ANA NJ
ACOUSTIC NEUROMA ASSOCIATION of NEW JERSEY

ANA/NJ Newsletter

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Acoustic Neuroma Association

of New Jersey

A Non-Profit Corporation

Website: www.ananj.org

Gamma Knife, Icon

Notices

● A reminder to please call Wilma if you would like to help the Executive Board in areas like program planning for future meetings, updating the ANA/NJ website, and/or improving our outreach on Facebook, among other things. Chat with Wilma about this at 609-510-9039 or 609-751-9501.

● Do you have important information about acoustic neuroma that you'd like to place here on the 'Notices' page? Or perhaps even a short article you've written about acoustic neuroma that you'd like to propose for the newsletter? Feel free to email any proposals to Dick Barker, editor, at rjbarker@centurylink.net, or regular mail to Dick at 115 Barbertown Pt Breeze Rd, Frenchtown, NJ 08825. Our Facebook page (Acoustic Neuroma Association of NJ) is another opportunity to share in the conversation about the AN experience. Both newly diagnosed and earlier ANers appreciate "hearing" in these special ways.

● An interesting website to visit on your computer is BrainFacts.org, edited by neuroscientists who provide a wealth of information about the anatomy of the brain and the functions of its different structures. There's a wide variety of articles, videos and interactive features. A search for "Hearing," for example, led to an informative article by Allen Ryan, UCA/San Diego, entitled "Closing the Gap between Cochlear Implants and Natural Hearing" (2014), a discussion of research to fine-tune the implant's ability to register differences in pitch and volume. There's also a good video presentation (transcript available) on "The Vestibular System: The Brain and Balance," by neuroscientist Stephen Lisberger, Duke University.

● Akouos ('Listen') is the name of the Boston-based biotech company established in 2016 that hopes to develop the first gene therapy to treat hearing loss. The co-founders are Dr. Emmanuel Simons (PhD in medical engineering from MIT) and Dr. Michael McKenna (MD, Professor of Otolaryngology at Harvard Medical School). For information about the company's gene therapy research, see www.akouos.com; and see also the Q&A with the co-founders entitled "Medical Report: Gene Therapy – An Emerging Strategy to Combat Acoustic Neuroma," in ANA *NOTES* (December 2019).

FDA Approval of Cochlear Implants for Single-Sided Deafness

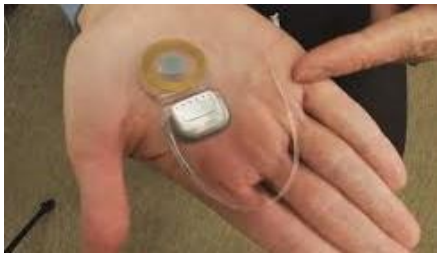
The U.S. Food and Drug Administration (FDA) has approved MED-EL's cochlear implant (CI) systems for single-sided deafness (SSD) and asymmetric hearing loss (AHL). MED-EL/Medical Electronics is a leader in implantable hearing solutions, including the Bonebridge bone-conduction implant. This is the first time the cochlear implant has been approved for the two indications.[1]

The approval of the MED-EL cochlear implant systems, including Synchrony and Synchrony 2, "was based on clinical data from a study at the University of North Carolina at Chapel Hill with 40 participants ages 18 or older to evaluate speech perception in quiet and noise, sound localization and quality of life. Trial participants had single-sided sensorineural profound hearing loss in one ear, or asymmetric hearing loss, for less than 10 years [average duration of 2.6 years] and had used a hearing aid regularly for at least some of that time. All of the people in the study had also tried some type of current hearing device to treat

[1] MED-EL (www.medel.com), Press Release, Durham, NC, July 22, 2019.

SSD, such as a hearing aid, bone-conduction device, or a CROS hearing aid. Subjects were implanted at the University of North Carolina at Chapel Hill with the MED-EL CONCERT or SYNCHRONY Cochlear Implant System . . . [with a standard 31 mm electrode array]."[2]

What's shown on the palm of the hand in the image on the right is the part of the Synchrony system (ca.1 inch wide by 1.7 inches long) implanted under the skin near the deafened ear. The electrode array is on the long wire which is inserted into the spiral-shaped cochlea during surgery The transmitter for an external audio processor is held in place on the head by a small internal magnet. On your computer, you can Google 'Synchrony 2 Cochlear Implant' for a helpful video illustrating all parts and explaining how the overall system works.[3]



The cochlear implant has the potential to bring back a deaf ear to hearing life. Are you currently thinking of having the cochlear implant for SSD?

Actually, it would appear that the way most Americans have dealt with the problem has been simply to *learn to live with it*. A Columbia/ Johns Hopkins University study on "Prevalence of Adult Unilateral Hearing Loss and Hearing Aid Use in the United States" concluded just recently that "Unilateral hearing loss (UHL) is common among U.S. adults [7.2%]. But hearing aid usage is very low (2%), even when there is perceived handicap. Public health education is needed to increase awareness of and auditory rehabilitation for UHL"[4]

Acoustic neuroma patients with SSD have been using CROS aids and Baha devices for quite a few years now. The Baha device was FDA approved back in 2002. Candidates for CI are expected to have had prior experience with such aids. Only recently have CI providers begun to advertise their advancing technology enticingly for "What You Can Do When Hearing Aids Aren't Enough?" FDA approval of the CI for SSD is likely to increase usage by encouraging medical insurers to give significant coverage more routinely for the implant. The process can be expensive, perhaps as much as $50,000-$100,000, considering the testing, surgical procedure and post-operative training with speech therapists and audiologists to assist in mastering the new way of hearing.

But first of all, are you an acceptable candidate for the CI? To even get started, you must have a useable cochlear nerve that connects the ear to the brainstem. Testing for the integrity of the nerve by MRI and EABR (electrical auditory brainstem response) will be needed. Then too, there's the question of how long

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[2] MED-EL, *op.cit.* For a report on the clinical trial, see Emily Buss et al, "Effects of Cochlear Implantation on Binaural Hearing in Adults with Unilateral Hearing Loss," *Trends in Hearing* (Jan-Dec 2018). This report is also available online at www.ncbi.nlm.nih.gov/pmc/articles/PMC59506/.

[3] At PubMed, see J.P.Roche (Univ of Iowa College of Medicine), "On the Horizon: Cochlear Implant Technology," *Otolaryngol Clin North Amer*, vol. 48(6)(Dec 2015). Online free full text copy available.

[4] J.S.Golub, F.R.Lin et al, in *Laryngoscope*, vol. 128(7) (July 2018).

the cochlear nerve has been out-of-service. Dr.Roland (NYU Langone) has observed: "Patients with a long-term deafened ear generally do not like or do not fully incorporate the cochlear implant signal early on and it takes a long period of time wearing the device and having the brain make sense of this new information."[5] Five to ten years is usually the limit set for the deafened ear, although benefit has been obtained in patients with long-standing UHL of more than 25 years.[6]

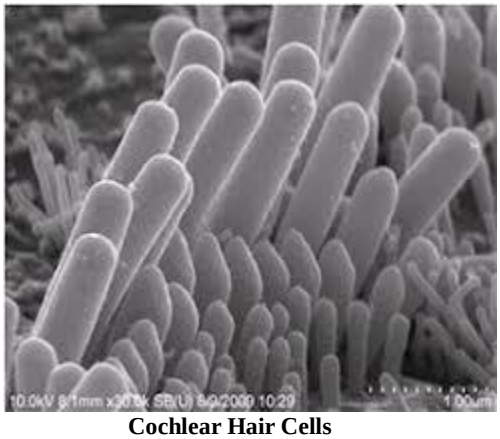
Prospective candidates for the CI also need to ask themselves if they will be able to handle the novelty of the device and persevere with the auditory therapy and device fine-tuning necessary to train the brain to hear again in the deafened ear. The MED-EL clinical trial showed that consistent device use and dedicated practice were very important for positive outcomes throughout the follow-up period. Most subjects in the trial reported wearing their CI between 10-12 hours per day; the two poorest performers reported using their CI unreliably for only 4 hours per day. Learning to hear again with the CI can be fatiguing.

One concern in the trial was the ability of the brain to sort out both acoustic and CI-electric stimuli, and the degree to which hearing from the CI might interfere with the acoustic signaling processing from the good ear. "Better ear dominance is one reason why clinicians and researchers often recommend postoperative rehabilitation with the CI alone for patients with SSD."[7]

The extent to which a CI has the potential to restore sound is highly individual.[8] After many years, it's hard to train parts of the brain that have not been stimulated regularly. The National Institute on Deafness and Other Communication Disorders recommends: "The decision to receive an implant should involve discussions with medical specialists, including an experienced cochlear-implant surgeon.

Research on Regrowing Hair Cells

Research at the University of Rochester and Harvard University, where scientists report on having been able to regrow sensory hair cells in the cochlea of neonatal mice, "could represent a new approach to cochlear regeneration [of hair cells] and, ultimately, restoration of hearing," writes Patricia White, associate professor at the Del Monte Institute for Neuroscience, Rochester School of Medicine.[9] The cochlea with its hair cells is where sound vibrations are converted into electrical signals, allowing people



Cochlear Hair Cells

[5] J.Thomas Roland, "Medical Report: Cochlear Implants and Acoustic Neuroma," *ANA NOTES* (December 2017).
[6] Emily Buss et al, *op.cit.*
[7] *Ibid.*
[8] On Facebook, check out "Cochlear Implant Experiences" for individual stories.
[9] "New Therapy for Hearing Loss?" *Rochester Review* (Fall 2018). See J.Zhang, P. White et al, "ERBB2 Signaling Drives Supporting Cell Prolif- eration in Vitro & Apparent Supernumerary Hair Cell Formation in the Neonatal Mouse Cochlea," *European Jour Neuroscience*,Vol 48(10) (Nov 2018).

to hear. “In mammals, cochlear hair cells are not regenerated once they are lost, leading to permanent hearing deficits.” The new research centers on the role of a specific growth receptor named ERBB2 which acts to foster the generation of new sensory hair cells. J.Zhang, associate in neuroscience in the Dept of Biology at the University of Rochester commented: “It’s funny, but mammals are the oddballs in the animal kingdom when it comes to cochlear regeneration. We’re the only vertebrates that can’t do it.” But will we be able to soon?

A Non-surgical Bone Conduction System

The global company MED-El (headquarters in Innsbruck, Austria) has also obtained FDA clearance for ADHEAR, a bone conduction hearing system that does not require implant surgery or a through-the-skin abutment. The ADHEAR employs “skin drive,” meaning that vibrations for hearing are passed across the skin to the bones of the skull.

An overview of the system in *Audiology Online* (Oct 2019), sponsored by MED-EL, describes how the system is comprised of two parts: an adhesive adapter and the audio processor (see above). “The adhesive adapter is placed on the hairless area of the mastoid behind the ear. The audio processor simply clicks into place on the adhesive adapter. The processor may be removed as desired throughout the day. The adapter stays in place typically for days at a time.”[10] The adapter is medical grade adhesive that is latex-free.

The contralateral ear to which the sound is being routed should have normal air and bone thresholds of 20 dB HL or better.

The audio processor features a lightweight design, automatic adaptive directional micro-phones, gain and compression controls, noise reduction, and feedback management. The design is symmetric so the device can be used on either ear. It is available in 3 colors: brown, silver and black.

[10] A.O’Donnell et al, “ADHEAR System by MED-El: An Overview of the Research and Clinical Experiences from the Field” (2019). There is a demo on UTube by Dr.Cliff Olson.

Cindy Rivkin in the Spotlight

The bottom line is that being deaf is hard and it's tiring. If it wasn't for a wonderful, supportive, close knit family, Cindy wouldn't have overcome so much. She had a close family growing up and her Mom is her best friend. She has a supportive husband and two grown children now and throughout her ordeal they held her together.

Cindy's troubles came packed together in way too short a time period to not be overwhelming. She had lost her left hearing gradually, probably due to a cold, so when she was diagnosed with an acoustic tumor in her right ear it was alarming. She held out as a 'wait-and-watch' for several years but when the tumor showed growth, she felt it was time to act.

Cindy was nothing if not thorough in her research and exploration of her options, but no doctors were especially enthusiastic about taking on the risk of her being plunged into total silence. In the end, after considering her options with doctors on both coasts, she chose Dr. Samuel Selesnick to do her retrosigmoid surgery, because if the 20% chance that she might lose her right hearing occurred, he could also do cochlear implants for her. Before she was able to do any of that, however, she was diagnosed with another medical problem which more urgently needed attention. She headed into the hospital for that surgery before she could proceed with the other. Thankfully it turned out not to be cancer as feared.

Six months later, Cindy was back in the hospital for her AN surgery. She couldn't risk radiation because the scarring it would cause would preclude a cochlear implant and she needed to retain that option. Surgery was done at Sloan Kettering Hospital: and indeed, it left her totally deaf. Despite that she insisted on going to her son's Junior High School performance in "The Beauty and The Beast." At her son's request, the school roped off prime seats for her. It was a silent performance for her, but she was there. Family is everything.

Cindy pushed hard to be scheduled quickly for a left cochlear implant (CI) wanting to move out of her silent world where communication was done at home with a white board. When the family gathered for meals, as they frequently did, her mom made sure to always provide pads, pens and markers at every seat ensuring she was included in the conversations. Her husband, kids, mom, stepdad, sisters-in-law, her niece and nephews, and her mother-in-law all went to unbelievable lengths for her. She is so very grateful.

The CI magnet was implanted at 6 weeks post AN surgery (on the opposite ear) and the hearing device was activated 2 weeks later. Amazingly, she could hear her mother right away, though not her husband, and she was unable to distinguish between her children. Her recovery from this first CI surgery was easy but the surgery later in the year for the AN side was much longer and more complicated due to scar tissue and complications from anesthesia. She had severe leg pain afterwards which turned out to be a blood clot requiring an emergency operation to save her life. She'd had seven surgeries in less than a year and a half! Hence, when she told her family, none of whom wanted a dog, that she wanted one, they didn't have the heart to say no to her. It soon became apparent that Jet, as he was named, was her best medicine giving her comfort, companionship, acceptance and love. Cindy is sure that in an emergency, with her devices off, Jet would alert her to danger.

It is now five years since Cindy received her Cochlear Implants. Over the first year there were many adjustments to correct the programming to improve her hearing. She was initially nauseated by high frequency sounds, but this was resolved through programming adjustments. There was hope that the second CI would eliminate her tinnitus, but although much improved, it never went completely away. Cindy does very well hearing and understanding in quiet environments when she can also see the speaker's face, but noisy environments remain excruciating. Sound and the constant work to interpret it is exhausting and causes brain fog. When Cindy takes off her CIs at night to sleep, she is plunged back into silence, which is unnerving. She has adjusted to a different life but has also been able to retain much of the good from her earlier life with necessary adjustments. There is little improvement these days in her hearing, which will never be like "the real thing". Adding sign language is not a realistic consideration, due both to the difficulty of learning this other language and the fact that those she would communicate with do not sign.

Cindy needs hearing breaks after a number of hours due to brain fatigue. She has sought support and answers to her questions from social media groups, including those for Acoustic Neuroma prior to her surgery and for Cochlear Implant users afterwards. The CI group helped her early on to realize things would improve over time and with fine tuning of her CIs. Members helped her to deal with her anxieties such as going for her first haircut after getting her CIs. She was self-conscious about her scars and worried knowing she'd be deaf during the hair wash and cut when the external parts for her CIs were removed.

In the real world, Cindy finds people have trouble understanding that her challenge with understanding speech is not a matter of volume but rather acuity, and that often it just requires patience and rephrasing a few words to enable her to understand. She doesn't like to call attention to her hearing loss and tries to manage without telling people. By all appearances, she is a normal, healthy young woman, but in reality, she is coping every day with the extra work of deafness.

Interview by Kristin Ingersoll
January 20, 2020

Spring Meeting

"Caring and Sharing"

Open Meeting for Acoustic Neuroma Patients, Family & Friends

Sunday , April 19, 2020
1 – 4 pm
Mercer County Library System
Lawrenceville Branch
2751 Brunswick Pike (Route 1 South)
Lawrenceville, NJ

Group Discussion Refreshments Social Time

(Questions or RSVP to Wilma at 609-510-9039)

Directions:

From North Jersey: Using Route 1 South. After the I-295 overpass and traffic light at Franklin Corner Rd, stay to the right to get onto Business Route 1. Follow until the traffic light and right turn onto Darrah Lane. The Library with parking will be there on the right.

From South Jersey/Trenton: Take Route 1 North to the Whitehead Rd Exit. Make a left onto Whitehead Rd and follow until the traffic light. Make a right onto Business Route 1 and continue about one mile. Immediately after the third traffic light, move into the jug-handle to cross Route 1 onto Darrah Lane. The Library with parking will be on the right.