The First ABI at the BNI: Case Report and Literature Review

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First introduced in 1979 by Drs. House and Hitselberger, auditory brain stem implants are capable of providing useful hearing for a patient suffering from bilateral retrocochlear deafness. The initial design of the ABI featured a single-channel ball-type electrode. ABIs have since evolved to include between 8 and 21 electrodes. In the year 2000, the Food and Drug Administration approved a multichanneled ABI for the treatment of neurofibromatosis type 2. Currently, more than 400 ABIs have been implanted throughout the world, including in the United States, Australia, Germany, Italy, the United Kingdom, Poland, and Croatia. Sound-only open-sets speech comprehension has been reported in a minority of patients who have received ABIs, but most patients derive maximum benefit from using the implants as an adjunct to lip reading. In this regard, ABIs perform as well as single-channel cochlear implants, on which their design has been based. We present our initial experience with ABIs and review the literature.

Key Words: acoustic neuroma, auditory brainstem implant, auditory prosthesis, neurofibromatosis type 2, vestibular schwannoma

Abbreviations Used: ABI, auditory brainstem implant; BCI, brain-computer interface; CT, computed tomography; FDA, Food and Drug Administration; MR, magnetic resonance; NF 2, neurofibromatosis type 2

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Much of the emphasis in current research on BCI technology is on restoration of neurological function. BCI technologies intended to extend human capabilities may have appeal for military applications. Thus far, however, all available examples of this technology have been developed with the goal of ameliorating the symptoms of disease, such as implantation of deep brain stimulation electrodes for the treatment of Parkinson's disease.

Of special interest is the potential for these devices to restore special sensory modalities, in particular, vision and hearing. Efforts to obtain viable prosthetic vision are still in their infancy.[6] In contrast, ABIs are available technology that provides prosthetic auditory information directly to the cochlear nucleus at the pontomedullary junction level of the brain stem. Specifically, in the year 2000, the FDA approved multichannel ABIs for use in patients with the diagnosis of neurofibromatosis type 2 (NF 2) who are 12 years of age or older and who have reasonable expectations about the device's utility.

At least five models of ABIs, including the Nucleus 24 Contour and Nucleus 22 ABIs (Cochlear Corporation, Englewood, CO), the MXM Digisonic ABI (Laboratoires MXM, Côte d'Azur, France), the Med-El ABI (Med-El Corporation, Research Triangle Park, NC), and the Clarion ABI (Advanced Bionics, Sylmar, CA), are available. To date, no evidence suggests superior performance of any particular model of ABI.

Regardless of the manufacturer, all ABIs share certain common features, including a digital speech processor, transmitter coil, and a receiver-stimulating unit that is connected by cable to an electrode array of 8 to 21 electrodes overlying the cochlear nucleus. The Nucleus 24, the most widely implanted device, features an electrode array of 21 platinum discs embedded in a Dacron mesh. The device is placed within the lateral recess of the fourth ventricle. The electrode is connected by a cable to a receiver stimulating unit, which itself is seated in the occipital region of the skull. An additional wire serves as a ground electrode and is implanted in the temporalis muscle. Therefore, the entire device is subcutaneous and communicates with the digital speech processor through a radio transmitter coil. This second component includes a microphone, which digitizes incoming sound, processes speech, transmits this digital signal through the radio transmitter coil to the receiver stimulator located underneath the scalp, and then transmits it from there to the brain stem.

Given the various number of channels that patients can use to interpret auditory stimuli, the utility of these devices relies on cooperation and training for the successful interpretation of the auditory input. With the Cochlear digital speech processor, the patient can adjust the speech processor through four user-selectable programs and can control the volume and sensitivity.

Indications

The FDA has approved ABIs for patients with NF 2 who are 12 years of age or older and who have reasonable expectations and motivation. NF 2 has an incidence of 1 in 40,000. The phenotype is associated with an autosomal dominant mutation of the merlin gene on chromosome 22, resulting in bilateral vestibular schwannomas. This latter feature is pathognomonic for the diagnosis. Vestibular schwannomas associated with NF 2 are more aggressive or more likely to invade the vestibulocochlear nerve than nonsyndromic schwannomas.

An additional set of characteristics establishes a patient as an optimal candidate for an ABI: good overall state of health, acceptable vision, an interest in spoken communication, and acceptable anatomical features. Vision is important because in terms of speech comprehension, the greatest utility is typically derived from using the ABI as an adjunct to lipreading. Perhaps most importantly, however, patients must exhibit high motivation and have sufficient family support.

Given the largely successful experience with ABI in the setting of NF 2, it is not surprising that the indications for ABI placement have evolved to include other conditions, including unilateral vestibular schwannoma (i.e., nonsyndromic) in an only hearing ear,[2] deafness as a result of bilateral skull base trauma,[1] cochlear nerve aplasia or hypoplasia,[4] and cochlear ossification.[7] In theory, any cause of bilateral retrocochlear deafness may suffice as an indication for implantation of an ABI. To date, however, experience with ABI placement for indications other than NF 2 is limited to Europe. The only absolute contraindication to the placement of a cochlear implant is the presence of active infection.

Case Report

A 56-year-old male was diagnosed with NF 2 at 21 years of age. The patient underwent a translabyrinthine resection of the vestibular schwannoma on the left side 20 years earlier at an outside institution. At the time of his evaluation at our institution, he had experienced progressive hearing loss on the right side. His medical history was significant for insulin-dependent diabetes mellitus, peripheral neuropathy related to the diabetes, and seizure disorder. His current medications included Neurontin, OxyContin, insulin, Skelaxin, omeprazole, and Capitrol. He also complained of difficulties with swallowing and clearing secretions and occasional bouts of urinary and bowel incontinence.

Physical examination revealed myoclonus, dystonic posturing of fingers, weakness of the intrinsic hand muscles, and bilateral foot drop. The patient's vision was excellent. He wore a hearing aid in the right ear and depended on lip reading for communication.

Preoperative audiography demonstrated an elevation of speech awareness threshold to 90 dB and a pure-tone discrimination threshold elevation of 106

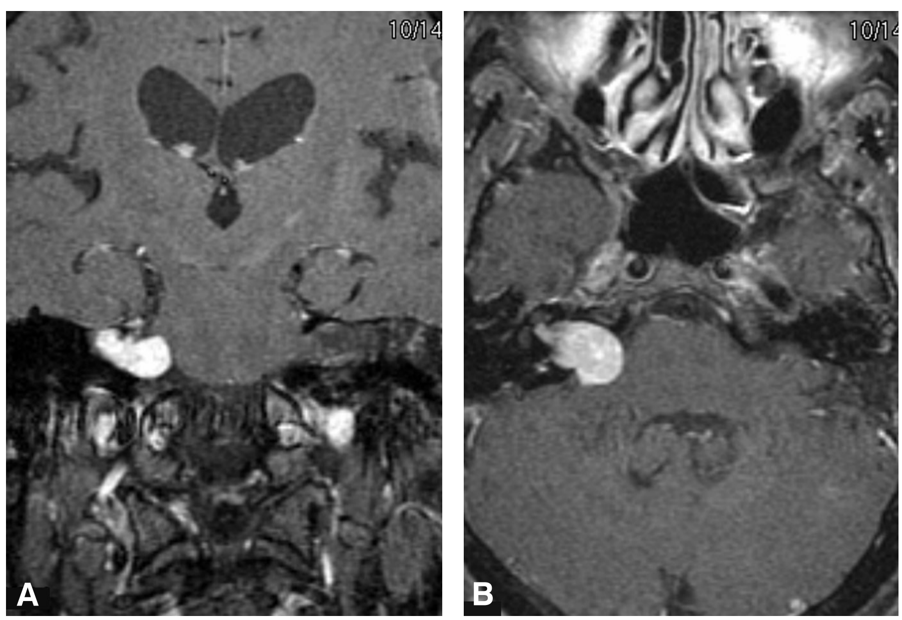
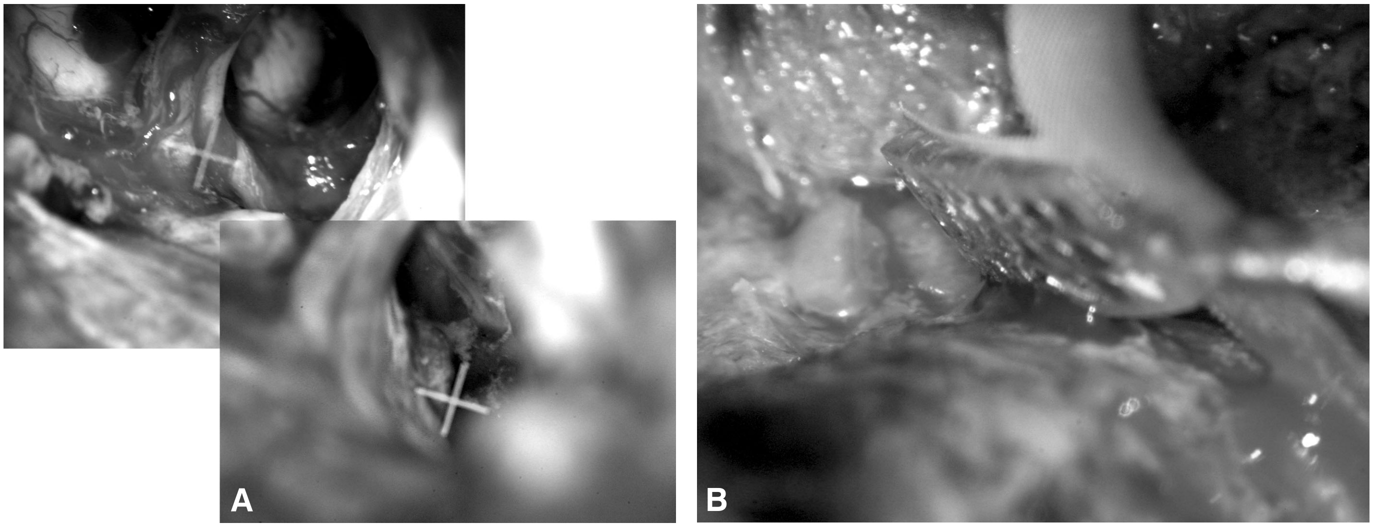


Figure 1. Preoperative (*A*) coronal and (*B*) axial contrast-enhanced MRIs show a homogeneously enhancing tumor of the right cerebellopontine angle with an 'ice-cream cone' profile characteristic of a vestibular schwannoma.



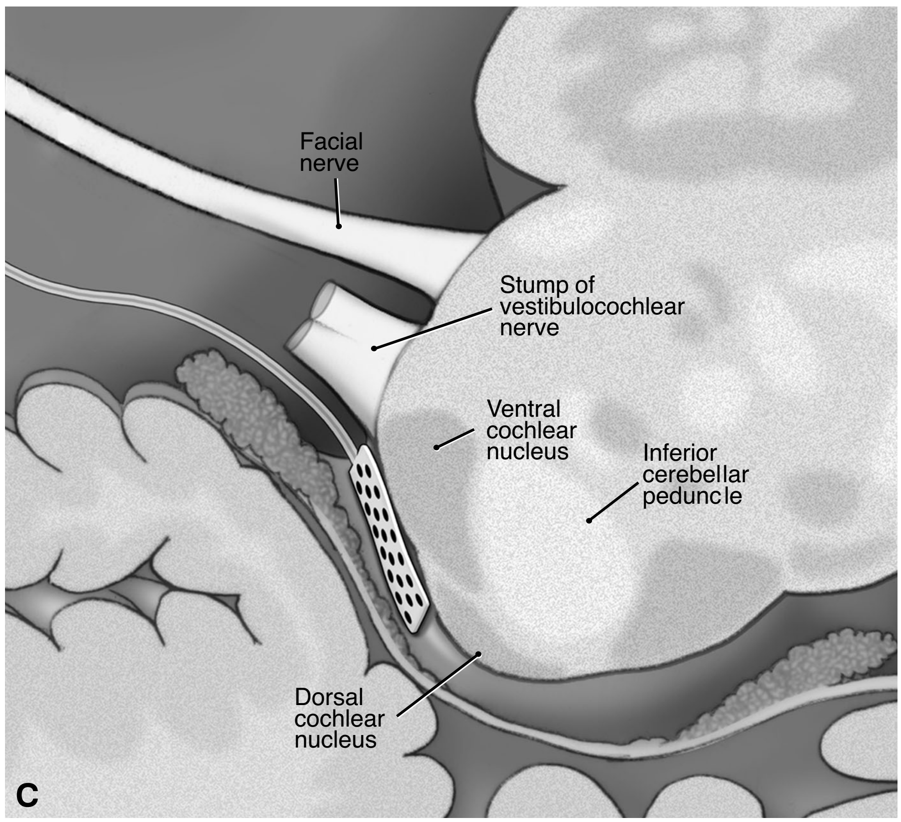


Figure 2. (*A*) Intraoperative photographic montage shows the seventh and eighth cranial nerve complex (*upper crosshairs*) and the ninth cranial nerve (*lower crosshairs*). (*B*) Intraoperative photograph of electrode array being advanced into the lateral recess of the fourth ventricle. (*C*) Schematic illustration of an ABI electrode in the lateral recess of the cochlear nucleus.

dB. Preoperative imaging (Fig. 1) showed a 13 x 20 x 12 mm acoustic schwannoma on the right side and surgical changes on the left side without evidence of tumor (the site of previous surgery). Comparison with MR images obtained 2 years earlier demonstrated modest tumor growth (previous size 12 x 15 x 10 mm).

The patient was offered the following options: (1) continued observation, (2) radiosurgery, or (3) resection of tumor with anticipated loss of hearing and placement of an ABI. During preoperative counseling, it is important to stress the risks, benefits, and alternatives to surgery. Deafness is likely to be universal in neurofibromatosis, but not all patients are ideal candidates for an ABI. Given that our patient was already anacoustic in the left ear, there was no chance of maintaining useful hearing regardless of treatment modality (i.e., surgery or radiosurgery). We believed that the patient's best chance of retaining useful hearing on the right side after resection of the tumor was an ABI. The patient elected to undergo surgical resection of the tumor and placement of an ABI. Informed consent was obtained.

The patient was positioned supine with his head turned left to expose the right ear. A standard translabyrinthine approach to the tumor was performed. The tumor was resected in the typical fashion. When resection of the tumor was completed, the facial nerve, the stump of the vestibulocochlear nerve at its entry to the brain stem, and the glossopharyngeal nerve were identified (Fig. 2). The lateral recess of the fourth ventricle was identified by following the tenia of the fourth ventricle and a small tuft of choroid plexus emerging from the foramen of Luschka. The implant was then advanced within the foramen of Luschka along the pontomedullary surface (Fig. 2B).

Despite the presence of significant signal artifact from the ABI (Fig. 3), postoperative MR imaging confirmed

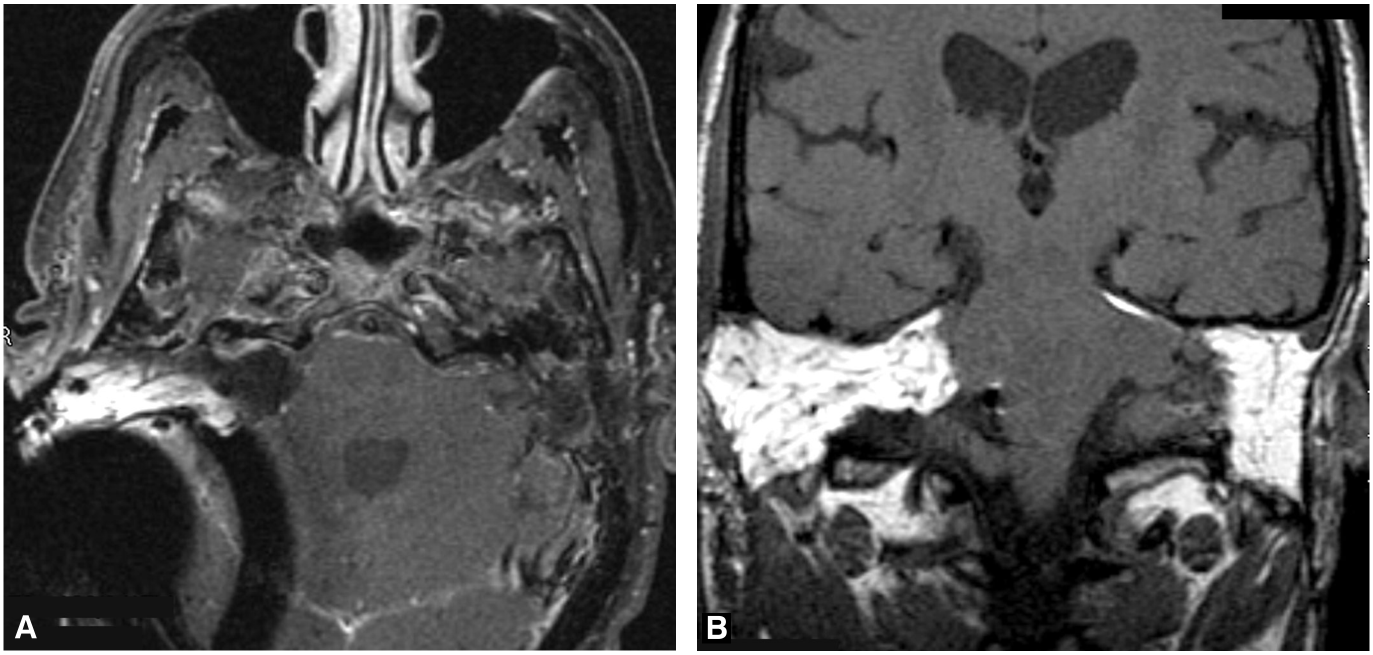


Figure 3. Postoperative (*A*) axial and (*B*) coronal MR images confirm complete removal of the tumor and show the fat packed in the translabyrinthine approach defect. The artifact on the images is caused by the ABI receiver-stimulator.

complete resection of the tumor with fat packed into the translabyrinthine defect. CT demonstrated satisfactory placement of the electrodes (Fig. 4). Pathological analysis confirmed the diagnosis of a typical vestibular schwannoma.

Approximately 8 weeks after surgery, the patient's ABI was turned on. During the initial programming with stimulation of a single channel, he reported the reception of auditory percepts on four channels. At his 15-month follow-up examination, the patient underwent 10 pitch-ranking maps. He is a daily user of his ABI, aware of environmental noises, and able to distinguish his wife's voice.

At the initial programming of the ABI, the patient had no auditory-only channels and auditory percepts were associated with a shock-like sensation. With subsequent programming, this nonauditory percept was eliminated. Formal speech-perception testing, as assessed by the Hearing in Noise Test presented via live voice, revealed speech comprehension rates of 12% (auditory only condition), 16% (visual only), and 40% (auditory + visual). Importantly, both the patient and his wife have noticed dramatic improvement in his hearing when he uses the device and at 24 months follow up were pleased with the results.

Discussion

In the English language literature, more than 180 patients with an ABI have been reported.[3,5,9,10,12] The largest single series is from the United States Clinical Trial,[5] which enrolled 92 patients whose average age at implantation was 33.9 years (range, 12.7 to 67.5 years). In two-thirds of the patients, their ABI was placed at the time of a second surgery. One-third received their ABI upon resection of their initial schwannoma (i.e., in the presence of a hearing ear on the contralateral side). Fifty-nine percent of the patients were female and 41% were male. Two patients died before the initial activation of their implants. Of the 90 remaining patients, 74 received auditory percepts upon stimulation (82.2%).

At the time of initial stimulation, most patients also experienced nonauditory sensations. The most common site for nonauditory percepts included the ipsilateral head and neck followed by the ipsilateral torso and upper and lower extremities (60%, 14%, 10%, and 11%, respectively). Although rare, the rate of contralateral percepts was 6%. Approximately 10% of patients received auditory sensations only, and another 7% received nonauditory percepts only. The origin of nonauditory percepts is attributed to the proximity of distal electrodes to the inferior cerebellar peduncle. In general, the character of nonauditory sensations has typically been described as tingling, dizziness, visual jittering, or muscle twitching.

In terms of improved speech comprehension, the results from the United States Clinical Trial mirror the percentage of patients who received auditory percepts. Eighty percent reported that they benefited from use of the device, and 75% reported wearing the device daily. When used in conjunction with lip reading, the ability of 85% of the patients to understand sentences after 3 to 6 months significantly improved based on the City University of New York (CUNY) score. Twelve percent of patients demonstrated clinically significant open-set sentence recognition in the auditory-only condition.

As recently reviewed,[8] these results have been replicated both domestically

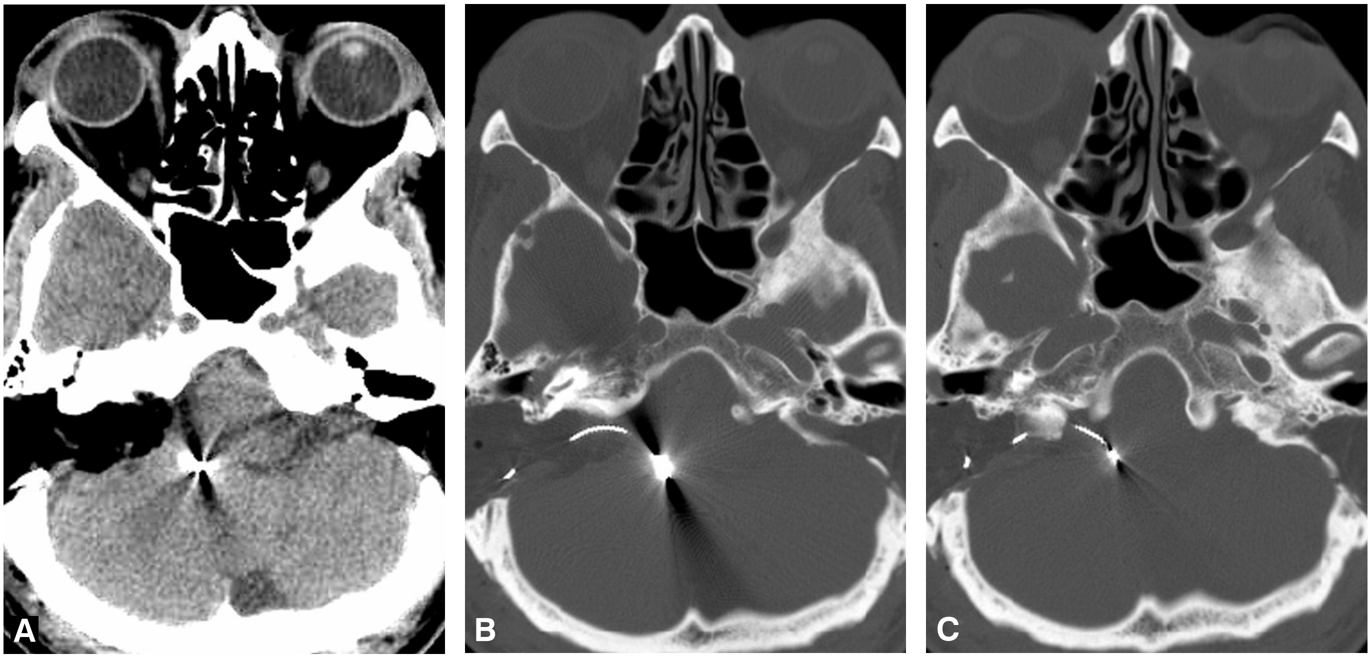


Figure 4. (*A*) Soft-tissue window and (*B*) bone window CTs of the head obtained immediately after surgery and (*C*) on the day of initial stimulation of the ABI confirm that the device did not migrate during the follow-up interval.

and internationally, including by the Digisonic Clinical Trial reported in the journal, *Otology Neurotology,* in 2002[12] and by European centers, including Verona,[3] Hannover,[9] and Freiburg.[10] The percentage of patients receiving auditory sensations ranged from 86% in the Digisonic study to 100% in the Hannover and Verona studies. Between 75 and 100% of patients were reported as daily users (the Digisonic trial and Verona study, respectively). There appears to be no association with the number of electrodes and efficacy of the device. The United States Clinical Trial was performed with the 8-electrode Nucleus 22 model. European data have been reported with either 14- or 21-electrode models. However, the results associated with the different devices have not varied significantly.

Preoperatively, it is important to counsel patients not to be discouraged or disappointed with the unnatural quality of the sound associated with their ABI. The sound has been likened to a muffled loudspeaker. However, the results of audiologic testing have continued to improve as long as 8 years after implantation. With appropriate expectations and patient cooperation, ABIs restore meaningful hearing to most patients. Based on the long-term follow-up from the experience at the House Ear Institute,[11] 87% of patients exhibited scores on closed-set testing of word recognition above the level of chance. Environmental sound discrimination was more than 50%, and sentence recognition scores improved by 26%. Importantly, even when sound-only open-set speech recognition is not obtained, patients benefit from the perception of environmental sound and their ability to read lips improves significantly.

Conclusion

Future designs of ABIs have centered on the development of the penetrating brain stem implant. However, initial results with this implant have been disappointing (Hitselberger W, personal communication, 2005). These results, which may reflect the development of perielectrode scar tissue, may not portend well for the use of other penetrating BCI strategies. Alternatively, the limited performance of the penetrating ABI may reflect a reliance on stimulation of deep cochlear nuclear structures, while providing enhanced tonotopic representation of pitch theoretically may diminish speech-recognition capability. Research into speech-processing algorithms may be another promising avenue for improvement of speech recognition. However, caution must be exercised in the exposure of the brainstem to charge. Hence, the ranges by which these parameters can be safely modulated may be limited.

As our experience with ABIs increases, innovations in electrode design and our improved understanding of plasticity in the ascending auditory pathway will be accompanied by improvments in the performance of these devices. Nevertheless, at present, ABIs truly reflect the state of the art of BCI technology.

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