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Hearing Rehabilitation in Neurofibromatosis Type 2 Patients: Cochlear versus Auditory Brainstem Implantation

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Key Words

Neurofibromatosis type 2 · Deafness · Cochlear implantation · Auditory brainstem implantation · Acoustic tumors

Abstract

Objective: We aimed to evaluate and compare the auditory performance of neurofibromatosis type 2 (NF2) patients with bilateral total deafness fitted with cochlear or auditory brainstem implants. ***Patients and Methods:*** A retrospective case review was performed. Nine patients suffering from NF2 who underwent hearing rehabilitation by means of cochlear (4 patients) or auditory brainstem (5 patients) implantation participated in the study. Postoperative auditory performance was assessed using closed- and open-set tests. ***Results:*** In the group of patients fitted with a cochlear implant, 3 subjects achieved open-set speech recognition abilities comparable to those of standard adult postlingual implant patients; the remaining patient scored 0% in all open-set format tests, reporting benefits only in environmental sound detection and lip-reading. Among the 5 patients who underwent auditory brainstem implantation, 1 reached good open-set speech recognition skills, scoring 70% in the common phrases comprehension test, and she was able to communicate on the telephone. Two other patients achieved open-set speech understanding (respectively, 33 and 41% in the common phrases comprehension test), reporting daily use of their device. The remaining 2 patients did not achieve any level of open-set speech perception, obtaining only improved access to environmental sound and lip-reading skills. ***Conclusions:*** Our study confirmed literature data reporting that cochlear implantation may offer open-set speech communication in NF2 patients. In this small cohort, cochlear implant patients performed better than auditory brainstem implant patients, even if variability in auditory performance was observed with both devices. More studies are needed in order to clarify the role and reliability of electrophysiological tests in predicting the residual functionality of the cochlear nerve after tumor removal.

Introduction

Neurofibromatosis type 2 (NF2) is an autosomal dominant disorder resulting from mutations in both alleles of a tumor suppressor gene, localized to the long arm of chromosome 22 and coding for a protein called merlin. The most common clinical feature of NF2 is the development of bilateral vestibular schwannomas (VS); the treatment options for VS include simple observation, microsurgical resection with or without hearing preservation and radiation therapy. Unfortunately, regardless of the

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treatment modality chosen, the great majority of NF2 patients [Jackler, 1994; Sanna et al., 1998] develop a total bilateral deafness. In the past, once a patient with NF2 had lost hearing, there was no possibility of regaining an auditory input. In 1979, House and Hitselberger [2001] first provided a deaf NF2 patient with a single ball auditory brainstem implant (ABI), altering the management of deafness in these patients. Over the past 25 years, ABI has undergone several improvements, ranging from the introduction of multichannel devices to increased capabilities of speech processing, and hundreds of patients have undergone implantation worldwide with variable hearing results [Nevison et al., 2002; Otto et al., 2002; Kanovitz et al., 2004; Colletti et al., 2005]. In the 1990s, Cueva [1992] reported on auditory sensations elicited by electrical promontory stimulation (EPS) in a patient with preserved cochlear nerve and deafness after VS resection, demonstrating the theoretical feasibility of cochlear implantation in deafened NF2 patients with intact cochlear nerve after VS removal. Since then, histological [Belal, 2001] and clinical studies [Cohen et al., 1992; Hulka et al., 1995; Tono et al., 1996; Graham et al., 1999; Ahsan et al., 2003; Nolle et al., 2003; Aristegui and Denia, 2005] confirmed that cochlear implantation is possible in at least a subset of this special population of patients. Today, we can offer 2 options for hearing restoration, cochlear or auditory brainstem implantation, to NF2 deafened patients with an anatomically preserved cochlear nerve. Currently, the most suitable treatment option for hearing rehabilitation when clinical, radiological and electrophysiological tests indicate that both devices can be used remains to be established. As reported in the literature, hearing results obtained with cochlear implantation seem to be more favorable than those achieved with ABI. However, to our knowledge, in the literature there are no studies that have compared auditory performance in NF2 patients fitted with a cochlear implant (CI) or an ABI in the same institution utilizing the same perception tests for all patients. To verify whether cochlear implantation allows better hearing performance than auditory brainstem implantation, we compared the hearing outcomes obtained in 9 NF2 patients, 4 fitted with a CI and 5 fitted with an ABI.

Materials and Methods

The database with prospectively collected data of the Gruppo Otologico of Piacenza-Roma (Italy) was searched for patients with NF2 who underwent cochlear or auditory brainstem implantation; information regarding 12 patients was retrieved from the database. As the aim of the study was to compare hearing performance between CI and ABI, 3 patients (2 fitted with a CI and 1 fitted with an ABI) with residual hearing in the nonimplanted ear were excluded because isolation of the implanted ear from the hearing ear could not be easily achieved. Thus, 9 patients participated in the study, 4 fitted with a CI (group A) and 5 fitted with an ABI (group B).

Group A patients were between 24 and 47 years old (mean age = 34.7); 2 of them were female and 2 were male. In 3 subjects cochlear implantation was performed 3 months after VS surgery (in 2 patients functional surgery via a retrosigmoid approach was attempted with unsuccessful hearing preservation, while in the remaining patient VS was removed via a translabyrinthine approach with cochlear nerve preservation). Before cochlear implantation, all 3 patients underwent high-resolution computed tomography of the temporal bone to check the patency of cochlear turns as well as EPS in order to acquire information about the functional integrity of the cochlear nerve. In the remaining patient, VS resection via a translabyrinthine approach and cochlear implantation were done simultaneously; after tumor removal, evaluation of the functional integrity of the cochlear nerve was performed by means of a cochlear nerve action potential (CNAP) study.

The Nucleus Promontory Stimulator (model Z10012, Cochlear, Melbourne, Australia) was used for all promontory stimulations. After topical anesthesia of the tympanic membrane with a Xylocaine solution, a needle electrode was placed transtympanically on the promontory and held in place by a foam earplug. Electrical stimulation by a 50-Hz square wave was initiated at 0 μA, with the current being slowly increased until the patient 'heard' or 'felt' the stimulus; the test was performed at 5 frequencies: 50, 100, 200, 400 and 800 Hz. The threshold level (the lowest current level at which the patient could perceive the stimulus) and maximum acceptable loudness (the current level at which stimulation was no longer comfortable) were determined and the dynamic range calculated (maximum acceptable loudness minus threshold level). The patients with auditory perceptions were asked to discriminate the pitches of the various stimuli at 50, 100, 200, 400 and 800 Hz. A positive response to EPS consisted of discrete tone perception and the ability to differentiate pitch, while the test was considered negative if the patient was not able to hear a tone, perceiving only discomfort from electrical stimulation.

In all 4 cases a Nucleus 24 Contour CI (Cochlear Corporation) was used. Table 1 shows the clinical and demographic findings of the 4 patients.

Group B patients were between 22 and 45 years old (mean age = 32.8), 3 of them were female and 2 were male. All these patients had been previously operated on for a VS on the contralateral side and in any case the cochlear nerve was preserved. During surgery, VS was removed via a translabyrinthine approach and, since it was not possible to maintain an intact cochlear nerve in any case, a Nucleus ABI 24 device (Cochlear Corporation) was positioned simultaneously with tumor removal. Table 2 shows the clinical and demographic findings of the 5 patients.

Postoperative auditory performance was assessed by means of the following tests in both closed-set (i.e. the patients must select their response from limited options) and open-set (i.e. understanding words or sentences without alternatives from which to choose the answer) formats: detection of environmental sounds, vowel and consonant identification, bisyllabic word recognition,

Table 1. Clinical and demographic findings of patients fitted with a CI (group A)

Case	Sex	Age years	Auditory deprivation months	Contralateral ear	Implanted ear	Electrophysiologic testing	Implant type	Device use	Telephone use
1	F	47	3	VS (4 cm) removal via TLA (cochlear nerve sacrificed)	VS (1.5 cm) removal via RSA: no hearing conservation	EPS: good response	Nucleus 24 Contour	daily	yes
2	F	24	0	Anacusis after VS (1.5 cm) removal via RSA	VS (2 cm) removal via TLA: intact CN (CI in the same stage)	CNAP: good response	Nucleus 24 Contour	daily	yes
3	M	32	3	VS (3.5 cm) removal via TLA (cochlear nerve sacrificed)	VS (2.5 cm) removal via TLA: intact cochlear nerve	EPS: good response	Nucleus 24 Contour	daily	yes
4	M	36	3	VS (4 cm) removal via TLA (cochlear nerve sacrificed)	VS (1.5 cm) removal via RSA: no hearing conservation	absent ABR; CNAP and EPS: doubtful response	Nucleus 24 Contour	daily	no

TLA = Translabyrinthine approach; RSA = retrosigmoid approach; ABR = auditory brainstem response.

Table 2. Clinical and demographic findings of patients fitted with an ABI (group B)

Case	Sex	Age years	Contralateral ear	Implanted ear	Implant type	Device use	Subjective comments	Telephone use
1	F	45	VS (4 cm) removal via TLA (cochlear nerve sacrificed)	VS (2 cm) removal via TLA (cochlear nerve sacrificed) + ABI insertion	Nucleus ABI 24	daily	sufficient benefit	no
2	F	22	VS (3.5 cm) removal via TLA (cochlear nerve sacrificed)	VS (3 cm) removal via TLA (cochlear nerve sacrificed) + ABI insertion	Nucleus ABI 24	daily	sufficient benefit	no
3	M	27	VS (4.5 cm) removal via TLA (cochlear nerve sacrificed)	VS (4 cm) removal via TLA (cochlear nerve sacrificed) + ABI insertion	Nucleus ABI 24	daily	scarce benefit	no
4	M	33	VS (4 cm) removal via TLA (cochlear nerve sacrificed)	VS (3.5 cm) removal via TLA (cochlear nerve sacrificed) + ABI insertion	Nucleus ABI 24	daily	scarce benefit	no
5	F	30	Anacusis after VS (1.5 cm) removal via RSA	VS (3 cm) removal via TLA (cochlear nerve sacrificed) + ABI insertion	Nucleus ABI 24	daily	excellent benefit	yes

TLA = Translabyrinthine approach; RSA = retrosigmoid approach.

sentence recognition and common phrases comprehension. The speech materials were presented in vision-only condition, auditory-only condition and auditory-plus-vision condition using monitored live voice through the sound field at a level of 70 dB sound pressure level. In this study, we present the results of hearing tests in auditory-only condition collected at 1-, 6- and 12-month postimplantation intervals. In the environmental sounds detection test, the patient had to respond to the presence or absence of sounds of different frequencies delivered at an intensity of 70 dB HL (drum for low frequencies, bell for medium frequencies, rattle for high frequencies).

In the closed-set consonant identification test, 13 meaningless consonant words (ABA, AGA, ATA) were read to the patient through live voice 4 times; in the closed-set vowel identification test, 10 mono-syllable vowels, composed of 5 long (BAAT, GAAT) and 5 short (BAT, GAT) ones, were read to the patient through live voice 4 times. In the open-set bisyllabic word recognition test, a list of 25 bisyllabic common Italian words was presented to the patient. The results of these tests were scored in terms of words correctly repeated by the patient. In the open-set sentence recognition test a list of 10 uncommon sentences was presented to the patient: each list contained 100 words and was scored for the total number of words correctly repeated. The open-set common phrases comprehension test was based on common and simple interrogative phrases such as 'How are you feeling?', to which the patient had to respond; test scoring was based on the percentage of correct responses.

Results

Group A (CI patients)

Patients 1 and 3 totally satisfied the criteria for a positive ESP, performed 1 month after surgery; patient 4 re-

Table 3. Hearing results in CI patients (group A)

Case	Vowel identification %			Consonant identification, %			Bisyllabic word recognition, %			Sentence recognition %			Common phrases comprehension, %		
months:	1	6	12	1	6	12	1	6	12	1	6	12	1	6	12
1	100	100	100	92	100	100	44	72	80	61	76	90	72	100	100
2	100	100	100	85	100	100	32	50	72	40	59	81	50	60	86
3	100	100	100	85	100	100	20	40	50	0	30	50	20	38	55
4	10	30	40	8	23	39	0	10	10	0	0	0	0	0	0

Table 4. Hearing results in ABI patients (group B)

Case	Vowel identification, %			Consonant identification, %			Bisyllabic word recognition, %			Sentence recognition, %			Common phrases comprehension, %			Active electrodes	
months:	1	6	12	1	6	12	1	6	12	1	6	12	1	6	12	intra-operative	12
1	20	40	70	15	39	70	0	20	30	0	0	31	0	0	33	14/21	15/21
2	30	55	90	23	46	85	0	20	40	0	32	38	0	29	41	15/21	17/21
3	10	20	50	8	15	39	0	0	0	0	0	0	0	0	0	18/21	12/21
4	20	60	80	15	54	70	0	0	20	0	0	10	0	0	0	9/21	12/21
5	90	100	100	85	92	100	20	50	70	0	30	54	0	42	70	19/21	20/21

ported sound perception only with 50- and 100-Hz stimuli and experienced pain with increasing stimulus intensity on 2 subsequent EPS, performed 1 and 3 months after surgery, respectively. After extensive counseling about the possible risks and results of cochlear and auditory brainstem implantation, this patient preferred the insertion of a CI.

On preoperative high-resolution computed tomography scans, no cases of cochlear ossification were observed and a full electrode array insertion into the scala tympani was achieved in all patients. In patient 2 (VS removal and CI in the same stage) a reliable CNAP was recorded confirming preservation of at least some cochlear nerve fibers after tumor removal. Thus, a CI was inserted into the scala tympani. The postoperative period was uneventful and a check radiogram confirmed the correct positioning of the implant in all subjects. The advanced combination encoder strategy was adopted in all cases. At switch on of the device all patients presented environmental sound detection.

Within this group, patients 1, 2 and 3 achieved good levels of auditory performance in open-set tests (table 3), scoring from 55 to 100% in comprehension tests at the 1-year follow-up. Patients 1 and 2 reached hearing outcomes similar to the best of standard postlingual adult implantees, also being able to easily understand phrases during telephone conversations. Patient 4 scored 0% in all open-set format tests and 10% in the bisyllabic word recognition test. He uses the implant daily, finding it useful only for lip-reading and detecting environmental sounds.

Group B (ABI Patients)

In all cases ABI was inserted under direct monitoring of the 7th, 9th and 11th cranial nerves and indirect monitoring of the 10th cranial nerve through electrocardiographic recording. After ABI insertion into the lateral recess, impedance telemetry and electrical auditory brainstem responses were performed to ascertain the correct position and function of the implant. Intraoperatively, the number of electrodes from which it was not possible to obtain electrical auditory brainstem response waves varied from 2 to 12. ABI activation was performed 1 month later in the operating room and the advanced combination encoder strategy was adopted. At switch on of the device all patients presented environmental sound

Table 5. Literature review on hearing results of cochlear implantation in NF2 patients (case reports)

Source	Cases	Age years	Surgical strategy	Implant type	Hearing results	Follow-up months	EPS	Notes
Hulka et al. [1995]	1	31	VS (0.6 cm) removal via RSA and CI 2 months later	Nucleus CI22	Pure tone thresholds of 30–40 dB (250–4000 Hz)	3	EPS positive 7 week after surgery	the patient uses the implant successfully
Tono et al. [1996]	1	31	VS (0.8 cm) removal via MCFA and CI 15 months later	Nucleus 22	62% on open-set SR test	12	EPS positive at 50 and 100 Hz 1 month after surgery	VS extending into the vestibular labyrinth
Graham et al. [1999]	1	44	VS (0.8 cm) removed via posterior fossa surgery and CI 7 years later	Magnetless Clarion 1.2 system	34% on BKB sentence and 97% on CUNY	6	EPS negative 2 weeks after surgery but round window stimulation positive 6 years after surgery	obliteration of the basal turn (0.5 cm)
Temple et al. [1999]	1	15	VS (0.5 cm) removal via MCFA and CI 9 months later	Med-el Combi 40	100% on SR, 70% on TWP, 60% on TFWP	12	EPS positive pre- and postoperatively	excellent and reproducible responses on EPS at 50, 100, 200, 400 and 800 Hz
Nolle et al. [2003]	1	16	VS removal via RSA and CI 2 years after	Nucleus 22 RCS	88% on open-set SR test	24	EPS positive	small residual tumor on the implanted side
Ahsan et al. [2003]	1	53	VS (0.7 cm) removal via TLA and concurrent CI	NA	No formal testing	NA	EPS positive	good speech understanding (subjective comment reported by the patient)
Aristegui and Denia [2005]	1	52	VS (4 cm) removal via TLA and concurrent CI	Med-el Combi 40	80% on VI, 90% on BWR, 100% on CID	18	not performed	on contralateral side CI was attempted after 1.5 cm VS removal via a TLA but total cochlear ossification was found

CID = Central Institute for the Deaf sentences of everyday speech; SR = sentence recognition; BKB = Bamford-Kowal-Bench; CUNY = City University of New York sentences in noise; TWP = two-word phrases; TFWP = three- to four-word phrases; VI = vowel identification; BWR = bisyllabic words recognition; MCFA = middle cranial fossa approach; TLA = translabyrinthine approach.

detection. Electrodes eliciting nonauditory sensations decreased over time and, at the 1-year follow-up, ranged from 1 to 9. In patients 3 and 4, ABI benefits remained limited to improvement in environmental sound detection and lip-reading skills. The remaining 3 patients achieved open-set speech understanding ranging from 33 to 70% in the common phrases comprehension test. The results of the hearing tests in this group of patients (1-year follow-up) are reported in table 4.

Discussion

Over the last 2 decades hearing rehabilitation in totally deaf NF2 patients evolved from no chances into the possibility to choose, at least in some cases, between 2 prosthetic devices, ABI or CI; moreover, in the past years, the auditory midbrain implant, a new central auditory prosthesis, has been under investigation to verify whether the central nucleus of the inferior colliculus could represent an alternative for auditory stimulation [Lenarz et al., 2006].

ABI was the first prosthetic device used in auditory rehabilitation of NF2 subjects. The reported hearing outcomes vary significantly, ranging from simple environmental sound awareness and assisting in lip-reading to open-set speech understanding, allowing conversation even in telephone communication. Since the 1990s, several individual case reports (table 5) have appeared in the literature describing satisfying, even if variable, rehabilitative results using a CI in NF2 subjects with deafness but preserved cochlear nerve after VS resection.

More recently, 2 multi-institutional studies reporting hearing outcomes in a series of NF2 patients who underwent cochlear implantation were published (table 6). Lustig et al. [2006] first reported on a series of 7 NF2 pa-

Table 6. Literature review on hearing results of cochlear implantation in NF2 patients (case series)

Source	Cases	Age years	Surgical strategy	Implant	Hearing results	Follow-up months	EPS	Notes
Lustig et al. [2006] Among the reported 7 patients, 3 had residual contralateral hearing at the time of implantation and 4 did not. In the table, only results for the 4 patients without residual contralateral hearing at the time of implantation are reported.	1	41	NA	NA	46% on MTS and 0% for all other tests	17	NA	improved environmental sound awareness + lip-reading
	2	28	NA	NA	0% on all speech tests	88	NA	improved environmental sound awareness + lip-reading
	3	50	NA	NA	46% on MTS, 98% on HINT	18	NA	excellent benefit
	4	57	NA	NA	12% on CNC, 35% on phonemes, 21% on HINT	9	NA	good benefit
Neff et al. [2007] (6 patients)	1	15	1.5 cm VS removal via MCFA and CI 9 months later	Med-el Combi 40	45% on 3SWLOS, 75% on 3SWLCS	120	EPS positive 6 weeks after surgery	telephone use
	2	30	VS removal via RSA and CI 4 months later	Nucleus Mini-22	98% on CID, 43% on CUNY, 91% on HINT	108	EPS negative 1 month after surgery and positive 8 weeks after surgery	telephone use
	3	59	Left VS removal via RSA	Nucleus Mini-22	100% on CID, 93% on CUNY, 96% on HINT	60	EPS positive 1 year after surgery	prior to CI in the left ear, ABI was inserted in the right ear concomitantly with 3 cm VS removal via TLA without hearing results
	4	38	1.8 cm VS removal via a RSA and CI few months later	Nucleus 22	90% on CID, 83% on HINT	96	immediate post-operative EPS negative; 2 months later EPS became positive	telephone use
	5	37	VS removal via a TLA and concurrent CI	Nucleus 24	100% on CID, 96% on HINT	60	not performed	telephone use
	6	31	8 mm VS removal via RSA and CI 2 months later	Nucleus 22	22% on CID, 0% on HINT	156	EPS positive 7 weeks after surgery	no telephone use

MTS = Monosyllable, Trochee, Spondee; HINT = Hearing in Noise Test; 3SWLOS = Three-Syllabic Word List, Open-Set; 3SWLCS = Three-Syllabic Word List, Closed-Set; CID = Central Institute for the Deaf sentences of everyday speech; CUNY = City University of New York sentences in noise; MCFA = middle cranial fossa approach; RSA = retrosigmoid approach; TLA = translabyrinthine approach; CNC = consonant-nucleus-consonant.

tients recruited from 3 centers. Three patients presented useful residual hearing in the contralateral ear at the time of implantation, while the remaining 4 subjects underwent implantation under more traditional criteria (0% speech reception score at the time of implantation). Within the latter group, the functional results varied considerably, ranging from excellent levels of open-set speech discrimination (46% on the Speech Discrimination Score and 98% on the Hearing in Noise Test in 1 patient) to simple improvement in environmental sound awareness and lip-reading skills. Regardless of the testing results, all patients reported benefits from cochlear implantation and used their device on a daily basis. In the other multi-institutional study Neff et al. [2007] reported on long-term hearing outcomes of cochlear implantation in 6 NF2 patients. Five patients achieved excellent functional results, scoring from 90 to 100% on Central Institute for the Deaf sentences and from 83 to 96% on the Hearing in Noise Test (average follow-up: 7.9 years). Interestingly, in 2 patients of this series EPS was negative in the immediate postoperative period but became positive 2 months after surgery. For this reason, Neff et al. [2007] stressed the importance of ESP in the assessment of the physiological viability of the cochlear nerve at the end of surgery, suggesting to delay electrophysiological evaluation 6–8 weeks after tumor removal. The same experience on initial negative result in EPS in the immediate postoperative period was described in the case reports published by Graham et al. [1999] and Hoffman et al. [1992]. In both studies the electrical stimulation of the cochlear nerve became successively positive and a successful cochlear implantation was performed in both patients. A period of neuropraxia was considered a possible cause of this phenomenon.

In the present study, hearing performance of subjects provided with a CI clearly surpassed that achieved by subjects fitted with ABI. At the 1-year follow-up, the performance in closed-set testing was similar: a mean vowel identification score of 85% in the CI group and 78% in the ABI group, and a mean consonant identification score of 85% in the CI group and 73% in the ABI group. A considerable difference resulted in open-set tests, with a mean common phrases comprehension score of 60% in the CI group and 29% in the ABI group, a mean sentence recognition score of 55% in the CI group and 27% in the ABI group, and a mean bisyllabic word recognition score of 53% in the CI group and 32% in the ABI group. In accordance with results reported in the literature, also in our study hearing performances with ABI were variable, ranging from the ability to understand speech by using only the sound from the device to simple improvement in environmental sound identification and lip-reading skills. Several factors related to the physiopathology of auditory pathway damage (excitability status of cochlear nerve and nuclei) as well as to the device properties (model, number of active electrodes, stimulation strategy) impact on ABI functional results.

Among the 4 patients fitted with a CI, 3 achieved open-set speech recognition abilities comparable to those of standard adult postlingual implant patients, while the remaining patient reported benefits only in environmental sound detection and lip-reading. Interestingly, this patient was the only one with a negative response on electrophysiologic testing. Although EPS testing presents some shortcomings, mainly because based on a subjective assessment, we agree with other authors [Friedmann et al., 1998; Marangos et al., 2000] in considering ESP as a useful test in the preoperative evaluation of NF2 patients selected for cochlear implantation. In the same way, when implantation is performed concomitantly with tumor removal, CNAP study can provide information about the residual functionality of the cochlear nerve, assisting in the choice between cochlear and brainstem implant. This is the case of patient 4 in the group of subjects fitted with a CI. Preoperatively, the patient was counseled and informed about the possibility to proceed to cochlear or auditory brainstem implantation depending on the presence or not of a residual functionality of the cochlear nerve at the end of tumor removal. Intraoperatively, there was convincing evidence of anatomical preservation of the cochlear nerve, as confirmed by the recording of a reliable CNAP. Thus, a CI was inserted into the scala tympani. This patient developed excellent open-set speech discrimination abilities and she was able to easily sustain a telephone conversation with unknown speakers. Electrophysiologic study is necessary but not sufficient to take the decision to provide NF2 patients already operated on for VS removal with a CI. Cochlear patency has to be confirmed by high-resolution computed tomography or magnetic resonance imaging, because rapid and progressive osteoneogenesis can occur into the cochlea after VS resection, mainly in the labyrinthectomized ear [Chen et al., 1988; Belal, 2001]. However, as reported in the literature, cochlear implantation has been successfully performed up to 18 months after labyrinthectomy [Kveton et al., 1989; Facer et al. 2000]. In our series, no ossification was observed in the 3 patients who underwent cochlear implantation in a second stage after tumor removal.

Another issue in cochlear implantation of NF2 patients concerns the possibility that hearing performance deteriorates over time if the cochlear nerve function declines as a result of postsurgical scarring or tumor regrowth. Currently, the only report on long-term results [Neff et al., 2007] demonstrated that hearing performance did not deteriorate over an extended postoperative time course. Whether or not the benefits of cochlear implantation in patients with NF2 remain stable over time, requires multi-institutional, prospective trials.

In conclusion, this study confirmed that a successful hearing rehabilitation in deafened NF2 patients is possible with both prosthetic devices. As expected, the hearing outcomes were better with CI than with ABI. Electrophysiologic testing plays a fundamental role in choosing which auditory prosthesis is more suitable when the cochlear nerve has been anatomically preserved after tumor removal. If a residual neural excitability can be demonstrated, CI should be preferred to ABI, not only because CI patients perform better than ABI patients but also because cochlear implantation presents minor surgical risks compared to auditory brainstem implantation. In the presence of an intraoperative positive CNAP, tumor removal and cochlear implantation can be performed in the same stage, avoiding a second surgical procedure. If intraoperative CNAP is absent, the viability of the cochlear nerve should be retested at least 2 months after surgery, since electrical stimulation can be negative in the immediate period after tumor removal. Cochlear implantation can be done if postoperative EPS demonstrates an excitable eighth nerve; when postoperative EPS is negative, ABI remains the best option for hearing rehabilitation.

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