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Review

Cochlear Implantation After Radiotherapy of Vestibular Schwannomas

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BACKGROUND AND OBJECTIVES: Vestibular schwannomas (VS) frequently lead to ipsilateral sensorineural hearing loss (HL) as part of its natural history or as a result of treatment. Cochlear implantation represents a well-documented treatment of profound HL that cannot be treated adequately with a conventional hearing aid, thus being offered to selected VS patients. A functional cochlea and cochlear nerve are prerequisites for sound perception with a cochlear implant (CI). The potential impact of radiotherapy on these structures is thus an important issue for subsequent CI hearing outcomes. The objective of this article is to present a case and to review the existing literature on the outcomes of cochlear implantation in irradiated VS patients systematically.

METHODS: A systematic literature review using preferred reporting items for systematic reviews and meta-analyses was conducted. Medline was searched systematically. Papers reporting ipsilateral CI outcomes after radiotherapy of VS were included. Additionally, results of CI after stereotactic radiotherapy in a 54-year-old male with neurofibromatosis type 2 are presented.

RESULTS: A total of 14 papers (33 patients) fulfilled inclusion criteria. Moderate preoperative HL was found in 11 patients. Six had moderate to severe HL, whereas 16 had severe HL or total deafness. Postoperative hearing outcomes varied from poor in 27% of patients to excellent in 19%, with remaining cases lying in between (mean follow-up of 19 months). Most patients achieved improvement in hearing and quality of life.

CONCLUSION: Despite variation in the degree of hearing outcome, CI after radiotherapy of VS appears to be effective in the majority of cases, as more than 70% of patients have good or excellent outcomes within 1-2 years post-implantation. Subjective benefits are considerable, even in cases with relatively poor objective outcome.

KEYWORDS: Acoustic neuroma, hearing rehabilitation, surgery, radiosurgery, auditory implant, hearing outcomes

INTRODUCTION

Vestibular schwannomas (VS) are benign intracranial tumors that develop from the Schwann cells of the vestibulo-cochlear nerve, accounting for roughly 10% of intracranial lesions. The incidence is increasing.[1,2] Common audio-vestibular symptoms include sensorineural hearing loss (HL), tinnitus, and disequilibrium, leading to reduced quality of life. VS occur most commonly in the sporadic, unilateral form, but approximately 5% are bilateral and associated with neurofibromatosis type 2 (NF2). NF2 is an autosomal dominant syndrome involving loss-of-function mutations of the NF2 tumor suppressor gene on chromosome 22q12.[3] As bilateral deafness is a threat especially for NF2 patients, any method to preserve or improve hearing is clinically relevant.

Current management of VS includes a conservative "wait-and-scan" observation regime, microsurgical resection, and irradiation.[4] Choice of management modality depends on patient preferences and characteristics (e.g., tumor size, tumor growth, cystic components, hearing, co-morbidity, etc.). Irradiation modalities include stereotactic radiosurgery (SRS)/radiotherapy, fractionated stereotactic radiotherapy (FSRT), and gamma knife surgery (GKS)/radiosurgery.[5] SRS targets the tumor with a single high dose of radiation originating from multiple sources, whereas FSRT involves multiple treatments with smaller individual doses as compared to SRS in order to reduce side effects. GKS utilizes precise gamma-ray radiation of the tumor as a single-shot treatment.

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Carlson et al.[6] reported that HL after radiotherapy in VS patients gradually progresses and at 10 years post-radiation, only 23% of VS patients have preserved serviceable hearing.[6] Since the combination of tumor on the vestibulo-cochlear nerve and radiotherapy leads to additive increase of the risk and occurrence of ipsilateral profound HL, cochlear implantation for the restoration of hearing is an obvious treatment option for some of these patients. However, the tumor and/or associated treatment may lead to damage of the cochlea and/or the cochlear nerve, thus compromising the transmission of electrical impulses from an implant in the cochlea to the brainstem. Reduced electrophysiological functionality of the cochlea and/or the cochlear nerve may thus hamper the hearing outcome after a cochlear implant (CI). The aim of this study was therefore to systematically review the literature for hearing outcomes after radiotherapy of VS and subsequent cochlear implantation and to present the objective and subjective outcomes of cochlear implantation in a radiated NF2 patient.

METHODS

The systematic literature review was conducted by adhering to the preferred reporting items for systematic reviews and meta-analyses (PRISMA) statement.[7] The systematic search for available literature was performed on June 5, 2019, using the Medline (PubMed) database and the following search string: "*(radiation OR radiation therapy OR radiosurgery OR stereotactic radiosurgery OR radiotherapy OR gamma knife surgery OR gamma knife radiosurgery OR gamma knife radiation) AND (vestibular schwannoma OR acoustic neuroma) AND (cochlear implantation OR cochlear implant)*."

The search resulted in a total of 39 hits that were imported into the Covidence software.[8] There was 1 duplicate. Title and abstract screening resulted in the exclusion of 24 titles. The remaining 14 full-text articles were assessed for eligibility, and 2 full-text articles were excluded, allowing 12 studies for qualitative analysis. These 12 studies represented a total of 69 patient cases, of which 44 cases were excluded due to irrelevancy to this study, leaving 25 relevant patient cases to be evaluated. Two additional papers identified through the retrieved 12 papers were also included. These 2 papers represented a total of 8 relevant cases. Thus, in total, 14 papers representing 33 patients were included. The systematic search process is illustrated in Figure 1.

Paper Inclusion Criteria

All studies (including case reports) on ipsilateral cochlear implantation outcomes after radiotherapy (all types) of VS (both sporadic and NF2) were included.

Paper Exclusion Criteria

Veterinary studies, review articles without original data, editorials, letters to the editor, conference abstracts, non-English articles, and studies of non-CI hearing rehabilitated individuals were excluded.

Data Extraction

The following patient data were registered: age, sex, sporadic or NF2 tumor, tumor side, tumor location, tumor size, information on radiation (type and number of fractions), CI side, additional relevant treatments, cochlear nerve testing procedures, preoperative hearing

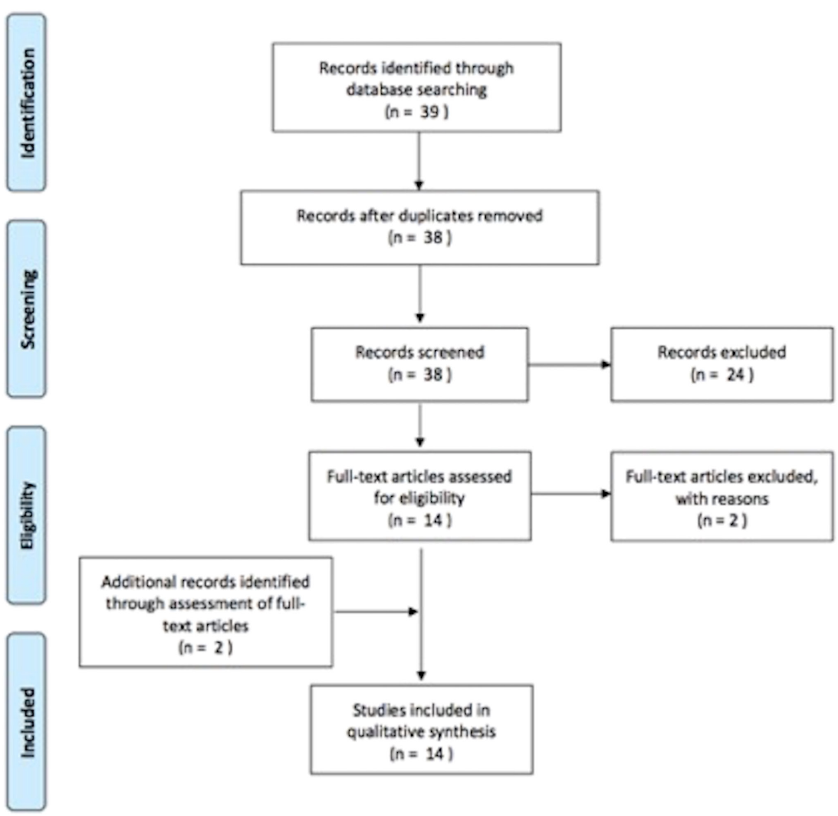

Figure 1. Preferred reporting items for systematic reviews and meta-analyses flowchart illustrating the systematic article selection process.

acuity, postoperative hearing outcomes including time at follow-up, manufacturer of implanted CI, and complications.

Bias

Most studies included were retrospective case reports or series, inherently representing a low level of evidence. In addition, the methods of reporting preoperative and postoperative hearing outcomes were heterogeneous. Uncertainties in postoperative hearing outcomes exist in cases where patients were unable to partake in follow-up hearing tests and thus only reported subjective effects without the use of questionnaires. Lastly, there may be reporting and publication bias involving the underreporting of unsuccessful patient treatments and thereby overestimation of the successfulness of the treatment.

Case Report

A male with NF2 and associated bilateral VS displayed progressive bilateral sensorineural HL. The patient had been a user of bilateral conventional hearing aids (HA) since the beginning of adulthood, and in 2014, at 49 years, the patient was fitted with a left-sided bone-anchored hearing system, which later became ineffective. He was however still able to benefit from a conventional HA on the left side. An MRI scan in early 2016 showed significant right-sided tumor growth. To stabilize the tumor, the patient was treated with FSRT (total 50 Gray over 30 fractions) in 2016, at 51 years. Prior to FSRT, the largest extrameatal tumor diameter was around 11 mm (Figure 2). After FSRT, hearing deteriorated further on the right side, and the patient was evaluated for cochlear implantation.

Preoperative Objective Hearing

The patient demonstrated a pure tone average (average of pure tone hearing thresholds at frequencies 0.5, 1, 2, and 4 kHz; PTA4) of 77.5 dB/80 dB on the right/left ear, respectively. Auditory steady-state response thresholds were in agreement with the psychoacoustic audiometry. Auditory brainstem response (ABR) test generated no identifiable potentials at maximum stimulation (100 dB). Monosyllabic speech discrimination score (DS) was 21%/34% with optimal amplification (conventional HA). Sentence test (Dantale) demonstrated a performance of 65% with lipreading in quiet, 59% without lipreading in quiet, and 14% without lipreading in noise. Without HA, the patient scored 6% in quiet and 0% in noise.

Preoperative Subjective Hearing

The patient's Nijmegen Cochlear Implant Questionnaire (NCIQ) total score was 259 with individual domain scores of 117 (Physical), 48 (Psychological), and 94 (Social). The Speech, Spatial, and Qualities of Hearing (SSQ12) total score was 9.1, with subunit scores being 3.4 (Speech), 2.0 (Spatial), and 3.8 (Quality). Their Tinnitus Handicap Inventory (THI) total score was 24, with subunit scores being 16 (Functional), 0 (Emotional), and 8 (Catastrophic thinking).

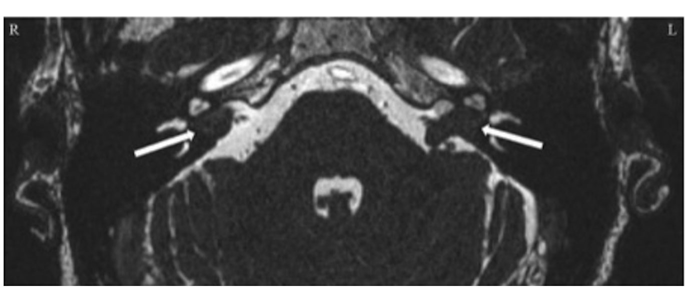

Figure 2. Posterior fossa magnetic resonance imaging demonstrating bilateral vestibular schwannomas (arrows) in a 54-year-old male with NF2. The patient underwent radiotherapy on the right side to stabilize the tumor and received subsequent ipsilateral cochlear implantation due to profound bilateral hearing loss. NF2, neurofibromatosis type 2; L, left; R, right.

Cochlear Implantation

The patient was elected for CI surgery and was implanted on his right ear with an Advanced Bionics (HiRes Ultra 3D Midscale) CI device at age 54. Intraoperative measurements of impedance were within normal range, and stapedial reflexes and compound action potentials could be elicited by stimulation.

Postoperative Objective Hearing

At the 6-month postoperative follow-up, the HL had progressed further on the left side, rendering HA use on this side redundant. The speech DS with the CI (right side) only was 88% with lipreading in quiet, 81% without lipreading in quiet, and 38% without lipreading in noise, indicating significant hearing improvement. The pre- and postoperative speech DS are shown in Figure 3.

Postoperative Subjective Hearing

The patient experienced significant benefit from the CI despite challenges in loud noise and abnormal perception of music. Tinnitus decreased immediately after implantation, and at 6-month follow-up, tinnitus was a problem only when the CI was deactivated. The patient is a daily user of his CI. Pre- and postoperative subjective hearing outcomes are illustrated in Figure 4 (NCIQ) and Figure 5 (THI).

SYSTEMATIC REVIEW

Results

Literature

The selection process led to inclusion of 14 studies published from 2006 to 2018. All included studies were retrospective. The 14 studies

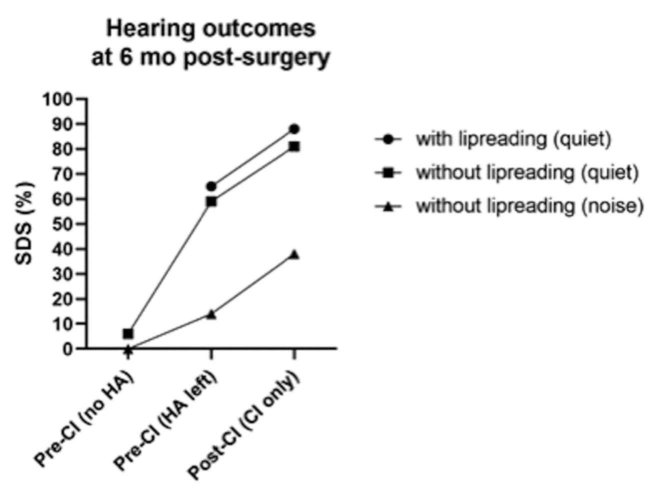

Figure 3. Objective hearing outcomes for the case report. Pre- and postoperative SDS at 6 months post-implantation for the case reported, a 54-year-old male with NF2, implanted on the right side following radiation of the VS on the same side. The patient improved in all 3 conditions (with lipreading in quiet, without lipreading in quiet, and without lipreading in noise). CI, cochlear implant; HA, hearing aid; NF2, neurofibromatosis type 2; SDS, speech discrimination score; VS, vestibular schwannomas.

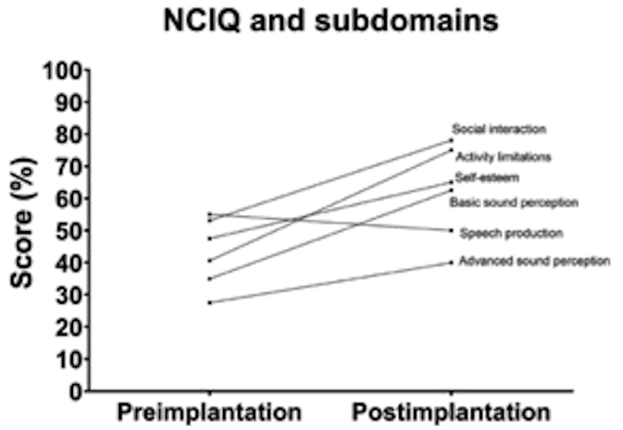

Figure 4. Subjective outcomes for the case report. Nijmegen Cochlear Implant Questionnaire with pre- and postoperative subdomain scores. Subjective improvements occurred in 5 subdomains, but deterioration in speech production was observed.

reported on a total of 33 patients undergoing cochlear implantation after VS radiotherapy.

Patient and Tumor Characteristics

Of the 33 patients included, 17 were female (52%), 15 were male (45%), and in 1 case the sex was not reported. A total of 32 cases (97%) had NF2, while 1 patient (3%) had a sporadic VS.

The mean patient age was 47.7 years (range 18-84 years). The 33 patients received a total of 34 CIs. The tumor locations were intrameatal (n = 6), extrameatal/cerebellopontine angle (n = 13), and not reported (n = 15).

The tumor size was reported for all but 5 tumors.[9-12] According to the Tokyo criteria, sizes of purely intrameatal tumors (n = 6) were tabulated as 0 mm.[13] For the reported sizes of extrameatal tumors (extrameatal or location not reported, n = 23), there was one Grade 1 tumor

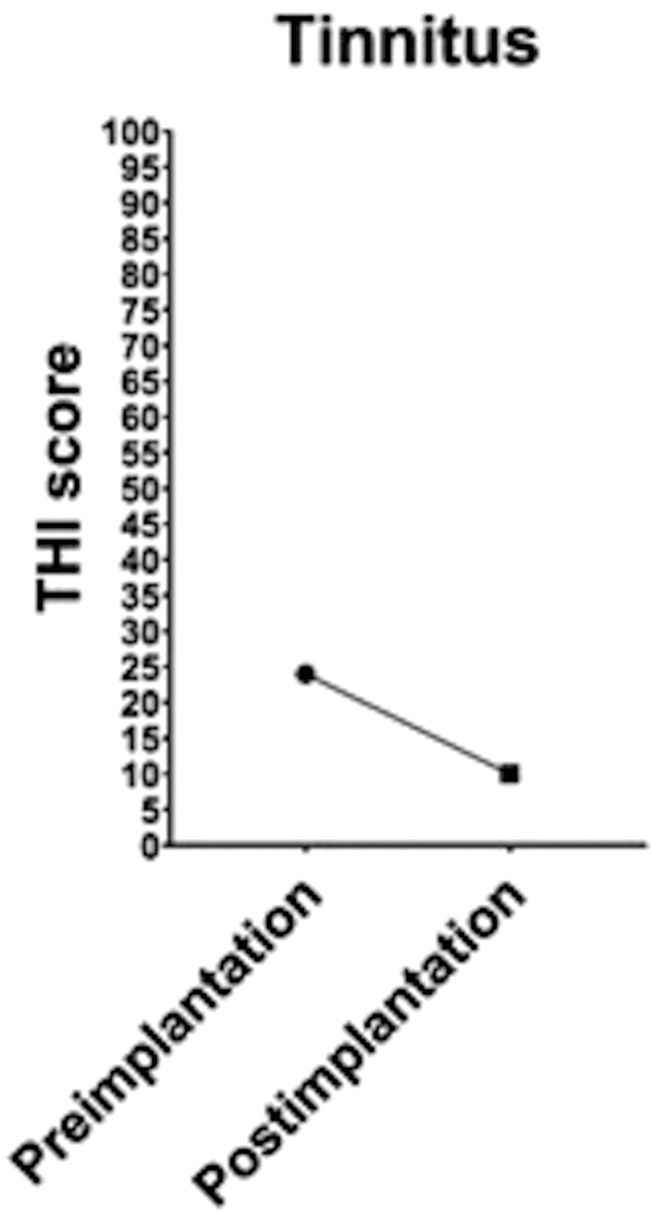

Figure 5. Subjective outcomes for the case report. Pre- and postoperative Tinnitus Handicap Inventory scores showing relief of tinnitus.

(1-10 mm),[14] ten Grade 2 tumors (11-20 mm),[15-19] nine Grade 3 tumors (21-30 mm),[12,16,18-21] and three Grade 4 tumors (31-40 mm).[14,16] The mean size of the extrameatal tumors was 20.5 mm [1-37 mm].

Contralateral Tumor and Other Interventions
Twelve patients had previously undergone surgical resection of the contralateral VS,[10-12,14,16,20,22] of which one patient had received a contralateral auditory brainstem implantation (ABI).[20] Two patients had received GKS,[17] and 1 patient FSRT on the contralateral VS.[23] One patient had received a partial resection of the ipsilateral VS prior to irradiation and CI.[18]

Radiation
The type of radiation used varied across included cases. Nine tumors received SRS,[9,14,15,18] 10 tumors received GKS,[11,17-22] and 6 tumors received FSRT.[12,18,19,23] In 9 cases, type of radiation was unspecified or not mentioned.[10,16,18] Four studies reported radiation dosage. Reported doses were mean 12.5 Gray,[14,20] to the 50% isodose line[14]; mean marginal dose 16 Gray [range 13-20] and mean maximum dose of 32 Gray [range 26-40][15]; mean dose 38.8 Gray [range 12-54] over a mean of 20 fractions [range 1-30], to a mean isodose line of 89% [range 87-90%].[12]

Preoperative Hearing Acuity
Hearing status was described heterogeneously. Moderate HL was described for 11 patients.[9,11,12,14,15,21-23] Six patients included had moderate to severe HL. Of these, 2 patients scored 0% for Bamford–Kowal–Bench (BKB) test without lip reading, but 8% and 20%, respectively, with lip reading.[16]

Two patients scored a mean of 2% for City University of New York (CUNY) Sentences Test in quiet and 19% for consonant nucleus consonant (CNC) word phonemes.[12] Another patient scored 2% and 16% on CNC words and CNC phonemes, respectively, as well as 3% for Arizona Biomedical Institute sentence list (AzBio) and 2% for word recognition score (WRS).[14] One patient scored WRS 0% and 16% unaided, and AzBio 0% and 2% for the left and right ears, respectively.[14]

Severe HL or total deafness (0% for various tests, e.g., CUNY, CNC, Hearing in Noise Test (HINT)) was described for 16 patients,[10,11,16-18,21,22] of which 1 patient had bilateral dead ears but scored 46% on BKB with lip reading.[16]

Cochlear Nerve Testing
Cochlear nerve testing was carried out in most studies (Table 1), in the form of either electrical promontory stimulation (n=9),[11,12,17-19,21] or electrically evoked compound action potential (n=4).[15] One study performed electrical auditory brainstem response.[9] Some studies described cochlear nerve responses, anatomical conditions, and impedances qualitatively.[10,16] Method of cochlear nerve testing was either not reported or reported to have not been carried out in 20 patients.

CI Model and CI Procedure
The CI model was reported in all but 8 patient cases across 4 studies.[10,14,19,23] The Cochlear Nucleus Freedom was the most encountered model used in 7 patients across 4 studies[12,16,18,22] (Table 1). Cochlear Nucleus 512 was used in 4 patients,[15,18] and Nurotron CS-10A was used in 2 cases.[17] Three cases were described as receiving "Cochlear Nucleus."[16] Other models included are found in Table 1.

One study reported the surgical procedure. Simultaneous bilateral cochlear implantation was reported in 1 study.[14]

Follow-up
Time of post-CI surgery or post-activation follow-up was reported in all but 7 patients.[11,12,18] Of the heterogeneously reported patients, 1 had a follow-up of 6 weeks post-CI activation,[9] and 2 patients had a mean follow-up of 20 months[17] (Table 2). Another 2 patients had a mean latest follow-up of 6 months post-cochlear implantation [range 3-9 months].[19] For all other patients, the mean follow-up was 18.5 months [range 6-56 months].

Postoperative Hearing
All postoperative hearing outcomes are summarized in Table 2. The hearing outcomes for monosyllables, phonemes, and sentences scores are plotted in Figure 6.

Hearing Outcomes by Test Applied
In order to provide an overview, the objective hearing results in aided condition of all included studies are grouped below according to the test(s) applied (Table 2). Most studies applied several tests.

Of the 14 studies included, 8 also reported subjective outcomes, for 21 of the 33 patients included in the review.[10,12,15,16-19,22] Twenty of the 21 patients reported subjective benefit, ranging from improved awareness of environmental sounds to telephone use and enjoyment of music (Table 2).

Speech/Word/Phoneme Recognition Tests
Hearing in Noise Test
Six of the 33 patients included in the review had the HINT test performed. Scores ranged from 34 to 98%. Mean score was 71%, and the median was 58/94% (Table 2).[9,10,14,15,21]

Bamford–Kowal–Bench Test
The BKB was most commonly reported by testing in noise, without lipreading. Eight of the 33 patients had this test performed. Scores ranged from 0 to 86% (mean 35% and median 26/54%) (Table 2).[16,18,19]

BKB in quiet without lipreading was reported for 4 patients, scoring a mean of 89% and a median of 82/90%.[16,18]

BKB in noise with lipreading was reported for 2 patients, scoring 36% and 68% (mean 52%).[16]

Consonant Nucleus Consonant Test
Six of the 33 patients had the CNC words test performed. Scores ranged from 28% to 86%. Mean score was 51%, and the median was 46/52% (Table 2).[9,14,15,21]

Table 1. Studies Included in the Systematic Review, Displaying Patient, Tumor, and Treatment Characteristics

Author	No. Patients Included	Mean Age	Sex	Type	Mean Tumor Size	Mean Tumor Grade	Location	Radiotherapy Type	CN Test	CI Device
Amoodi, 2012[9]	1	26	F = 1	NF2	NR	NR	NR	SRS	EABR (0)*	Med-El Sonata
Carlson, 2012[15]	4	48 [37-61]	F = 1 M = 3	NF2	10 [0-14]	2	IAC = 1 NR = 3	SRS	ECAP	Cochlear Nucleus 512, RE, and RCA
Costello, 2016[22]	1	57	F = 1	NF2	8	1	IAC = 1	GKS	NR	Cochlear Nucleus Freedom
Harris, 2017[18]	6	57.8 [41-80]	F = 2 M = 4	NF2	19.5 [11-27]	2.7	NR	GKS = 3; FSRT =2; NR =1	EPS	Cochlear Nucleus Freedom CI512, Medel Concerto Flexsoft
Lustig, 2006[10]	2	46 [41-50]	F =1 M = 1	NF2	NR	NR	CPA = 1; NR = 1	NR	*	NR
Mukherjee, 2013[16]	6	39 [18-72]	F = 5 M = 1	NF2	18 [0-37]	2.6	IAC = 1; NR = 5	NR	*	Cochlear Nucleus 5 Freedom, series 3
Pai, 2013[19]	2	59.2 [42.1-76.3]	M = 1 NR = 1	NF2 = 1; Sporadic VS = 1	21.5 [20-23]	2.5 [2-3]	NR	GKS = 1; FSRT = 1	EPS	NR
Peng, 2018[20]	1	25	F = 1	NF2	30	3	NR	GKS	NR	Cochlear (Multichannel and device)
Pimentel, 2015[23]	1	50	M = 1	NF2	10	1	IAC	FSRT	NR	NR
Pisa, 2017[14]	2	58 [38-77]	F =1 M = 1	NF2	21.8 [1-32.8]	3	NR	GKS	NR	NR
Roehm, 2011[21]	1	60	F = 1	NF2	25	3	NR	GKS	EPS	Cochlear Nucleus 24 RCA
Tan, 2018[17]	2	28 [20-36]	F = 2	NF2	8 [0-16]	2	IAC = 1; NR = 1	GKS	EPS	Nurotron CS-10A
Tran Ba Huy, 2009[11]	1	26	M = 1	NF2	NR	NR	NR	GKS	EPS	Cochlear Esprit 3G processor
Trotter, 2010[12]	3	59 [41-84]	F = 1 M = 2	NF2	30 (NR = 2)	3	IAC = 1; NR = 2	FSRT	EPS	Cochlear Nucleus Freedom

Square brackets represent range. Mean tumor size/grade according to Kanzaki et al.[13] Grade 1: 1-10 mm, Grade 2: 11-20 mm, Grade 3: 21-30 mm, Grade 4: 31-40 mm.
CI, cochlear implant; CNC, consonant nucleus consonant; CPA, cerebellopontine angle; EABR, electrical auditory brainstem response; ECAP, electrically evoked compound action potential; EPS, electrical promontory stimulation; F, female; FSRT, fractionated stereotactic radiotherapy; GKS, gamma knife surgery; IAC, internal auditory canal; M, male; N, number relevant for this review; NF2, Neurofibromatosis Type 2; NR, not reported; SRS, stereotactic radiosurgery. *Studies reporting cochlear nerve testing results, but not method used.

In the CNC phoneme test, scores ranged from 37% to 93% for 7 patients, with a mean of 66% and a median of 67%.[12,14,15,21]

City University of New York Sentences Test

CUNY in quiet was reported for 6 patients, scoring from 36% to 100%, with a mean of 78% and a median of 72%/96% (Table 2).[12,18,19,22]

Other Speech/Phoneme Recognition Tests

The WRS was reported by 2 studies, for 4 patients.[14,17] Scores ranged from 0% to 60% for monosyllabic words (mean 24% and median 8%/28%), and from 5% to 78% for disyllabic words (n = 2; mean 42%).

Five publications included other tests more rarely applied in this context (Table 2).[10,11,18,20,23]

Pure Tone Thresholds

PTAs were reported by 4 studies, including 8 patients.[10,14,15,17] Thresholds ranged from 18 to 80 dB, with a mean of 35 dB and a median of 25 dB.

Complications

Complications were only scarcely reported. A prominent side effect of radiotherapy was subsequent HL and/or deafness.[10-12,15] Transient imbalance after radiotherapy was reported in 1 patient.[14] Two patients experienced progressive tumor growth after radiotherapy.[15,16] One of these patients had to undergo tumor resection and CI explantation and subsequently received an ABI. In 1 case, surgery led to bilateral cochlear fibrosis.[14]

Table 2. Postoperative Hearing Outcomes (Last Available) for the Cases Included in the Review

Study/Author	Patient Case in Text	Follow-Up Time	Postoperative Hearing Outcome
Amoodi et al.[9]	2	6 weeks	HINT: 94%, CNC: 52%
Carlson et al.[15]	6	22 months	CNC words: 46%; CNC phonemes: 67%; CUNY: 100%; HINT: 95%. OSP achieved. CI PTA/SRT: 18. Subjective benefit.
	7	56 months	CNC words: 86%; CNC phonemes: 93%; AzBio: 95%; BKB-SIN: 9.75 dB. OSP achieved. CI PTA/SRT: 20. Daily user.
	8	25 months	PTA/SRT: 19. Immediate subjective benefit but performance decline and no sound perception 1 year after CI.
	9	12 months	No sound perception despite high stimulation levels. Not daily user.
Costello et al.[22]	1	12 months	CUNY: 36%. Subjective improvement.
Harris et al.[18]	2	N/A	CUNY with LR: 100%. BKB in quiet without LR: 82%. BKB in noise without LR: 54%. Enjoys music; uses phone.
	3	N/A	CUNY with LR: 100%. BKB in quiet without LR: 2011: 84%, 2015: 100%; BKB in noise without LR: 2011: 68%, 2015: 86%. Music and phone use.
	6	N/A	Awareness of voice and environmental sounds only.
	7	N/A	With LR, sentence score 28%. Subjective benefit.
	9	N/A	BKB without LR: 90% (in quiet) and 26% (in noise)
	12	6 months	Speech discrimination: sentence score with LR: 56%. Subjective benefit.
Lustig et al.[10]	4	17 months	MTS recognition: 46%. 0 for all other tests. PTA 55 dB. Improved environmental sound awareness; uses LR
	6	18 months	SDS[17]: 46%. HINT: 98%. PTA 35 dB.
Mukherjee et al.[16]	1	12 months	Blind patient: no OSP without LR. Subjective benefit.
	2	12 months	BKB in noise without LR: 54%; BKB in quiet without LR: 82%. BKB live voice with LR: 100%.
	3	3 years	Environmental sound only at 3 years.
	4	6 months	BKB without LR: 0%; BKB with LR: 68%.
	5	6 months	BKB without LR: 0%; BKB with LR: 36%.
	6	12 months	Only environmental sound awareness present. Subjective benefit.
Pai et al.[19]	4	3 months	BKB: 63%, CUNY with LR: 94%. Daily user.
	7	9 months	BKB 13% (1 week), 0% (9 mo), CUNY with LR: 22% (1 week), 61% (9 mo). Phone user. Daily user.
Peng et al.[20]	5	3 years	NU-CHIPS: 100%; Iowa Consonants: 78%; Iowa Vowels: 96%
Pimentel et al.[23]	1	12 months	100% discrimination for Ling's Six Sound Test, vowels, monosyllables, and sentences in open context.
Pisa et al.[14]	1	12 months	PTA 18/20 dB. WRS in quiet: 8%. CNC phonemes: 37%, CNC words: 24%. HINT: 45%. Right PTA: 25, Left PTA: 25.
	2	12 months	WRS in quiet: 28%, CNC phonemes: 55%, CNC words: 28%, HINT: 34%. PTA: 22
Roehm et al.[21]	4	36 months	CNC words: 68%, CNC phonemes: 87%. CUNY in quiet: 92%, CUNY in noise: 90%, HINT in quiet: 58%.
Tan et al.[17]	4	Mean 20 months*	PTA with CI: 80 dB. mWRS 0%, dWRS 5%, SRS 5% in quiet without LR. Subjective benefit. Daily user. No telephone use.
	5	Mean 20 months*	PTA 25 dB. mWRS 60%, dWRS 78%, SRS 82% in quiet without LR. Daily user and user of telephone.
Tran Ba Huy et al.[11]	1	N/A	1y post: Phonemes: 100%; Closed-set words: 100%; Open-set words: 96%; Open-set sentences without LR in quiet: 96%; Open-set sentences without LR in noise: 91%.
			4y post: Phonemes: 95%; Closed-set words: 100%; Open-set words: 100%; Open-set sentences without LR in quiet: 97%; Open-set sentences without LR in noise: 89%.
Trotter et al.[12]	1	3 years	CUNY in quiet 96%, CUNY in noise 79%. CNC phonemes 79%. Subjective improvement in communication with LR. Daily user.
	2	12 months	CUNY in quiet 72%. CNC phonemes 45%. Subjective benefit.
	3	N/A	Daily user. Subjective good benefit.

AzBio, Arizona Biomedical Institute sentence list; Bi, bisyllables; BKB, Bamford–Kowal–Bench; CI, cochlear implant; CNC, consonant nucleus consonant; CUNY, City University of New York sentences; dB, decibel; dWRS, disyllabic word recognition score; LR, lipreading; HINT, Hearing in Noise Test; MTS, Monosyllable, Trochee, Spondee; mWRS, monosyllabic word recognition score; NU-CHIPS, Northwestern University Children's Perception of Speech; OSP, open-set speech perception; PTA/SRT, pure tone average/speech recognition threshold; SDS, speech discrimination score; SRS, speech recognition; WRS, word recognition score. *Mean time at follow-up: 20 months [range of all patients in Tan H et al.[17]: 12-60 months].

DISCUSSION

The NF2 patient case presented above received a CI in a tumor ear that had undergone radiotherapy to stabilize the tumor. Six months after implantation, the patient had excellent performance as measured objectively by speech discrimination. Subjectively, the patient experienced substantially improved hearing-related quality of life and a considerable decrease of tinnitus burden. The case illustrates that despite compromised functionality of the cochlea and the cochlear nerve by both the tumor and radiotherapy, good hearing outcomes may be achieved with a CI.

Although heterogeneous in many aspects, the scrutinized literature confirms that the majority of radiated patients will improve their hearing status after cochlear implantation, with some patients obtaining ceiling scores on postoperative hearing tests.[11,15,18,20,23] However, some patients only experience small or no improvements,[15-18] with postoperative hearing test scores showing very limited to no speech perception at all.[15-18] Yet again, it is remarkable that in general, low-performance patients are still daily users and report improved hearing-related quality of life, despite poor objective results.[12,15-18] This proves to show that very small improvements of hearing may have a great significance for this group of patients.

Interestingly, several of the included papers present both patients who score exceptionally high and patients who score very poor in the postoperative hearing tests, reflecting the unpredictability of results, even within individual treatment centers. Tan et al.[17] reported that good contralateral hearing status correlates with poor speech perception outcomes from the CI ear. However, Lundin et al.[24] found no significant relationship but a positive trend between bilateral preoperative and postoperative hearing outcomes.

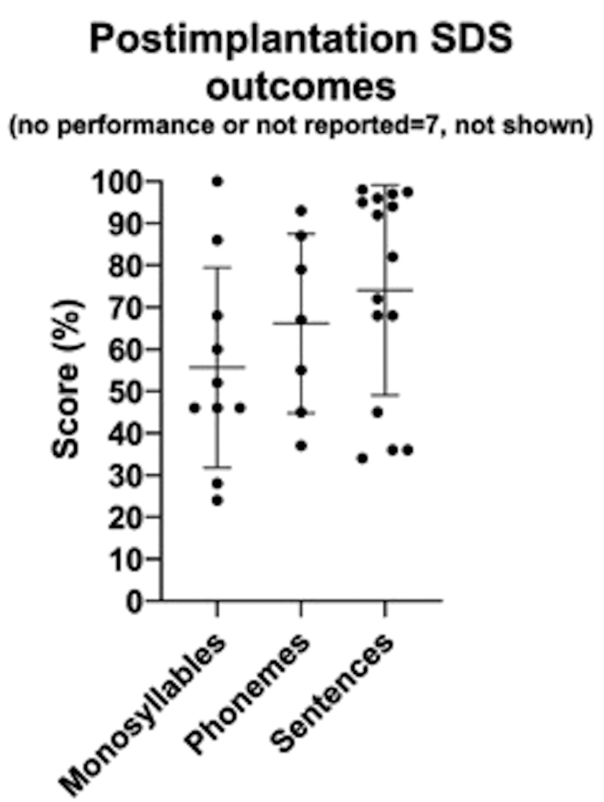

Figure 6. Reported speech discrimination after cochlear implantation in 26 radiated VS patients. Seven of the 33 patients identified in the literature review could not be plotted, as they were not reported in a way to allow plotting or because of no performance. VS = vestibular schwannomas.

Tumor grade does not seem to predict post-intervention hearing outcomes. The material is however too small and heterogeneous for a proper statistical analysis. Equivalently, it is not possible to make robust conclusions concerning the effect of type, field, and dose of radiation on outcomes after CI. Other potential relationships worthy of investigation with regard to postoperative hearing outcomes include patient age at implantation, preoperative cochlear nerve functionality, cochlear fibrosis/ossification, duration of HL prior to radiation, duration of HL between radiation and CI, location and size of tumor, and time at postoperative follow-up. Especially noteworthy for the reviewed literature is the variability concerning the use of preoperative cochlear nerve testing, and the method used for this. Cochlear nerve test results are likely to be the most precise predictor of postoperative hearing outcomes.

Although long-term hearing deprivation prior to CI is an established negative predictor of outcome,[16,25] it should not be an absolute contraindication for implantation, as good outcomes have been reported despite decades of deprived hearing.[26] Lin et al.[27] suggested that older patients with lower preoperative speech discrimination experience less postoperative benefit from CI surgery, and Chatelin et al.[28] concluded that postoperative performance improvement is slightly less for elderly CI patients. However, other papers report no significant correlation between age and postoperative auditory performance,[29,30] and it should be stressed, of course, that the mechanism of HL is altogether different for radiated VS patients as compared to conventional CI candidates. In this review, the mean age of the 7 cases across 5 papers that scored exceptionally high on postoperative hearing outcomes was 47 years.[11,15,18,20,23] The mean age of 7 cases that scored poorly was 48 years.[15-18]

All but 1 patient identified through the systematic literature review was classified as having NF2. This is not surprising, as the risk of significant bilateral HL is much higher than for unilateral, non-NF2 cases. Single-sided deafness is however becoming an established indication for CI,[31] and an increasing number of patients with radiated unilateral tumors should thus be expected as future CI candidates. In a recent systematic review on outcomes after microsurgical VS resection and simultaneous cochlear implantation, unilateral cases comprised 61%.[32]

As it is well known from systematic reviews in general, the reviewed literature lacks homogeneity concerning reporting, follow-up, and tests used. In addition, all studies included in the review were retrospective case reports or case series, and the strength of evidence is therefore inherently low. Furthermore, we estimate that underreporting of unsuccessful patient treatments may be an issue, thus implicating reporting and publication bias, leading to overestimation of the success of treatment.

CONCLUSION

The presented case and systematic literature review provide evidence that the majority of radiated VS patients will benefit from a CI, both objectively and subjectively. Although most patients have a good or even excellent hearing outcome, some perform poorly. This variability is likely to reflect patient and tumor-related characteristics, as well as radiation type, field, and dose. The exact impact of these variables is unclear and needs to be addressed in future research.

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