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Surgical experiences in 58 cases using the Nucleus 22 multichannel auditory brainstem implant

W.-P. Sollmann, R. Laszig*, N. Marangos*

Abstract
Patients with bilateral total deafness due to lesions of the vestibulocochlear nerve can be treated by electrical stimulation of the second auditory neuron. A 22-channel multi-electrode implant with transcutaneous transmission was developed that allows the selection of the most useful electrodes. Acoustic neuromas were removed from 49 out of 58 patients by ENT surgeons and neurosurgeons working in collaboration and using either a translabyrinthine or suboccipital approach. The central electroauditory prosthesis was implanted in the same procedure. Six patients were deaf after previous acoustic neuroma surgery without recurrence, three had diagnoses other than neurofibromatosis type 2 (NF2). There were no complications due to the implantation procedure. Side effects could be excluded by stimulation of the auditory electrodes alone. Most of the patients used their ABI daily. They reported perception of different sounds and frequencies, enhancement of lip-reading ability, and three of the patients were able to use the telephone.

Key words: Neuroma, Acoustic; Brain Stem; Prosthesis Implantation; Electric Stimulation; Cochlear Nucleus; Deafness; Neurofibromatosis 2

Introduction

Patients with bilateral total deafness due to lesions of the vestibulocochlear nerve can be treated by electrical stimulation of the second auditory neuron. An auditory brainstem implant (ABI) was first successfully carried out in 1979 by House and Hitselberger,[1–3] after Simmons *et al.* had failed in stimulating the inferior colliculus.[4] The first multichannel ABI was implanted by Laszig and Sollmann in 1992.[5,6] For successful ABI surgery, there are some important issues to respect: patient selection, choice of device, choice of approach and technique of tumour removal, knowledge of the micro-anatomical variations, intra-operative identification of the cochlear nucleus, and prevention of complications.

Indications

The indication for ABI is bilateral neural deafness, especially in NF2-patients with bilateral acoustic neuromas. The best candidates are young patients with small or moderately-sized bilateral acoustic neuromas, who have no other significant problems from their NF2 disease and a long life expectancy (Figure 1). These patients recover very fast from their AN surgery, and the ABI is no additional surgical risk. They are usually highly motivated, intelligent and have an active social environment.

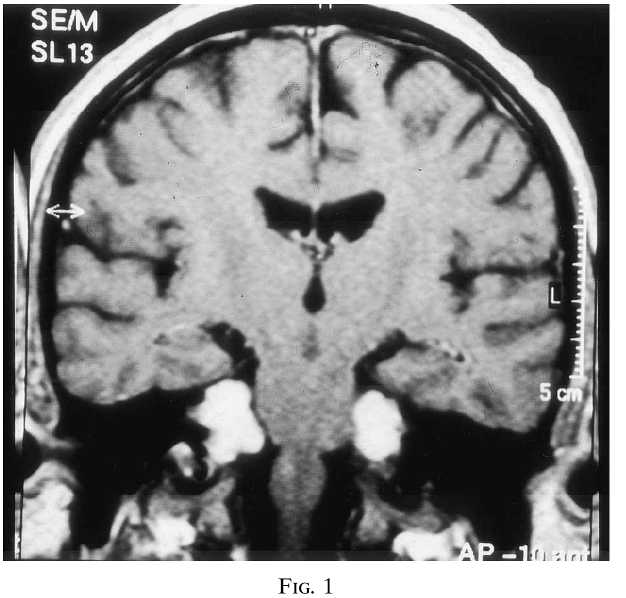

Fig. 1

Bilateral acoustic neuromas in a patient with neurofibromatosis type 2. Although there is contact with the pons, the pontomedullary junction with the cochlear nucleus is free of tumour.

We have had similar good results in patients having no tumour recurrence and well-preserved anatomy after previous AN surgery. Because ABI surgery has been free of complications so far, we can

From the Neurochirurgische Klinik, Städtisches Klinikum Braunschweig, Braunschweig, and the *HNO-Klinik, Universität Freiburg, Freiburg, Germany.

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FIG. 2

Intra-operative view from suboccipital approach; patient with axonal neuropathy. Cranial nerves VII to X are exposed. The auditory brainstem implant has to be positioned in the direction of the arrow following the cochlear nucleus vein. The AICA-loop is dissected free in the lateral recess and positioned on the back of the implant.

recommend it even for patients free of tumour. A longer-lasting period of deafness is no contra-indication – we had several patients with excellent results after 14–16 years of hearing loss.

One 17-year-old boy suffered from a congenital axonal neuropathy. His ear had completely normal anatomy, and the exposure of the landmarks at the cochlear nucleus and the electrode implantation was optimal (Figure 2). Thus he had all 21 electrodes working without any side-effects. In another patient with an ossified cochlea the anatomy was normal as well, although the implantation was difficult due to a functionally closed lateral recess.

However, there are many other patients who wish to have some benefit from an ABI, although their results may not be as good.

In very large tumours the landmarks can be distorted, or the cochlear nucleus may be damaged by pressure or ischaemia. In patients operated on several times because of recurrences, there may be scar formation or invasion of the brainstem, or some cranial nerves may be lost after previous surgery. Thus, some important landmarks may be completely missing. After gamma knife irradiation, surgery is more difficult, because the facial nerve is softer and more vulnerable, and there may be more adhesions around the tumour. The cochlear nucleus may be damaged by direct irradiation or occlusion of its arterioles leading to a delayed failure of an initially working ABI.

Patients with symptomatic multiple schwannomas may have less energy for successfully undergoing a training programme; they may be severely disabled especially by spinal or optic tumours and their life expectancy may be short. They may be disappointed as soon as they realize that the ABI is a very limited aid in only one aspect of their disabling disease.

ABI is no substitute for sacrificed hearing in a pre-operatively hearing patient. If there is a last functional hearing ear, the risk of surgery should not be taken more lightly because of the option of ABI. Surgery may be carried out in the opposite ear, if there is a tumour recurrence and hearing loss in the last ear is to be expected in the next few years. Only if there is a critical tumour growth in the last functioning acoustic nerve causing brainstem compression, may complete hearing loss be accepted. Tumour growth is sometimes much more than the average 2–4 mm per year. Some tumours double their size within one year. So in this situation there is no alternative to surgery, even if a small degree of hearing is still present.

Choice of device

The initial concept of the European ABI was to have a completely implantable multichannel device good for long-lasting daily use and multiple stimulation options taking advantage of the tonotopy of the cochlear nucleus. This was already given by the original research device with 20 channels and two reference electrodes.[7] These implants were individually cut and shaped and worked quite well, although now it is thought that the electrode surface of 0.5 mm was quite small.[8–10] Some modifications improved the Nucleus 22 ABI: the size, shape and flexibility of the electrode carrier were optimized for the anatomy of the lateral recess, the electrode surface was enlarged to a diameter of 0.7 mm and the number of electrodes enlarged to 21, the dacron mesh on the tail of the elctrode carrier was designed as a wing, and the cable was shortened and softened. So now we have a device that is comfortable to implant in a very delicate region of the brainstem, that safely keeps its position over time and that up until now has never had a technical failure. This means safety for the patient and success of the procedure, and we should not easily risk changing to any other implant that does not have the background of continuous development and safe high quality industrial production in a large series.[7,11,12]

Choice of approach and technique of tumour removal

The decision whether to use a retrosigmoid or translabyrinthine approach depends on the concept and experience of the centre. Most ENT clinics use the translabyrinthine approach for AN with complete functional hearing loss. The advantages are early identification of the facial nerve, no cerebellar retraction even in large tumours and better access to the lateral recess. The patients recover surprisingly fast, and anaesthesiology is rarely a problem. Disadvantages are the limited exposure of cranial nerves and vessels in the posterior fossa and the difficulties arising if there is a small mastoid and high jugular bulb or large sigmoid sinus. In completely deaf patients with first stage surgery, with the tumour far lateral in the meatus and convenient anatomy of the mastoid, the translabyrinthine approach is the best choice. It offers the best chance for preservation of facial nerve function and balance.

The lateral suboccipital approach is traditionally preferred by neurosurgeons. It is fast, safe and offers very good exposure of the lateral posterior fossa

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showing the whole extent of the tumour and its relationship to the cranial nerves and vessels. However, it may cause more cerebellar compression and atrophy the translabyrinthine approach. The facial nerve is identified at a later stage of tumour resection. The angle to the lateral recess and the direct vision of the entrance is more difficult than in the translabyrinthine approach. However, in patients with no tumour the lateral suboccipital approach is even better for the preservation of the facial nerve and is a good method for opening the lateral recess within scars because of the early and complete identification of the cranial nerves.

The choice of positioning is also different between the centres. Most surgeons use the supine position, that has the advantage of less loss of CSF, less traction of bridging veins, and less risk of air embolism and fluid shifts especially in elderly patients. The sitting position may be superior for hearing and facial nerve preservation in larger tumours but with a greater tendency for bleeding. Tumours up to 2 cm can be well controlled and dissected in a small retrosigmoid approach in the supine position, which is comfortable for both the patient and the surgeon.

For successful ABI implantation the choroid plexus and the exit of the cochlear nerve should be identified carefully early during tumour removal, and their positions marked. Both structures may retract or shrink during completion of the tumour excision close to the brainstem and may be missing later as landmarks for the cochlear nucleus.

Electrode implantation

Acoustic neuromas, even large ones, usually originate cranially to the pontomedullary junction (Figure 1). So the secret of ABI implantation in large tumours is to remove the tumour completely and to find normal anatomy caudally to the tumour bed. For the translabyrinthine approach this means creating a large opening including exposing the jugular bulb and the dura of the middle fossa, because otherwise the angle of vision through the microscope will not allow a view of the lateral recess, and instruments for dissection and implantation may not pass the bony edges of the craniotomy.

To expose the landmarks: retract the flocculus together with the choroid plexus, do not dissect between them. Identify the exits of the cranial nerves VII, VIII and IX. Cranial nerves VII and IX should be identified and tracked by electromyography. From known anatomical studies, one of the landmarks should be found, and the others including the cochlear nucleus will then be within an area of 6 mm. Learn the direction needed to reach the next landmarks from the first one within the triangle, and identify as many landmarks as possible. Then open the arachnoid and elevate the vessels over the lateral recess. The taenia of the choroid plexus sometimes has to be cut. Frequently an AICA-branch or a big vein has to be lifted from the entrance to the lateral recess. This is easier for arteries than fragile veins. Do not seal small vessels close to the cochlear nucleus but cover them with gelfoam and cottonoids and gently compress for a few minutes. Enter the lateral recess with a 2 mm dissector without touching the cochlear nucleus. Control the outflow of CSF out of the lateral recess while the anaesthetist ventilates with elevated airway pressure or carries out a Valsalva manoeuvre. Do not push the electrode carrier into the retro-olivary fossa or into the brainstem, and only implant if you have safely identified the opening of the lateral recess and the contour of the cochlear nucleus.

Results

Since 1992, ABI surgery has been performed on 58 patients with our cooperation in 18 centres in nine European and one Asian country. Acoustic neuromas have been removed from 49 NF2 patients using a translabyrinthine or suboccipital approach and a central electroauditory prosthesis implanted. Six patients were free of tumour after previous AN-surgery, one suffered from a recurrent Lindau-tumour, one from an axonal neuropathy and one from an ossified cochlea. Their mean age was 33 years, and the tumour size was 1.0 to 6.0 cm. The translabyrinthine approach was used in 37 and the lateral suboccipital approach in 21 patients. The tumours were completely removed in all cases, and the facial nerve anatomically preserved in 56 out of 58 patients. The intra-operative positioning of the electrode carrier was guided by anatomical landmarks (preserved cranial nerves VII, VIII and IX, choroid plexus, and exit of the foramen of Luschka) and was estimated as easy in 35 cases, difficult after extensive dissection in 22 and not possible in one case (Figure 3).

Intra-operative monitoring of the auditory brainstem evoked potentials helped in optimizing the position in questionable or gave security in very accurate implantations. The monitoring was not safe and reliable in all cases. If there are no auditory brainstem responses at all and the position of the electrode looks good, it should be left as all of these patients had hearing function. It may be that

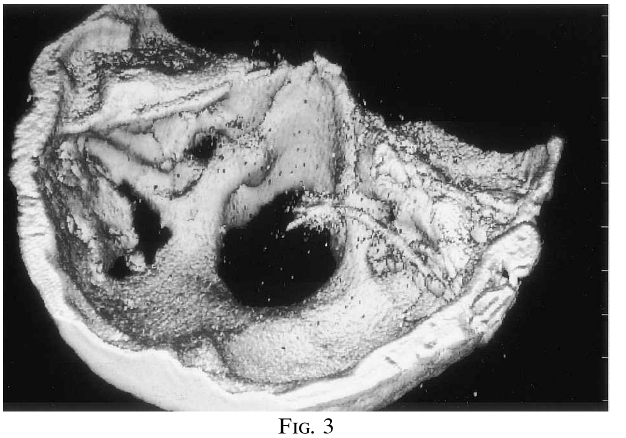

FIG. 3

Position of the Nucleus 22 auditory brainstem implant after implantation (3D-CT).

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sometimes the cochlear nucleus is irritated by manipulation of the tumour, heating or irrigation but will recover later.

In all but three of the 54 patients tested there was some hearing in between two and 21 patients, on average 9.4 electrodes. Forty-eight patients are using the device daily. The patients have different sound and frequency impressions. In tests most of the patients were able to recognize different words. The device facilitates lip-reading and is useful in daily life for environmental awareness. The patient suffering from a bilateral recurring Lindau-tumour had no hearing function at all because of direct lesion of the cochlear nucleus by tumour invasion. Two patients with gamma knife irradiation prior to ABI had late failures of their implant function months and years after initially successful surgery. One patient died on the third post-operative day due to pulmonary embolism, and one, two years later after spinal surgery in a foreign clinic. One patient frequently fell on his implant due to ataxia, and the uncovered device had to be removed after several attempts at plastic surgery. There were no other complications due to the implant itself. Three patients are not satisfied with the quality of hearing and are only poor users.

A constant finding was the oblique direction of the positive electrodes over the electrode carrier. It seems that only a part of the neurons suited for stimulation lie superficially in the lateral recess, while others are covered by the spinocerebellar tract. The horizontal part of the cochlear nucleus lies in an oblique angle to the cranio-caudal course of the lateral recess.

Most of the patients showed a tonotopic distribution of the stimulation responses with high frequencies on the dorsal position of the tip of the electrode carrier and deep frequencies on the ventral side. Furthermore, there is not only one tonotopy but up to three regions with tonotopic organization corresponding to the different intersections of the cochlear nucleus.

Conclusions

Multi-electrode stimulation of the second auditory neuron by using transcutaneous signal transmission is possible.[1–3,5,6] The best results are obtained in patients with stable NF2 disease and primary surgery of small acoustic neuromas. ABI may be successful in tumour recurrences and with an interval of hearing loss up to 16 years or even more. The first results with diseases other than NF2 are promising.[5,6,12]

The surgery should be carried out in selected centres in different countries by an established surgical technique, respecting the need for successful ABI implantation after complete tumour removal. The implantation will be most accurate if the surgeon removing the tumour preserves the anatomy perfectly and the ABI specialist is included at a stage where there still is a chance of identifying the landmarks and finding the direction of the cochlear nucleus. Up until now, no navigation will aid and the neuromonitoring is only helpful if we are already very close to the functionally intact target.

Most patients are using their implant daily. They have different sound and frequency impressions and improvement of lip-reading, some have a limited open speech discrimination. Mapping of the stimulation effects gives reproducible information about the tonotopic organization of the cochlear nucleus and the functional anatomy of the surrounding of the cochlear nucleus.

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Address for correspondence:
Prof. Dr. med. Wolf-Peter Sollmann,
Neurochirurgische Klinik,
Städtisches Klinkum Braunschweig,
D-38126 Braunschweig,
Germany.

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