

Cochlear Implants in NF2

In January 2009, Marie Drew, NF2 Representative on the NF, Inc. Board of Directors, posed a series of questions on behalf of the NF2 Crew to Derald E. Brackmann, M.D. of the House Ear Institute in Los Angeles.

- What can a patient do to convey to the doctor the thought that saving the auditory nerve is as important to them or more important, depending on the individual, than saving the facial nerve?
- Does it matter what the patient makes plain to the doctor, or does it all depend on the size and adherence of the tumor, and what the surgeon is required to do?
- Could you improve your chances of being able to use the Cochlear Implant (CI) by having surgery rather than doing the “watch and wait” routine?
- Are chances of utilizing the CI better or worse with radiosurgery or microsurgery?

Dr. Brackmann's Response:

First of all, as you noted, the auditory nerve must be intact and also functional for a cochlear implant to work. This is often very difficult to achieve in NF2. The tumors in NF2 behave differently than unilateral vestibular schwannomas. With unilateral vestibular schwannomas, the cochlear nerve is displaced to the surface of the tumor whereas in NF2, the cochlear nerve is incorporated within the tumor in all but the smallest tumors. Dr. Linthicum in our Histopathology Laboratory has shown this in NF2 patients.

When tumors are operated when they are very small, before they involve the cochlear nerve, we may actually remove them and save the hearing; and this is what we recommend if tumors can be discovered when they are very small. Despite saving the cochlear nerve in these patients, sometimes the hearing is lost due to the blood supply to the cochlea being involved with the tumor. This causes degeneration of the hair cells and in some of those patients, the cochlear nerve remains intact and functional. These patients would then benefit from a cochlear implant. Unfortunately, this is a very small percentage of the total number of patients with NF2.

This does support the idea, however, of operating the tumors when they are small so that even if the hearing is not preserved, the cochlear nerve might be, so that a cochlear implant would be possible.

Other than making a decision to have surgery when the tumor is small, behavior of the tumor determines the ability to save the cochlear nerve rather than the surgical technique. As tumors become bigger, the cochlear nerve is entirely involved with the tumor and removal of the tumor will remove the cochlear nerve, making a cochlear implant impossible.

In regard to radiosurgery, the cochlear nerve is preserved with radiosurgery. Many of these patients lose their hearing, not because of non-functioning of the cochlear nerve, but because the radiation affects the blood supply to the inner ear causing degeneration of the hair cells. Some of these patients are excellent candidates for cochlear implants. I, personally, have 3 patients who have undergone radiosurgery with stabilization of the tumors. We then do a test called promontory stimulation. If patients perceive auditory sensations to the electrical stimulation, we consider them to be a good candidate for the cochlear implant. These 3 patients are functioning at a very high level with their cochlear implants, much like a cochlear implant patient without a tumor would do.

Unfortunately, there are some patients who have had radiation who do not receive auditory sensations from electrical stimulation on the promontory stimulation test. Apparently, in some cases, the cochlear nerve is also affected and a cochlear implant is not possible.

I now consider the possibility of a cochlear implant in all of my NF2 patients. In general cochlear implant patients perform better than ABI patients, although there are some exceptions to this. Some of our ABI patients actually do as well as some cochlear implant patients.

To summarize: unfortunately, there is not a great role for cochlear implantation in NF2 because the cochlear nerve is intimately involved with the tumor in most cases. The role of radiosurgery in NF2 is still debated but there are cases where cochlear implantation has been successful following hearing loss with radiosurgery where the tumors have remained stable.

“My response to you does not, and cannot, provide individual medical advice, but rather is for general informational purposes only. My response is not intended to be a substitute for individualized medical advice, diagnosis or treatment by a physician who is aware of your medical history and has had an opportunity to examine you. Do not rely on my response in place of seeking professional medical advice.”

Derald E. Brackmann, M.D.
House Ear Institute
Los Angeles
dbrackmann@hei.org