ANA NJ
ACOUSTIC NEUROMA ASSOCIATION of NEW JERSEY

ANA/NJ Newsletter
Vol. XV, No. 1, September 2015

The 20th Anniversary chapter meeting, April 26, 2015, was held at the University Medical Center of Princeton, One Plainsboro Road, Plainsboro, NJ. There was excellent attendance of 42 AN patients and care givers. The celebration of ANA/NJ's founding twenty years ago may have taken some new AN patients by surprise, but everyone seemed to participate happily and enjoy the cake and coffee. We wish to extend our thanks to staff of the Princeton Center for their special assistance throughout the meeting.

Wilma Ruskin welcomed everyone and Kathy Cecere introduced the five panelists for the afternoon session on "Living with Acoustic Neuroma: A Peer Panel Discussion." The panelists, who are listed in the ANA/NJ Directory, each spoke briefly about their special experiences with AN.

Andy Gregg, who was one of the first founder-members of ANA/NJ, began by describing how he has been Wait-and-Watch for a 1.7cm tumor for 22 years; there was only one worrisome time when an MRI reading 'seemed' to indicate growth – but it was a false alarm. Mike Illuzzi spoke next about his Wait-and-Watch experience with a 1.0cm tumor for the past 8 years; he reported how he has adopted a healthier lifestyle that has helped reduce his migraines, and he has used Neuromonics therapy to moderate his long-standing tinnitus.[1]

The next three panelists reported on their experiences with retrosigmoid surgery: Cindy Rivkin, for a 7mm tumor (2014); Ken Sacharin, for a 3.2cm tumor (2009); and Elizabeth Snyder, for a 2cm tumor (2001).

Elizabeth's tumor was discovered incidentally during the course of an already sufficiently troublesome bout with Lyme disease. She reported on her post-surgery problems of facial paralysis (now mostly resolved) and single-sided deafness (SSD) which she has countered by having a Baha bone-anchored hearing device. She described having difficulty adjusting to the Baha.

Ken's was the largest tumor and he reported on a wide variety of unanticipated postop conditions. He was prepared for SSD and some facial paralysis, but has also confronted really 'annoying' problems with

[1] See Andy Gregg, "A Case of 'Wait-and-See'," *ANA/NJ Newsletter* (May 1998); Kristin Ingersoll, "Mike Illuzzi in the Spotlight," *ANA/NJ Newsletter* (April 2009), available online at www.ananj.org.

eyelid closure, loss of taste and smell, continual runny nose, nostril narrowing interfering with breathing, balance issues, headaches … and cognitive deficits (e.g., poor word recognition, memory lapse).[2]

Cindy's was the smallest tumor, surgically treated after 3 years of Wait-and-Watch when the tumor began to show growth. The decision to intervene surgically was difficult since Cindy's tumor ear was also her only hearing ear. Unfortunately, the surgery resulted in deafness in that ear as well. Cindy decided her only option for hearing was to have a cochlear implant (CI). This was placed about a year ago on her surgery side. Cindy commented after the meeting that the CI is "working wonderfully," even though "it's definitely not 'regular' hearing, and it's something that takes time to get used to. . . The good thing is that even in the year that I've had it, there have been unbelievable advances (like bluetooth for my cell phone) that make life easier with it."

It's good to learn of the availability and success of the CI for patients who have lost hearing in both ears. Discussion at the meeting about coping with hearing problems, however, focused rather on devices for SSD: in particular, the Baha bone conduction system (minor implant surgery, external abutment, detachable sound processor) and the Widex Cros and Phonak Cros systems, both of which entail no surgery but do require having a device in each ear -- a microphone/transmitter for the deaf ear, and a receiver/amplifier for the good ear.[3]

The response to the evaluation form distributed at the meeting showed that at least 10 attendees were Wait-and-Watch. The number of AN patients selecting this treatment option has grown over the years. MRI imaging has been able to identify many more small ANs that can be 'managed conservatively' by Wait-and-Watch, and doctors are now more likely to call attention to the option. Most notably, in Denmark "more than 90% of ANs. . . are allocated [by the socialized healthcare system] to wait and scan if they are less than 2cm." The quote is from Dr. Michael Link's review of the valuable Danish research contribution to our knowledge of tumor growth behavior and the consequent pros and cons of Wait-and-Watch.[4]

My Cochlear Implant Experience

by

Cindy Rivkin

Today, social media is the way to go. You can't live today without knowing about Google, Face-book and Twitter. I think they're wonderful sources of information – when they're used correctly. You have to sort through the information in terms of what to keep and what to throw away, and then also know when to stop looking.

When I was told I had an acoustic neuroma, I hopped online to find out what was going on with my brain. I immediately joined an AN group on Facebook and became incredibly well-informed. I spoke to different people and learned what they had gone through, what questions they asked, and what results they had. I chose to do Wait-and-Watch, and when my tumor began to show growth after 3 years, I signed on to Facebook again to inquire about surgeries. Deciding on surgery was an incredibly difficult decision for me because I had already lost my hearing in one ear due to a common cold, and now I was facing possible loss on the other side. (It's not very reassuring either when brain surgeons in our Tristate area and all the way out to California tell you that you're a rare case and they're not sure what to tell you!)

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[2] See Miranda Warren Sacharin, "A Caregiver's Story: Long-Term Complications Following Treatment, But Eventual Healing," ANA *Notes* (March 2014).

[3] See "Thinking About Hearing Devices for SSD," in ANA/NJ Newsletter (March 2013); also *Hearing Loss Rehabilitation for Acoustic Neuroma Patients* (ANA Booklet, revised January 2013).

[4] Michael Link, M.D. (Mayo Clinic), "Comparison of Surgery, Radiation and Natural History," ANA *Notes* (September 2011), available online at www.ANAUSA.org., Members Section.

Anyway, after an exceedingly long and painful information-gathering 'era,' I opted for retrosigmoid surgery. Unfortunately, I lost all of my remaining hearing.

I was left in total silence for about six weeks. It was horrible. The only 'light at the end of the tunnel' was to learn that I was a candidate for a cochlear implant. I plunged online to look into the technological websites for information. Along the way, I found that Facebook also had a wonderful site for CI (cochlear implant) Users. So I became a member of two Facebook groups, the AN and the CI.

I also found a useful 'Fact Sheet"[5] for cochlear implants issued by the National Institute on Deafness (NIH), which had this to say about the hearing experience with CI:

> *A cochlear implant is very different from a hearing aid. Hearing aids amplify sounds so they may be detected by damaged ears. Cochlear implants bypass damaged portions of the ear and directly stimulate the auditory nerve. Signals generated by the implant are sent by way of the auditory nerve to the brain, which recognizes the signals as sound. Hearing through a cochlear implant is different from normal hearing and takes time to learn or relearn. However, it allows many people to recognize warning signals, understand other sounds in the environment, and enjoy a con-versation in person or by telephone.*

That being said, I had my work cut out for myself. I called in the cavalry. I ordered a book by Arlene Romoff about her experience with the CI. I also sought out Facebook members, and I spoke to everyone I knew, hoping someone would at least know someone who had a CI (apparently very rare!). I didn't know what to expect for my hearing once my cochlear implant would be activated, and I was terrified.

I still wonder if all the research I did helped me, because I was really surprised when my CI was finally activated. I guess I had totally mixed emotions. I was able to hear my Mom's voice immediately. I seemed to struggle with my husband's (I guess there could be a lot of reasons for that!) I seemed to have an easier time with women's voices than men's. I couldn't tell my son and daughter apart – both teenagers. The first time I heard popcorn popping, I dove under the couch for cover, thinking we were being attacked. When something on the stove sizzled, it sounded like I was in the pot itself. Yet, when the doorbell rang, I didn't hear it.

Fine tunings are done at my audiologist's office. They're done at regular intervals for about a year after activation of the implant, and then they're continued annually. With these devices, nothing can done by yourself; it all works with the help of the audiologist. There are different programs on the CI to go through before more 'in depth' adjustments can be made. It's an experience that basically takes over your life, because now every ounce of energy is being put into actually listening. This is something you've always been able to do without thinking – something that comes naturally. Now it doesn't work that way. People with normal hearing are hearing and processing and forming responses naturally. This is not the case with me. I have to put a lot of energy and concentration into actually listening. It's especially hard in group situations. For a normal hearing person, the brain automatically knows what background noise to ignore, but my brain has to be retrained to do that. If I'm in a crowded restaurant with music playing, people talking, and sirens going by, my head can explode at any moment.

I also find that I should no longer say "Excuse me," or something like that, when I don't hear part of what someone has said. People usually respond by repeating everything, whereas what I really need is a restatement using different words. With a CI, a lot of words sound alike. For example, "that may be an amusing story" could sound to me like "that baby in a music store." And louder is not better for me. I heard you speaking, I just didn't understand what you said. Also, new voices are a challenge; I find it takes a while to get used to a new voice.

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[5] NIDCD Fact Sheet, www.nidcd.nih.gov.

I've now been implanted on the left ear for 16 months, and on the right ear for 9 months. My entire life has changed since my first surgery in February 2014. Things have gotten better gradually with the two CIs. No, it's not normal hearing, but I don't find it robotic-sounding, and people no longer sound like Mickey Mouse or Alvin and the Chipmunks. But every day is a struggle in a different way. It's tiring to hear, strange as that may sound. Sometimes it just gets overwhelming and I need to take time off from the world. I suppose, though, that everyone has that feeling now and then.

You come across all kinds of people in this world. Some are patient and take the time to look directly at you when they speak, some need to be asked but respond kindly to your request, and some just can't be bothered. It's been my experience that most people are patient and willing to take their time. But it's still hard to be 'that person' with the hearing problem.

Thinking about 'Residual Tumor'

Doctors are using the term 'residual tumor' to refer to remnants of an acoustic neuroma left intentionally during surgery to help preserve good facial nerve function for the patient. In other words, residual tumor is not tumor that was missed or could not be seen during a surgery where the objective was to "get it all," as in a gross total removal (GTR). Rather, the remnant is left purposefully as part of a facial nerve-sparing surgery – as with a near total removal (NTR) or subtotal removal (STR) -- in line with today's increased concern for postop quality of life.

The following two published medical journal articles (PubMed abstracts) reported on outcomes for facial nerve-sparing surgeries:

- M.S.Schwartz et al (House Clinic, Los Angeles), "Evaluation of the Increased Use of Partial Resection of Large Vestibular Schwannomas: Facial Nerve Outcomes and Recurrence/Regrowth Rates," *Otol Neurotol*, Vol 34 (October 2013).

  This is a report on 400 acoustic neuroma patients with ANs of 2.5cm or greater who underwent translabyrinthine surgery. There were 325 gross total removals (GTR), 44 near total removals (NTR) and 31 subtotal removals (STR).

  At 1-year follow-up, higher rates of facial nerve preservation were seen for NTR (97%) and STR (96%) compared with GTR (77%). But for follow-ups averaging ca. 3-5 years, higher rates of tumor regrowth were seen for NTR (21%) and STR (22%) compared with GTR (3%). Need for further treatment, although infrequent, was at a higher rate for NTR (2%) and STR (10%) compared with GTR (0%).

  The report concluded that near total and subtotal removals for large tumors are viable treatment options to maintain facial nerve function. Long-term follow-up is recommended to check for regrowth of the residual after NTR and STR.

- Z.Chen et al (Shanghai JT Univ Hospital), "The Behavior of Residual Tumors and Facial Nerve Outcomes after Incomplete Excision of Vestibular Schwannomas," *Jour Neurosurg*, Vol 120 (June 2014). This is a report on 111 patients who underwent translabyrinthine surgery. Tumor sizes were not stated. There were 73 near total removals (NTR) and 38 subtotal removals (STR). The residual tumor was left on the facial nerve alone in 62 patients, on facial nerve and vessels in 2, and on facial nerve and brainstem alone in 25.All had follow-up of 7-10 yrs as wait-and-scan patients.

  For the 105 patients who began with normal facial nerve function, postoperative function was House-Brackmann Grade I-II for 51, Grade III for 34, and Grade IV-VI for 20.

Seven patients who had STR showed residual tumor regrowth. No NTR patient exhibited regrowth, while having almost the same result in terms of facial nerve function. The report recommended that NTR and STR patients undergo follow-up MRIs for at least 7-10 years.

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These two reports show that cases of regrowth of residual tumor following facial nerve-sparing surgery are a possibility. That is, some residuals are likely to regrow and become in effect recurrent tumors. The following report that comes to us from Copenhagen, Denmark, indicates that tumor remnant size is probably the greatest determinant of regrowth:

• C.H.Hahn et al, "Residual Tumor after Vestibular Schwannoma Surgery," *Jour Laryngol Otol*, Vol 127 (June 2013).

This is a report on 1,143 patients who underwent translabyrinthine surgery during 1976-2008. There were 978 gross total removals (GTR), 140 near total (NTR) and 25 subtotal (STR). The mean preop size of tumors having GTR was significantly smaller than for NTR and STR. At 1-year follow-up, 65% of small tumor remnants and 11% of large tumor remnants were not visible on MRI. Revision surgery was performed for 14 patients (1.2%), of whom 2 had received GTR, 5 NTR and 6 STR, initially. The report concluded: few patients with a small residual will require revision surgery or secondary radiotherapy. Most residual tumors disappear spontaneously, probably due to de- vascularisation.

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The above report speaks of 'revision surgery' for dealing with regrowths of residual tumor. Another study presents the observation: "Revision surgery is usually considerably more difficult than primary surgery, with a higher complication rate, particularly with regard to the facial nerve."[6] The procedure now widely used for treatment of residuals is stereotactic radiosurgery (Gamma Knife). One important recent report on this procedure is:

• R.Haque, M. Sisti et al (Columbia Univ), "Efficacy of Facial Nerve-Sparing Approach in Patients with Vestibular Schwannoma," *Journal Neurosurg*, Vol 115 (Nov 2011).

For preserving good facial nerve function in patients with vestibular schwannoma, the report recommends a combination of facial nerve-sparing microsurgery and, as needed, Gamma Knife radiosurgery for residual tumor.

151 patients underwent microsurgery, either gross total removal (GTR) or subtotal removal (STR), with excellent facial nerve preservation rates of 96% and 97%, respectively.

However, 20 of the 151 microsurgery patients (13.2%) required postoperative re-treatment for significant growth of residual tumor. Subtotal removal (STR) patients carried a higher risk of requiring re-treatment, compared with gross total (GTR). In the 20 patients, who were re-treated with Gamma Knife (dose to the tumor margin was 12Gy), the rate of facial nerve preservation was 95%. The report concluded: "A facial nerve-sparing resection followed by [Gamma Knife radiosurgery] for any significant regrowth provides excellent facial nerve preservation rates."

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Dr. Michael Link (Mayo Clinic) has reported in ANA *Notes* (March 2009) on "Subtotal Tumor Removal Followed by Stereotactic Radiosurgery." In this article he discussed reasons why a subtotal removal might be appropriate (e.g., pre-surgery facial nerve weakness; patient age of >70 years). He also addressed the issue of when to proceed with the radiosurgery treatment. He wrote that "we often elect to just follow a small remnant. However, if there is evidence that the tumor was actively growing on serial images prior to surgery we often proceed directly to radiosurgery after a 3-12 month recovery period after subtotal surgical removal. We prefer to let any facial weakness that might ensue after surgery recover as much as possible. . . Fortunately, we haven't seen any progressive facial weakness after radiosurgery for tumor remnants. . . ."

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[6] S.R.Freeman et al (Manchester, UK), "Revision Surgery for Residual or Recurrent Vestibular Schwannoma," *Otol Neurotology*, Vol 28 (December, 2007).

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Dr. Samuel Selesnick (Weill Cornell) has advised us that multiple-session fractionated radiotherapy is also being used for treatment of residual tumor with promising results (MSKCC, Harvard), although there are as yet no published reports on outcomes for facial nerve preservation. Dr. Fred Barker (Mass General) commented on his preference for fractionated radiation for residuals at his session on "Recurrent Tumors" at ANA's 2013 Symposium.[7]

As doctors continue using less aggressive treatments of acoustic neuroma for purposes of facial nerve-sparing (or hearing preservation), cases of residual tumor will increase. Long-term data on re-treatment modalities and their outcomes will be needed.

"The BRAIN Initiative"

> ▪ *With nearly 100 billion neurons and 100 trillion connections, the human brain remains one of the great mysteries in science and one of the greatest challenges in medicine. . . Despite the many advances in neuroscience in recent years, the underlying causes of most neurological and psychiatric conditions remain largely unknown, due to the vast complexity of the human brain.*
>
> (National Institutes of Health, June 2014)

Knowledge of how the brain actually works has eluded us for centuries. It's a fundamental gap in our knowledge that is today holding up the necessary full understanding and means of effective treatment of major neurological disorders such as Alzheimer's and Parkinson's. For acoustic neuroma patients, attempts to deal definitively with problems of tinnitus have been frustrated. We are now able to map and image neural circuits in the brain, but the crucial next step is still missing: "We must understand how these circuits work to capture the full sense of what is happening in the healthy brain – and what goes awry in disease."

Launched by the Obama administration on April 2, 2013, The BRAIN Initiative (Brain Research through Advancing Innovative Neurotechnologies) has the potential to end the centuries-long search to discover what has been called 'the language of the brain.' For learning about this ambitious project, Helen Shen's article "Neurotechnology: Brain Storm," *Nature* (Nov 6, 2013) is an informative examination of its inception in 2013; and "The New Century of the Brain," *Scientific American,* 310 (Feb 2014), by Rafael Yuste (Columbia Univ) & George M. Church (Harvard Univ), picks up the story a year later. See also Sara Reardon, "Ambitious Plans for BRAIN Project Unveiled," *Nature* (June 6, 2014). For basic information and current updates, consult the website of the National Institutes of Health (search BRAIN Initiative).

Zebrafish

[7] Available at ANAUSA.org (Members Section).

The Zebrafish (*Zebra danio*, a popular aquarium starter fish) will have its important role to play in the BRAIN Initiative. Some people may be a bit skeptical about scientists using fish as research subjects, but this small freshwater striped minnow, which has a brain similar to our own, and is conveniently transparent in the larval stage, and breeds and develops rapidly, has been a favorite neurobiologists' study-organism for whole brain imaging. The little fish's 100,000 nerve cells (neurons) have just recently been made to light up showing every blip of brain activity.[8] Studies of neuronal activity in zebrafish (100,000 neurons) are an important step forward in the development of new imaging technologies and varied sensors that can be applied for mice (75 million neurons), Rhesus monkeys (6 billion) and ultimately humans (100 billion

Notices

● A reminder that ANA/NJ continues its invitation for new board members. If you are interested in helping out with the association's support group activity, please call Wilma Ruskin (609-799-4442) and chat with her about joining the board.

● Sonitus Medical of San Mateo, CA, the manufacturer of the SoundBite hearing system, filed for bankruptcy January 15, 2015. This followed a decision by the Centers of Medicare & Medicaid Services (CMS) to "restrict coverage of novel solutions for hearing loss."

● Would you like to participate in a long-term mailed questionnaires survey by the Mayo Clinic to investigate how the diagnosis and treatment of acoustic neuroma may affect quality of life? Go to www.ANAUSA.org for information about the study and how to get started.

● Annual reports for 2012-14 are available on the ANAUSA website (click on 'About ANA'). The report for 2014 shows: 61 active support groups nationwide; 128 meetings held, involving an overall total of 2,007 patients; distribution of 19,321 copies of the quarterly *Notes* newsletter; distribution of 1,153 complimentary general information packets and 7,240 information booklets for 8 special topics; and over 800 patients enrolled in the Yale University AN Causation Study directed by Dr. Elizabeth Claus (the goal is to survey 1,000 patients).

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[8] See "The New Century of the Brain" (cited above); also Ariel Sabar, "Flashes of Genius," *Smithsonian*, July/August 2015, a report on the zebrafish research conducted by Professor Florian Engert, Harvard neuroscientist.

● *Scientific American* (June 2015, p. 20) reports on the development at Washington University School of Medicine, St. Louis, of 'fluorescent goggles' to help surgeons spot microscopic bits and pieces of tumor that might otherwise go undetected and increase the likelihood of tumor recurrence.

● Sam Kean (a science writer who reviews books for *The Wall Street Journal*) has been called "science's premier story teller." Has anyone read his most recent story of 16th-century medicine, *The Tale of the Dueling Neurosurgeons* (2014), examining the careers of Amboise Parė (d.1590) and Andreas Vesalius (d. 1564)? Intriguing title!

A Few More "Hospital Chart Bloopers"

- I saw your patient today, who is still under our car for physical therapy.
- Patient was alert and unresponsive.
- While in ER, she was examined, x-rated and sent home.
- Patient has two teenage children, but no other abnormalities.
- The lab test indicated abnormal lover function.
- Occasional, constant infrequent headaches.
- Patient has been depressed since he began seeing me in 1993.
- Patient is numb from the toes down.
- She stated that she had been constipated for most of her life, until she got a divorce.
- Patient is tearful and crying constantly. Also appears to be depressed.

Some Websites for AN Patients

ANA, National Acoustic Neuroma Association
www.anausa.org

ABTA, American Brain Tumor Association
www.abta.org

ALDA, Association for Late Deafened Adults
www.alda.org

ATA, American Tinnitus Association
www.ata.org

BAHA, Bone Anchored Hearing System
www.cochlear.com

HEC, House Ear Institute , Los Angeles
www.hei.org

HLA, Hearing Loss Association of America
www.hearingloss.org

IRSA, International Radiosurgery Association
www.irsa.org

NHGR, National Human Genome Research Inst
www.genome.gov

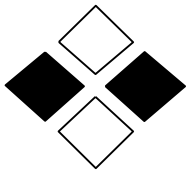
NIH, National Institutes of Health
www.nih.gov

NFI, Neurofibromatosis Inc.
www.nfinc.org

NLM, National Library of Medicine
www.pubmed.gov

SHHH, SelfHelp for Hard of Hearing People
www.shhh.org

VEDA, Vestibular Disorders Association
www.vestibular.org



Fall 2015 Chapter Meeting

"Acoustic Neuroma: An Open Discussion of Diagnosis, Growth Rates, Treatment Options and More"

Dr. Richard M. Hodosh

Medical Director, Atlantic Brain & Spine Institute

Neurosurgeon (Morristown and Overlook Hospitals)

Medical Advisory Board, ANA/NJ

Sunday, October 18, 2015, 1-4pm

Morristown Medical Center, Auditorium B

Refreshments Social Time

Directions to Morristown Medical Center, 100 Madison Ave, Morristown, NJ

From Northeastern NJ. Take I-80 West to I-287 South to Exit 35, marked Madison Ave. Turn left at the light onto Madison Ave. Make a left at the next light to hospital main entrance and parking garage.

From Newark area. Take I-78 to Route 24 West to I-287 South, and follow the directions above for Northeastern NJ.

From Central and Southern NJ. Take I-287 North to Exit 35, marked South Street. Turn left at the light. Bear right onto the access road toward Madison Ave and turn right onto Madison Ave. Make a left at the next light to the hospital main entrance and parking garage.

▪ **Note:** For **Auditorium B**, enter the hospital at the main entrance, go straight through the lobby (passing the information desk on the left) and continue down the hallway. Turn left when you see the Gift Shop to go to the elevators. Take the elevator down to Level B and take a quick left, and then right, to Auditorium B. Look for ANA/NJ signs. Discount coupons for the parking garage will be distributed at the meeting.

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