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Cochlear implantation for hearing rehabilitation in single-sided deafness after translabyrinthine vestibular schwannoma surgery

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Abstract The aim of the study was to investigate the option of cochlear implantation (CI) in resultant single-sided deafness associated with unilateral translabyrinthine resection of sporadic vestibular schwannoma (VS). This is a retrospective study performed at Tertiary Care Academic Centre. Following extensive counselling regarding the potential for delayed CI, translabyrinthine VS resection was performed and an intracochlear placeholder was inserted to allow later CI in 11 patients who showed intraoperative microscopic confirmation of preserved cochlear nerve anatomy. Follow-up magnetic resonance imaging (MRI) and promontory testing were performed 1 year after surgery to confirm the absence of VS recurrence and viable cochlea. Confirmed CI candidates underwent a second procedure where the placeholder was removed and the CI inserted (4/11). Preimplant unaided and CI-aided evaluations at 12 and 24 months were performed for subjective and objective hearing outcomes. Tinnitus suppression was also measured for implant on and off effects. Available audiological data for three patients demonstrated significant hearing benefits for 'speech from deaf/implanted side, noise from the normal-hearing side' in all three patients and localisation ability improved for 2/3 patients. Subjective findings presented similar results. For the two patients with preimplant tinnitus, complete suppression occurred during active CI. CI is beneficial for hearing rehabilitation and tinnitus reduction in SSD patients with remaining viable cochlear nerve after translabyrinthine VS surgery. Counselling on the risks of intracochlear placeholder insertion and the inherent limitations for ongoing MRI investigations of VS recurrence is essential.

Keywords Vestibular schwannoma · Acoustic neurinoma · Single-sided deafness · Cochlear implantation · Placeholder

Introduction

The sporadic vestibular schwannoma (VS) is a benign tumour and most commonly originates close to the vestibular ganglion at the junction of the central and peripheral myelin near the fundus of the internal auditory canal (IAC) at the Schwann cell–glial junction. It can be found anywhere along the nerve from the IAC to the terminal ends of the eighth cranial nerve within the vestibule, cochlea, or semicircular canals [1–3]. VS resection can cause several physical impairments such as vestibular disorders with unsteady gait, persistent postoperative headache, and might precede complications such as cerebrospinal fluid leak or facial nerve dysfunction. The postsurgical care often focuses only on these problems. Although most VS patients will have suffered from ipsilateral hearing impairment, most would have had at least residual hearing abilities before translabyrinthine surgery. After translabyrinthine surgery, they immediately sustain either asymmetric hearing loss (AHL) or single-sided deafness (SSD) similar to patients with sudden hearing loss. The impact of SSD or AHL can be very debilitating in daily function. These patients experience significant disabilities in many situations, especially when communicating in competing background noise where speech arrives at

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the poorer ear [4]. They also experience the inability to identify the location of sound and the loss of binaural advantages gained from binaural summation, binaural squelch and the head shadow effect [5, 6].

Two types of amplification systems providing pseudo-binaural hearing are available for SSD/AHL patients that transfer signals arriving at the side of the head with the poorer ear to the better hearing ear via contralateral routing of sound (CROS). This is achieved either externally using a conventional CROS hearing aid or transcranially, providing amplification via a bone-conductor device on a soft band. Nevertheless, CI is the only rehabilitation option that can potentially provide binaural hearing benefits to patients suffering from single-sided deafness in the presence of a viable cochlea on the affected side [7].

Several studies on patients with conventional SSD/AHL aetiologies (such as sudden hearing loss, endolymphatic hydrops or temporal bone fracture) have demonstrated significant improvement for speech understanding in noise and localisation abilities, and for subjective measures of hearing improvement, tinnitus suppression, and enhancement of quality of life with CI [7–14]. The circumstances for VS patients, however, are special since the VS removal can cause extensive trauma to the labyrinth. Experience with cochlear implantation following VS resection has been reported for patients with bilateral hearing loss due to bilateral (neurofibromatosis type 2, NF 2) and unilateral sporadic VS in the only or better hearing ear [15–17]. Several authors have shown that in approximately 60–70 % of these patients CI can provide open-set discrimination as long as anatomic integrity of the cochlear nerve during tumour excision is maintained [17–19]. It should be pointed out that translabyrinthine resection of VS, unlike the retro-mastoid sub-occipital and transtemporal approach, always includes a (partial) labyrinthectomy and, thus, can be highly traumatic to the delicate inner ear structures, including the spiral ganglion cells and cochlear nerve. Furthermore, both arterial blood supply and venous drainage are endangered during tumour removal; consequently, fibrosis and ossification will affect the cochlea postoperatively [20]. An intracochlear placeholder can reduce fibrotic changes inside the cochlea and allow CI placement during a later surgery [15].

Studies of translabyrinthine VS resection in the only hearing ear with concurrent ipsilateral hearing loss and simultaneous ipsilateral cochlear implantation have shown good functional outcome with CI [15, 21]. The above-mentioned reports have shown that CI is a viable hearing rehabilitation treatment for patients with translabyrinthine VS given residual cochlear function; however, to our knowledge, there is no current published literature on CI treatment in patients with SSD due to translabyrinthine VS resection.

The aim of this retrospective study, therefore, was to investigate the specific procedure of simultaneous VS resection and intracochlear placeholder insertion with a two-stage cochlear implantation after at least 1 year in patients with SSD due to translabyrinthine VS surgery, and the evaluation of the objective and subjective hearing outcome with CI.

Materials and methods

Subjects

All patients between June 2011 and May 2015 operated at the University Hospital in Freiburg implant facility who had undergone translabyrinthine resection of an intracanalicular VS (defined as tumour limited to the IAC = tumour stadium T1), intralabyrinthine schwannoma, or intralabyrinthine schwannoma with intracanalicular extension and who had given preoperative consent for intracochlear placeholder insertion were included in the study if intraoperative examination under microscope indicated preservation of the cochlear nerve anatomy.

Patients were counselled about possible complications associated with translabyrinthine surgery including progressive ossification of the cochlea and reduced rehabilitation results with loss of binaural hearing, as well as treatment options for the postoperative resultant SSD or AHL (conventional versus bone-anchored CROS, Bi-CROS and CI). In the event patients considered CI as a treatment option, they were advised that to prevent cochlear ossification an intracochlear placeholder would be inserted during the surgery if the integrity of the cochlear nerve could be confirmed microscopically.

The intracochlear placeholder is a silicone-covered electrode array without any attached implantable parts. It has a basal diameter of 0.8 mm and apical diameter of 0.5 mm. The pre-curved design for perimodiolar placement is supported by a stylet to keep the placeholder straight during the initial insertion phase ("Depth Gauges"; Cochlear Ltd., Lane Cove, Australia). At the authors' tertiary centre, intracochlear placeholders are also used for intracochlear insertion sampling in cases of presumed ossification and simultaneous electrode array insertion. The insertion and the intracochlear placeholder itself are currently an "off-label use". The application of intracochlear placeholders is only recommended in expert hands. Detailed information about the risks associated with the intracochlear placeholder insertion; e.g. infection, foreign body reaction, misplacement, as well as the lack of approval especially for continuous use in the human body must be provided to the patient during the pre-surgical counselling phase. All patients signed written consent form

for the placeholder. During VS surgery, the desired insertion depth was 22 mm for all patients. After placeholder insertion, an autologous fat graft was implanted in the translabyrinthine approach. Furthermore, the antrum, the posterior tympanotomy and opened air cells at the facial recess were sealed with fascia of the temporalis muscle and fixed with fibrous glue in each case to avoid cerebrospinal fluid leakage.

Twelve months after VS surgery, the first magnetic resonance imaging (MRI) was performed to check for VS recurrence as recommended in the literature [22, 23]. Within the same time frame, in all patients included a promontory test (PT) was performed to evaluate the integrity of the cochlear nerve by placing a needle electrode transtympanically on the basal turn of the cochlea (Cochlear Promontory Tester, Cochlear LTD, Melbourne, Australia). The PT aims to elicit an auditory perception in response to an electrical stimulation [24]. Depending on the result of the PT, patients were counselled on possible implantable hearing amplification options. In case of a positive PT, cochlear implantation was suggested in addition to conventional CROS hearing aids and bone-anchored CROS. In case of a negative PT result, patients were advised against hearing rehabilitation with a CI, and conventional CROS and bone-conduction CROS devices were recommended. During pre-surgical examination for CI evaluation, a 3-week trial with conventional CROS or Bi-CROS hearing aids and a bone-anchored CROS (bone-conduction device mounted on a soft band) was performed.

For potential CI candidates, hearing outcomes were assessed with a battery of objective and subjective tests in aided and unaided and postimplant CI-aided listening conditions.

Preimplant (post-VS resection surgery and prior to CI) unaided and postimplant open-set, speech recognition in noise tests with CI at 12- and 24-months were performed. The Oldenburg sentences test (OlSa) was used with an adaptive procedure in three test conditions with background noise set at 65 dB SPL: S_0N_0 (noise and speech from front), SnhNssd (speech from the normal-hearing side, noise from deaf/implanted side) and SssdNnh (speech from deaf/implanted side, noise from the normal-hearing side). The test setup is described in detail by Arndt et al. [7].

Localisation measurements were performed in the sound field with a frontal, semi-circle (2 m in diameter) consisting of seven, equidistant loudspeakers in a horizontal plane at intervals of 30° positioned at the subjects' head level as described by Arndt et al. [7]. The speech stimuli, OlSa sentences, were presented at a sound level of 65 dB SPL. The ability to localise was recorded by localisation error in degrees (°). The chance level for correct speaker localisation is 14.3 %.

Subjective assessment

Subjective assessment was evaluated using the standardised Speech, Spatial and Qualities of Hearing (SSQ) questionnaire at preimplant and at 12 and 24 months, post-first fitting. Questions in each of the three subcategories related to speech understanding, spatial hearing, and quality of hearing are scored from 0 to 10 where 0 represents 'unable to hear' and 10 indicates 'hears perfectly'. Tinnitus distress was measured with the visual analogue scale (VAS) preimplant and at 12 and 24 months after CI activation with patients rating tinnitus intensity on a scale from 0 (no tinnitus) to 10 (maximum strength). Furthermore, anecdotal information about daily CI operating times was gathered.

Data analysis

For speech perception in noise using the OlSa in an adaptive procedure, a difference of 1 dB or greater ($p > 0.05$) in speech reception threshold (SRT_{L50}) is taken to be outside of normal test–retest variation and considered as an intra-individual significant difference [25, 26].

Results

The MRI findings obtained 1 year after VS resection surgery ruled out the recurrence of VS in all 11 patients. All patients presented with sensorineural deafness in the unilateral ear as a result of the translabyrinthine surgery (SSD in 8; AHL in 3). Pure-tone average (PTA) thresholds of the contralateral ear and subsequent classification into SSD and AHL subgroups are presented in Table 1.

Transtympanic PT was offered to all patients with four patients rejecting the option to undergo further examination. The reasons for rejection were persistent vertigo after VS surgery in two cases. The remaining two patients requested more time after the VS surgery. Three out of four patients that rejected PT decided against any hearing amplification for the time being. One subject opted for conventional Bi-CROS because of fears associated with another surgery.

The PT was performed on the remaining seven subjects. Negative PT results were obtained in two patients and one of these patients with SSD received a conventional CROS system; the other patient with AHL opted for a Bi-CROS.

Positive PT test results were found in five subjects who performed a test trial with pseudo-binaural hearing amplification systems (conventional and bone-anchored CROS) and were counselled about CI again. A total of four patients underwent cochlear implantation. All four subjects went with CI surgery because they did not experience subjective

Table 1 Patient demographics with pure-tone average (PTA_4 contralateral at 500, 1000, 2000, and 4000 Hz)

Subject	Side	PTA_4 contralateral (dB)	SSD/ AHL	Age at VS surgery (years)	Intracochlear placeholder	Tumour extension	PT	Amplification chosen (time interval in-between VS and CI surgery in brackets)
1	Left	10	SSD	32	Yes	T1	pos	CI (14 months)
2	Left	12.5	SSD	53.7	Yes	Intralabyrinthine schwannoma with intracanalicular extension	pos	CI (13 months)
3	Right	15	SSD	48.4	Yes	Intralabyrinthine schwannoma	pos	CI (19 months)
4	Right	47.5	AHL	57	Yes	Intralabyrinthine schwannoma	*	NIL
5	Right	7.5	SSD	58	Yes	T1	pos	CROS
6	Left	40	AHL	64.6	Yes	T1	*	Bi-CROS
7	Left	35	AHL	51	Yes	T1	neg	Bi-CROS
8	Right	11.25	SSD	49.8	Yes	T1	*	NIL
9	Right	8.75	SSD	54	Yes	T1	*	NIL
10	Left	25	SSD	63	Yes	T1	pos	CI[a] (13 months)
11	Left	12.5	SSD	69	Yes	T1	neg	CROS

VS vestibular schwannoma, *T1* tumour classification stage, *PT* transtympanic promontory test, *pos* PT testing positive, *neg* PT testing negative, *CI* cochlear implantation, *CROS* contralateral routing of signal, *Bi-CROS* binaural contralateral routing of signal

* Patient rejected PT

[a] Patient underwent CI surgery April 2015; no rehabilitation data are currently available

benefit with conventional CROS or Bi-CROS hearing aids and bone-anchored CROS during test trial. The remaining patient with positive PT decided on conventional CROS hearing aid due to fears of a renewed surgery. For detailed demographic information, see Table 1. No complications were observed due to implantation of the intracochlear placeholder, to date, in all subjects.

As of this writing, three patients have attained 12- and 24-month CI rehabilitation data. One patient who had underwent T1 VS resection received CI surgery in May 2015 so that no rehabilitation data could be collected. During CI surgery, posterior tympanotomy was detected by following the outer auditory canal wall since it had already been performed during previous surgery. Intracochlear electrode was inserted directly after extraction of the placeholder. Explanation of the intracochlear placeholder and CI surgery was uneventful in all four subjects.

The following presents individual data for three patients with long-term CI outcomes ($n = 3$) after translabyrinthine schwannoma surgery.

Subject 1

Subject 1 (S1) suffered from an intracanalicular (T1) VS. CI surgery (Nucleus CI24 RE (CA), Cochlear Ltd., Lane Cove, Australia) was performed 14 months after initial translabyrinthine VS resection with simultaneous placeholder insertion. The intracochlear placeholder extraction was without complications and the CI electrode was inserted without resistance. No postoperative complications occurred. Age at implantation was 34 years and duration of subjective deafness was 21 months (presurgical deafness of 7 months plus 14 months of deafness after surgery).

The OlSa sentence critical SNR_{L50} results unaided at the preimplant, 12 and 24 months intervals with CI are illustrated in Fig. 1. Twelve months after CI surgery, speech comprehension ability in competing background noise conditions improved significantly from SNR_{L50} 0.9 dB unaided to −7.8 dB, −7.6 after 24 months with CI in the $S_{SSD}N_{NH}$ situation, respectively ($p > 0.05$). For coincidental speech in noise testing (S_0N_0), a significant improvement of speech recognition SNR_{L50} with CI was identified at 12-month postimplant (SNR_{L50} −5.2 dB) and 24 months (SNR_{L50} −5.9 dB) compared to the unaided situation (SNR_{L50} −4.1 dB). In the $S_{NH}N_{SSD}$ condition, there was a significant improvement from unaided SNR_{L50} −11.7 to −14.3 dB at 12-month postimplant ($p > 0.05$) followed by a significant deterioration at 24-month postimplant (SNR_{L50} −12.7 dB; $p > 0.05$). S1 showed good performance in localisation testing, resulting in a decrease of localisation error from 27.4 unaided compared to 20.5 at 12 months and 18.9 at 24-month postimplant (Fig. 2). Subjective assessment revealed a remarkable

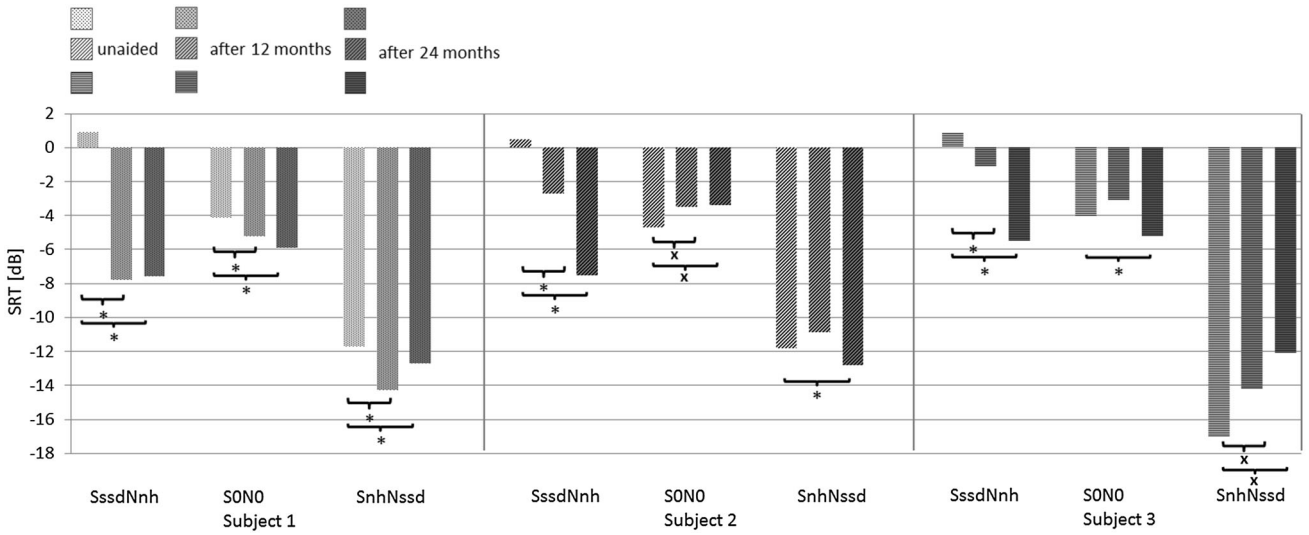

Fig. 1 Critical SNR_{L50} of the three subjects obtained with the OLSA sentence in noise test for the unaided condition and the aided condition with CI after 12 and 24 months. *SnhNssd* = speech from the normal-hearing side, noise from deaf/implanted side; S_0N_0 noise and speech from front; *SssdNnh* speech from deaf/implanted side, noise from the normal-hearing side. *Significant improvement compared to unaided condition ($p < 0.05$). [x]Significant deterioration compared to unaided condition ($p < 0.05$)

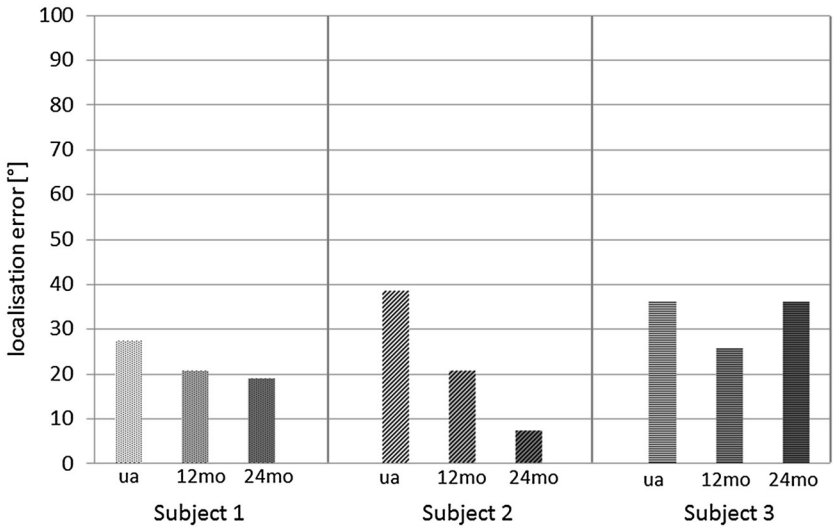

Fig. 2 Localisation error unaided (ua), 12- and 24-month postimplant results in SSD patients after VS surgery

improvement in the speech understanding section as well as in the spatial section at 12- and 24-month postimplant compared to the unaided condition (Fig. 3). S1 did not suffer from tinnitus at any time. The patient reported that she enjoys wearing her CI all day and does not notice any major differences in sound and speech quality in comparison to her healthy ear, suggesting that both ear signals are integrated forming one speech signal.

Subject 2

Tumour expansion of VS in S2 revealed an intralabyrinthine schwannoma with extension in the lateral semicircular duct as well as the anterior semicircular duct and the vestibulum with proximity to the stapes footplate. Endoscopic control showed no residual tumour inside the vestibulum and entry to cochlea. Intracochlear place holder was inserted without any resistance since cochlear nerve could be preserved. VS surgery and postoperative course were uneventful.

CI surgery (Nucleus CI24 RE (CA), Cochlear) with uneventful extraction of intracochlear placeholder was performed 13 months after translabyrinthine VS resection. Age at implantation was 53.7 years. Duration of subjective unilateral deafness until CI surgery was 19 months.

Individual speech test outcomes are shown in Fig. 1. In the $S_{SSD}N_{NH}$ configuration, SNR_{L50} significantly improved from 0.5 dB in the unaided situation to −2.7 dB after

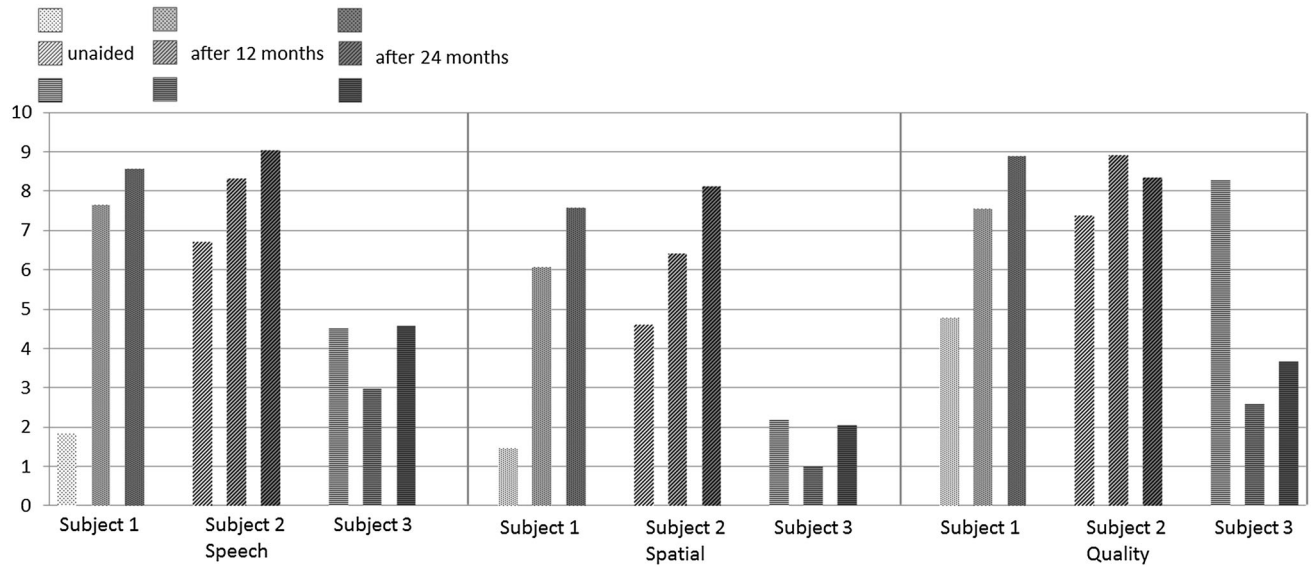

Fig. 3 Subjective self-assessment by SSQ for patients suffering from SSD after VS surgery unaided preoperative, 12 and 24 months after cochlear implantation

12 months, respectively, −7.5 dB after 24 months when listening to the binaural condition with CI. In the S_0N_0 configuration, SNR_{L50} deteriorated significantly from unaided −4.7 to −3.5 dB after 12 month and to −3.4 dB at 24-months postimplant ($p > 0.05$). In the $S_{NH}N_{SSD}$ condition, there was non-significant deterioration at 12-month postimplant (SNR_{L50} −10.9 dB) compared to baseline (SNR_{L50} −11.8 dB). After 24 months, a significant recovery was seen (SNR_{L50} −12.8 dB; $p > 0.05$). S2 presented with good performance in localisation testing, resulting in a notable decrease of localisation error from 38.6 unaided compared to 20.6 at 12-month and 7.2 at 24-month postimplant (Fig. 2).

Subjective evaluation revealed a substantial improvement at 12- and 24-month postimplant in the speech and spatial section compared to baseline. Only little change could be found in the 'quality of hearing' section (Fig. 3). S2 suffered from unilateral severe tinnitus after translabyrinthine VS surgery on the same ear, which decreased after surgery and is completely suppressed when the CI speech processor is activated (Fig. 4).

S2 anecdotally reports that she wears her CI more than 8 h per day. She points out that speech understanding in group situations with CI is much better than without CI. There are no major differences in sound and speech quality in comparison to her healthy ear.

Subject 3

S3 suffered from an isolated intralabyrinthine schwannoma. During translabyrinthine surgery, the tumour was

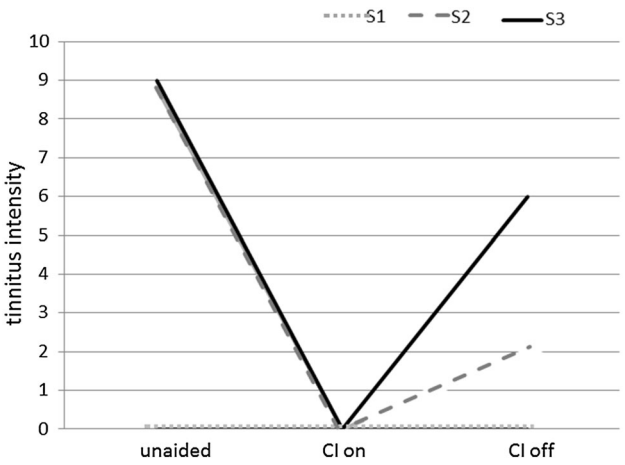

Fig. 4 VAS tinnitus loudness of three subjects before CI implantation (unaided), 12 months after CI surgery, with CI switched on and off. Patients rated tinnitus intensity on a scale from 0 (no tinnitus) to 10 (maximum strength). S1 did not suffer from tinnitus. *S* subject

resected via partial labyrinthectomy from cochlear basal and second turn, as well as from the vestibulum. The intracochlear placeholder was inserted and the promontorium was reconstructed with bone meal and fibrin adhesive. Intraoperative X-ray demonstrated correct positioning of intracochlear placeholder. No intra- or postoperative complications occurred. Twelve months after intralabyrinthine schwannoma surgery, MRI showed no recurrence of VS and the PT was positive. Age at implantation was 50 years and duration of subjective deafness was 96 months. S3 ruled out both pseudo-binaural hearing amplification options after test trials and decided for CI surgery. Extensive counselling had been done considering

the invasive surgery and the long duration of deafness. CI surgery [Nucleus CI24 RE (CA), Cochlear] with uneventful extraction of the intracochlear placeholder was performed 19 months after initial tumour surgery for which no intraoperative or postoperative complication occurred. Postoperative RT showed correct positioning of the CI electrode (Fig. 5).

Audiological test results preoperative unaided, and 12- and 24-month interval data using OlSa sentence critical SNR_{L50} are illustrated in Fig. 1. OlSa speech threshold testing in the spatially separated test configuration $S_{SSD}N_{NH}$ shows a significant improvement from SNR_{L50} 0.9 dB unaided to SNR_{L50} −1.1 dB after 12 months and to SNR_{L50} −5.5 dB after 24 months with CI ($p > 0.05$). For the S_0N_0 presentation configuration, significant improvement can only be found after 24 months with CI (SNR_{L50} −5.2 dB) compared to the unaided condition (SNR_{L50} −4.0 dB; $p > 0.05$). For the spatially separate test configuration, $S_{NH}N_{SSD}$, significant deterioration of SNR_{L50} was demonstrated after 12 (SNR_{L50} −14.2 dB; $p > 0.05$) and 24 months (SNR_{L50} −12.1 dB; $p > 0.05$) compared to baseline (SNR_{L50} −17.0 dB) (Fig. 1). Localisation error decreased from 36.0 (unaided) to 25.7 (12 months) and 18.9 at 24-month postimplant (Fig. 2). The SSQ scores showed deterioration in all three sections after 12 months. In the speech and spatial sections, these values returned to baseline at the 24-month interval (Fig. 3). The 'quality of hearing' subsection was rated minimally better for 24 months compared to 12-month results.

S3 suffered from incapacitating tinnitus before CI surgery. Tinnitus was completely suppressed with CI activated, as shown by VAS ratings in Fig. 4.

Anecdotal reports revealed full-time CI usage, quality of hearing with CI was described initially as partly distorted, so that CI caused some distraction to speech understanding in noisy environments. After a change of the coding strategy to MP3000^{TM} [27] in 04/2014, less distortion was reported.

Discussion

Hearing rehabilitation in patients with SSD due to translabyrinthine VS resection is particularly challenging since choices have to be made in advance of the VS surgery. Besides pseudo-binaural hearing amplification, cochlear implantation and perhaps a placeholder might be an option for these patients, as long as anatomic preservation of the cochlear nerve is achieved.

The translabyrinthine pathway induces postsurgical ossification of the cochlea, which hinders later consideration of electrode insertion for successful hearing rehabilitation through cochlear implantation [20]. The exact time of ossification of the cochlea following trauma to the temporal bone irrespective of its cause remains unknown [28]. In a human temporal bone study, Belal [20] observed total cochlear ossification with almost total degeneration of the spiral ganglion cells and cochlear nerve with postoperative survival times varying from 4 to 11 years. Therefore, he recommended simultaneous tumour removal and cochlear implantation during translabyrinthine surgery [20]. If cochlear implantation is planned in a second stage, an intracochlear placeholder to reduce fibrotic changes inside the cochlea can be inserted to allow later CI placement [15].

Successful application of intracochlear placeholder to prevent cochlear ossification prior to cochlear implantation has already been demonstrated in patients suffering from labyrinthitis ossificans [29]. In this study, the intent of initial placeholder insertion and 'wait and see' procedure for the first year after VS removal instead of simultaneous CI surgery during VS resection lays in the fact of possible, undetected microscopic deposits left on crucial points such as the facial nerve, the preserved cochlea nerve, or the fundus of the internal auditory canal, which could be possible causes for a recurrence [30]. If hearing rehabilitation with CI for consecutive SSD/AHL was requested, our patients were advised to undergo implantation of an

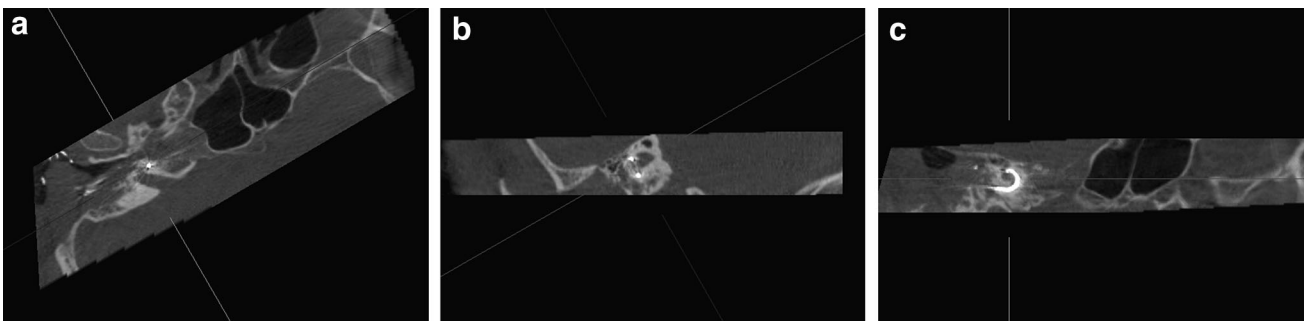

Fig. 5 Postoperative rotational tomography scan showing correct positioning of the electrode. Image display of oblique angle reoriented RT (oriented towards the reconstructed cochlear) to display the electrode. **a** Oblique axial section with inner ear canal; **b** midmodiolar reconstruction, only first cochlear turn; **c** cochlear view orientation

intracochlear placeholder during VS surgery to prevent cochlear obliteration and allow MRI recurrence check and CI placement at a later date.

Recurrence rates after translabyrinthine VS resection have been reported to be up to 9 % [23]. Ahmad et al. [30] presented an extremely low rate of 0.05 % during 1 and 13 years after the initial surgery; however, this was for an adaptive enlarged translabyrinthine approach. Gadolinium-enhanced MRI represents the gold standard imaging modality for VS post-treatment follow-up. First postoperative MRI scan for recurrence check should be performed 12 months after VS surgery [22, 23]. Accessibility to assess VS recurrence in MRI was not decreased by intracochlear placeholder in the present study. There was no VS recurrence detected in any of the 11 patients after 12 months. Time delay in cochlear implantation might be crucial due to possible spiral ganglion cell loss after translabyrinthine VS removal; nonetheless, early stage recurrence in critical regions can occur and would necessitate further therapy. Detection of VS recurrence in MRI after cochlear implantation with a CI magnet in place can become challenging since artefacts can hinder realistic assessment of auditory-vestibular structures [31].

Transtympanic PT was recommended to all 11 patients for additional diagnostic evaluation since it can give evidence of survival of cochlear nerve fibres relative to intact spiral ganglion cells if an auditory perception in response to an electrical stimulation is present [20, 24]. Controversy exists as to the prognostic value of the PT results [32] since negative responses on PT do not exclude the ability to make functional use of the cochlear implant signal [33–37]. However, transtympanic PT can help when counselling CI candidates about expected outcomes and alternative pseudo-binaural treatment options. In the present study, 2/7 patients agreeing to undergo evaluation had a negative PT result despite intraoperative microscopic anatomical examination suggesting a preserved cochlear nerve. We would not recommend CI treatment for patients with negative PT after translabyrinthine VS resection since functional benefit from CI use remains less likely. Furthermore, SSD/AHL patients continue to receive auditory signals in the contralateral ear, which makes the situation more difficult than in patients with bilateral hearing loss due to VS. Therefore, hearing rehabilitation would be reduced to pseudo-binaural options such as conventional CROS and bone-conducted CROS in case of negative PT. Special care in counselling must be given since these patients had earlier agreed to a placeholder and may have the expectation of receiving a CI and hearing rehabilitation. Regular attendance to the clinic is recommended to check on possible side effects due to the intracochlear placeholder. To date, no long-term complications have been seen in the remaining seven patients with permanent placeholder.

Four (4/5) patients with positive results in PT underwent CI surgery. Complete long-term audiological data were assessable for three patients, resulting in significant benefit to CI for all three implantees after 24 months in the most challenging test configuration for SSD patients, when sound is presented to the CI ear and noise to the normal-hearing ear. Consistent with this, is the significant benefit with CI for this spatial presentation already reported in SSD patients with conventional aetiologies (sudden hearing loss, endolymphatic hydrops, post-trauma) and shorter follow-up times [7, 9]. Needless-to-say, the results of Vermeire and Van de Heyning [9] cannot be compared directly with our results because of their more difficult presentation setup (loudspeakers were separated from each other by 90°).

Inconsistent results were demonstrated in both the other configuration presentations for the three SSD patients with CI after VS resection. The summation effect obtained from the S_0N_0 presentation configuration was significant only after 24 months in two patients (S1 and S3) with non-significance after 12 months, but in subject 2 the effect significantly deteriorated after 12 and 24 months. Literature on summation effect in SSD patients with conventional aetiologies presents inconsistent results, as well. While Vermeire and Van de Heyning [9] described a small deterioration of the critical SNR and Arndt et al. presented non-significant improvement in this presentation configuration, Távora-Vieira et al. showed the CI to be significantly beneficial for the summation effect compared to the unaided condition [7, 9, 14]. It remains unclear which factors contribute to a significant improvement in the summation effect in SSD patients. Our results showed a significant improvement in speech perception in noise scores when speech was presented to the CI ear and noise to the normal-hearing side at 24 months, postimplant, for two patients (S1 and S2) while subject 3 presented with significant deterioration. Other authors did not show significance [7, 9] or presented significant benefit of adding the CI for their SSD groups with conventional aetiologies [10, 14]. As mentioned, direct comparison may be affected by the audiological setups that were more difficult since loudspeakers were separated by 90° [9, 10, 14]. It is important to note that there was no deterioration of the SNR_{L50} in our two cases when the noise was presented to the CI side indicating that the CI did not hinder performance.

Assessment of localisation abilities in the present report revealed a substantial improvement for two subjects (S1 and S2) at the 24-month interval with CI. Subject 3 showed improvement at the 12-month interval but demonstrated a return to baseline values after 24 months. Literature shows a consistent improvement of localisation abilities with CI for SSD and AHL patients with conventional aetiologies [7, 12, 38].

Subjective assessment measures in SSQ questionnaires were consistent with objective audiological benefit of speech recognition. All three patients with CI following VS resection demonstrated superior scores for speech understanding when using the CI compared to the unaided condition. For S1 and S2, scores were better in the spatial hearing subsection (after 12 and 24 months) and qualities of hearing subsection (only after 12 months for S2) compared to the unaided situation; S3 showed no improvement for spatial hearing and reported deterioration for the quality of hearing subsection. The favourable results for S1 and S2 in all subsections of SSQ with the CI are in agreement with Vermeire and Van de Heyning [9] and Távora-Vieira et al. [14]. Arndt et al. [7] were able to prove a significant subjective benefit with the CI in the speech understanding and spatial section compared to the unaided situation while non-significance was shown for the 'qualities of hearing' section.

Cochlear implantation in SSD patients was initially used as a treatment option for debilitating tinnitus reduction [8]. Two patients presented in this study suffered from preoperative tinnitus. A complete suppression of tinnitus was obtained with activated speech processor. There was a recurrence of tinnitus after switching off the CI in both patients, in agreement with the reports in published literature [7, 13]. A significant reduction of tinnitus loudness or distress after cochlear implantation in SSD patients has also been shown by other authors [7, 8, 39, 40]. Tinnitus suppression seems to represent a very encouraging side effect for patients with SSD after translabyrinthine VS surgery and subsequent cochlear implantation.

The reason for the overall inferior results for S3 might lie in the long time interval in between VS resection and CI surgery as well as in the harmful effects of the tumour removal surgery due to the extension of the VS. In translabyrinthine VS resection, especially in case of intracochlear tumours, a complete removal without ganglion cell damage cannot be achieved. Postoperative fibrosis with ossification and further degeneration of spiral ganglion cells is predictable. Furthermore, early electrical stimulation by the cochlear implant is known to improve the survival of spiral ganglion cells and can prevent retrograde nerve degeneration [41, 42], which is supported by the good 24-months results of S1 and S2. Therefore, the consideration for simultaneous VS resection with CI surgery with the risk of restricted control of tumour recurrence via MRI should be considered in the future to achieve the earliest electrical stimulation possible. After translabyrinthine VS removal in patients with bilateral hearing loss and bilateral VS due to NF2 or unilateral sporadic VS, this has already been presented with functional hearing outcome [15].

Most contemporary cochlear implants are "MRI conditionally safe" with or without magnet removal depending on the specific conditions [43]. In addition, Todt et al. [44] has introduced alternative CI surgery procedures that proved to optimise image quality in 3-Tesla MRI with CI magnet in place. A nasion-external ear canal angle of 90° or 160° and a distance of 9 cm in relation to the external ear canal and specific MRI sequences make a complete assessment of the internal auditory canal and labyrinth possible [44]. Still, counselling of patients with sporadic VS and intended translabyrinthine resection must include that VS surgery necessitates follow-up imaging. An MRI after cochlear implantation for tumour recurrence check should still be carefully considered. It depends on the type of CI and its MRI compatibility. Furthermore, artefacts due to magnetic parts of the CI may be a factor. Computed tomography (CT), which is the gold standard for the evaluation of bony structures, can be performed in case of suspected, recurring VS instead of MRI. A CT can depict extensions and erosions of porous and internal acoustic meatus as an indirect sign of tumour regrowth [45, 46]. Patients must be counselled in advance that MRI might be less effective after CI surgery due to artefacts, at least until new procedures such as the ones currently presented by Todt et al. [44] have proven themselves and are integrated in clinical routine. Although highly useful, a CT might not be as reliable in the early detection of recurrent VS.

More data on the outcome of CI patients suffering from SSD/AHL due to translabyrinthine VS surgery are needed in support of the long-term benefit of hearing rehabilitation with CI. Furthermore, comparison of audiological outcome with CI to that with conventional CROS and bone-anchored CROS hearing systems is needed to indorse the superiority of CI which has already been demonstrated for other patient collectives [7]. The advantage of VS resection with simultaneous placeholder insertion and CI surgery with time delay in favour of a possibly better early stage recurrence verification must be weighed against the audiological benefits of early electrical stimulation in case of simultaneous CI surgery. A larger number of subjects are needed to allow general conclusions and to provide valid statistical analysis.

Conclusion

In the present study, cochlear implant rehabilitation as a treatment for SSD due to translabyrinthine VS surgery was evaluated for the first time. Overall, objective and subjective measures indicate that these patients may be able to make use of some of the benefits associated with binaural hearing. Furthermore, our data suggest that CI can suppress tinnitus after translabyrinthine VS surgery. Critical selection of candidates as well as extensive counselling is required to ensure awareness of risks in case of intracochlear placeholder insertion, about the different treatment options that include conventional CROS and bone-

conducted CROS, as to the potential binaural benefits of CI, and also possible MRI incompatibility or imaging limitations after CI surgery. Explicit declaration of possible neuronal injury to the auditory pathway during VS resection and afterwards is absolutely essential to avoid disappointment and provide reasonable expectations.

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Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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