



ANA/NJ Newsletter
Vol. XV, No. 2, April 2016

Chapter Meeting, Morristown
October 18, 2015



Our Fall chapter meeting was held at the Morristown Medical Center. Twenty-five AN patients and caregivers attended. The meeting featured an open discussion presided over by Dr. Richard M. Hodosh, a dedicated neurosurgeon at Morristown and Overlook Hospitals for many years and long-time member of our Medical Advisory Board. Dr. Hodosh played a key role in the organization and success of our association's debut conference for acoustic neuroma in New Jersey held at Morristown in 1996; and more recently he has been a mainstay on doctors' panels for our biennial mini-conferences.

Dr. Hodosh began the Oct 18 meeting with some reflections about the success of microsurgery for AN today compared to the early 1900s when total tumor removal was generally considered to be impossible and doctors hoped their patients would be able even to survive acoustic neuroma surgery. Those were the days of Dr. Harvey Cushing (1869-1939), one of the pioneers of modern brain surgery at Harvard and Yale, who is often called the 'father of neurosurgery'.¹ Dr. Hodosh recalled that his own first acoustic neuroma surgery was done in 1975, by which time surgeons had the advantages of the operating microscope and proven surgical techniques for AN. He said that as a young neurosurgeon he enjoyed the challenge of going for total removals. Eventually, however, with the development of MRI imaging and advances in radiation technology, total removals became less the standard of care at least for large tumors. With facial nerve and hearing preservation in mind, partial removals by microsurgery followed by radiation treatment of 'residuals' (tumor remnants) began to be done.² In this regard, Dr. Hodosh coordinated with Dr. Louis Schwartz at the Overlook Hospital CyberKnife Center.

Discussion during the meeting focused on experiences with small tumors and residuals, Wait-and-Watch, and surgery vs. radiosurgery vs. radiotherapy as treatment options. Noticeably, those asking questions were quite knowledgeable about acoustic neuroma and its treatment. They sought support for decisions mostly already made after a good deal of investigation and thought. Dr. Hodosh was happy to provide input based on his clinical experience over many years.

The meeting appeared to confirm something that Wilma Ruskin has written just recently: "We feel that the 'face' of acoustic neuroma patients has changed dramatically over the past 20 years, from mostly post-surgery patients with severe problems, to newly diagnosed patients who are looking for information that will help them make informed decisions about their treatment options. This is a remarkable 'change of face' which, we believe, has been advanced significantly by the work of ANAUSA and local groups like ANA/NJ all around the country."

¹ See *Harvey Cushing: A Life in Surgery*, by Michael Bliss (Oxford Univ Press, 2005).

² See below, "More about Residuals," p.6. Dr. Hodosh spoke about partial removals at our chapter meeting at Overlook Hospital, April 3, 2005.



Jane Meryll in the Spotlight

Imagine what it must be like for someone whose life and livelihood is music and the performing arts to be told she has an acoustic tumor. To make matters worse, her first doctor went on to tell Jane that if she didn't have surgery to remove her 8 mm tumor as soon as possible, she could die!



The year was 1996 and Jane, aged 52, had been experiencing transient vertigo, dizziness and seeing spots. Although she immediately scheduled the surgery, from which she was told she would lose her hearing, she began researching her condition. Her cousin, a neurosurgeon told her not to have the surgery and advised her to wait-and-watch to see if the tumor grew. She'd read that in Europe this was a common practice and that if a small tumor didn't grow in the first 12 months, it likely never would. She cancelled her surgery and received a letter from the diagnosing doctor confirming that without the surgery she might die.

Jane became very involved in the NY Chapter of The Acoustic Neuroma Association and served on its board. She became a patient of Dr. Samuel Selesnick because he was willing to follow her as a wait-and-watch patient having checkups every few years. She expressed emphatically that she is not an alarmist and she was determined not to let her diagnosis impinge on her life. She had come to NY in 1968 from Boston to pursue a professional career in music as a singer and pianist. She married a producer and worked in his business as a freelance studio musician doing advertising music and recording as a pianist, singer, composer, producer and arranger. Theirs was a 30-year collaboration, so when they divorced, she was left to pursue a career on her own. She added a masters degree in education and psychology to her earlier attendance at the Berklee College of Music so that she could teach young people, particularly in special education. She moved into coaching and giving private lessons in piano, voice, and overcoming stage fright (to learn more visit www.janemeryll.com). She continues to work approximately 25 hours a week between her adjunct professor work and her private studio work and she plans to produce a legacy CD of her compositions. She feels composing is something she can and will do until the end of her life and is grateful to have her hearing so she is able to hear her compositions, unlike Beethoven.

About a year ago Jane experienced some serious health issues including shingles and the flu and she again experienced vertigo. She had an MRI and went to see Dr. Selesnick who advised that her tumor is now "a non- issue" and that she will not need to see him for it again. Since diagnosis, there has been no change in it whatsoever. Her hearing was tested and she was found for all intents and purposes to have normal hearing. Although she has lost some of the acuity on her tumor side, particularly in noisy environments, she has been able to make minor adaptations to address this. Jane was told, however, that she had BPPV (Benign Paroxysmal Positional Vertigo) which was the cause of her vertigo. She did some exercises for this for a few days and it relieved. So far it hasn't reoccurred.

Twenty years after diagnosis, Jane is still going strong believing her earlier decision not to have surgery has been the right decision for her. She feels prepared for whatever life brings her and would encourage anyone diagnosed with an acoustic neuroma to research, research, research and to make the right decision for him/herself. She encourages patients to believe in their own informed insight, not to be intimidated by the title of doctor. She says "no one knows what's better for you than you. You must be your own best advocate!"

Interview by Kristin Ingersoll
February 15, 2016

NIDCD Statement on the Cause of Acoustic Neuroma

The following is the current statement on the cause of acoustic neuroma issued by the National Institute on Deafness and Other Communication Disorders (NIH):

Unilateral vestibular schwannomas [aka. acoustic neuromas] affect only one ear. They account for approximately 8 percent of all tumors inside the skull; one out of every 100,000 individuals per year develops a vestibular schwannoma. Symptoms may develop at any age but usually occur between the ages of 30 and 60 years. Unilateral vestibular schwannomas are not hereditary.

Bilateral vestibular schwannomas affect both hearing nerves and are usually associated with a genetic disorder called 'neurofibromatosis type 2' (NF2). Half of affected individuals have inherited the disorder from an affected parent and half seem to have a mutation for the first time in their family. Each child of an affected parent has a 50 percent chance of inheriting the disorder. Unlike those with a unilateral vestibular schwannoma, individuals with NF2 usually develop symptoms in their teens or early adulthood. In addition, parents with NF2 usually develop multiple brain and spinal cord related tumors. They can also develop tumors of the nerves important for swallowing, speech, eye and facial movement, and facial sensation. Determining the best management of the vestibular schwannomas as well as the additional nerve, brain and spinal cord tumors is more complicated than deciding how to treat a unilateral vestibular schwannoma. Further research is needed to determine the best treatment for individuals with NF2.

Scientists believe that both unilateral and bilateral vestibular schwannomas form following the loss of function of a gene on chromosome 22. (A gene is a small section of DNA responsible for a particular characteristic like hair color or skin tone.) Scientists believe that this particular gene on chromosome 22 produces a protein that controls the growth of Schwann cells. When this gene malfunctions, Schwann cell growth is uncontrolled, resulting in a tumor. Scientists also think that this gene may help to control the growth of other types of tumors. In NF2 patients, the faulty gene on chromosome 22 is inherited. For individuals with unilateral vestibular schwannomas, however, some scientists hypothesize that this gene somehow loses its ability to function properly.

Scientists are working to better understand how the gene works so they can begin to develop gene therapy to control the overproduction of Schwann cells in individuals with vestibular schwannomas. Also, learning more about the way genes help control Schwann cell growth may help to prevent other brain tumors.

(The NIDCD provides a directory of organizations to contact for further information on vestibular schwannomas at www.nidcd.nih.gov/directory.)

Funding Big Science: How Much?

Our Sept 2015 issue called attention to the US "Brain Initiative" launched in 2013 and intended to end the centuries-long search for how the brain actually works. The European Union has had its own "Human Brain Project" since 2012. An article by Stefan Theil in *Scientific American* (Oct 2015) comparing investments in these and other projects raises the question of the degree of human commitment to serious study of the brain. Note the following:

Project	Funding
US Brain Initiative, 2013-15	\$300 million
EU Brain Project, 2012-23	\$1.63 billion
Human Genome, 1990-2003	\$4.73 billion
Apollo Program, 1960-1973	\$104 billion
Space Station, 1998-2001	\$140 billion
F-35 Fighter Jet (as of 2014)	\$391 billion
Manhattan Project, 1942-45	\$23 billion
Atomic bomb	

Tinnitus Research Update

Researchers at Georgetown University Medical Center (GUMC) in Washington, D.C., and the Technische Universität München (TUM), Germany, have announced that they have uncovered the brain malady responsible for tinnitus and chronic pain. “Identifying the problem,” they observe, “is the first step to developing effective therapies for these disorders, which afflict millions of people.”³

The lead researchers are Josef Rauschecker, PhD,DSc, director of the Laboratory for Integrative Neuroscience & Cognition at GUMC, and Markus Ploner, MD, PhD, Professor of Human Pain Research at TUM.



Dr. Rauschecker

According to the September 2015 news release by GUMC: “The scientists describe how the neural mechanisms that normally ‘gate’ or control noise and pain signals can become dysfunctional, leading to a chronic perception of these sensations. They traced the flow of these signals through the brain and showed where ‘circuit breakers’ should be working – but aren’t. . . Tinnitus can occur after the ears are damaged by loud noise, but even after the brain reorganizes itself, it continues to ‘hear’ a constant hum or drum. . . Areas of the brain [in the limbic system] responsible for these errant sensations act as a central gatekeeping system for perceptual sensations, whether produced externally or internally. . . Tinnitus and chronic pain occur when the system is compromised.”

Possible therapy? “Because these systems rely on transmission of dopamine and serotonin between neurons, drugs that modulate dopamine may help to restore sensory gating,” states Dr. Rauschecker.

Increase in Incidental Findings

Dr. Mark E. Robson , who is the Director of the Genetics Service Clinic at Memorial Sloan Kettering Cancer Center (MSKCC) in New York, has reported that genome analysis of cancer tumors has begun to reveal patient risks for other diseases (*Center News*, April 2014). These incidental genetic findings are being dubbed “incidentalome.” He notes: “Most clinical testing of [cancer] tumors has been for a relatively limited number of specific mutations, not the full genome. But soon we’re going to be testing for a much broader panel of genes, increasing the chances of incidental findings.”

Dr. Robson’s report reminded us of how the introduction of MRI imaging also resulted in finding “incidentalomas,” including small and/or asymptomatic acoustic neuromas that are so important to identify early on for best treatment outcomes (See “Incidental Acoustic Neuroma,” in *ANA/NJ Newsletter*, March 2012).

For Dr. Robson and researchers at MSKCC, the incidental genetic findings have raised the ethical question of the extent to which clinicians should inform patients of their possible health risks. Do all patients want to know everything? It’s a complicated issue for doctors and researchers. But meantime, advances in cancer research and genome analysis will work to the benefit of acoustic neuroma patients. “All boats are lifted” is a good way to think of how basic research acts as a rising tide for advancement in all areas of medical science.

³ See “Neuroscientists Uncover Brain Abnormalities Responsible for Tinnitus and Chronic Pain,”

www.gumc.georgetown.edu/news, 10/17/2015;

Josef Rauschecker et al, “Frontostriatal Gateway of Tinnitus & Chronic Pain,” *Trends in Cognitive Sciences*, 19 (October 2015).

More About ‘Residuals’

As it was 90 years ago, the question of partial versus total tumor resection surgery, at least for medium to large tumors, is now up for debate.

Dr. Marc S. Schwartz

Our Sept 2015 newsletter article entitled “Thinking about Residual Tumor” looked at some early reports on outcomes for ‘facial nerve-sparing surgery’, a procedure entailing partial removal of large acoustic neuromas followed (as needed) by stereotactic radiation treatment for ‘residuals’, the tumor remnants left behind. The basic goal of the approach is to maximize both facial nerve function and patient quality of life. The article concluded with the recommendation that long-term data on outcomes for partial resection surgery will be needed.

Anyone considering having partial removal of a medium or large tumor would do well to read Dr. Marc Schwartz’s report on “Gross Total vs. Near Total vs. Partial Resection Surgery” in *ANA Notes*, Issue 135 (Sept 2015). Briefly, Dr. Schwartz (House Clinic, Los Angeles) reports: “There is as of yet little short-term and no reliable long-term data showing the results of stereotactic radiation following partial resection of larger acoustic neuromas.” Secondly, “[although] long-term results are not known, . . . it is probably reasonable to expect that facial nerve function preserved at 1-year will be maintained long-term in most patients.” On the other hand, thirdly: “Stereotactic radiation carries significant risk to hearing, so if this is carried out either directly after surgery or at a later time, its risk to hearing must be factored in. Thus in regard to hearing, the risks of surgery itself, presence of tumor on the [cochlear] nerve, and subsequent radiation, taken together, make the long-term chance of hearing preservation quite low.”

Although supportive of quality of life goals, Dr. Schwartz remains skeptical that treatment outcomes for acoustic neuroma are improved significantly by less aggressive procedures. “Today,” he writes, “the risk of major neurological complications with total resection of even large and giant acoustic neuromas is very low in the hands of experienced, high volume surgical teams.” He expresses concern that less experienced surgeons may oversell the benefits of partial resection surgery.

Hearing Rehab Webinar

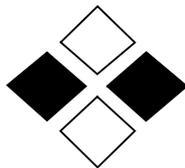
ANAUSA’s recent webinar entitled “*Hearing Rehabilitation for Acoustic Neuroma Patients*,” by Dr. Brian Neff, Professor of Otolaryngology at the Mayo Clinic, is an excellent, up-to-date review of the various options for hearing restoration following treatment for acoustic neuroma.

Dr. Neff’s presentation with slides is well-organized and informative. Listeners are able to ‘Pause’ the audio for leisurely viewing of text summaries and graphics. A main theme of the presentation is that tumor size and pre-operative hearing capacity are two key variables predictive of hearing outcomes and the type and level of hearing rehab technology that may be needed following treatment. Dr. Neff presents data from his study at the Mayo Clinic dealing with “variables influencing audiometric decline.”⁴ There is a good discussion of CROS technology (Contralateral Routing of Sound) for SSD, e.g., the Widex system. The Baha bone-anchored system is reviewed and compared in its effectiveness with the Cochlear Implant (CI); testing has shown the CI to be better with sound localization and for dealing with noisy places. Medicare still does not give coverage for the CI for SSD patients. There’s a heads-up for SSD patients: besides avoiding loud noises, be sure to check your medications for possible Hearing loss side-effects.

⁴ *Journal of Neurosurgery*, 118 (March 2013)

Notices

- ANA/NJ has received a grant from the Lincoln Family Foundation. Board members have voted to use part of the grant to reduce the registration fee for attendance at the Mini-Conference scheduled for April 24 (See below).
- In *Journal of Neurosurgery, Neurosurgical Focus Section*, 33 (Sept 2012), Drs. James Liu, R. Jyung, R. Schmidt et al have published “Incidental Vestibular Schwannomas: A Review of Prevalence, Growth Rate and Management Challenges”; also, Drs. James Liu, Derald Brackmann et al have published “An Update on Unilateral Sporadic Small Vestibular Schwannoma.” Text available at www.ANAUSA.com, Members Section, Journal Articles.
- Jane Huck has called attention to a report on research by Scottish scientists (Univ of Strathclyde) testing a new hearing aid design for eliminating unwanted background noise based on studies of how insects hear sounds. For more, go to www.meddeviceonline.com/insect-inspired-hearing-aids.
- Neurologists Peter Goadsby (UCSF) and David Dodick (Mayo Clinic Arizona) believe they have identified drugs – monoclonal antibodies – specifically designed to prevent migraine headaches from occurring rather than stopping the attacks once they have started. Science writer David Noonan reports on the story in *Scientific American* (December 2015); see also his report on vertigo in the August 2015 issue. For more on migraine, visit the website of the American Academy of Neurology, www.aan.com/patients.
- Most funding for acoustic neuroma research is by the National Institutes of Health (www.nih.gov). For example, \$21 million was granted for NF2 research projects for each of the years 2014 and 2015. For 2016, an estimated total of \$4 billion will be granted for research in the category of ‘brain disorders’; \$22 million has been designated for NF2 studies. The American Hearing Research Foundation is a private non-profit organization (www.american-hearing.org) that makes small grants of up to about \$20,000 for acoustic neuroma research. The organization is interested mainly in funding research in the area of early detection of hearing disorders. The current president is Dr. Alan Micco, associate professor of otolaryngology at Northwestern Medical School, Evanston, IL. Donations in support of research are welcome.



ANA/NJ Mini Conference

“Acoustic Neuroma 2016: Life Before, Life After”

Sunday, April 24, 2016 9:30 a.m. – 4:00 p.m.

The Conference Center at JFK Medical Center
65 James Street, Edison, NJ

Program

Registration & Coffee	9:30-10:30
Welcome by Wilma Ruskin, President ANA/NJ	10:30
Welcome by Dr. Joseph C. Landolfi, JFK NeuroScience Institute	
Doctors's Panel: Diagnosis Acoustic Neuroma! What Next?	10:45-12:00
▪ Dr. James K. Liu, <i>Moderator</i> (NJMS/Rutgers)	
▪ Dr. Christopher J. Farrell (Thomas Jefferson University)	
▪ Dr. John G. Golfinos (NYU Langone Medical Center)	
▪ Dr. Joseph C. Landolfi (JFK NeuroScience Institute)	
Lunch	12:15-1:00
~ <i>Afternoon Meetings</i> ~	
Balance and Cognitive Issues	1:15-2:15
▪ M. Lucia Jimenez, DPT, MA (Elmhurst Physical Therapy and Balance Center, Queens, NY)	
Meditation Workshop	
▪ Nancy Rothman, PhD (Meditation Practitioner)	2:30-3:30
Social Time	3:30-4:00

Directions to JFK

From Newark & north: Garden State Pkwy south to Exit 131. Right off the Exit to Rt 27. South on Rt 27 to the traffic light at James St (Dunkin Donuts on the right). Turn right onto James St to the JFK Med Center seen on the left. **NOTE:** The **Conference Center** is just opposite on the right side of James St. Turn right for the Conference Center and its separate parking area. Look for ANA/NJ sign.

From Philadelphia & south: NJ Turnpike to Exit 10 to Route 1 North. On Rt 1 North. At the Menlo Park Mall, exit on the right to go around the jughandle onto **Parsonage Road**. Take Parsonage Rd, past the mall and on through the underpass, to the traffic light at Rt. 27 (landmark is a Dunkin Donuts seen on the far right). Go straight across Rt 27 (Parsonage Rd now becomes James Street) to the JFK Medical Center on your left. **NOTE:** The **Conference Center** is just opposite on the other side of James Street. **Turn right** to the **Conference Center** and its separate parking area. Look for ANA/NJ sign.

From North & West using Rt 287: Take 287 South to Exit 3 (New Durham Rd). Bear left at the light onto County Rd 501/New Durham Rd toward Metuchen. In one mile, the road bears left, becomes Middlesex Ave, and then Rt 27 North. Continue on Rt 27 N to the light at James Street. Turn left onto James St to the JFK Medical Center on the left. **NOTE** (as above): The **Conference Center** is just opposite on the right side of James St.

