

REFLECTIONS

An Otolologist's Experience as a Cochlear Implant Patient—The Power of Neuroplasticity

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I knew it was time to consider getting a cochlear implant when I realized that patients I was providing with implants had better hearing than I did. This resulted in a somewhat unique situation: an otologist with an implant. The saga of surgery, programming, and relearning how to hear with an implant has given me some valuable insights—especially regarding the neuroplasticity of the human brain—that, I believe, would otherwise be lost to the implant community. What follows are the lessons learned.

I have an autosomal dominant, progressive, sensorineural hearing loss that was diagnosed when I was a young child. Fortunately, normal speech development was possible for me; however, as time went on, my ability to hear high frequencies diminished to the point that even the strongest hearing aids were of little help. By age 50 years, I was understanding speech with only low tones remaining in both ears (250 Hz and 500 Hz), corresponding to vowel sounds. Speech without consonants sounds very much like Charlie Brown's teacher. I was able to use lip reading and other strategies to get by with my family, friends, and patients but was definitely experiencing a diminished quality of life.

There is no question that I had the same trepidations as other potential implant recipients, such as losing residual hearing and relying on an external device, but my hesitancy was also founded on my knowledge of the variability in implant performance in the patients I had treated: not everyone was universally satisfied. My concerns were eventually outweighed by the increasing barriers I was experiencing with my hearing loss, such as trouble talking on the phone, the challenges posed by air travel (not being able to hear announcements or gate changes), and the difficulty fielding questions at conferences.

Choice of Device. In my practice as an implant surgeon, I let the patient choose the implant. There are 3 manufacturers: Cochlear Inc, MED-EL, and Advanced Bionics, all of which offer comparable products. My audiogram showed preservation of low tones in both ears, but since I really did not have any hearing at the high frequencies (1000 Hz or above), I was—audiometrically speaking—a perfect candidate for a hybrid device, which uses electrical acoustic stimulation (EAS). In theory, EAS provides the best of both worlds for implant recipients because it combines both types of hearing (electric from the implant for high tones and acoustic from the remaining low tones in the same ear).

My concern was that residual hearing in my ear would eventually go away, and I would reach a point where I would require full electrical stimulation. Therefore, I opted for complete insertion of the longest electrode available (a nonhybrid device), so that the end of the cochlea, where the low tones are stimulated, could be reached. My surgical experience implanting long, straight electrodes showed preservation of residual hearing in a high percentage of patients.

If this were the case with my own implant, I would be able to use my preserved low tones until I could no longer hear these tones acoustically, at which time the deeper electrodes would be stimulated (via programming).

I underwent the implantation procedure as an outpatient, and the surgery was uneventful. The first thing I did when I woke up from anesthesia was tap on the plastic cup ear dressing, allowing me to confirm that I did have preserved hearing. However, although I experienced the usual reduction in hearing due to full insertion of the electrode, the postoperative audiograms showed that this was not the expected sensorineural loss. Instead, it was apparently an inner ear conductive loss. The reasons for this phenomenon are currently being investigated and could have implications for future electrode design. Images taken at the time of surgery had shown successful insertion of all the electrodes. The implant would be turned on after 2 weeks of recuperation—I was good to go!

Implant Programming: Let the Brain Do the Work. Any implant surgeon will tell you that the true work begins after the surgery, in the audiologist's office. This is where the implant is activated and the programming occurs. Programming adjustments continue at numerous follow-up visits during subsequent months, at which time the audiologist will adjust the loudness for each frequency for each electrode to give the patient optimal hearing through the device.

There have been many heartfelt moments in my experience as an implant surgeon when that switch was first turned on, say for an infant patient, eliciting a cry from the infant and tears from the infant's parents. When my implant was activated, I could have cried too, but for different reasons. Although the tones above 1000 Hz were fairly clear and distinct, they sounded much like beeps and whistles. Everything below 1000 Hz was nearly absent or distorted. I think this is why many implant recipients with poor low-tone discrimination describe the first input after activation as sounding like Mickey Mouse. Below 750 Hz, the perceived sounds were akin to someone running sandpaper over wood. It was not tonal at all and was really not pleasant. Thus began my saga of getting it just right.

I was a vigilant implant recipient—doing all the aural rehabilitation exercises and using appropriate software aids on a regular basis. Although I was noticing sounds that I had not heard for a long time, like the clicking a car engine makes when it cools off, these all sounded bad to me, and most seemed too loud, especially high tones that I had not heard in a long time. Since I could not recall what normal sounds were, I would need to ask what something was before I could hear it correctly. Certain consonants sounded different from each other, but it took a while to determine, for example, which was a “p” and which was a “d”; this ability came with use, like learning a new language. I was progressing ad-

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equately with high-frequency sounds, but I could not differentiate between men's and women's voices.

One of the frustrating things for me was when the audiologist would ask how a particular program sounded. It is difficult for someone who has not heard in a long time to answer this question. After all, how would I know what something was supposed to sound like? In my opinion, everything sounded kind of bad, and all the new sounds I was hearing seemed way too loud. However, repeated adjustments to the programming, to make things sound better—I would ask the audiologist to turn down the sounds that I perceived as too loud only to realize that the resulting tones were then not loud enough—would cause a 2-week setback in my ability to understand speech. This appeared to be how long the neuroplasticity of my brain needed to readjust. Thus, endless tweaking of the program only made things worse because my brain had to relearn how to hear the new input each time. After about 6 weeks of repeatedly manipulating the programming, I realized what was happening and asked the audiology team to program every frequency straight across at 30 dB, since that is how the normal cochlea works. This is the program that remains in my implant 4 years later, and I have not adjusted it since that time.

About 1 or 2 months after the final programming, a curious thing happened: I was able to distinguish a man's voice on the radio. This ability lasted for just a minute or 2 and then went away. I thought there was something wrong with the radio, and I was very intrigued but disappointed with my fleeting ability to hear the low sounds. However, the next day this phenomenon lasted longer, and over a period of 3 to 4 days, my ability to perceive low tones, which had been pretty much nonexistent, had come back. My brain appeared to latch onto this new stimulation, and I could discriminate a man's voice from a woman's and my own voice sounded more normal. Since this change happened without any additional programming, I began to believe that outcome in cochlear implantation is more related to neuroplasticity of the brain than to the device or to the cochlea. And I was amazed how fast it all happened once it began.

As soon as I could hear low tones in my implanted ear, however, an additional problem surfaced. This was due to the remaining hearing in my other ear. Using a tone generator on my phone and comparing it between the residual tones in my nonimplanted ear, I was able to determine that 250 Hz in the implant was being heard as 220 Hz. Sounds at 500 Hz came over as 550 Hz. Since the implanted ear was not correctly perceiving the low tones, I experienced a dissonance from the correct tones (250 Hz and 500 Hz) being heard in my contralateral ear. The result was that any speech in the low tones sounded like the creepy possessed girl from *The Exorcist*. Not great quality.

Over the next 2 to 3 months, or 4 to 6 months after the final programming, I compared each ear with my tone generator from week to week. I found that 220 Hz in the implant ear slowly came up to match 250 Hz in the hearing ear. The same thing happened at 500 Hz. The tone discrepancies resolved, and voice input sounded correct for both low and high tones after 6 to 9 months. While my own voice still sounds like I am talking on a microphone—hearing aids had that same effect for me—I think other things sound pretty normal now.

This experience makes me skeptical of the advice we currently give to implant recipients, which is to plug up the hearing ear and

only use the implanted ear to hear. I did that for a while, but it turned out that my other ear helped the implanted ear to reprogram to the correct frequency. Additionally, although EAS was initially a little bit better than full electrical stimulation, after I adapted to the low tones, I opted to use full electrical stimulation over EAS. This was primarily because the noise reduction software that is part of the implant worked best when all frequencies were being stimulated electrically. I was also very pleased to remove the earpiece needed for EAS.

Music Appreciation. You cannot expect good outcomes with an implant unless you stimulate the ear with what you want to hear. I had worked on speech first, followed by voices on the phone, and then television. This process worked with time for nearly everything, except music. I used to play the trombone, and the smooth glissando using the slide of this instrument did not sound quite right with the implant, sounding more like individual keys on a piano: I could hear each step from each electrode channel activation.

The complexity of music also affects my ability to appreciate it. Hearing rhythm was good from the beginning, and a jazz piece with just a few instruments does not sound bad; however, a full orchestra sounds like rain on a tin roof. Although I no longer wear a hearing aid in my other ear, preserved low tones in that ear have helped with music appreciation. Also, there seems to be some auditory memory involved, since once I am told the name of a piece I am listening to, I can then "hear" it. It begs the question whether I am really hearing a song or if my brain is just filling in the blanks. That said, music through a cochlear implant, at least in my experience, sounds like a transistor radio from the 1970s, but it is still better than nothing.

A Word on Tinnitus. As with many who have a hearing impairment, I have had lifelong tinnitus. Although my tinnitus was a little bit worse when I first received the implant, once my brain started adapting to the low tones, the tinnitus went away. After about 6 months, the tinnitus was nearly completely suppressed. The interesting thing is that the tinnitus comes back when the implant is off, almost within 2 to 3 minutes, but even then, it is not as loud as it was before, and it almost instantaneously goes away when the implant is turned back on. My theory is that the electrical stimulation from the cochlea reverses some of the maladaptive sensory reorganization that occurs in the brain that causes the tinnitus. Continuous electrical stimulation from the cochlea, when the implant is on, is necessary for more complete resolution of the tinnitus.

Conclusions. This has been one of the most fascinating experiences I have ever had, both personally and professionally, and I feel that I have learned more from this 1 cochlear implant in my head than the 1000 plus devices I have implanted in others. I had always thought that outcome was related to many factors, but primarily to the condition of the cochlea and spiral ganglion cells and to the design of the implant itself. I had no idea how important neuroplasticity of the brain was and how powerful it could be when learning how to hear with an implant. This leads to many questions in my mind. What is the mechanism of these changes, and how can we augment them? Why does this neuroplastic change not occur in everybody with similar auditory profiles? Should we include routine assessments of cognitive function for implant candidates and, if so, which ones? What is the mechanism in children who have never heard, and is it different than in adults? This is medicine's first successful attempt to replace a special sense, and we have so much more to learn!