ANA/NJ Newsletter
Vol. XII, No. 3, April 2011

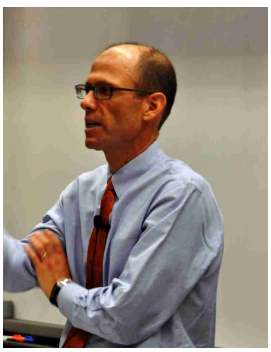
ANA/NJ Mini-Conference
October 24, 2010

Among those in attendance, there was overwhelming agreement that the 2010 Mini-Conference "Diagnosis Acoustic Neuroma: What Next?" held at the Summit Medical Group's campus in Berkeley Heights, NJ, was one of the best meetings presented by ANA/NJ for acoustic neuroma patients, family and friends. It certainly was a very busy, information-packed, and we think rewarding day for the 80-90 attendees.

Wilma Ruskin opens the Conference

At the morning session, ANA/NJ president Wilma Ruskin welcomed everyone, and Victor Mankoski, the program chairperson, introduced the members of the Doctors' Panel who were present to discuss "Patient Profiles – Treatment Options." As shown in the photo above (from left to right), the panelists were: Dr. Philip E. Stieg (Weill Cornell), Dr. John Golfinos (NYU), Dr. James Liu(UMDNJ) and Dr. Louis Schwartz (Overlook). The panel moderator was Dr. Jed A. Kwartler (Summit Medical Group). Dr. Kwartler's orchestration of the panel discussion was outstanding.

The panelists were confronted with a series of AN patient profiles and associated MRIs, and they were asked about possible treatment options, to include wait-and-watch, the surgical approaches, single-dose radiation (radiosurgery) and fractionated radiation (radiotherapy). Case No.1, for example, was that of a young male patient, age 38, who presented with no useful hearing, facial nerve weakness, and a giant acoustic tumor, as shown in the MRI projected on the large screen at the front of the room. The flow of ideas this case provoked was fascinating to follow. Briefly (1) Dr. Steig – radiation not an option in view of tumor size; wait-and-watch out because of young age; retrosigmoid surgery recommended, but could be translab (2) Dr. Golfinos – translab surgery the way to go; MRI shows the tumor one of the "soft" ones, a "sticky" problem for removal (3) Dr. Liu – a patient type that gets into trouble; translab surgery best, he's young, try to get it all (4) Dr. Schwartz – agree the tumor too large for radiation; hearing is not an issue, consider surgery to decompress.



Dr Kwartler

The interchange among medical professionals was fascinating to witness, and it became even more intriguing as Dr. Kwartler began to alter the patient profiles & MRIs: thus, Case No.2, young male, no hearing, small tumor; Case No.3, young female, moderate hearing loss, small tumor; Case No.4, large left and right side tumors, neurofibromatosis/ NF-2. A few of the many ideas generated by and discussed for these cases were:

• Hearing "preservation" for tumors more than 2.0 cm is always problematic. Radiotherapy perhaps has greater success than radiosurgery in saving useful hearing.
• In Norway the general practice is: wait-and-watch for tumors less than 2.0 cm; "dealer's choice" for 2-3.0 cm; surgery for all 3.0 cm and over.
• Malignant transformation of acoustic tumors by radiation treatment is a myth. • The head frame for Gamma Knife treatment is too uncomfortable. Perhaps for some, but isn't accuracy during radiation treatment the more important concern?
• Treatment of acoustic neuroma should not be expected to cure tinnitus.
• NF-2 is a really tough disease to treat, and maybe doctors are learning to just wait it out for many cases, with the understanding that new and better treatments are on the way.
• Monitoring hearing during surgery is still not as good as it should be.


Dr. Rosenberg

After a break for a very good lunch, the afternoon began with a presentation on "Vestibular Rehabilitation" by Dr. Michael Rosenberg, NJ Neuroscience Institute, JFK.

A diagnosis of acoustic neuroma often involves some experience with imbalance, dizziness, vertigo or visual disorientation, before or after treatment, and Dr. Rosenberg was excellent in sorting out the nature and causes of such problems. Rehabilitation, he emphasized, is a way of helping the brain "compensate for a dysfunctional vestibular system." It takes time, focus and just the right amount of practice to retrain the brain. Too much practice – a "no pain, no gain" policy – does not apply, he advised. There were many questions for Dr. Rosenberg, but he deferred these to the "Informal Discussion Session for Patients & Physicians" which he planned to attend.

The discussion session involved more medical professionals than originally anticipated. Present were not only Dr. Rosenberg, but also Dr. Schwartz from the morning panel, as well as Dina Leyden (physical therapist) and Dr. Reza Momeni (plastic surgeon) from the Summit Medical Group. Dr. Kwartler agreed to moderate the session. The day ended in discussions about various imbalance problems, tinnitus, sound directionality issues related to hearing loss, hearing loss and fractionated radiation, and how to find a good acoustic neuroma physician.


Afternoon Discussion Session

Notices

● Dr James K. Liu has been elected to membership on ANA/NJ's Medical Advisory Board. Dr Liu is Assistant Professor and Director of Skull Base & Pituitary Surgery at the Neurological Institute of NJ, UMDNJ. He is Director of the Brain Tumor Center.

● The Executive Board has invited the members of our Medical Advisory Board to contribute to a series of brief introspective articles on *Acoustic Neuroma Today* for the Newsletter. Dr. Samuel Selesnick's excellent article, "The Wait and Scan Approach: A Treatment Choice with Benefits and Risks," (see below) is the first in the series. We look forward to presenting other contributions to *Acoustic Neuroma Today.*

● A Winter "Share & Care" meeting was held January 23, 2011 at the Community Recreation Center, William Birch Park, Fort Lee, NJ. The meeting was hosted by Alice Lolas. Board members Donna Carides and Debbi Bifulco helped coordinate. In spite of snow and cold weather, nine people attended and there was much lively discussion.

● A second Winter "Share & Care" meeting, hosted by Board members Tim & Karen Reid, was held on February 26, 2011, at the Ocean County Library, 101 Washington Street, Toms River, NJ. Twenty people attended, including Board members Jane Huck (and husband Charlie), Victor Mankoski, and our President Wilma Ruskin.

● For its Focusing on Treatment series, the American Brain Tumor Association has published a new 15-page pamphlet entitled "Stereotactic Radiosurgery." The pamphlet can be read at www.abta.org, or to request a copy call 1-800-886-2282 or e-mail to info@abta.org.

New Research on Regrowing Hair Cells

Researchers at Stanford University School of Medicine led by Dr. Stefan Heller have reported using stem cells from mouse embryos to produce functional hair cells that can be used for experimentation. The scarcity of human hair cells for experimentation has impeded research into the biochemical basis for the inner ear's inability to regrow hair cells. More is known about sensory cells in the eye or the nose, where such cells are "plentiful, renewable and easy to extract." But now, Dr. Heller and his colleagues have been able to produce groups of cells with the ability to respond electrochemically to sound vibrations. "Short range, [the goal of the scientists] is to grow abundant numbers of working hair cells for further research. Long range, they hope to use what they discover to restore the ear's ability to regenerate hair cells. (Findings reported in the journal *Cell*, May 14, 2010. See www.nidcd.nih.gov, "Researchers Cook Up First Recipe for Functional Hair Cells from Stem Cells.")

The research at Stanford was funded by the National Institute on Deafness and Other Communication Disorders (NIDCD), one of the 27 Institutes, Centers & Offices of the National Institutes of Health in Bethesda, Maryland. Established in 1988, NIDCD supports biomedical and behavioral research on the processes of hearing, balance, smell, taste, voice, speech and language. The Institute supports and conducts approximately 600 research projects. It sponsored the 1991 NIH Consensus Development Conference on "Acoustic Neuroma"; and the 1995 Consensus Development Conference on "Cochlear Implants in Adults and Children." Special Workshops on "Tinnitus" were organized for December 2005 and August 2009. At the Institute's very informative website, www.nidcd.nih.gov, numerous free publications on hearing, balance, etc. can be ordered. There is an archived newsletter. Interested persons can also examine the Institute's budget, research plans and annual reports to Congress.

NIH: Some Insights

In his book *The Art and Politics of Science* (2009), Dr. Harold Varmus provides us with valuable insights into the growth and successful functioning of the National Institutes of Health (NIH), the world's largest agency for biomedical research. Dr. Varmus, who was director of NIH from 1993 to 2000, and most recently president of Memorial Sloan-Kettering Cancer Center, explains the origins of NIH's 27 Institutes, Centers and Offices (each with its own appropriated funds). He writes: "Each. . .was legislated into being by members of Congress, commonly working together with citizen advocates, who believe that some aspect of biomedical research – a specific disease (like cancer or arthritis), a specific organ (like the heart or lung), a time of life (like aging or childhood), or a discipline (like nursing or bioengineering) – can benefit from the creation of a unit of the NIH devoted to it." During his tenure as director, Dr. Varmus tried to restrain further proliferation, for example by observing how proposed new activities (e.g., alternative medicine, minority health, and imaging) could be assigned to preexisting agencies or coordinating bodies. (See Varmus, "Proliferation of National Institutes of Health," *Science*, vol 291, March 9, 2001) But resistance to the trend proved especially difficult because of the political popularity of medical research. "Few government agencies," Dr. Varmus writes, "have the reputation, promise, and political appeal of the NIH. Everyone, in government or the electorate, is worried about one disease, or all diseases." Dr. Varmus discusses how on occasion NIH experiences political pressures for disease-specific spending. A presidential request for special funding will be honored. But Dr. Varmus notes that NIH appropriations bills "rarely contain the kinds of 'earmarks' that direct an agency [of NIH] to spend its funds on a favored project." The NIH maintains and Congress supports a rigorous system of peer review for proposed research projects. The NIH works to guarantee that some research is going on in all important areas.

ACOUSTIC NEUROMA TODAY

The Wait and Scan Approach: A Treatment Choice with Benefits and Risks

By Dr.Samuel H.Selesnick

As we move further into the twenty first century, increasing numbers of acoustic neuromas are being diagnosed. Several reasons have led to this change. First, otolaryngologists and non-otolaryngologists have become more aware of the potential for acoustic neuroma diagnosis in patients presenting with suspicious symptoms. Secondly, even more sophisticated MRI technology is now available. Present day scanners can identify tumors as small as several millimeters in size. Given these factors it is not surprising that there is an increasing cohort of patients who have small tumors that may or may not be symptomatic. Oddly, however, there continues to be patients diagnosed with sizeable tumors. At this juncture there are three general forms of treatment for acoustic tumors: serial observation, radiosurgery in some form and microsurgery by one of three approaches. This article will focus on the first option, serial observation, also known as the wait and scan approach.

There are five important parameters to consider when deciding which treatment option is optimal for any given individual. The first is the patient's age, yet even age can be a nebulous concept. For example, an individual who is sixty years old, takes no medications, has no illnesses and exercises frequently is clearly different than an individual who is sixty years old, has diabetes, cardiac disease and multiple prior surgeries. The second factor to consider is the tumor size. The third is the tumor location, specifically regarding both the internal auditory canal and the cerebellopontine angle portion of tumor. For example, if the tumor penetrates deeply down the internal auditory canal, the ability to save hearing with surgery decreases. Further, where the tumor is located in the cerebellopontine angle is important in that if the tumor is centered more towards the back of the head there is less pressure on the facial nerve but more pressure on the brain and vice versa if the tumor is centered more forward. Greater brainstem compression makes radiosurgery options less appropriate. The fourth consideration is the patient's hearing, and the fifth is the presence of other symptoms, specifically dizziness, headache or worrisome symptoms consistent with hydrocephalus or increased pressure in the head. When factoring in all of these issues, there are certainly times when serial observation may be a reasonable treatment option.

Once the decision is made to begin with serial observation, the first question that arises is: what is being observed? In general, there are three observations being made. One is clinical symptomatology. Two is hearing results on audiogram and three is tumor size measurements. If there is an increase in the patient's symptoms such as worsened dizziness, if there is a drop in the patient's hearing on audiogram or if there is evidence of growth on MRI scan, intervention is usually strongly considered. But even identifying growth on MRI scan is not necessarily straight forward. There is, in principle, a standard agreement of which dimensions of the tumor should be measured on MRI. The sensitivity required for measurement is only available with MRI scans and not CAT scans. There may be some error involved in

measurement and this can be manifest in two ways: the first is the actual MRI software sensitivity, and the second is the reproducibility of the individual reading the scan. Given these variables, most would agree that 2 millimeters of growth can be reliably identified, and that less than 2 millimeters of growth of a tumor may not be consistently proven. While measurement of three dimensional tumor volume far more accurately represents the tumor size, nearly all clinicians use a two dimensional, linear set of measurements.

The one fundamental question that serial observation as a management strategy tries to define is: what is the growth rate of this particular acoustic neuroma? Simple logic and common sense are no replacements for scientific studies but it would make sense that regardless of what the tumor size is at diagnosis, the tumor size at the onset of growth is zero, so that the tumor had to have grown to attain whatever size is measured at the time of presentation. So every tumor has shown the capacity to grow, and, in fact, would be expected to grow. So, why would one expect a tumor to simply stop growing? Are acoustic neuromas fundamentally different than most other types of tumors in that they have a stop signal at some point, in some patients? Again this flies in the face of common sense but there are many studies that have proposed this. Further compounding the problem is the fact that some researchers have found that if a tumor is located only in the internal auditory canal it will be less likely to grow than if the tumor is in the internal auditory canal and in the cerebellopontine angle. Again this flies in the face of logic. Can a tumor biologically change when it extends from one location into an additional location? While it does not make sense, data has proven that this may be the case in some patients.

Examining the medical literature on this subject is confounding because there is considerable variation in the amount of follow up during the observation period. For example, there is far less meaning of a tumor not growing over a one-year period than over a ten-year period. In addition, it has been shown that there can be variability in terms of tumor growth since acoustic neuromas may not grow a small regular amount every day. Rather there can be periods of no growth, followed by active periods of growth. In other words, if a tumor does not grow for a certain period of time, this fact does not insure that it will continue to lie dormant with longer follow up. In addition, the literature is confounded by the fact that most people who have been observed are older. So the question arises: do tumors behave the same in younger people as they do in older individuals?

Perhaps the most thorough review of the literature on this topic was published in the journal *Otology and Neurotology* by Nikolopoulos and coworkers in 2010. This group examined 3,330 papers having to do with acoustic tumors and found that 41 papers met the inclusion criteria. In these 41 papers, over 3,000 subjects were evaluated. There was considerable variability in terms of the amount of follow up, ranging from six months to 65 months. On average, follow up was around one year. These authors found that tumor growth occurred anywhere from 6% of patients up to 73% of patients, so there was again tremendous variability. The median data point was 38% of patients showing growth. These authors concluded that tumor growth is unpredictable and can vary from no growth to rare cases of very rapid growth. These authors also note that most tumor growth occurs in the first 5 years but that tumors that are stable for long periods may also begin to grow. For this reason there is no clear indication when to discharge a patient from interval MRI scans.

Another key question involving the wait and scan approach is: does the wait and scan approach have risks associated with it? The answer is yes. The risks can be divided into short term and long term risks. Short term risks primarily revolve around the development of further hearing loss. In general, there is a parallel between tumor growth and hearing loss, but this is a very loose relationship which, for any individual, may or may not hold true. For example, there can be evidence of tumor growth on serial observation yet no change in the hearing. Sadly however, there can be no tumor growth, yet sudden deafness during the observation period. Hearing loss in this situation is thought to be due to compression of the blood vessels that supply the inner ear but, in fact, the actual cause is not entirely clear. This hearing loss would not improve with treatment.

Stangerup and colleagues from Denmark published an interesting article in the journal *Otology and Neurotology* in 2010 examining this issue. They used the standard four-level grading scale that is employed to classify hearing in patients with acoustic tumors. Grade A hearing is the highest level and is a range from normal to mildly impaired but "aidable". These researchers found that those patients starting with Grade A hearing lost their grade A hearing one quarter of the time after one year of follow up. By five years however, almost half lost their grade A hearing. So that this study makes clear that there is risk associated with the serial observation treatment modality, and that risk is primarily the potential loss of hearing over a relatively short term.

An additional question of great importance for the serial observation treatment modality is: how good are patients at remaining compliant with their follow up appointments? This question was addressed in an article published from the University of Pittsburg in 2010. They looked at 122 patients who were undergoing the wait and scan treatment modality. Their average age was 69 years. 43% of these patients failed to meet follow up criteria. This is an important finding because if the wait and scan method is employed, its success is predicated on a reliable patient returning for appropriate evaluations, so that tumor growth does not occur unrecognized.

To conclude, serial observation, or the wait and scan treatment method, is a management option with both benefits and risks. Clearly this form of treatment makes sense for an older patient with other health problems and who has a small tumor and poor hearing. But does this treatment option make sense for patients who are younger, in relatively good health and have moderate sized tumors? At this point that answer is unclear, but further research in this area will hopefully further clarify this issue in the future.

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Spring Meeting

"An Open Discussion: Everything You Ever Wanted to Know about Acoustic Neuroma"

Dr. Samuel H. Selesnick

Professor and Vice Chairman, Department of Otorhinolaryngology
Weill Cornell Medical College, NY Presbyterian Hospital
Department of Neurological Surgery, MSKCC

April 10, 2011

1-4 pm

Summit Medical Group

Lawrence Pavilion, One Diamond Hill Road

Berkeley Heights, NJ

Refreshments **Social Time**

Directions to Summit Medical Group, Berkeley Heights, NJ

The most direct way to Summit Medical Group is via Route 78.

From **Route 78 East**, take Exit 43 (Berkeley Heights/Watchung). Follow the exit road to the light at **Valley Road** and turn left onto Valley Road. Take Valley Road to the next **light** and turn left onto **Diamond Hill Road**. Follow Diamond Hill Rd to the light at **Mountain Avenue**. Go left on Mountain Ave for a short distance to the entrance to Summit Medical Group on the left. You will see Lawrence Pavilion and parking straight ahead as you enter. In the Lawrence Pavilion lobby, take the elevator down to 1R, the Cafe/Conference area (Note: there is another entrance to Summit Medical Group on the left just before the Mountain Avenue light. Follow the signs for Lawrence Pavilion/Parking Lots 1&2.

From **Route 78 West**, take Exit 43 (New Providence/Berkeley Heights). Bear right onto Diamond Hill Rd. Follow the instructions above for Summit Medical Group, Lawrence Pavilion.

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