Aus der Klinik für Hals-Nase-Ohrenheilkunde der

Medizinischen Hochschule Hannover

Long-term hearing outcome of cochlear Implantation in cases of simultaneous intracochlear schwannoma resection

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1. Summary:

Intracochlear Schwannomas (ICS) are rare tumors arising completely inside the cochlea, posing a therapeutical challenge, as their natural history and surgical removal invariably lead to anacusis. Historically little attention has been paid to hearing restoration after surgery, until the feasibility of extirpation followed by or combined with successful cochlea implantation was recently explored. However, the risk of recurrence in the follow-up remains unclear.

The aim of our retrospective study was to analyze the hearing performance of ICS patients treated with simultaneous tumor extirpation and cochlear implantation in the long term (36 months) compared with an age matched non-tumor single-side deafness cochlear implant patient cohort, also evaluating the use of the speech recognition tests in suspecting tumor recurrence.

In our experience, cochlear implantation is the strategy of choice in hearing rehabilitation in case of ICS, granting excellent results in the long term. The speech recognition tests can be effectively used to monitor for recurrence. Tumor extirpation via enlarged cochleostomy with simultaneous cochlear implantation offers a sufficient degree of radicality without compromising the cochlear integrity, granting excellent hearing rehabilitation results in the long term. The technique allows for revision if required.

2. Introduction

a. Vestibular Schwannoma and its current treatment

Sporadic vestibular schwannomas are benign nerve sheet tumors arising from the Schwann cells coating the neuronal axons along the vestibulo-cochlear nerve (NC VIII) from the membranous labyrinth to the brainstem. The etiology of the disease remains unclear, but it is supposed that defect on chromosome 22q may be responsible for the development of both sporadic and bilateral vestibular schwannomas as seen in Neurofibromatosis Type 2, which is determined by a mutation in the NF2 gene encoding the tumor suppressor merlin. As schwannomas develop inside the nerve sheath, they compress rather than infiltrate the nerve itself, thereby leaving a cleavage plane between the nerve fibers and the tumor, thus facilitating its surgical removal.

Vestibular schwannomas remain mostly asymptomatic for a long time, slowly growing and filling the space inside the internal auditory meatus, eventually protruding in the cerebello-pontine angle. The tumor invariably exerts pressure on the cochleovestibular nerve, leading to acute or progressive irreversible sensorineural hearing loss and recurrent vertigo spells. In case of major intracranial extension, they can eventually compress the brainstem, the facial nerve and the trigeminal nerve and occlusion of liquor circulation, requiring urgent removal and decompression. Due to their slow growth and the relatively

abundant free space around them, it is not unusual to diagnose vestibular schwannomas when they have already reached sizeable volumes[1].

As the diagnostic work-up for sudden hearing loss or vertigo includes nowadays a MRI scan of the skull base to rule out pathologies along the cochleovestibular nerve and in the brainstem, there has been a relative increase in the reported incidence of vestibular schwannoma in the last decades [2].

The current treatment of vestibular schwannomas is based on their size, location, clinical appearance and residual function of the cochleovestibular and facial nerve. Different treatment algorithms have been developed and are under constant scientific examination and discussion. As an example, all tumors still inside the internal auditory meatus without signs of growth and still preserved hearing function can be yearly controlled by means of magnetic resonance imaging[3]. In case of tumor growth or symptoms[4], the patient can be treated with radiotherapy[5] (in the form of stereotactic radiotherapy or gamma knife) or microsurgical extirpation[6]. There is an ongoing debate on the advantages and disadvantages of the two treatment modalities[7-10].

In case of radiotherapy or surgical treatment, there is a high risk of losing the residual hearing function due to progressive post-treatment cochlear fibrosis or damage of the auditory neurons, arising the question of hearing rehabilitation in vestibular schwannoma[11,12]. Traditionally hearing aids or CROS hearing aids (rerouting the hearing stimulation received by the affected ear to the contralateral side) have been the only options to rehabilitate the patients. In

recent years, the application of cochlear implants to substitute the damaged hearing organ has gained momentum[13,14]. The results are promising, but are generally less performing as compared with other forms of single-side deafness[15].

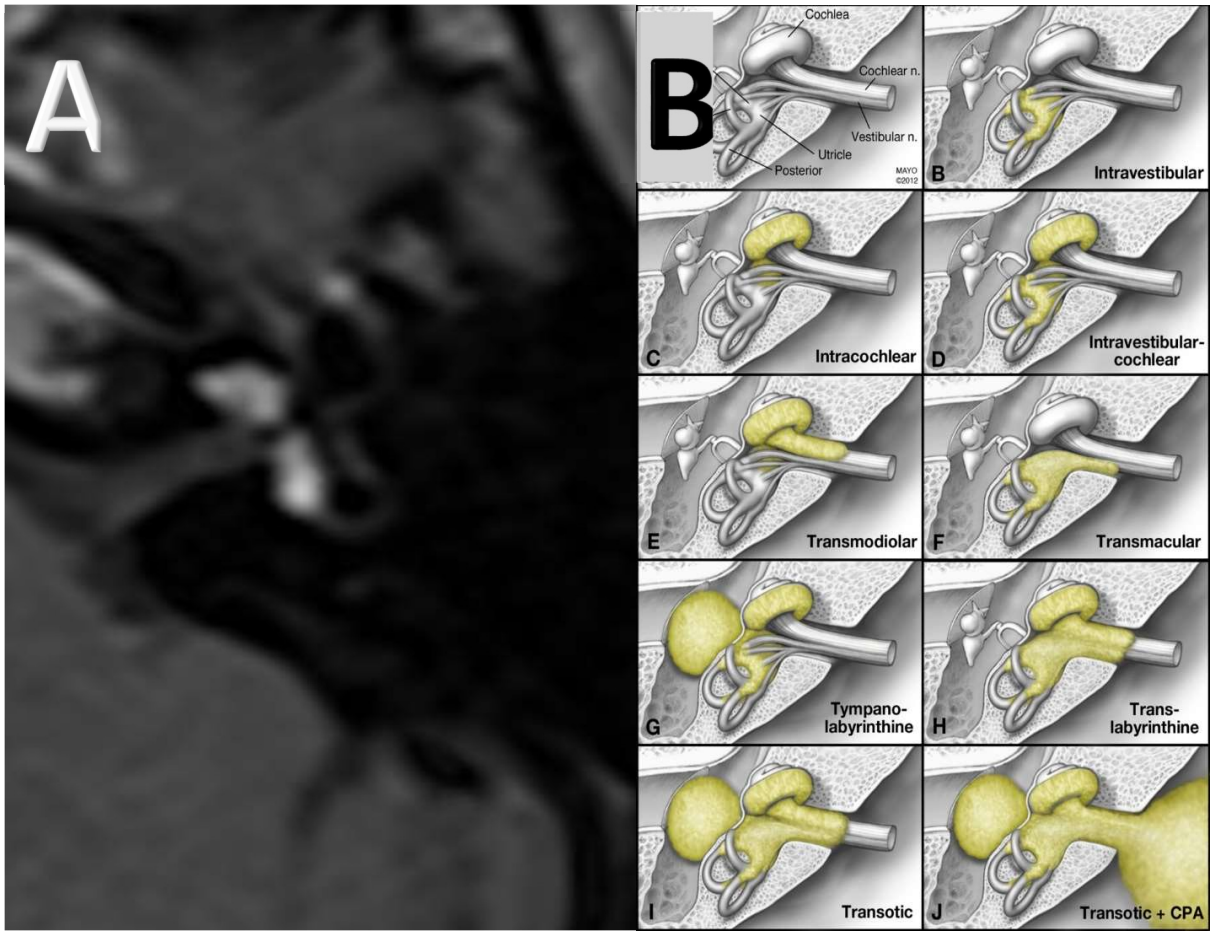

Fig.1: A) Example of an intralabyrinthine Schwannoma, specifically an intravestibulocochlear one. (Courtesy of MHH, HNO Abteilung)
B) Salzmann -Van Abel Classification of intralabyrinthine schwannomas
Courtesy of Van Abel, K.M., et al., *Primary inner ear schwannomas: a case series and systematic review of the literature.* Laryngoscope, 2013. 123(8): p. 1957-66.

b. Intracochlear Schwannoma Natural history and epidemiology

Intracochlear Schwannomas (ICS) represent the most common form of intralabyrinthine Schwannomas (ILS), which are currently anatomically

subclassified into seven groups based on the affected inner ear structures: intravestibular, intracochlear, intravestibulocochlear, transmodiolar, transmacular, transotic, and tympanolabyrinthine[16,17]. ICSs arise from the Schwann´s cells of the cochlear and vestibular nerve axons interspersed in the osseous spiral lamina and modiolus, proximal to the spiral ganglion[18,19]. A schwannoma arising from these cells will progressively occupy the scala media in the cochlear basal turn, invade the modiolus, erode the cribriform area of the IAC and then spread into the vestibule. The hearing loss may be determined by a combination of compression of the cochlear nerve, depleted vascular supply, mechanical interference with hearing function, or even progressive changes in the metabolic status of inner ear fluids[19-21]. There is epidemiological evidence that ILS are more common than previously thought [22] and ICS represent up to 50 % of ILS[18,19]. Moreover, there is the possibility, especially in neurofibromatosis patients, of combined tumors arising in the IAC and the labyrinth at the same time. These extremely small benign tumors pose a special clinical and therapeutical challenge to the neurotologist, as their natural history and surgical removal invariably lead to deep sensorineural hearing loss due to destruction of the cochlear anatomy.

c. Clinical Presentation of intracochlear schwannomas

The clinical presentation of intracochlear schwannoma may vary, but it is usually characterized by sensorineural hearing loss (94.5%), together with

Tinnitus (69.1% to 95.8%), vestibular symptoms (59.1%)[23] and aural fullness. The natural history of the disease is progressive, with recurrent sudden hearing loss episodes eventually leading to severe pantonal sensorineural hearing loss with poor speech perception, sometimes resembling the audiological presentation of Meniere's disease. If invasion and compression of the vestibule is present, the tumors may cause endolymphatic hydrops, resulting in disequilibrium and vertigo. Occasionally, a mixed-type hearing loss can occur, presumably due to increased intracochlear mechanical interference. In either way, the diagnosis is possible preoperatively only by means of a high-resolution magnetic resonance imaging. Unfortunately, there is a large delay between emergence of the symptoms and diagnosis, averaging 72.5 months, as the lesions are usually small, growth rate is slow, subtle progressive symptoms are easily compensated and a high resolution MRI is not always performed in case of SNHL and vertigo[24]. The time to functional deafness remains, however, unpredictable and a case-to-case approach should be adopted. Usually, the patient has already lost his serviceable hearing by the time of the first diagnosis and is in need of hearing rehabilitation. Although hearing loss may initially still be amendable with hearing aids, in case of not serviceable hearing on the affected side and acceptable surgical risk the indication for cochlear implantation is given. Indicatively all ICS are good candidates for cochlear implantation, although tumor removal may prove challenging.

d. **Radiological appearance of intracochlear Schwannomas**

Performing a high-resolution MRI in case of progressive or sudden sensorineural hearing loss facilitates the identification of ICS. Typically, an ICS is enhanced on T1-weighted images with gadolinium with sharply delineated edges and shows a signal loss in the high-intensity intralabyrinthine fluid on T2-weighted images. These MRI findings may mimic infectious or inflammatory processes in the labyrinth, making labyrinthitis the principal differential radiological diagnosis. However labyrinthitis usually shows an enhancement, which commonly affects the complete cochlea or the vestibular system, receding in the follow-up and there are no filling defects such as in ICS [18,25,26]. In cases of doubt, the application of temporal bone computer tomography to rule out intralabyrinthine fibrosis and ossification may be of assistance. Other rare differential diagnosis are intracochlear hemorrhage, which has a variable signal on T2-weighted images but appears as a high signal intensity lesion on T1-weighted images without contrast enhancement, and lipoma, which could also present as hyperintense lesion on T1-weighted images and hypointensity on fat-suppression sequences. Therefore, T1-weighted postcontrast enhanced images are pivotal to confirm that a lesion is a schwannoma [16,27].

e. Therapeutical options for intracochlear schwannoma

Once diagnosed, ICSs may not necessarily require immediate surgery and hearing rehabilitation. Although always leading to sensorineural deafness due to infiltration and destruction of the inner ear structures in the long term, ICS remains rather unpredictable. The tumor may remain dormant and paucisymptomatic for many years, whereas in other cases it shows a rapid increase in size and consequent decrease of the hearing capabilities. The treatment is however invariably connected with profound hearing loss; therefore, careful planning and counselling of the patient are required. The tumor's size, evidence of tumor's growth in successive imaging, degree of hearing loss, intractable vestibular symptoms, patient´s concerns and other concurring medical conditions must be taken into consideration to tailor the therapy. Although hearing loss may initially still be amendable with hearing aids, in case of not serviceable hearing on the affected side, unbearable vestibular symptoms and acceptable surgical risk the indication for cochlear implantation is given. Indicatively all ICS are good candidates for cochlear implantation, although tumor removal may prove challenging. At the moment, there are four possible treatments for ICS.

d.1 Wait-test-scan

The traditional wait-test-and-scan approach with serial MRI rescanning every 12 months is advisable if the patient presents with minor otological complaints

and a good residual hearing (e.g. Word recognition score >60%). Should the situation remain unchanged, and no tumor growth be registered, the follow-up can be prolonged indefinitely. There is however clinical evidence that the tumor will eventually growth and become symptomatic [18,19], making this strategy time limited. As a symptom, hearing loss is tightly correlated to tumor growth.

d.2 Radiotherapy

Radiotherapy offers a treatment option for the comorbid patient who has a progressively enlarging schwannoma but may not be eligible for surgery. The aim of radiation therapy is to inhibit further tumor growth while preserving the surrounding structures. In the specific case of ICS experience with radiotherapy is still limited and anecdotal [28-30]. General principles can be derived from the radiotherapy for the more common internal auditory canal schwannomas. In IAC Schwannoma hearing preservation rates after fractioned radiotherapy (FRT) or stereotactic radiosurgery (SRS) range between 70-80% at two years and 55-75% at five years without statistical difference between the two methods [31-33], however recent protocol advancements and single dose reduction to 12 Gy seem to have better results [31]. Fractioned proton therapy (FPT) has been also applied with promising growth control results, although at the cost of more local complications and hearing loss (up to 42 % at one year) caused by the scattering of the proton bean through the mastoid structure[31,34,35]. It is however clear that hearing deterioration inexorably increases in the follow-up on a multifactorial

base, in particular because of post radiation sclerosis, continuous compression by the residual tumor mass and simple aging. Hearing rehabilitation via cochlear implantation is still a possible solution in post-irradiated patients [5,36,37]. In the specific case of ICS it remains difficult to achieve the recommended cochlear tolerance doses of approximately 5 Gy in SRS and 35 Gy in FSRT, given that the required therapeutic doses are 12 Gy in SRS and 50.5-54Gy in FSRT[31,32] and the hearing damage after radiation is indirectly proportional to the cochlea total irradiation[32] . Irrespective of the radiotherapy technology (SRS via gammaknife or cyberknife, FSRT via linear accelerator, Adron therapy), the risk of irreparably damaging the bodies of the spiral ganglion neurons is high and restrictions for later rehabilitation with CI have been reported [38]. Furthermore, there are anecdotical cases of malignant transformation of schwannoma after radiation, a sensible issue for young patients undergoing radiation therapy [39-41].

d.3 Cochlear implantation without tumor extirpation

A stiff electrode can be applied to directly perform a cochlear implantation without removal of the tumor. The surgical concept of addressing the hearing deficit with a CI and leaving the tumor in place was initially proposed for Neurofibromatosis Type 2 patients, as a recurrence or second tumor are almost granted. The reported cases are mostly transmodiolar tumors where a radical extirpation was not feasible without removing the modiolus itself, which is a

conceptually different case as compared with purely isolated ICS in our study. The insertion of a perimodiolar electrode employing an enlarged cochleostomy and a late stylet release technique in case of resistance proved successful, although tip fold-overs were registered. This approach was recently extended to patients with suspected ILS on imaging not willing to undergo concomitant tumor resection[42]. The reported hearing rehabilitation results at 6 and 12 months are satisfying, however it remains unclear if the electrode performance will remain stable in the long term or will eventually decrease due to displacement, loss of function and tumor growth[43].

d.4 Tumor Extirpation in combination with cochlear implantation

Finally, microsurgical extirpation with or without immediate hearing restoration with CI could be considered in the already functionally deaf patient. The operation itself invariantly leads to anacusis and various surgical technique have been described, ranging from enlarged cochleostomy to more radical cochleoectomy. Historically, a dummy electrode was inserted in the cochlear remnants after simple extirpation to prevent obliteration and serial MRI were performed in the follow-up to rule out recurrence before proceeding with cochlear implantation[44]. More recent studies advocate a simultaneous combined approach to minimize the risk of postoperative sclerosis.

f. Aims of the study

- Surgical feasibility of simultaneous tumor extirpation with cochlea implantation
- Comparison of the rehabilitation results with non-tumor cochlear implant patients
- Long term hearing results in implanted patients
- Evaluation of the use of the speech recognition tests in detecting tumor recurrence
- Indications, results, and limits of the microsurgical extirpation combined with cochlear implantation

g. Methods:

d.1 Study cohort

In order to study this rare disease, we enrolled all patients who underwent extirpation of ICS with simultaneous cochlea implantation at the Medizinische Hochschule Hannover between 2014 and 2021. The preoperative audiological and radiological finding together with the intraoperative findings and hearing results were retrospectively analyzed. All patients concluded their rehabilitation in the Deutsche Hörzentrum and were followed up for at least 36 months after surgery. The preoperative neurotological evaluation included clinical examination, pure tone audiometry, Freiburger monosyllabic word recognition

test at 65 dB (Fb65), promontorial test, caloric test, and dynamic posturography. All patients underwent a preoperative MRT Scan 1.5 Tesla and a high-resolution CT-scan or cone-beam CT of the temporal bone. We indicated surgery only in case of functional single-side deafness or unbearable tinnitus and vertigo; otherwise, a wait-test-and-scan approach was preferred.

d.2 The surgical technique

Surgery consisted in mastoidectomy with extended posterior tympanotomy with remotion of the incus buttress, followed by an enlarged cochleostomy if the tumor was limited to the basal turn, otherwise a second cochleostomy was performed in the middle turn and the tumor was extirpated with push through or pull-through techniques. A canalplasty was never required. After diagnosis was confirmed by intraoperative frozen section, the electrode of the cochlear implant was placed under direct visual control in the same surgical procedure (Fig 2, C-F). The cochleostomy was sealed with temporal muscle. Four weeks after surgery the cochlear implant was activated, and the patients underwent rehabilitation.

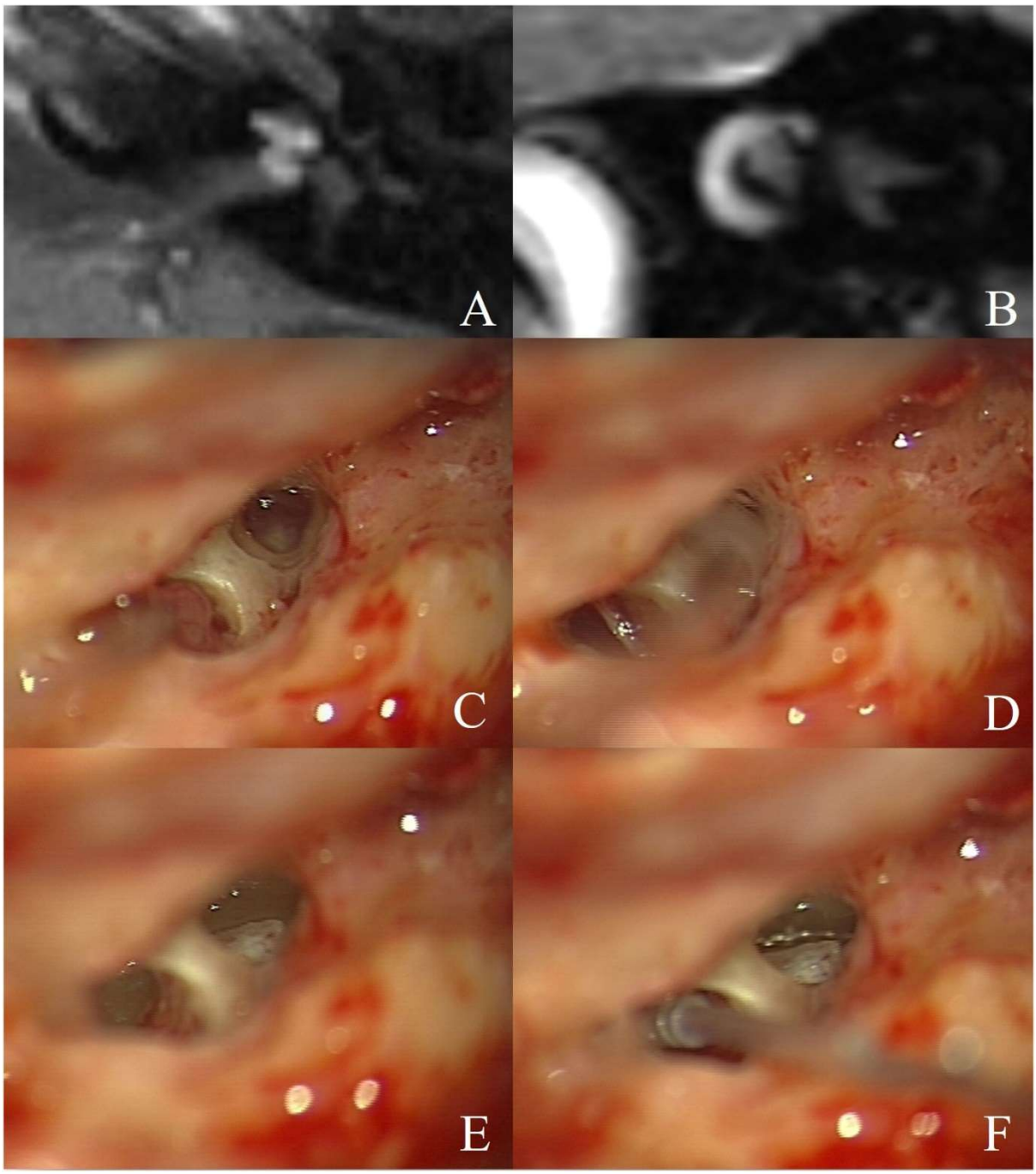

Fig 2. T1 MRI scan of ICS with involvement of basal and middle turn of the cochlea (A,B). Microsurgical extirpation via push-through (C) and pull-through (D) techniques via basal turn enlarged cochleostomy and middle turn cochleostomy through posterior tympanotomy. The skeletonized bony modiolus remains intact and the lateral wall is visualized (E). A perimodiolar electrode (Nucleus 612) is positioned under direct visualization (F).

d.3 The hearing evaluation

The hearing results were measured by means of speech recognition tests in German language, precisely Fb65 and the Hochmaier-Schulz-Moser Test in silence (HSM) and noise (HSMn, 10-decibel white noise) at 1, 6, 12, 24, 36 months after the operation. After descriptive statistics and normality test, we compared the values at different time-points by means of one-way ANOVA.

d.4 The comparison group

To evaluate the CI performance in ICS patients we compared their hearing results at 12 months (Fb65, HSM, HSMn) with a control group based on our historical single-side deafness cochlear implants cohort. The inclusion criteria for the control group were: having received cochlea implantation for SSD, having completed the rehabilitation at our institution between 2008 and 2015, complete audiological data at 12 months after activation and no evidence of vestibular schwannoma or neurofibromatosis. The hearing results of the two resulting groups were compared with a Unpaired T-Student test with Welch correction.

d.5 The statistical analysis

Summary statistics were collected, and normality tests, analysis of variance, t tests were performed with Graphpad Prism Version 9.0 (GraphPad Software, Inc., Boston, USA). Statistical significance was set at $p<0.05$.

d.6 Ethic committee protocol

The responsible ethics committee approved the protocol for using the patient's data for this retrospective study (Project identification code 1897-2013). All patient data were anonymized and de-identified prior to the retrospective analysis.

h. Results:

According to our inclusion criteria we collected 15 patients (seven males and eight females) with a purely monolateral intracochlear schwannoma who underwent a simultaneous cochlear implantation with tumor extirpation between 2014 and 2021.

The mean age at the time of diagnosis was 50.07 y (SD +/-14.15 y). Mean duration of deafness was 6.0 y (SD +/-3,65 y), ten patients reported compensated tinnitus and only two dizziness. The vestibular examination proved normal in all cases. In 9 cases the tumor was limited to the basal turn in MRT scan, in 5 cases there was involvement of both basal and middle turn, and one patient presented a tumor confined in the middle and apical turn. An enlarged cochleostomy was performed in 9 cases, whilst a second cochleostomy in the middle turn was necessary in all other cases. The diagnosis was always confirmed by the pathological examination and the radicality of extirpation was directly ascertained intraoperatively. For the electrode selection, we opted for a

Cochlear Nucleus 512 in 9 cases, Cochlear Nucleus 612 in 2, Medel Flex 28 in 3, AB HiRes Ultra Hifokus mid scala in one. The surgery was uneventful, and auditory nerve compound action potential could be registered in all cases. No postoperative complication was registered.

All patients started their rehabilitation 4 weeks after surgery. The monosyllable word recognition test (FB65) showed a statistically significant improvement between first fitting and after 12 months of use (one way ANOVA, p=0,004). Although there were patient specific variations between the successive measurements, we found no statistical difference between the results at one year and those at 24 months (p=0.81) and 36 months (p=0.41). The mean Fb65 at 12 months was 55.33% (SD +/-20.39). For the HSMq and HSMn there was also a steady improvement between the start of the rehabilitation and 12 months, afterwards the results remained stable. The HSMq assessed 74.09% (SD +/-25.09) at 12 months, the HSMn 43.02% (SD +/-27.56). There was no statistically significant difference between the speech recognition results of perimodiolar electrode patients and the lateral wall ones, although the small number of subjects makes this assumption questionable. Electrical impedances remained stable over the observation period. As the tumor recurrence is supposed to dislocate the electrodes causing a decrease in hearing performance, stable hearing rehabilitation results over the long-term indirectly confirmed the radicality of the resection and the absence of a clinically relevant recurrence.

We did not experience patient who were not using their cochlear implant during the observation time (so called non-users).

One patient, implanted with an Advanced Bionics HiRes Ultra Hifocus Midscala in winter 2016, suffered after three years of use an unexplained drop of performance in both FB65 (35% to 20%) and HSMq (78.5% to 40.57%) HSMn (37.3% to 0%) with persistent dizziness, arising suspicion for recurrent disease or implant malfunction. The magnet was removed, an MRI scan proved inconclusive and revision surgery was performed. The cochleostomy was reopened and the intraoperative biopsy showed no evidence of spindle cells pathognomonic for Schwannomas, ruling out recurrence. An implant hard failure was detected. Reimplantation was performed with a Nucleus 612 and open speech recognition was once again achieved after one year, thus demonstrating the feasibility of revision surgery and reimplantation with the adopted surgical technique.

When compared to the MHH historical cohort of postlingually deafened non tumor patients[15], 6 ICS patients categorized as good performer (FB65 >65%), 6 as average performer (FB65> 35%) and 3 as poor performers (FB65<35%) after one year of use. It is worth considering that in our study ICS patients classify as single-side deafness ones. In SSD the hearing rehabilitation results are usually inferior as compared with bilateral postlingual deaf patients due to various

reasons, such as decrease acceptance of the cochlear implant, and conflict with normal hearing ear [15]. As a consequence, we compared the CI performance of ICS patients with a control group based on our historical cohort of SSD patients from 2008 to 2015. 52 patients, (21 males, 31females) respected the inclusion criteria. The mean age at implantation was 50.02 y (SD +/-11.21), the mean duration of SSD before implantation was 5 y (SD +/-7,04). The mean Fb65 in 1 year was 55.33% (SD+/- 20.13), the mean HSMq 74.09% (SD +/-25.09) and the mean HSMn 43.02% (SD +/-27.56). Although the ICS patients performed generally better, there was no statistically relevant difference between them and non-tumor SSD ones in Fb65 ($p=0.21$) and HSMq ($p=0.64$). A significant difference was registered for the HSMn ($p=0.01$), where the ICS group performed better (Fig.3). Thus, we demonstrated that the hearing rehabilitation results in case of monolateral intracochlear schwannoma are not inferior to those of non-tumor patients. Moreover, the surgical extirpation does not reduce the rehabilitation potential of the cochlear implantation, making the combination of the two procedures in one surgical time a valid therapy for these rare tumors.

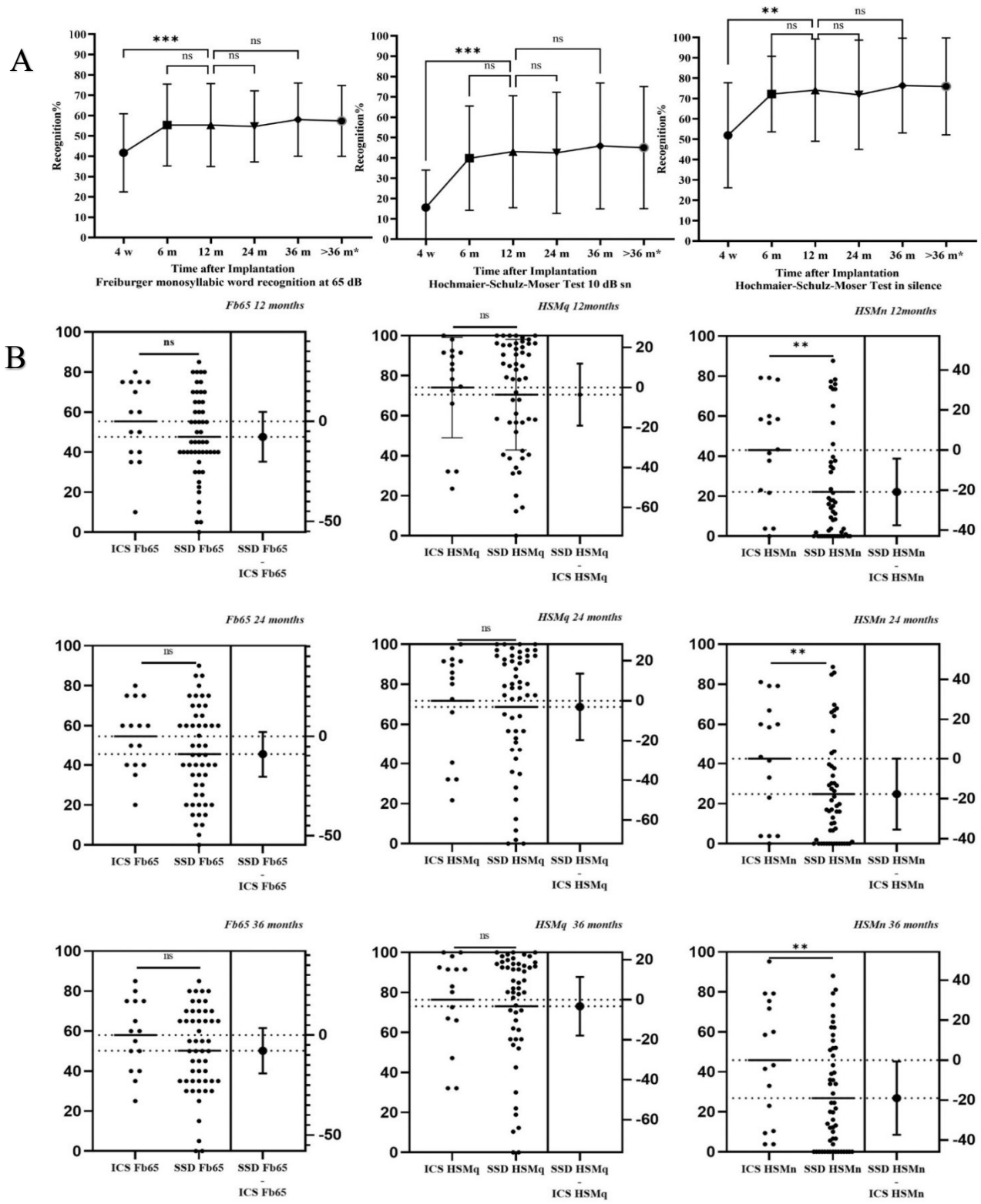

Fig 3 A) Speech recognitions scores (Fb65, HSMq, and HSMn) of the 15 ICS patients after surgery. >36*: not all the 15 patients were observed after 36 months.

B) Speech recognition scores (Fb65, HSMq, and HSMn) comparison at 12, 24, and 36 months between ICS and SSD patients. At all three time points, there was no statistically relevant difference between the two groups in Fb65 and HSMq. A significant difference was registered for the HSMn ($p < 0.05$).

3. Publication with reference number

Title of the publication:

Long-term hearing outcome of cochlear Implantation in cases of simultaneous intracochlear schwannoma resection

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The Laryngoscope

Long-Term Hearing Outcome of Cochlear Implantation in Cases with Simultaneous Intracochlear Schwannoma Resection

Riccardo Di Micco, MD; Rolf Salcher, MD; Anke Lesinski-Schiedat, MD; Thomas Lenarz, MD

Objectives: The aim was to analyze the long-term hearing results after simultaneous microsurgical extirpation via enlarged cochleostomy and cochlear implantation in intracochlear schwannoma as compared with non-tumor single-side deafness patients.

Methods: Microsurgical extirpation via enlarged cochleostomy with simultaneous cochlear implantation was performed in 15 cases of intracochlear schwannoma between 2014 and 2021. Speech recognition tests in German language and impedance performances were collected over 36 months of observation and compared with an internal cohort of 52 age matched non-tumor single-side deafness patients. Retrospective cohort study in a tertiary referral center.

Results: The surgery proved feasible and uneventful in all cases. In the case of intracochlear schwannoma, the hearing rehabilitation results were highly satisfactory and comparable to those of the non-tumor single-side deafness cohort. The speech recognition performance improved steadily in the first 12 months; afterward, it remained stable, providing indirect evidence against tumor recurrence during the follow-up. One patient required implant revision surgery related to device failure, but no recurrence was registered in the 36 months of observation.

Conclusions: Cochlear implantation is the strategy of choice for hearing rehabilitation in case of intracochlear schwannomas in the long term. In particular, the combination of tumor extirpation via cochleostomy with a cochlear implantation in the same surgical time offers a viable therapy for intracochlear schwannoma, granting a sufficient degree of radicality without compromising the cochlear integrity. This technique allows for revision surgery if required.

Key Words: cochlear implantation, intracochlear schwannoma, otology-neurotology, sensorineural hearing loss, vestibular schwannoma.

Level of Evidence: 4

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INTRODUCTION

Once a rare finding during cochlear implantation surgery or autopsy, incidence of intralabyrinthine schwannomas (ILS) has increased thanks to advances in diagnostic evaluation using high-resolution MRI.[1–5] ILSs originate from the Schwann cells interspersed in the neural tissue of the vestibule, cochlea, and semicircular canals and are classified based on the affected inner-ear structures.[6,7] They represent up to 10% of all vestibular schwannomas, and in roughly 50% of cases, they are completely intracochlear.[6] There is epidemiological evidence that ILS are more common than previously thought.[8] These benign tumors pose a special clinical and therapeutical challenge, as their natural history and surgical removal invariably lead to anacusis. The feasibility of extirpation followed by or combined with successful cochlear implantation has been demonstrated[5,9]; however, the risk of recurrence in the follow-up remains unclear. The aim of the current retrospective study is to analyze the hearing performance of intracochlear schwannoma (ICS) patients treated with simultaneous tumor extirpation via enlarged cochleostomy and cochlear implantation in the long term (36 months) compared with non-tumor single-side deafness cochlear implant patients, evaluating the use of the speech recognition tests in suspected tumor recurrence.

METHODS

We enrolled patients who underwent extirpation of ICS with simultaneous cochlea implantation in our institution between 2014 and 2021. The preoperative audiological and radiological finding together with the intraoperative findings and hearing results were retrospectively analyzed. All patients concluded their rehabilitation in our institution and were followed up for at least 36 months after surgery. The preoperative neurotological evaluation included clinical examination, pure tone audiometry, Freiburger monosyllabic word recognition test at 65 dB (Fb65), promontorial test, caloric test, and Equi-test. All the patients underwent a preoperative MRT Scan 1.5 Tesla (Fig. 1A,B) and a high-resolution CT scan or cone-beam CT of the temporal bone. We indicated surgery only in case of functional single-side deafness or unbearable tinnitus and vertigo; otherwise, a wait-test-and-scan approach was preferred.

From the Department of Otorhinolaryngology (R.D.M., R.S., A.L.-S., T. L.), Medizinische Hochschule Hannover, Hannover, Germany.

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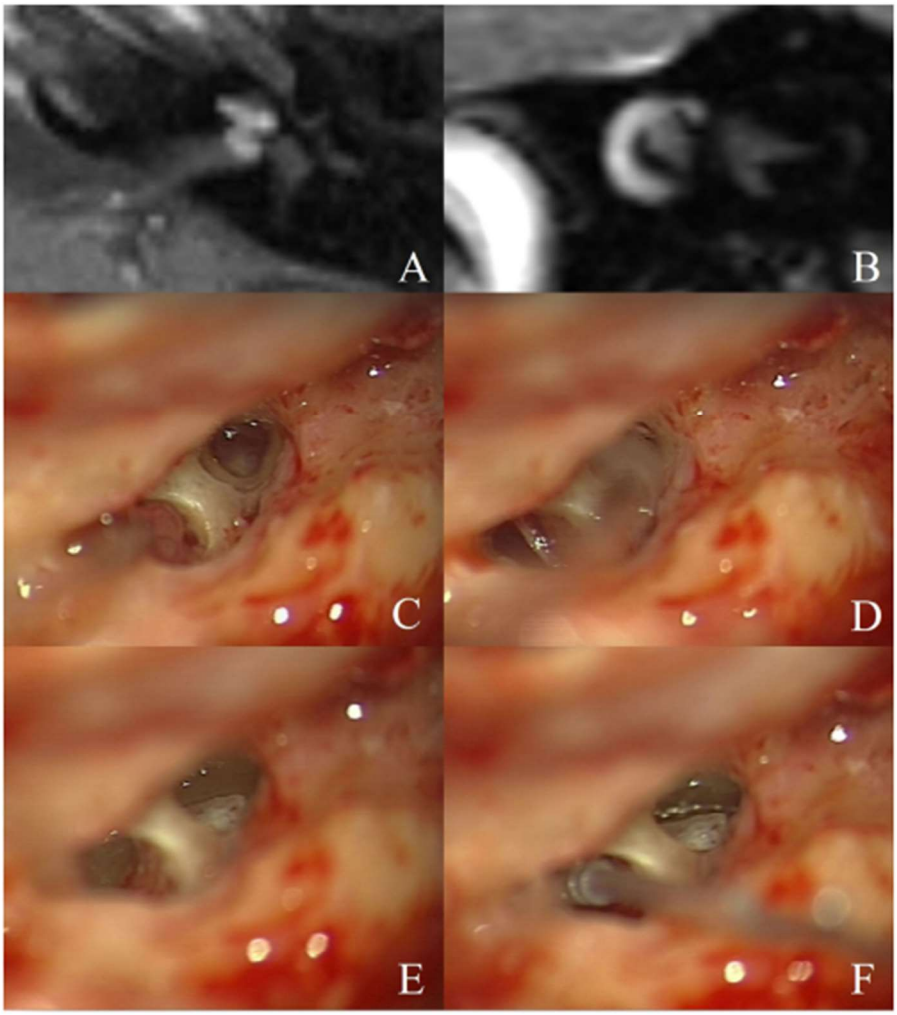

Fig. 1. T1 MRI scan of ICS with involvement of basal and middle turn of the cochlea (A,B). Microsurgical extirpation via push-through (C) and pull-through (D) techniques via basal turn enlarged cochleostomy and middle turn cochleostomy through posterior tympanotomy. The skeletonized bony modiolus remains intact and the lateral wall is visualized (E). A perimodiolar electrode (Nucleus 612) is positioned under direct visualization (F). [Color figure can be viewed in the online issue, which is available at www.laryngoscope.com.]

Surgery consisted in mastoidectomy with extended posterior tympanotomy with remotion of the incus buttress, followed by an enlarged cochleostomy if the tumor was limited to the basal turn; otherwise, a second cochleostomy was performed in the middle turn, and the tumor was extirpated with push-through or pull-through techniques. A canalplasty was never required. After diagnosis was confirmed by intraoperative frozen section, the electrode of the cochlear implant was placed under direct visual control in the same surgical time (Fig. 1C–F). The cochleostomy was sealed with temporal muscle. Four weeks after surgery the cochlear implant was activated, and the patients underwent rehabilitation. The hearing results were measured by means of speech recognition tests in German language, precisely Fb65 and the Hochmaier–Schulz–Moser (HSM) Test in silence and noise (HSMn) at 1, 6, 12, 24, 36 months after the operation.

To evaluate the CI performance in ICS patients, we compared their hearing results at 12 months (Fb65, HSM, and HSMn) with a control group based on our historical single-side deafness cochlear implants cohort. The inclusion criteria for the control group were having received cochlea implantation for SSD, having completed the rehabilitation at our institution between 2008 and 2015, complete audiological data at 12 months after activation, and no evidence of vestibular schwannoma or neurofibromatosis.

Summary statistics were collected, and normality tests, analysis of variance, and t tests were performed with GraphPad Prism Version 9.0 (GraphPad Software, Inc., Boston, USA). Statistical significance was set at $p < 0.05$. The responsible ethics committee approved the protocol for using the patient's data for this retrospective study (project identification code 1897–2013). All the patient data were anonymized and de-identified prior to the retrospective analysis.

RESULTS

Between 2014 and 2021, we identified 21 patients with ILS with already established functional unilateral deafness. Of them, 17 presented with a purely intracochlear tumor

and 15 (7 M, 8F) underwent a simultaneous cochlear implantation with tumor extirpation. The mean age at the time of diagnosis was 50.07 y (SD ± 14.15 y). All the patients complained of deafness on the affected side for a mean of 6.0 y (SD ± 3.65 y); ten patients reported compensated tinnitus and only two dizziness. The vestibular examination proved normal in all cases. In nine cases, the tumor was limited to the basal turn in the MRT scan, in 5 cases there was involvement of both basal and middle turn, and one patient presented a tumor confined in the middle and apical turn. An enlarged cochleostomy was performed in 9 cases, whilst a second cochleostomy in the middle turn was necessary in all other cases. For the electrode selection we opted for a Cochlear Nucleus 512 in 9 cases, Medel Flex 28 in 3, Cochlear Nucleus 612 in 2, AB HiRes Ultra Hifokus midscale in one. The surgery was uneventful, and all implants proved functional during the implantation with positive answers in the electrocochleography. No postoperative complication was registered.

All the patients started their rehabilitation 4 weeks after surgery. The monosyllable word recognition test (FB65) showed a statistically significant improvement between implant activation and after 12 months of use (one way ANOVA, $p = 0.004$). Although there were patient specific variations between the successive measurements, we found no statistical difference between the results at one year and those at 24 months ($p = 0.81$) and 36 months ($p = 0.41$). The mean Fb65 at 12 months was 55.33% (SD ± 20.39). For the HSMq and HSMn, there was also a steady improvement between the start of the rehabilitation and 12 months; afterward, the results remained statistically constant. The HSMq assessed 74.09% (SD ± 25.09) at 12 months, the HSMn 43.02% (SD ± 27.56) (Fig. 2). There was no statistically significant difference between the speech recognition results of perimodiolar electrode patients and the lateral wall ones. Electrical impedances remained stable over the observation period. We did not experience nonusers.

One patient, implanted with an AB HiRes Ultra Hifokus Midscala in winter 2016, suffered after three years of use an unexplained drop of performance in both FB65 (35% to 20%) and HSMq (78.5% to 40.57%) HSMn (37.3% to 0%) with persistent dizziness, arising suspicion for recurrent disease or implant hard failure. The magnet was removed, an MRI scan proved inconclusive, and revision surgery was performed. The cochleostomy was reopened and the intraoperative biopsy showed no evidence of spindle cells, ruling out recurrence. An implant hard failure was detected. The implant was substituted with a Nucleus 612 and open speech recognition was once again achieved after one year.

When compared with our historical cohort of postlingually deafened non-tumor patients,[10] six ICS patients categorized as good performer (FB65bB >65%), six as average performer (FB65 > 35%), and three as poor performers (FB65 < 35%) after one year of use. To evaluate the special issue of SSD, we compared the CI performance of ICS patients with a control group based on our historical cohort of SSD patients from 2008 to 2015. Fifty-two patients (21 M, 31 F) respected the inclusion criteria. The mean age at implantation was 50.02 y (SD ± 11.21), the mean duration of SSD before implantation was 5 y (SD ± 7,04). The mean Fb65 at 1 year was 55.33% (SD± 20.13), the mean HSMq was 74.09% (SD ± 25.09), and the mean HSMn was 43.02% (SD ± 27.56). Although the ICS patients performed generally better, there was no statistically relevant difference between them and non-tumor SSD ones in Fb65 ($p = 0.21$) and HSMq ($p = 0.64$). A significant difference was registered for the HSMn ($p = 0.01$) (Fig. 3). The ICS cohort proved not inferior to the non-tumor one.

DISCUSSION

ICSs arise from the Schwann's cells of the cochlear and vestibular nerve axons interspersed in the osseous spiral lamina and modiolus, proximal to the spiral ganglion.[11,12] ICSs progressively occupy the scala media, invade the modiolus, erode the cribriform area of the IAC, and then spread into the vestibule. The clinical presentation of ICSs is characterized by sensorineural hearing loss (94.5%), together with Tinnitus (69.1% to 95.8%),

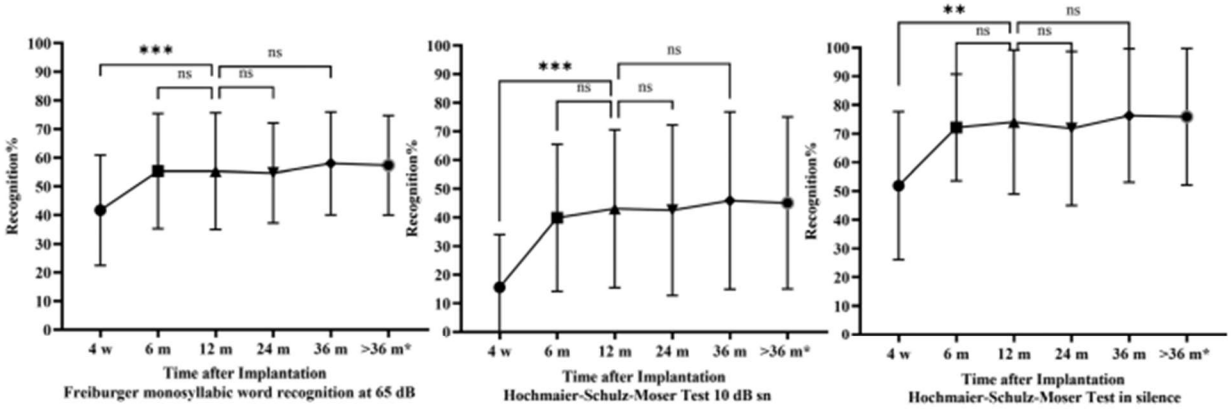

Fig. 2. Speech recognitions scores (Fb65, HSMq, and HSMn) of the 15 ICS patients after surgery. >36*: not all the 15 patients were observed after 36 months.

vestibular symptoms (59.1%), and aural fullness.[13] The natural history of the disease is progressive, eventually leading to severe pantonal SNHL with poor speech perception. If invasion and compression of the vestibule is present, the tumors may cause endolymphatic hydrops, resulting in disequilibrium and vertigo. Due to the small size of the lesions, slow growth rate, and easily compensated symptoms, there is a large delay between emergence of the symptoms and diagnosis, averaging 72.5 months.[14] Usually, the patient has already lost his serviceable hearing by the time of the first diagnosis; however, the time to functional deafness remains unpredictable.

ICSs are detected with high-resolution MRI during diagnostic workup for SNHL. ICSs are enhanced on T1-weighted images with gadolinium with sharply

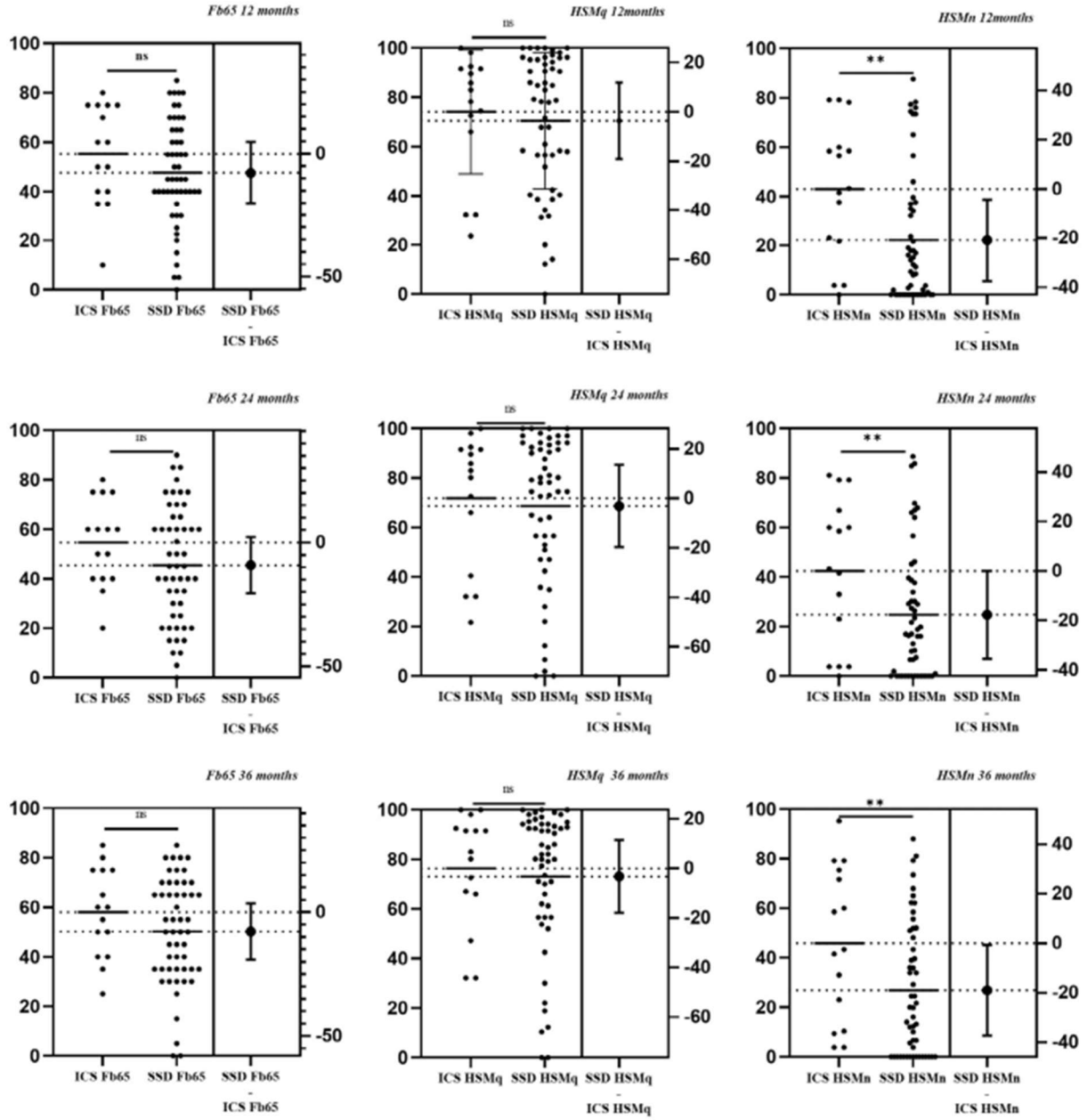

Fig. 3. Speech recognition scores (Fb65, HSMq, and HSMn) comparison at 12, 24, and 36 months between ICS and SSD patients. At all three time points, there was no statistically relevant difference between the two groups in Fb65 and HSMq. A significant difference was registered for the HSMn ($p < 0.05$).

delineated edges and show a delineated signal loss in the high-intensity intralabyrinthine fluid on T2-weighted images. These MRI findings mimic infectious or inflammatory processes in the labyrinth, making labyrinthitis the principal differential diagnosis.[11,15,16] Once diagnosed, evidence of tumor's growth in successive imaging, degree of hearing loss, intractable vestibular symptoms, patient's concerns, and other concurring medical conditions must be taken into consideration to tailor the therapy. Regarding hearing rehabilitation, hearing loss may initially still be amendable with hearing aids, but in case of not serviceable hearing on the affected side and acceptable surgical risk, the indication for cochlear implantation is given. Indicatively all ICS are good candidates for cochlear implantation, although tumor removal may prove challenging.

The traditional wait-test-and-scan approach with serial MRI rescanning every 12 months is advisable if the patient presents with minor otological complaints and good residual hearing. Should the situation remain unchanged, and no tumor growth be registered, the follow-up can be prolonged indefinitely. There is however clinical evidence that the tumor will eventually growth and become symptomatic,[11,12] making this strategy time-limited.

Radiotherapy offers a treatment option for the ICS comorbid patient who has a progressively enlarging tumor but may not be eligible for surgery. In the specific case of ICS, experience with radiotherapy is still limited[17-19] and it remains difficult to respect the recommended cochlear tolerance doses of approximately 5 Gy in stereotactic radiosurgery (SRS) and 35 Gy in fractioned radiotherapy (FSRT), given that the required therapeutic doses are 12 Gy in SRS and 50.5-54Gy in FSRT.[20,21] It is however clear that the hearing damage after radiation is indirectly proportional to the cochlea total irradiation[21,22] and hearing deterioration increases in the follow-up on a multifactorial base. Irrespective of the radiotherapy technology applied, the risk of irreparably damaging the bodies of the spiral ganglion neurons is high,[9] making future rehabilitation via cochlear implantation challenging.[23-25]

Alternatively, a stiff electrode can be applied to directly perform a cochlear implantation without removal of the tumor. This technique was initially developed to treat mostly transmodiolar schwannomas in Neurofibromatosis Type 2 patients were a radical extirpation was not feasible without removing the modiolus itself.[26] The insertion of a perimodiolar electrode employing an enlarged cochleostomy and a late stylet release in case of resistance proved successful, although tip fold overs were registered. The approach has been recently applied to patients with suspected ILS on imaging not willing to undergo concomitant tumor resection.[27] The hearing rehabilitation results at 6 and 12 months are satisfactory; however, it remains unclear if the electrode performance will remain stable in the long term or will eventually decrease due to displacement, loss of function, and tumor growth.[28]

Finally, microsurgical extirpation with or without immediate hearing restoration with CI could be considered in the already functionally deaf ICS patients. Historically, a dummy electrode was inserted in the cochlear remnants after simple extirpation to prevent obliteration and serial MRI were performed in the follow-up to rule out recurrence before proceeding with cochlear implantation.[29] In our experience, this approach proves unpractical as the patients tend to drop out of the follow-up or refuse the second operation. Furthermore, the feasibility of the implantation rapidly declines due to progressive sclerosis of the cochlear and nerve remnants. In case of ICSs, the affected turn of the cochlea, the involvement of the anterior or posterior side of the modiolus, and the size of the tumor determine the surgery of choice. We adopted an extended cochleostomy technique, as this surgical approach allows a good visualization of the tumor, guarantees acceptable radicality, preserves the cochlea structure, and does not endanger the bony modiolus. The more aggressive cochleoectomy, requiring a total drill out of the modiolus followed by direct placement of the electrode on the skeletonized remnant and sealing with a cartilage perichondrium transplant, may be adopted to further medially expose the tumor and to achieve maximal radicality.[9,30,31] This technique involves extensive exposure of the entire cochlear anatomy, with risk of cerebrospinal fluid leakage and damage of the spiral ganglion itself.[30-32] In case of transmodiolar extension only incomplete extirpation is possible.[33]

ICSs show good hearing rehabilitation results with CI, comparable with the non-tumor postlingually single-side deafened cochlear implant patients of the same age.[10,34,35] The reasons behind such a good hearing performance remain poorly understood. A possible answer is good fitting of the electrode inside the cochlea and the reduced spread of electric field after partial cochleoectomy [36]; however, even in case of CI without tumor removal, the results are above average.[26] Newly established SNHL responds more favorably to cochlear implantation; however, ICS are usually identified in already long-time established deafness. As ICSs develop directly inside the cochlea, the damage is limited to the sensory part of the inner ear, disrupting the architecture of the Corti's organ but sparing the long axons of the cochleovestibular nerve. There is histological evidence that ICS initially involves Type A 1 bipolar neurons only in their myelinated osseous spiral lamina part,[37,38] thus preserving the neuron body in the modiolus, without subsequent damage to the neural conduction potential. Furthermore, the extirpation of ICS does not involve the nerve itself, avoiding major axon lesions; consequently, the nerve can continue unhindered to transport the information provided by the electrical stimulation of the inserted cochlear implant.[39] Microscopic involvement of the modiolus remains difficult to demonstrate in the clinic and is unclear if the unsatisfactory results of CIs by ICS are related to it.

All things considered, the hearing rehabilitation after simultaneous tumor resection and placement of a CI in ICS shows solid results despite large opening of the cochlea. The challenges include radicality of the resection,[40] long-term tumor control and follow-up.

The recurrence rate in internal auditory canal schwannomas depends on follow-up time and completeness of the tumor removal[41]; however, there are no available data for ICSs. ICSs are not always easy to resect; nevertheless, it is essential to spare the modiolus, even if infiltrated, as its partial removal greatly jeopardizes the CI performance.[42] Counting on the slow growth rate of the potential residual tumor, a subtotal resection preserving the cochlear anatomy should be considered in favor of better chances for long-term hearing rehabilitation with a CI, without however completely sacrificing disease control. Trying to preserve the cochlear anatomy during tumor extirpation may prove even more advantageous in the long term, as it allows for effective surgical revision in case of reimplantation. On the contrary, an aggressive resection may put at risk the hearing restoration without any significant clinical gain. In case of unresectability, the cochlear implantation alone remains a worthy therapy in selected cases.[26]

On the clinical side, the follow-up is based on serial MRI scans and the overall performance of the cochlear implant. MRI-compatible cochlear implants allow the visualization of the modiolus[26]; nevertheless, imaging is not always sufficient alone to rule out recurrence due to artifacts. On the other hand, changes in the electrophysiological measurements of the CI (impedance, eABR, and eCAP performance) can indirectly indicate tumor regrowth,[28,43] as the tumor mass should reduce the performance of the cochlear implant by electrode interference or progressive damage of the spiral ganglion. It remains however challenging to differentiate tumor regrowth from fibrosis and ossification. In our clinical experience, word recognition scores peaked at 12 months after activation and remained stable during the 36 months of observation, providing indirect evidence of absence of recurrence of the disease. An unexpected decrease in performance should prompt a thorough diagnostic workup to detect a possible recurrence or implant disfunction, possibly leading to revision and reimplantation. Thus, the yearly evaluation of impedance and hearing performances acquires even more importance and should be indefinitely pursued.

CONCLUSIONS

Our long-term observations show that in case of ICS, simultaneous tumor extirpation and cochlear implantation is not only a feasible surgical intervention, but also a valid hearing rehabilitation strategy, providing satisfactory hearing results and no evidence of clinically significant recurrence in the long term. Further research is required to properly identify preoperative prognostic factors to select those patients who will profit the most from the CI rehabilitation, to define the role of residual tumor in the long term, and to effectively compare the different therapeutical and surgical approaches already developed.

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4. Discussion

a. Microsurgery of intracochlear schwannomas

The affected turn of the cochlea, the involvement of the anterior or posterior side of the modiolus and the size of the tumor determine the surgery of choice. If only the basal turn is affected, an extended round window approach via posterior tympanotomy is feasible and in most cases sufficient, otherwise an extended cochleostomy with resection of the promontory and part of the cochlear wall may be used. In case of extension into the middle or apical turn, a second cochleostomy offers a good alternative to the total drill-out of the cochlea. Once safely exposed the tumor is removed through push-through or pull-through techniques, afterwards the electrode of the cochlear implant can be placed under direct visual control. The operation may be completely performed through a posterior tympanotomy, and the plugging of the damaged structure can be performed with fascia tissue or temporal muscle. This surgical approach allows a good visualization of the tumor, preserves most of the cochlea structure and does not endanger the bony modiolus. On the basis of this consideration, we adopted this procedure in our case series. A more radical approach, the cochleoectomy, has been proposed to expose the tumor also in its medial aspects to achieve maximal radicality [38,45,46]. For apically located tumors, a partial cochleoectomy may be performed with resection of the second turn and apex of the cochlea, in case of basal and middle turn involvement a total

cochleoectomy requires drilling out of the entire modiolus but in its basal turn. The electrode of the CI is then placed directly on the modiolus remnant, and everything is sealed with a cartilage perichondrium transplant. This technique requires a transcanal approach to visualize the cochlea and involve alteration of the entire cochlear anatomy, with risk of cerebrospinal fluid leakage and damage of the spiral ganglion itself, although the actual ganglional damage may be in practice limited[45-47]. Recently an endoscope assisted variant of this technique has been described[48]. In case of transmodiolar extension, only incomplete extirpation is possible [49]. Should the tumor extends into the vestibulum of IAC or present as ILS a classical open [50]or endoscopic transcanal transpromontorial endoscopic approach labyrinthectomy [51] must be considered

b. Cochlea Implantation by Intracochlear Schwannoma: Technical issues and follow-up

Historically, a dummy electrode was inserted in the cochlear remnants after simple extirpation to prevent obliteration and serial MRI were performed in the follow-up to rule out recurrence before proceeding with cochlear implantation[44]. In our experience, this approach proves unpractical, as the patients tend to drop out of the follow-up or refuse the second operation. Furthermore, the feasibility of the implantation rapidly declines due to progressive sclerosis of the cochlear and nerve remnants. As the tumors are known for their extremely slow growth

rate and their extension is mostly limited to accessible areas of the cochlea, a combined approach is preferable.

A perimodiolar electrode has usually been preferred in order to minimize the distance between the surviving spiraliform ganglion and the electrical impulse and improve the electrical performance of the implant, as preservation of the inner ear structure is not always completely possible. The use of a stylet also helps during the insertion in case of residual tumor. Preformed, custom-made electrode could be beneficial in some cases of complete cochleoectomy [52].

All things considered, the hearing rehabilitation after simultaneous tumor resection and placement of a CI in ICS shows solid results despite large opening of the cochlea. The challenges include radicality of the resection[48], long term tumor control and follow-up. The recurrence rate in internal auditory canal schwannomas depends on follow-up time and completeness of the tumor removal[53], however there are no available data for ICSs. ICSs are not always easy to resect, nevertheless it is essential to spare the modiolus, even if infiltrated, as its partial removal greatly jeopardize the CI performance[54]. Counting on the slow growth rate of the potential residual tumor, a subtotal resection preserving the cochlear anatomy should be considered in favor of better chances for long-term hearing rehabilitation with a CI, without however completely sacrificing disease control. According to our experience with a

suspected recurrence and a hard failure of the cochlear implant, trying to preserve the cochlear anatomy during tumor extirpation may prove even more advantageous in the long term, as it allows for effective surgical revision. On the contrary, an aggressive resection may put at risk the hearing restoration without any significant clinical gain. In case of unresectability the cochlear implantation alone remain a worthy therapy in selected cases[55].

On the clinical side, the follow-up is based on serial MRI scans and the overall performance of the cochlear implant. MRI-compatible cochlear implants allow the visualization of the modiolus[55], nevertheless imaging is not always sufficient alone to rule out recurrence due to artefacts. On the other hand, changes in the electrophysiological measurements of the CI (impedance, eABR, eCAP performance) can indirectly indicate tumor regrowth [43,50], as the tumor mass should reduce the performance of the cochlear implant by electrode interference or progressive damage of the spiral ganglion. It remains, however, challenging to differentiate tumor regrowth from fibrosis and ossification. In our clinical experience, word recognition scores peaked at 12 months after activation and remained stable during the 36 months of observation, providing indirect evidence of absence of recurrence of the disease. An unexpected decrease in performance should prompt a thorough diagnostic work-up to detect a possible recurrence or implant disfunction, possibly leading to revision and reimplantation. Thus, the yearly evaluation of impedance and hearing

performances acquires even more importance and should be indefinitely pursued.

c. Cochlear Implantation results in intracochlear Schwannomas

ICSs show good hearing rehabilitation results with CI, comparable to the non-tumor postlingually single-side deafened cochlear implant patients of the same age [15,56,57]. The reasons behind such a good hearing performance remain poorly understood. A possible answer is good fitting of the electrode inside the cochlea and the reduced spread of electric field after partial cochleoectomy[58], however even in case of CI without tumor removal the results are above average[55]. Newly established SNHL responds more favorably to cochlear implantation, however ICS are usually identified in already long-time established deafness. A possible explanation is the special anatomical location of the tumor (Fig.4).

As ICSs develop directly inside the cochlea, the damage is limited to the sensory part of the inner ear, disrupting the architecture of the Corti's organ but sparing the long axons of the cochleovestibular nerve (Fig.5).

There is histological evidence that ICS initially involves Type A 1 bipolar neurons only in their myelinated osseous spiral lamina part [59,60], thus preserving the neuron body in the modiolus, without subsequent damage to the neural conduction potential. Furthermore, the extirpation of ICS does not involve the

nerve itself as in classical vestibular schwannomas, avoiding major axon lesions, consequently the nerve can continue unhindered to transport the information provided by the electrical stimulation of the inserted cochlear implant [61]. Microscopic involvement of the modiolus remains difficult to demonstrate in the clinic and is unclear if the unsatisfactory results of CIs by ICS are related to it.

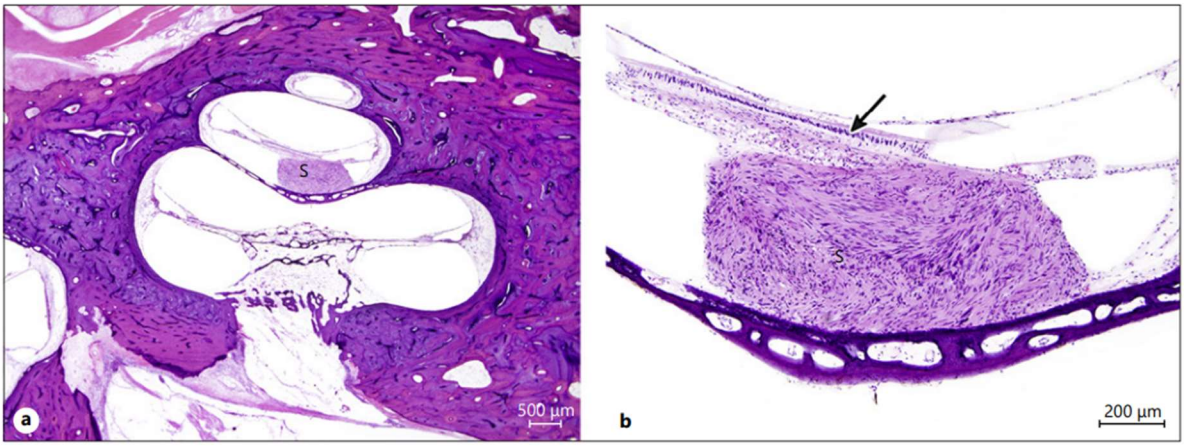

Fig. 4: Histological section of an intracochlear Schwannoma, note the completely intracochlear extension with destruction of the Corti´s organ with spiral ganglion sparring.
Courtesy of *Bagattini M, Quesnel AM, Röösli C. Histopathologic Evaluation of Intralabyrinthine Schwannoma. Audiol Neurootol. 2021*

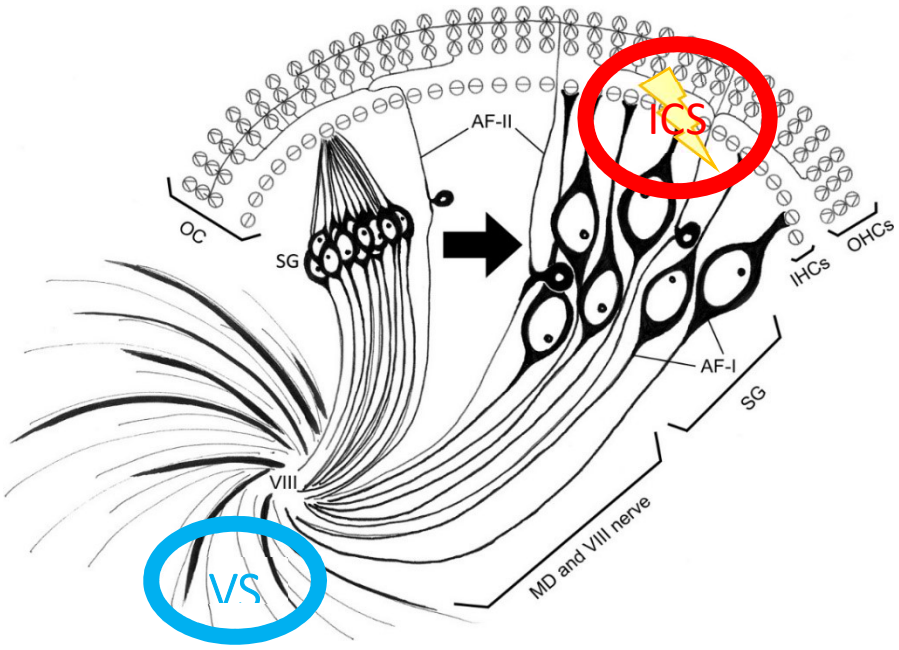

Fig. 5. Schematic section of spiral ganglion with the AF-II neurons, unmyelinated and connected with the outer hair cells (OHC) and the bipolar afferent neurons AF-I, myelinated and connected to the inner hair cells, responsible for the hearing sensation. The intracochlear Schwannomas (ICS) arise from the Schwann cells surrounding the short afferent axons, resulting in less damage in the acoustic information transport, or at least a damage easily overcome with a cochlear implantation. On the contrary the classical schwannoma (VS) invariantly damages the long efferent axons,
Modified from *The Anatomical Record, Volume: 302, Issue: 3, Pages: 463-471, First published: 16 April 2018, DOI: (10.1002/ar.23815)*

5. Conclusions

- Our long-term observations show that in case of ICS simultaneous tumor extirpation and cochlear implantation is not only a feasible surgical intervention, but also a valid hearing rehabilitation strategy, providing satisfactory hearing results and no evidence of clinically significant recurrence in the long term. Radiotherapy should be considered only in patients who do not qualify for surgery, while the wait-test-and-scan approach conserves its value in case of sufficient residual hearing.
- When compared to non-tumor single-side deaf patients the CI rehabilitation results in ICS patients are not inferior, on the contrary these patients usually perform surprisingly well. The reasons for such a satisfactory performance and the correlated prognostic factors remain unclear. A possible answer is the proximity of the electrode to the modiolus or the alteration of the cochlear anatomy after the operation.
- Even in the long term, the hearing results in implanted patients remains satisfactory and stable, indirectly demonstrating the absence of a clinically relevant recurrence after tumor resection.
- As cochlear implantation results in reduced sensitivity of the traditional MRI screening to detect tumor recurrence in the follow-up, regular evaluation of the speech recognition tests may be valid substitute. An unexpected decrease in performance should prompt a thorough

diagnostic work-up to detect a possible recurrence or implant disfunction, possibly leading to revision and reimplantation. Thus, the yearly evaluation of impedance and hearing performances acquires even more importance and should be indefinitely pursued.

- Although various surgical techniques have been proposed to effectively remove ICS, in our experience enlarged cochleostomy combined with simultaneous cochlear implantation proved secure, efficient, and highly performant. This approach allows for revision surgery and reimplantation if needed. In case of extracochlear and transmodiolar extension partial resection or more traditional labyrinthectomy should be considered.
- Further research is required to properly identify preoperative prognostic factors to select those patients who will profit the most from the CI rehabilitation, to define the role of residual tumor in the long term and to effectively compare the different therapeutical and surgical approaches already developed. Due to recent advances, a standardized differentiated approach for this once rare pathology is already possible.

6. Abbreviations:

1. ICS: Intracochlear Schwannoma
2. ILS: Intralabyrinthine Schwannoma
3. VS: Vestibular Schwannoma
4. CI: Cochlear implant
5. Fb65: Freiburger speech recognition test at 65 dB
6. HMSq: Hochmaier-Schulz-Moser Test in silence
7. HMSn: Hochmaier-Schulz-Moser Test in noise, 10 dB
8. dB: Decibel
9. SD: Standard deviation
10. SSD: Single side deafness
11. SNHL: Sensory neural hearing loss
12. IAC: Internal auditory canal
13. MRI: Magnetic resonance imaging
14. FRT: Fractioned radiotherapy
15. SRS: Stereotactic radiosurgery
16. FPT: Fractioned proton therapy

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Pubikationen

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J. Clin. Med. **2023**, *12*, 6973.

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9. Erklärung

Erklärung nach § 2 Abs. 2 Nr. 7 und 8 PromO

Ich erkläre, dass ich die der Medizinischen Hochschule Hannover zur Promotion eingereichte Dissertation mit dem Titel

„***Long-term hearing outcome of cochlear Implantation in cases of simultaneous intracochlear schwannoma resection“***

in der Klinik für Hals- Nasen- und Ohrenheilkunde der Medizinischen Hochschule Hannover unter Betreuung, Unterstützung und in Zusammenarbeit mit Professor Dr. med. Thomas Lenarz ohne sonstige Hilfe durchgeführt und bei der Abfassung der Dissertation keine anderen als die dort aufgeführten Hilfsmittel benutzt habe.

Die Gelegenheit zum vorliegenden Promotionsverfahren ist mir nicht kommerziell vermittelt worden. Insbesondere habe ich keine Organisation eingeschaltet, die gegen Entgelt Betreuerinnen und Betreuer für die Anfertigung von Dissertationen sucht oder die mir obliegenden Pflichten hinsichtlich der Prüfungsleistungen für mich ganz oder teilweise erledigt.

Ich habe diese Dissertation bisher an keiner in- oder ausländischen Hochschule zur Promotion eingereicht. Weiterhin versichere ich, dass ich den beantragten Titel bisher noch nicht erworben habe.

Die Ergebnisse der Dissertation sind im Publikationsorgan Laryngoscope veröffentlicht.

Hannover, den _____