CNAP To Predict Functional Cochlear Nerve Preservation in NF-2: Cochlear Implant or Auditory Brainstem Implant

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ABSTRACT

The ideal management of bilateral vestibular schwannomas (VSs) involves complete tumor resection with preservation of hearing in at least one ear. While auditory brainstem implants (ABIs) have represented a significant advance in the management of neurofibromatosis type 2 (NF-2) patients, hearing rehabilitation is far from ideal. More recently, cochlear implantation has been used in selected cases of NF-2, following tumor removal, where the patient is left with bilateral profound hearing loss but with anatomical continuity of the cochlear nerve. In selected cases, cochlear implant (CI) has given superior results to ABI. The Gruppo Otologico experience in managing NF-2 patients consists of 29 patients treated between December 1996 and December 2007 out of a total of 1723 VSs. Thirty-nine tumors have been removed, with 10 patients having had bilateral tumor removed. Ten ABIs have been implanted and 5 CIs placed. We present a case that illustrates some of the difficulties encountered in the management of NF-2 VS and provides a basis for discussion of a technique to assist intraoperative decision-making to achieve optimal hearing rehabilitation. In the setting of a negative fast auditory brainstem response (ABR), but preserved cochlear nerve action potential (CNAP), the option of a CI can be considered. However, where the fast ABR and the morphology and amplitude of the CNAP are significantly degraded at the brainstem, the placement of an ABI should be strongly considered at the time of tumor removal. This technique must be further evaluated, but it represents an adjunct in this difficult area for the neurotologist. To this end we propose that a multicenter trial be undertaken to further elucidate the role of intraoperative monitoring to determine the functional preservation of the cochlear nerve.

KEYWORDS: Intraoperative monitoring, NF-2, fast ABR, CNAP, hearing rehabilitation

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Received: December 6, 2007. Accepted: December 6, 2007.
Published online: February 19, 2008.
DOI 10.1055/s-2008-1043753. ISSN 1531-5010.

The management of the neurofibromatosis type 2 (NF-2) patient is a difficult task. The ideal management of bilateral vestibular schwannomas (VSs) involves complete tumor resection with preservation of hearing in at least one ear. Unfortunately, the advanced size of tumors at presentation often precludes attempted hearing preservation.

Hearing preservation surgery in the setting of NF-2 also appears to be less favorable than in sporadic cases, although more recent reports tend to suggest similar results.¹⁻⁵ Despite this, however, bilateral profound hearing loss has been reported to develop in 96% of cases.⁵

The first auditory brainstem implant (ABI) was placed by House and Hitselberger in 1979. Despite significantly improving hearing rehabilitation in the management of NF-2 patients affected by bilateral tumors of the cerebellopontine angle, most patients only report environmental noise and sound cues that provide assistance while lip-reading.⁶⁻⁹

We present a case report that illustrates some of the difficulties encountered in the management of NF-2 VS and provides a basis for discussion of a technique to assist intraoperative decision-making to achieve optimal hearing rehabilitation.

CASE HISTORY

A 24-year-old man with bilateral VSs was first seen in 2002. At that time he had a right 1-cm VS with 100% speech discrimination score (SDS) and a pure tone audiometry (PTA) of 20 dB, and a left 3.8-cm VS with 100% SDS and a PTA of 20 dB (Fig. 1).

A left translabyrinthine approach (TLA) with trans-apical extension was performed in October 2002.¹⁰ A decision was undertaken to terminate the procedure due to significant intraoperative bleeding, resulting in an approximate 50% reduction in tumor size.

Three months later a type A transcochlear approach was used to maximize anterior control,

allowing complete removal of the residual VS and the small trigeminal tumor. At this time the patient was referred for lip-reading training.

A left anastomosis of facial nerve to hypoglossal nerve was performed in April 2003 prior to addressing the contralateral tumor. This was to ensure that there was no possibility of a bilateral facial nerve paralysis. The patient recovered to House-Brackmann Grade III.

In September 2004 a retroigmoid-retrolabyrinthine (RS-RL) approach with attempted hearing preservation was conducted on the right VS, which had increased in size to 2 cm.¹¹ Hearing function was monitored with a combined fast auditory brainstem response (ABR) and cochlear nerve action potential (CNAP) technique (MK12 Amplaid, Italy). Continuous electromyographic facial nerve monitoring of the orbicularis oculi and orbicularis oris was applied (NIM-Pulse, Xomed-Trace, Nashville, TN). Anatomic preservation of the cochlear and facial nerves was achieved. During tumor removal wave V could not be clearly identified and the CNAP performed at the fundus and brainstem entry zone revealed a wide variance in amplitude and depolarization patterns of the waves P1 and N1 (Fig. 2).

Postoperatively, the patient was left with complete bilateral deafness. Promontory stimulation was performed, first 45 days postoperatively with no response, and then at 90 days postop revealing only vibratory sensation at maximal stimulation.

In November 2004 a right cochlear implant (CI) was performed with very poor results. The patient scored 0% in all open-set format tests. At this stage the patient does not want any further hearing rehabilitation.

In retrospect, the correct decision would have been to insert an ABI at the time of resection of the right VS. The presence of a CNAP was thought to provide evidence of a functioning cochlear nerve, allowing CI as a staged procedure. On review, however, the significant decline in amplitude of the CNAP at the brainstem heralded the extremely poor results of CI.

Figure 1 Magnetic resonance imaging scans of the patient before treatment.

In addition, an argument could be made for the insertion of a “sleeper” ABI after the first tumor removal.

DISCUSSION

Decision-Making Process in NF-2

The Gruppo Otologico experience in managing NF-2 patients consists of 29 patients treated between December 1996 and December 2007 out of a total of 1723 VSs. Thirty-nine tumors have been removed, with 10 patients having had bilateral tumor removed. Ten ABIs have been implanted, and 5 CIs placed.

The management plan is highly complex and the patient with his or her family must be made aware of all the options. Obviously it is not only the bilateral VSs and hearing that must be considered, but the optimal management of the coexisting manifestations of NF-2.

While ABIs have represented a significant advance in the management of NF-2 patients, hearing rehabilitation is far from ideal. More recently cochlear implantation has been used in selected cases of NF-2 following tumor removal, where the patient is left with bilateral profound hearing loss but anatomical continuity of the cochlear nerve. In selected cases CI has given superior results to ABI. However, successful CI in this setting is predicated on a functioning cochlear nerve.^{12–15} Reports of positive promontory

Figure 2 (A) CNAP tracing taken from the fundus. Note the normal morphology of the CNAP, and a 25 microAmp scale. (B) Note at the level of the brainstem the morphology of the CNAP has deteriorated, indicating poor depolarization of the nerve. This is predominantly related to N1. There is also a significant diminution of the CNAP amplitude compared with measurements taken at the fundus (the amplitude scale is 1 microAmp).

stimulation following labyrinthectomy and TLA VS removal^{16,17} predicted that successful cochlear implantation could be achieved after the removal of a VS on the same side.^{18–21} Arriaga and Marks were the first to report simultaneous VS resection and CI implantation,¹⁹ with Ahsan et al reporting the first simultaneous translabyrinthine VS resection and CI insertion in an NF-2 patient.¹³ Small case series have also reported favorable results with CI in NF-2.^{12,14,15} The superior hearing results reported by Neff et al confirm that the presence of a positive promontory stimulation 8 weeks after tumor removal predicts an improved outcome following CI in this setting. They also report that long-term benefit was achieved in these patients, negating concerns that cochlear nerve fibrosis postoperatively would lead to a decline in function.¹⁴

The treatment of the only hearing ear in NF-2 presents many difficulties. If the tumor fits the criteria for hearing preservation surgery, this is attempted. A fast ABR and CNAP are utilized through an RS-RL approach. If the monitoring remains favorable, there is a high likelihood of hearing preservation.²² Alternatively, if the cochlear nerve is disrupted an ABI should be implanted simultaneously. If a TLA approach is used, or fast ABR is lost, but the cochlear nerve is preserved, an intraoperative method to assess functionality of this nerve is desirable to determine suitability of CI as opposed to ABI. Arriaga and Marks have suggested that an intraoperative promontory stimulation may be of some benefit in this setting,¹⁹ but we and others have found that intraoperative promontory stimulation is unreliable.²³ Its unreliability is strengthened by reports of early postoperative false-negatives occurring with the use of this technique.^{14,18}

The most significant problem with the plan to place CI in the setting of a preserved cochlear nerve is the lost opportunity to place an ABI if the cochlear nerve function is inadequate to sustain a CI. This is due to the very poor results with placement of an ABI as a second-stage procedure. A further important consideration is to minimize the

Figure 3 The Gruppo Otologico proposed protocol for hearing rehabilitation of the only hearing ear in the NF-2 patient. NF2, neurofibromatosis type 2; RS-RL, retrosigmoid-retrolabyrinthine; ABR, auditory brainstem response; CNAP, cochlear nerve action potential.

number of major interventions in the NF-2 patient. We propose, however, combining the predictive power of a fast ABR and CNAP to guide the intraoperative decision. If during surgery the fast ABR is lost, but preservation of CNAP with good amplitude and morphology is maintained, a CI can be performed. If the CNAP is absent, or the amplitude and morphology significantly altered, then an ABI should be placed (Fig. 3). This schema can be used in treating either the first or second tumor in NF-2, when there is bilateral severe-profound hearing loss.

We have previously reported the predictive value of CNAP and fast ABR in relation to post-operative hearing, with the loss of fast ABR and CNAP correlating to very poor postoperative hearing.²⁴ This allows us to be sure that we are not potentially sacrificing residual hearing by placing a CI. A potential problem exists in the setting of a false-negative CNAP.^{23,25} This result would trigger the placement of an ABI, where a CI would have provided a superior hearing result. While early false-negative promontory stimulation tests have reversed at delayed testing, the higher sensitivity of CNAP makes this less likely.

Until further research can truly determine the false-negative rate of CNAP with an anatomically

preserved cochlear nerve, we recommend the placement of an ABI simultaneously.

It is hoped that this will minimize the risk of creating a situation where the only chance of hearing rehabilitation involves the placement of an ABI as a second-stage procedure, as in the scenario that we have presented here.

TECHNICAL ISSUES OF TUMOR DISSECTION

The preservation of a functioning cochlear nerve is central to this discussion. Injury to the hearing pathway can occur from labyrinthine damage, vascular interruption to the cochlea and/or cochlear nerve, and direct mechanical trauma to the cochlear nerve. Tumors arising from, infiltrating, or severely effacing the cochlear nerve make the possibility of preserving function very remote. This is especially the case with tumors greater than 1.5 cm. The histological nature of NF-2 VSs insinuating among the neural fibers also plays a role.^{26,27} These factors give support to the aim of early intervention in the management of NF-2 VS due to increased likelihood of cochlear nerve preservation, if not hearing preservation itself. It is paramount, however, to correlate the parameters of physiological monitoring in conjunction with the physical status of the nerve following dissection.

TECHNICAL ASPECTS OF CNAP MONITORING

The reliability of the CNAP as a prognosticator is based both on the recorded amplitude and the morphology of the wave. The amplitude of the response is proportional to the number of functioning cochlear nerve fibers, and thus can predict the results of CI.²⁸ The morphology of the CNAP is also a critical measure, with depolarization of the nerve indicated by the relationship of P1 and N1.

Degradation of the morphology thus represents a desynchronization of neural activity.

The ideal situation to allow analysis of changes in CNAP is one where a recording can be taken at the proximal cochlear nerve prior to tumor removal of the intracranial portion of the nerve. This allows comparison of amplitude before and after tumor removal. This is, however, impossible in larger tumors, and it is difficult to ensure that the electrode remains in an identical position to allow direct comparison.⁵ Following tumor removal, comparison of the CNAP at the fundus and the brainstem entry zone should be performed. A significant difference between the fundus and brainstem indicates a loss of function of the cochlear nerve itself.

As presented in this case, while a CNAP was recorded following tumor removal, both the amplitude was significantly reduced and the morphology was abnormal. In retrospect this predicted the negative promontory stimulation and poor CI result. Given these intraoperative findings, an ABI should have been placed at the time of tumor removal.

CONCLUSION

We propose a technique that allows the intraoperative decision to be made as to whether a preserved cochlear nerve will result in acceptable hearing rehabilitation using a CI. In the setting of a negative fast ABR, but preserved CNAP, the option of a CI can be considered. However, where the fast ABR and the morphology and amplitude of the CNAP are significantly degraded at the brainstem, the placement of an ABI should be strongly considered at the time of tumor removal.

This technique must be further evaluated, but it represents an adjunct opportunity for treatment in this difficult area for the neurotologist. To this end, we propose that a multicenter trial be undertaken to further elucidate the role of intraoperative monitoring to determine the functional preservation of the cochlear nerve.

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