Note from the Director

I am pleased to present to you the inaugural issue of Health Letter of the Mount Sinai Hearing and Balance Center. In this newsletter, we hope to familiarize the public with the members of our staff, and with the latest trends in otology and hearing healthcare. As disorders of hearing and balance have become increasingly prevalent, our ability to diagnose and treat these disorders has advanced as well.

The contemporary management of hearing and balance problems is by necessity a team approach. The diagnostic side consists of medical, audiologic, and radiologic specialists. The treatment of hearing impairment now includes medical, surgical, and rehabilitative therapies which include hearing aids, implanted hearing devices and cochlear implants.

Balance disorders can now be diagnosed with unprecedented accuracy. Medical therapies and vestibular rehabilitation have made it possible to offer relief to the majority of patients who suffer from vertigo. Our colleagues in neurology and rehab medicine have been a great asset in this regard. Collaborative efforts with colleagues in neurosurgery, interventional radiology, and intensive care medicine have also made Mount Sinai a world-renowned center for acoustic neuroma and skull base surgery.

Our goal in these newsletters is to educate both the public and general medical practitioners including general otolaryngologists, medical physicians, pediatricians, and hearing health professionals. We hope that our readership will contribute suggestions, critiques, and original ideas to this newsletter and we look forward to your input.

Eric Smouha, MD
Director of Otology-Neurotology

Avoiding and Managing Facial Nerve Complications

The facial nerve is vulnerable to injury in surgery of the ear, head and neck, and skull base. Despite advances in surgical technique, facial nerve weakness cannot always be prevented. This important topic was the subject of a miniseminar at the last annual meeting of the American Academy of Otolaryngology-Head and Neck Surgery in Chicago, whose aim was to review the surgical anatomy of the facial nerve, the role of facial nerve monitoring, intraoperative management, and treatment of consequences of facial nerve injury. Dr. Eric Smouha moderated the seminar, and noted experts in the field contributed; including Barry Schaitkin from the University of Pittsburgh, Sam Marzo from Loyola University, Chicago, and Fred Fedok from Penn State Medical Center at Hershey.

The proper diagnosis of facial nerve problems is vital to preventing complications. Bell's palsy is the most common cause of facial paralysis. It develops quickly (days), is usually accompanied by pain in or around the ear, and always recovers at least partially. However, other more serious conditions are sometimes misdiagnosed as Bell's palsy. It is important to recognize that slowly progressive paralysis, pain, isolated branch weakness, multiple cranial nerve involvement, and failure to recover function are signs of tumor, and this fact highlights the need for MRI or CT imaging in atypical cases.

Knowledge of facial nerve anatomy is essential for safe surgery, however the anatomy may be hidden by disease. A familiarity with normal and pathological surgical anatomy is imperative in mastoid, parotid, skull base, and even facial plastic surgery, where the facial nerve can be at risk. Complications most often occur when there is distorted anatomy, as in cholesteatoma, or unanticipated pathology, such as a facial nerve tumor presenting as a benign parotid mass. Managing a facial nerve injury intraoperatively requires clear surgical judgment; the steps that are taken in an operative crisis will often determine the final outcome.

continued on page 2
Reconstructed Canal Wall in Cholesteatoma Surgery

Cholesteatoma is an abnormal growth of skin inside the ear that has the capacity for infection and bone destruction. The ideal treatment for cholesteatoma continues to be the subject of debate. The goals of cholesteatoma surgery are to eliminate the disease, restore hearing, and create a safe dry ear. For the last generation, discussions of management of cholesteatoma have centered around “canal-wall-up” vs. “canal-wall-down”. Canal-wall-up surgery preserves the normal ear canal anatomy, heals rapidly, avoids the need for water precautions, and readily permits the use of a hearing aid. However, recurrent cholesteatoma remains relatively common, and so a second operation is sometimes necessary. Canal-wall-down surgery, on the other hand, provides better surgical access and nearly eliminates the chance of recurrence, but the creation of a mastoid cavity necessitates periodic cleaning, water precautions, and the risk of mucosal drainage.

The ideal treatment for cholesteatoma would be a single-stage operation with very low recurrence rate and no need for maintenance. The technique of canal wall reconstruction attempts to bridge the gap between canal-wall-up and canal-wall-down, by combining the virtues of both approaches (several methods have been tried over the years but they have had their drawbacks). For the past few years at Mount Sinai, we have utilized a novel technique of canal wall reconstruction in which the bony ear canal is first removed en bloc (in one piece) in order to provide wide unobstructed view of the middle ear and mastoid. The disease can then be removed completely, with no compromise to surgical exposure. The canal wall is then replaced, and the ear canal skin is rearranged over it, to create a small, manageable surgical cavity.

We have used this method selectively in a series of patients with excellent results, and we presented our findings at the 2007 AAO-HNS meeting [G. Wanna and E. Smouha, “Beyond canal wall up/canal wall down: Replacing the canal wall in cholesteatoma surgery”]. We have achieved a very low complication rate, with no instances of recurrent disease and only one case needing revision surgery, and with very little need for aftercare in the office.

Of course, cholesteatoma is not a one-size-fits-all disease, and treatments must be individualized. Our reconstructed canal wall method is best suited for patients with large, well-developed mastoids who have disease in hard-to-reach areas, and the decision to perform this procedure is generally determined at the time of surgery.

At Mount Sinai, our goal is to try to treat cholesteatoma with a single operation whenever possible. Reconstructed canal wall has provided an important option for patients who classically would have required two-stage surgical management.

Avoiding and Managing Facial Nerve Complications

Facial nerve monitoring has been widely adopted in otologic and parotid surgery. Although this has increased the margin of safety, it has certain limitations. The information gained from facial nerve monitoring must be taken in the context of the surgery, such as when applying traction on soft tissue or when drilling on bone. Monitoring will probably not warn against a sudden transection injury however. Intraoperative facial nerve stimulation can help to locate a nerve covered by disease, confirm the integrity of a nerve during dissection, and determine the degree of conduction block after injury, making it a valuable surgical tool.

The postoperative management of the patient with facial nerve injury offers great potential to improve outcome. Immediate and delayed electrical testing plays a role in decision-making and treatment selection. Protection of the eye from dryness is of paramount importance. Primary facial nerve repair with grafting is sometimes feasible, but plastic and reconstructive surgical interventions are many and can give gratifying results when necessary.
Introducing the Staff of Mount Sinai’s Hearing and Balance Center

ERIC SMOUHA, MD, FACS is an Associate Professor of Otolaryngology and Director of Otology and Neurotology of Mount Sinai School of Medicine. Dr. Smouha is board-certified in both Otolaryngology and Neuro-Otology. With over twenty years of experience, his clinical and academic interests span all aspects of otology and neurotology.

Dr. Smouha has performed over four hundred operations for cholesteatoma and has given instructional courses on cholesteatoma and complications of otitis media at the American Academy of Otolaryngology–Head and Neck Surgery and in Graduate Education at Mount Sinai. With expertise on hearing preservation, he has performed hundreds of successful stapedectomy operations for the treatment of otosclerosis, developed innovative techniques aimed at conserving function in cholesteatoma surgery, as well as newer surgically-implanted hearing devices including BAHAs.

His publications include the management of cholesteatoma in the normal hearing ear, the time course of recovery of benign positional vertigo, and the medical and surgical treatment of Meniere’s disease. He has also conducted basic research on matrix metallo-proteinase enzymes in cholesteatoma, and on three-dimensional imaging of the temporal bone. Additionally, Dr. Smouha has done pivotal basic research in hearing preservation surgery of the inner ear and has re-established a Cochlear Implant program at Mount Sinai.

As a neurotologic surgeon, Dr. Smouha has expertise in the treatment of acoustic neuromas and diseases of the skull base. He is a fellow of the American Academy of Otolaryngology–Head and Neck Surgery and the American College of Surgeons; and a member of the American Otological Society, the American Neurotological Society, the Triological Society, the North American Skull Base Society; and, a recipient of the Certificate of Honor of the American Academy of Otolaryngology-Head and Neck Surgery.

Recently voted Teacher of the Year by the resident staff, his devotion to training physicians has further established excellence in the otology teaching programs. His contributions to the field of Neuro-Otology at Mount Sinai have established him as a leader in his field and an outstanding clinician.

DEBRA FRIED
Diagnostic Audiology and Cochlear Implantation

Debra Fried, M.S., CCC-A received her Master of Science degree from Teachers College, Columbia University. Prior to joining the Mount Sinai Medical Center, she worked at Saint Louis University Hospitals. In addition to providing diagnostic audiology services, Debra manages Mount Sinai Hospitals newborn hearing screening program and she is actively involved in the Cochlear Implant Program. Since 2004, she has been traveling to Jinotega, Nicaragua several times a year as a volunteer on medical missions providing diagnostic and hearing aid services to the indigent residents in that region of the country.

KAREN SIEGEL
Audiology—North Shore Medical Group

Karen E. Siegel, MA CCC-A received her Bachelor of Science and Masters of Arts degrees in Audiology from State University of New York at Plattsburgh. She is currently involved in the distanced learning Au.D. Program at the Arizona School of Health Sciences and will receive her doctorate degree in audiology in June 2009. Her work experience over the past ten years includes working at private practices in Connecticut, contracting audiological services with convalescent homes in Connecticut and New York, opening and managing a hearing aid business and more recently joining the Mount Sinai School of Medicine in January of 2008. Her expertise includes hearing aid dispensing and vertigo and balance testing/therapy. Her personal interests include composing music, singing, and playing piano and guitar. She also enjoys traveling and learning about new cultures.

JENNIFER DONATH
Diagnostic and Rehabilitative Audiology

Jennifer E. Donath, MS CCC-A received her Bachelor and Master of Science degrees in Audiology from State University of New York at Plattsburgh. She is currently working part-time at the North Shore Medical Group. Jennifer has expertise in the management ofpatients with vestibular disorders and monitoring those receiving ototoxic medications. In her free time, she enjoys reading, photography and spending time with her family.

ALLISON GREGG
Audiology at the Center for Advanced Medicine

Allison Gregg Baile received her Bachelor’s degree from West Virginia University in 2000 and her Master’s degree from Hunter College in 2002. In June 2008, she finished her coursework from Salus University to receive her Doctorate in Audiology (Au.D.). She is currently working part-time at Mount Sinai Medical Center performing patient diagnostic testing and part time in private practice. She is interested in working with patients with vestibular disorders and monitoring those receiving ototoxic medications. In her free time, she enjoys reading, photography and spending time with her family.

If you, or someone you know, would like information about supporting our work, please contact Kim Woodward in Mount Sinai’s Development Office at 212-659-1594 or at kim.woodward@mountsinai.org.
It is now well known that early detection of hearing loss leads to better outcomes. Statistically, at least 1 out of every 1000 babies are born with significant permanent hearing loss in both ears, and 2 of 1000 babies have permanent unilateral hearing loss. Without early intervention, these hearing-impaired children will have delays in language and social development, and have educational and employment disadvantages later in life.

The goal of universal newborn hearing screening is to detect hearing problems as early as possible. The Joint Commission on Infant Hearing has recommended that all newborns have their hearing screened by 1 month of age and start rehabilitation by 6 months of age. Our program aims to detect all forms of hearing impairment, including congenital bilateral or unilateral sensory or conductive hearing loss as well as auditory neuropathy. Children identified with hearing loss at birth who receive intervention by 6 months of age can achieve normal language comprehension, expression and social development by early childhood (prior to newborn screening, the average age of detection of hearing loss was over 2 years).

In New York State, mandatory newborn hearing screening has been in effect since 2002. Large medical centers like Mount Sinai are required to provide the hearing screenings prior to discharge, and the results are reported to the Department of Health on a quarterly basis.

We use fully automated screening equipment that does not require a behavioral response from the infant. Test outcomes are recorded as pass or fail. The testing program is designed with a high sensitivity, that is, children who truly have hearing loss are very unlikely to slip through the cracks. There will be a few false-positives that will need repeat testing in outpatient follow up.

At Mount Sinai, with 6,000 births per year, we have been conducting hearing screenings since 2002. Two full-time trained technicians provide screening services 6 days per week, and one audiologist, Debra Fried, manages the program. Two types of audioligic testing procedures are used, either alone or in combination. Otoacoustic Emissions (OAE) are a measure of sound waves generated by the inner ear in response to clicks. It is quick and simple to administer, and can detect even mild hearing impairment. Automated Auditory Brainstem Response testing (ABAER) is used in babies who fail the OAE screening. This test measures electrical brain activity in response to click sounds, and takes a bit longer to perform. Using this 2-tiered test approach (OAE for everyone and ABAER for confirmation) achieves a high sensitivity with few false-positive tests.

Each year, we identify several new babies with hearing loss using these methods of early detection. With modern techniques of hearing rehabilitation (including the option of cochlear implantation), hearing-impaired children are now able to maximize their speech, language, and reading skills. When verbal communication is chosen, these children can expect to be mainstreamed into the regular school system by the time they reach the first grade.