Hearing restoration with auditory brainstem implants after radiosurgery for neurofibromatosis Type 2

Report of three cases

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The auditory brainstem implant (ABI) is designed to restore useful auditory sensations in patients with neurofibromatosis Type 2 (NF2). The implantation is usually performed at the time of tumor removal in patients who do not undergo radiation treatment. The authors evaluated the performance of ABIs in three patients with NF2 in whom vestibular schwannoma continued to grow after radiation treatment. These three patients with NF2 received a 21-channel ABI; a translabyrinthine approach was used for both the tumor removal and the ABI placement. The interval between radiosurgery and the tumor removal plus device implantation ranged from 2 to 11 years. In all cases, the tumor was growing and the patients presented with total deafness. The mean number of active electrodes in these three patients was equivalent to the average results reported in other patients who received ABIs. The patients in this study used the ABI regularly for everyday life and obtained useful levels of environmental sound recognition. It is concluded that hearing function can be rehabilitated using ABIs in patients with NF2, even if radiosurgery fails to control the tumor growth.

KEY WORDS • acoustic neuroma • neurofibromatosis Type 2 • auditory brainstem implant • vestibular schwannoma • radiosurgery

Abbreviations used in this paper: ABI = auditory brainstem implant; CPA = cerebellopontine angle; EABR = evoked auditory brainstem response; GKS = gamma knife surgery; LINAC = linear accelerator; MR = magnetic resonance; NF2 = neurofibromatosis Type 2; VS = vestibular schwannoma.

Until now, it has been thought that ABI placement should not be considered after a radiosurgical procedure, because Slattery and Brackmann21 have reported the absence of auditory response after placement of this device, which was associated with poor postoperative facial nerve function after radiosurgery in two cases. These failures may be related to the degenerative effect of irradiation on the structure of the cochlear nuclei, which reduces the possibility of electrical induction of auditory sensation. In this work, we report on auditory function in three patients with NF2 who received implants after radiosurgery and who had uncontrolled VS growth and/or total hearing loss.

Clinical Material and Methods

Patient Population

This report includes three French-speaking patients with NF2 who received 21-channel ABIs (Nucleus; Cochlear Corp., Lane Cove, Australia). All three patients were considered to be medically and psychologically suitable for the investigational protocol of the Nucleus ABI European clinical investigation.10 Two patients acquired good lip-reading skills before implantation. All patients received the implant during the second tumor removal; these tumors had initially been irradiated to preserve hearing in the other ear. The delay between irradiation and ABI placement ranged...
from 2 to 11 years. Table 1 summarizes the general characteristics of these patients. Postoperative facial nerve grades were assigned according to the House and Brackmann system.6

**Auditory Brainstem Implant**

The 21-channel ABI is based on the Nucleus 22-channel cochlear implant system (Cochlear Corp.). It contains a receiver/stimulator package, an electrode lead, and an electrode array. This array is composed of 21 platinum electrodes arranged in three rows of seven pads in a silastic carrier (3 × 8 mm). The electrode diameter is 0.7 mm. A Dacron mesh is attached to the electrode carrier, forming a T to prevent movements after the placement of the device. The 11-cm-long electrode lead is made of a specially compressed microcoil. The implanted receiver/stimulator is provided with a removable magnet that can be replaced through a silicone plug after application of local anesthetic. This allows patients to undergo MR imaging when necessary. The external parts of the device include an antenna, microphone, and processor (Fig. 1).

**Brainstem Implantation Procedure**

The translabyrinthine approach was used in all cases because it provided direct access to the lateral recess and the surface of the cochlear nuclei along anatomical landmarks: the seventh and eighth cranial nerves and the choroid plexus, confirmed by the outflow of cerebrospinal fluid with a Valsalva maneuver.13,14,26 Electrically evoked auditory brainstem responses (EABRs) and the facial and glossopharyngeal nerve activities were monitored by surface electrodes during surgery. For EABRs, a conventional auditory brainstem response recording system (MIPC; Bio- digital, Paris, France) was used in combination with the diagnostic programming system (Cochlear Corp.) in which the ABI functions as an electrical stimulus generator. For the facial and glossopharyngeal nerves, a continuous electromyographic monitoring system (NIM II; Xomed, Jacksonville, FL) was used. Possible electrical stimulation of other cranial nerves was minimized by this monitoring.

The appropriate placement of ABI electrodes for the activation of the auditory system was verified intraoperatively by using the EABR recordings.13,14,26,27 Typical EABRs after ABI stimulation were characterized by two or three positive peaks within the first few milliseconds after the stimulus.27

**Processor Fitting, Programming, and Speech Perception Evaluation**

A computerized tomography scan was obtained the day before the device was activated to evaluate the correct localization of the electrode in the lateral recess. The activation was performed 6 weeks postoperatively and because of possible risks associated with the stimulation of brainstem structures, it was performed in the critical care unit while the patient underwent electrocardiographic monitoring. Threshold and maximum comfort levels were assessed for each electrode to select the optimal electrode configuration. Channels producing unpleasant sounds or nonauditory effects were inactivated. Two electrodes were stimulated in a random sequence, and the patient was instructed to determine if the first or second sound was higher in pitch. After assessment of all pairs, the channels were arranged in an appropriate tonotopic order.

During the 1st postoperative year, speech perception and auditory training was conducted by the speech therapist three times per week. Reprogramming and speech therapy evaluation were performed every 2 weeks during the first 3 months, every 3 months during the 1st year, and annually thereafter. The speech perception evaluation main-

![Fig. 1. Schematic representation of the ABI device comprising the implanted part and the external part (processor, microphone, and antenna). Inset A shows the position of the electrode pad in the lateral recess, and inset B depicts the electrode array.](image-url)
ly included recognition of environmental sounds, vowels, consonants, monosyllabic words, disyllabic words, and sentences without context (open sentence). The implant’s microphone can be turned off by the patient or the speech therapist during the tests. All tests for word and sentence recognition were performed in sound-only mode (implant turned on), sound plus vision mode (implant turned on), and vision-only mode (implant turned off). Scores obtained in each condition were compared to define lip-reading enhancement and device-only benefit. Although no formal assessment was made of environmental awareness, on the performance questionnaire the ABI recipients rated environmental awareness and communication abilities with familiar speakers in quiet settings as the most significant benefits received from the device.

Case Reports

Case 1

This 49-year-old woman presented with a right-sided VS in 1979. This lesion was removed at another center via a suboccipital approach, resulting in a Grade 5 postoperative facial nerve function. The diagnosis of mild NF2 was established by the discovery of a left-sided VS in 1987. This lesion was irradiated in a single fraction by using the stereotactic LINAC procedure. Data concerning the radiation dose were not available. In 1994, the patient was referred to our department for fluctuating left-sided hearing loss that required internal auditory meatus decompression via a middle fossa approach. Subsequently, a high-powered hearing aid was used in the left ear for 2 years. From 1987 to 1997, the size of the left-sided VS was stable on successive MR images (Fig. 2 left). In 1998, the patient developed total deafness in the left ear due to rapid growth of the previously irradiated tumor (Fig. 2 center). She quickly acquired a good lip-reading ability. The left-sided VS was removed via a translabyrinthine approach, followed by ABI placement on the left side (Fig. 2 right). The dissection of facial nerve from the tumor was performed easily. Histological examination of the tumor demonstrated a typical VS. The patient had Grade 2 facial nerve function 1 year postsurgery.

Case 2

This 55-year-old woman presented with a diagnosis of mild NF2 with bilateral VS and multiple cervical schwannomas made in 1978. Several operations were performed to remove the right-sided VS and multiple cervical schwannomas. In 1983, she became deaf bilaterally and acquired a good lip-reading ability. The left-sided VS (12 mm, located in the CPA) was irradiated in 1996 by using GKS. The mean radiation dose to the tumor margin was 11 Gy. Despite GKS, the VS continued to grow at a rate of 4 mm per year (Fig. 3). In 1998, the patient was referred to our department and underwent surgery via the translabyrinthine approach, with total tumor removal and ABI placement on the left side. The facial nerve dissection was difficult because it was surrounded by scar tissue and there was no arachnoid plane. Despite moderate arachnoiditis, the electrode plate was successfully inserted into the lateral recess.
Auditory brainstem implants after radiosurgery for NF2

The histological findings corresponded to a typical VS. The postoperative period was uneventful, and postoperative facial nerve function was classified as Grade 5 at 1 year.

Case 3

This 17-year-old woman presented with a diagnosis of severe NF2 made in 1989. In 1996, a large left-sided VS was removed via a translabyrinthine approach and a right-sided VS (20 mm, located in the CPA) was irradiated using GKS with 12 Gy on the 50% isodose line. Her hearing loss worsened slowly on the right side to the point of deafness the 2 years following GKS. Meanwhile, the patient did not develop satisfactory lip-reading skills. On successive MR images obtained after GKS, the right-sided VS increased slightly in volume (Fig. 4). The patient was referred to our department and underwent surgery via a translabyrinthine approach, with total removal of the lesion and ABI placement on the right side. The immediate postoperative period was uneventful; however, the patient had a skin necrosis over the implant’s receiver–stimulator device 4 months after the ABI was placed, which required two surgical repairs in which musculocutaneous flaps were used. The ABI setup was delayed until 12 months postsurgery. At this time, postoperative facial function was classified as Grade 4.

Auditory Results

Intraoperative auditory responses were satisfactory in the first two cases and poor in the last one. Active electrodes ranged from five to 14. Nonauditory effects included mainly dizziness and tactile sensations in the face, auricle, arm, shoulder, and the leg (Table 2).

Figure 5 provides a summary of the last follow-up results 2 years after the device was first switched on. All three patients reported a benefit for speech perception and said that they used the ABI regularly for ordinary life. All patients reported improved environmental sound awareness on the questionnaires and showed improved lip-reading ability during the tests in sound-only compared with sound plus vision modes. One patient achieved functional open-sentence speech understanding. This patient achieved speech recognition in sound-only mode (with no possibility of lip reading) when using the implant, in comparison with no ability when the implant was turned off.

Discussion

Most patients with NF2 who have bilateral VSs will experience total deafness as a result of the progression of the tumor and/or surgical procedures. Large tumors (> 3 cm) that are located in the CPA and compress the brainstem are treated by resection. The goal of the treatment in these cases is the preservation of life and neurological function, including the facial nerve function, but not hearing.19 In only a few patients with NF2 who had small lesions could useful hearing be preserved after tumor removal.19,22

The alternative treatment for small VSs (< 2 cm) in the CPA is stereotactic radiosurgery by either LINAC or GKS.8,17,18,24-25 The advantage of these treatments is the avoidance of surgery, with facial nerve function preserved in 80 to 90% of cases.8,16,17,23,24 Moreover, the rate of hearing preservation, as high as 33 to 43% in patients with NF2 who were treated using these procedures, appears interesting despite some deterioration during the ensuing 6 years.3,7,8,17,18,24

The main criticism about radiosurgery, however, is the unknown long-term overall tumor control rate. In current studies a 98 to 100% tumor control rate is reported with a median follow-up period of only 3 to 7 years.8,17,18,24,25 Our first case demonstrates possible tumor evolution long after radiation treatment by LINAC. Indeed, the tumor was stable for 11 years after radiosurgery, and this appears to be a success from the radiosurgical point of view. Nevertheless, resumption of the tumor growth led to a total hearing loss. This hearing deterioration was probably delayed by the internal auditory meatus decompression completed via a middle fossa approach.

Another reported criticism of radiosurgery is the fact that the microsurgical resection of a VS after radiation treatment is technically difficult, with poor cranial nerve preservation.1,16,22 A frequent intraoperative finding in these cases is tumor adherence to the facial nerve, with scar tissue and no identifiable arachnoid planes. This was observed in the two cases of VS in which GKS had been performed previously. In these cases, surgery yielded a lower postoperative facial function grade than that usually obtained by the same surgical team for nonirradiated VS of the same volume.22

The ABI placement was successful in all of our patients.
The results indicate that accurate placement of the electrodes in the lateral recess is possible in patients who have previously undergone radiation treatment. The intact facial nerve may serve as a guide to locate the lateral recess but it should be carefully dissected to avoid postoperative deterioration of facial nerve function. In a case of ABI placement after radiosurgery, no arachnoiditis was observed in the lateral recess and the electrode was placed without difficulty. Intraoperative electrical monitoring was helpful for proper placement of the electrode array. The EABRs generated by the ABIs were important for the confirmation of an accurate intraoperative placement of the electrode in two patients.

The original feature of this report is the ABI placement in three patients who were previously treated by radiosurgery, which had resulted in a lack of tumor control and/or total deafness. All three patients reported improved speech perception when using their ABI regularly for everyday life. The global auditory performance in these three patients who had previously undergone radiosurgery was slightly lower compared with our other patients who received ABIs and the other reports of patients in whom the devices were implanted during the second tumor removal. All patients showed environmental sound awareness and improved their lip-reading ability. The benefit from environmental awareness is likely related to the reduced sense of social isolation related to profound or total hearing loss. The ABI improved lip reading by yielding information on speech rhythm and intonation. In our first reported case, the functional results were better than in the other two cases because the first patient gained the ability to communicate in a sound-only mode.

Several factors may influence the results and make comparisons with other patients difficult. Factors such as the duration of deafness (Case 2, 15 years of deafness) and the preoperative level of lip-reading ability (Case 3) are dependent on the patient. Others depend on anatomical configuration such as the accurate placement of the electrodes in contact with cochlear nuclei. In fact, the auditory performances with ABIs after radiosurgery were not related to the number of active electrodes: the mean number of active electrodes in our three patients was equivalent to that recorded in the Nucleus ABI European Clinical Investigation (9.3 compared with 8.6, unpublished data). A degenerative effect of irradiation on the structure of the cochlear nuclei might account for the lower performances in our patients, but still an ABI is an irreplaceable aid for these disabled patients.

Conclusions

Placement of an ABI is feasible after stereotactic radiosurgery of a VS. The results, however, show lower ABI efficiency than that achieved in patients who received implants at the time of the second VS removal.

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