



Hearing Restored After Adult-Onset Deafness with an Auditory Brainstem Implant: A Narrative Review of Rehabilitation and Everyday Communication Outcomes

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Abstract

An auditory brainstem implant bypasses the cochlea and the auditory nerve entirely, delivering stimulation to the cochlear nucleus. It is offered to people for whom a cochlear implant cannot work. What it delivers varies enormously, and the variation is not random.

Objectives: *To review the published evidence on auditory brainstem implantation in adults: what auditory outcomes are achieved, how sharply they differ between tumour and non-tumour aetiology, what rehabilitation contributes, and how the results should be described to a candidate before surgery.*

Material and Methods: *Narrative review of case series, systematic reviews, single-centre cohorts and device studies indexed in PubMed and the major otolaryngology and audiology journals. No new patient data were generated; every estimate quoted is one published by the original investigators.*

Results: *In the largest single-centre adult series with at least one year of follow-up, open-set speech perception averaged 59 per cent in 48 non-tumour adults, with a range of 10 to 100 per cent, against an average of 10 per cent in 32 patients with neurofibromatosis type 2, range 5 to 31 per cent ($p = 0.0007$). A systematic review of 36 studies covering 662 tumour and 267 non-tumour patients reported weighted mean speech recognition in tumour patients of 39.2 per cent for closed-set words, 23.4 per cent for open-set words and 21.5 per cent for open-set sentences.*

Conclusion: *For patients with neurofibromatosis type 2 the realistic expectation is environmental sound awareness and substantial enhancement of lip-reading rather than telephone use. For adults deafened by cochlear or nerve pathology with an intact central pathway, open-set speech is achievable in a majority. Conflating the two produces expectations that the device cannot meet.*

Abbreviations: *ABI – Auditory Brainstem Implant, CAP – Categories of Auditory Performance, CI – Cochlear Implant, CUNY – City University of New York sentence test, NF2 – Neurofibromatosis Type 2, SD – Standard Deviation.*

Keywords: *Auditory Brainstem Implant; Neurofibromatosis Type 2; Speech Perception; Aural Rehabilitation; Patient Expectations*

Introduction

Cochlear implantation restores useful hearing to most people with severe to profound sensorineural deafness, but it depends on a functioning auditory nerve [1,2]. Where the nerve is absent, destroyed by bilateral vestibular schwannoma surgery, or where the cochlea cannot be implanted because of ossification or malformation, the implant has nothing to stimulate [3,4]. Where the nerve is preserved at surgery, cochlear implantation may still be possible and is preferred [28,29].

The auditory brainstem implant was developed for that situation, placing an electrode array on the surface of the cochlear nucleus in the lateral recess of the fourth ventricle [5,6]. It was first used in patients with neurofibromatosis type 2, in whom hearing loss follows removal of bilateral tumours, and licensed on that basis [7,8].

Indications subsequently widened to non-tumour adults and children with cochlear or cochlear nerve pathology and an intact central auditory pathway [9,10,11]. That expansion produced the central observation of this literature: the same device, in the same anatomical position, performs very differently depending on why the patient became deaf. This review sets out that difference and what it means for counselling.

Material and Methods

Search strategy: PubMed and the archives of the major otolaryngology, otology and audiology journals were searched for studies of auditory brainstem implantation. Search terms combined auditory brainstem implant with neurofibromatosis type 2, cochlear nerve aplasia, cochlear ossification, speech perception, open-set and rehabilitation. Reference lists of surgical and device reviews [5,12] were screened by hand for primary sources.

Eligibility: Studies were included if they reported auditory or speech perception outcomes in five or more implanted patients, or characterised the surgical technique, complications or rehabilitation associated with the device. Paediatric series are cited where they inform the aetiological comparison and are identified as such.

Data extraction and handling: Aetiology, sample size, duration of follow-up, the speech test used and the reported result were extracted as published. Aetiology and the test format are reported alongside every outcome figure, because the two account for most of the apparent disagreement between series: closed-set and open-set scores are not interchangeable, and tumour and non-tumour populations should not be pooled. No pooling or re-analysis was performed. Figures 1 to 4 and Tables 1 and 2 reproduce published values only. This is a narrative synthesis and was not registered as a systematic review.

Results

The aetiological divide: The largest single-centre experience reported on 112 implanted patients, of whom 80 had reached at least one year of follow-up: 32 with neurofibromatosis type 2 and 48 with non-tumour disease. Open-set speech perception scores ranged from 10 to 100 per cent in the non-tumour adults, with a mean of 59 per cent, against 5 to 31 per cent in the tumour group with a mean of 10 per cent (Figure 1). The difference was statistically significant ($p = 0.0007$) [13,14]. All patients had a functioning implant and no operative or long-term complications were reported in that series.

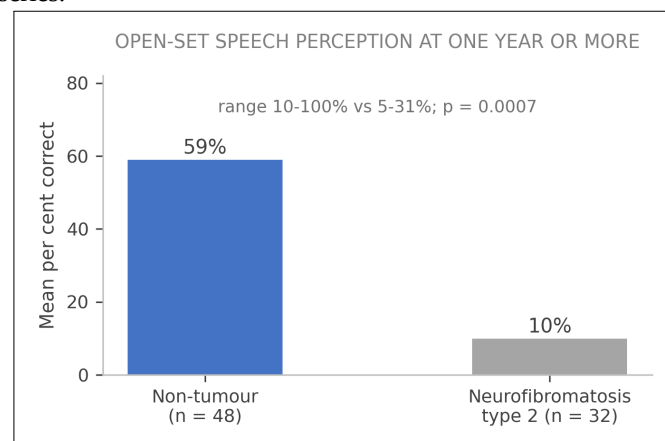


Figure 1: Mean open-set speech perception in non-tumour adults and in patients with neurofibromatosis type 2 [13,14]

What pooled data show: A systematic review of 36 studies extracted data on 662 tumour and 267 non-tumour patients. In tumour patients, weighted mean speech recognition was 39.2 per cent for closed-set words (range 19.6 to 83.3), 23.4 per cent for open-set words (17.2 to 37.5) and 21.5 per cent for open-set sentences (2.7 to 48.4), with a mean Categories of Auditory Performance score of 3.1 (Figure 2) [15]. Non-tumour patients in that review comprised cochlear nerve aplasia (31.3 per cent), cochlear ossification (22.7 per cent), cochlear malformations (14.1 per cent) and cochlear nerve hypoplasia (13.8 per cent), and were predominantly prelingually deafened (83.2 per cent) with a mean age of 15.6 years [15].

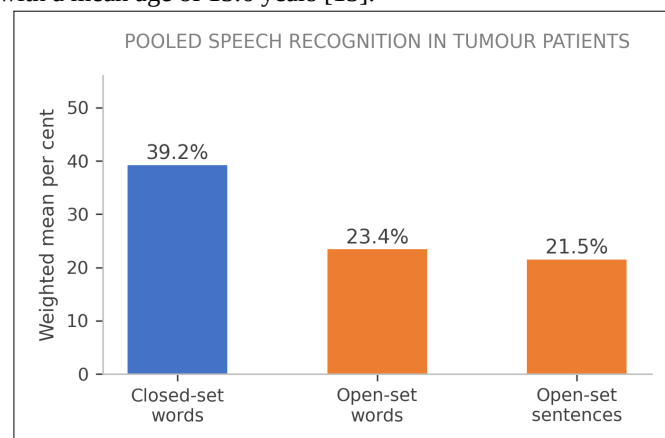


Figure 2: Weighted mean speech recognition in tumour patients across 36 studies [15]

Where the benefit actually lies in tumour patients: A review of 86 implanted patients with neurofibromatosis type 2 found that 13 did not respond to stimulation and one device was explanted. Among the 60 with six-month data, mean sentence scores were under 10 per cent with the implant alone, 30 per cent with vision alone, and close to 55 per cent with implant and vision together (Figure 3) [16]. That combination figure is the one that matters clinically. The implant is not restoring speech understanding; it is roughly doubling what lip-reading alone achieves, and some patients exceed 90 per cent when both are combined.

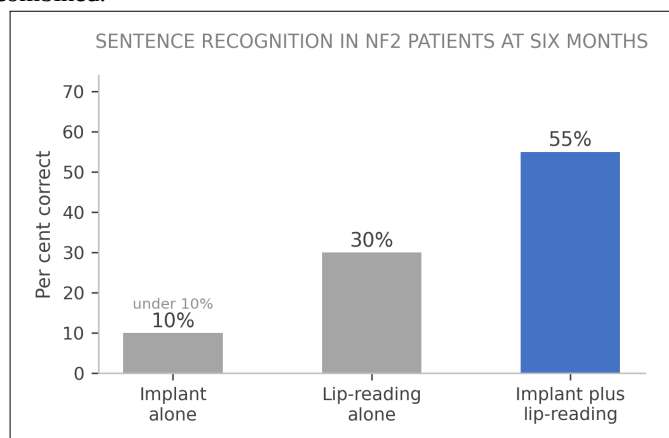


Figure 3: Sentence recognition with implant alone, lip-reading alone, and both combined, in patients with neurofibromatosis type 2 [16]

The size of the evidence base: The pooled review covered 36 studies and 929 patients in total, 662 with tumour and 267 without (Figure 4) [15]. For a device implanted for four decades that is a modest literature, and it is dominated by case series from a small number of high-volume centres [5,9,17]. The paediatric non-tumour literature has grown separately and reaches similar conclusions about aetiology [31,32,33], and the surgical approach has been described in detail [34].

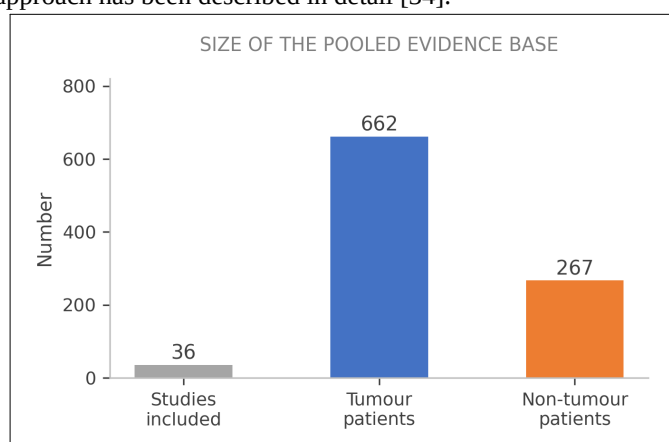


Figure 4: Studies and patients contributing to the pooled analysis of auditory brainstem implant outcomes [15]

Study	Design and cohort	Principal reported finding
Colletti et al. 2009 [13]	Single centre, 80 adults with 1-year follow-up	Open-set 59% non-tumour vs 10% NF2 (p = 0.0007)
Colletti 2006 [14]	80 patients, 26 NF2 and 54 non-tumour	Non-tumour 12-100% (mean 59%); NF2 5-30% (mean 11%)
Garcia et al. 2024 [15]	Systematic review, 36 studies, 929 patients	Tumour: closed-set 39.2%, open-set sentences 21.5%, CAP 3.1
NF2 series of 86 [16]	Tumour patients with 6-month data	Implant alone <10%; with lip-reading ~55%; 13 non-responders
Grayeli et al. 2007 [17]	23 NF2 patients	Good or excellent result in more than half
Sennaroglu et al. 2011 [11]	Non-tumour paediatric and adult series	Established outcomes in cochlear nerve deficiency
Sanna et al. 2012 [18]	Single-centre surgical series	Surgical approach, complications and audiological outcome

Table 1: Principal studies reviewed, with the finding as reported by the original investigators

Why the difference exists: Two explanations are offered and are not mutually exclusive. Tumour growth and its removal damage the cochlear nucleus and adjacent brainstem, distorting the anatomy the array must sit on and injuring the neural population it must stimulate [13,19]. Non-tumour patients have a structurally intact nucleus and an undamaged central pathway. Electrode position and the number of electrodes producing auditory rather than non-auditory sensation differ accordingly, and non-auditory side effects from spread to adjacent structures are a recognised constraint on programming [20,21,35].

Rehabilitation and programming: Fitting an auditory brainstem implant is considerably more complex than fitting a cochlear implant. Electrodes must be tested individually for pitch and for non-auditory effects, and the map is built from those that produce useful auditory sensation [21,22]. Structured aural rehabilitation, including lip-reading training, is integral rather than optional given where the benefit lies for tumour patients [23,24]. Device use over time is variable: in one long-term series, some recipients became regular users while others reverted to sign language [25], and updated single-institution experience reports the same pattern [30].

What patients report: Outcome measurement in this field is dominated by speech testing in quiet, which is not the environment in which the device is used. The smaller literature on patient-reported outcomes emphasises environmental sound awareness, safety, and reconnection to a spoken-language world for people who had always been hearing [16,26,27]. For an adult deafened in mid-life, that reconnection may matter more than a percentage on a sentence test, and it is poorly captured by the measures the literature reports.

Aetiology	Realistic expectation	Evidence
Neurofibromatosis type 2	Sound awareness; lip-reading enhanced	Mean open-set 10%; combined with vision ~55% [13,16]
Cochlear nerve aplasia	Variable; open-set achievable in some	Largest non-tumour subgroup in pooled data [15]
Cochlear ossification	Often good open-set speech	Intact nerve and central pathway [14,15]
Head trauma	Among the better non-tumour outcomes	Reported in single-centre series [13]
Auditory neuropathy	Poorer than other non-tumour causes	Central pathway may also be affected [13]
Any aetiology	13 of 86 NF2 patients had no response at all	Non-response must be discussed beforehand [16]

Table 2: Expected auditory outcome by aetiology and the evidence supporting each

Discussion

The single most important thing this literature establishes is that auditory brainstem implant outcomes cannot be quoted as a single figure. A mean of 59 per cent open-set speech and a mean of 10 per cent describe the same operation with the same device, and the difference between them is why the patient became deaf [13,14]. Any consent conversation that uses an undifferentiated figure is misleading in one direction or the other.

For the patient with neurofibromatosis type 2, the honest description is not that the implant will restore speech understanding. It is that it will probably provide awareness of environmental sound and will approximately double what lip-reading alone achieves, taking sentence recognition from around 30 to around 55 per cent when the two are combined [16]. Framed that way, the benefit is real and worth having, particularly for someone who has spent their life in a hearing world. Framed as hearing restoration, it invites disappointment.

There is also a figure that belongs in every such conversation and is easy to leave out. In the series of 86 tumour patients, 13 obtained no auditory response at all, and one device was explanted [16]. That is roughly one in seven undergoing brainstem surgery and receiving nothing. It should be stated before surgery, not discovered at activation.

The non-tumour results are genuinely impressive and warrant caution of a different kind. A mean of 59 per cent conceals a range from 10 to 100 per cent [13]. Some adults achieve results comparable with good cochlear implant users; others gain very little. The predictors are incompletely worked out, and aetiology within the non-tumour group matters, with auditory neuropathy performing worse than cochlear ossification or head trauma [13,15]. Prediction at the individual level remains poor, which is the honest position rather than a failure of this review.

Study limitations

The limitations belong to the underlying literature. There are no randomised trials, and there will not be, since the alternative

to implantation is no hearing at all. Evidence is dominated by case series from a small number of high-volume centres, with the attendant risk that results reflect surgical and programming expertise not generalisable elsewhere [5,13,17]. Speech tests differ between series and between languages, and closed-set and open-set results are not comparable [15]. Follow-up duration varies widely and outcomes change with experience of the device [25]. The pooled review mixed adults and children and predominantly prelingually deafened patients, which limits its application to an adult deafened in mid-life [15]. Non-auditory side effects and device non-use are inconsistently reported. And patient-reported outcomes, which are arguably what matters most here, are largely absent [26,27].

Conclusion

An auditory brainstem implant does different things depending on why the patient became deaf, and the gap is wide enough that the two situations should be discussed as separate operations. For an adult deafened by cochlear or cochlear nerve pathology with an intact brainstem, open-set speech understanding is a realistic hope and around half achieve substantial results. For a patient deafened by neurofibromatosis type 2, the device is a powerful aid to lip-reading and a reconnection to environmental sound, roughly one in seven gains nothing, and telephone conversation is not a reasonable expectation. Both of those are worth having. Only one of them is hearing restoration, and saying so before surgery is part of doing the operation properly.

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Conflict of Interest

The authors declare no competing interests.

Ethics Statement

This article reviews previously published studies only. No new patient data were collected and no participants were recruited, so ethical approval was not required.

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