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Cochlear implants in the management of hearing loss in Neurofibromatosis Type 2

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Objective: Review of cochlear implant (CI) outcomes in patients with Neurofibromatosis Type 2 (NF2), implanted in the presence of an ipsilateral vestibular schwannoma (VS). Hearing restoration was combined in some cases with a Bevacizumab regime.

Method: Retrospective review of 12 patients, managed over the period 2009–2016, at a tertiary referral multidisciplinary NF2 clinic. The patients are grouped by hearing outcomes to explore likely protective factors, and to generate a proposed decision-making tool for the selection of either CI or Auditory Brainstem Implant (ABI).

Results: Four of the 12 patients achieved speech discrimination without lip-reading. In these individuals there is reason to think that the mechanism of their hearing loss was cochlear dysfunction. A further four patients received benefit to lip-reading and awareness of environmental sound. For such patients their hearing loss may have been due to both cochlear and neural dysfunction. Two patients gained access to environmental sound only from their CI. Two patients derived no benefit from their CIs, which were subsequently explanted. Both these latter patients had had prior ipsilateral tumour surgery, one just before the CI insertion.

Conclusion: Cochlear implantation can lead to open set speech discrimination in patients with NF2 in the presence of a stable VS. Use of promontory stimulation and intraoperative electrically evoked auditory brainstem response testing, along with case history, can inform the decision whether to implant an ABI or CI.

Keywords: Cochlear implants, Acoustic neuroma, Vestibular schwannoma, Neurofibromatosis Type 2, Bevacizumab

Introduction

Neurofibromatosis Type 2 (NF2) is an autosomal dominant condition resulting in multiple benign intracranial and spinal tumours. The incidence of this genetic disorder is one in 25 000–33 000 (Ferner *et al.*, 2014). NF2 typically presents with bilateral vestibular schwannomas (VS) (Evans *et al.*, 1992). Hearing loss is thought to be caused by a combination of neural loss (compression of the cochlear nerve by growing VS) and cochlear dysfunction. However, recent work by Dilwali *et al.* (2015) suggests that some VS may act specifically on cochlear function. Profound bilateral hearing loss has a major impact on quality of life for NF2 patients, leading to social isolation and loss of independence (Neary *et al.*, 2010). As of January 2016, 166 cases with NF2 are being managed in our unit. Of these, 56% currently experience significant hearing disability, as indicated by maximum speech discrimination scores of less than 50% in at least one ear using AB word lists (Boothroyd, 1968; Markides, 1978). Hearing loss assumes greater impact in the presence of wider disability, such as vision or mobility issues.

Surgical management of VS and options for hearing preservation/restoration are presented by Tysome *et al.* (2012). Options include: full or partial excision of tumours, radiotherapy, or drug treatment with Bevacizumab. Each of these can be accompanied by attempts to preserve hearing. Surgical success at preserving the cochlear nerve when excising VS is improving. This leads to the possibility of restoring hearing with a CI as an alternative to Auditory Brainstem Implant (ABI) (Amoodi *et al.*, 2012; Celis-Aguilar *et al.*, 2012; Lassaletta *et al.*, 2016; Lloyd *et al.*, 2014; Pai *et al.*, 2013; Roehm *et al.*, 2011). Radiotherapy, often advocated as a hearing preservation treatment in patients presenting with sporadic

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VS, does not have the same protective effect for patients with NF2 (Mallory *et al.*, 2014). Bevacizumab (Avastin ®), a monoclonal antibody that targets the Vascular Endothelial Growth Factor protein, has been associated with stabilising or improving hearing in the majority of cases (Morris *et al.*, 2016; Plotkin *et al.*, 2012). Once functional hearing is lost, consideration of the likely mechanism of the hearing loss will determine the best option for hearing rehabilitation. If the cause of hearing loss is considered to be cochlear dysfunction, then a CI may have more benefit than an ABI (Bento *et al.*, 2013; Monteiro *et al.*, 2012; North *et al.*, 2016; Tysome *et al.*, 2012).

We present the outcome of 12 NF2 patients who underwent cochlear implantation and we discuss possible reasons why their outcome is so varied. This case series has helped us formulate a potential counselling tool, for determining which cases may be better to proceed to ABI or to CI.

Method

A retrospective review of 12 NF2 patients who underwent CI, 2009–2016. Selection criteria for CI in NF2 cases at our tertiary referral multidisciplinary NF2 clinic are as follows:

- stable VS, or VS controlled by Bevacizumab, or removal of VS with cochlear nerve preservation;
- unilateral or bilateral severe-profound hearing loss – thresholds higher than 90 dB HL at 2000 and 4000 Hz;
- poor functional communication skills in at least one ear – less than 50% accuracy on the standardised Bamford–Kowal–Bench (BKB) sentence repetition tasks without the aid of lip-reading (Bench *et al.*, 1979);
- for those without response on behavioural hearing test, possible promontory stimulation (carried out pre-operatively) or electrically evoked auditory brainstem response (eABR) (conducted intraoperatively);
- for those undergoing VS removal, positive response to eABR.

Promontory stimulation (in the clinic) involves inserting a trans-tympanic needle electrode located at the round window niche, and a small electrical current is used to stimulate the cochlear nerve. The patient is invited to report the presence or the absence of auditory percept when the nerve is stimulated. EABR when conducted intraoperatively can help determine the response of the cochlear VIII nerve to electrical stimulation. A single use temporary electrode (based on a Med-El Combi 40+ implant) is inserted into the cochlea, and an external receiver stimulator used to stimulate one of the three available electrodes (Kasbekar *et al.*, 2012). However, the measurements from eABR may not correlate well with the hearing outcome post-operatively.

The patients were grouped by their speech discrimination outcomes as follows:

- Group A: Open set function: Able to listen without lip-reading to sentences spoken out of context. This was measured by score ≥ 65% on the BKB test;
- Group B: Aid to lip-reading and environmental sound awareness: Able to score highly on the CUNY sentence task, using lip-reading and sound to interpret spoken sentences (Boothroyd *et al.*, 1985);
- Group C: Awareness of environmental sounds only;
- Group D: No benefit from CI.

Results

The mean VS intracranial tumour diameter was 20 mm (standard deviation 5.3 mm) averaged over the 11 patients who received an implant with tumour *in situ*. The remaining patient received CI immediately after excision of a 30 mm tumour. Five patients had radiotherapy to their VS between 14 and 19 years prior to CI. Two patients had partial tumour resection at another unit prior to joining our care; the remaining four cases had no prior surgical or radiotherapy management to the VS. Age at implant ranged from 27 to 80 years, and duration of profound deafness from six months to more than 20 years (Table 1). Five of the 12 patients had significant pre-existing facial weakness (House Brackmann grades 3–6) on the contralateral side. This was a consideration, as some wished to avoid risk of further facial weakness from radical surgery and therefore regarded cochlear implantation a better option than auditory brainstem implantation. Four of the 12 had promontory stimulation as part of the CI assessment; of these two patients gave definite report of sound; one response was inconclusive and one patient could not report sound. Full insertion of the electrode array was achieved for all 12 cases. Pre-implant MRI identified three cases that had tumour in the cochlear duct. A decision was taken to implant the Nucleus Contour device with the stylet *in situ* rather than advance off stylet, so that the electrode array could be pushed through tumour bulk. No patients complained of additional disturbance to taste, balance or facial function after the CI insertion (Table 2).

Non-Auditory Sensation (NAS) was found in five patients, including facial twitching, eye movements and a sense of dizziness. Reduced dynamic range limited optimal programming. Approaches taken to optimise auditory perception and minimise NAS included:

- widening pulse width;
- changing stimulation rate;
- changing programming strategy and stimulation modes (in the case of the Cochlear device, moving between monopolar and pseudo-monopolar stimulation modes; and changing from the default programming strategy ACE™ to SPEAK™);

Table 1 NF2 case series 2009–2016: pre-implant status

Case	Age at CI; gender (F = Female) (M = Male)	Year CI; device	Size of ipsilateral VS (max diameter) (mm)	Management of ipsilateral VS	Facial function on ipsilateral side (and contralateral) House Brackman score	Promontory stimulation nt = not tested + = positive response	Duration profound deafness implanted ear	Sentence repetition task – no lip-reading (BKB score pre-implant) (%)	Hearing Thresholds pre-implant in dB (HL) at: 250,500,1000,2000, 4000 Hz
1	60; F	2009; Nucleus CI512	13	No treatment and stable tumour	HB 1; (HB1)	+ 2009	Six months	0	90, 95, 105, 105, 110
2	54; F	2010; Nucleus CI512	13	1992 Gamma knife then stable tumour	HB1; (HB4)	+ 2010	13 years	0	110, 120, 120, 120, 110
3	64; M	2011; Medel Concerto Flexsoft	11	1992 Gamma knife then stable tumour	HB 1; (HB1)	Nt	Approx 20 years	0	80, 95, 120, 120, 120,
4	52; F	2012; Nucleus Freedom (contour advance)	24	Active tumour; Bevacizumab commenced after CI	HB1; (HB 6)	Nt	Nine years	0	80, 85, 110, 120, 120
5	32; F	2012; Nucleus Freedom (contour advance)	24	Active tumour that grew post CI to 30 mm; subsequent tumour control on Bevacizumab	HB 1; (HB1)	+ 2011	Two years	Not tested; mother tongue not English	105, 85, 90, 80, 70
6	80; M	2012; Nucleus Freedom (contour advance)	27	Partial tumour resection 2006; Radiotherapy 2011	HB 2; (HB 3)	Nt	Two years	0	120, 120, 120, 120, 120
7	41; F	2012; Nucleus Freedom (contour advance)	23	Stereotactic radiotherapy 1996 then stable tumour	HB1; (HB1)	Tested with inconclusive response	Eight years	0	95, 95, 90, 90, (unreliable at 4kHz)
8	27; M	2012; Nucleus Freedom (contour advance)	20	Initially stable followed by growth after CI; tumour control on Bevacizumab	HB1; (HB2)	Nt	Four years	0	105, > 105, 110, 100, 80
9	64; M	2014 Nucleus Freedom (contour advance)	22	Fractionated stereotactic radiotherapy in 1998 then stable tumour.	HB1; (HB1)	Nt	14 years	36	85, 80, 85, 80, 95

Continued

Table 1 *Continued*

Case	Age at CI; gender (F = Female) (M = Male)	Year CI; device	Size of ipsilateral VS (max diameter) (mm)	Management of ipsilateral VS	Facial function on ipsilateral side (and contralateral) House Brackman score	Promontory stimulation nt = not tested + = positive response	Duration profound deafness implanted ear	Sentence repetition task – no lip-reading (BKB score pre-implant) (%)	Hearing Thresholds pre-implant in dB (HL) at: 250,500,1000,2000, 4000 Hz
10	34; F	2012 Nucleus Freedom (contour advance)	30	Surgical excision of tumour at time of implantation		Nt	Six months	0	Sudden loss of hearing from moderate to total loss, no recordable responses 6 months prior to CI. Not recovered despite steroids.
11	45;F	2015 Nucleus CI512	23	Retrosigmoid debulking 2004 and stable remnant	HB 1, (HB4)	2015 negative	12 years	0	All > 120
12	44; M	2015 Nucleus CI512	21	Gamma knife 2001 then slow growing tumour	HB1; (HB3)	Nt	10 years	0	105, > 120, > 120, > 120, > 110

- altering the gain of the input signal and loudness growth functions (i.e. manipulating the pre- and post-processing parameters);
- deactivating selected electrodes.

Deactivation of selected electrodes was used only after all other options had been exhausted. Apart from two of the strong performers (Cases 2 and 3), the programming options under user control were kept to one setting only, so that there was a consistent sound input, enabling the user to establish their listening confidence.

Outcomes

The CI recipients are presented here as four groups, according to putative mechanism of hearing loss (Tables 3–6). Better outcomes are associated with patients presumed to have predominantly cochlear loss, rather than neural dysfunction.

Group A (Table 3; 4 of 10 CI users) achieved open set speech discrimination. The slowly progressive nature of their hearing loss, together with presence of either subjective or objective hearing response, is strongly suggestive of a cochlear origin to the hearing loss. Thus, the CI has been able to restore hearing to a good level, as the nerve has not been compromised by the VS.

Group B (Table 4; 4 of 10 users) show more modest outcomes, giving sound support for lip-reading and awareness of voice and environmental sounds. Disease severity ratings were higher, with one case rated moderate and three rated severe. Here, we might speculate that the hearing loss is partly cochlear in origin and part neural compromise. We note that one case initially had benefit to lip-reading but his hearing outcome then declined following further tumour growth. Indeed, the drop in his sound perception and reduced speech discrimination were early indicators prompting repeat scanning and revision of his Bevacizumab drug regime.

Group C (Table 5; 2 of 10 users) represents poor outcome with aid to environmental sounds only. These cases are instructive. Although Case 1 had mild tumour load, and positive response to promontory stimulation, her hearing loss had been sudden and total. We might now presume that her hearing loss was more neural in origin. She was subsequently lost to follow up after transferring away. Likewise Case 6 had mild disease severity, but with both prior partial resection and irradiation of his tumour; the use of the implant was limited to eight active electrodes giving sound percept. NAS was a major limiting factor when programming his implant. He still wears the processor full time to support his environmental sound awareness. A pre-operative promontory stimulation test would have been informative in this case.

Table 2 NF2 case series 2009–2016: Surgical and audiological status

Case	Insertion of CI	Facial function on operated side	Balance or taste disturbance?	Electrodes active	Electrodes inactive due to NAS; other factors	Sound field aided thresholds: range across 250–6000 Hz
1	Full insertion	Unchanged	None	21 of 22	None	≥40 dB
2	Full insertion; tumour noted in cochlear duct	Unchanged	None	22 of 22	None	≥30 dB
3	Full insertion	Unchanged	None	12 of 12	None	≥35 dB
4	Full insertion; tumour noted in cochlear duct	Unchanged	None	12 of 22	NAS	≥35 dB
5	Full insertion	Unchanged	None	22 of 22	None	≥35 dB
6	Full insertion; tumour noted in cochlear duct	Unchanged	None	8 of 22	All electrodes cause NAS. 14 inactive.	≥35 dB
7	Full insertion	Unchanged	None	13 of 22	1–6; 20–22	≥35 dB
8	Full insertion	Unchanged	None	19 of 22	1–2; 15.	≥40 dB
9	Full insertion	Unchanged	None	20 of 22	1–2	≥25 dB
10	Full insertion	Unchanged	None	No response to CI	n/a	n/a
11	Full insertion	Unchanged	None	No response to CI	n/a	n/a
12	Full insertion	Unchanged	None	19 of 22	1–3 Out of compliance	≥30

Group D (Table 6; two cases, non-users) received no percept from the implant, confirming their hearing loss as neural in origin. They represented challenging surgical presentations, being cases with moderate or severe ratings for disease severity. Case 10 was implanted after full resection of a large 30 mm VS; she was pre-operatively counselled on the low probability of success. Already an ABI user on the left, with only modest benefit to lip-reading, she consented to CI for the right ear, in the hope she would gain more functional benefit. During VS removal, cochlear nerve function was monitored electrically by performing continuous eABR. Stable Wave eV responses were obtained throughout tumour removal; however, amplitude dropped during mobilisation of a small remnant of tumour from the fundus of the internal

Table 3 Group A – CI giving open set speech discrimination; presumed cochlear dysfunction

Case number	Disease severity rating (mild/ moderate/ severe)	Electrodes active	Aid to lip-reading? Score on sentence level CUNY task	Enjoys music?	Use the phone?	Speech discrimination; BKB score, no lip-reading	Tumour status since implant
2	Mild	22 of 22	Yes; 100%	Yes, able to enjoy familiar music	Yes	82% in quiet; 54% in noise	
3	Mild	12 of 12	Yes; 100%	Yes, enjoys both familiar and new music	Yes	2011: 84% in quiet; 68% in noise. 2015: 100% in quiet, 86% in noise	
5	Moderate	22 of 22	Yes; estimated 89% (non-standardised task via interpreter)	Yes, better with time	Yes	82% estimate (non-standardised task via interpreter), uses phone in native language; needs lip-reading support in English	
9	Mild	20 of 22	Yes, not formally tested	Music not enjoyable from CI; lost access to music when he had radiotherapy	Uses phone in contra ear with residual natural hearing	90% in quiet, CI only. 26% in noise, rising to 80% if bimodal with hearing aid	

Table 4 Group B – CI giving benefit to aid lip-reading; presumed mixed cochlear/neural hearing loss

Case number	Disease severity rating	Electrodes active	Aid to lip-reading?	Enjoys music?	Use the phone?	Speech discrimination; Sentence score, *with lip-reading and sound together*	Tumour status since implant
4	Severe	12 of 22	Yes	No	No	2012: 90% 2016: 96%	2016 Bevacizumab therapy continues; tumour size 27 × 17 mm
7	Moderate	13 of 22	Yes	Yes	No	28%	
8	Severe	19 of 22	Yes – until benefit lost	No	No	58%; dropping by 12 months to 18%.	2016 Bevacizumab therapy continues; tumour size 28 × 20 × 21mm
12	Severe	19 of 22	Yes	Yes, developing	No	At six months: 56%	2016 Bevacizumab continues; tumour size 21 × 17 × 21mm

auditory meatus. Papavarine was topically applied to the cochlear nerve and Wave eV returned but not to its original amplitude. It was decided that cochlear implantation should be performed despite lower Wave eV amplitude. The temporary electrode was removed and cochlear implantation was performed with a Nucleus Freedom (Contour Advance), but subsequent Wave eV responses using the Freedom implant were absent. At switch on there was no sound percept.

Case 11 presented as a potential candidate for ABI after a 12 year history of profound hearing loss following retrosigmoid debulking of VS. The patient was highly reluctant for ABI (due to the higher surgical risk to facial function). As an alternative, cochlear implantation was performed despite a negative promontory stimulation outcome (which led to counselling that a poor/nil sound perception outcome was likely). No sound percept was achieved. Both cases have since been explanted, to allow for easier disease monitoring through MRI.

Discussion

This study supports CI for NF2 patients where the hearing loss is considered to be cochlear in origin; this is often those patients with stable tumours or those tumours that can be controlled with drug treatment.

Our results indicate that the key positive predictive factors in hearing outcome after CI in NF2 are as follows:

- stable VS;
- clear evidence of a functioning cochlear nerve either by recording a subjective auditory response or by auditory response to promontory stimulation;
- slowly progressive onset of hearing loss in the presence of a stable tumour.

The clinical team's view at the time was that promontory stimulation was not always a strong predictor of cochlear nerve function (e.g. Bento *et al*., 2013), so this test was not carried out for all patients. Furthermore, a negative promontory stimulation did not deter the attempt at CI. This position was then later reconsidered in the light of the weaker outcomes for those without positive promontory stimulation response.

We suggest that the response of the cochlear nerve to the electrical stimulation may be accounted for by the mechanism of the hearing loss in each case. The less aggressive tumours often present as gradual hearing loss over time; where the hearing loss perhaps arises from cochlear dysfunction rather than by direct tumour compression of the VIII nerve. It is our observation that patients who present with slowly progressive hearing loss in the presence of an ipsilateral stable tumour tend to achieve a good outcome. In this scenario, the cochlear nerve is preserved sufficiently to allow stimulation through CI. Likewise, patients who have had radiotherapy control of a growing tumour associated with some hearing preservation also seem to perform well after CI.

Table 5 Group C – CI giving only awareness of environmental sounds; loss dominated by neural component

Case number	Disease severity rating [of 3]	Electrodes active	Aid to lip-reading?	Enjoys music?	Use the phone?	Speech discrimination; sentence score, with lip-reading;
1	Mild	21 of 22	No – suspect awareness of voice and environmental sounds only	No	No	Not able to test
6	Mild	8 of 22	Very limited; awareness of voice and environmental sounds only	No	No	Not able to test

Table 6 Group D – CI giving nil outcome; neural loss

Case number	Disease severity rating [of 3]	Electrodes active	Tumour	Surgical comment
10	Moderate	None	30 mm VS – removed prior to CI	CI inserted after positive intraoperative eABR
11	Moderate	None	23 mm; prior debulking (2004) and then CI inserted in 2015.	Normal insertion

CI treatment and Bevacizumab

Bevacizumab is now established as a treatment option to control rapidly growing schwannomas, and can in some cases lead to mild hearing improvement (Plotkin *et al*., 2012), possibly as a result of tumour shrinkage relieving VIII nerve conduction block across the site of tumour compression. We present the first a series of patients who have undergone CI on the side of a VS which is also controlled by the administration of Bevacizumab.

Surgical factors

One major factor limiting outcome seems to be a history of previous surgery even if that surgery was performed with hearing preservation in mind. Two patients underwent previous hearing preservation surgery at other units many years before cochlear implantation. Case 6 derived a limited awareness of sound from the CI while Case 11 did not achieve any sound percept. In these more complex cases, we would now recommend greater emphasis be placed on pre-operative promontory stimulation. We would suggest CI has limited prospect if there is no recordable hearing and no positive promontory stimulation result, especially if there is previous surgical intervention. Planned tumour removal and simultaneous CI should also be considered (Lloyd *et al*., 2014). One of our series underwent cochlear nerve preservation surgery with the help of eABR. The Wave eV was lost during resection of tumour from the fundus and although a waveform of smaller amplitude returned after application of Papavarine, the recipient derived no benefit from the CI. Thus, loss or significant reduction in eABR amplitude must lead to consideration of ABI insertion.

These comments are summarised in Fig. 1, being a proposed counselling and decision-making tool when considering CI/ABI treatment options.

The advantages of CI over ABI are clear: More likelihood of open set speech discrimination outcome, less likelihood of non-auditory stimulation, and lower

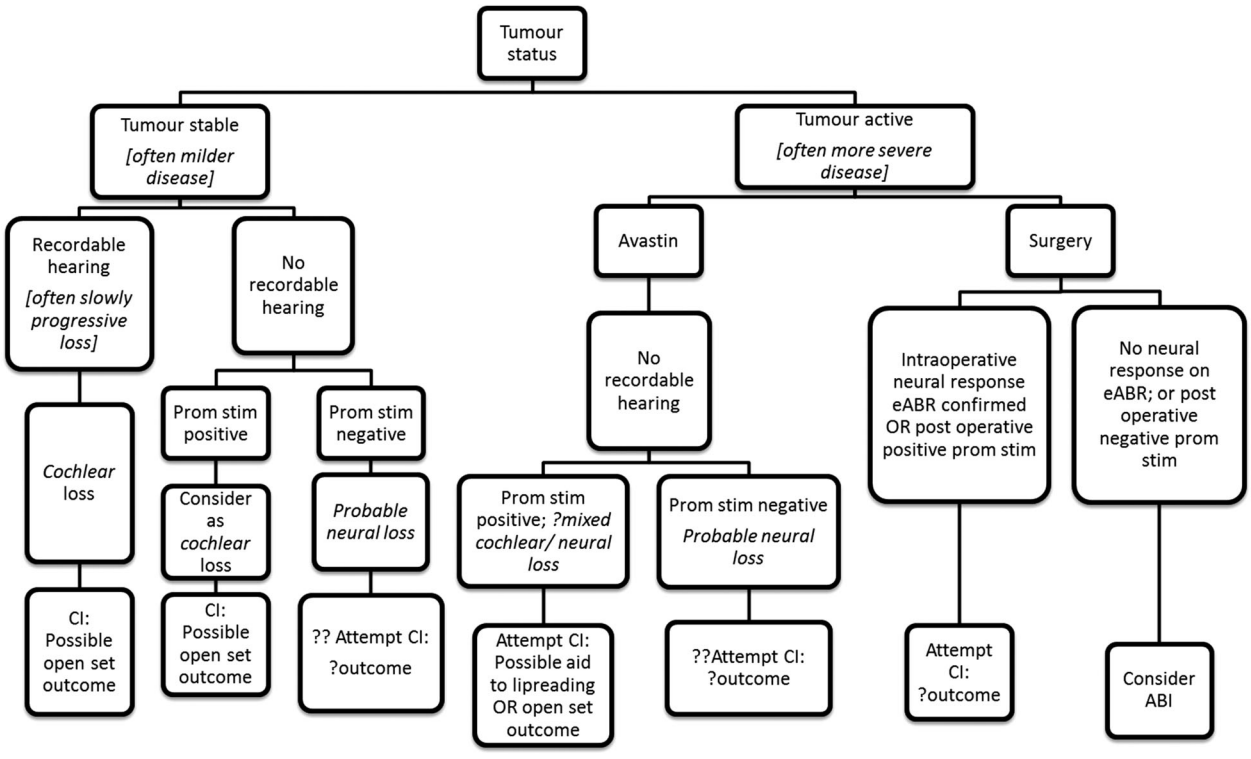

Figure 1 Decision-making/counselling tool for selection of CI or ABI in NF2 cases.

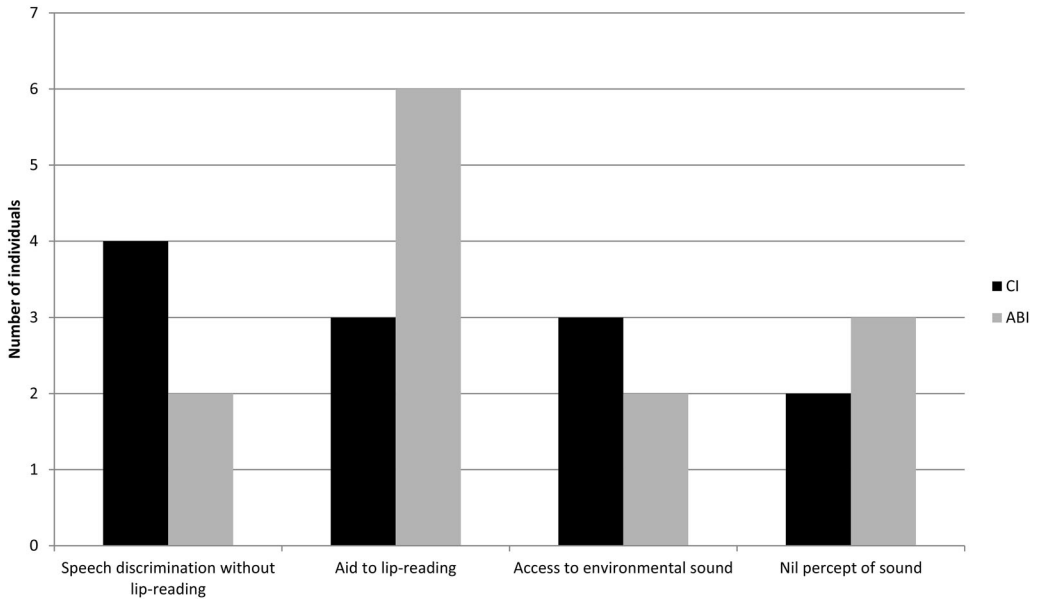

Figure 2 Outcomes of CI versus ABI.

surgical risk. The advantage of CI over ABI for speech outcomes is captured in Fig. 2, using data on ABI patients at our unit, 1999–2016.

Benefit of CI over time

The benefit of CI in NF2 may change over time. The on-going disease progression may lead to progressive cochlear nerve dysfunction; this in turn could lead to a change in sound perception from the CI. Further, VS surgery (if the tumours start to grow) will mean that the implant has to be removed.

MRI and CI

With the development of a safe scanning procedure with the magnet *in situ*, our unit now monitors NF2 disease using MRI scanning without magnet removal (Walton *et al*., 2014).

Conclusion

CIs provide a valuable option for hearing rehabilitation in NF2, even in the presence of an ipsilateral VS. CI combined with ipsilateral VS surgery is likely to lead to poor outcome unless there is evidence of cochlear nerve function either by the presence of some acoustic hearing or a positive promontory stimulation test prior to surgery. Intraoperative eABR monitoring can also give a good indication of whether the cochlear nerve has survived sufficiently intact to make use of a CI.

The encouraging early Bevacizumab outcomes offer the opportunity of inserting a CI in the presence of a growing tumour that is subsequently made stable. A counselling tool is proposed to guide the clinician, making use of objective measures of neural function such as promontory stimulation and eABR. Outcomes from CI appear to be at least as good as those from ABI in giving aid to lip-reading, usually with a better quality of sound. Some NF2 CI users achieve excellent speech discrimination ability with open set speech discrimination, which exceeds that expected from an ABI. Recent developments in surgical technique and the use of Avastin now open up more choices for proactive hearing management and hearing restoration in the NF2 population.

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