



ANA/NJ Newsletter
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Acoustic Neuroma Association

of New Jersey

A Non-Profit Corporation

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ANA/NJ Mini-Conference
April 22, 2018

Our Mini-Conference held at the JFK Medical Center in Edison, NJ, on April 22, 2018, was well attended and rated as highly successful by those who completed the evaluation form. This was the third mini-conference hosted for us by JFK. It was the first held jointly with the NYCity Acoustic Neuroma Support Group. Miranda Sacharin spoke for the NYCity group during the 'Welcome' beginning the conference. Unusual greeters this day were two very friendly 'Hearing Dogs,' arranged with our thanks by Melanie Riordan. Present throughout the day was a representative for Oticon Medical, Alison Sabbar, who demonstrated the new Ponto 3 bone-anchored hearing system.

The morning session of the conference featured a distinguished Doctors' Panel for the topic "Wait-and-Scan, Partial Removals, Fractionated Radiation: Exploring the Pros and Cons." Dr. Jed Kwartler (Summit Medical Group) was the moderator. The other members of the panel were Dr. Christopher



Dr. Kwartler

Farrell (Thomas Jefferson University), Dr. Joseph Landolfi (JFK Medical Center), Dr. John G. Golfinos (NYU Langone Medical Center) and Dr. Philip Stieg (Weill Cornell), as shown from left to right at the speakers' table in the photo below.



Dr. Kwartler began with a brief slide-based review of basic factors influencing treatment decisions for acoustic neuroma: e.g., presenting symptoms such as hearing loss, tinnitus and vertigo; tumor size and location; patient age; concerns about avoiding damage to the 7th cranial facial nerve. The final slide in this review showed a One-Way sign with a large arrow pointing to *Surgery!* But this was only to remind us of what used to be the single, standard treatment for acoustic neuromas. Today's patients and their doctors have a wide range of treatment choices, and increasingly with emphasis on preserving quality of life.

The panelists discussed treatment choices for several 'typical' AN patients. Sample MRIs to initiate discussion were projected on screen for everyone to see. As it proved, surgery was recommended for the first case, a 38 year old man with a very large tumor. Hearing in the affected ear would be lost by the surgery; the main concern was to preserve facial nerve function. With this concern in mind, should complete resection of the tumor be attempted? Or wouldn't it be safer to do a partial, subtotal removal? But if subtotal, how little 'residual' tumor could be left? Was it even possible to determine this beforehand? There was much to and fro among the panelists about post-surgery outcomes. And then too, since the patient's hearing in the affected ear would be lost, could a Baha hearing device be implanted

during the surgery? Perhaps it would be better to wait and do this separately later? To implant later would be a minor outpatient procedure.

A second case was that of a 38 year old woman with a small tumor, mild hearing loss, tinnitus and vertigo. ‘Wait-and-Scan’ was one recommendation because of the small size of the tumor, and provided the vertigo was tolerable. But there was discussion of the need for more data about long-term outcomes for patients electing this option. Middle Fossa surgery was also recommended: that is, remove the tumor while it was still small, and thereby help remedy the vertigo, and let this young woman get on with her life. Here, for a time, the conversation among the doctors became highly professional and technical. It brought to mind a question of whether or not so-called “shared decision-making” is possible in all cases among patients and doctors?¹

Radiation treatment was not discussed as a possibility for this second case. Types of radiation available, for example, would be single session treatment by Gamma Knife or Linac (linear accelerator); 3-5 session hypofractionation by CyberKnife (Linac); or 30 session hyperfractionation by Linac or Proton Beam. Just recently (2017), a new “Edge” (Linac) radiation system by Varian (Palo Alto, CA) was announced. The Varian brochure states: “A growing body of clinical evidence is demonstrating the benefits of delivering high doses in a small number of fractions. . . Stereotactic Radiosurgery (SRS) is progressing to treat targets previously not considered candidates for high-dose, hypofractionated delivery.”² But the panelists did not get involved comparing radiation systems or the values of different levels of dose fractionation. They expressed concern mainly about dangers of radiation of large tumors growing near the cochlea or into the brain stem. It was noted that these critical areas cannot sustain high doses of radiation.

Following a good lunch, the conference continued with the two scheduled afternoon sessions. The first session dealt with “Balance Issues and Vestibular Therapy.” The two presenters were Dr. Michael Rosenberg, director of Neuro-Ophthalmology at JFK, and Dina Leyden, a specialist in physical therapy at the Summit Medical Group (Berkeley Heights). Dr. Rosenberg began the session with a slide presentation that helped to explain and distinguish between dizziness problems such as light-headedness, imbalance and vertigo. He discussed what he likes to call the “Visual Vestibular Mismatch Syndrome” that develops when coordination between vision and vestibular functioning goes awry. For the lively Q&A that followed, Dr. Rosenberg and Dina Leyden team-answered very nicely a variety of interesting questions related to individual experiences with balance problems and therapy.



Dr. Rosenberg

The second afternoon session dealt with “Tinnitus Issues and Hearing Assistive Devices.” It was presented by Dr. Virginia Toth, who is the manager of audiology programs at JFK. Dr. Toth gave an ex-

¹ See “Critical Decisions (A Look at Shared Decision-Making),” in the ANA/NJ Newsletter (June 2013).

² See Edge Radiosurgery System, www.Varian.com.



cellent description of the nature of single-sided deafness (SSD) and its problems. She cautioned that for many people “One ear is really not good enough.” She reviewed the variety of assistive hearing devices available for SSD today.

As for tinnitus, no recently discovered ‘cure’ could be brought to our attention. But for help ‘getting used to it’, Dr. Toth recommended proven habituation systems such as Tinnitus Retraining Therapy (TRT) or Neuromonics, and perhaps trying a hearing aid. Some people have tried acupuncture, hypnosis or B vitamins, but there is little evidence that these work. She warned definitely against using ear muffs or ear plugs. Among the things that can be helpful, Dr. Toth recommended: investigating coping strategies; participating in a tinnitus support group; avoiding irritants and stress; trying soft music to mask the tinnitus; and limiting alcohol consumption.³

MRI Advisory

The Gold Standard for diagnosis of acoustic neuroma is magnetic resonance imaging (MRI). Using ‘thin slice’ sectioning and gadolinium dye contrast helps to identify even the smallest of tumors. This is important especially for spotting rare cases of bilateral ANs (NF2) where one tumor is often very small.

Acoustic neuroma patients need to be aware that the strong magnetic fields generated by MRI may not be safe for implanted medical or electronic devices and other metal objects in the body. There’s a long list of things that MRI technologists need to know about and assess for potential risk before treatment can begin. The list includes cardiac pacemakers, stents for AAA (abdominal aortic aneurysm), cochlear implants, vascular clips, insulin pumps, gastric reflux devices, ‘Triggerfish’ contact lenses, shrapnel. Dr. Frank Shellock maintains an online detailed list at www.MRISafety.com. A website by the University of California at San Francisco (UCSF) also provides safety guidelines.

Immunotherapy Advisory

An article in the *Washington Post* (January 2, 2018), by health and medicine writer Laurie McGinley, provides a helpful ‘heads up’ for patients treated by newly introduced immunotherapy drugs. “The new therapies,” she writes, “offer a tantalizing chance for survival for patients with advanced melanoma and hard-to-treat cancers of the bladder, kidney and lung.” However, a problem that has emerged for immunotherapy patients is that many physicians outside of major academic medical centers are not yet up to speed for recognizing all the side effects of the new drugs. McGinley reports, for example, on the case of a woman treated by her ophthalmologist with laser surgery for a supposedly torn retina. But the black specks she was seeing in her eyes were actually a side effect of the immunotherapy drug she was on for advanced lung cancer. Or again: “One expert said a patient who had an immunotherapy-caused rash was

³ For a list of tinnitus support groups and phone ‘help network’ volunteers in NJ, see the Sept 2014 issue of the Newsletter.

diagnosed in the emergency room with a skin infection and given an antibiotic, which was useless. Another said ER doctors sometimes tell patients with diarrhea to take medications such as Imodium, whereas actually steroids might be needed to calm the immune system.” Immunotherapy patients may at times contribute to the confusion by telling doctors, erroneously, that they are on chemotherapy, which has a completely different side-effect profile. McGinley notes that side effects occur in 15 to 70 percent of immunotherapy patients depending on which drug is used. Most problems are usually mild; dangerous ones are extremely rare. But meantime, steps are being taken to narrow the knowledge gap on immunotherapy. “Professional groups such as the Society for Immunotherapy of Cancer and the Association of Community Cancer Centers are writing recommendations on side effects and conducting programs for doctors and nurses.” “The last thing you want to do is scare people away from lifesaving treatments,” states Jeffrey Bluestone, immunologist at UCSF.

Guidelines for the Treatment of Vestibular Schwannomas (VS)

For any long-term endeavor, there comes a time to step back and review what’s been done, what’s working (or not), and make adjustments and shape plans for future efforts. Two major American medical associations, the Congress of Neurological Surgeons (CNS) and the American Association of Neurological Surgeons (AANS), have determined that this time has come for the diagnosis, management and treatment of vestibular schwannomas. The Joint Guidelines Committee of the associations states: ⁴

“Although much information has been garnered from current large retrospective clinical data series, it appears we have maximized the information to be gained by simply increasing the numbers of patients treated in the same fashion and adding to the duration of follow-up.

“A useful starting point to improving the current knowledgebase is to define the benchmarks of our current knowledge regarding VS management using evidence-based techniques in order to allow meaningful points of departure for future scientific and clinical research.

For this purpose, committees of experts in the field in the USA were organized to review and assess the voluminous medical literature on VS with emphasis on the period 1990 through 2014. Doctors from the Icahn School of Medicine at Mount Sinai in our area participated. Questions for assessment of the evidence were designed and writing groups for final reports for 9 major topics were formed, including: Audiologic Screening, Nerve Monitoring, Imaging/Wait-and-Scan, Surgical Resection, Radiosurgery and Radiotherapy, Hearing Preservation, Emerging Therapies. The following are “Summary of Results” for three of these nine final reports. The topics selected are ones dealt with by the Doctors’ Panel at the April 22 mini-conference at JFK.

Imaging/Wait-and Scan

Summary of Results. 25 full-text articles were reviewed; 15 were included as evidence. “Class 3 evidence [mainly retrospective] supports the conclusion that about two-thirds of patients with VSs may not exhibit measureable growth, while one-third demonstrated growth, defined as either any increase in size or a

⁴ Congress of Neurological Surgeons Systematic Review and Evidence-Based Guidelines on the Treatment of Adults with Vestibular Schwannomas: Introduction and Methods (*Neurosurgery*, February, 2018), www.cns.org/guidelines.

change in diameter. Intrameatal [in the canal] tumors are less likely to grow. While large literature surveys suggest average growth rates of 1.2 to 1.9 mm/year, separate analysis of actively growing tumors reveal faster rates. Early growth may predict future growth; however, late growth after 5 years of quiescence may occur. An MRI 6 months after tumor discovery may identify tumors likely to continue growing; otherwise, scans may be obtained annually for 5 years, and scan intervals should be lengthened if no growth is detected.”

Partial Removals

Summary of Results. 17 full-text articles were reviewed; 13 (all retrospective) were included as evidence. “There is insufficient evidence to support [that] subtotal resection followed by SRS [stereotactic radiosurgery] provides comparable hearing and [facial nerve] preservation to patients who undergo a complete surgical resection. . . . When a VS is treated with subtotal resection followed by radiosurgery either primarily or because of tumor remnant growth, tumor control rates are consistently 93% to 96% with >90% of patients maintaining normal or near normal facial function. This tumor control rate is similar to that of series on gross total resection. . . . Future studies directly comparing gross total resection to subtotal resection plus radiosurgery [are needed]. In addition, it would be highly valuable for a ‘lowest acceptable’ percentage of surgical resection to be determined that could still be followed by radiosurgery with comparable results to gross total resection.”

Fractionated Radiation

Summary of Results. 202 studies were reviewed; 15 (those using MRI for radiographic follow-up) were included as evidence. “As there is no difference in radiographic control and clinical outcome using single or multiple fractions, no recommendation can be given. . . . High rates of tumor control (i.e., generally >90%) were afforded by single fraction, hypofractionated, and traditional hyperfractionated schemes. As compared to [the high] tumor control, lower rates of hearing preservation were reported, and hearing preservation rates lessened with longer follow-up assessment and for larger tumors. Rigorous evidence supporting a single fraction approach compared to others for preserving hearing seems lacking. Further clinical investigation will be required to determine an optimal fractionation approach for VS patients. However, a one-size-fits-all approach is not likely to be ascertained, and an optimal approach may vary based upon various factors, including tumor size (or volume). There is no recommendation that can be given based on the available data regarding the schemes of the fractionation and which patient population will benefit from that.”

As will be noted, these ‘Summary of Results’ sections of the reports are very cautious. Each is based on evidence provided from a select number of clinical studies determined to be unbiased and also in other ways decided acceptable for inclusion in the final report. Data from the studies selected for inclusion is presented in some detail just before the summary statements. Full bibliographies are provided for the medical journal articles that were reviewed.

These “Evidence-based Guidelines for the Treatment of Vestibular Schwannomas” should prove useful for new VS patients wanting to learn more about the treatment options they are considering. Reading the section dealing with Wait-and-Scan, for example, would help patients to ask informed questions about this option when meeting with their treating physician.⁵

The ‘Evidence-based–Guidelines’ can be heavy reading. After all, they were prepared to the attention mainly of doctors specializing in the treatment of VS. New patients may prefer to look first at more

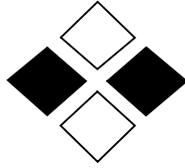
⁵ See the Newsletter (June 2013) for “Critical Decisions: A Look at Shared Decision Making,” a review of Peter Ubel’s book, *Critical Decisions: How You and Your Doctor Can Make the Right Medical Choices Together* (2012).

introductory-type decision aids, such as Dr. Ted McRackan's recent webinar for ANAUSA entitled "Evolution in the Management of Acoustic Neuroma: Developing an Evidence-based Treatment Approach" (March 21, 2018).⁶ Dr. McRackan is Assistant Professor in the Department of Otolaryngology at the Medical University of South Carolina. His webinar provides a helpful overview and orientation for the treatment of VS. The presentation is clear and well-paced with excellent graphics and up-to-date data. He begins with an interesting review of the early history of the treatment of VS during the 1920s when subtotal vs. total resection was definitely a key issue. He comments on today's trend toward subtotal removals as a sort of revival of the issue. The increase in Wait-and-Scan is discussed: he notes how he recommends this conservative management for over 50% of his patients. Gamma Knife single-session radiosurgery is looked at carefully, but he does not touch on the pros and cons of fractionated radiotherapy treatments.

Notices

- An important reminder! ANA/NJ continues to welcome volunteers to serve on the executive board for planning and helping with scheduled meetings and special services for acoustic neuroma patients and caregivers. Please call Wilma Ruskin at 609-510-9093 or email ananjinc@aol.com.
- Past issues of newsletters by ANA/NJ and ANAUSA are available online and are a valuable source for articles written by doctors specializing in the diagnosis and treatment of acoustic neuroma. For example, the ANA/NJ Newsletter (available 2004 to date) has important articles about 'Wait-and-Scan' (April 2011, Dr. Selesnick) and 'CyberKnife' (June 2013, Dr. Lipani). The ANA/NJ website has an index of major articles. ANAUSA's *Notes* (available 2014 to date) provides featured medical articles about 'Radiosurgery' (June 2014, Dr. Chang) and 'Subtotal Removals' (June 2016, Dr. Montfared).
- Currently there is no FDA-approved drug for preventing the growth of vestibular schwannomas. The search goes on. Jessica Sagers, Konstantine Stankovic and other researchers at the Massachusetts Eye & Ear Infirmary, Harvard Medical School, have recently announced that they are "cautiously optimistic about the therapeutic potential of [the drug] 'mifepristone' for patients with vestibular schwannomas, either from NF2 or those arising sporadically." A phase 2 clinical trial is being planned. (See "Computational Repositioning and Preclinical Validation of Mifepristone for Human Vestibular Schwannoma," *Scientific Reports*, vol 8 (2018).
- A first meeting of the Facial Paralysis Support Group of Greater New York, hosted by the NYU Langone Facial Paralysis & Reanimation Center, was held August 8, 2018, at the NYU Langone Medical Center, 435 E. 30th Street, New York, NY.
- 'Signia' is the new name for CROS/BiCROS hearing aid systems manufactured by Siemens, the German company headquartered in Berlin and Munich. The company recommends its new Signia CROS and BiCROS systems for people with hearing loss in one ear. The BiCROS solution is for people who also have some hearing impairment in their 'good' ear.

⁶ Go to www.ANAUSA, Webinar Library.



Fall Meeting

“Caring and Sharing”

Open Meeting for Acoustic Neuroma Patients, Family & Friends

Sunday, October 14, 2018

1:00 – 4:00 pm

Holman Conference Center at the Virtua Voorhees Hospital

100 Bowman Drive

Voorhees, New Jersey 08043



Group Discussion

Q&A

Refreshments

Social Time

Directions:

Virtua Voorhees Hospital in Voorhees, NJ, is located to the SE of Camden/Cherry Hill and a bit north of the towns of Berlin and Winslow.

From the north, take I-295 south to Exit 36A and merge onto Route 73 South. Turn rt off Route 73 onto Dutchtown Rd, and then rt onto Howe Blvd for less than 1 mile to Bowman Dr and the Hospital. **From the south** go north on I-295 to the interchange for Route 73 south; or (e.g., from Winslow) take Route 73 north to Cedar Hill Dr, which becomes Bowman Dr at the roundabout. The Hospital is about 0.1 miles past Howe Blvd.

At the Hospital, **park in Lot B**. Enter at lobby B and ask for directions to the Holman Conference Center and the ANA/NJ meeting. Look for ANA/NJ signs.

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