and implemented in accordance with the Essential Areas and Policies of the Accreditation Council for Continuing Medical Education through the joint sponsorship of the American College of Surgeons and the American Otological Society. The American College of Surgeons is accredited by the ACCME to provide continuing medical education for physicians.

AMA PRA Category 1 Credits™

The American College of Surgeons designates this live activity for a maximum of 6.75 *AMA PRA Category 1 Credits™*. Physicians should claim only the credit commensurate with the extent of their participation in the activity.



American College of Surgeons
Division of Education

American Otological Society, Inc.

Mission Statement

Purpose

The American Otological Society, created in 1868, is dedicated to fostering a dialog on and dissemination of, information pertaining to advances in evidence based diagnosis and management of otologic and neurotologic disorders. The focus on otologic and neurotologic disorders and scientific advances are translated to the provision of quality care that is consistent with the ACGME general competency areas and the Institute of Medicine competencies.

Target Audience

The primary target audience for the educational efforts of the American Otological Society is the current and potential members of the society. These members are physicians, otologists, residents, fellows, and researchers in the fields of otology and neurotology. Educational activities are also open to nurses, occupational and speech therapists and other healthcare professionals who are involved in the care of patients with otologic and neurotologic conditions.

Activities

The primary activity of the American Otological Society is the Annual Meeting that focuses on the advancement of the scientific and clinical evidence that supports advances in otologic and neurotologic care to patients. Additionally, non certified educational support and resources include the publication and dissemination of peer reviewed and evidence-based content through the Otology & Neurotology Journal and support for research in otology/neurotology and lateral skull base surgery and related disciplines.

Content

The content for the Annual Meeting and other related educational efforts are limited to the otologic and neurotologic evidence based science, clinical standards of care, and effects on communication.

Expected Results

The expected results are focused on enhancing knowledge translation and promoting competence for the membership and other identified target audiences. The Annual Meeting, the CME certified annual activity of the society, and the other scholarly activities such as the publication of the Journal and support for research provide a rich and robust environment for self assessment and reflection, access to resources for lifelong learning and opportunities for discussion and re-evaluation

2014 AOS Spring Meeting CME Activity Planning

The American Otological Society (AOS) is committed to improving public health care through the provision of high-quality continuing medical education (CME) to our members. The overall goal is to provide CME activities that will address the knowledge gaps and enhance the clinical competence of the participants.

Planning an educational activity that meets the needs of our members is of the utmost importance to the leadership of the AOS. At the close of each annual meeting, we ask that you complete an exit-evaluation. The evaluation is used as a tool to determine the success of the CME program in meeting program objectives, addressing professional practice gaps and educational needs. Your responses provide extensive feedback on what was learned from the Program as well as what you would like to see in the future. The responses are peer-reviewed by the Council prior to the next meeting to assist the Program Committee in developing future AOS continuing medical education programs. The educational program is designed to address the topics identified as practice gaps through individual presentations and in depth panel discussions. Based on the response, the following data regarding professional practice gaps among attendees were noted:

- Most practitioners perceive that there are no treatments for tinnitus, and have little understanding of the pathophysiology.
- Practitioners face ethical dilemmas in their practices but have not thought about how to approach these or discuss with patients.
- Current treatment is driven more by past experience than knowledge of contemporary studies and evidence-based medicine.
- Varying surgical methods for dealing with incus necrosis in revision stapes surgery, with a variety of outcomes.

AOS President, Dr. John W. House, selected Dr. David A. Moffat as the 2014 AOS Guest of Honor. Dr. Moffat will kick off the scientific program on Friday at 1:00 P.M. with his presentation entitled, "*Ethical Dilemmas in Otology*". Other highlights of the 147th AOS annual meeting include the Basic Science lecture entitled, "*The Gray Area - Tinnitus and the Brain*", presented by Dr. Josef P. Rauschecker on Saturday. Dr. Carol A. Bauer put together a top-notch panel to discuss "*Tinnitus: Models, Mechanisms and New Management Strategies*" on Friday. A second panel entitled, "*Single-Sided Deafness: Why Not a CI?*" moderated by Dr. Bruce J. Gantz accompanied by a superb panel of experts will take place on Saturday. Dr. Neal M. Jackson and Dr. Nathan H. Calloway were both selected as recipients of an *AOS Resident Research Travel Award*.

In addition, there are a vast number of oral presentations exploring the latest otological research and findings. Be sure to visit the Octavius Ballroom where you will find an outstanding display of AOS poster submissions. Posters will be available for viewing on Friday & Saturday, 9:00-4:00. Recipients of the AOS/ANS combined poster awards will be announced at the close of the AOS Scientific program on Friday, May 16th at 5:00 P.M. The Combined Poster Reception/Meet the Authors will take place Friday evening from 5:30-7:00 P.M.

To close the identified practice gaps, participants of this activity will need to learn:

- Attendees will learn about the pathophysiology of tinnitus (specifically how the brain changes in response to hearing loss and how tinnitus is generated), and how specific characteristics of tinnitus patients may make them a better candidate for one type of treatment so that treatment choices can be targeted to patient needs.
- Attendees will have the opportunity to consider ethical dilemmas in their own practice and formulate a methodology to approach these issues in a thoughtful way. They may change the way that they interact and communicate with patients in these circumstances as a result.
- A number of studies using intratympanic treatments and reports of new delivery methods will be presented. Participants need to learn the state of contemporary scientific knowledge about the evidence basis for use of these treatments to optimize patient care.
- Revision stapes surgery is required for dislocation of the stapes prosthesis. The use of tissue glue has made some reconstructions easier and more long lasting. Participants will learn when these techniques may be most useful and how to apply them to their surgical practice.

Patient outcomes will be improved by:

Practitioners will develop a more thorough understanding of and approach to patients who have targeted conditions, including tinnitus, otosclerosis, and Meniere's disease, which are very common in the population of patients we treat. They will thoughtfully consider their approach to ethical issues that arise in otologic practice.

Patient outcomes will be improved in the following ways:

1. Offering the best understanding of the tinnitus condition and recommendations for the best treatments to patients suffering from this condition.
2. Perform most appropriate treatments for Meniere's disease.
3. Perform best surgical technique for treatment of displaced stapes prosthesis.

Learning Objective(s) - At the end of this activity, participants will be able to:

- Explain current state of knowledge on pathophysiology of tinnitus
- Evaluate and implement appropriate treatments available for tinnitus
- Apply the use of bone cement for treatment of incus necrosis in revision stapes surgery

Position Statement: Any presentations, conversations, exhibits, or other meeting communications, including descriptions of the use of drugs or devices, does not imply or constitute endorsement of any company, product, application, or use by the American Otological Society.

The following statement was read, submitted, and signed by every individual connected with this educational activity. Failure to comply disqualifies the individual from planning or speaking at any AOS Continuing Medical Education program.

In compliance with ACCME Accreditation Criteria, the American College of Surgeons, as the accredited provider of this activity, must ensure that anyone in a position to control the content of the educational activity has disclosed all relevant financial relationships with any commercial interest. All reported conflicts are managed by a designated official to ensure a bias - free presentation.

In accordance with the ACCME Accreditation Criteria, the American College of Surgeons, as the accredited provider of this activity, must ensure that anyone in a position to control the content of the educational activity has disclosed all relevant financial relationships with any commercial interest. Therefore, it is mandatory that both the program planning committee and speakers complete disclosure forms. **Members of the program committee were required to disclose all financial relationships and speakers were required to disclose any financial relationship as it pertains to the content of the presentations.** The ACCME defines a 'commercial interest' as "any entity producing, marketing, re - selling, or distributing health care goods or services consumed by, or used on, patients". It does not consider providers of clinical service directly to patients to be commercial interests. The ACCME considers "relevant" financial relationships as financial transactions (in any amount) that may create a conflict of interest and occur within the 12 months preceding the time that the individual is being asked to assume a role controlling content of the educational activity.

AOS is also required, through our joint sponsorship partnership with ACS, to manage any reported conflict and eliminate the potential for bias during the activity. All program committee members and speakers were contacted and the conflicts have been managed to our satisfaction. However, if you perceive a bias during a session, please report the circumstances on the session evaluation form.

Please note we have advised the speakers that it is their responsibility to disclose at the start of their presentation if they will be describing the use of a device, product, or drug that is not FDA approved or the off - label use of an approved device, product, or drug or unapproved usage.

The requirement for disclosure is not intended to imply any impropriety of such relationships, but simply to identify such relationships through full disclosure, and to allow the audience to form its own judgments regarding the presentation.

Disclosure Information

PUBLICATION STATEMENT

The material in this abstract, (Name of Abstract), has not been submitted for publication, published, nor presented previously at another national or international meeting and is not under any consideration for presentation at another national or international meeting. The penalty for duplicate presentation/publication is prohibition of the author and co - authors from presenting at a COSM society meeting for a period of three years. Submitting Author's Signature (required)

All authors were advised that the submitted paper becomes the property of *Otology & Neurotology* and cannot be reprinted without permission of the Journal.

*****Disclosures *****

American Otological Society, Inc. Statement

All authors, presenters, panelists, guest lecturers, Council members, Program Advisory Committee members, Administrative staff and any other contributing individuals who may be in a position to control content of a CME activity were required to complete a Disclosure/Conflict of Interest/Attestation declaration prior to consideration for presentation or appointment to a CME planning Committee. All potential conflicts of interest were resolved prior to participation in the planning of this activity.

Authors were instructed to read and sign the following Attestation statement.

1. I will disclose all relevant financial relationships to the AOS, disclose this information to learners verbally (for live activities) and in print.
2. The content and/or presentation of the information with which I am involved will promote quality or improvements in healthcare and will not promote a specific proprietary business interest of a commercial interest. Content for this activity, including any presentation of therapeutic options, will be well - balanced, evidence - based and unbiased.
3. I have not and will not accept any honoraria, additional payments or reimbursements beyond that which has been agreed upon directly with the AOS.
4. If I am presenting at a live event, I am aware that a CME monitor will attend the event to ensure that my presentation is educational, and not promotional, in nature. If presentation is found to be promotional in any way, I understand I will be ineligible to participate in an *AOS/ACS* jointly sponsored CME accredited activity for a period up to two years.
5. If I am providing recommendations involving clinical medicine, they will be based on evidence that is accepted within the profession of medicine as adequate justification for their indications and contraindications in the care of patients. All scientific research referred to, reported or used in CME in support of justification of a patient care recommendation will conform to the generally accepted standards of experimental design, data collection and analysis.
6. If I am discussing specific healthcare products or services, I will use generic names to the extent possible. If I need to use trade names, I will use trade names from several companies when available, not just trade names from any single company.
7. If I am discussing any product use that is off label, I will disclose that the use or indication in question is not currently approved by the FDA for labeling or advertising.
8. If I have been trained or utilized by a commercial entity or its agent as a speaker (e.g., speaker's bureau) for any commercial interest, the promotional aspects of that presentation will not be included in any way with this activity.
9. If I am presenting research funded by a commercial company, the information presented will be based on generally accepted scientific principles and methods, and will not promote the commercial interest of the funding company.

*****FACULTY DISCLOSURES***
(in alphabetical order)**

American Otological Society Council 2013-2014

The following Council Members disclose:

Samuel H. Selesnick, MD

Medtronic ENT-Royalty

D. Bradley Welling, MD, PhD

J. Morita-Speaker

The following Council Members have nothing to disclose:

Roberto A. Cueva, MD

John W. House, MD

Herman A. Jenkins, MD

Paul R. Lambert, MD

Steven A. Telian, MD

Debara L. Tucci, MD

Program Advisory Committee 2014

The following Committee Members disclose:

John P. Carey, MD

Otonomy-Advisory panel

Pfizer-Advisory panel

Sujana S. Chandrasekhar, MD

Scientific Development & Research Inc.-Shareholder, board member

Cochlear Corp-Consultant

MED-EL-Consultant

Bruce J. Gantz, MD

Advanced Bionics-Consultant

Cochlear Corp-Consultant

Medtronic-Consultant

Bradley W. Kesser, MD

Nasco, Inc-Royalty (patent holder)

Lawrence R. Lustig, MD

Advanced Bionics-Medical advisory board

MED-EL-Surgical advisory board

Samuel H. Selesnick, MD-(see above)

The following Committee Members have nothing to disclose:

J. Douglas Green Jr., MD

John W. House, MD

John K. Niparko, MD

Debara L. Tucci, MD

AOS Administrative Staff has nothing to disclose:

Kristen Bordignon

Ashley Westbrook

*****AOS Disclosures—Oral Presentations *****
Friday, May 16, 2014
Scientific Session
Oral Presentations: Authors/Presenters
& Panel Participants
(listed in order of presentation)

1:15pm - GUEST OF HONOR

The following individual has nothing to disclose:
David A. Moffat, PhD, FRCS

1:42pm

The following individuals have nothing to disclose:
M. Jennifer Derebery, MD (Primary)
Courtney Voelker, MD, PhD
Audrey Calzada, MD
Laurel M. Fisher, PhD

1:50pm

The following individuals have nothing to disclose:
Catherine M. Kelso, BS (Primary)
Hirobumi Watanabe, PhD
Zhen Jason Qian, BS
Joseph Wazen, BS
The following individuals disclose:
Jeffrey W. Kysar, PhD
Columbia University-Patent pending related to microneedle
Anil K. Lalwani, MD
Advanced Bionics-Medical advisory board
Columbia University- Patent pending related to microneedle

1:58pm

The following individuals have nothing to disclose:
Il Joon Moon, MD, PhD (Primary)
Justin S. Golub, MD
Jay T. Rubinstein, MD, PhD

2:06pm

The following individuals have nothing to disclose:
Daniel Schuster, MD (Primary)
Louis B. Kratchman, BS
The following individual disclosed:
Robert F. Labadie, MD, PhD
Ototronix-Medical advisory board
Ototronix-Royalty

2:14pm

The following individuals have nothing to disclose:
Amit J. Sood, BA (Primary)
Paul R. Lambert, MD
Ted A. Meyer, MD, PhD
The following individual disclosed:
Shaun A. Nguyen, MD, MA
Olympus, Inc-Consultant
Global Life Technologies, Inc-Consultant and research support
Angiotech, Inc-Consultant

2:22pm

The following individuals have nothing to disclose:
Bryan A. Lanzman, MD (Primary)
Benjamin J. Rubinstein, MD
Daniel S. Chow, MD
Angela Lignelli, MD
William Millar, MD
The following individual disclosed:
Anil K. Lalwani, MD
Advanced Bionics-Medical advisory board

*****Disclosures—Oral Presentations***
Friday, May 16, 2014
Scientific Session**

2:30pm - Resident Research Travel Award

The following individuals have nothing to disclose:

Neal M. Jackson, MD (Primary)
Laveil M. Allen, MD
Anagha Kakade, MSC, DBM, PDCR
Anita Jeyakumar, MD, MS
Christopher Arcement, MD
Yu-Lan Mary Ying, MD

The following individual disclosed:

Moises A. Arriaga, MD, MBA
Elsevier-Royalties

3:14pm

The following individuals have nothing to disclose:

Stéphane Tringali, MD, PhD (Primary)
Kanthaiah Koka, PhD
Herman A. Jenkins, MD
Daniel J. Tollin, PhD

3:23pm

The following individuals have nothing to disclose:

Daniel J. Roche, MD, JD (Primary)
Debara L. Tucci, MD
Jeffrey Marcus, MD
Jay McClennen, AOCA, CCA, CFm
David M. Kaylie, MD

3:31pm

The following individuals have nothing to disclose:

J. Eric Lupo, MD (Primary)
John W. House, MD
John C. Goddard, MD

3:39pm

The following individuals have nothing to disclose:

Scott K. Hudson, MD (Primary)
Richard K. Gurgel, MD
The following individual disclosed:
Clough Shelton, MD
Cochlear Corp-Research funding

3:47pm

The following individuals have nothing to disclose:

Daniel Q. Sun, MD (Primary)
Linda Zhou
Frank R. Lin, MD
Howard W. Francis, MD, MBA
John P. Carey, MD
Wade W. Chien, MD

3:55pm

The following individuals have nothing to disclose:

Louisa Ha, AuD (Primary)
Samuel Trosman, MD
Neel Patel, MD
Gabrielle Cager, AuD
Miriam I. Redleaf, MD

*****Disclosures—Oral Presentations***
Friday, May 16, 2014
Scientific Session**

4:08pm - PANEL

The following individuals have nothing to disclose:

Carol A. Bauer, MD - Moderator

Gregory P. Lekovic, MD, PhD

The following individuals disclose:

Michael D. Seidman, MD

NIH-Research funding

Body Language Vitamin Co-founder

Visalus Sciences-Royalties

Intellectual Property-Patents

Richard S. Tyler, PhD

Cochlear-Grant

Micro Transponder-Consultant

Sound Cure-Consultant

Fan-Gang Zeng, PhD

Sound Cure-Founder, stocks

Neurotron-Founder

*****Disclosures - Oral Presentations ***
Saturday, May 17, 2014
Scientific Session
Oral Presentations: Authors/Presenters
& Panel Participants
(listed in order of presentation)**

1:05pm

The following individuals have nothing to disclose:

Aaron K. Remenschneider, MD, MPH (Primary)

Sarah A. Lookabaugh, MD

Jacob R. Brodsky, MD

Avner Aliphaz, MD

Selena E. Heman-Ackah, MD

Daniel J. Lee, MD

Alicia M. Quesnel, MD

1:13pm

The following individuals have nothing to disclose:

Susan D. Emmett, MD (Primary)

Howard W. Francis, MD, MBA

1:21pm

The following individuals have nothing to disclose:

Matthew L. Bush, MD (Primary)

Bryan Hardin, BS

Christopher Rayle, BA

1:29pm

The following individuals have nothing to disclose:

Andrew J. Layman, BS (Primary)

Carol Li, BS

Luigi Ferrucci, MD, PhD

Danielle Kauffman, BS

John P. Carey, MD

Yuri Agrawal, MD

1:37pm

The following individuals have nothing to disclose:

Garrett G.A. Casale, BS (Primary)

Brian D. Nicholas, MD

Bradley W. Kesser, MD

*****Disclosures—Oral Presentations***
Saturday May 17, 2014
Scientific Session**

1:45pm

The following individuals have nothing to disclose:

Matthew G. Crowson, MD (Primary)

Katherine Callahan

James E. Saunders, MD

1:53pm

The following individuals have nothing to disclose:

Jeffrey D. Sharon, MD (Primary)

Shariq S. Khwaja, MD, PhD

Andrew Drescher, MD

The following individuals disclose:

Hiram Gay, MD

Augmenix, Inc-Consultant

ROCS-Medical Reviewer

Richard A. Chole, MD, PhD

Checksite Medical Inc-Patent

2:01pm

The following individuals have nothing to disclose:

Robert J. Yawn, MD (Primary)

Matthew L. Carlson, MD

Alejandro Rivas, MD

The following individual disclosed:

David S. Haynes, MD

Advanced Bionics-Advisory board

Cochlear Corporation-Advisory board

MED-EL-Advisory board

Grace Medical-Consultant

Stryker-Consultant

Synthes/Anspach-Consultant

2:16pm - BASIC SCIENCE LECTURE

The following individual has nothing to disclose:

Josef P. Rauschecker, PhD, DSc

3:15pm

The following individuals have nothing to disclose:

Kathryn M. Van Abel, MD (Primary)

Douglas P. Sladen, PhD

Brian A. Neff, MD

Charles W. Beatty, MD

The following individual disclosed:

Colin L.W. Driscoll, MD

Cochlear Corp-Consultant

MED-EL-Consultant

Advanced Bionics-Consultant

*****Disclosures—Oral Presentations***
Saturday May 17, 2014
Scientific Session**

3:23pm - Resident Research Travel Award

The following individuals have nothing to disclose:

Nathan H. Calloway, MD (Primary)

Adam P. Campbell, MD

The following individuals disclose:

Claire Iseli, MBBS, FRACS

Cochlear Corporation-Contractual research support

Craig A. Buchman, MD

Advanced Bionics-Consultant, Contractual research support

Cochlear Corporation-Contractual research support

MED-EL-Consultant, Contractual research support

Cochlear Americas-Consultant

Douglas C. Fitzpatrick, PhD

Advanced Bionics-Contractual research support

Cochlear Corporation-Contractual research support

MED-EL-Contractual research support

NIH-NIDCD-Research grant

Oliver F. Adunka, MD

Advanced Bionics-Consultant, Contractual research support

Cochlear Corporation-Contractual research support

MED-EL-Consultant, Contractual research support

3:31pm

The following individuals disclose:

Nathaniel T. Greene, PhD (Primary)

Cochlear Boulder LLC-Consultant

James R. Easter

Cochlear Boulder LLC-Employee

The following individuals have nothing to disclose:

Travis J. Pfannenstiel, MD

Herman A. Jenkins, MD

Daniel J. Tollin, PhD

3:39pm

The following individuals have nothing to disclose:

Scott H. Troob, MD (Primary)

Alison Singleton, AuD

Abbas A. Anwar, MD

Anthony M. Tolisano, MD

Yixin Fang, PhD

The following individuals disclose:

J. Thomas Roland Jr., MD

Cochlear Americas-Advisory board, Consultant

Advanced Bionics-Advisory board, Consultant

Susan B. Waltzman, PhD

Cochlear Americas-Consultant

3:47pm

The following individuals have nothing to disclose:

Michael V. Certo, BFA, MM (Primary)

Gavriel D. Kohlberg, MD

Dean M. Mansuco, AuD

Divya A. Chari, BS

The following individual disclosed:

Anil K. Lalwani, MD

Advanced Bionics-Medical advisory board

*****Disclosures—Oral Presentations*****

Saturday May 17, 2014

Scientific Session

4:02pm - PANEL

The following individuals disclose:

Bruce J. Gantz, MD - Moderator

Cochlear Corporation-Consultant

Advanced Bionics-Consultant

René H. Gifford, PhD

Advanced Bionics-Audiology advisory board

MED-EL-Audiology advisory board

Cochlear Americas-Audiology advisory board

Lawrence R. Lustig, MD

Advanced Bionics-Medical advisory board

MED-EL-Medical advisory board

Jay T. Rubinstein, MD, PhD

Cochlear Ltd-Consultant, P.I.

Advanced Bionics-Mentor

J. Thomas Roland Jr., MD

Advanced Bionics-Speaker

Cochlear America-Speaker

Colin L. W. Driscoll, MD

Cochlear Corp-Consultant

MED-EL-Consultant

Advanced Bionics-Consultant

David S. Haynes, MD

Advanced Bionics-Advisory board

Cochlear Corporation-Advisory board

MED-EL-Advisory board

Grace Medical-Consultant

Stryker-Consultant

Synthes/Anspach-Consultant

The following individual has nothing to disclose:

Marlan R. Hansen, MD

*****AOS Disclosures—Poster Presentations *****

(in numerical order E001-E030)

Effect of CHD7 Mutation on Inner Ear Structure

The following individuals have nothing to disclose:

Ilene Chiu, MD (Primary)

Seema R. Lalani, MD

Christine M. Eng, MD

John W. Belmont, MD, PhD

Jeffrey T. Vrabec, MD

Safety of Temperatures Generated by Endoscopes in Transcanal Endoscopic Ear Surgery

The following individuals have nothing to disclose:

Tsukasa Ito, MD, PhD (Primary)

Toshinori Kubota, MD, PhD

Akira Takagi

Tomoo Watanabe, MD, PhD

Kazunori Futai, MD, PhD

Takatoshi Furukawa, MD, PhD

Seiji Kakehata, MD, PhD

Vestibular Schwannoma Cells are viable 3-Years after Stereotactic Radiation Therapy: A Case Report with Implications for Pre-procedure Counseling

The following individuals have nothing to disclose:

Terry N. Platto, BA (Primary)

Suzu Igarashi, BS

Maki Niihori, PhD

Allison M. Dunn, BA

Audriana Hurbon, BS

Abraham Jacob, MD

Tympanic Membrane Manipulation to Treat Symptoms of Patulous Eustachian Tube

The following individuals have nothing to disclose:

Matthew D. Brace, MSc, MD (Primary)

Peter Horwich, BSc

David Kirkpatrick, MD, FRCS

Manohar Bance, MB, ChB, MSc

Endolymphatic Hydrops Reversal on 3D FLAIR 3T MRI after Diuretic Treatment

The following individuals have nothing to disclose:

Nopawan Vorasubin, MD (Primary)

Kevin A. Peng, MD

Ali R. Sepahdari, MD

Sameer Ahmed, MD

Akira Ishiyama, MD

A Novel Bone Conduction Hearing System - Long-term Safety and Efficacy Trial

The following individuals have nothing to disclose:

Richard K. Gurgel, MD (Primary)

Clough Shelton, MD

Development of an Algorithm for the Automatic Otitis Media Image Diagnosis

The following individuals have nothing to disclose:

Pa-Chun Wang, MD, MSc, MBA (Primary)

Te-Yung Fang, MD

Men-Tzung Lo, PhD

The following individuals disclose:

Chuen-Kai Shie, MS

HTC Corporation-Computation technology support

Ming-tien Lin, MS

HTC Corporation-Computation technology support

Hao Ting Chang, MS

HTC Corporation-Computation technology support

Third Window Lesions and Endolymphatic Hydrops

The following individuals have nothing to disclose:

Kevin A. Peng, MD (Primary)

Nopawan Vorasubin, MD

Ali R. Sepahdari, MD

Akira Ishiyama, MD

Measuring Sentence Recognition Outcomes among Cochlear Implant Recipients: Variability by Age

The following individuals have nothing to disclose:

Ahmad F. Mahmoud (Primary)

Michael J. Ruckenstein, MD

The One-cut Meatoplasty: Novel Surgical Technique and Outcomes

The following individuals have nothing to disclose:

Kristen L. Hovis, BA (Primary)

Matthew L. Carlson, MD

George B. Wanna, MD

Alex D. Sweeney, MD

The following individual disclosed:

David S. Haynes, MD

Advanced Bionics-Advisory board

Cochlear Corporation-Advisory board

MED-EL-Advisory board

Grace Medical-Consultant

Stryker-Consultant

Synthes/Anspach-Consultant

*****Disclosures—Poster Presentations*****

Treatment of Ear and Bone Disease in the Phex Mouse Mutant with Dietary Supplements

The following individuals have nothing to disclose:

Cameron C. Wick, MD (Primary)

Sharon J. Lin, BS

Heping Yu, BS

Qing Y. Zheng, MD

Cliff A. Megerian, MD

Apogeotropic Positional Nystagmus in Horizontal Semicircular Canal Vertigo: Case Series and Review

The following individuals have nothing to disclose:

Kimanh Nguyen, MD (Primary)

Sameer Ahmed, MD

Akira Ishiyama, MD

Robert W. Baloh, MD

Gail Ishiyama, MD

Totally Transcanal Endoscopic Retrograde Mastoidectomy on Demand using Electrically Powered Instruments

The following individuals have nothing to disclose:

Seiji Kakehata, MD, PhD (Primary)

Tomoo Watanabe, MD, PhD

Tsukasa Ito, MD, PhD

Takatoshi Furukawa, MD, PhD

Toshinori Kubota, MD, PhD

Kazunori, Futai, MD, PhD

Quality Improvement in Delivery of Care for Vestibular Disorders

The following individual discloses:

David R. Friedland MD, PhD (Primary)

MED-EL-Surgeons Advisory Board

The following individuals have nothing to disclose:

Christy Erbe, BS

Sergey Tarima, PhD

Alexia Miles, MPT

Michael Stadler, MD

Outcomes of Cochlear Implantation in Patients 80 years and Older

The following individuals have nothing to disclose:

Eleanor P. Kiell, MD (Primary)

Carolyn E. Witman, BA

Meagan P. Lewis, AuD

John S. May, MD

Eric R. Oliver, MD

Temporomandibular Joint Osteomyelitis Associated with Otitis Externa

The following individuals have nothing to disclose:

Candace E. Hobson, MD (Primary)

Andrew A. McCall, MD

Barry E. Hirsch, MD

Yael Raz, MD

Canal Wall Reconstruction Tympanomastoidectomy with Mastoid Obliteration for the Treatment of Cholesteatoma in Children

The following individuals have nothing to disclose:

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*****Disclosures—Poster Presentations*****

Curved Adjustable Fiberoptic Laser for Endoscopic Cholesteatoma Surgery

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Hossein Mahboubi, MD, MPH
Hamid R. Djalilian, MD

New Possibilities in Middle Ear Implants with Direct Acoustic Cochlea Stimulation

The following individuals have nothing to disclose:

Henryk Skarzynski, Prof, MD, PhD (Primary)
Lukasz Olszewski, MSc
Arkadiusz Wasowski, MSc, PhD
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Anterior-posterior Diameter of the Incus in Stapes Surgery

The following individual has nothing to disclose:

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Eustachian Tube Point: A Novel Clinical Gesture in Otolaryngology

The following individuals have nothing to disclose:

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Paraneoplastic Syndrome: A Masquerade of Autoimmune Inner Ear Disease

The following individuals have nothing to disclose:

Jacqueline J. Greene, MD (Primary)
Michael W. Keefe, MD
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The following individual discloses:

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Otonomy Inc-Equity and consultant

Systematic Review of Cochlear Implantation's Impact on Cognition in the Elderly

The following individuals have nothing to disclose:

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Carol L. Howe, MD, MLS
Mindy Fain, MD
Abraham Jacob, MD

Short-Term Clinical Performance of the Direct-Drive Hearing Device: A Pilot Study

The following individuals have nothing to disclose:

Hossein Mahboubi, MD, MPH (Primary)
Yaser Ghavami, MD
Amy Y. Yau, MD

The following individuals disclose:

Peyton Paulick, MS
Intellectual property-Patent pending
Mark Bachman, PhD
Intellectual property-Patent pending
Hamid R. Djalilian, MD
Intellectual property-Patent pending

Cochlear Implantation Requiring a Retrofacial Approach to the Round Window

The following individual has nothing to disclose:

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The following individuals disclosed:

Loren J. Bartels, MD

Advanced Bionics-Advisory board, Grant/research support
MED-EL-Grant/research support

Brandon Isaacson, MD

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Advanced Bionics-Medical advisory board
Stryker-Course instructor

Two Types of Endoscopic Approach for Congenital Middle Ear Cholesteatoma

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Tsukasa Ito, MD, PhD

Takatoshi Furukawa, MD, PhD

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Seiji Kakehata, MD, PhD

Radiologic Quandary: Giant Cell Tumor of the Temporal Bone Masquerading as Osteomyelitis with Abscess

The following individuals have nothing to disclose:

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Biofilm as a Cause of Cochlear Implant Extrusion

The following individuals disclose:

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Grace Medical, Inc-Product royalties

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Facial Nerve Paresis as Presentation of Recurrent Non-Hodgkins Lymphoma

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Useful Cases of Multiple Cholesteatoma by non-EPI DW-MRI and MRC Images

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**THE AMERICAN OTOLOGICAL SOCIETY WOULD
LIKE TO THANK THE FOLLOWING MEMBERS
FOR THEIR CONTRIBUTION TO THE
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Mark your calendar!

**Combined Poster Reception
AOS/ANS/ASPO/ARS**

Friday, May 16, 2014

5:30 pm - 7:00 pm

Octavius Ballroom

AOS President's Reception & Banquet

Saturday, May 17, 2014

Reception - 6:30 pm

Pompeian Ballroom III/IV

Dinner/Dance - 7:30 pm

Pompeian Ballroom I/II

Formal attire/Black tie optional
{Advanced ticket purchase required
Members & Invited Guests only}

Upcoming meetings

**AOS 148th Annual Meeting
(in conjunction with COSM)**
April 24–26, 2015
Sheraton Boston
Boston, MA

2014 AAO-HNS Annual Meeting & OTO Expo
September 21-24, 2014
Orange County Convention Center
Orlando, FL

The Abstract deadline for the AOS 148th Annual meeting is Wednesday, October 15, 2014.

Abstract Instructions and submission form will be available on website after July 7, 2014.
Website - www.americanotologicalsociety.org

All primary and contributing authors are required to complete a disclosure/conflict of interest statement at time of abstract submission in order for the abstract to be considered by the Program Advisory Committee.

Journal Requirements/Instructions to Primary Authors
Manuscripts are required of ALL ORAL presentations. Manuscripts must be submitted online a **minimum of four weeks** prior to the annual meeting, via the journal's website. Instructions for registering, submitting a manuscript, and the author guidelines can be found on the Editorial Manager site:
<https://www.editorialmanager.com/on/>

The journal of **OTOLOGY & NEUROTOLOGY** does not accept paper manuscripts. Manuscripts will be peer reviewed prior to the Annual meeting for conflict of interest review and resolution.

Failure to comply with the guidelines & requirements of the American Otological Society and the O&N Journal will result in the disqualification of your presentation.

For Society business, please forward all inquiries to:

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The following ACGME competency areas will be addressed throughout this CME activity.

Patient Care that is compassionate, appropriate, and effective for the treatment of health problems and the promotion of health.

Medical Knowledge about established and evolving biomedical, clinical, and cognate (e.g. epidemiological and social-behavioral) sciences and the application of this knowledge to patient care.

Practice-Based Learning and Improvement that involves investigation and evaluation of their own patient care, appraisal and assimilation of scientific evidence, and improvements in patient care.

Interpersonal and Communication Skills that result in effective information exchange and teaming with patients, their families, and other health professionals.

Professionalism as manifested through a commitment to carrying out professional responsibilities, adherence to ethical principles, and sensitivity to a diverse patient population.

Systems-Based Practice as manifested by actions that demonstrate an awareness of and responsiveness to the larger context and system of health care and the ability to effectively call on system resources to provide care that is of optimal value.

Friday, May 16, 2014

- 12:30 Business Meeting (Members Only)**
Room: Roman Ballroom I/III
New member introduction
Secretary-Treasurer Report
AOS member photograph-Roman I/III
- 1:00 Scientific Program**
(Open to registered Members and Non-members)
Badge required for admittance)
Room: Roman Ballroom I/III
- 1:00 Welcome & Opening Remarks by the President**
John W. House, MD
- O&N Impact Award**
Erika A. Woodson, MD
- NIH funding-K Awards**
Yuri Agrawal, MD
David Horn, MD
Akihiro J. Matsuoka, MD, PhD
- 1:05 Presidential Citations**
Robert K. Jackler, MD
Paul R. Lambert, MD
Richard T. Miyamoto, MD
Clough Shelton, MD
Fred F. Telischi, MD
- 1:15 GUEST OF HONOR LECTURE**
“Ethical Dilemmas in Otology”
David A. Moffat, PhD, FRCS
- 1:35 DISCUSSION**
- 1:42 An Open Label Study to Evaluate the Safety and Efficacy of Intratympanic Golimumab Therapy in Patients with Autoimmune Inner Ear Disease**
M. Jennifer Derebery, MD
Courtney Voelker, MD, PhD
Audrey Calzada, MD
Laurel M. Fisher, PhD
- 1:50 Creation of Microperforation Significantly Enhances Diffusion across Round Window Membrane**
Catherine M. Kelso, BS
Hirobumi Watanabe, PhD
Zhen Jason Qian, BS
Joseph Wazen, BS
Jeffrey W. Kysar, PhD
Anil K. Lalwani, MD
- 1:58 Endolymphatic Shunt Surgery and Intratympanic Gentamycin: An Uncontrolled Retrospective Comparison**
Il Joon Moon, MD, PhD
Justin S. Golub, MD
Jay T. Rubinstein, MD, PhD

- 2:06** **Characterization of Basilar Membrane Rupture Forces in Fresh Human Cadaveric Cochleae**
Daniel Schuster, MD
Louis B. Kratchman, BS
Robert F. Labadie, MD, PhD
- 2:14** **Endolymphatic Sac Surgery for Meniere's Disease: A Systematic Review and Meta-Analysis**
Amit J. Sood, BA
Paul R. Lambert, MD
Shaun A. Nguyen, MD, MA
Ted A. Meyer, MD, PhD
- 2:22** **Pöschl Reconstruction in Assessment of Vestibular Aqueduct**
Bryan A. Lanzman, MD
Benjamin J. Rubinstein, MD
Daniel S. Chow, MD
Angela Lignelli, MD
William Millar, MD
Anil K. Lalwani, MD
- 2:30** **RESIDENT RESEARCH TRAVEL AWARD**
The Relationship of Age on Radiographic Incidence of Superior Semicircular Canal Dehiscence in Pediatric Patients
Neal M. Jackson, MD
Laveil M. Allen, MD
Anagha Kakade, MSC, DBM, PDCR
Anita Jeyakumar, MD, MS
Christopher Arcement, MD
Moises A. Arriaga, MD, MBA
Yu-Lan Mary Ying, MD
- 2:38** **DISCUSSION**
- 2:45** **BREAK WITH EXHIBITORS**
- 3:15** **Sound Location Modulation of Electrocochleographic Responses in Chinchilla with Single Sided Deafness and Fitted with an Osseointegrated Bone-conducting Hearing Prosthesis**
Stéphane Tringali, MD, PhD
Kanthaiah Koka, PhD
Herman A. Jenkins, MD
Daniel J. Tollin, PhD
- 3:23** **Osseointegrated Implants for Auriculectomy and Microtia Defects: Operative Techniques and Complication Management**
Daniel J. Rocke, MD, JD
Debara L. Tucci, MD
Jeffrey Marcus, MD
Jay McClennen, AOCA, CCA, CFm
David M. Kaylie, MD

- 3:31 Management of Incus Necrosis in Revision Stapedectomy using Hydroxyapatite Bone Cement**
J. Eric Lupo MD
John W. House, MD
John C. Goddard, MD
- 3:39 Revision Stapedectomy with Bone Cement: Are Results Comparable to Standard Techniques?**
Scott K. Hudson, MD
Richard K. Gurgel, MD
Clough Shelton, MD
- 3:47 Racial Differences in Cochlear Melanocytic Content Are Associated with Hearing Loss**
Daniel Q. Sun MD
Linda Zhou
Frank R. Lin MD
Howard W. Francis MD
John P. Carey MD
Wade W. Chien MD
- 3:55 Subjective Hearing Loss in Adults with Normal Thresholds May Be Improved by Auditory Training**
Louisa Ha, AuD
Samuel Trosman, MD
Neel Patel, MD
Gabrielle Cager, AuD
Miriam I. Redleaf, MD
- 4:03 DISCUSSION**
- 4:08 PANEL – “Tinnitus: Models, Mechanisms and New Management Strategies”**
Carol A. Bauer, MD - Moderator
Gregory P. Lekovic, MD, PhD
Michael D. Seidman, MD
Richard S. Tyler, PhD
Fan-Gang Zeng, PhD
- 5:05 ADJOURNMENT**
- 5:30 COMBINED POSTER RECEPTION**
AOS/ANS/ASPO/ARS
Octavius Ballroom
- 7:00 PAST PRESIDENTS DINNER**
(Invited guests only)

Saturday, May 17, 2014

- 12:30 Business Meeting (Members Only)**
Room: Roman Ballroom I/III
Committee Reports
Board of Trustees of the Research Fund
American Board of Otolaryngology
Award of Merit Committee
American College of Surgeons
AAO - HNS
AAO - HNS Board of Governors
Audit Committee
O&N Journal
Membership Development
Nominating Committee
Unfinished Business
New Business
- 1:00 Scientific Program**
(Open to registered Members and Non-members)
Badge required for admittance)
Room: Roman Ballroom I/III
- 1:00 Remarks by the President**
John W. House, MD
- 1:05 Otologic Outcomes Following Blast Injury-
The Boston Marathon Experience**
Aaron K. Remenschneider, MD, MPH
Sarah A. Lookabaugh, MD
Jacob R. Brodsky, MD
Avner Aliphass, MD
Selena E. Heman-Ackah, MD
Daniel J. Lee, MD
Alicia M. Quesnel, MD
- 1:13 The Socioeconomic Impact of Hearing Loss
in US Adults**
Susan D. Emmett, MD
Howard W. Francis, MD, MBA
- 1:21 Rural Barriers to Early Diagnosis of Infant
Hearing Loss**
Matthew L. Bush, MD
Bryan Hardin, BS
Christopher Rayle, BA
- 1:29 Impact of Vestibular Dysfunction on Gait
Characteristics in Older Individuals:
Preliminary Data from the Baltimore
Longitudinal Study of Aging**
Andrew J. Layman, BS
Carol Li, BS
Luigi Ferrucci, MD, PhD
Danielle Kauffman, BS
John P. Carey, MD
Yuri Agrawal, MD

- 1:37 Cholesteatoma in Congenital Aural Stenosis**
Garrett G.A. Casale, BS
Brian D. Nicholas, MD
Bradley W. Kesser, MD
- 1:45 Review of Otorrhea Microbiology: Is There a Pathogenic Role of Corynebacter?**
Matthew G. Crowson, MD
Katherine Callahan
James E. Saunders, MD
- 1:53 Osteoradionecrosis of the Temporal Bone: A Case Series**
Jeffrey D. Sharon, MD
Shariq S. Khwaja MD, PhD
Andrew Drescher, MD
Hiram Gay, MD
Richard A. Chole, MD, PhD
- 2:01 Lateral-to-Malleus Underlay Tympanoplasty: Surgical Technique and Outcomes**
Robert J. Yawn, MD
Matthew L. Carlson, MD
David S. Haynes, MD
Alejandro Rivas, MD
- 2:09 DISCUSSION**
- 2:16 BASIC SCIENCE LECTURE**
“The Gray Area - Tinnitus and the Brain”
Josef P. Rauschecker, PhD, DSc
- 2:36 DISCUSSION**
- 2:45 BREAK WITH EXHIBITORS**
- 3:15 Early Hearing Preservation Outcomes following Cochlear Implantation**
Kathryn M. Van Abel, MD
Douglas P. Sladen, PhD
Brian A. Neff, MD
Charles W. Beatty, MD
Colin L.W. Driscoll, MD
- 3:23 RESIDENT RESEARCH TRAVEL AWARD**
Intracochlear Electrocochleography in Cochlear Implant Patients
Nathan H. Calloway, MD
Adam P. Campbell, MD
Claire Iseli, MBBS, FRACS
Craig A. Buchman, MD
Douglas C. Fitzpatrick, PhD
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- 3:31 Peak Intracochlear Pressure and Ossicular Displacement at Very High Sound Intensities**
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Travis J. Pfannenstiel, MD
Herman A. Jenkins, MD
Daniel J. Tollin, PhD
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- 3:39 Predicting Performance Outliers in Adult Cochlear Implant Recipients**
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Anthony M. Tolisano, MD
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J. Thomas Roland Jr., MD
Susan B. Waltzman, PhD
- 3:47 Modulation of Reverberation Time Influences Musical Enjoyment with Cochlear Implants**
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Dean M. Mansuco, AuD
Divya A. Chari, BS
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- 3:55 DISCUSSION**
- 4:02 PANEL – “Single-Sided Deafness: Why Not a CI?”**
Bruce J. Gantz, MD - Moderator
Rene H. Gifford, PhD
Lawrence R. Lustig, MD
Jay T. Rubinstein, MD, PhD
J. Thomas Roland Jr., MD
Colin L. W. Driscoll, MD
David S. Haynes, MD
Marlan R. Hansen, MD
- 5:00 INTRODUCTION OF INCOMING AOS PRESIDENT**
D. Bradley Welling, MD, PhD
- 5:05 ADJOURNMENT**
- 6:30 AOS PRESIDENT'S RECEPTION & BANQUET**
(Members and Invited Guests -advance ticket purchase required)
Pompeian Ballroom I/IV

An Open Label Study to Evaluate the Safety and Efficacy of Intratympanic Golimumab Therapy in Patients with Autoimmune Inner Ear Disease

*M. Jennifer Derebery, MD; Courtney Voelker, MD, PhD
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Objective: To evaluate the safety and efficacy of intratympanic golimumab (GLM), a TNF alpha inhibitor, as a steroid-sparing agent for patients with steroid-dependent autoimmune inner ear disease (AIED).

Study Design: Open label

Setting: Tertiary referral center

Subjects & Methods: Ten patients with steroid-dependent AIED were enrolled. Stage 1 consisted of 3 patients given a single intratympanic GLM injection, followed for 30 days assessing for adverse events. Ten patients were then enrolled for four injections of GLM during which they were also taken completely off of oral prednisone. The average patient age at enrollment was 59, with an average of 12.5 years from the start of bilateral hearing loss symptoms. The average dose of daily prednisone was 18mg.

Main Outcome Measure: The primary endpoint, hearing loss progression (treatment failure), defined as either an increase in pure tone thresholds OR a decrease in word recognition score.

Results: Nine patients experienced stable pure-tone thresholds in the injected ear, while seven of the ten had stable word recognition scores. Two patients experienced an improvement in word recognition scores. Seven were able to completely taper off of steroids, and all 7 experienced stable pure-tone thresholds, while five had stable word recognition (71%). These results exceeded the hypothesized 30% showing stability ($p=0.0001$). There were no serious adverse events.

Conclusion: The results indicate that intratympanic GLM could potentially provide AIED patients with a steroid-holiday, but additional, randomized studies are needed.

Define Professional Practice Gap & Educational Need: Pressing need for alternative treatments in autoimmune inner ear disease

Learning Objective: Present an alternative therapy for autoimmune inner ear disease

Desired Result:

- 1) Understand more about diagnosing and treating autoimmune inner ear disease,
- 2) Understand empirical evidence for golimumab treatment

IRB Approved

Creation of Microperforation Significantly Enhances Diffusion across Round Window Membrane

*Catherine M. Kelso, BS; Hirobumi Watanabe, PhD
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Jeffrey W. Kysar, PhD; Anil K. Lalwani, MD*

Hypothesis: Creation of microperforations in round window membrane(RWM) will allow reliable and predictable intracochlear delivery of pharmaceutical, molecular or cellular therapeutic agents.

Background: A means for reliable delivery into the inner ear for therapeutic purposes remains a formidable challenge. The RWM is an attractive target for intracochlear delivery. However, simple diffusion of medications across an intact RWM is limited by what material can be delivered, size of material to be delivered, difficulty with precise dosing, timing, and precision of delivery over time. Further, absence of reliable method for measuring diffusion across RWM in vitro is a significant experimental impediment.

Methods: A novel model for measuring diffusion across a guinea pig RWM, with and without microperforation, was developed and tested: the cochlea, sparing the RWM, was embedded in a 3D-printed acrylic holder using hybrid dental composite and light cured to adapt the round window niche to a 3ml Franz Cell. Microperforation was created with a 100 μ m diameter needle and examined with light microscopy. Diffusion of 10 μ M rhodamine B across RWM in a Franz Cell was measured via fluorometry.

Results: The Franz Cell apparatus provided reliable and replicable measurement of diffusion across RWM. The 100 μ m needle produced a 240 μ m x 460 μ m elliptical perforation in the membrane and was associated with a 1000-fold enhancement in diffusion ($p < 0.05$).

Conclusion: Microperforation in RWM is an extremely effective means of increasing diffusion across the RWM. However, better microneedle design, based on the mechanical properties of RWM, is needed for creating precise microperforation without tearing.

Define Professional Practice Gap & Educational Need: Inner ear therapy based on diffusion across an intact round window membrane is associated variable efficacy.

Learning Objective:

1. To understand the normal function of the round window membrane in the inner ear.
2. To understand how creations of microperforation can enhance inner ear delivery of therapeutic agents.

Desired Result: Development of technology to facilitate inner ear delivery of therapeutic agents will lead to more reliable patient outcome.

IRB Approved

Endolymphatic Shunt Surgery and Intratympanic Gentamycin: An Uncontrolled Retrospective Comparison

*Il Joon Moon, MD, PhD; Justin S. Golub, MD
Jay T. Rubinstein, MD, PhD*

Objective: To report audiovestibular outcomes of endolymphatic shunt surgery (ELS) and intratympanic gentamicin injections (ITG) in patients with Meniere's disease (MD) Study Design: Retrospective review

Setting: Tertiary referral center

Patients: Patients with MD refractory to medical management between 2004 and 2012: 30 patients underwent ELS by a single surgeon and all had outcomes available while 25 patients underwent ITG and 21 had outcomes available. Mean follow-up duration for the ELS and ITG groups were 23.1 and 20.0 months, respectively.

Interventions: ELS or ITG

Main Outcome measure: Successful control of vertigo, pure-tone average (PTA; 0.5, 1, 2 and 4 kHz), word recognition score (WRS), and complications of treatment Results Control of vertigo was achieved in 80.0% of the ELS group and in 71.4% of the ITG group. The mean PTA and WRS in the ELS group before and after surgery changed from 43.9 dB and 69.6% to 40.0 dB and 78.3%, respectively. The mean PTA and WRS in the ITG group before and after treatment changed from 60.7 dB and 44.2% to 63.6 dB and 37.4%, respectively. The change in WRS for the ELS group (+8.7%) was significantly favorable to the ITG group (-6.8%) (P=0.011). Post-treatment unsteadiness was reported in 42.9% of the ITG group and remained chronic in 23.8%. This symptom was not reported in the ELS group.

Conclusions: ELS provided successful vertigo control at least as well as ITG with a lower rate of audiovestibular complications. Surprisingly, a small but statistically significant improvement in word recognition was observed.

Define Professional Practice Gap & Educational Need: Lack of contemporary knowledge to provide optimum treatment to patients with medically refractory Meniere's Disease

Learning Objective: To report audiovestibular outcomes of endolymphatic shunt surgery and intratympanic gentamicin injections in patients with Meniere's disease

Desired Result: Endolymphatic shunt surgery provided successful vertigo control at least as well as intratympanic gentamicin injection in patients with medically refractory Meniere's Disease with a lower rate of audiovestibular complications.

IRB Approved

Characterization of Basilar Membrane Rupture Forces in Fresh Human Cadaveric Cochleae

*Daniel Schuster, MD; Louis B. Kratchman, BS
Robert F. Labadie, MD, PhD*

Hypothesis: To develop a method to measure the forces required to rupture the basilar membranes (BM) of fresh human cochleae.

Background: Damage to BM during cochlear implantation may lead to suboptimal audiological outcomes. Prior work investigating the rupture force of human BM comes from a single study conducted on isolated, ex-vivo cadaveric specimens which did not account for in-vivo structural support.

Methods: Fresh (post mortem <120 hours), non-fixed, never-frozen human temporal bones underwent preparation consisting of surgical isolation of the cochleae and exposure of the BM by removing bone covering scalae tympani and vestibuli without disrupting attachments to the spiral ligament or osseous spiral lamina. Each isolated cochlea was mounted to a six-axis load cell using a positionable mounting stage. A 300 μ m diameter ball-tipped probe was attached to a piezoelectric linear motor and advanced at 1mm/sec through the BM while recording force from the load cell concurrent with video.

Results: Five BM's were successfully exposed. The range of rupture forces was 42 to 107 mN with a mean of 89 mN. Four of the five specimens failed via rupture with rapid release of strain energy upon rupture (linear loading) while one failed by the BM being pulled away from its attachment to the osseous spiral lamina.

Conclusions: Our novel technique and data suggest that the average force required to rupture the BM is less 100 mN. Correlation to human perception ability is necessary to determine if this force is detectable during cochlear implant surgery.

Define Professional Practice Gap & Educational Need:

1. Lack of knowledge about basilar membrane rupture characteristics

Learning Objective:

1. To understand rupture characteristics of the basilar membrane as they apply to cochlear implantation

Desired Result:

1. Attendees will have an increased understanding of the mechanical properties of the basilar membrane and apply this knowledge during cochlear implantation.

IRB Exempt

Endolymphatic Sac Surgery for Meniere's Disease: A Systematic Review and Meta-analysis

*Amit J. Sood, BA; Paul R. Lambert, MD
Shaun A. Nguyen, MD, MA; Ted A. Meyer, MD, PhD*

Objective: To analyze current endolymphatic surgery techniques and quantify their efficacy in controlling vertigo and maintaining hearing in the short and long term.

Data Sources: A comprehensive literature search using the PubMed-NCBI database from 1970-2013.

Study Selection: Articles on sac decompression and mastoid shunt (with and without silastic) were included. Included studies had to report data using the 1985 or 1995 AAO-HNS guidelines, describe surgical technique in detail, include a minimum of 10 patients, and have minimum 12 months follow-up.

Data Extraction: Endpoints were vertigo control and hearing preservation using AAO-HNS guidelines. Analysis included short term (> 12 months) and long term (>24 months) follow-up.

Data synthesis: Data analysis was performed using MedCalc 12.7.0. Each article was weighted according to number of patients treated. Analysis of pooled proportion was performed, and Freeman-Tukey transformation was utilized to correct for probably variance. A T-Test (of proportions) was performed to compare differences between groups.

Conclusions: Endolymphatic sac surgery (sac decompression or mastoid shunt) is effective at controlling vertigo in the short term (>1 year follow-up) and long term (>24 months) in at least 75% of Meniere's patients who have failed medical therapy. Sac decompression and mastoid shunting techniques provide similar vertigo control rates. Mastoid shunting, with and without silastic, also provides similar vertigo control rates. Non-use of silastic, however, appears to maintain stable or improved hearing in more patients compared to silastic sheet placement. The data suggests that once the sac is opened, placing silastic does not add benefit and may be deleterious.

Define Professional Practice Gap & Educational Need:

Endolymphatic sac surgery is the most common operative procedure performed for medically intractable Meniere's disease in the United States. Currently, however, no objective systematic analysis on endolymphatic sac surgery exists in the literature. This study was undertaken to systematically analyze current endolymphatic sac surgery (sac decompression and mastoid shunt) procedures using the AAO-HNS guidelines to better quantify their efficacy in controlling vertigo and preserving hearing in the short and long term.

Learning Objective:

1. To analyze current endolymphatic surgery techniques and quantify their efficacy in controlling vertigo and maintaining hearing in the short and long term.
2. To determine if there are any differences amongst current endolymphatic sac surgery techniques in controlling vertigo and preserving hearing in the short and long term.

Desired Result: 1. Providers should be able to inform patients undergoing endolymphatic sac surgery of expected outcomes (vertigo control, hearing preservation) in the short and long term.

2. Providers should be able to better understand any differences amongst current endolymphatic sac surgery techniques in controlling vertigo and preserving hearing in the short and long term.

IRB Exempt

Pöschl Reconstruction in Assessment of Vestibular Aqueduct

*Bryan A. Lanzman, MD; Benjamin J. Rubinstein, MD
Daniel S. Chow, MD; Angela Lignelli, MD
William Millar, MD; Anil K. Lalwani, MD*

Hypothesis: The vestibular aqueduct is more easily identified and measured with Pöschl reconstructions (parallel to the superior semicircular canal) compared to traditional axial planes.

Background: Large vestibular aqueduct (VA) is a common inner ear malformation associated with SNHL. Generally, it is considered to be large if $>1.5\text{mm}$ at its midpoint between the external aperture and the common crus on axial plane. However, determination of midpoint can be difficult as the structure curves in the shape of “J” and the exact threshold for “large” ($>1.0\text{vs}1.4\text{vs}1.5\text{vs}2.0$) remains controversial.

Methods: The size of VA in both axial and Pöschl planes for each ear was measured. Additionally, the ease of visualization for each VA was scored from 1-4 (1 difficult to visualize and 4 easy to visualize). Differences between measurements and visualization score was assessed using paired t-tests.

Results: A total of 96 VAs were assessed from 48 consecutive patients. The mean diameter of the VA between axial and Pöschl plane was 0.71 cm and 0.70 cm, respective ($p = 0.74$). Ease of visualization was significantly higher for Pöschl view (2.9/4.0), where the entire course of VA could be visualized in a single image, compared to axial planes (2.2/4.0) ($p < 0.001$).

Conclusions: The Pöschl reconstruction is a useful tool in visualizing the vestibular aqueduct. In comparison with axial images, visualization of the VA was significantly easier and no significant difference in midpoint measurements was observed. Pöschl reconstruction has the potential to be used alone or in combination with axial measurement in defining LVA.

Define Professional Practice Gap & Educational Need:

1. The definition of large vestibular aqueduct remains controversial.
2. Vestibular aqueduct visualization and measurement is difficult on axial images of CT of temporal bones.

Learning Objective:

1. To understand the difference between the axial view and Pöschl reconstruction.
2. To understand the role of Pöschl reconstruction in evaluation of vestibular aqueduct.

Desired Result: The participant will understand the utility of Pöschl reconstruction in evaluation of vestibular aqueduct.

IRB Approved

RESIDENT RESEARCH TRAVEL AWARD

**The Relationship of Age on Radiographic Incidence
of Superior Semicircular Canal Dehiscence
in Pediatric Patients**

*Neal M. Jackson, MD; Laveil M. Allen, MD
Anagha Kakade, MSC, DBM, PDCR
Anita Jeyakumar, MD, MS; Christopher Arcement, MD
Moises A. Arriaga, MD, MBA; Yu-Lan Mary Ying, MD*

Objective: To determine if age affects radiographic incidence of superior semicircular canal dehiscence (SSCD) in pediatric patients.

Study Design: Retrospective Case Review

Setting: Tertiary Children's Hospital

Patients: Patients (0-18 years) with high-resolution computer tomography (CT) temporal bone scans from April 2001 to February 2013

Interventions: Diagnostic high-resolution computer tomography (CT) temporal bone scans

Main Outcome Measures: Findings of dehiscent, thin or normal superior semicircular canal (SSC) on CT scans (including reconstructed views). Inter-observer radiographic interpretation rate between neuroradiologist and otologist.

Results: Seven-hundred CT scans (1400 ears) were reviewed, and 1188 ears were acceptable for analysis. Twenty-three ears (1.9%) had dehiscent SSC, 185 ears (15.6%) had thin SSC, and 980 ears (82.5%) had normal SSC. Median ages of dehiscent, thin, and normal canals were 5, 7, and 9 years, respectively, (p value <0.05). As age increased, the incidence of dehiscent and thin SCC cases decreased; for example, dehiscent or thin canal existed in 51.4% of children less than 12 months, 17.5% of children between 1-2 years, 18.5% of children between 3-10 years, and 10.9% of children between 11-18 years. The Kappa value of agreement between neuroradiologist and otologist was 0.814, demonstrating a high value of agreement ($p < 0.05$).

Conclusions: Pediatric SSCD, though uncommon, appears to be more prevalent in younger children, especially infants less than 12 months. This suggests that the SSC may develop more bony covering with age.

Define Professional Practice Gap & Educational Need: Lack of knowledge about relationship of age on incidence of superior semicircular canal dehiscence in pediatric population.

Learning Objective: Compare incidence of dehiscent, thin, or normal semicircular canal among different pediatric age groups.

Desired Result: Attendees will better understand the effect of age on the incidence of dehiscent and thin superior semicircular canals in pediatric patients.

IRB Approved

Sound Location Modulation of Electrocochleographic Responses in Chinchilla with Single Sided Deafness and Fitted with an Osseointegrated Bone-conducting Hearing Prosthesis

*Stéphane Tringali, MD, PhD; Kanthaiiah Koka, PhD
Herman A. Jenkins, MD; Daniel J. Tollin, PhD*

Hypothesis: Bone anchored hearing systems (BAHS) provide sound-location dependent input to the normal ear for reducing the head shadow effect in the case of single sided deafness (SSD).

Background: Patients suffering from SSD can be fit with a BAHS positioned on the impaired side. Despite successful outcomes, and some reports of spatial hearing capabilities, little data is available regarding the physiological performance of BAHSs in response to free field sounds.

Methods: Cochlear microphonics (CM) were recorded from 5 chinchillas before and after destruction of one cochlea. A BAHA® (Cochlear) was fitted on the deafened side. CM measurements were made in response to tones, with and without the BAHA, to free-field sounds presented ipsilateral to the SSD, on the side of the normal ear, and along the midline. Stimuli were also presented directly through the BAHA and an earphone to generate sounds with inter-aural time (ITD) and level (ILD) differences approximating free-field sounds.

Results: With the BAHA, CM thresholds were decreased (re: no BAHA) by ~10 dB for sources ipsilateral to the SSD, ~14 dB for midline sources and ~5 dB for sources contralateral to the SSD. Changes in CM amplitudes and thresholds were sound location dependent. CM amplitudes were modulated by ITDs and ILDs generated by the interaction of BAHA and acoustic signals.

Conclusions: This study suggests that BAHS can provide input to the normal ear that is modulated by sound location which serves to reduce the head shadow effect and may also offer cues to sound location.

Define Professional Practice Gap & Educational Need: Bone anchored hearing systems applied to cases of single sided deafness are reported to reduce the head shadow effect for sounds on the deaf side and some studies report increased sound localization capabilities.

Learning Objective: Provide a physiological understanding of how bone vibration and acoustic inputs are combined during free-field sound stimulation at the normal hearing cochlea.

Desired Result: Improve our understanding of how bone anchored hearing systems in cases of single sided deafness reduce the head shadow effect and possibly provide spatial location dependent information.

IRB Approved

Osseointegrated Implants for Auriclectomy and Microtia Defects: Operative Techniques and Complication Management

*Daniel J. Rocke, MD, JD; Debara L. Tucci, MD
Jeffrey Marcus, MD; Jay McClennen, AOCA, CCA, CFm
David M. Kaylie, MD*

Objective: Auriclectomy defects and microtia are challenging to reconstruct with native tissue. We describe operative techniques and complication management for patients undergoing osseointegrated implants for auriclectomy defects and microtia.

Setting: Tertiary referral center.

Patients: All patients at Duke University Medical center with auriclectomy defects or microtia treated with Vistafix osseointegrated implants from January 1, 2010 until September 16, 2013.

Interventions: Vistafix osseointegrated implantation for auriclectomy and microtia defects

Main outcome measure: Description of operative techniques, complications, and complication management.

Results: 16 patients met inclusion criteria. Five patients had microtia and atresia. Two of these patients had bilateral microtia and atresia and underwent bilateral simultaneous implantation of both Vistafix and BAHA. Two other microtia/atresia patients underwent simultaneous unilateral Vistafix and BAHA. 11 patients had unilateral defects from either trauma or skin cancer resection. Two patients received adjuvant radiation prior to implantation. Complications included tissue overgrowth requiring revision surgery (two patients), inadequate bone stock requiring split calvarial bone graft and later implantation (one patient), loss of implant secondary to osteoradionecrosis requiring hyperbaric oxygen therapy, and skin infection requiring antibiotic therapy.

Conclusions: Reconstruction of auriclectomy defects and microtia is difficult using native tissue. Complications are common, and results are often inadequate. Osseointegrated implantation offers an outstanding alternative for reconstructing these defects. We examine operative techniques and focus on the unique challenges of simultaneous and bilateral simultaneous Vistafix and BAHA implantation, as well as implantation in a post-radiated field. We also examine common complications and management options.

Define Professional Practice Gap & Educational Need:

1. Lack of published knowledge on osseointegrated implants for auriclectomy and microtia defects, especially in regards to difficult situations like bilateral simultaneous implantation or implantation in a radiated field.

Learning Objective:

1. Describe the benefits of osseointegrated implants for auriclectomy and microtia defects.
2. Recognize difficulties and common complications with osseointegrated implants.

Desired Result:

1. Attendees will be able to employ osseointegrated implants for rehabilitating auriclectomy and microtia defects.
2. Attendees will be able to recognize difficult implantation situations and manage complications associated with implantation.

IRB Approved

Management of Incus Necrosis in Revision Stapedectomy using Hydroxyapatite Bone Cement

*J. Eric Lupo MD, MS; John W. House, MD
John C. Goddard, MD*

Objective: Review audiological outcomes after revision stapedectomy using hydroxyapatite bone cement.

Study design: Retrospective case review.

Setting: Tertiary neurotological referral center.

Patients: Twenty-seven cases of previously treated otosclerosis where incus erosion was observed during revision stapedectomy.

Intervention(s): Hydroxyapatite bone cement was used to rebuild the eroded incus and stabilize the prosthesis during revision stapedectomy.

Main outcome measure(s): Pre- and post-operative bone conduction (BC-PTA) and air conduction (AC-PTA) pure tone averages (0.5, 1, 2, 3 kHz) including high tone bone conduction (1, 2, 3 kHz), air-bone gap (ABG), and speech discrimination scores (SDS) were measured. Short term (three weeks) and longer-term (median, 12 months) hearing outcomes were measured. Data are reported according to the most recent guidelines of the AAO-HNS Committee on Hearing and Equilibrium.

Results: Among 27 ears undergoing revision stapedectomy with use of hydroxyapatite bone cement, the AC-PTA was 64.4 dB pre-operatively and 35.7 dB post-operatively. The mean postoperative ABG was 9.0 dB (st dev. = 8.18, range -2.5 – 32.5 dB). Mean overclosure of 2.5 dB (st dev = 5.9, range -8.33 – 18.33 dB) of the high tone BC-PTA was observed. No further revisions or adverse events were noted during the follow-up period.

Conclusions: Hydroxyapatite bone cement is useful to reconstruct and stabilize the prosthesis in revision stapedectomy when erosion of the long process is encountered. Short and longer-term hearing results are favorable to previously reported results of revision stapedectomy.

Define Professional Practice Gap & Educational Need: Revision stapedectomy is a challenging procedure with reported results inferior to that of primary stapedectomy

Learning Objective: To learn a new technique of revision stapedectomy incorporating the use of hydroxyapatite bone cement

Desired Result: The attendee will be able to apply the knowledge from the presentation to improve outcomes in their own revision stapedectomy procedures.

IRB Approved

Revision Stapedectomy with Bone Cement: Are Results Comparable to Standard Techniques?

*Scott K. Hudson, MD; Richard K. Gurgel, MD
Clough Shelton, MD*

Objective: To examine hearing results in revision stapedectomy using hydroxyapatite bone cement for incus necrosis.

Study Design: Retrospective case series.

Setting: Tertiary-care academic medical center.

Patients: Eligible patients underwent revision stapedectomy for otosclerosis utilizing hydroxyapatite bone cement to reconstruct incus necrosis, between March 2007 and August 2013. Patients were excluded for insufficient audiometric data or an operative indication other than otosclerosis.

Intervention: Revision stapedectomy performed with standard stapes piston prostheses, utilizing hydroxyapatite bone cement to reconstruct the distal incus and stabilize the prosthesis.

Main Outcome Measure: Air bone gap (ABG), air conduction and bone conduction pure tone audiometry thresholds, and speech discrimination scores (SDS) were obtained preoperatively and postoperatively. The preoperative result closest to the date of surgery was compared with the postoperative result farthest from the date of surgery.

Results: Twenty-seven patients (age 32-83) met criteria and were included in the study. Average duration of follow up was 7 months (range 1-39 months). Postoperative closure of the ABG to within 10 dB was achieved in 77.8% of cases. Closure to within 20 dB was achieved in 96.3%. The average postoperative ABG was 8.1 dB (range 0-24 dB). The average SDS change was +7.4% (-12% to +92%). There were no cases of significant postoperative sensorineural hearing loss by bone conduction PTA.

Conclusions: Use of hydroxyapatite bone cement for treatment of incus necrosis in revision stapedectomy provides excellent hearing outcomes, and is a reasonable alternative to total ossicular reconstruction prosthesis or malleovestibular prosthesis techniques.

Define Professional Practice Gap & Educational Need:

1. Lack of awareness of management options available for revision stapedectomy.
2. Lack of awareness of expected hearing results with the technique described.

Learning Objective:

1. Attendee will learn of an alternative management strategy for revision stapedectomy.
2. Attendee will be aware of expected hearing outcomes in the described technique.

Desired Result:

1. Attendee will make an informed decision when considering the technique described for management of revision stapedectomy cases.

IRB Approved

Racial Differences in Cochlear Melanocytic Content Are Associated with Hearing Loss

*Daniel Q. Sun, MD; Linda Zhou; Frank R. Lin, MD
Howard W. Francis, MD; John P. Carey, MD
Wade W. Chien, MD*

Hypothesis: Racial differences in age-related hearing loss are associated with differential melanocytic content in the cochlea.

Background: Race is a well-known risk factor for hearing loss, with black individuals having 40-60% lower odds of hearing loss compared to white individuals. Studies have suggested that this difference in hearing loss is mediated through skin color and possibly melanocyte content in the inner ear. However, detailed analysis of melanocyte location and content is lacking in human ears.

Methods: Human temporal bone specimens from 20 age- and gender-matched black and white individuals were studied. These individuals were over 60 years of age and had audiologic evidence of presbycusis. The melanin content in the mid-modiolar section of each temporal bone was quantified using image analysis software (Image J, Bethesda MD). Statistical analysis was done using the 2-tailed t-test and a p-value of <0.05 was considered statistically significant.

Results: Melanocytes are found in highest quantity in the stria vascularis and Rosenthal's canal. In the stria vascularis, black and white individuals had average melanocytic contents of 0.1mm^2 and 0.03mm^2 , respectively ($p<0.001$). Similarly, in Rosenthal's Canal, the average melanocytic content was 0.3mm^2 and 0.05mm^2 for black and white individuals, respectively ($p<0.001$). Audiometrically, black individuals had lower pure tone averages compared to white individuals across all frequencies, reaching statistical significance at 4000Hz ($p=0.006$) and 8000Hz ($p=0.009$).

Conclusions: Black individuals have significantly higher melanocyte content in the cochlea compared to age-matched white individuals, suggesting that inner ear melanocytes may have a protective role in age-related hearing loss.

Define Professional Practice Gap & Educational Need:

1. Lack of contemporary knowledge in the mechanisms through which risk factors for presbycusis lead to hearing loss

Learning Objective:

1. Describe the characteristics locations of melanin in the cochlea
2. Understand racial differences in cochlear melanin content
3. Understand the potential importance of cochlear melanin in presbycusis

Desired Result:

1. Appreciate the role of various risk factors in presbycusis
2. Trigger interest and additional research in the biology and role of melanin in the cochlea

IRB Approved

3:55 pm

Subjective Hearing Loss in Adults with Normal Thresholds May Be Improved by Auditory Training

*Louisa Ha, AuD; Samuel Trozman, MD
Neel Patel, MD; Gabrielle Cager, AuD
Miriam I. Redleaf, MD*

Objective: To improve satisfaction with hearing performance in adults with normal thresholds but subjective hearing loss.

Hypothesis: Adult with normal audiograms who nonetheless complain of impaired hearing may have central auditory processing disorders (CAPD) which can be ameliorated through auditory training.

Subjects: All 8 adults (age 24-52) with subjective hearing impairments and normal audiograms who enrolled in our program of testing and training, completed it. Thirteen other candidates initially volunteered, but withdrew upon learning the degree of commitment.

Methods: Subjects with normal pure tone thresholds underwent pre- and post-training APHAB questionnaires along with 7 tests of central processing in each ear. They spent 15-20 minutes per day, 5 days per week, for 4 weeks using the Listening and Communication Enhancement training program.

Results: On average of all subjects, 13 of 14 CAPD test scores showed improvement after training. The scores of 60 of the 112 CAPD test were improved, while 21 of 112 were lower. The averaged change in APHAB scores showed improvement (11.49) (range= -2.83 - +22.53). Six of the 8 subjects rated their hearing as improved, while 2 of 8 felt it had deteriorated on the APHAB.

Conclusion: Adults with normal audiograms who complain of hearing impairment can show improvement in tests of central auditory processing and on hearing handicap questionnaires after auditory retraining.

Define Professional Practice Gap & Educational Need: Lack of awareness

Learning Objective: Describe subjects that may benefit from auditory training and rehabilitation.

Desired Result: Attendees will be able to identify patients that may benefit from auditory training based on presenting symptoms and auditory testing.

IRB Approved

Otologic Outcomes Following Blast Injury- The Boston Marathon Experience

*Aaron K. Remenschneider, MD, MPH
Sarah A. Lookabaugh, MD; Jacob R. Brodsky, MD
Avner Aliphas, MD; Selena E. Heman-Ackah, MD
Daniel J. Lee, MD; Alicia M. Quesnel, MD*

Objective: Ear related trauma was the most common physical injury sustained following the dual explosions at the Boston Marathon on April 15th, 2013. The goal of this study is to detail the types of otologic injury, describe how subspecialty care was delivered across multiple institutions, and report on outcomes from patients undergoing otologic treatment secondary to blast injury.

Study Design: Multi-institutional, prospective cohort study

Methods: Multi-institutional IRB approval was obtained. Children and adults seen for otologic complaints in relation to the Boston Marathon bombings were invited to participate in this prospective study. Participants underwent otologic evaluation and audiometry. They completed a current symptom assessment, otologic/noise exposure history, and quality of life questionnaires at initial visits and six months after the bombing event. Subjects are being followed clinically out to one year.

Results: Over 150 patients from seven major medical campuses have been evaluated for blast-related otologic injuries. Intense collaboration among staff at all Boston sites has allowed for rapid access to otologic care, coordination of follow-up and de-identified data sharing. Patients admitted with coincident injuries tended to have near total tympanic membrane perforations. The most common outpatient presenting complaints were hearing loss and tinnitus; audiograms have demonstrated both conductive and sensorineural losses. Over twenty patients with non-healing perforations have undergone successful tympanoplasty following a period of watchful waiting. Short term follow up demonstrated closure of the perforation in all but one.

Conclusion: Blast-related otologic injuries constitute a major source of morbidity following the Boston Marathon bombings. Close follow up of this cohort is currently underway.

Define Professional Practice Gap & Educational Need:

1. Lack of awareness of the extensive otologic morbidity of the Boston Marathon Bombings
2. Lack of knowledge on how to best care and follow patients with significant acoustic trauma
3. Need to better understand how large scale clinical and research collaboration can be performed between academic ORL departments

Learning Objective:

1. Detail the extent of otologic injuries sustained at the Boston Marathon Bombings
2. Describe how sub-specialty care was delivered, including outcomes on patients undergoing surgical intervention
3. Explain how to perform large scale research and patient care collaboration across academic centers

Desired Result:

1. Attendees should gain insight into the expected otologic morbidity among civilians following blast injury
2. Attendees should gain knowledge on how to best care for the blast-affected patient
3. Attendees should leave with new information on how to conduct a large multicenter prospective study

IRB Approved at all Institutions

1:13 pm

The Socioeconomic Impact of Hearing Loss in US Adults

Susan D. Emmett, MD; Howard W. Francis, MD, MBA

Objective: To evaluate the associations between hearing loss and educational attainment, income, and unemployment/underemployment in US adults.

Study design: National cross-sectional survey.

Setting: Ambulatory examination centers.

Patients: Adults aged 20-69 years who participated in the 1999-2002 cycles of the National Health and Nutrition Examination Survey (NHANES) audiometric evaluation and income questionnaire (n = 3377).

Intervention(s): Pure tone audiometry, with hearing loss defined by World Health Organization criteria of bilateral pure tone average >25 decibels (0.5,1,2,4 kHz).

Main outcome measure(s): Low educational attainment, defined as not completing high school; low income, defined as family income less than \$20,000/year, and unemployment or underemployment, defined as not having a job or working less than 35 hours per week.

Results: Individuals with hearing loss had 3.26 times higher odds of low educational attainment (95% CI: 2.24-4.74; p <0.001) compared to normal-hearing individuals. Controlling for education, age, sex, and race, individuals with hearing loss had 1.56 times higher odds of low income (95% CI: 1.14-2.13; p = 0.007) and 1.97 times higher odds of being unemployed or underemployed (95% CI: 1.36-2.85; p = 0.001) compared to normal-hearing individuals.

Conclusions: Hearing loss is associated with low educational attainment in US adults. Even after controlling for education and important demographic factors, hearing loss is independently associated with economic hardship, including both low income and unemployment/underemployment. The societal impact of hearing loss is profound in this nationally representative study and should be further evaluated with longitudinal cohorts.

Define Professional Practice Gap & Educational Need: The relationship between hearing loss, educational attainment, and socioeconomic status in US adults has not been well defined.

Learning Objective: After this presentation, attendees will be able to describe the associations between hearing loss, education, and socioeconomic status.

Desired Result: Attendees will better understand the socioeconomic impact of hearing loss

IRB Approved

1:21 pm

Rural Barriers to Early Diagnosis of Infant Hearing Loss

*Matthew L. Bush, MD; Bryan Hardin, BS
Christopher Rayle, BA*

Objective: In spite of Early Hearing Detection and Intervention (EHDI) programs, the diagnosis of congenital hearing loss is often delayed. Rural residents, from areas of healthcare disparity such as Appalachia, may face multiple barriers to timely care. The purpose of this study is to assess regional barriers in the diagnostic and therapeutic process following abnormal newborn hearing screening testing.

Study Design: Cross sectional questionnaire study

Setting: Tertiary referral center

Patients: Parents of infants who failed newborn hearing screening from 2009-2011
Intervention and Main Outcome Measures: In collaboration with state EHDI agencies, subjects were identified and mailed a questionnaire assessing demographic information, county of origin, and perceived barriers to infant hearing screening.

Results: Of the 410 participants, 28% of parents were from a rural Appalachian region. Of the non-Appalachian subjects, 72.5% were from metro regions. 30% of Appalachian parents considered the diagnostic process difficult, compared with 16.7% of non-Appalachian parents. A higher percentage of Appalachian parents were lacking a high school diploma (OR 1.69, $p=0.028$). Medicaid insurance was more common in Appalachians (51.3% versus 29.6%) ($p=0.000$). Distance from the diagnostic/therapeutic center represented was a significant barrier for Appalachian parents (OR 3.00, $p=0.001$). Appalachian parents expressed a strong interest in telemedicine (56.8% versus 34.1%) ($p=0.000$). Of those children diagnosed with hearing loss, 52.6% of Appalachian children were diagnosed after 3 months of age compared with 39.5% of non-Appalachian children.

Conclusions: Multiple barriers can affect timely diagnosis and treatment of congenital hearing loss. Educational and telemedicine programs may benefit parents in rural regions.

Define Professional Practice Gap & Educational Need: There is a lack of knowledge of rural pediatric hearing healthcare disparities and barriers that parents face in obtaining timely care.

Learning Objective: At the conclusion of this presentation, the participants should be able to

1) Discuss factors that may prevent early identification and intervention of infant hearing loss and 2) Explain strategies that may address barrier to timely care.

Desired Result: The attendee will have an increased awareness of barriers that may impede the hearing healthcare in children from rural regions and strive to develop culturally appropriate methods to address these barriers.

IRB Approved

Impact of Vestibular Dysfunction on Gait Characteristics in Older Individuals: Preliminary Data from the Baltimore Longitudinal Study of Aging

*Andrew J. Layman, BS; Carol Li, BS
Luigi Ferrucci, MD, PhD; Danielle Kauffman, BS
John P. Carey, MD; Yuri Agrawal, MD*

Objective: To investigate the association of semicircular canal (SCC) and otolith function with age-related gait impairment.

Study design: Cross-sectional analysis within the Baltimore Longitudinal Study of Aging (BLSA), a longitudinal prospective cohort study.

Setting: Vestibular testing and gait laboratory within an acute care teaching hospital.

Patients: Community-dwelling participants age ≥ 60 without a history of vestibular disease, who did not use assistive devices.

Intervention(s): Vestibulo-ocular reflex (VOR) assessment using video head-impulse testing to measure horizontal SCC function, cervical and ocular vestibular-evoked myogenic potentials (VEMPs) as measures of saccular and utricular function respectively, and gait assessment with 3D motion capture.

Main outcome measure(s): VOR gain (ratio of eye to head velocity), presence of VEMP responses, and gait speed and variability as measures of age-related gait impairment.

Results: We have analyzed 9 subjects so far (mean age 75.1 years, 4 females). Participants with present vs. absent CVEMP responses had significantly higher gait speeds (1.30 vs. 0.93m/s, $p=0.0005$) and lower step length variability ($p=0.0565$). Gait speed was positively associated with VOR gain, and in age-adjusted analyses participants above a VOR gain threshold of 0.7 had higher gait speeds than participants below (1.24 vs. 0.95m/s, $p=0.084$). OVEMP responses were not associated with gait parameters.

Conclusions: Although vestibular function is known to decline with age, these preliminary analyses demonstrate the functional correlates of this decline: lower gait speed and increased gait variability, which are markers of mobility disability and increased fall risk. Further studies are ongoing to confirm and extend these findings in a larger sample.

Define Professional Practice Gap & Educational Need:

1. Incomplete understanding of the influence of vestibular dysfunction on gait outcomes
2. Lack of evidence-based management of vestibular disorders

Learning Objective:

1. Demonstrate correlation between vestibular dysfunction and gait characteristics
2. Propose diagnostic vestibular thresholds influencing gait parameters

Desired Result:

1. Recognize the association between specific vestibular and gait parameters
2. Understand the clinical relevance of vestibular testing to improving gait outcomes

IRB Approved

Cholesteatoma in Congenital Aural Stenosis

*Garrett G.A. Casale, BS; Brian D. Nicholas, MD
Bradley W. Kesser, MD*

Objectives: To estimate the prevalence of external auditory canal (EAC) stenosis and cholesteatoma in patients with congenital aural atresia/stenosis, to identify risk factors that may herald the development of ear canal cholesteatoma, and to report postoperative outcomes for patients with atresia/stenosis.

Study Design: Retrospective chart review

Setting: University tertiary referral center

Patients: Data from 673 patients (770 ears) with congenital aural atresia were evaluated. From this population, patients with EAC stenosis with and without canal cholesteatoma were identified.

Main outcome measure(s): Demographic data, symptoms at presentation, and audiometric data were compared between those with and those without cholesteatoma to identify risk factors for cholesteatoma. Postoperative outcomes for these patients were also tabulated.

Results: Of the 673 patients evaluated (770 ears), 87 (12.9%) were found to have at least one stenotic EAC (101 ears). Of this group with ear canal stenosis, 17 patients (19.5%) had a concurrent ear canal cholesteatoma with one patient having bilateral cholesteatomas. Demographic, clinical, and audiometric parameters showed no difference between stenotic ears with and without cholesteatoma. One month postoperative audiometric data for the cholesteatoma group was comparable to the non-cholesteatoma group and to published postoperative atresia repair outcomes.

Conclusion: Approximately 1 in 5 patients with congenital aural stenosis are at risk for the formation of cholesteatoma. Basic audiometric parameters provide no diagnostic utility in distinguishing ears with cholesteatoma from those without cholesteatoma. Audiometric outcomes are comparable between groups.

Define Professional Practice Gap & Educational Need: Identifying the presence of ear canal cholesteatoma in patients with congenital aural stenosis or atresia is critical to prevent long-term damage to the ear. This paper retrospectively reviews patients with aural stenosis to identify risk factors that may lead to the suspicion of cholesteatoma and describes the management of these patients.

Learning Objective: To estimate the prevalence of external auditory canal (EAC) stenosis and cholesteatoma in patients with congenital aural atresia/stenosis, to identify risk factors that may herald the development of ear canal cholesteatoma, and to report postoperative outcomes for patients with atresia/stenosis.

Desired Result: Attendees will understand the nature of cholesteatoma in congenital aural stenosis and be able to evaluate and manage this rare problem.

IRB Approved

Review of Otorrhea Microbiology: Is there a Pathogenic Role of Corynebacter?

*Matthew G. Crowson, MD; Katherine Callahan
James E. Saunders, MD*

Objective: To investigate the prevalence and pathogenicity of non-diphtheroid *Corynebacteria* spp. in commonly encountered purulent and mucopurulent otologic infections.

Study design: Retrospective case review

Setting: Academic tertiary referral center

Patients: Review of 140 microbiology cultures from adults and children presenting with purulent and mucopurulent otologic infections from 2010 to 2012.

Intervention(s): Clinical history and demographics, microbiology cultures with speciation and antibiotic susceptibility analyses.

Main outcome measure(s): Microbiological speciation and census, species association with clinical characteristics, antibiotic susceptibility to oral and systemic agents, therapy frequency and agent selection.

Results: Patients presenting with clinically significant otologic infections grew *Corynebacteria* spp. second to *Staphylococci* spp. in frequency. *Corynebacteria*-positive infections were significantly associated with pre-diagnosis use of hearing aids, and increased frequency of chronic myringitis diagnoses. *Corynebacteria* isolates were highly resistant to first-line fluoroquinolone therapy. The majority of patients with *Corynebacteria*-positive infections harbored at least one resistant strain, and patients with strains resistant to first-line topical therapy had a non-significant trend for an increase in antibiotic upgrade after antibiotic susceptibility assay data became available.

Conclusions: Identification of a causative pathogen in otologic infections assists with maximizing therapy efficacy particularly in refractory and chronic cases. Current standards for treatment of otologic infections include anti-staphylococcal, anti-streptococcal, and anti-pseudomonal antibacterial agents on the assumption that these targeted species are the most common pathogens. However, other species, such as *Corynebacteria* spp., may be underestimated in their potential to cause clinically significant otologic infections. Our results indicate a potential need for expanding surveillance for *Corynebacteria* spp. that are not routinely cultured.

Define Professional Practice Gap & Educational Need: Lack of contemporary knowledge and research of *Corynebacteria* spp. as potential pathogens in otologic infections.

Learning Objective:

To demonstrate the prevalence of non-diphtheroid *Corynebacteria* spp. in otologic infections.

To demonstrate species association with clinical characteristics, antibiotic susceptibility to oral and systemic agents, therapy frequency and agent selection.

To discuss the potential benefit of expanding surveillance for *Corynebacteria* spp. in otologic infections, particularly in patients with hearing aids, chronic infections, and infections that are refractory to first line therapy.

Desired Result: It is the authors' hope that attendees will consider the role of *Corynebacteria* spp. as potential pathogens in patients with otologic infections, and consider expanding surveillance for *Corynebacteria* spp. that are not routinely cultured from these patients.

IRB Approved

Osteoradionecrosis of the Temporal Bone: A Case Series

*Jeffrey D. Sharon, MD; Shariq S. Khwaja, MD, PhD
Andrew Drescher, MD; Hiram Gay, MD
Richard A. Chole, MD, PhD*

Objective: To study osteoradionecrosis (ORN) of the temporal bone

Study Design: Retrospective case review

Setting: Academic medical center

Patients: Patients were included who had previously undergone radiation to the head and neck and then developed exposed necrotic bone within the ear canal that persisted at least three months

Intervention(s): Patients were treated with a variety of modalities, including conservative therapy with antibiotic ear drops and in-office debridements, hyperbaric oxygen therapy and surgery.

Main Outcome Measure(s): To describe the presentation and management of patients with temporal bone osteoradionecrosis.

Results: 33 patients with temporal bone osteoradionecrosis were included. The most common site of primary tumor was the parotid gland (n=11), followed by the nasopharynx (n=7). The time to development of ORN varied between 1 and 22 years, with mean 7.9 years. The mean radiation dose was 62.6 Gy to the primary tumor, 53.1 Gy to the affected temporal bone, and 65.2 Gy to the affected tympanic bone. The most common symptoms of ORN were otorrhea (n=15), hearing loss (n=13), and otalgia (n=12). 15 patients had bacterial superinfection, most commonly *S. aureus* (n=9). Conservative therapy was successful at managing symptoms but not in eradicating exposed bone in most patients. Surgery was used for recalcitrant pain, infection, cholesteatoma, cranial neuropathies, and intracranial complications.

Conclusions: Osteoradionecrosis is a rare complication of radiation to the temporal bone. Management should be aimed at relief of symptoms, eradication of superinfection, and treatment of other commonly present radiation effects like cholesteatoma and hearing loss.

Define Professional Practice Gap & Educational Need: Osteoradionecrosis of the temporal bone is a rare complication of radiation to the head and neck. As such, there is little information available about its presentation and management.

Learning Objective: To use information gathered from the largest case series of osteoradionecrosis (ORN) of the temporal bone to date to review the presentation, risk factors, associated symptoms, pathology, microbiology, clinical course, and treatment of ORN of the temporal bone.

Desired Result: Attendees will be able to better diagnose and manage osteoradionecrosis of the temporal bone.

IRB Approved

Lateral-to-Malleus Underlay Tympanoplasty: Surgical Technique and Outcomes

*Robert J. Yawn, MD; Matthew L. Carlson, MD
David S. Haynes, MD; Alejandro Rivas, MD*

Objective: The lateral-to-malleus underlay tympanoplasty involves dissection of the tympanic membrane remnant from the malleus with subsequent graft placement medial to the annulus but lateral to the malleus. The objective of the current study is to describe the clinical outcomes using the lateral-to-malleus underlay tympanoplasty technique.

Study Design: Retrospective series

Setting: Single tertiary otologic referral center

Patients: One hundred thirty patients undergoing lateral-to-malleus underlay tympanoplasty. In order to isolate the effects of tympanoplasty on audiometric outcome, only cases with an intact and mobile ossicular chain were evaluated; ears with ossicular discontinuity, fixation and/or ossicular chain reconstruction were excluded.

Intervention(s): Lateral-to-malleus underlay tympanoplasty

Main Outcome Measures: 1) Change in air-bone gap and bone conduction thresholds; 2) tympanic membrane lateralization; 3) primary and delayed graft failure

Results: One hundred thirty tympanoplasties were analyzed. In the early postoperative period, 129 of 130 (99%) grafts were intact and at a mean of 2.8 years following surgery, 111 (85%) remained without significant retraction or re-perforation. There were no cases of early or delayed graft lateralization. The mean preoperative and most recent postoperative air-bone gap was 22.9 dB and 14.0 dB, respectively ($P < 0.0001$). Overall, there was less than a 0.5 dB decline in bone conduction thresholds following surgery ($p = 0.88$).

Conclusions: The lateral-to-malleus underlay tympanoplasty technique offers improved exposure of the tympanic space and a low rate of graft failure. The risks of sensorineural hearing loss with ossicular chain manipulation and tympanic membrane lateralization with graft placement lateral to the malleus are very low.

Define Professional Practice Gap & Educational Need: There currently exists a lack of contemporary knowledge concerning the lateral-to-malleus underlay tympanoplasty. There is anecdotal belief that this technique is associated with an increased risk of graft lateralization and/or sensorineural hearing loss. Elevating the tympanic membrane off of the malleus affords improved exposure of the tympanic space, and graft placement lateral the malleus but medial to the annulus provides an addition point of graft support.

Learning Objective: The learning objective of this study is to understand clinical outcomes using lateral-to-malleus underlay tympanoplasty. Specifically, the authors demonstrate that this technique carries a very low risk of graft lateralization, and sensorineural hearing loss and affords reliable and durable repair.

Desired Result: The desired result is that attendees will understand the true risks and benefits of lateral-to-malleus underlay tympanoplasty so that this technique can be applied to cases that involve pathology anterior to the malleus, where conventional underlay tympanoplasty provides limited access.

IRB Approved

3:15 pm

Early Hearing Preservation Outcomes following Cochlear Implantation

*Kathryn M. Van Abel, MD; Douglas P. Sladen, PhD
Brian A. Neff, MD; Charles W. Beatty, MD
Colin L.W. Driscoll, MD*

Objective: Evaluate early hearing preservation in patients undergoing implantation with the cochlear implantation

Study design: Prospective cohort.

Setting: Tertiary academic institution.

Patients: Patients undergoing cochlear implantation with a preoperative PTA of ≤ 80 dB from 5/9/2012-4/9/2013.

Interventions: Cochlear implantation.

Main outcome measure(s): Surgical and audiometric data were analyzed. Audiometric variables included hearing thresholds and shifts, speech perception, absolute hearing loss (average PTA at 250 and 500 Hz >120 dB), and functional hearing preservation (average PTA at 250 and 500 Hz <80 dB, <85 dB).

Results: 26 patients met criteria (average age 65 years; range: 13-85). Insertion was round window in 25 and cochleostomy in 1. A single dose of IV dexamethasone was given intraoperatively in 23/26, Healon was used in 13/26, and depth of insertion was to the first marker in 6, between the first and second markers in 6 and to the second marker in 14 patients. Six-month results show 26.9% maintained better than 80dB HL PTA and 42.3% maintained better than 85dB HL PTA. Complete hearing loss has occurred in 5/26 participants. Average CI sentence understanding score is 75%.

Conclusions: New technology allows the surgeon to combine a thin, flexible electrode with a minimally traumatic round window insertion. Hearing preservation is possible in the majority of patients, with functional hearing preservation achievable in over 1/3. The potential for combining acoustic and electrical cochlear stimulation in these patients postoperatively is promising. More study is needed to determine optimal surgical and medical management to maximize hearing preservation.

Define Professional Practice Gap & Educational Need: Lack of awareness regarding hearing preservation in cochlear implantation and options available for hearing preservation implantation.

Learning Objective:

- 1) To review the potential for hearing preservation in patients undergoing cochlear implantation with functional preoperative hearing.
- 2) To discuss our experience with regard to hearing preservation implantation.

Desired Result: Attendees will have a better understanding of hearing preservation cochlear implantation, will be better able to make informed decisions, and will gain an appreciation for the future potential for attaining hybrid cochlear stimulation (acoustic and electrical).

IRB Approved

RESIDENT RESEARCH TRAVEL AWARD

**Intracochlear Electrocochleography
in Cochlear Implant Patients**

*Nathan H. Calloway, MD; Adam P. Campbell, MD
Claire Iseli, MD; Craig A. Buchman, MD
Douglas C. Fitzpatrick, PhD; Oliver F. Adunka, MD*

Hypotheses: Electrocochleography to acoustic stimulus is feasible with an intracochlear electrode. Responses demonstrate greater amplitudes than at the round window (RW) and can be used to detect intracochlear damage during cochlear implant (CI) electrode insertions.

Background: Cochlear responses to sound are present in nearly all CI recipients when measured at the RW. The goal of this study is to compare RW to intracochlear recording sites, and to determine if cochlear function can be monitored during insertion of the electrode array.

Methods: Auditory stimulation (500 Hz tones at 85-95 dB HL) and ECoG recordings were made intraoperatively in patients receiving a CI from the RW and from just inside the scala tympani (n=24). Further recordings were obtained during 20 mm insertions of a custom-designed temporary electrode (n=6). Response magnitudes were measured as the sum of the first and second harmonics.

Results: All patients had measureable responses at the RW. The response magnitude increased by an average of 6.54 dB with intracochlear electrode placement. In 4 of the 6 longitudinal insertions, increasing signal amplitudes were observed with increasing depth of insertion. In these cases, a return to baseline was observed with withdrawal of the electrode. Two tracks showed peak amplitudes during the insertion and decreasing amplitudes with increasing depth of insertion, and the response was below baseline after the electrode was withdrawn, indicating cochlear damage.

Conclusions: Intracochlear ECoG responses are typically greater than those measured at the RW. Increasing response magnitudes indicate non-damaging insertions, while reductions in magnitude might suggest cochlear trauma.

Define Professional Practice Gap & Educational Need: Cochlear implantation is currently performed for hearing loss by placing an electrode at a theoretically optimal distance in the cochlea. Aside from tactile and distance information, the surgeon is left with little adjunct feedback to ensure optimal electrode placement. There is also a lack of knowledge of the electrocochlear environment during electrode implantation. Intracochlear electrocochleographic information has the potential to allow the surgeon a more optimal electrode placement and, hopefully, a greater patient response to cochlear implantation.

Learning Objective: The reader will learn how intracochlear electrocochleography is performed and how to interpret the information obtained from these analyses.

Desired Result: Attendees will attain a greater degree of knowledge of intraoperative electrocochleography during cochlear implantation.

IRB Approved

Peak Intracochlear Pressure and Ossicular Displacement at Very High Sound Intensities

*Nathaniel T. Greene, PhD; Travis J. Pfannenstiel, MD
Herman A. Jenkins, MD; Daniel J. Tollin, PhD
James R. Easter*

Hypothesis: That intracochlear (IC) pressure is directly proportional to stapes displacement when exposed to very high intensity sounds.

Background: The stapes is fixed over the oval window by the stapedial annular ligament, which limits total peak-to-peak displacement of the stapes to 50-100 μ m in humans. At moderate sound levels IC pressure is directly proportional to stapes displacement, thus current models (e.g. AHAAH) of middle ear function predict that peak IC pressure will vary linearly with stapes displacement up to this displacement limit; however, no test of this hypothesis has been performed.

Methods: Human cadaveric temporal bones were prepared by mastoidectomy and extended facial recess to expose the ossicular chain. Pressure measurements were made in scala vestibuli (SV), tympani (ST), and external auditory canal (EAC), concurrently with velocity (scanning laser Doppler vibrometry) measurements along the ossicular chain. Stimuli were high intensity, low frequency tones (~150dB SPL; 20-1280Hz) and impulses (simulated blast waveforms; ~170dB peak SPL).

Results: IC pressure varied linearly with stapes displacement over a range of moderate to high intensity stimuli, with SV transfer functions showing 10-20 dB gain with respect to the stimulus, consistent with prior measurements at lower sound levels. Stapes displacement reached its limit (50-100 μ m) in response to impulsive stimuli (>170dB SPL peak), with the corresponding SV pressure exceeding 2 kPa.

Conclusion: Results confirm that IC pressure is directly proportional to stapes motion, indicating that current models of acoustic hazard faithfully represent middle-ear energy transfer for sound intensities below the level eliciting maximal stapes displacement.

Define Professional Practice Gap & Educational Need: It is not currently known how high intensity acoustic energy is transmitted through the middle and into the inner ear.

Learning Objective: This paper begins to address this need by simultaneously measuring the ossicular motion and intracochlear pressure in response to very high energy sound presentation in cadaveric human specimens.

Desired Result: Elucidation of this mechanism will inform the development of devices designed to protect against injury resulting from such high intensity sounds.

IRB Approved

Predicting Performance Outliers in Adult Cochlear Implant Recipients

*Scott H. Troob, MD; Alison Singleton, AuD
Abbas A. Anwar, MD; Anthony M. Tolisano, MD
Yixin Fang, PhD; J. Thomas Roland Jr., MD
Susan B. Waltzman, PhD*

Objective: Numerous factors have been associated with performance outcomes in adult cochlear implant recipients. Variables often cited are length of deafness and cochlear anatomy; however, unexplained performance variability exists. This study sought to determine if performance outliers possess identifiable factors predictive of performance.

Study design: A retrospective review of adult cochlear implant patients examining demographic and performance variables including length of hearing loss, length of profound deafness, preoperative aided CNC, hearing aid use, medical history, etiology of deafness, radiographic data, electrode type and coding strategies, and postoperative performance. Preoperative prognosis was assigned as good when short length of deafness and normal cochlear anatomy were present, and poor in patients with long length of deafness or abnormal cochlear anatomy.

Setting: Tertiary referral hospital.

Patients: 777 adult patients implanted between 2000-2010.

Intervention: Therapeutic.

Main outcome measures: CNC monosyllabic word test.

Results: Twenty-two cases were identified where postoperative performance was poor despite a good preoperative prognosis (Group 1), and 19 cases where those with a poor prognosis performed well (Group 2). The control group consisted of 35 patients with a poor prognosis and poor postoperative performance. Multivariate analyses revealed that the only significant variable predictive of performance in both Groups 1 and 2 was age at onset of hearing loss.

Conclusion: A younger age at the onset of hearing loss was the only statistically significant predictor of performance. In the future, age at onset of any amount of hearing loss should be considered when counseling cochlear implant candidates regarding possible post-implantation outcomes.

Define Professional Practice Gap & Educational Need:

1. Inconsistency regarding how candidacy for cochlear implantation determined
2. Lack of awareness that occasionally patients perform either better or worse than expected preoperatively

Learning Objective:

1. Understand how prognosis for postoperative performance is assigned to candidate cochlear implant recipients.
2. Review of large series of adult cochlear implant recipients, with specific focus to factors that were predictive of performance outliers.

Desired Result:

1. After presentation attendees are expected to be able to more fully counsel cochlear implant candidates regarding expected postoperative performance.
2. Consider that age at onset of any degree of hearing loss may be predictive of postoperative cochlear implant performance.

IRB Approved

Modulation of Reverberation Time Influences Musical Enjoyment with Cochlear Implants

*Michael V. Certo, BFA, MM; Gavriel D. Kohlberg, MD
Dean M. Mansuco, AuD; Divya A. Chari, BS
Anil K. Lalwani, MD*

Objective: To identify factors that enhance the enjoyment of music in cochlear implant (CI) recipients. Specifically, we assessed the hypothesis that variations in reverberation time (RT60) may be linked to variations in the level of musical enjoyment in cochlear implant users.

Methods: Normal hearing adults (N=20) were asked to rate a novel, 20-second melody on three enjoyment modalities: musicality, pleasantness, and naturalness. Participants listened to seven different instruments play the melody, each with five levels (0.2sec, 1.6sec, 3.0sec, 5.0sec, 10.0sec) of RT60, both with and without CI simulation processing. Linear regression analysis with ANOVA was used to assess the impact of RT60 on music enjoyment.

Results: Without CI simulation, music samples with RT60=3.0sec were ranked most pleasant and most musical, while those with RT60=1.6sec and RT60=3.0sec were ranked equally most natural (all p-values < 0.005). With CI simulation, music samples with RT60=0.2sec were ranked most pleasant, most musical, and most natural (all p-values < 0.005). Samples without CI simulation show a preference for middle-range RT60, while samples with CI simulation show a negative linear relationship between RT60 and musical enjoyment, with preference for minimal reverberation.

Conclusion: Minimization of RT60 may be a useful strategy for increasing musical enjoyment under CI conditions, both in altering existing music as well as in composition of new music.

Define Professional Practice Gap & Educational Need:

1. Lack of qualitative and quantitative assessments of music enjoyment in patients with cochlear implants.
2. Delineation of factors that affect music enjoyment in cochlear implantees.

Learning Objective:

1. To understand factors that impact upon music enjoyment in patients with cochlear implants.
2. To identify ways to enhance music enjoyment in cochlear implantees.

Desired Result: To understand strategies to improve the music listening experience for the cochlear implant user.

IRB Approved

ABSTRACTS OF SELECTED POSTERS

NO. E001

Effect of CHD7 Mutation on Inner Ear Structure

*Ilene Chiu, MD; Seema R. Lalani, MD
Christine M. Eng, MD; John W. Belmont, MD, PhD
Jeffrey T. Vrabec, MD*

Hypothesis: Inner ear structure in individuals with clinical features of CHARGE syndrome is defined by the presence or absence of mutations in CHD7.

Background: CHARGE syndrome is a constellation of developmental anomalies including Coloboma, Heart anomalies, Choanal Atresia, Retardation of growth and/or development, Genitourinary anomalies, and Ear anomalies. Inner ear anomalies are recognized in these patients, though findings have not been correlated with genetic data.

Methods: Complete gene sequencing of CHD7 was performed in 59 individuals with clinical criteria suggestive of CHARGE syndrome.

Results: The prevalence of CHD7 mutations was 45.8% (27/59). Nine variants remain unclassified. Two siblings had the same CHD7 mutation, while all others were unique. Those with CHD7 mutations had a uniform appearance of the inner ear including a small vestibule, absence of the semicircular canals, and oval window anomaly. Cochlear nerve deficiency was more prevalent in individuals with CHD7 mutations ($p < 0.001$). The location of the mutation did not predict degree of inner ear malformation or hearing level. The positive predictive value for presence of CHD7 mutation in CHARGE syndrome patients with this inner ear appearance is 100%.

Conclusions: Inner ear structure is predictive of CHD7 mutation in those with features of CHARGE. The degree of hearing impairment was variable, but a high prevalence of cochlear nerve deficiency makes these individuals guarded candidates for cochlear implantation.

Define Professional Practice Gap & Educational Need: Lack of awareness of the inner ear structural anomalies present in CHD7 CHARGE syndrome patients.

Learning Objective: To inform physicians of the constellation of inner ear findings that can be predictive of CHD7 mutation status in a patient with CHARGE syndrome and its implications for cochlear implantation candidacy

Desired Result: Attendees will use the knowledge when evaluating CHARGE syndrome patients and determining if the patients would be favorable candidates for cochlear implantation.

IRB Approved

**Safety of Temperatures Generated by Endoscopes
in Transcanal Endoscopic Ear Surgery**

*Tsukasa Ito, MD, PhD; Toshinori Kubota, MD, PhD
Akira Takagi; Tomoo Watanabe, MD, PhD
Kazunori Futai, MD, PhD; Takatoshi Furukawa, MD, PhD
Seiji Kakehata, MD, PhD*

Hypothesis: Heat generated by endoscopes during transcanal endoscopic ear surgery (TEES) falls within safe levels.

Background: TEES can be performed using endoscopes with a 2.7-mm or 4-mm outer diameter coupled to either a conventional xenon or newer light-emitting diode (LED) light source. However, the combined heat generated by the endoscope tip and light source must fall within safe levels. We thus measured tympanic cavity temperatures during simulated TEES using a 3D-model of a human temporal bone.

Methods: The 3D-model was made by rapid-prototyping technology using CT data. The endoscope tip was fixed at the tympanic annulus during TEES simulation. The light sources were tested at both clinical (30% for xenon and 40% for LED) and 100% settings. Temperature was measured using thermocouples attached to three points within the tympanic cavity: promontory, horizontal portion of the facial nerve and lateral semicircular canal, as well as the endoscope tip.

Results: The maximum temperature measured within the tympanic cavity was less than 31°C at clinical settings, while the temperature rose to 47°C using a 4-mm endoscope with a xenon light source set at 100%. Temperatures measured at the tip were all safe at clinical settings, but rose dramatically to 146.2 °C for the 4-mm endoscope again when xenon was set at 100%.

Conclusion: The endoscope can be safely used in TEES at clinical settings. However, operators should not exceed clinical settings, particularly with 4-mm endoscopes with a xenon light source, to ensure temperatures generated within the tympanic cavity stay within safe levels.

Define Professional Practice Gap & Educational Need: Lack of contemporary knowledge about temperatures within the tympanic cavity generated by endoscopes in transcanal endoscopic ear surgery (TEES)

Learning Objective: To determine whether heat generated by endoscopes falls within safe levels during TEES Transcanal endoscopic ear surgery

Desired Result: A minimally invasive, secure and functional technique, will be safely and widely indicated for cholesteatoma patients.

IRB Approved

Vestibular Schwannoma Cells are viable 3-Years after Stereotactic Radiation Therapy: A Case Report with Implications for Pre-procedure Counseling

*Terry N. Platto, BA; Suzu Igarashi, BS
Maki Niihori, PhD; Allison M. Dunn, BA
Audriana Hurbon, BS; Abraham Jacob, MD*

Objective: To demonstrate that vestibular schwannoma (VS) cells removed years after stereotactic radiation are viable despite cystic degeneration of solid tumor on serial MRI scans.

Setting: Academic medical center

Patient: A 53 y/o male VS patient was treated with stereotactic radiation 3-years prior to presenting with facial numbness, dysphagia, and pyramidal motor tract dysfunction. Serial MRI demonstrated involution of solid tumor but enlargement of cystic regions in the mass. The patient underwent translabyrinthine craniotomy for marsupialization of cysts and subtotal disease removal. Neurological symptoms were immediately improved by surgery. Schwannoma cells harvested during surgery were cultured and immuno-stained for S-100 and p53 markers.

Intervention: Stereotactic radiation; translabyrinthine craniotomy; cell culture; immunohistochemistry.

Main Outcome Measures: Immunofluorescence staining of cultured schwannoma cells; comparison of radiated tumor with 2 non-radiated human VS specimens.

Results: Radiated schwannoma cells grew in culture with similar kinetics to non-radiated specimens. Both radiated and non-radiated tumors were S-100 positive. Radiated tumor cells were rounded in their morphology and demonstrated 100% p53 positivity. Only a small number of non-radiated tumors were p53 positive.

Conclusions: The use of radiation to treat benign tumors remains controversial. Our case demonstrates that viable S100 positive schwannoma cells are present years after radiation for VS, even when the solid component appears to be involuting by MRI. These residual tumor cells may acquire additional oncogenic hits at an accelerated rate. Both findings increase risk for delayed radiation failure or malignant transformation and must be specifically discussed for informed consent.

Define Professional Practice Gap & Educational Need:

1. There is a lack of awareness about the sustained viability of Vestibular Schwannomas after stereotactic radiation and the possibility of subsequent malignant transformation.
2. Inconsistencies for appropriate patient counseling:

Learning Objective:

1. To inform that Vestibular Schwannomas can remain viable after stereotactic radiation and undergo malignant transformation.
2. Address patient counseling standards with regards to aforementioned information in addition to presentation of possible unknown outcomes.
3. Implementation of standardized patient counseling protocols.

Desired Result:

Physicians will modify patient counseling to include information about the sustained viability, possible malignant transformation and limited time frame of evidence that is available regarding stereotactic radiation treatment for Vestibular Schwannomas.

IRB Approved

Tympanic Membrane Manipulation to Treat Symptoms of Patulous Eustachian Tube

*Matthew D. Brace, MSc, MD; Peter Horwich, BSc
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Objective: Patulous eustachian tube (PET) can have a significant negative impact on a patient's quality of life. Previous work has demonstrated that temporarily mass loading and stiffening the tympanic membrane significantly reduces these symptoms. This study examined KTP laser myringoplasty (LM) and cartilage tympanoplasty (CT) as a means to manipulate the tympanic membrane in order to alleviate PET symptoms.

Study Design: Retrospective case review

Setting: Academic tertiary care referral hospital

Patients: Patients were identified from the senior authors' specialty eustachian tube disorders clinic. Patients met previously established diagnostic criteria for PET. All patients had a clinically apparent flaccid segment of the eardrum, and had symptom improvement after simple mass loading of their eardrum in clinic.

Interventions: Patients in this study either received KTP LM or cartilage tympanoplasty to treat their flaccid eardrum segment in an attempt to alleviate PET symptoms.

Main Outcome Measures: Pre-operative and post-operative questionnaire scores and tympanometry measurements were compared.

Results: Patients undergoing cartilage tympanoplasty for PET had a significant reduction in their symptoms of autophony ($p \leq 0.001$), conducted breath sounds ($p = 0.001$), and aural fullness ($p = 0.009$). KTP LM did not significantly reduce symptoms.

Conclusion: Cartilage tympanoplasty provides a safe and accessible surgical option for the treatment of PET and significantly reduces the symptoms of autophony, conducted breath sounds, and aural fullness. Other surgical approaches may be necessary to alleviate loud environmental sounds caused by PET.

Define Professional Practice Gap & Educational Need: Lack of a simple satisfactory solution for patients seeking symptomatic relief of patulous Eustachian tube.

Learning Objective: To present a simple and effective novel treatment for symptoms of patulous Eustachian tube.

Desired Result: Attendees will be able to describe a novel approach to treating symptoms of patulous Eustachian tube and how to identify candidate patients.

IRB Approved

**Endolymphatic Hydrops Reversal on
3D FLAIR 3T MRI after Diuretic Treatment**

*Nopawan Vorasubin, MD; Kevin A. Peng, MD
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Akira Ishiyama, MD*

Objective: (1) To present two cases of radiographic reversal of endolymphatic hydrops (ELH) after diuretic treatment (2) Review the literature on ELH imaging with clinical correlation.

Study design: Retrospective chart review

Setting: Tertiary referral center

Patients: Two patients presented with long-standing histories of episodic vertigo, unilateral tinnitus and aural fullness and asymmetric hearing loss.

Intervention(s): Patients were evaluated with audiogram, electronystagmography and 3D FLAIR 3Tesla MRI 4 hours after intravenous gadolinium. Results were consistent with ELH involving the vestibule. Both patients were given a trial of diuretic treatment and reimaged 3-6 months later.

Main outcome measure(s): On follow-up, subjective audiovestibular symptoms were reassessed and post-treatment scans were obtained.

Results: After diuretic treatment, vestibular symptoms, tinnitus and aural fullness resolved while hearing level remained stable. Post treatment MRI revealed complete resolution of ELH. Previous treatment with intratympanic gentamicin and betahistine failed to demonstrate change in ELH on MRI, while isosorbide is the only other medical therapy to produce visualized resolution of ELH. Studies to correlate degree of ELH on MRI have shown correlation with audiogram and vestibular evoked myogenic potentials but not with calorics or electrocochleography. MRI imaging of ELH is a relatively novel technique undergoing rapid improvement, allowing for more precise differentiation between perilymph and endolymph. As the diagnostic accuracy of this tool improves, it can serve as an invaluable means to investigate efficacy of different therapeutic modalities.

Conclusions: ELH is a dynamic process that can demonstrate reversal after diuretic treatment on MRI.

Define Professional Practice Gap & Educational Need: Currently endolymphatic hydrops is a difficult diagnosis that most primary care physicians overlook, frequently misdiagnosing it as BPPV. Establishment of a consistent diagnostic modality that primary care providers have access to will aid in accurate diagnosis. Developing an objective means to measure disease response for correlation with subjective outcomes can serve as an invaluable tool in investigating efficacy of different treatment methods.

Learning Objective: To become familiar with a new method of diagnosing endolymphatic hydrops and hows these imaging characteristics correlate with patient symptoms, objective audiovestibular testing and responses to therapy

Desired Result: To encourage further investigation of this imaging modality in diagnosing and following patients with endolymphatic hydrops to develop a means to assess disease progression and response to therapy.

IRB Approved

**A Novel Bone Conduction System -
Long-term Safety and Efficacy Trial**

Richard K. Gurgel, MD; Clough Shelton, MD

Objective: To determine the safety and efficacy of an in-the-mouth bone stimulator (ITM) for patients after a twelve month period of use.

Study design: Prospective, multi-site, non-randomized patient enrollment with outcomes based on audiometric profile and self-reported assessment

Setting: Multi-institutional: private, hospital-based, and tertiary care academic medical center.

Patients: Adult patients with unilateral, acquired profound sensorineural hearing loss of any etiology

Intervention: The ITM system using a behind-the-ear (BTE) microphone which transmits acoustic information to an in-the-mouth bone stimulator. Sound conducts from the ITM, coupled with the dentition, to the contralateral ear.

Main outcome measure(s): Efficacy as measured by the Abbreviated Profile of Hearing Aid Benefit (APHAB) questionnaire and safety as measured by adverse events.

Results: 78 patients completed the study. Pre-trial average aided threshold was 17.1 dB and the post-trial average aided threshold was 18.4 dB. Mean APHAB scores improved for ease of communication (16.6 points), background noise (26.7 points), reverberation (21.2 points), and global benefit (21.9 points, $p < 0.001$ for all variables). There were eight device-related non-major adverse events including three related to BTE component, and five related to the ITM component, all of which resolved either spontaneously or by adjusting the ITM.

Conclusions: The ITM bone stimulator is a hearing prosthesis that delivers acoustic information through bone conduction coupled with the dentition. The ITM provided improvement in ease of communication, hearing in background noise, sound reverberation, and global hearing benefit. There was no detrimental effect to aided pure tone thresholds and few minor adverse events.

Define Professional Practice Gap & Educational Need:

Lack of contemporary knowledge of a novel ITM bone stimulation device

Learning Objective: To report on the safety and efficacy of ITM hearing system

Desired Result: After the presentation, attendees will understand the hearing benefits and adverse reactions associated with using an ITM hearing system

IRB Approved

**Development of an Algorithm for the Automatic
Otitis Media Image Diagnosis**

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Hypothesis: The otitis media diagnosis algorithm successfully diagnosed otitis media with accuracy higher than 85%.

Background: Otitis media (OM) is a disease prevalent among pediatric, and adult population as well. For parents or even primary care physicians, otoscopic diagnosis for OM can be a challenging task because of ear drum's complex anatomy and delicate pathologic changes during disease process. In this study, we aim to establish an OM diagnosis algorithm using image analysis and computation technologies.

Methods: We retrospectively reviewed and analyze 1000 digitized otoscopic images from 500 OM patients. The otoscopic images were analyzed through pre-processing, segmentation, noise and artifact removal, histogram adjustment, contour definition, feature extraction, image refinement, and classification steps. The OM otological features, such as injection, perforation, bulging, discharge, fluid, air bubble, cholesteatoma, translucency, or retraction, were identified and scored by 2 senior otologists. The critical patient information, key clinical history, and image findings were entered into an expert system.

Results: We proposed the automatic algorithm for predicting diagnostic categories of acute OM, acute OM with otorrhea, OM with effusion, chronic OM with perforation, and chronic OM with cholesteatoma. Moreover, we used the machine learning method to identify specific features for different categories of OM. A decision tree-like structure method is built based on these features to differentiate OM of different categories. The system successfully diagnosed OM with accuracy higher than 85%.

Conclusion: In the era of telemedicine, the algorithm established by this study, in conjunction with mobile device technology, may contribute to develop a remote automatic diagnosis system for OM.

Define Professional Practice Gap & Educational Need: lack of a diagnosis algorithm using image analysis

Learning Objective: how to develop an otitis media diagnosis algorithm from image analysis to diagnose otitis media

Desired Result: the algorithm established by this study may contribute to develop a remote automatic diagnosis system for otitis media patient care

IRB Approved

Third Window Lesions and Endolymphatic Hydrops

*Kevin A. Peng, MD; Nopawan Vorasubin, MD
Ali R. Sepahdari, MD; Akira Ishiyama, MD*

Objective: To describe a previously unreported association between radiological findings of endolymphatic hydrops and third window lesions of the inner ear.

Study design: Retrospective case review.

Setting: Tertiary referral center.

Patients: Eighty-one patients suspected of having endolymphatic hydrops (EH) underwent magnetic resonance imaging (MRI) of the inner ear; of these, 18 additionally underwent computed tomography (CT) of the temporal bones. For statistical analyses, contralateral ears were treated as independent subjects.

Interventions: MRI imaging sequences included delayed intravenous-enhanced three-dimensional fluid-attenuation inversion recovery (DIVE-3D-FLAIR), and “cisternographic” three-dimensional T2. Endolymphatic structures occupying more than 50% of the vestibule on 3D MIP images or asymmetries in perilymph signal on 3D-FLAIR were interpreted as EH. Temporal bone CTs were reviewed for evidence of third window lesions.

Main outcome measure: Statistical association between radiological findings of EH and third window lesions.

Results: Four ears in four patients were found to have simultaneous radiological evidence of EH and third window lesions (semicircular canal dehiscence). Fisher’s exact test revealed a nonrandom association between EH and third window lesions in our study population ($p = 0.0017$).

Conclusions: EH likely represents the common final pathway of a variety of inner ear maladies, including infection and trauma. We found a strong statistical association between EH and third window lesions of the inner ear. This corroborates previous observations that the bony covering of the labyrinth is thinner in ears affected by EH. Further investigation of the causality of this association, and its implications for the diagnosis and management of hydrops, is needed.

Define Professional Practice Gap & Educational Need:

1. Lack of contemporary knowledge regarding association between endolymphatic hydrops and third window lesions
2. Lack of awareness of use of MRI as a robust diagnostic tool for endolymphatic hydrops

Learning Objective:

1. To discuss the nonrandom association between endolymphatic hydrops and third window lesions discovered in our study population
2. To discuss state-of-the-art MRI protocols for use in diagnosing endolymphatic hydrops

Desired Result:

1. To become aware of overlap between clinical presentation and radiographic findings of endolymphatic hydrops and third window lesion
2. To implement MRI as a diagnostic tool for endolymphatic hydrops and Meniere’s disease

IRB Approved

Measuring Sentence Recognition Outcomes among Cochlear Implant Recipients: Variability by Age

Ahmad F. Mahmoud; Michael J. Ruckenstein, MD

Objective: We seek to identify the effects of age at cochlear implantation on sentence recognition as measured by HINT and AzBio testing.

Study Design: Retrospective chart review.

Setting: Tertiary referral center.

Patients: The records of 121 postlingually deafened adults who received cochlear implants (CIs) were reviewed.

Intervention(s): Unilateral cochlear implantation.

Main outcome measure(s): Stratification by age at implantation (AAI) was performed comparing postoperative HINT and AzBio scores under quiet, +10 dB, and +5 dB SNR conditions. Patients age 65 and older (elderly group) were compared to adult patients under 65 (control group). Preoperative PTA and duration of deafness were also recorded and analyzed.

Results: Stratification analysis revealed that patients with AAI of 60 – 79, and 80 and older performed significantly poorer than patients with AAI of 20 – 39 on HINT testing under quiet conditions. Patients were less likely to score above 90% on HINT testing under quiet, and +5 dB SNR conditions with increasing AAI. Elderly patients performed significantly worse than control group on HINT testing under +5 dB, and +10 dB SNR. The difficulty of each sentence recognition test was found to be linearly correlated with the p value for each group analysis of elderly vs. control groups; easier tests yielded a more statistically significant p value.

Conclusions: AzBio testing was found to be less dependent on AAI than HINT. These results suggest that more difficult sentence recognition testing should be utilized in the elderly population due to its relative resistance to age-dependent variation.

Define Professional Practice Gap & Educational Need: 1. Lack of knowledge regarding sentence recognition as measured by various tests among aging cochlear implant recipients.
2. Lack of contemporary knowledge regarding patient outcomes among elderly cochlear implant recipients using AzBio and HINT tests.

Learning Objective: 1. Participants will be able to describe how various sentence recognition tests are affected by patient's age at cochlear implantation.
2. Participants will be able to explain the advantages and disadvantages of the AzBio and HINT tests in measuring sentence recognition among elderly cochlear implant recipients.

Desired Result: 1. Participants will be better informed in counseling their patients on how sentence recognition outcomes following cochlear implantation will be affected by the patient's age at implantation.
2. Participants will be better informed to choose the most appropriate sentence recognition test based on the patient's age following cochlear implantation.

IRB Approved

The One-cut Meatoplasty: Novel Surgical Technique and Outcomes

*Kristen L. Hovis, BA; Matthew L. Carlson, MD
George B. Wanna, MD; Alex D. Sweeney, MD
David S. Haynes, MD*

Objective: Meatoplasty is an often-overlooked part of canal wall down (CWD) surgery but has significant consequences when performed poorly. Most traditional techniques incorporate a sizeable external ear canal meatus to support adequate cavity ventilation and permit access for in office examination and cleaning. These techniques often achieve a self-cleaning mastoid cavity but are associated with reduced cosmesis, frequent caloric disturbances, and are often challenging to fit with a conventional hearing aid. We present our results using a novel modification to conventional meatoplasty.

Study Design: Case series

Setting: Tertiary academic referral center

Patients: Consecutive patients undergoing primary CWD tympanomastoidectomy by the senior author between 2009 and 2013.

Intervention(S): Modified CWD tympanomastoidectomy incorporating the one-cut meatoplasty technique

Main Outcome Measures: 1) stenosis requiring revision meatoplasty; 2) otorrhea; 3) patient reported cosmesis

Results: Thirty ears met inclusion criteria and were analyzed. All patients underwent a primary CWD tympanomastoidectomy in conjunction with the one-cut meatoplasty technique. There were no cases of postoperative meatal stenosis and in office cavity exposure was adequate in all ears. Intermittent drainage developed in 15% of cases but was remedied by conservative measures. None of the patients reported dissatisfaction with aesthetic appearance. Operative technique and postoperative photos will be presented.

Conclusions: The one-cut meatoplasty is an effective alternative to traditional techniques, and in our experience, results in shorter convalescence and superior cosmesis than conventional meatoplasty without compromising cavity aeration or exposure. The current series demonstrates a high level of patient satisfaction with rare stenosis or need for revision surgery.

Define Professional Practice Gap & Educational Need: There currently exists a lack of contemporary knowledge concerning alternative meatoplasty techniques. Conventional meatoplasty incorporates a sizeable external ear canal meatus that is often associated with reduced cosmesis, frequent caloric disturbances, and difficulty with hearing aid fitting.

Learning Objective: The learning objective of this talk is to describe a novel modification to conventional meatoplasty and to report clinical outcomes using this technique.

Desired Result: The desired result is that surgeons will be able to use this simple and practical meatoplasty technique during canal wall down tympanomastoidectomy in order to improve cosmesis and improve ease of hearing aid fitting.

IRB Approved

**Treatment of Ear and Bone Disease in the
Phex Mouse Mutant with Dietary Supplements**

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Cliff A. Megerian, MD*

Hypothesis: We investigated the effect of phosphorous and vitamin D (calcitriol) on the otic capsule of the Phex male mouse, a murine model for endolymphatic hydrops (ELH), and whether bony changes alter the postnatal development of ELH and sensorineural hearing loss (SNHL).

Background: Phosphorus and calcitriol treatments improve the dysmorphic bone associated with X-linked hypophosphatemic rickets (XLH). Male Phex mice have XLH, which includes osteomalacia of the otic capsule and development of ELH and SNHL by postnatal day (P) 21. The therapeutic effect of these supplements, however have never been investigated on the otic capsule.

Methods: Water supplemented with phosphorus was available ad libitum. The prevention group was administered calcitriol from P7 to P40, while the rescue group received a punctuated calcitriol treatment from P20 to P40. Wild-type and Phex negative controls did not receive any supplementation. All mice underwent auditory brainstem response (ABR) testing prior to sacrifice at P40. Temporal bone histology and serum analyses were posthumously performed.

Results: Histologic analysis showed improved mineralization with less osteoid deposition in the otic capsule in both treatment groups. Serum alkaline phosphatase and fibroblast growth factor 23 (FGF-23) were elevated in all mutant mice. The rescue group showed a reduction in alkaline phosphatase compared to the Phex negative control ($p < 0.01$). Preliminary data show that progression of SNHL and ELH were not significantly affected.

Conclusions: Supplementation with phosphorus and calcitriol improves otic capsule bone morphology in the Phex male mouse but does not significantly alter development of ELH or SNHL.

Define Professional Practice Gap & Educational Need: There remains a knowledge gap regarding the etiology of endolymphatic hydrops and Meniere's Disease.

Learning Objective: The learner should have a better appreciation for the Phex murine mouse model of endolymphatic hydrops and the characteristics of X-linked hypophosphatemic rickets.

Desired Result: Attendees can apply this knowledge to better serve their patient population with Meniere's Disease via further insight into the disease etiology, relationship with bone disorders, and potential therapeutic options.

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Apogeotropic Positional Nystagmus in Horizontal Semicircular Canal Vertigo: Case Series and Review

*Kimanh Nguyen, MD; Sameer Ahmed, MD
Akira Ishiyama, MD; Robert W. Baloh, MD
Gail Ishiyama, MD*

Objective: The horizontal variant of benign positional vertigo which presents with apogeotropic nystagmus can be difficult to diagnose and treat. Horizontal semicircular canal (HSC) cupulolithiasis or canalolithiasis is believed to be associated with apogeotropic nystagmus; the exact location of the particles relative to the cupula of the horizontal canal is unknown. The objective of this study is to describe the detailed clinical presentation of the HSC apogeotropic variant and the maneuvers to treat this often misdiagnosed entity.

Data Source: Case series from patients who underwent vestibular testing who demonstrated the HSC apogeotropic variant. This study was exempt from IRB approval.

Study Selection: Patients with apogeotropic positional nystagmus examined at a tertiary medical center were compared to reported cases in the literature.

Data Extraction: Information about patient's gender, age, clinical history, physical examination, and quantitative vestibular testing were extracted.

Data Synthesis: Relevant clinical data were analyzed and compared to data from studies in the literature.

Conclusion: Although less commonly seen, the apogeotropic variant of HSC vertigo is a distinct clinical entity that requires further understanding. Cupulolithiasis of the HSC is implicated as the underlying mechanism. We hypothesize that the apogeotropic nature of the nystagmus requires that particles are on the ampullary side of the cupula, and thus, further away from the utricle.

Define Professional Practice Gap & Educational Need: The mechanism of apogeotropic nystagmus in HSC variants is not fully understood.

Learning Objective: To better understand the mechanism of apogeotropic nystagmus in HSC variants of vertigo.

Desired Result: To recognize and treat these clinical variants of HSC vertigo more effectively.

IRB Approved

**Totally Transcanal Endoscopic Retrograde
Mastoidectomy on Demand using
Electrically Powered Instruments**

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Toshinori Kubota, MD, PhD; Kazunori Futai, MD, PhD*

Background: Transcanal endoscopic ear surgery (TEES) provides fewer anatomical dead corners and is becoming a viable alternative to a conventional microscopic approach. However, its safe use in the antrum is a challenge that still needs to be addressed.

Objective: We examined the feasibility of transcanal endoscopic retrograde mastoidectomy on demand (RMOD) to access pathologies in the attic and antrum using electrically powered instruments, including the ultrasonic bone curette (UBC) and a high-speed curved burr in cholesteatoma surgery.

Study design: A prospective case series

Setting: Tertiary referral center

Patients: TEES was performed on 58 patients with primary cholesteatoma between September 2011 and September 2013.

Intervention: Transcanal endoscopic RMOD was performed using a Sonopet UBC (Stryker) and a high-speed curved burr (Medtronic). These instruments are designed to help protect critical anatomy from mechanical injury.

Results: Transcanal endoscopic RMOD was performed on 47 cases, including 28 cases with atticotomy and 19 cases with minimum atticotomy, using the electrically powered instruments. TEES made it possible to remove only that bony tissue necessary to visualize the pathology and with absolutely no collateral damage to surrounding tissue seen with standard drills. The cholesteatoma was completely removed from the attic and antral mucosa under clear endoscopic visualization. This procedure resulted in greater mastoid preservation than is possible with conventional microscopic approach.

Conclusions: The transcanal endoscopic RMOD approach to the antrum using the powered instruments proved to be less invasive and more functional. This powered TEES has opened the door to totally TEES procedures in restricted surgical spaces.

Define Professional Practice Gap & Educational Need: Lack of awareness that totally transcanal endoscopic ear surgery is used safely in the antrum .

Learning Objective: To learn that the transcanal endoscopic retrograde mastoidectomy on demand approach to the antrum using the powered instruments proved to be less invasive and more functional.

Desired Result: To understand that this powered TEES has opened the door to totally TEES procedures in restricted surgical spaces.

IRB Approved

Quality Improvement in Delivery of Care for Vestibular Disorders

*David R. Friedland MD, PhD; Christy Erbe, BS
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Michael Stadler, MD*

Objective: To improve the quality of health care delivery to patients with vestibular disorders.

Study design: This QA project assessed the ability to efficiently and effectively triage vestibular patients using a secure application (REDCap) normally employed to support data analysis for research studies.

Setting: Tertiary referral center.

Patients: New patients with chief complaint of dizziness.

Intervention: Answers from an intake vestibular disorders questionnaire (VDQ) were input into REDCap.

Main outcome measure: We assessed the utility of transposing clinical data into a research database and the effect on patient care.

Results: In the past 12 months 307 new VDQs were returned. This 10-page questionnaire was readily recreated in REDCap providing for real-time data analyses include profiles of demographics, main complaints, headache profile, hearing status profile, and medical history. As an example of real-time program adjustment, after 117 records were reviewed we identified 62.7% of patients with complaints of dizziness with standing-up and 54.5% of patients with dizziness when lying or turning in bed. Our vestibular therapy evaluation protocol now consists of both Dix-Hallpike maneuver and orthostatic assessment. Other adjustments included new education material on vestibular migraine as 38.4% of new vestibular patients associated headache with their dizziness. Logistic regression and classification and regression trees (CART) are also being employed to refine a clinical decision making algorithm.

Conclusions: Use of technology to track patient clinical profiles in real-time will allow for greater provider efficiency and increase value in healthcare delivery for vestibular disorders.

Define Professional Practice Gap & Educational Need:

- 1) Inconsistencies in care for vestibular patients
- 2) Lack of evidence for care plans

Learning Objective:

To provide an evidence-based clinical pathway algorithm for care of vestibular patients

Desired Result: Improved practice management for vestibular patients

IRB Exempt

IRB approval is not needed at our institution for quality initiatives and their presentations

**Outcomes of Cochlear Implantation in Patients
80 years and Older**

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Eric R. Oliver, MD*

Study design: Retrospective case review of 21 ears that underwent CI.

Setting: Tertiary referral center.

Patients: 20 patients who had 21 ears implanted at or older than 80 years of age.

Intervention(s): CI.

Main outcome measure(s): Pre-operative frequency-specific, aided testing compared to post-operative frequency specific testing. AZ Bio and Hearing In Noise Testing (HINT) scores pre implantation compared to post-implantation. Surgical complications. Self-reported vestibular symptoms.

Results: All ears implanted experienced an improvement in pure tone thresholds, with an average improvement of 65 dB ($p < 0.0001$). Post-implant sentence testing (AZ Bio and HINT) scores were significantly improved over pre-implant sentence testing (AZ Bio and HINT) scores ($p < 0.0001$). All but 2 patients tested experienced post-implant sentence comprehension $> 53\%$.

There was only one patient who suffered device failure, and one patient who experienced a wound complication. Of those with vestibular data, there is no change in subjective vestibular complaints after CI.

Conclusions: Elderly patients experience a similar improvement in audiometric measures after CI, relative to younger patients. There is minimal morbidity associated with undergoing CI, even in older patients with significant co-morbidities. Elderly patients who undergo CI do not seem to experience worsening of perceived vestibular deficits.

Define Professional Practice Gap & Educational Need:

1. Lack of data demonstrating outcomes in cochlear implantation in the extreme elderly patient population

Learning Objective: Identify audiometric outcomes and complications of cochlear implantation (CI) in elderly patients. Identify any changes in subjective vestibular symptoms from pre- to post-CI.

Desired Result: Consider cochlear implantation in elderly patients even with co-morbidities since risks are minimal and outcomes are comparable to other age groups.

IRB Approved

**Temporomandibular Joint Osteomyelitis
Associated with Otitis Externa**

*Candace E. Hobson, MD; Andrew A. McCall, MD
Barry E. Hirsch, MD; Yael Raz, MD*

Objective: The goal of this study is to report our experience with the management of temporomandibular joint (TMJ) osteomyelitis with associated otitis externa.

Methods: Retrospective case review of all cases of TMJ osteomyelitis with associated otitis externa between 2006 and 2013 at a tertiary care center.

Results: Nine cases were identified, but only 7 had sufficient data for analysis. The average patient age was 59 years old. Six patients had malignant otitis externa with extension into the TMJ, and one patient developed an infection of a TMJ prosthesis, with extension of the infection into the external auditory canal (EAC). Six of the seven (85%) patients were treated surgically after failed medical management. One patient, who failed conservative management with antibiotics and hyperbaric oxygen, eventually underwent canal-wall-down mastoidectomy and partial mandibulectomy with temporalis flap. The patient with the infection originating from an infected TMJ prosthesis underwent removal of hardware, excision of EAC granulation tissue, with skin graft coverage of the EAC defect and later total joint reconstruction with a prosthetic joint.

Conclusions: Osteomyelitis of the TMJ with associated otitis externa is a rare infection, which may originate in either the ear canal or the TMJ. All but one of the patients in our series had TMJ osteomyelitis as a result of MOE, and 85% patients were treated surgically. This illustrates that while MOE is a disease that is typically managed medically, extension of this disease into the TMJ often warrants surgical intervention.

Define Professional Practice Gap & Educational Need: TMJ osteomyelitis that is associated with otitis externa is a rare clinical entity of which many otolaryngologists lack awareness. The management of TMJ osteomyelitis associated with otitis externa is inconsistent among practitioners.

Learning Objective:

To increase awareness of TMJ osteomyelitis associated with otitis externa.

To provide recommendations for the management of this disease.

Desired Result: Attendees will have a better understanding of the presentation and appropriate management of TMJ osteomyelitis associated with otitis externa.

IRB Approved

**Canal Wall Reconstruction Tympanomastoidectomy
with Mastoid Obliteration for the Treatment
of Cholesteatoma in Children**

*P. Cody Buchanan, DO; Justin S. Golub, MD
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Objective: To determine the outcomes of canal wall reconstruction tympanomastoidectomy with mastoid obliteration (CWR/MO) for cholesteatoma in the pediatric population.

Setting: Tertiary pediatric referral center

Patients: Twelve patients ranging in age from 5-17 years old (mean 11.8 years) with cholesteatoma who were treated between 2006 and 2013. Six of these patients had surgery for cholesteatoma elsewhere prior to presentation.

Interventions: All patients underwent CWR/MO. Patients were re-evaluated with a second look tympanoplasty 6-12 months later. Long-term follow up ranged from 11-83 months (mean 42 months).

Main Outcome Measures: Rate of recurrent cholesteatoma and the need for future canal wall down tympanomastoidectomy (CWD).

Results: Five of 12 patients had residual or recurrent disease during the second look. One patient had a retraction pocket, one had 2 epithelial pearls on the stapes, and 3 had frank cholesteatoma. Of these 5 patients, one underwent CWD and one underwent an endaural atticotomy. During the subsequent long-term follow up period, one out of the original 12 patients had recurrent cholesteatoma and underwent revision canal wall up tympanomastoidectomy.

Conclusions: CWR/MO is an alternative to CWD without creation of a mastoid bowl. A second look is recommended to eliminate any residual or recurrent disease. In the pediatric population, this technique resulted in a recurrence rate of 8%, comparable to the CWD technique.

Define Professional Practice Gap & Educational Need: Cholesteatomas in children are typically treated with either a canal wall down or canal wall intact tympanomastoidectomy. There is a general lack of awareness among clinicians regarding alternative procedures, such as the canal wall reconstruction tympanomastoidectomy with mastoid obliteration technique.

Learning Objective:

1. To understand the canal wall reconstruction tympanomastoidectomy with mastoid obliteration technique.
2. To be able to apply knowledge of this technique to pediatric patients within clinical practice.
3. To be able to predict successful outcomes using this technique.
4. To know how to manage and prevent common complications associated with this technique.

Desired Result: The physician will be familiar with the canal wall reconstruction tympanomastoidectomy with mastoid obliteration technique and will be able to utilize it in clinical practice.

IRB Approved

**Curved Adjustable Fiberoptic Laser
for Endoscopic Cholesteatoma Surgery**

*Amy Y. Yau, MD; Hossein Mahboubi, MD, MPH
Hamid R. Djalilian, MD*

Hypothesis: Endoscopic cholesteatoma removal can be performed efficiently and safely using a curved and adjustable fiberoptic based laser.

Background: The endoscope allows for visualization around corners in the middle ear and mastoid. Angled instruments are required in endoscopic ear surgery to access recesses of the middle ear without extra drilling. Lasers are effective at ablating visible and microscopic cholesteatoma matrix and removing granulation tissue. We present the novel use of a curved, adjustable fiberoptic-based laser carrier for endoscopic cholesteatoma surgery.

Methods: The Iridex Adjustable and Intuitive Endo Ocular Probe, which is original designed for retina surgery, was used in 6 cases of endoscopic-assisted cholesteatoma surgery. This probe is fiberoptic-based, is available in multiple gauges, and can be used with multiple laser systems. The tip of the laser through the handle has a 45 degree curve and the length of the tip is adjustable. After cholesteatoma was removed with curved suctions and curved blunt hooks, residual cholesteatoma was identified by the endoscope. Due to the proximity of the cholesteatoma to the sinus tympani, stapes suprastructure, oval window, attic, or Eustachian tube orifice, an Argon laser with the 23 gauge Endo Ocular Probe was used to ablate the cholesteatoma in a precise and contact-free manner.

Results: In these 6 cases, the cholesteatoma was removed completely with no injury to surrounding structures and with no evidence of residual cholesteatoma with a follow up period of up to 18 months.

Conclusions: The curved laser probe allows for precise removal of cholesteatoma endoscopically.

Define Professional Practice Gap & Educational Need: Lack of adequate surgical instruments in endoscopic ear surgery

Learning Objective: To show that endoscopic cholesteatoma removal can be performed safely using a curved and adjustable fiberoptic based laser.

Desired Result: Use of a curved fiberoptic based laser will assist in the removal of cholesteatoma, granulation tissue, and adhesions during endoscopic ear surgery.

IRB Approved

**New Possibilities in Middle Ear Implants with
Direct Acoustic Cochlea Stimulation**

*Henryk Skarzynski, Prof, MD, PhD; Lukasz Olszewski, MSc
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Profound mixed hearing losses are a great challenge in modern audiology and otosurgery. Available treatment options do not provide satisfying results in many cases. Both, surgical and non-invasive approaches are inefficient or impossible to apply in many cases. A new partially implantable direct acoustic cochlear stimulator may be a prospective alternative to current treatment solutions.

The aim of the study is to present audiological results of the patients with profound mixed hearing loss using the direct acoustic cochlear system after 18 months post surgery.

5 patients (aged between 43 and 71 y.o.) were implanted unilaterally with a direct acoustic cochlear stimulator in 2012. Surgeries were conducted as part of a multicenter clinical study. Audiological assessment included impedance audiometry and pure tone audiometry (under headphones and in free field). Monosyllabic speech tests in noise and in quiet and the adaptive sentence test in noise were applied as well. Additionally to the multicenter study we extend test with results of acoustical loudness scaling in free field, using narrowband noise and 50-points loudness scale measured in the device only condition are presented. The frequencies measured ranged from 500Hz to 6300Hz and intensity levels ranged from 40 to 90 dB SPL.

Hearing was preserved in 100% of cases. 4 out of 5 patients showed a statistically significant improvement of speech recognition scores with the device in quiet and the speech reception threshold (SRT) in noise was significantly better in comparison to preoperative results with conventional hearing aids. The authors also observed better free field thresholds with the device compared to preoperative free field thresholds tested with hearing aids.

Obtained results allow to state that the new implantable direct acoustic cochlear stimulator system may be an alternative to other treatment methods applied in cases of profound mixed hearing losses.

Define Professional Practice Gap & Educational Need: lack of knowledge on direct acoustic cochlear stimulator - audiological results

Learning Objective: Participants will gain knowledge on the audiological results of patients implanted with direct acoustic cochlear stimulator.

Desired Result: Information about possible use of new device.

IRB Approved

**Anterior-posterior Diameter of the Incus
in Stapes Surgery**

Norman Wendell Todd Jr., MD

Hypothesis: In clinically normal incudes, the anterior-posterior diameter of the incus at the crimping site has a wide range, is bilaterally symmetrical, and correlates with the small mastoid indicator of childhood otitis media.

Background: Stabilization of stapes prosthesis where it is crimped onto the incus, is a continuing problem in stapes surgery. If too loose, there is a conductive hearing loss; if too tight, the long process of the incus may erode. Anecdotally there is great variability of incus bulk at the crimping site, though rigorous study non-fixed non-decalcified clinically normal incudes could not be found. Erosive loss of the long process of the incus is the most common ossicle problem of patients suffering chronic otitis.

Methods: From 41 cadavers with clinically normal ears, incudes were digitally photographed, with the incudes oriented lateral-up. The anterior-posterior diameter of each long process was measured at 1.4mm from the tip of the incus, where prostheses are typically attached. Mastoid pneumatization size was quantified on radiographs.

Results: Incus anterior-posterior diameters were similar right and left, medians 0.76 and 0.74 mm respectively, ranges 0.55 to 1.10 and 0.55 to 1.00 respectively. Though right-left symmetry of diameters was found ($r=.64$, 95% CI .35 to .82), no correlation with mastoid size was found.

Conclusions: The loop of a stapes prosthesis should accommodate incudes with a two-fold range of anterior-posterior diameters.

Define Professional Practice Gap & Educational Need: Lack of awareness of the range of anterior-posterior diameters of the incus where a stapes prosthesis is mounted.

Learning Objective: Know that the range of anterior-posterior diameters of the incus, where a stapes prosthesis is stabilized, has a wide range, 0.55 to 1.10mm.

Desired Result: Better results of stapes prosthesis stabilization to the long process of the incus.

IRB Exempt

Eustachian Tube Point: A Novel Clinical Gesture in Otolaryngology

Elliot D. Lieberman, MD; Miriam I. Redleaf, MD

Objective: While eustachian tube dysfunction (ETD) remains widely prevalent in both children and some adults, not all patients present with a clinical history of infections. Instead, the physical examination and tympanogram may be normal. The sole indicator of ETD may be the patient's spontaneous gesture to the location directly below the auricle, at the apex of the jugulodigastric region, over the styloid process as the focus of otalgia. We postulated that this precise location, the "eustachian tube point," signifies ETD.

Methods: Twenty four patients at a tertiary care otolaryngology clinic spontaneously indicated otalgia that localized to the "eustachian tube point." Physical exam and tympanograms were normal. In 6 patients, nasal endoscopy with gentle palpation of the eustachian tube orifice was performed and in 5 (83%) this maneuver reproduced their pain.

Results: All 24 patients were offered either nasal steroid spray or trial myringotomy followed by tympanostomy tube. Four declined intervention. Four selected nasal steroids and all (100%) patients experienced less otalgia. Sixteen patients elected myringotomy trial of whom 14 (88%) experienced resolution of otalgia. Among all 24 "eustachian tube point" patients pursuing one of the treatment options, 18/20 (90%) achieved otalgia improvement.

Conclusion: Our report is the first study to identify the "eustachian tube point," which suggests a novel referred pain pathway for ETD. This study also introduces a new otolaryngologic gesture in physical examination that will aid in the diagnostic evaluation and therapeutic management of patients with eustachian tube point and normal examinations.

Define Professional Practice Gap & Educational Need:

1. Vast differential diagnosis for otalgia
2. Multitude of vague complaints that support eustachian tube dysfunction
3. Paucity of clinical signs to distinguish eustachian tube dysfunction from other causes of otalgia

Learning Objective:

1. Recognize the otolaryngologic clinical gesture, the "eustachian tube point"
2. Implement new methodologies and practices for eustachian tube dysfunction

Desired Result: Otolgia is a pervasive complaint in the otolaryngology clinic. Knowledge of the "eustachian tube point" will help physicians in the diagnostic evaluation of otalgia and will help patients in the therapeutic management of eustachian tube dysfunction. Highly successful treatment options will be shared by the presenter with the attendees.

IRB Approved

**Paraneoplastic Syndrome: A Masquerade of
Autoimmune Inner Ear Disease**

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Jeffrey P. Harris MD, PhD; Akihiro J. Matsuoka, MD, PhD*

Rare and diagnostically challenging, paraneoplastic syndrome can appear months to years prior to detection of the underlying neoplasm and is associated with rapidly progressive neurologic deficits including cochleovestibulopathy and death.

Less than 20 cases of paraneoplastic cochleovestibulopathy have been reported in the online database Pubmed.

Three recent cases of paraneoplastic cochleovestibulopathy are presented: 1) a 42 year old man with a history of B-cell follicular lymphoma who developed dermatomyositis and hearing loss prior to detection of lymphoma recurrence in his anterior chest wall, 2) a 42 year old woman with sudden asymmetrical hearing loss refractory to steroids that was ultimately found to have a 12 cm renal mass prior to her death, and 3) a 40 year old woman with fluctuating bilateral hearing loss that was ultimately found to have a thymoma.

Although characterized as type VI (non-immune rapidly progressive sensorineural hearing loss) within the Harris autoimmune inner ear disease classification system, the mechanism of paraneoplastic cochleovestibulopathy is not well understood. While specific anti-neuronal antibodies such as anti-Hu may be associated with other paraneoplastic neurologic disorders, these antibodies have limited diagnostic utility with paraneoplastic cochleovestibulopathy. Steroids have limited efficacy with regards to hearing recovery, while intravenous immunoglobulin (IVIG) has been shown to be of benefit.

These recent cases and prior literature are presented with the goal of further understanding the clinical presentation of paraneoplastic cochleovestibulopathy and the need for screening and detection of the underlying malignancy given the associated risk of rapid decline and mortality.

Define Professional Practice Gap & Educational Need: lack of awareness about paraneoplastic syndrome cochleovestibulopathy

Learning Objective: Present rare cases of paraneoplastic syndrome cochleovestibulopathy

Desired Result: Increased awareness about paraneoplastic syndrome cochleovestibulopathy

IRB Exempt

Systematic Review of Cochlear Implantation's Impact on Cognition in the Elderly

*Gina S. Miller, BS; Craig Miller, BS
Carol L. Howe, MD, MLS; Mindy Fain, MD
Abraham Jacob, MD*

Objective: Hearing loss (HL) is the 3rd most prevalent chronic condition faced by the elderly population and has been linked to difficulties in speech perception, activities of daily living, and social interaction. Recent studies have suggested a correlation between duration/severity of hearing loss and an individual's cognitive function; however, it was unclear whether a causative link has been established. Because prolonged auditory deprivation is common prior to cochlear implantation, we performed a systematic review to determine whether cochlear implantation's potential influence on cognition in the elderly population has been studied.

Data sources: 3,886 articles related to cochlear implants, cognition, and older adults were reviewed.

Study selection: Inclusion criteria were as follows (1) elderly patients >65, (2) intervention with cochlear implantation, and (3) cognition as the primary outcome measure of implantation.

Data extraction: Systematic review. Out of 3,886 studies selected, 3 met inclusion criteria.

Data synthesis: No statistical techniques required.

Conclusions: While many publications have shown that cochlear implants improve speech perception, social functioning, and overall quality of life, we found no studies in the English literature that have prospectively evaluated changes in cognitive function after cochlear implantation in the elderly. Because prolonged auditory deprivation is common in older adults before they receive a cochlear implant, investigating whether cochlear implantation improves cognition may help to establish a causative link between hearing and cognitive function.

Define Professional Practice Gap & Educational Need:

1. Lack of evidence of cochlear implantation's effect on cognition in the elderly.

Learning Objective:

1. Understanding the many psychological advantages of cochlear implantation in the elderly.
2. Recognizing that hearing loss in the elderly is correlated with cognitive decline.
3. Understanding the clinical need for evidence as to whether cochlear implantation in the elderly has any effect on cognition; either stopping or reversing decline.

Desired Result: Attendees will be aware that hearing loss in the elderly can lead to many psychological issues as well as cognitive decline. Cochlear implantation has been shown to improve many psychological aspects of hearing loss, but it is unclear as to how it affects cognition. Attendees will recognize the need for evidence of cognitive benefit in cochlear implantation and then apply this evidence when deciding whether cochlear implantation benefits will outweigh the costs in the elderly.

IRB Exempt

**Short-Term Clinical Performance of the
Direct-Drive Hearing Device: A Pilot Study**

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Yaser Ghavami, MD; Amy Y. Yau, MD
Mark Bachman, PhD; Hamid R. Djalilian, MD*

Objectives: This is our first report of testing the Direct-Drive Hearing Device (DHD), a novel completely-in-the-canal hearing aid prototype, in a subject. Our objectives were to determine the voltages required to match desired loudness levels at different frequencies, and potential short-term side effects of device placement.

Setting: Tertiary referral center

Patients: A healthy adult was recruited.

Intervention(s): The DHD was inserted into the canal and placed in touch with different portions of the tympanic membrane and malleus.

Main Outcome Measures: Sounds at 0.5, 1, 2, 4, and 8 kHz were presented to the subject through the device. Using an audiometer, sounds with the same frequencies were presented to the left ear at different loudness levels and the subject matched the loudness between the ears. Music perception was compared between the two ears. Once data was collected, the device was removed and the tympanic membrane (TM) and ear canal were inspected for potential side effects.

Results: The subject tolerated the device well and reported that he was able to hear the sounds through the direct-drive mechanism and that the device had a high fidelity for all frequencies tested. Voltage levels between 27 and 250 mV were able to produce sounds equivalent to 40 to 80 dB HL across various frequencies. No desquamation, bleeding, perforation, or infection occurred.

Conclusions: DHD is capable of enabling a subject to hear with relatively low voltage levels. No significant side effects were noted. Long-term clinical outcomes of the device remain to be studied in the future.

Define Professional Practice Gap & Educational Need: Inconsistencies within approaches for selection of different hearing aids and their advantages and disadvantages.

Learning Objective: To better learn the differences between air conduction and implantable hearing aids, and the possibility of combining the advantages of both into one device.

Desired Result: Develop a better understanding of the novel approach to design a completely-in-the-canal hearing aid.

IRB Approved

Cochlear Implantation Requiring a Retrofacial Approach to the Round Window

*Kyle P. Allen, MD, MPH; Brandon Isaacson, MD
Loren J. Bartels, MD*

Objective: To describe two cases of cochlear implantation requiring a retrofacial approach to the round window due to an anteriorly positioned vertical facial nerve.

Study Design: Retrospective case review

Patients: Two patients with sensorineural hearing loss undergoing cochlear implantation.

Interventions: Cochlear implantation via a retrofacial approach to the round window.

Results: During cochlear implantation for sensorineural hearing loss, two patients were found to have inadequate visualization of the round window via a standard posterior tympanotomy approach. A retrofacial approach was performed to provide exposure of the round window to ensure correct placement of the electrode array. Pre- and postoperative imaging and intraoperative photographs are presented to better demonstrate the need for, and performance of, the retrofacial approach.

Conclusions: When an anteriorly positioned facial nerve obscures visualization of the round window, a retrofacial approach can be utilized to visualize the round window.

Define Professional Practice Gap & Educational Need: Lack of awareness of atypical approaches to the round window in patients with anomalous positioning of the facial nerve.

Learning Objective: Demonstrate the retrofacial approach to the round window when a standard posterior tympanotomy does not provide adequate visualization.

Desired Result: Attendees will be able to apply the retrofacial approach when faced with an anteriorly positioned facial nerve.

IRB Exempt

Two Types of Endoscopic Approach for Congenital Middle Ear Cholesteatoma

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Objective: Congenital middle ear cholesteatomas are characteristically located in the anterosuperior quadrant (ASQ) of the middle ear. When this cholesteatoma is removed via a microscopic approach, the attachment of the cholesteatoma is hidden because of the presence of the malleus. To solve this problem, we performed the transtympanic approach or the transcanal approach with endoscopic ear surgery (EES). The advantages of these approaches are examined.

Study design: A prospective case series.

Setting: Tertiary referral center

Patients: We performed either approaches in 6 patients.

Intervention: The transtympanic procedure: Tympanic membrane fenestration was performed at the ASQ with a CO2 laser (diameter: 2 mm). The cholesteatoma was carefully elevated from its attachment, and removed through the tympanic membrane fenestra. The fenestra was then covered with artificial dermis and a Chitin wound dressing.

The Transcanal Procedure: A tympanomeatal flap was elevated from the superior region of the external auditory canal (EAC). The tympanic membrane was elevated from the malleus handle. The cholesteatoma was carefully freed from its attachment, and then removed. The tympanomeatal flap was repositioned. The EAC was packed with gelatin sponge.

Result: When a cholesteatoma located only in the ASQ (Potosic stage I), a transtympanic procedure was selected. For a cholesteatomas (Potosic stage II) extended to other quadrant(s) or recurrent cholesteatoma, a transcanal procedure was performed. The cholesteatomas were successfully removed from the attachment under clear endoscopic visualization of the entire cholesteatoma.

Conclusion: The cholesteatoma of stage I and II can be successfully removed under clear endoscopic visualization.

Define Professional Practice Gap & Educational Need: Congenital middle ear cholesteatomas are characteristically located in the anterosuperior quadrant (ASQ) of the middle ear. When this cholesteatoma is removed via a microscopic approach, the attachment of the cholesteatoma is hidden because of the presence of the malleus.

Learning Objective: To use two type of endoscopic approach, we would like to remove a cholesteatoma at the ASQ under clear endoscopic visualization.

Desired Result: The cholesteatoma at ASQ can be successfully removed under clear endoscopic visualization.

IRB Approved

Radiologic Quandary: Giant Cell Tumor of the Temporal Bone Masquerading as Osteomyelitis with Abscess

*Rachel B. Cain, MD; David M. Barrs, MD
Peter A. Weisskopf, MD*

Objectives: Present a diagnostically challenging case of temporal bone destruction. Highlight unusual radiographic features of a temporal bone giant cell tumor.

Setting: Academic tertiary referral center

Patients: A 39-year-old man presented with a 2-year history of temporomandibular joint clicking and a 1-month history of temporal swelling, serous otitis media, and a mild asymmetric conductive hearing loss. CT showed bony destruction superior to the external auditory canal, mastoid, infratemporal fossa, and zygomatic process. A 3.4 cm rim-enhancing fluid collection in the mastoid raised the concern of osteomyelitis with abscess formation. MRI showed a rim-enhancing fluid collection abutting dura with mild dural enhancement.

Interventions: A mastoidectomy was recommended for diagnosis. Intraoperatively, cortical destruction was seen after opening a postauricular incision. The mastoid cortex was soft, rubbery, and easily curetted. Biopsy of bone and a cyst wall revealed giant cell tumor of bone.

Main outcome measures: The varying clinical and radiographic features, treatment options and surgical approaches for tumors of this type will be presented.

Results: Giant cell tumor of the temporal bone is rarely encountered. These generally benign tumors have the potential for locally aggressive behavior and malignant degeneration. Gross total resection is the first line of treatment. However, this is not always possible, as evidenced by the extensive skull base involvement in this case. The benefit of postoperative radiotherapy following subtotal resection is controversial.

Conclusions: Tumors of the temporal bone have variable clinical and radiographic presentations. The rarely encountered giant cell tumor of temporal bone may masquerade as infection.

Define Professional Practice Gap & Educational Need:

1. Lack of widespread clinical familiarity with the radiographic variability of temporal bone tumors.
2. Lack of awareness regarding treatment modalities and efficacy of temporal bone giant cell tumor.

Learning Objective: Present a diagnostically challenging case of temporal bone destruction. Highlight unusual radiographic features of a temporal bone giant cell tumor.

Desired Result: Attendees will have a greater appreciation of presenting features and imaging characteristics of temporal bone tumors, as well as increased knowledge regarding the rarely encountered temporal bone giant cell tumor.

IRB Exempt

Biofilm as a Cause of Cochlear Implant Extrusion

*Christopher J. Linstrom, MD; Prayag Patel
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Objective: To review the identification and treatment of cochlear implants affected by biofilm formation.

Study design: Retrospective review of cases involving suspected or proved biofilms.

Setting: Tertiary referral center.

Patients: Individuals who received cochlear implants, healed and without apparent cause had delayed infection and extrusion of the device.

Intervention(s): Intravenous antibiotics, explantation, prophylaxis against skin flora from eczematoid dermatitis.

Main outcome measure(s): Healing, sound perception and successful use of a cochlear implant.

Abstract: Several cases of successful cochlear implantation followed by delayed infection and extrusion pointing to biofilm formation will be presented. One case is illustrative: A 49M with progressive hearing loss L>R was evaluated for and received a LEFT cochlear implant. He healed uneventfully and rehabilitation commenced 4 weeks after his surgery. All was well until 3 months post-op when the wound started to break down. He received 12 weeks of intravenous antibiotics, but the functioning implant remained infected, became exposed and was removed. He underwent a second LEFT implant about 3 months later that healed. He decided to have the RIGHT ear implanted about 1.5 years later and the same event occurred: initial healing followed by wound break down and extrusion/removal at 3 months. At this time, it was apparent that he had severe eczematoid dermatitis which proved to be the cause of his infection. For the revision on the RIGHT ear, the patient had a very short haircut pre-operatively, used Hexachlorophene (PhisoHex®) shampoo daily for 2 weeks prior to implantation and had a successful, healed implant of a Cochlear Americas N-5 device that failed, had to be removed, but using the above prophylaxis, ultimately healed. He uses bilateral cochlear implants.

Other cases of delayed infection after normal healing will be summarized which illustrate the need to assure that the skin surrounding the implant field must be carefully inspected pre-operatively and that perfect sterile technique must be assured to help avoid biofilm formation, delayed infection and extrusion.

Results: N/A in this small series.

Conclusions: Biofilms may form on large foreign bodies such as cochlear implants and may become the cause for delayed infection and extrusion. This small study investigates possible causes for and reasonable means to prevent biofilm formation.

Define Professional Practice Gap & Educational Need:

1. Lack of awareness of biofilm formation as a cause of cochlear implant infection and extrusion.
2. Lack of awareness of conditions that predispose to biofilm formation.
3. Lack of knowledge of common methods of prevention of biofilm formation.

Learning Objective:

1. Knowledge of the entity and pathophysiology of biofilm formation.
2. Knowledge of the common causes of biofilm formation and how biofilms can affect cochlear implants.
3. Knowledge of means of prevention of biofilms.

Desired Result:

1. Improved wound healing and function of cochlear implants.
2. Prevention of biofilm formation in cochlear implants.
3. Pre-operative recognition of patients at risk for biofilm formation.

IRB Exemption

**Facial Nerve Paresis as Presentation of
Recurrent Non-Hodgkins Lymphoma**

*Mark A. Fritz, MD; Paul E. Hammerschlag, MD
Alec S. Goldenberg, MD*

Objectives: Present a patient with facial paresis and magnetic resonance imaging (MRI) findings consistent with brain abscess found to be due to recurrent non-Hodgkins lymphoma. Literature describing lymphoma in the temporal bone is reviewed.

Methods: Case report from tertiary care center.

A 71-year-old female presented with right aural fullness, otalgia, and conductive hearing loss. Her symptoms resolved after one week of Ciprodex, Prednisone, and Ciprofloxin. One month later, she developed a right facial paresis. Two retroauricular –occipital skin nodules were noted and thought to be cutaneous lymphoma to be treated with chemotherapy. Computed tomography (CT) demonstrated mastoid and middle ear opacification, and her MRI showed a 1.0cm smooth, well-defined nodular enhancing lesion in the right posterior temporal lobe consistent with brain abscess of otogenic origin. Below the right mastoid was a 3cm tender mass concerning for a possible Bezold's abscess.

Results: The patient underwent an urgent right tympanomastoidectomy. The mastoid intraoperatively contained pale fibrinous granulation tissue, consistent with ongoing response to preoperative antibiotic treatment and prednisone. Postoperative pathology revealed non-Hodgkins lymphoma, follicular center cell, consistent with the previous lymphoma and the culture was negative. Facial nerve paresis completely resolved within one month after mastoidectomy. Positron emission tomography (PET) imaging of the temporal lobe lesion demonstrated it to be consistent with lymphoma metastasis. Proton beam therapy eradicated this nodular enhancing lesion in the temporal lobe.

Conclusion: Non-Hodgkins lymphoma metastasis should be considered in the evaluation of middle ear and mastoid metastasis opacification associated with facial nerve paresis and concurrent possible otogenic brain abscess.

Define Professional Practice Gap & Educational Need:

1. Lack of awareness
2. Lack of description in the literature

Learning Objective: To demonstrate the occurrence of non-hodgkin's lymphoma and the multiple different presentations that it can emulate and to describe the prior literature of non-hodgkin's lymphoma in the temporal bone.

Desired Result: Attendees should be able to include non-Hodgkin's lymphoma in their differential diagnosis when a similar patient presents to their practice.

IRB Exempt

**Useful Cases of Multiple Cholesteatoma
by non-EPI DW-MRI and MRC Images**

*Takatoshi Furukawa, MD, PhD; Tomoo Watanabe, MD, PhD
Tsukasa Ito, MD, PhD; Yasuhiro Abe, MD, PhD
Toshinori Kubota, MD, PhD; Kazunori Futai, MD, PhD
Seiji Kakehata, MD, PhD*

Objective: When transcanal endoscopic ear surgery (TEES) is performed to treat cholesteatoma, it is particularly important to conduct pre-operative diagnostic imaging to confirm the extent of the cholesteatoma progression.

Study design: Case reports

Setting: Tertiary referral center

Patients: Two cases with multiple cholesteatoma

Intervention: Cholesteatoma surgery

Main outcome measure(s): 1-mm slice non-EPI diffusion-weighted magnetic resonance imaging (DW-MRI) as a way to preoperatively diagnose cholesteatoma, and evaluated the cholesteatoma progression by fusing these images with those of the Magnetic Resonance Cholangiopanc-reatography (MRC).

Results: Case 1: Fusion images confirmed that the mastoid antrum was a high intensity area. Surgically, we first completely extracted the epithelium that had extended from the flaccid area using TEES. And we discovered an independent sac-like cholesteatoma in the mastoid antrum, and extracted this endoscopically using attico-antrotomy. Case 2: Fusion images showed high intensity regions from the flaccid region to the mastoid antrum, as well as an independent high intensity region dorsal to this. We performed TEES to completely extract the epithelium from the flaccid region up to the mastoid antrum. We then extracted a peripheral cholesteatoma by performing a microscopic postauricular mastoidectomy.

Conclusion: In the present two cases, we went into surgery with the pre-operative fusion images of multiple cholesteatoma in mind, which allowed us to address all of them without missing any. We hereby conclude that the detailed evaluation made possible by image fusion between non-EPI DW-MRI and MRC images can serve as an effective pre-operative diagnosis for peripheral cholesteatoma.

Define Professional Practice Gap & Educational Need: Lack of awareness of pre-operative diagnostic imaging

Learning Objective: Confirmation of the extent of the cholesteatoma progression

Desired Result: Effective pre-operative diagnosis for peripheral cholesteatoma Patient Care

IRB Exempt

AOS Clinician-Scientist Award Progress Report
Project: Spectral and Temporal Resolution in Infants with Cochlear Implants
PI: David Horn, MD, MS

There are two specific aims for this study: 1) to determine the rates at which auditory spectral and temporal resolution emerge in CI infants relative to normal hearing (NH) infants, and 2) to assess the relationship between early spectral and temporal resolution and later speech and language development in CI infants. During the first 6-months of the grant cycle, work has focused primarily on collecting data for the experiments for Aim 1. In experiment 1, spectral resolution was measured by measuring the ability to discriminate a spectral shift in a broad-band noise (Spectral Ripple Discrimination, SRD). Ripples refer to periodic modulation of the noise in the spectral domain. Inversion of the peaks and troughs of the spectral modulation creates a change in spectral shape that becomes harder to discriminate as ripples are spaced closer together. The SRD threshold is the highest ripple density at which the listener can detect the peak/trough inversion. Previously, we had shown that although 7-9 month old NH infants performed more poorly than NH adults, performance of both infants and adults improved at similar rates with increasing ripple amplitude depth suggesting that spectral resolution was similar between age groups.

In experiment 1, we have hypothesized that spectral resolution will mature in both NH and CI infants between 3 and 7 months hearing age. We have collected data from 20 of the planned 40 NH infants (10 3-month olds and 10 7-month olds) and have single test-interval data from 3 CI infants. SRD thresholds at two different ripple depths were collected from each infant. All were tested using the observer-based psychoacoustic procedure (OPP) in an adaptive method to arrive at the SRD threshold. Data from 10 NH adults and 3 CI adults have also been obtained using the above methods and test procedures. Over 85% of infants and all adults could complete the protocol. For NH listeners, significant main effects of ripple depth (higher SRD at higher ripple depths) and of age (higher SRD at higher ages) were seen. Independent sample t-tests for each age-group comparison showed that 3-month old SRD thresholds were lower than adults across ripple depths but that 7-month old thresholds were lower than adults only at 10dB and adult-like at 20dB. These results suggest that spectral resolution is more mature at 7 months than 3 months of age in NH infants. Data from 3 adult long-term CI users and 4 CI infants with 3-5 months of listening experiments have been obtained. Although preliminary, these CI infants show adult-like SRD thresholds across ripple depths, suggesting similar spectral resolution as CI adults.

We have begun to collect data for experiment 2, which examines temporal skills of NH and CI infants and adults using an amplitude modulation detection task (AMD). Listeners are asked to discriminate steady broad-band noise from amplitude modulated noise at decreasing modulation depths until the threshold for discrimination is reached. AMD thresholds at a low (10Hz) and high (150 Hz) are obtained from each listener to get a sense of AM detection at two disparate points of the temporal modulation transfer function.

(CONT)

To date, we have completed testing with 7 3-month old infants and in the same 8 CI users who were tested in experiment 1. Preliminary results suggest that 3 month-old NH infants show near floor-performance at 150Hz but more adult-like performance at 10Hz, suggesting that temporal resolution is immature at 3 months of age in NH listeners. CI infants, on the other hand, seem to show adult-like performance at both modulation rates, suggesting that maturation of temporal resolution in CI infants may occur during the first 3-5 months of CI use.

Further data collection from CI infants is ongoing for both experiments 1 and 2. For the Aim 2 experiments, speech perception and language measures will be collected from CI infants after 12 months of CI use. This work will begin during the second 6-months of the grant period. Correlations between SRD and AMD thresholds and speech perception and language measures will be examined.

As part of the career development portion of the grant, the PI completed a graduate course in Auditory Development this fall. During the winter quarter, independent study of predictors of language development in typical and atypical populations will ensue under the guidance of Dr. Truman Coggins. The PI is extremely pleased to report that a 5-year K23 was recently awarded to continue this research and training plan. This funding period will begin July 1, 2014.

AOS Clinician-Scientist Award Progress Report
Project: Nanotechnological Regeneration of Spiral Ganglion
Neurons with Human Stem Cells
PI: Akihiro J .Matsuoka, MD., PhD

Many meritorious efforts are under way to develop strategies for hair cell regeneration in the hope that it will allow some early interventions for patients with severe to profound sensorineural hearing loss (SNHL). However, any patient with any degree of subacute to chronic severe to profound SNHL will be accompanied by trans-synaptic, retrograde degeneration of spiral ganglion neurons (SGNs), which could inhibit hair cell regeneration from accomplishing the ultimate goal of restoring hearing. While a very long term goal for those in this filed might involve the regeneration of both hair cells and SGNs, a more parsimonious and practical approach involves the regeneration of SGNs in conjunction with a cochlear implant (CI). Further, this approach also would likely be beneficial to the population of patients who already have a CI that has not provided hoped benefits likely due to SGN degeneration. One method to regenerate SGNs is to use human pluripotent stem cells (hPSCs), including both human embryonic stem cells (hESCs) and induced pluripotent stem cells (iPSCs), thereby re-establishing the neural circuit required for CI function. However, clinical use of PSC-based therapies has been hindered by a number of critical hurdles, including safety, dosage, long-term survival, neuronal differentiation, and functional efficacy. Recent studies have shown that the survival and growth of transplanted stem cells are mostly regulated by dynamic, bidirectional interactions with the local tissue environment in the immediate vicinity of the cell, termed the "stem cell niche", which is characterized by matrix stiffness, architecture, and neurotrophic factors. We proposed that recreating the stem cell niche is crucial for further advancement of stem cell-based therapies for the inner ear. Self-assembling peptide amphiphiles (PA) created at Northwestern University would potentially provide a unique tool to solve this issue. This material can be injected as a liquid into the inner ear where it self-assembles to form an artificial extracellular matrix. The matrix material can be engineered to display signaling sequences (i.e., BDNF and NT-3) that support SGN survival and health. By utilizing this material in conjunction with transplantation of hESC-derived otic neuronal progenitors (ONPs), we proposed to establish and validate an effective therapeutic application of hESCs to regenerate SGNs. The successful establishment of this novel technology would not only improve CI technology and potentially broaden the population of patients that can derive benefits, but also will be critical for future hair cell generation therapy since the success of both treatment modalities relies upon survival of a critical number of SGNs following deafness. The two aim of this funded study were 1) to establish a method for the controlled generation of hESC-derived SGNs using diffusible ligands and overexpression of relevant human transcription factors at developmentally relevant time points in a stepwise fashion. 2) to develop an appropriate stem cell niche for hESC-derived ONPs to efficiently differentiate them into SGNs.

With regard to the specific aim #1, we first differentiated undifferentiated hESCs to non-neuronal ectoderm (NNE) using BMP4 on a Matrigel-coated plate in serum-free medium. The hESC-derived NNE-like cells were then differentiated into preplicodal ectoderm (PPE)-like cells with a treatment of Noggin. The hESC-derived PPE-like cells expressed a transcription factor of SIX1, SIX4, and EYA1, which are the main markers of PPE. As hESC-derived PPE-like cells also expressed PAX6, which is a marker of the central neurons, the PPE-like cells were then undergone cell sorting using MACS (magnetic activated cell sorting system) to positive-sort with p75, which is a marker for peripheral neurons. After the sorting, the cells were further differentiated toward an otic lineage using neurotrophic factors, Wnt, FGF, and EGF for 14-21 days. These cells expressed FOXG1, nestin, GATA3, PAX2, and PAX8, which are markers for ONPs on immunohistochemical analyses. (CONT)

The challenge we are facing now is how to increase efficiency in generating ONPs. One way to increase the efficiency is to increase the population of peripheral neurons in the first stage when we are generating the NNE.

The second method is to increase PPE population and decrease neural crest population by using a Wnt inhibitor. A Wnt inhibitor has been mainly used in a field of cancer stem cell research and as such an effect on hESC is unknown at this point. We are planning to create a dose-response curve using quantitative PCR to identify the suitable concentration of several Wnt inhibitors to maximize generation of hESC-derived PPE-like cells.

The second part of this aim was to examine whether SGNs could also be generated through controlled expression of specific human genes. Based on murine embryology, the transcription factors PAX2 and PAX8 were chosen as Pax2/Pax8 double mutants arrest ear development at the otic vesicle stage highlighting their important role in ONP differentiation. We performed nucleofection of hESC-derived PPE-like cells with PAX2 and PAX8 using an Amaxa® Cell Line Optimization Nucleofector® Kit (Lonza, Cologne, Germany). Our preliminary result of nucleofection of hESC-derived PPE-like cells with PAX2 and PAX8 indicated that transfected hESCs were expanded and selected using Neomycin for PAX2 and Hygromycin B for PAX8, respectively. Many of Neomycin or Hygromycin B-resistant cells exhibit intense GFP-fluorescence and red-fluorescence demonstrating hESC-derived PPE-like cells stably expressing the expression constructs. We are hoping to increase efficiency of nucleofection by using different nucleofection kit.

With regard to the specific aim #2, we have successfully cultured hESC-derived ONPs in IKVAV-PA gels. This experiment indicated that cultured hESC-derived ONPs extended neurite in the IKVAV-PA gel that may be used in the inner ear to provide grafted hPSCs with a framework for cellular regeneration and differentiation. Also, preliminary data shows viable stem cells with evidence of neural differentiation at greater than two weeks using PA gels in vitro. We also used human cadaveric temporal bone to prove the technical feasibility of a transmastoid approach for intracochlear injection and inner ear stem cell delivery. In vitro live/dead cell viability assays and EdU proliferation assays were also performed in PA gels with cultured hPSCs. Two human cadaveric temporal bones were used to test the feasibility of using IKVAV-PA gels. The resultant vestibulocochlear nerve complex was harvested and sectioned for transmission electron microscopy. Cell viability assays in IKVAV-PA gels demonstrated adequate hPSC viability on days 5, 7, and 14 with minimal apoptosis. EdU cell proliferation assays on day 5 showed no proliferating cells, with nestin positivity on day 15 supporting hPSC maturation into a neuronal phenotype. In a cadaveric temporal bone study, fluorescence corresponding to both IKVAV-PA gel and TRA 1-81 (a marker for an undifferentiated hESC) stained hESCs was noted along the internal auditory canal following intracochlear injection. These preliminary results demonstrated that re-creating a “stem cell niche” by providing IKVAV-PA gels with the appropriate neurotrophins in the inner ear may promote cell survival and neuronal differentiation. This portion of work was presented on a podium at Combined Triological Society Meeting (01/10/2014, Miami Beach, FL) and one of our lab members, Zafar Sayed, M.D. won the Walter Work, MD Resident Research Award (the first prize) for his work. Additionally, we performed further immunohistochemical characterization of ONPs in the IKVAV gel. This additional experiment indicated that the nestin, FOXG1, and PAX8 were expressed in ONPs in IKVAV-PA gels, however, markers for neurons such as TUJ1 and GLUT2/4 were negative, suggesting that external cues for ONPs will be required for neuronal differentiation.

Finally, I was just notified that my application to NIH K08 award will be funded starting on 07/01/2014. I would like to thank American Otological Society for allowing me to generate sufficient preliminary data to be able to win this award from NIH.

AOS Research Grant Award Progress Report
Progress Report: Toward Pathophysiologic Standards for
Gentamicin Pharmacotherapy
Principal Investigator: Larry F. Hoffman, PhD

Intratympanic gentamicin has become a widely used treatment for Meniere's disease. Though numerous studies have attempted to establish correlates between tests of vestibular function and treatment efficacy, standards for the relationship between the extent of neuroepithelial lesion and functional output, which will critically ameliorate objective tests of vestibular function, do not exist. Such standards will enable rigorous objective evaluation of the intended vestibular paresis and the efficacy of intratympanic gentamicin treatment for eliminating the symptoms of Ménière's disease.

We are continuing our investigations to establish these pathophysiologic standards through the administration of defined quantities of gentamicin in an animal model. As reported previously, we are using doses that correspond to total gentamicin quantities of 0.5 and 0.75 μg . Our studies in the past year have been instrumental in confirming the histopathologic status of the vestibular neuroepithelia subsequent to these doses.

In our work with 1 μg total gentamicin administration we found that afferent calyces were more labile to structural compromise than hair cells. Therefore, we use calyx morphology as a sensitive index of gentamicin-induced lesions. We have found that 0.5 μg , which corresponds to approximately 65 μM in the 16 μl perilymph volume of the animal model in use, does not produce an observable lesion in either hair cells or afferent calyces. Neuroepithelia from these treated labyrinths exhibit the full complement of hair cells and calyces. However, single neuron electrophysiologic recording studies have revealed a functional deficit in the discharge characteristics of these afferents. We found that the temporal fidelity of the output of these neurons has been compromised, revealed through analyses of the coherence between stimulus input and afferent discharge output. These data indicate that functional changes can result in conditions for which the morphologic features of the epithelia appear intact.

The situation was much different with a slightly higher gentamicin dose. At a total gentamicin dose equivalent to 0.75 μg (corresponding to 98 μM), we found extensive lesions in the vestibular epithelia, where approximately 20% of hair cells were lost and virtually all calyces were lost in the central zones of the semicircular canal cristae. The utricular striola was similarly affected. Afferent discharge in these specimens exhibited similar functional changes as observed for the 0.5 μg specimens, with the decrease in stimulus/response coherence reflecting a compromise in the temporal fidelity of the afferent system. Therefore, the afferents that were characterized reflected a similar functional deficit as found for the lower doses. These data demonstrate that functional afferents can exist in epithelia exhibiting significant morphologic lesions from gentamicin administration. The more extensive morphologic lesion at this dose suggests that some afferents (namely, the calyx only afferents projecting to the crista central zones) are likely to have lost their inputs and ability to respond (i.e. without a calyx), further suggesting a far greater effect in terms of the input to the central nervous system.

(CONT)

We are in the process of completing development of the system to record the vestibulo-ocular reflex in our animal models to test how these two different lesions manifest in vestibular-related behaviors. The turntable system is in place, and we are implementing the infrared video system to record and analyze stimulus-evoked eye movements. This system will be used to test the hypothesis regarding the behavioral deficits associated with low-dose gentamicin administration conditions.

In this past year we have also conducted experiments to determine whether some key molecular components within hair cells or afferents are particularly labile to gentamicin. Such experiments could contribute to identifying the mechanism of the subcellular dysfunction that could explain our results with the low gentamicin doses. We are currently focusing on otoferlin, a protein that is critical to stimulus-evoked transmitter release (Dulon et al., 2009).

We have also conducted analyses of the degree to which gentamicin is retained by hair cells and afferent dendrites long after initial administration using the small administered quantities of gentamicin in our studies. These analyses are important in view of the persistent physiologic effects that we have observed. The retention of gentamicin has been previously reported (Imamura and Adams, 2003; Lyford-Pike et al., 2007), but not in the context of precise dose and with physiologic outcome measures. We have found that even specimens receiving the lowest dose exhibit gentamicin within calyces 2 months post-administration, presenting the problem that any intrinsic mechanisms for functional recovery may be prevented by the residual gentamicin.

The experiments enabled by support from the Research Fund of the American Otological Society were critical to our laboratory, and provided the needed pilot data in order to submit a comprehensive proposal for funding from the NIDCD. This proposal was submitted February 2014.

Dulon D, Safieddine S, Jones SM, Petit C (2009) Otoferlin is critical for a highly sensitive and linear calcium-dependent exocytosis at vestibular hair cell ribbon synapses. *J Neurosci* 29:10474-10487.

Imamura S, Adams JC (2003) Distribution of gentamicin in the guinea pig inner ear after local or systemic application. *J Assoc Res Otolaryngol* 4:176-195.

Lyford-Pike S, Vogelheim C, Chu E, Della Santina CC, Carey JP (2007) Gentamicin is primarily localized in vestibular type I hair cells after intratympanic administration. *J Assoc Res Otolaryngol* 8:497-508.

AOS Clinician-Scientist Award Progress Report

PI: Yuri Agrawal, MD

Project: Influence of Vestibular Dysfunction on Fall Risk in Older Individuals

The aims of this study were: 1) to characterize the vestibular physiologic changes associated with the aging process, 2) to determine the association between specific vestibular physiologic deficits and fall risk in older individuals, and 3) to develop a valid, reliable measure of fall risk that is sensitive to changes in vestibular function in older individuals.

With respect to Aim 1, we have entered into a highly productive collaboration with the Baltimore Longitudinal Study of Aging (BLSA), which is a longitudinal survey funded by the National Institute on Aging that has been running continuously since 1958. At present, 1116 participants over the age range of 20-103 years are actively enrolled in the BLSA. Currently, 22% of participants are <60 years (N=246), 52% are 60-79 years (N=580) and 26% are ≥80 years (N=290). The BLSA tests about 500- 600 subjects per year. Analytic strengths of the BLSA include the large sample size across a broad age range (20-103 years) and longitudinal study design, allowing for assessment of population- and individual-level variation in vestibular physiologic function and associated outcomes.

We have added vestibular testing to the test battery that all BLSA participants undergo. Vestibular testing consists of video-head impulse testing (vHIT) in each semicircular plane as a measure of canal function, cervical vestibular-evoked myogenic potential (cVEMP) as a measure of saccular function, and ocular VEMP (oVEMP) as a measure of utricular function. A major strength of the BLSA is that other rigorous, comprehensive measures are also collected. Notably, the BLSA has a state-of-the-art gait laboratory, consisting of a 10-meter walkway and a 3D Vicon motion capture system for detailed gait analysis. Additionally, important potential confounders are also rigorously measured in the BLSA as follows:

- 1) Vision: contrast and glare sensitivity, visual acuity (with an auto-refractor), visual fields (with a frequency doubling technology perimeter), color vision, and stereopsis (depth perception);
- 2) Sensorimotor function: ankle proprioception testing (quantitative ankle plantarflexion and dorsiflexion proprioception using a custom dual-footpedal threshold test), strength testing (of knee flexion and extension with the Biodex system), quantitative ankle, knee and hip range of motion testing;
- 3) Neurological and cognitive testing: an extensive battery of cognitive testing is performed that measures cognitive processing speed, executive function, verbal fluency, and general intelligence.

Additionally, non-contrast brain MRI's are obtained on all participants at each visit;

- 1) Audiometry: pure-tone and speech audiometry.

This collaboration with the BLSA represents an unprecedented opportunity to leverage existing NIA infrastructure and rigorously characterize the changes in semicircular canal and otolith function that occur with aging, in a large well-studied cohort of older adults followed longitudinally. We intend to fully explore the richness of the BLSA data, which include extensive neurocognitive testing, broad biomarker and genetic assays, and brain MRI scanning *on all participants every 1-5 years.*

(CONT)

For example, in addition to the aims outlined above, we are interested in examining whether age-related vestibular loss may lead not only to physical decline but also to dementia, consistent with clinical observations that patients with dizziness often present with concurrent difficulties with thinking and memory. We will use sophisticated analytic strategies including longitudinal data analysis and structural equation modeling to explore this rich array of data. We will generate and test conceptual models that will establish the direction for the next set of research questions.

With respect to Aim 2, the BLSA also collects self-reported falls data. We will make use of the longitudinal study design to evaluate whether age-related vestibular loss at a given time point is an independent predictor of incident falls that occur in the subsequent observation period. Finally, with respect to Aim 3, we are currently working on coding and analysis of the gait laboratory data. Our goal will be to identify whether certain quantitative parameters of gait are associated with fall risk.

I am very pleased to report that my K23 proposal to the NIDCD was recently funded, with the award period starting July 1, 2014. The preliminary data for this proposal was generated using funds from the AOS Clinician-Scientist Award, and I am very grateful to the Society for your support during this very critical period at the start of my career.

Publications from our research group since the last progress reports include:

Willis JR, Vitale SE, Agrawal Y, Ramulu PY. Visual Impairment, Uncorrected Refractive Error, and Objectively Measured Balance in the United States. *JAMA Ophthalmol.* 2013 Jun 6:1-7.

Ward BK, Agrawal Y, Hoffman HJ, Carey JP, Della Santina CC. Prevalence and Impact of Bilateral Vestibular Hypofunction: Results From the 2008 US National Health Interview Survey. *JAMA Otolaryngol Head Neck Surg.* 2013 Aug 1;139(8):803-10.

Agrawal Y, Davalos-Bichara M, Zuniga MG, Carey JP. Head Impulse Test Abnormalities and Influence on Gait Speed and Falls in Older Individuals. *Otol Neurotol.* 2013 Aug 6.

Mantokoudis G, Schubert MC, Saber Tehrani AS, Wong AL, Agrawal Y. Early Adaptation and Compensation of Clinical Vestibular Responses After Unilateral Vestibular Deafferentation Surgery. *Otol Neurotol.* 2013 Aug 20.

Agrawal Y, Schubert MC, Migliaccio AA, Zee DS, Schneider E, Lehen N, Carey JP. Evaluation of Quantitative Head Impulse Testing Using Search Coils vs. Video-oculography in Older Individuals. *Otol Neurotol.* 2013 Sep 27.

Agrawal Y, Bremova T, Kremmyda O, Strupp M, MacNeilage PM. Clinical Testing of Otolith Function: Perceptual Thresholds and Myogenic Potentials. *JARO.* 2013 Sept 27.

Davalos-Bichara M, Agrawal Y. Normative Results of Healthy Older Adults on Standard Clinical Vestibular Tests. *Otol Neurotol.* 2013 Oct 16.

Agrawal Y, Ward BK, Minor LB. Vestibular dysfunction: Prevalence, impact and need for targeted treatment. *J Vestib Res.* 2013 Jan 1;23(3):113-7.

Bremova T, Bayer O, Agrawal Y, Kremmyda O, Brandt T, Teufel J, Strupp M. Ocular VEMPs indicate repositioning of otoconia to the utricle after successful liberatory maneuvers in benign paroxysmal positioning vertigo. *Acta Otolaryngol.* 2013 Dec;133(12):1297-303.

Baxter M, Agrawal Y. Vestibular dysfunction in Turner syndrome: a case report. *Otol Neurotol*. 2013 (In press).

Sun D, Davalos-Bichara M, Zuniga MG, Carey JP, Agrawal Y. Evaluation of a bedside test of utricular function – the bucket test – in older individuals. *Acta Otolaryngol*. 2013 (In press).

The K08 (basic research) and K23 (patient oriented research) are mentored research training grants for physicians that are the standard launch for a junior faculty clinician-scientist career. Most physician R01's are awarded to former recipients of these awards. An appropriate goal of the AOS is to assist promising junior faculty in obtaining the preliminary data needed to obtain these now extremely competitive awards which are \$230,000 per year over 5 years to defray both salary and supply costs. The mentors on these awards must be experienced, NIH-funded investigators.

AOS should be exceedingly proud that in 2014, all three of its 2013 AOS Clinician-Investigator awardees have been awarded K grants. An astounding accomplishment in the current federal funding climate.

Their K awards allow them to relinquish their AOS funding so that resources are now freed to fund other promising young clinician-scientists.

The American Otological Society, Inc., through its Research Fund offers Research Grant Awards, an Award for a Clinical Trial, full-time Research Training Fellowships, and a Clinician-Scientist Award.

Research supported by the Research Grant Awards and by the Award for a Clinical trial must be focused on the study of otosclerosis or Ménière's disease. A description of the relationship between the research study and otosclerosis or Ménière's disease is an integral part of the application.

The full-time Research Training Fellowships and the Clinician-Scientist Award can support the training of the recipient in research on any topic related to ear disorders.

These grant awards and fellowships are for work conducted in United States or Canadian institutions only. Additional details may be found on the AOS website at www.americanotologicalsociety.org

The deadline for grants and fellowship applications must be received by January 31, 2015 for consideration of the July 2015 – June 2016 funding cycle.

*John P. Carey, MD
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H. Alexander Arts, MD (Active 2001)
Ann Arbor, MI

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Tokyo, Japan

Michel Portmann (Honorary 1983)
Bordeaux, France

Naoaki Yanagihara, MD (Honorary 2008)
Matsuyama, Japan

IN MEMORIUM (in alphabetical order)

The AOS Administrative office was notified of the following members death since the last Spring meeting.

Joseph C. Farmer, Jr., MD - member since 1984

William E. Hitzelberger, MD - member since 1997

Robert A. Jahrsdoerfer, MD - member since 1982

Claude L. Pennington, MD - member since 1973

Please take a moment of silence to remember these outstanding colleagues & friends.

We need your help, do you know this member?

The AOS Administrative Office has lost contact with the following members:

- Roger Boles, MD
- John M. Epley, MD
- Richard L. Ruggles, MD
- Robert A. Butler, MD
- Isamu Sando, MD

If you know of the whereabouts of any of the above members, please contact the AOS Administrative office at 217-638-0801 or by email: administrator@americanotologicalsociety.org