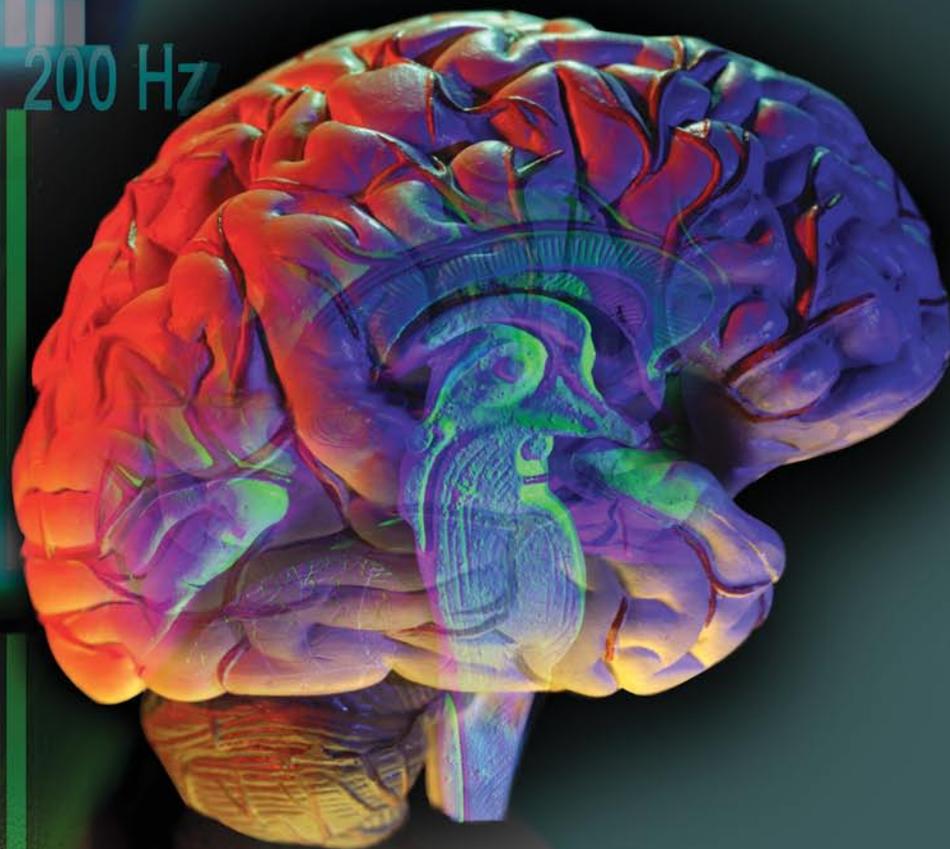




Hearing with Our Brain

Karen's Journey Back to the World of Sound

What is an auditory brainstem implant (ABI) and who is a candidate? Here is a scientific and personal perspective. The ABI is cleared in the United States by the FDA for individuals 12 years of age or older. There are about 400 individuals with ABIs in the United States and approximately 1,000 in the world. The House Ear Institute has the largest number of any place in the world with approximately 260 implantees.



By Barbara Liss Chertok

The House Ear Institute (HEI) in Los Angeles was established in 1946 as a non-profit organization by Howard P. House, M.D. for the exploration of the auditory system from the ear canal to the cortex of the brain. Institute scientists investigate hearing loss and ear disease at the cellular and molecular levels, as well as the complex neurological interactions between the auditory system and brain.

They are working to improve hearing aids and auditory implants, diagnostics, clinical treatments, and intervention methods. House researchers work with House Clinic physicians to integrate medicine and science through clinical and research trials that may directly benefit patients.

A pioneer in the development of the cochlear implant that has restored at least partial hearing in more than 150,000 children and adults with profound bilateral hearing loss, HEI has redefined the frontiers of hearing science. For more information go to www.hei.org.

There are many causes of hearing loss: genetic, noise-induced, age-related, autoimmune, ototoxic or drug-related, but it is neurofibromatosis which poses a particularly challenging problem for the patient.

Auditory Brainstem Implant (ABI) for NF2

Neurofibromatosis Type II (NF2) is a life-threatening, genetic disease of the nervous system characterized by bilateral, non-cancerous fibrous tumors—also referred to as vestibular schwannomas or acoustic neuromas—that grow on the vestibular nerves.

Current treatment options include surgery that usually requires severing the hearing or auditory nerve. If the auditory nerve is cut, a cochlear implant cannot be used to treat an individual with NF2 because the auditory nerve is then not able to carry signals from the cochlea to the cochlear nucleus in the brainstem. In such cases, an NF2 patient may benefit from an auditory brainstem implant (ABI), designed to stimulate the auditory portion of the cochlear nucleus in

the brainstem and send sound signals directly to the brain.

The current FDA approved ABI provides most recipients benefit through increased sound awareness, and a small percentage have achieved near cochlear implant levels of performance. ABI candidates are thoroughly counseled about what to expect from an ABI. An ABI always works best in combination with lipreading cues, and regular use of the ABI contributes greatly to long-term benefit. ABI recipients have shown the ability to continue to improve in performance for ten years or longer.

Karen's Journey

Karen Lichtefeld (56) has been married for 21 years to Paul, a commercial building developer. Karen has three stepsons, three daughters-in-law, and six grandchildren. Her hobbies are travel, yoga, golf and walking. They reside in Kentucky and spend winters in Florida.

Karen worked for more than 20 years in the insurance industry, the health care marketing field and in sales. She also worked as an admissions director for Sullivan College of Technology and Design in her hometown of Louisville, Kentucky.

At age 27, while on the job at an insurance company, she noticed having difficulty hearing on the phone with her left ear. A visit to an ENT and testing showed a 50 percent decrease in her hearing and a CT scan showed a 1.5 cm acoustic neuroma. Surgery to remove the tumor was performed by Dr. Michael Glasscock of the Otology Group in Nashville, Tennessee.

The tumor removal resulted in facial paralysis lasting about six months—a normal occurrence with acoustic neuroma surgery. During this time, Karen lost the remaining hearing in that ear. She returned to work five weeks later and her facial expression did return in about six months.

"I considered myself very lucky, she says, and did not even notice that I no longer had bilateral hearing."

Fifteen Years Later

Karen retired wanting to travel more and continue her volunteer work. After returning from a trip by air, her right ear became blocked and

“My whole world changed that day in March of 1995...I remember literally dropping the phone when my doctor called to give me the results of the MRI. He was very blunt about my diagnosis and told me that they could not predict how long my hearing would remain, only that at some point I would be completely deaf.”

the hearing distorted. This time, she visited an ENT in Louisville. Noticing her history, he immediately ordered an MRI which showed an acoustic neuroma in her right ear, and she was diagnosed with NF2.

"My whole world changed that day in March of 1995," she recalls sadly. "I remember literally dropping the phone when my doctor called to give me the results of the MRI. He was very blunt about my diagnosis and told me that they could not predict how long my hearing would remain, only that at some point I would be completely deaf."

Karen went through a very dark period of panic, losing weight and not sleeping. Her husband tried to calm her by helping with research and scheduling appointments with specialists. Each physician confirmed the diagnosis and the dire consequences of NF2.

After a month, she visited her surgeon, Dr. Glasscock, and he assured

her that although her situation was serious, she had time to watch and wait. He also told her that because acoustic neuroma tumors have such a slow growth rate and since her tumor was about 1.5 cm, she could have up to five years before she would have to undergo surgery.

"It was like being thrown a life-saver!" Karen remembers. "Now, I had a goal to work on. I am an organized person and for months I had not been in control or functioning. I had lost ten pounds and was a virtual wreck. I decided to take my life back and learn everything I could about my disease and what options I had to beat this."

Karen Takes Life Back

Karen signed up for classes in speechreading (lipreading) and sign language. She credits her husband, family and super network of girlfriends who kept her going.

It was at this time that she became acquainted with two organizations that changed her life dramatically. The first was the Acoustic Neuroma Association (www.anausa.org). She saw an article in their newsletter a year after her diagnosis that the House Ear Institute in Los Angeles was looking for newly-diagnosed patients with NF2 for a six-year study. She immediately contacted them and met all the criteria for the research group.

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Left to right: Donna Kiley, Karen Lichtefeld, Theresa Kidwell and Michelle Kiley—all members of the Hearing Loss Association of Kentuckiana and HLAA National—at the Grand Old Opry during the 2009 HLAA Convention in Nashville.

Hearing with Our Brain

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"I was to be a 'lab rat' and I was so encouraged," she smiles.

The second organization was HLAA. "I attended my first meeting five years ago and have met the most wonderful people. The members were not only friendly but they really understood the world of hearing loss and the daily problems that entails. I have learned so much from them and information about the technology which is available and improving daily. I wish I had known about them before my first surgery," she admits.

Karen had now met her five-year "wait and watch" goal of annual MRIs and the mountain of tests and paperwork to comply with the HEI study. She was beginning to experience severe tinnitus, one of the first signs that the tumor is putting pressure on the acoustic nerve. She met with her research team and Dr. Derald Brackmann at HEI.

"I was terrified before going there," she recalls, "because I had put my life back to my 'new normal' of denial with plenty of hearing left. I was going to have to face the awful diagnosis again of deafness."

Penetrating Auditory Brainstem Implant (PABI)

"I remember my first meeting with Dr. Brackmann like it was yesterday. He said to me, 'You will NOT be deaf,' and I believed him! He then proceeded to tell me about the groundbreaking research they were doing with the auditory and penetrating brainstem implants (ABI/PABI)."

Because she was already in the NF2 Research study, Karen was a candidate for the new PABI which had about five recipients.

"I can't say enough about the positive reinforcement and excellence of care at HEI. The entire staff exudes professionalism and hope to persons with hearing loss. The entire facility is dedicated to training and research. For the first time in years, I had hope of continuing the hearing life that I had known," Karen relates.

In January 2006, Karen's tumor was about 2 cm by then and her residual

hearing had dropped very low even with a powerful hearing aid. Dr. Brackmann said it was time to think about a surgery date to remove her tumor and implant a PABI.

"I had surpassed my five-year goal to keep hearing and had been training like an athlete to be physically ready for the more than eight-hour surgery," she says.

On September 12, 2006, Karen underwent surgery with Drs. Derald Brackmann and William Hitselberger for a PABI at HEI. The operation was a success and her facial nerve was spared.

"It was very important for my vision to remain clear in order to speechread while I could not hear."

She was in ICU for 24 hours followed by eight more days in the hospital. The surgery rendered Karen completely deaf for the first time in her life. She returned to Louisville after two weeks.

"It was very scary," she says, "but I completely trusted my team and knew this would only be a two-month wait before my hookup."

This period was a special time of healing, introspection and hope for Karen. She had been dreading the eventual surgery that she knew awaited her for more than ten years. She had survived and was now starting a new life of electronic hearing. She felt energized by it.

Another New Life

That November before Thanksgiving, she returned to HEI for the hookup or initial activation of her device.

"I didn't know what to expect or how many of my electrodes would function. There were six people watching to see if I would hear the beeping sounds being introduced by the audiologist," she remembers. And she did.

Karen sums up her ABI/PABI experience this way: "It has taken several years and has been a wonderful journey of relearning the sounds of everyday life. I am still hearing more each month. I just received a new processor and the sound quality is so much better than my old one," she beams.

"I am so grateful for the physicians and the technology that has allowed me to hear again. The research and development of new treatments for NF2

are being discovered every day. I want everyone who is living with this disease to know there is help available and so much hope for the future."

"The time right before my surgery and the year after were the most difficult. The loss of hearing along with the surgery and subsequent relearning to hear were very stressful. I was depressed and discouraged for a time and sought counseling for that year. It absolutely helped me, along with my faith, massage and any holistic practice I could find. It takes time and determination to accept that your life will be completely different but wonderful again," Karen shares.

Karen is completing a two-year term as vice-president and social chair of the Hearing Loss Association of Kentuckiana Chapter. She also serves on a Study Group Panel with the Kentucky Commission for the Deaf and Hard of Hearing (KCDHH).

"We are trying to introduce a House Bill in Kentucky to ensure that open captions or Rear Window™ captioning is made available in theaters to the entire state, now only available in our two largest cities. Last June, I and two other chapter members met with our Congressional Representative John Yarmuth of the Third District of Kentucky to ask him to co-sponsor HR 1646, the Hearing Aid Assistance Tax Credit Bill. He is a personal friend and after our meeting, he became the 89th Representative to sign on as co-sponsor. It made me realize," she adds, "the amount of work that goes into getting a House Bill to the floor and how much work still needs to be done." ■■■

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Barbara Liss Chertok is a freelance writer and a former lipreading/speechreading teacher. She lost her hearing suddenly at age 21 due to an autoimmune disorder and

is a bilateral cochlear implant user. She joined HLAA in 1979 and is active in Hearing Loss Association of Sarasota, Florida. She serves on the board of the American Hearing Research Foundation. She can be reached at barbchert@aol.com.

Talking with Karen Lichtefeld

By Barbara Liss Chertok

Has anyone else in your family had NF2?

No. My diagnosis of NF2 was a spontaneous occurrence as opposed to hereditary.

How difficult was the decision to go ahead with the implant?

It took me about a year to make the final decision after much prayer and meditation.

When your device was activated, what did you hear?

I heard mostly sounds and they were not very clear. But I was just so excited to hear anything after what I had been through. My audiologist, Steve Otto, turned on the water and it sounded like Niagara Falls. Speech took several months of concentration and lipreading to become clearer.

Now after three years, how is your speech comprehension with your implant?

The higher pitched voices of women seem to be easier for me to hear and I hear familiar voices better than someone I just met because I have the memory of

voice. In a quiet setting, I can comprehend about 90 percent of the conversation with lipreading.

How important are your lipreading skills in general?

My lipreading skills actually improved as I was losing my hearing and during the two silent months before my implant was activated. I still use lipreading daily and especially in noisy environments like restaurants, malls and large gatherings.

Can you understand conversation on the telephone?

I can't hear well enough on the phone to carry on a conversation so I use the CapTel® 800i at home and WebCapTel® with my cell phone.

[Note: The CapTel® 800i is a new model captioned telephone that provides captions for both outgoing and incoming calls and uses only one phone line.]

Which environmental sounds can you hear and which ones help keep you safe?

In my home, I can easily hear my phone, doorbell, coffeemaker and even when my central air conditioning is running. One morning last week, I asked my husband what that incessant sound was that I kept

hearing. My bedroom window was open and it was the crickets chirping!

In my car, I feel much safer being able to hear sirens, horns honking and my turn signal. When I walk in the park, I can hear birds, road noises, and people around me and even carry on a conversation when walking with someone.

Have you attended any formal auditory rehab sessions or practiced on your own?

I have attended several local seminars presented by Cochlear Americas. The practice drills—listening to someone read aloud and noticing sounds and memorizing them—have been very helpful. I have received many wonderful tips and encouragement from cochlear implant recipients in our HLA-Kentucky Chapter.

How does music sound with your implant?

I am still trying to relearn music, but it sounds more mechanical than melodic. The first songs I recognized after my implant were the *Star Spangled Banner* and the *Jeopardy* television jingle. That was exciting! 🎵🎵

Karen Lichtefeld can be reached at kalich@bellsouth.net.

Three Perspectives on Auditory Brainstem Implants

Auditory Brainstem Implantation *By Derald E. Brackmann, M.D.*

The auditory brainstem implant is placed following translabyrinthine or retrosigmoid craniotomy to remove an acoustic neuroma. The majority of the patients at the House Ear Clinic have been done via the translabyrinthine approach. In either case, following tumor removal, the auditory brainstem implant is placed.

The receiver stimulator is placed in a pocket above and behind the ear. The electrodes are then placed onto the brainstem. There is a naturally occurring opening called the lateral recess of the fourth ventricle. The cochlear nucleus lies just below this recess. The electrode is placed into this recess and then held in position with some Teflon felt material. The device is stimulated intra-operatively to make sure that it is in the correct position. The incisions are then closed in the usual manner. There have been no complications related to ABI placement.

Ten PABI surgeries were performed as a study. That study is now on hold because the penetrating electrodes did not appear to add significant benefit to the surface device.

To expand on the PABI, we were hoping that an electrode that penetrated into the cochlear nucleus would provide more specific pitch information at lower thresholds. There were a few patients who did demonstrate these characteristics but only on a few electrodes. We found it very difficult to correctly place the penetrating electrode into the small cochlear nucleus. Overall, the PABI patients did not do significantly better than the surface electrode patients and we therefore put the project on hold. 🎵🎵

Derald E. Brackmann, M.D., is Karen's surgeon. In addition to his work at the House Ear Institute, he is clinical professor of otolaryngology and neurosurgery, USC School of Medicine and clinical professor of otolaryngology, University of California, Irvine School of Medicine. His current projects include: co-principal investigator—Auditory Brainstem Implant; Management of Neurofibromatosis Type II; Management of Acoustic Neuromas; and Management of Skull Base Tumors.

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ABI Auditory Program *By Steven R. Otto*

The Nucleus ABI uses a pad-shaped surface electrode array approximately 2.5 by 8.5 mm in size made of silicone and dacron mesh with 21 platinum-iridium disk electrodes on it. This is connected to the regular Nucleus receiver/stimulator, basically the same thing that's used for cochlear implants (CIs).

The penetrating auditory brainstem implant (PABI), which we studied in 10 patients and is now on hold, had two different configurations. The first model had 14 surface electrodes and eight penetrating electrodes, and the second model had 12. The penetrating electrodes were needle-like and embedded in a silicone button about 2.5 mm in diameter. The penetrating electrodes lengths ranged from 1 to 2.5 mm. The purpose of the penetrating array was to obtain hearing at lower levels of current and with more focused current fields. We achieved both of these goals, and those electrodes improved speech recognition in combination with surface electrodes.

We hoped the penetrating electrodes would provide better speech recognition than the surface electrodes but we didn't see that. If we had been able to get a higher number of penetrating electrodes that gave hearing, we might have seen more of an effect in that regard. As it turned out, only about 25 percent of penetrating electrodes gave hearing, while more than 60 percent of surface electrodes gave hearing. It was more difficult to accurately target the precise location for implantation with the small needle electrodes, and being off by one mm could mean the difference between hearing and not hearing.

ABI recipients achieve substantial auditory benefit approximately 16 percent of the time, even allowing some recognition of words in sentences. About 9 percent of ABI recipients do not get any hearing sensations at all, getting only non-auditory sensations, such as tingling, dizziness, eye-related, etc. The most notable reason probably being related to neuro-anatomical difficulties.

ABI candidates are thoroughly counseled about what to expect from an ABI, including the fact that the ABI always works best in combination with lipreading cues, and that regular use of the ABI contributes greatly to long-term benefit. ABI recipients have shown the ability to continue to improve in performance for ten years or longer.

In Karen's program one, she has six surface electrodes active and two penetrating electrodes active. Altogether, there are 12 surface electrodes and 10 penetrating electrodes. Karen can use at least one penetrating electrode in combination with her surface electrodes, and she likes this configuration and has shown the best performance with it. This has been true of at least two other PABI recipients as well.

There are approximately 18 ABI centers in the United States. Cochlear Americas is the only company with an ABI available in the United States. Med-El produces an ABI that is marketed in Europe only. Advanced Bionics does not produce an ABI at this time. 

Steven R. Otto, M.A., is Karen's audiologist and has been at the House Ear Institute (HEI) for 21 years. An advanced research associate at HEI, he has the major responsibility for the day-to-day management of the ABI program.

PABI: The Auditory Brainstem Implant with Penetrating Microelectrodes

By Robert V. Shannon, Ph.D.

The original auditory brainstem implant (ABI) was developed at the House Ear Institute (HEI) in 1979 to provide hearing sensations to patients who could not benefit from a cochlear implant (CI). These patients have bilateral acoustic tumors caused by a genetic disorder called Neurofibromatosis Type II (NF2). When the tumors are removed the auditory nerve is usually cut and so there is nothing for a cochlear implant to stimulate.

The ABI is like a CI, but the electrode is designed to be placed on the surface of the brainstem to activate a structure called the cochlear nucleus, which is the first relay station for hearing information on the way to the brain. Patients with the original ABI could hear different pitch sensations on each electrode and generally found the device to be useful in everyday life as an aid to lipreading and for awareness of environmental sounds.

Research groups at HEI and the Huntington Medical Research Institutes (HMRI) in Pasadena, California, received a contract from the National Institutes of Health to develop a new generation of ABI to improve performance. The new design, developed in collaboration with Cochlear Americas included penetrating microelectrodes (PABI) to improve the selectivity of the stimulation. Both cochlear implants and the original ABI stimulate neurons with a broad electrical field that activates a broad area of neurons. The PABI electrodes could activate a very small cluster of neurons and should theoretically produce clearer pitch sensations and higher pitch sensations than the surface electrodes.

After more than 15 years of development and safety testing we initiated clinical trials of the PABI in NF2 patients. We achieved good pitch sensations on some penetrating electrodes, but this improvement does not seem to improve overall speech recognition. The project is on hold while we evaluate the outcomes in the first 10 patients. In the meantime, better speech recognition was observed in Italy when the ABI was used in patients who did not have NF2 (Colletti and Shannon, 2005).

Some of those non-NF2 patients were able to hear on the telephone like CI patients. This surprising result suggests that the limitation in ABI and PABI performance is due to NF2 and not to the design or placement of the ABI electrode. We continue to do research to improve the ABI device and outcomes. 

Robert V. Shannon, Ph.D., is head, Auditory Implant Research, at the House Ear Institute.