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Auditory Brainstem Implants in NF2 Patients: Results and Review of the Literature

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Objective: Neurofibromatosis Type 2 (NF2) patients have multiple central nervous system tumors and, specifically, bilateral vestibular schwannomas (VSs) causing bilateral deafness. If the cochlear nerve is not preserved during tumor removal, the only hearing rehabilitation in these patients could be via an auditory brainstem implant (ABI).
Study Design: Retrospective case study and literature review.
Setting: Tertiary referral cranial base center.
Patients: In 24 NF2 patients, 25 ABIs were placed in the lateral recess of the fourth ventricle after VS surgery via a translabyrinthine approach.
Results: In this series, a large range of results are observed: from open speech and use of the telephone to no ABI use, because of the poor sound identification ability. Of the 24 patients, 19 use their ABI on a daily basis, 4 are nonusers, and 1 died of NF2 progression. A multivariate analysis did not reveal a good predictor for ABI outcome. In literature, the results of ABI in NF2 are difficult to compare, and the overall outcome was poor compared with cochlear implantation results.
Conclusion: Auditory brainstem implantation in NF2 patients directly after tumor removal is a safe procedure and the best means of hearing rehabilitation if the cochlear nerve is not preserved. The results in NF2 cases in the literature and these series are poor compared with cochlear implantation. If a cochlear implant is possible, it has the preference over an ABI, also in NF2. Nevertheless, the majority of the patients have benefit of the ABI during daily life particularly in combination with lip reading. **Key Words:** Brainstem—Hearing rehabilitation—Implantation—Neurofibromatosis—Review—Vestibular schwannoma.

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In neurofibromatosis Type 2 (NF2), the development of bilateral vestibular nerve schwannomas is the distinctive feature. NF2 patients develop next to their cerebellopontine angle tumor multiple spinal cord, brain and peripheral nerve tumors throughout the course of their life. Because of pressure of the tumor in the internal auditory canal, the intracanalicular portion of the cochlear nerve is prone to decrease in function causing progressive sensorineural hearing loss. Also, tinnitus, poor discrimination, and dysequilibrium could be symptoms of a vestibular schwannoma. Once the tumor is compressing the brainstem or compromising the lower cranial nerves, surgery is inevitable for these bilateral tumors. However, such a treatment often necessitates sacrifice of the VIIIth cranial nerve, consequently impeding hearing restoration by means of a cochlear implant. NF2 patients are rarely cochlear implant (CI) candidates; although if the cochlear nerve is preserved, they could greatly benefit from this (1–5).

Hearing rehabilitation in the absence of a cochlear nerve is demanding a different approach to the auditory pathway, which led to the development of a single-channel auditory brainstem implant (ABI). After Simmons et al. (6) had failed in stimulating the inferior colliculus, the first successful brainstem implant was performed in 1979 by House and Hitselberger (7,8). The first multi-channel implant was presented in 1991 (9) and was slightly modified in 1993 (10). Currently, there are several brainstem implants on the market.

One would expect that the outcomes with an ABI would progressively increase as the number of users and experience increases. However, a rising number of ABI user is not equivalent to a better result, as the main indication (NF2) embraces more than just the placement of an implant. Surprisingly, the outcomes with an ABI are much more variable than with cochlear implantation. The question arising from this situation is what factors in ABI placement predominantly influence the outcome?

The aim of this study was to assess the ABI outcomes in NF2 patients at our center, discuss the preoperative,

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perioperative, and postoperative special care in NF2 ABI patients and to compare our results with the results in literature. Factors that play a role in the outcome of ABI hearing rehabilitation are discussed.

PATIENTS AND METHODS

Patients

From October 1986 through July 2010, we operated in the Gruppo Otologico Piacenza–Roma 2313 acoustic neuromas in NF2 patients and non-NF2 patients. Of the 46 NF2 patients, 24 NF2 patients with a median age of 35 years (18–69 yr) have received 25 ABIs. Five NF2 patients received a cochlear implant. All ABI recipients met the following inclusion criteria: diagnosis of NF2, requiring a tumor removal at the first or second side, and having oral language competency. Furthermore, they had reasonable expectations and psychological stability and are able to follow rehabilitation. All patients were informed (orally and in writing) of the risks and benefits of the procedure.

Twenty-five implants were used in 24 patients (Table 1). Twenty-four Nucleus ABI24M implants (Cochlear, Sydney, Australia) and 1 Digisonic SP ABI (Neurolec MXM, France). One patient received a second implant at the time of the second tumor removal, after dislocation of the first implant (Patient 12).

Auditory Brainstem Implant

The Nucleus ABI24M auditory brainstem device (Cochlear, Sydney, Australia) has a flat silicone plate electrode carrier (3 × 8.5 mm) with 21 plate electrodes (each 0.7 mm) and a removable magnet. The Digisonic SP ABI (Neurolec MXM, France) has 15 electrodes (0.7 mm) on an electrode carrier (3 × 8 mm) but no removable magnet. In patients with NF2, the removal of the magnet can be important as continuous follow-up with magnetic resonance imaging (MRI) is necessary. The ABI (both devices mentioned) is MRI compatible (at 1.5T) without removal of the magnet from the receiver–stimulator but with special precautions (head bandage and information about the pulling sensation) and creating an artifact on the magnetic resonance (MR) scan. A MR scan with a higher magnetic field or less artifact can only be achieved with removal of the magnet. In the United States, the rules about the scanning with the magnet in place are more strict and have led in NF2 patients to remove the magnet and use an adhesional disk to the scalp (11). Other companies do not have an ABI with the ability to remove the magnet, which makes them less favorable.

The external part includes a microphone headset, the Nucleus SPrint sound processor, and a transmitter coil. The processor, also used with cochlear implantation, uses the Nucleus SPEAK spectral peak speech coding strategy. In this strategy, only 20 of the 21 electrodes are used. In the Neurolec MXM ABI, we used the MPIS mean peak interleaved sampling strategy, in which all 15 electrodes are used.

Surgery

The surgical approach for implantation of ABI has been via the modified translabyrinthine route in all patients (12). In our experience, this approach provides the most direct access to the site of the cochlear nuclei, the best visualization of the relevant anatomy and is the best way to remove a vestibular schwannoma. Even in large size tumors, the translabyrinthine approach (TLA) is preferable (12).

Further surgical and postoperative information, including preservation of the cochlear nerve, surgical landmarks, exact placement of the electrode, avoiding cerebrospinal fluid (CSF) leak, and other surgical instructions, is provided as Supplemental Digital Content, available at http://links.lww.com/MAO/A98.

Intraoperative Monitoring and EABR

At the time of electrode array placement, proper positioning is confirmed via evoked auditory brainstem response (EABR) audiometry. Stimulation is administered via electrodes at the

TABLE 1. *Auditory brainstem implant patient characteristics*

Patient	Age (yr)	Sex	Tumor size (cm)	Side of auditory brainstem implant	Auditory brainstem implant	Follow-up (mo)	User
1	34	Female	3.0	Second	ABI N24	53	Yes
2	43	Female	2.5	Second	ABI N24	44	Yes
3	33	Female	4.4	First	ABI N24	40	Yes
4	35	Male	0.5	First	ABI N24	46	Yes
5	41	Male	4.0	Second	ABI N24	33	Yes
6	39	Male	8.0	First	ABI N24	30	Yes
7	69	Female	1.5	First	ABI N24	23	Yes
8	26	Female	2.0	First	ABI N24	18	Yes
9	20	Male	5.0	First	ABI N24	†	†
10	30	Female	3.0	First	ABI N24	3	Yes
11	20	Female	3.0	Second	ABI N24	12	Yes
12a	52	Male	5.0	First	ABI N24	24	No
13	44	Female	*5.0*	Second	ABI N24	4	No
14	18	Female	2.5	First	ABI N24	2	Yes
15	39	Male	3.0	First	ABI N24	12	No
16	18	Male	2.0	First (residual)	ABI N24	16	Yes
17	26	Female	4.0	Second	ABI N24	6	Yes
18	51	Female	3.0	First (residual)	ABI N24	8	No
19	28	Female	2.5	Second	ABI N24	10	Yes
20	51	Male	1.0	Second	ABI N24	7	Yes
21	25	Female	5.0	First	ABI N24	4	No
22	20	Female	2.0	First	ABI N24	7	Yes
23	26	Male	3.0	First	ABI MXM	36	Yes
12b	54	Male	2.1	Second	ABI N24	7	Yes
24	44	Female	3.0	First	ABI N24	2	Yes
Average	**35 years**		**3.0 (±1.7) cm**				

4 corners of the array, and the presence or absence of EABRs at each location is used to guide any repositioning that may be necessary. Attention should be paid as this could be a time-consuming task and requires an experienced electrophysiologist, who provides feedback to the surgeon. EABR typically comprise 2 or 3 visible waves from an acoustic ABR. The 3 wave responses probably represent Wave III (the CN itself) and Waves IV and V, whereas 2-wave responses may be Waves III and IV or Waves IV and V (Fig. 1) (13).

Monitoring of the facial and glossopharyngeal nerves is performed simultaneously to detect undesirable stimulation of these nerves. Indirect monitoring of the Xth cranial nerve can be done through electrocardiographic recording; direct monitoring can be done via a special tube with monitoring vocal fold contractions.

Device Fitting and Programming

Fitting and adjustment were scheduled at 1, 3, 6, 12, and 24 months. At initial stimulation, there was a setting of the threshold and comfort levels, evaluation and management of nonauditory stimulation, and pitch scaling. Monopolar stimulation was administered via each electrode, and the auditory and nonauditory sensations are recorded. In some cases, bipolar stimulation had less nonauditory side effects and is therefore activated. In our series, we stimulate 18 patients with monopolar stimulation and one in the variable modus.

The next step is to pitch scale the electrodes with auditory responses by comparing the electrodes and order them pitch-like accordingly. Electrodes without an auditory sensation were eliminated, and the pitch gaps were filled by simultaneous stimulation of low- and high-sounding electrodes, creating a ''new'' virtual intermediate electrode.

Speech and Sound Perception Measures

The following speech and sound perception measures were taken in an open-set (i.e., understanding words or sentences without alternatives from which to choose the answer) format: detection of environmental sounds (sounds), vowel and consonant identification, bisyllabic word recognition (word), common phrases comprehension (sentence), and open-set speech recognition (speech). 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, and rattle for high frequencies). The speech materials were presented using monitored live voice through the sound field at a level of 70 dB sound pressure level and signal-to-noise ratio of 15 dB. The setup is presented in Figure 2. Postoperative auditory speech performance was measured by using the following tests: Italian version of the Northwestern University Phonetically Balanced Word List (NU 6) and Central Institute for the Deaf Everyday Sentence List.

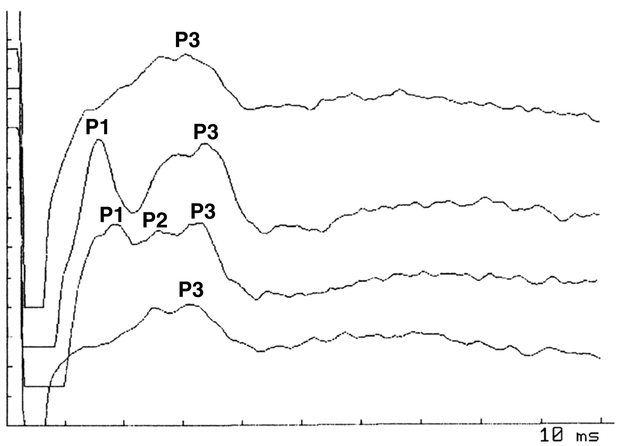

FIG. 1. Intraoperative EABR typically comprise 1, 2, or 3 visible waves from an acoustic ABR. The 3 wave responses probably represent Wave III (the CN itself) (P1), Wave IV (P2), and Wave V (P3), whereas 2-wave responses may be Waves III and V or Waves IV and V.

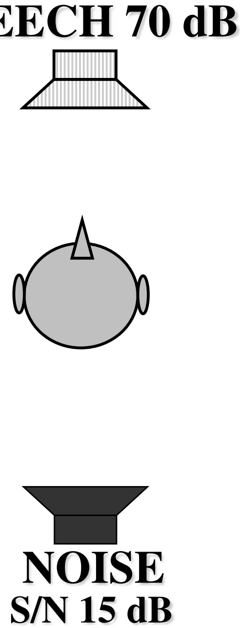

FIG. 2. Setup for the audiologic speech test (words, sentences, and questions) in acoustically isolated room. The test setup is with a recorded voice in front and with noise in the back with signal-to-noise relationship S/N 15 dB.

The open-set common phrases comprehension test was based on common and simple interrogative phrases (e.g., ''How are you feeling?'') to which the patient had to respond; test scoring was based on the percentage of correct responses. We always ask our patients simple questions such as the following: do you use your device daily? Can you use telephone? If you can use telephone, can you use it just with people whose voices are familiar for you (like your family, your friends etc.) or with all people? In the open-set speech 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. In this study, we present the results of hearing tests in auditory-only condition collected at the latest follow-up after implantation.

Literature Review

A literature review was performed using search themes in PubMed and Medline: ''neurofibromatosis Type 2,'' ''NF2,'' ''vestibular schwannoma,'' ''acoustic neuroma,'' ''auditory brainstem implant,'' ''ABI,'' and all the combinations. A selection was made based on English language and, especially, if the article reported on NF2 patients who received an ABI. Single case reports on NF2 and ABI placement were not taken into account. Some articles of the same center report on the same patients consecutively, sometimes in different articles simultaneously or in multiple articles with or without the outcomes of patients of other centers. Some series report on all their ABI patients as one group with various indications, instead of presenting a NF2 group alone. The above-mentioned reporting issues are the reason why comparison or estimation on the amount and results of ABI in NF2 patients in general is redundant and imprecise.

RESULTS

Twenty-four patients have received 25 ABIs (Table 1). One patient had already an implant in the contralateral ear at referral (Patient 11). Average age was 35 years (18–69 yr), average tumor size 3.0 cm (SD ± 1.7). The patients received their ABI (24 Nucleus 24M ABI, 1 Digisonic SP ABI) at the removal of the first vestibular schwannoma (14 times), at the removal of the residual first tumor (2 times), or at the removal of the second tumor (9 times). Two of these patients (Patients 11 and 12) received an ABI at first and second side. First side operation was of Patient 11 was done in another center and, therefore, will not be calculated. Both patients had dislocation of their first ABI. We implanted a second ABI at the contralateral ear with better results.

Preoperative, intraoperative, and postoperative problems were encountered more regularly than in the normal vestibular schwannoma patients or compared with ABI placements in nontumor cases. Preoperative problems related to the NF2 were unilateral and bilateral facial nerve paralysis (Patients 6 and 18) and voice and swallowing problems due to lower cranial nerve palsy (Patient 20). Preoperative hearing loss was variable. Some had a normal hearing on both sides, whereas bilateral deafness was common (Table 2).

Intraoperative and Postoperative Outcomes

We have not encountered CSF leaks (49), pseudomeningoceles, or lower cranial nerve palsies due to the surgery or other complications, like vascular damage or

TABLE 2. *Electrodes, side effects, and auditory outcomes*

No.	Hearing preoperatively	Follow-up (mo)	Electrode activation	Side effects at activation	Electrode used	User	Sound	Words	Sentence	Speech
1	Bilaterally deaf	53	21	None	21	Yes	100	90	100	100[a]
2	Bilaterally deaf	44	14	XI ipsilateral	19	Yes	100	70	72	80[a]
3	Contralaterally normal	40	20	Vertigo, XI	18	Yes	100	80	70	75[a]
4	Bilaterally deaf	46	13	XI ipsilateral	13	Yes	70	0	0	0
5	Bilaterally deaf	33	17	VII	19	Yes	60	0	0	0
6	Contralaterally severe hearing loss	30	11	IX	10	Yes	90	30	15	20
7	Severe hearing loss	23	11	Vertigo	13	Yes	80	10	0	0
8	Contralaterally normal	18	17	VII, IX, XI	12	Yes	100	30	35	30
9	Bilaterally deaf	†				†				
10	Contralaterally normal	3	5	IX, XI ipsilateral	10	Yes	90	65	35	40
11	Auditory brainstem implant contralateral	12	7	XI ipsilateral	6	Yes	80	10	0	0
12	Contralaterally normal	24	0	IX, XI ipsilateral	0	No				
13	Bilaterally deaf	4	0	XI ipsilateral	0	No	0	0	0	0
14	Bilaterally normal	2	8	Vertigo and headache	8	Yes	70	0	0	0
15	Moderate hearing loss, contralaterally normal	12	0	XI ipsilateral	0	No	0	0	0	0
16	Bilaterally deaf	6	21	None	21	Yes	100	60	65	70[a]
17	Bilaterally deaf	6	13	XI ipsilateral	13	Yes	40	0	0	0
18	Bilaterally deaf	8	0	IX, XI ipsilateral	0	No	0	0	0	0
19	Bilaterally deaf	10	12	None	12	Yes	85	20	10	0
20	Severe hearing loss	7	5	Vertigo, headache	5	Yes	60	0	0	0
21	Contralaterally severe hearing loss	4	0	Vertigo/vertigo and acoustical	0	No	0	0	0	0
22	Bilaterally deaf	7	17	None	17	Yes	75	0	0	0
23	Bilaterally deaf	36	9	XI	9	Yes	70	0	0	0
12b	Contralaterally auditory brainstem implant, moderate hearing loss	7	11	Vertigo	11	Yes	85	20	15	20
24	Bilaterally deaf	2	21	None	20	Yes	65	25	0	0
			12		13					

Mean electrodes used.

Patient 9 died of the progressive NF2 (†) during follow-up.

Sounds = detection of environmental sounds (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, and rattle for high frequencies). Words = bisyllabic word recognition (Italian version of the Northwestern University Phonetically Balanced Word List (NU 6).

Sentence = common phrases comprehension. Test scoring was based on the percentage of correct responses. Speech = open-set speech recognition: a list of 10 uncommon sentences of the Central Institute for the Deaf Everyday Sentence List was presented to the patient: each list contained 100 words and was scored for the total number of words correctly repeated.

Sounds/words/sentences/speech, audiologic tests as explained in the text; Roman numericals correspond to corresponding cranial nerve stimulation as side effect.

[a]Patient uses telephone.

hemorrhage. A facial nerve graft was performed in 1 patient because it was impossible to preserve the facial nerve (Patient 16). There were no other intraoperative problems encountered. A patient (Patient 9), which was radiated twice before surgery, had a malignant tumor (confirmed on histopathology), which was removed at the time of the implantation. The patient died 3 months later after normal postoperative outcome.

In each case, a computed tomographic (CT) scan was performed in the postoperative phase to rule out displacement of the ABI electrode (Fig. 3).

In the postoperative period, 1 patient had a CSF collection beneath the skin, which was resolved by aspiration and a compression head bandage for several days (Patient 8). Another patient developed an infection, and the ABI was extruded and had to be surgically removed (Patient 13). In 3 cases (Patients 12, 15, and 17), we have seen a dislocation of the electrode array. Another patient with no hearing abilities was lost to follow-up after 19 months of rehabilitation (Patient 7). Lower cranial nerve stimulation or vertigo due to activation of other cranial nerve nuclei was seen in many patients (Table 2). After altering the proper fitting and sometimes disactivating electrodes, these problems disappeared.

Evoked Auditory Brainstem Response

As already mentioned to assist the placement of the ABI, an intraoperative EABR is essential. A 1-, 2-, or 3-peak response was observed in all of the cases. Time consuming was the EABR assessment of all 21 electrodes (Nucleus 24M ABI) in each surgery (in Neurolec-MXM ABI 15 electrodes). A strategy of a ''cross-sectional check'' for peaks was applied in which the first 4 electrodes were measured and subsequently adjacent electrodes. A continuous feedback to the surgeon was given, mostly by means of a schematical rough drawing (Fig. 4: drawing of

FIG. 3. Three-dimensional CT scan of the postoperative result after placement of a right-side ABI. The CT scan is performed to check if no displacement has occurred.

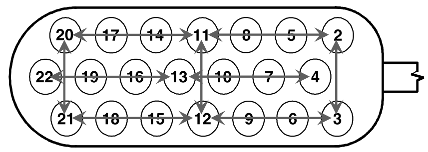

FIG. 4. Schematic drawing of the electrode testing intraoperative. Scheme of stimulation bipolar of the different electrodes to get a correct positioning of the array. The first electrode test is 13 toward 22 and after 13 toward 4, then 11 toward 20 and 11 toward 2 and, subsequently, the other electrodes.

Nucleus cross-sectional testing) and as soon as most of the electrodes had a clear EABR closure of the TLA was started.

Electrodes and Channels

At initial stimulation, median of 12 electrodes of the available 21 electrodes gave auditory responses (Patient 23: 9 of the available 15 electrodes; Table 2). The electrodes with no response or nonauditory response were deactivated. In the weeks and months after initial activation, we have encountered sometimes a shift from nonauditory to auditory or vice versa requiring deactivation of those electrodes. This effect, probably because of the repositioning of the brainstem after removal of the tumor, is seen up to 2 months after surgery. An increased number of electrodes showed an auditory response over time (mean of 12 turned to 13 electrodes used, Table 2), which is remarkable compared with other studies (13).

In the counseling of the patient, we found it very helpful to stress the fact that the sounds can be very disappointing in the beginning but with time and training, the sound quality will slowly but surely increase.

Audiologic Results

The overall results are unpredictable, 19 of the 23 patients we could follow-up on are users; 19 have sound recognition, 11 some kind of word recognition and only 8 speech recognition (Fig. 5). Of the few with speech recognition, some are very good users, with 4 with more than 50% speech discrimination and 4 with even telephone use and 75% to 100% speech discrimination (Table 2 and Fig. 5).

Tumor Size

We have analyzed the outcomes of the patients comparing the group with tumor size below 3 cm with 3 cm and above. The average sentence and speech scores turned out to be 20% and 20% in the group with tumor size less than 3 cm and 17% and 18% with tumor size of 3 cm or greater. Therefore, a relation between tumor size and ABI outcome remains uncertain.

Multivariate Analysis to Reveal Predictors of Good ABI Performers

A multivariate analysis (logistic regression, SPSS®, IBM® version 15) was performed with the question ''what factors influence the outcome of ABI?'' Patients with a

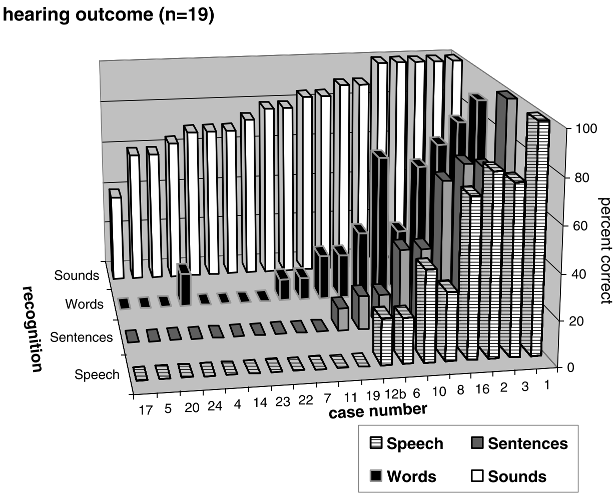

FIG. 5. Audiologic outcomes of the patients who are users of their ABI. *x* axis represents the number of the case/patients in our series and matches patient characteristics in Tables 1 and 2. *y* axis is the test used, either sound recognition test, word recognition test, sentences test, and speech recognition test. *z* axis percentage of correct answers.

speech outcome of 70% or more were compared with the other users with less speech performance.

The following four factors were analyzed: age, size of the tumor, the number of electrodes at activation/with good response on eABR during surgery, and the number of electrodes used/with audiologic response during rehabilitation (Table 2).

All of the factors were not significant in a forward selection process, except factor 4 (number of electrodes used during rehabilitation). When plotted in a regression model also, this factor did not have enough significance (p value = 0.114). In conclusion, we were not able to identify statistical significant factors that influence the outcome of ABI performance. Therefore, it remains hard to predict the outcome of an individual NF2 patient receiving an ABI.

Literature

In comparing the ABI hearing abilities of NF2 patients with other publications, we encountered difficulties because of methodologic differences: a diversity of tests, graph presentation, and a variable group selection. Then, the way of presenting the selected patients, the number of users, the side effects, the number of electrodes used, and other specifications also were numerous. Tables 3 and 4 show most of the variables as distilled from the articles from each center/group. Most of the articles present their audiologic outcome differently, which makes them hard to compare. Of the patients who had follow-up (excluding deceased and nonactivated), the majority had auditory sensations and, mostly, an improvement in communication

TABLE 3. *Literature review and methods used in auditory brainstem implant case series concerning neurofibromatosis Type 2 patients*

Institute	First author	Year	Cases	Age	Implant	Approach	Technique ABI
Paris	**Grayeli (14)**	2008	23	17–65	Nucleus 21	TLA	
MHH	**Lenarz (15)**	2001	14	24–61			
MHH	**Lenarz (16)**	2002	14	24–62	Nucle4s 22 (n = 8), 24 (n = 1), Clarion (n = 5)	TLA (n = 1) and RS (n = 13)	Lateral recess EABR, probe
MHH	**Lesinski-Schiedat (17)**	2000	8	24–52			
EUR	**Vincent (18)**	2002	14	14–56	MXM Digisonic ABI	TLA (n = 11), RS (n = 3)	Choroidea EABR
EUR	**Nevison (13)**	2002	27	13–58	Nucleus 20/21	TLA (n = 21)/RS (n = 6)	Lateral recess EABR NRT
HEI	**Otto (19)**	2008	10	19–53	PABI	TLA	Choroidea EABR
HEI/US	**Ebinger (20)**	2000	92	12–67	Single and N22	TLA	Lateral recess EABR NRT
HEI	**Otto (21)**	2002	61	12–71	N22	TLA	Lateral recess EABR NRT
HEI	**Schwartz (11)**	2008	230	nm	Nucleus 8 and 21	TLA	Lateral recess EABR NRT
Fulda	**Behr (38)**	2006	20	18–56	Med-El Combi 40+	RS	Lateral recess ABR NRT glue
Ver	**Colletti (22)**	2006	14				
Ver	**Colletti (23)**	2005	10	17–70	Nucleus 22/24	RS	Lateral recess ABR NRT
Ver	**Colletti (24)**	2005b	10				
Frei	**Laszig (10)**	1995	9	nm	N22 and N24	TLA	Lateral recess ABR NRT
Frei	**Marangos (25)**	2000	15	17–58	N20 (n = 2), N21 (n = 11), N24 (n = 1)	TLA	
Frei	**Sollmann (26)**	2000	55	33 mean	N24	TLA/RS	Lateral recess ABR NRT
NY	**Kanowitz (27)**	2004	18	15–55	N22 (n = 12) and N24 (n = 6)	TLA (n = 16), RS (n = 2)	Follow c.n. NVIII, probe
Mel	**Maini (28)**	2009	11	17–46	Nucleus 22 and 24	TLA (n = 9), RS (n = 1)	Lateral recess EABR NRT
Piac	**This study**	2010	23	18–69	Nucleus 24 (n = 22), MXM (n = 1)	TLA (n = 23)	Choroid plexus EABR[a]

Choroid plexus, choroidea, and lateral recess are surgical landmarks for the ABI placement.

EABR indicates evoked/electrical auditory brainstem response; EUR, European ABI research group; Frei, University of Freiburg, Germany; Fulda, University of Marburg, Fulda, Germany; Glue, use of glue for fixation of ABI; HEI, House Ear Institute, Los Angeles, U.S.A.; Mel, Melbourne Cochlear Implant Clinic, Australia; MHH, Medical University Hannover, Germany; MXM, MXM Digisonic ABI; N8, N20, N21, N22, N24, multichannel Nucleus ABI systems; NY, NYU Medical Center, New York, U.S.A.; PABI, penetrating ABI; Paris, Hôpital Beaujon, Clichy, France; Piac, Gruppo Otologico, Piacenza–Rome, Italy; Probe, to verify place of cochlear nucleus before inserting ABI; RS, retrosigmoid approach; Single, single channel ABI; TLA, translabyrinthine approach; US, United States ABI research group; Ver, University of Verona, Italy.

[a]EABR monitoring c.n. VII, IX, X, XI, and XII more important than neural response telemetry (NRT).

TABLE 4. *Literature review and outcomes of auditory brainstem implant case series in neurofibromatosis Type 2 patients*

Institute	First author	Year	Daily users	Nonauditory side effects	Open-set sentence recognition (auditory alone)	No auditory response	Auditory response	Follow-up	Active electrodes	Complications
Paris	**Grayeli (14)**	2008	70% (16/23)			22% (5/23)	"Out the 16 'ABI users': 8 had open speech >50% discrimination, 5 limited to speech and sound and 3 only sound awareness. Furthermore, 5 no auditory awareness, 1 not activated, and 1 died of NF2."	1–10 yr		CSF leak, meningitis, embolism, CPA hematoma, death of NF2 progression
MHH	**Lenarz (15)**	2001	93% (13/14)		0% (0/14)	8% (1/13)	"Combination of lip reading and ABI produced an improvement in speech tracking in all patients over time" "Open-set speech recognition in auditory alone mode was not common"	1–41 m		Migration of ABI (n = 1)
MHH	**Lenarz (16)**	2002								
MHH	**Lesinski-Schiedat (17)**	2000			13% (1/8)				5–15	
EUR1	**Vincent (18)**	2002	86% (12/14)	7% electrode in 36% (5/14)	21% (3/14)	7% (1/14)	"5 of 9 scored 70% open-set sentence correct answers using sound and vision, 3 patients improved speech perception in sounds only mode, and 1 was able to use the telephone"			Repositioning of ABI after RS
EUR2	**Nevison (16)**	2002	88% (23/26)	92% (24/26)	8% (2/26)	4% (1/26)	"Although most subjects did not achieve any functional auditory alone open speech understanding, two subjects (of 26) were able to use the ABI in conversation without lip reading"		8.6 ± 4.2	Embolism and death, death of NF2, epileptic seizures
HEI	**Otto (19)**	2008		>20% (2/10)	2.6% (~3.3)		"Even after three years of experience, patients using penetrating electrodes did not achieve improved speech recognition compared with surface electrodes."		0–8 penetrating, 0–14 surface	
HEI/US	**Ebinger (20)**	2000	97%	66%	<5%	85% (75/88)	"Most listeners were unable to recognize words in sentences by using sound only" For most patients, the benefit in communication is used in conjunction with lip reading."			
HEI	**Otto (21)**	2002	90% (55/61)	Yes	~5% (0%–65%)	9% (6/61)		1–8 yr		CSF leak, meningitis
HEI	**Schwartz (11)**	2008		24% electrode	5% (CID)	15% (13/86)		1–8 yr		

Fulda	**Behr**	2006	90% (18/20)	11% electrode in 45% (9/20)	42% (n = 8)		‘‘lip reading was improved.’’ ‘‘For many patients, comprehension of open-set speech was restored to a useful level.’’			Dislocation of the ABI (n = 1), pseudomeningocele (5/20), hydrocephalus, facial paresis (6/20), swallowing problems
Ver	**Colletti (22)**	2006					A good auditory response of NF2 patients could not be distracted from the articles because of the report on inhomogeneous groups (adults, children), tumor (NF2 and VS) and various nontumor indications.			
Ver	**Colletti (23)**	2005		69% (20/29)	5%–35%				12–21	Myocardial infarction,
Ver	**Colletti (24)**	2005b								
Frei	**Laszig (10)**	1995	44% (4/9)		0%	11% (1/9)	Only data of 3 patients, no patients with open-set speech recognition in auditory alone mode			Death due to infections,
Frei	**Sollmann (48)**	2000	87% (48/55)				‘‘most patients are able to recognize different words... the device facilitates lip-reading and useful in daily life (1995).’’ ‘‘three of the 48 patients were able to use the telephone (2000)’’		2–21	nVII sacrifice (n = 2), late failure of ABI after radiotherapy (n = 2), pulmonary embolism death (n = 1)
NY	**Kanowitz (27)**	2004	61% (11/18)	Yes, minor	0% (CID)	6% (1/18)	‘‘No patients demonstrated an open-set speech recognition’’ ‘‘Auditory performance was consistently better with the Nucleus N24 device (n = 3) compared with the N22 device (n = 8)’’	2–78 m	Median 4/8 (N22 ABI) and 10/22 electrodes (N24 ABI)	CSF leak (n = 1), wound infection, facial nerve paresis (n = 2), facial nerve sarcrifice, seroma (n = 1), died of NF2
Mel	**Maini (28)**	2009	70% (7/10)	20% (2/10)	<5% (CUNY)	10% (1/10)	‘‘In the long-term, ABI provides an additional 35%–40% speech perception in the combined mode compared to lip reading alone.’’	1–12 yr		died of NF2

Legend of institutes, see Table 3.
ABI indicates auditory brainstem implant; CID, Central Institute for the Deaf sentence test; CPA, cerebellopontine angle; CSF, cerebrospinal fluid; CUNY, City University of New York sentence test; NF2, neurofibromatosis Type 2; RS, retrosigmoid approach; VS, vestibular schwannoma.

skills, especially in combination with lip reading. Nevertheless, compared with the results as seen with cochlear implantation, the outcome is, in general, poor.

In the management of NF2, the decision whether to place an ABI or CI is based on preoperative parameters, such as ipsilateral and contralateral tumor dimensions, and on intraoperative findings. Final decision is made on the basis of surgeon's awareness of anatomic maintenance of the VIIIth nerve (1). Furthermore, decision making during tumor removal via TLA or retrosigmoid approach is not easy. If there is any possibility in hearing preservation (tumor dimensions less than 1.5 cm and good hearing), a combined retrosigmoid-retrolabyrinthine approach is attempted, and complete cochlear nerve neuromonitoring (ABR and cochlear nerve action potential [CNAP]) is used to obtain information on the functional status of the CN. In case of TLA, the decision to implant a CI is based mainly on intraoperative preservation of neural integrity.

DISCUSSION

ABI speech perception results do not match with the good results seen in modern cochlear implantation, but the auditory sensations provided by ABI can be very helpful in facilitating oral communication of the NF2 patient. When asked in a questionnaire, many say they benefit greatly from their ABI (11). We have seen the same variable results including some very good users (4 with telephone use). Such a result is new in NF2 ABI recipients. Still, we have to be modest because the overall result is very variable, and 19 of the 23 patients we could follow up on are users, with only 8 patients who have speech recognition. The reasons for these variable outcomes are multiple. First, patient-related reasons: the patient could die of progression of the disease/other reasons or the patient could have multiple handicaps interfering with their audiologic training and outcome. Or the patient still has serviceable hearing on the contralateral ear. Second, there are implant-related reasons: stimulating other nuclei or nonfunctional electrodes. Third, surgical reasons: the implant could be displaced, the wound and implant could get infected and extruded/removed, or other complications would interfere with a ''normal'' placement or use of the ABI. Although we have not found a correlation between tumor size and ABI performance, it seems reasonable to presume that the size of the tumor and inevitably the displacement/distortion of anatomical landmarks could lead to an inconsistency between intraoperative measurements and postoperative results. In general, the results are unpredictable, and the counseling with the patient is therefore extremely important to minimize the expectations.

Overall literature is stating that the ABI in NF2 patients will enhance communication in addition to lip reading. This is in contrast to the results with a cochlear implant as they are much better and more predictable (Table 5). Arriaga and Marks (29) were the first to report a simultaneous cochlear implantation and vestibular schwannoma resection. They and others observed good results

TABLE 5. *Neurofibromatosis Type 2 patients who received a cochlear implant*

#	Sex	Age (yr)	Contralateral ear	Implanted ear	Electrophysiologic testing	Implant type	Device use	Sentence recognition (%)	Common phrases comprehension (%)	Telephone use
1	Female	47	VS (4 cm) removal via TLA (cochlear nerve sacrificed)	VS (1.5 cm) removal via RS: no hearing conservation	EPS: good response	Nucleus 24 Contour	Daily	90	100	Yes
2	Female	24	Anacusis after VS (1.5 cm) removal via RS	VS (2 cm) removal via TLA intact CN (CI in the same stage)	CNAP: good response	Nucleus 24 Contour	Daily	81	86	Yes
3	Male	32	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	50	55	Yes
4	Male	36	VS (4 cm) removal via TLA (cochlear nerve sacrificed)	VS (1.5 cm) removal via RS: no hearing conservation	Absent ABR; CNAP and EPS: doubtful response	Nucleus 24 Contour	Daily	0	0	No
5	Female	72	Intrameatal VS with sparing of IAC fundus	VS (2 cm) removal via TLA and EAC closure: intact cochlear nerve	CNAP: good response, NRT: good response	Nucleus 24 Contour	Daily	85	93	No

Most of the patients with a CI have better results with their cochlear implant than the majority of the NF2 patients who received an ABI.
Some details of this table are previously published by Vincenti et al. (5) and Odat et al. (1).
ABR indicates auditory brainstem response; CNAP, cochlear nerve action potential; EAC, external auditory canal; EPS, electrical promontory stimulation; IAC, internal auditory canal; NRT, neural response telemetry.

with cochlear implantation after vestibular schwannoma surgery (1,30,31), also in NF2 patients (2–4,32,33–36). Taking this fact into account, it seems clear that cochlear implantation will be the rehabilitation of choice in all NF2 cases with an intact cochlear nerve and cochlea.

However, how can we predict an intact and functional cochlear nerve? Arriaga and Marks (29) have suggested that an intraoperative promontory stimulation may be of some benefit in this setting, but we have found that intraoperative promontory stimulation is unreliable (37). Its unreliability is strengthened by reports of early postoperative false negatives occurring with the use of this technique (3,33). The CNAP seems a more helpful tool in the decision making between CI or ABI in NF2 patients (37). Although the numbers are limited, the results of using the CNAP are hopeful (Table 4) (1).

With the knowledge that the majority of the patients with NF2 will progress to bilateral deafness sooner or later, a rehabilitation of the hearing is necessary (11). In concordance with most centers (11,13,14,38), we advocate implantation at the time of the first-side tumor removal. A small percentage will not respond to their ABI, and these patients could have a second chance at the second removal. Patients should be carefully counseled as the results can vary considerably, and the device tuning and rehabilitation can take much longer than patients expect. Also important to mention are the complications as these seem higher than in the vestibular schwannoma patients as a whole, as most ABI centers, except one (27), report. We have not encountered pseudomeningoceles (38) or CSF leaks (11) but do share the experience of a higher incidence of aspiration and dysphagia (11). Ipsilateral and contralateral lower cranial nerve problems should therefore be carefully investigated.

The approach to remove the tumor and place the implant is possible via a retrosigmoid or a TLA. Because of several reasons, we prefer and promote the (enlarged) TLA: first, the complete tumor can be made visible; second, there is no limit in the size of the tumor, which can be removed in this manner (39); third, the facial and cochlear nerve can be closely followed during the whole surgery; fourth, the opening is wider and the choroid plexus and nerve endings come easily in view; and fifth, a cochlear implant, if needed, can be performed in the same approach. Placement of the ABI electrode is a difficult procedure. We tend to insert almost the entire length of the array into the recess and follow the choroid plexus as main landmark, as explained earlier.

In literature, the arguments of the different approaches are not discussed in great detail. Nevison et al. (13) state that the approach is not a major factor; still, they explain their preference for the TLA approach: ''A better angle to the lateral recess and the better orientation, as large tumors give compression and displacement of the brainstem which hampers ABI placement orientation.''

Which brand of ABI implant provides the best results? This is hard to tell as the numbers still are small. The only implants clinically compared are the Nucleus ABI N22 and N24 (Cochlear, Sydney, Australia) with similar outcomes in a larger series (11) or in favor of the newer N24 ABI in a smaller series (27). A penetrating ABI did not fulfill the high expectations (19). In the selection of an ABI implant, we advocate in favor of the removable magnet in the Cochlear N24 ABI (Cochlear, Sydney, Australia). The magnet creates an artifact covering the cerebellopontine angle on an MR image. In NF2 patients, sequential MRI is inevitable, and making a good strategy regarding ABI magnet removal is essential.

There is a growing understanding of true and false indications of ABI and also experience in other indications than NF2. The reports on the use of an ABI in nontumor patients are growing (40–45) but debatable if these results are overall better than those in tumor patients. Even some indications are questionable (46) as bilateral nerve avulsion seems more to be a theory as traumatic cochlear sensorineural hearing loss seems the true cause of the deafness. Cochlear implantation in these cases is always possible. Furthermore, we have operated some cases, who received an ABI in another center without hearing results, and they showed excellent results just by implanting a cochlear implant in the contralateral side (47). In our series, we have seen several postmeningitis cochlear ossifications with amazing results (44) but also with poor results. The key to success and a predictable outcome in ABI surgery has not been clarified till now. Some search for better options at a different location in the auditory pathway, for example, the midbrain implant (48) at the colliculus inferior, after earlier attempts of Simmons in 1964 (6), but results with this type of implant has not overthrown the ABI results.

CONCLUSION

Auditory brainstem implantation in patients with NF2 provides good support in the communications skills of the deafened patients, especially in combination with lip reading. Although the results of brainstem implantation are unpredictable, some patients achieve open-set speech discrimination and even telephone use. Because of the unpredictable results in our and other ABI NF2 series, a cochlear implant should be the rehabilitation of first choice; still therefore, a functional and intact cochlear nerve is necessary (49).

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