The ABI and NF2:

How Advancements in Auditory Technology Benefit the Patient

Using information gathered over decades of research to develop and improve the cochlear implant, a prosthetic device designed to provide some sound and speech information for deaf people, scientists at HEI devised an auditory brainstem implant (ABI) to provide similar benefits for those suffering from NF2. Neurofibromatosis Type II (NF2) is a rare genetic disease characterized by tumors growing on both vestibular nerves that adversely affect the auditory and facial nerves. We recently discussed the ABI device and how it benefits patients suffering from NF2 with Bob Shannon, Ph.D., and Derald E. Brackmann, M.D. Together, these experts were able to elucidate both the technology and the disease.

Q: How is the auditory brainstem implant (ABI) device different from the cochlear implant?

A: The cochlear implant stimulates the cochlea or hearing organ and sound is then conducted along the auditory nerve to the brain. The ABI is the first device specifically designed to bypass the cochlea and the auditory nerve to transmit sound directly to the brainstem, allowing the recipient to receive environmental sounds and speech information.

Q: Who can benefit from an auditory brainstem implant (ABI)?

A: NF2 patients and other people who have damaged or severed auditory nerves, can benefit from the ABI. After implantation, patients work with audiologists to map and adjust their speech processors as they learn to understand and interpret new sound signals.

Q: Where is the ABI positioned?

A: The ABI is placed directly on the nerve center (cochlear nucleus) at the base of the brain, typically during surgery to remove tumors.

Q: How does an ABI help people communicate?

A: The ABI device is designed to bypass the auditory nerve and transmit sound directly to the brainstem. When the ABI user receives the processed electrical signal via the external digital signal processor, they are able to recognize some environmental sounds and some speech.

Q: Who invented the auditory brainstem implant (ABI)?

A: William Hitselberger, M.D., and William F. House, M.D., developed the Central Electroauditory Prosthesis, the forerunner of the ABI, at the House Ear Institute in the 1970s.

Q: I understand that the ABI device was invented in hopes of restoring partial hearing to NF2 patients. What is NF2 and how common is it?

A: Neurofibromatosis Type II (NF2) is a rare genetic disorder that occurs in one in 40,000 live births causing bilateral tumors to develop on the vestibular (balance) nerve as well as in other parts of the central nervous system. If NF2 is identified, the children of affected individuals also could be at risk of developing the disorder. The mutant gene occurs in 50% of the offspring of an NF2 patient. Early screenings of these individuals with MRI and/or blood tests can be highly effective in minimizing the effects of NF2.

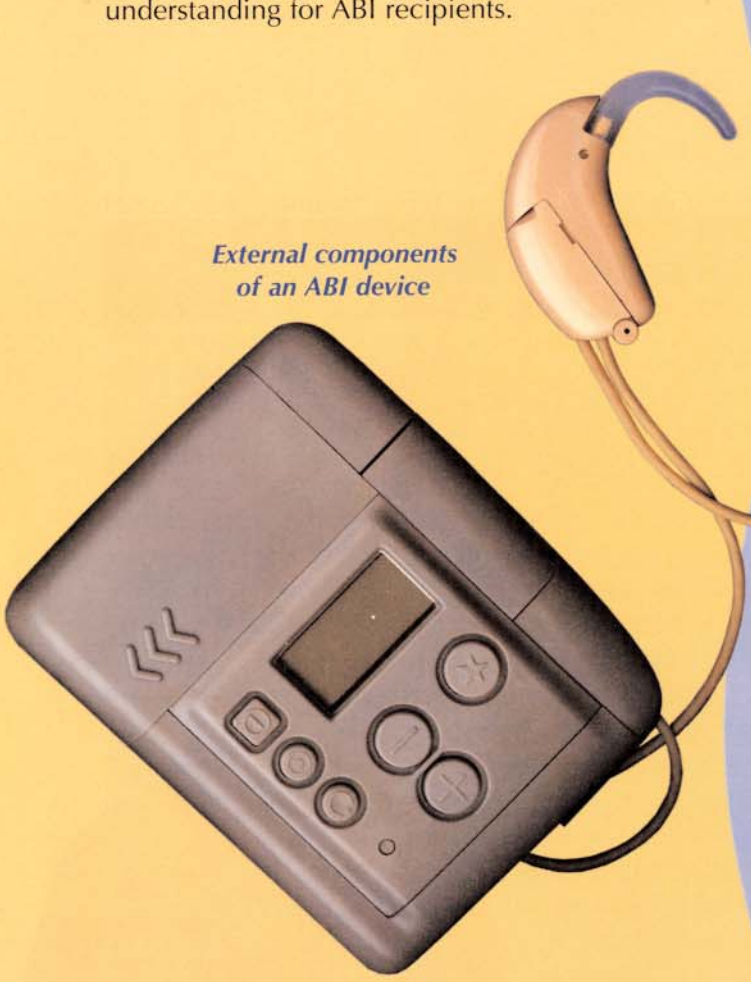
Q: If tumors from NF2 grow on the balance nerve, how does this affect hearing?

A: The stato-acoustic nerve is made up of two branches, the acoustic (hearing) branch and the vestibular (balance) branch. These nerves travel through the same narrow canal at the base of the skull. Pressure created by the vestibular schwannomas (tumors) can compress the nerves, resulting in hearing loss. Some patients may experience a loss of balance in the involved ear, but it is possible for the other ear to compensate over time. *(continued on page 10)*

(continued from page 4)

Q: How have general improvements in technology worldwide advanced ABI science, and what does the future hold for improved ABI technology?

A: New models of ABIs have benefited from the high-tech development of materials and electronics by researchers in the field of neural prosthetics and speech perception. Future generations of the ABI will actually penetrate the cochlear nucleus in the brainstem with a penetrating electrode array. It is theorized that this may result in improved speech understanding for ABI recipients.



External components of an ABI device

Bob Shannon, Ph.D., is Director of HEI's Department of Auditory Implants and Perception (DAIP) and Auditory Implant Research Laboratory. Derald E. Brackmann, M.D., limits his practice at the House Ear Clinic to Otology and Neurotology. Dr. Brackmann specializes in diseases of the ear and facial nerve, dizziness and acoustic tumors. Both experts are members of HEI's Auditory Brainstem Implant (ABI) research program.

Q: Is hearing loss inevitable for NF2 patients?

A: If tumors are discovered when they are small, they may be removed with preservation of hearing. Unfortunately, many tumors are only discovered when they are large, where hearing preservation is not possible. In most cases of NF2, growth of the tumors has eventually necessitated surgery to remove any threat to patients' lives, and complete tumor removal often results in both hearing nerves being severed during surgery, resulting in profound deafness. Hearing aids or cochlear implants would be of no assistance to these patients, but an auditory brainstem implant (ABI) can be highly beneficial.

Q: Are all patients with NF2 treated with ABIs?

A: Sometimes very small tumors can be removed without severing the auditory nerve. Another surgical treatment available to some patients is "decompression," which makes room for the tumor so it is less likely to hurt the hearing nerve. A somewhat controversial treatment is the use of x-ray therapy to shrink the tumors. X-ray therapy frequently causes hearing to deteriorate and may not arrest the growth of the tumor. Subsequent surgery is more difficult, particularly in regard to preservation of the facial nerve. We do not recommend radiotherapy for treatment of tumors in NF2.

Q: How many ABI operations have been performed at the House Ear Institute/House Ear Clinic, and is the ABI procedure offered anywhere else?

A: More than 125 patients with NF2 have been implanted with ABIs at HEI. The ABI is also available at an expanding number of other medical facilities nationwide, each staffed with a surgical and audiology team who received training from the House Ear Institute/House Ear Clinic. ❖