The emerging role of hearing loss rehabilitation in patients with vestibular schwannoma treated with Gamma Knife Radiosurgery. Literature review.

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INTRODUCTION

Vestibular schwannoma (VS) is typically a benign, slow-growing tumor that originates from the vestibulo-cochlear nerve (VIII cranial nerve). VS correspond to approximately 6-10% of all intracranial neoplasms **[1].** Most VS are sporadic, but 5-13% are associated with neurofibromatosis type 2 (NF2) and its growth has a negative impact on a patient's quality of life. **[1].**

The management of VS includes: observation with serial magnetic resonance imaging (MRI) and audiograms, microsurgical resection, fractioned radiotherapy, and stereotactic radiosurgery (SRS). In NF2 patients; chemotherapy with bevacizumab, a monoclonal antibody directed against vascular endothelial growth factor, has also been reported **[2]**.

Although microsurgery has been the primary active treatment of VS for many years, a substantial number of studies have reported the safety and efficacy of radiosurgery for VS, many patients prefer radiosurgery to surgical resection due to the significantly lower morbidity of the procedure while ensuring similar rates of long-term tumor control. The tumor control rate after SRS range from 87 to 98.4% and the overall morbidity and mortality rate range from 0% of mortality to 6% of morbidity compared to microsurgical resection **[Lee CC, Yen YS, Pan DH, et al Delayed microsurgery for vestibular schwannoma after gamma knife radiosurgery. J Neurooncol 2010;98:203-12], [Yang I, Aranda D, Han SJ et al. Hearing preservation after stereotactic radiosurgery for vestibular schwannoma: A systematic review. J Clin Neurosci 2009; 16:742-7], [Maniakas A, Saliba I. Microsurgery versus stereotactic radiation for small vestibular schwannoma: A meta.analysis of patients with more than 5 years' follow-up.Otol Neurotol 2012;33:1611-20],** [**Nicola Boari, M.D., Michele Bailo, M.D., Filippo Gagliardi, M.D., Alberto Franzin, M.D., Marco Gemma, M.D., Antonella del Vecchio, Ph.D., Angelo Bolognesi, M.D., Piero Picozzi, M.D., and Pietro Mortini, M.D. Gamma Knife radiosurgery for vestibular schwannoma: clinical results at long-term follow-up in a series of 379 patients. J Neurosurg (Suppl 2) 121:123–142, 2014**]. Comparative studies have shown an increase of 30–45% in the risk of deterioration of quality of life after VS surgery **[11].** Reported hearing preservation rates after observation, radiosurgery, and microsurgery for small to medium size VS were in a large series respectively 58.9%, 60.2% and 4.3%, whereas corresponding rates of tumor controls were respectively 71.1%, 97% and 94.3% **[12].**

Among delivery techniques for VS radiosurgery, most of the available evidence of efficacy and safety in VS treatment with SRS are with GKSRS **[13].** However, despite the high accuracy and the use of low margin doses (<13 Gy) that significantly reduce cranial nerve (CN) morbidity, hearing deterioration still remains a crucial and disabling side effect [**9**]. GKSRS post-treatment hearing loss, has an unclear and multifactorial pathophysiology, **[14].**

VS has a natural history of hearing loss, which is caused by a combination of auditory nerve compression and cochlear dysfunction due to ischemia, infarction, or invasion. A common audiometric sign of VS is the progressive speech discrimination decline that is worse than expected for the degree of hearing loss. Hearing loss resulting from bilateral vestibular schwannomas (VSs) has a devastating impact on the quality of life of patients with NF2. NF2 patients report that their greatest problem is deafness, which results in communication difficulties in their social life with close partners, family and friends. Patients also describe a relationship between general mood changes and hearing difficulties, social communication problems, balance difficulties and mobility problems**[15]**. Although NF2 patients may have significant cochlear nerve dysfunction and are less likely to achieve the hearing levels experienced in the general population, cochlear implant (CI) and auditory brainstem implant (ABI) remain the main therapeutic approaches to NF2 related deafness **[16]**

While in the presence of bilateral VS is obvious, the negative impact on the quality of life of unilateral deafness in patients with sporadic VS and normal contralateral hearing (single side deafness, SSD) is mainly the loss of binaural hearing, which is a three-dimensional perception needed for good comprehension of speech in noisy environments, localization and orientation to sounds (Ching et al, 2006). Patients with substantial unilateral hearing loss report a reduced quality of life (Wie et al., 2010). Analysis of SSQ questionnaires provided great details of how mode of unilateral input differentially influenced listening in a variety of scenarios and contexts, affecting all aspects of daily communication (Dwyer et al., 2014)

The effects of unilateral hearing loss are: the loss of summation effect (binaural loudness summation, which increases the audibility of a target signal), the reduction of the squelch effect (the ability of the brain to separate noise and speech coming from different locations), and the loss of the head shadow effect (improved speech discrimination in the shielded ear when the head is between the source of the sound and the noise).

CI rehabilitation in SSD leads to partial restoration of binaural hearing with an improvement in speech comprehension in noise and in sound localization, and partial suppression of subjective incapacitating tinnitus **[17]**.

Despite many study describe the possible causes or influencing factors of hearing loss after SRS for VS, only few authors only a few authors have tried to fill this gap proposing the use of devises for hearing rehabilitation in VS patients.

Data reported in the literature concerning CI outcomes after SRS are reported but are still limited to NF2 patients, and results are affected by many patient and tumor variables and a correlation with radiation dose and acoustic drops is not possible. Data concerning CI outcomes in patients after sporadic VS surgical resection are also very limited, but promising. An improvement of speech perception in noise, in sound localization with tinnitus suppression has been observed **[18]**. Therefore, CI has shown to be a viable solution also for patients with sporadic VS.
The aim of the present study was to highlight the role of hearing rehabilitation after VS-SRS treatment.
A descriptive review of literature was performed in order to define the role of hearing rehabilitation after GKRS treatment in VS patients. Clinical case reports of patients undergoing CI surgery after VS-SRS were collected and analyzed in order to evaluate the audiological outcomes in sporadic and NF2 VS treated by GKRS.
A second aim of the study was to evaluate the possible role of CI in sporadic VS treated with GKRS, establishing a plan for a more evidence-based approach for patients suffering from SSD or deafness.

MATERIALS AND METHODS

A literature review of the current clinical data was performed searching scientific literature databases. Relevant references of published articles were screened. The Medline search was conducted on Pubmed, Scopus, Embase, ISI-web, Cochrane library and Web of Science. Keywords and mesh terms searched were: cochlear implants AND acoustic neuroma; cochlear implant and vestibular schwannoma; cochlear implants AND radiation therapy AND acoustic neuroma; cochlear implants AND radiation therapy AND vestibular schwannoma; cochlear implants AND radio-surgery, Stereotactic radio-surgery AND cochlear implants; hearing restoration AND radiosurgery.
Articles reporting clinical and audiological outcomes in NF2 patients were included. Case reports, case series, editorials, technical reports, case–control studies, cohort studies, retrospective studies, meta-analyses or clinical trials reporting clinical and audiological outcomes after GKRS treatment were considered. Systematic reviews were thoroughly screened for possible inclusion. Non-English articles and animal studies were excluded. Other exclusion criteria were: papers dealing with GKSRS-VS patients not treated with CI, and with less than five months of follow up (FU); patients with CI in only observed or surgical treatment VS. The search results were independently screened by two of the authors (CM and PM); disagreements were resolved by consensus.
Selected articles reporting CI patients affected by VS and treated with radiotherapy and radiosurgery, the following data were extracted from each article, if reported:

year of publication, number of patients included in each study, number of patients treated with radiotherapy out of the total, epidemiological and demographic data of the patients (gender, age, diagnosis of NF2), preoperative symptoms, time to implantation, median interval between tumor intervention and implantation, the median ipsilateral duration (MO) of deafness before CI, hearing in the contralateral ear at the time of CI, preoperative audiological data (PTA WRS), tumor data (size), tumor treatment (only radiosurgery/radiotherapy or radiosurgery/radiotherapy and surgery), dose of radiation therapy, type of cochlear implant used, complications, average follow up, postoperative clinical, and audiological outcomes.

The collected data were analysed by means of descriptive statistics (absolute and relative frequencies). Continuous variables were expressed as mean ± standard deviation (SD), and categorical variables as percentages.

RESULTS

The search of the articles was performed on Pubmed, Scopus, Embase, ISI-web, Cochrane library and Web of Science: the research was complementary in all of these libraries

A literature search using Pubmed obtained 738 full text papers. The following keywords were used to search the Pubmed database:

- cochlear implant AND vestibular schwannoma: 274 papers
- cochlear implants AND acoustic neuroma: 160 papers
- cochlear implants AND radiation therapy AND vestibular schwannoma: 21 papers
- cochlear implants AND radiation therapy AND acoustic neuroma: 19 papers
- Gamma Knife AND cochlear implant: 26 papers
- stereotactic radiosurgery AND cochlear implants: 12 papers
- hearing restoration AND radiosurgery: 3 papers
- hearing preservation after Gamma Knife radiosurgery: 211 papers
- audiological outcome AND Gamma Knife radiosurgery: 13 papers

Literature review results were depicted in the PRISMA (**P**referred **R**eporting **I**tems for **S**ystematic Reviews and **M**eta-**A**nalyses) flow diagram **(Fig 1)**.

From all of the articles found, 169 non-duplicate studies were initially selected and 130 articles were excluded. Exclusion criteria results were: only surgical treated VS (42), clinical retrospective series of SRS or surgical treated VS without CI with < 5 months follow-up (58), cases or series with CI without VS treatment (1), CI in non VS tumors (4), and descriptive or review articles (25).

From the remaining thirty-nine (39) studies, sixteen (16) case reports or case series describing patients affected by sporadic or NF2 VS that underwent CI after VS radiosurgical treatment were selected. Out of a total of 138 patients described in the 16 included papers, 44 subjects were treated with radiosurgery (18 male, 19 female and in 7 cases the sex was not specified; 43 NF2 and 1 sporadic VS) and were included in the analysis.

The median age of the patients (when reported) was 45.4 years (SD +/- 17.2). Audiological data were reported as hearing threshold pure tone audiometry (PTA) (16 studies) in dB or as AAOHNS (American Academy of Otolaryngology-Head Neck Surgery) hearing class ranging from A-D (normal to not-serviceable hearing), speech perception in quiet (12 studies), speech perception in noise with a variable fixed signal/noise ratio (SNR) (14 studies).

In studies where speech in quiet or SNR were assessed, it was uncommon to find description of signal and noise presentation or SN ratio, and therefore outcomes will be generically accounted as pre-post SRS: improvement, deterioration or unchanged hearing outcome **(Table 1-4) [19-34].**

Tumor dimensions were reported in 14 articles (37 cases). According to ~~Sami~~ Koos grading classification **[35]** all treated tumors were small to medium-size lesions (≤ Koos T3A-B; 25-30 mm in cisternal diameter). ~~Sami~~ Koos stage T1 (tumor confined to IAC or <10 mm) was described in 5 cases. ~~Sami~~ Koos stage T2-T3A (>10 mm <20 mm in cisternal diameter) was described in 14 patients, and 18 cases were T3A T3B. In 7 cases the tumor size was not reported.

The interval between tumor intervention and cochlear ipsilateral implantation was described in 11 articles (25 patients) and the median time was 71 months.

The median ipsilateral duration of deafness before CI was reported in 13 articles (30 patients) and it was 87.5 months. The hearing in the contralateral ear at the time the CI was reported in 12 articles was described as complete deafness or profound hearing loss (GR hearing class D).

All patients underwent radiation treatment on the ipsilateral implanted tumor. The dose of radiation therapy was reported in 5 studies (14 patients). Stereotactic radiosurgery with Gamma Knife was performed in 9 patients. In 5 cases fractionated radiotherapy was done.

The mean duration of follow-up after CI was 24.42 months.

Audiological assessment:

Preoperative audiological assessment (PTA, WRS) showed profound or severe-profound hearing loss in all cases. PTA 25-4000 HZ DB was 51 dB in only one case (Sporadic AN). Speech perception in quiet was described in 12 papers (21 cases) and the mean value was 6% (word 0-65%-average 8.3%, sentences 0%); speech perception in noise was reported in 10 papers (19 cases) ranging from 0-36% (average 2.16%).

When described (11 articles), intraoperative telemetry was normal in 13 cases. A poor neural response telemetry (NRT) was recorded in 3 patients and in 2 cases no intraoperative electrode response was detected.

Post-implant audiological assessment showed an improvement related to PTA dB value in 6 papers (20 patients), a no response in quiet for 13 patients, and no response in noise in 16 cases.

WRS was recorded in 18 patients ranging from 0-100% (average 52.1%).

The sentence score in quiet was recorded for 26 patients, ranging from 0-100% (average 62.5%).

Sentence score in noise was described for seventeen (17) patients, ranging from 0-98% (average 52.04%).

Open-set speech perception was tested in nineteen (19) patients: 8 patients recorded good or excellent recognition; no sound perception was defined in 2 patients; only environmental sound or lip reading was recorded for 7 patients; one patient was unable to perform the test.

The clinical outcome at the last follow up showed an audiological improvement in twenty-five (25) of the forty-four (44) patients. The audiological outcome was unchanged in 16 cases. Audiological deterioration was recorded in 3 cases.

In the specific literature which addresses hearing loss after SRS, it is classified according to the Gardner-Robertson (GR) hearing scale and modification of the Silverstein and Norell classification in five classes: Class I, good or no hearing deterioration; Class II, serviceable; Class III, non-serviceable hearing; Class IV, poor; Class V, deaf; anacusis or not sufficiently evaluable hearing **[36].**

According to the head and neck classification by the American Academy of Otolaryngology, each GR class is associated to a value of pure tone average threshold (PTA) and to a percentage of speech discrimination (SDS) **[37]**.

GR class 1 correspond to PTA of 30 dB or less with a SDS of 70% or greater.

GR class 2 correspond to PTA of 30 dB or less with a SDS between 50-69%.

Class 1 and 2 correspond to serviceable functional hearing.

GR class 3 correspond to PTA ≥ 51 dB but more then 30 dB with a SDS between 5-49%.

GR class 4 correspond to PTA ≥ 91 dB with a SDS between 1-4%.

In GR class 5 PTA is not testable and SDS is 0%.

DISCUSSION

The present study is a review on CI audiological outcomes in patients treated with GKRS for VSs, experiencing post-treatment deafness. Although GKRS has been established as a first-line treatment option for small to medium-size VSs, it remains difficult to preserve hearing function [Tveiten et al with

PMID 25850598.]. Detailed data concerning long-term hearing function at follow-up appointments (> 5 years) after GKS are scarce, and hearing preservation remains an underestimated problem.

When functional hearing deteriorations occur, the benefits of binaural hearing are lost. The audiological impact of sporadic VS in patients with normal contralateral hearing is mainly the loss of binaural hearing, which induces decreased audibility, worsening of perception in noisy environments, of orientation to sounds, and localization of target signals [17].

The mechanisms of hearing deterioration after radiosurgery for VS are not yet fully understood **[39-42]** On one side there is a compressive effect on the cochlear nerve, a vascular occlusion of internal auditory artery or *vasa nervorum,* and the biochemical alterations of the inner ear fluids **[38].** Also radiation effect on the cochlea can explain hearing loss , therefore only low marginal (<13 Gy) and modiolous (4 Gy) are accepted.

Hearing deterioration within 3 years after GKS could be explained by ischemic or mechanical damage to the cochlear nerve due to dose irradiation or nerve compression by transient tumor expansion. This transient tumor expansion can be observed during the first 2 years after treatment, and is a consequence of post-actinic citotoxical oedema **[9]**. Even if in the first 2 years after treatment the mean PTA loss is higher (7.03 dB/year), as has been demonstrated in many articles that hearing acuity continues to deteriorate even beyond 5 years (2.39 dB/year) corresponding with hearing preservation rates of 47–77% at 3 years, 28–64% after 5 years and 23–45% at 10 years **[9,44, 51-53].**

The causes of this progressive hearing deterioration at long term follow-up are not known.

It seems likely that improvement after CI in SRS treated patients could also be conditioned by different variables such as radiotherapy modality, dose and pre-treatment audiological characteristics of patients. Therefore a synthetic overview of SRS effects on hearing is given.

The role of GKRS in VSs and hearing preservation

Since the first VS radiosurgical treatment in 1979 **[43],** many studies have investigated and described GKRS as an effective and safe treatment. Actually, many patients prefer radiosurgery to resection because of lower morbidity and shorter hospitalization length compared to surgical resection **[9-10, 44-45]**.

Before radiosurgical treatment, signs and symptoms should be accurately collected. Before and after treatment, hearing function can be evaluated according to Gardner-Robertson (GR) modification of the Silverstein and Norell classification **[36-37].**

Tumor morphology, tumor volume, the length between VS intracanalicular portion and the fundus of the IAC, and the distance between VS and the cochlea are calculated and investigated from pre and post-treatment MRI scans using GammaPlan software (Elekta).

The maximal inner dose should vary from 20 to 32.6 Gy (median 26 Gy), the marginal dose from 11 to 15 Gy (median ≤ 13 Gy). According to some authors the maximum cochlear dose should not exceed 4 Gy to ensure hearing preservation **[46]**. The isodose line for the tumor margin varied from 40-60% (on average 50%). The number of isocenters can vary from 1 to 41 (on average 12). The application of a low-dose irradiation (marginal dose 11–13 Gy at 50% isodose line), results in 89–99% overall tumor control rates (93–95% at 5 years and 86–95% at 10 years after GKS) **[9,47-49]**. The use of lower margin doses (≤ 13 Gy) with a median mean cochlear dose of 4.0 Gy should reduce the risk of cochlear damage but tumor control can be achieved with longer follow up **[47-49]**.

Among the effects on hearing loss, no significant difference between single and fractionated doses are reported in the literature **[50**].

Radiotherapy and hearing outcomes

Many investigators have reported relatively long-term hearing outcomes after SRS, but these results are still unsatisfactory, showing an overall serviceable hearing preservation rate of ≤ 50% over longer periods. [**9,39**].

The use of conformal and selective irradiation with avoidance of excessive irradiation of the adjacent structures minimizes the risk of treatment-related complications, including trigeminal and facial neuropathy **[14].** Nevertheless, hearing deterioration represents the main unresolved drawback of its procedure especially in younger patients **[51-53].**

Many factors, including a patient's age, Gardner-Robertson hearing class before irradiation, Koos tumor stage, extension of the intrameatal part of the neoplasm up to fundus, tumor shape, nerve of tumor origin, presence of cystic changes in the neoplasm, cochlear dose, marginal dose or treatment modality (single section or fractionated), were widely investigated, but only a small amount of data are statistically significant **[9,39].**

A comprehensive analysis of Yang et al. and a systematic meta-analysis of Arthurs et al. revealed a statistically significant correlation among serviceable hearing preservation and the use of marginal dose < 13 Gy **[39, 54**]. In their literature review hearing preservation was observed in 50-60% of cases during 35-71 months of follow-up.

Yang et al. also reported that among 4234 included patients (followed up on average 44.4 months, median 35 months) the overall preservation of functional hearing was 51% (60.5% in patients receiving ≤ 13 Gy and 50.4% in patients receiving > 13 Gy) **[39]**.

A retrospective series of Boari et al. described an overall hearing preservation rate of 49% during a mean follow up of 59.9 months. They showed in the multivariate regression model that pre-GKS PTA, difference between bilateral pre-GKS PTA, mean cochlear dose, fundus obliteration, use of a 4mm collimator to the intracanalicular portion, and distance from the fundus and the tumor end were significant for hearing preservation, but the only independent factor found to be significant at the multivariate analysis for GR class loss and functional hearing loss was the age >55 years. [**9**].

Frischer et al. showed with an univariated regression analysis, that after 2 years follow-up of the GR hearing class prior to GKRS, Koos grade at GKRS, median cochlear dose, age at GKRS, prescription dose, radiation time, and target volume seemed to have a significant impact on the GR hearing class at the last follow-up. However, in the multivariate regression model, only the GR class prior to GKRS and the median dose to the cochlea were found to be independent predictors of the GR class at follow-up ($p < 0.001$ and $p = 0.029$, respectively) **[44].**

Tumor volume and its morphology are other possible relevant parameters to predict the probability of hearing preservation after GKRS **[38-41].**

Tumors without whole IAC involvement, with a greater distance from the cochlea, and less brainstem compression showed a better hearing preservation after GKRS **[38-41].**

Conversely tumors with projection into intracanalicular portion (pear and linear type) are associated with a greater tumor length in the IAC and a worse hearing outcome **[38]**.

Hearing preservation rates after radiosurgery have been correlated with total radiation dose delivered to the cochlea. Therefore, a median marginal dose ≤ 13 Gy and the maximum cochlear dose of 4 Gy are accepted values in order to enhance the probability of hearing preservation **[46]**. **(Table 5).**

CI rehabilitation in SRS treated VS

When hearing preservation is not possible, hearing rehabilitation is an important step to provide for a better quality of life for these patients.

In patients affected by sensorineural hearing loss with retro-cochlear pattern such as in VS, the conventional hearing aid amplifications are often ineffective and auditory brainstem implants (ABI) infrequently permit high-level open-set capacity **[20]**.

Historically, in the presence of sensorineural hearing loss due to retro-cochlear lesions, CI was considered a contraindication **[20].** However, few studies have reported favourable hearing outcomes of CI in patients affected by NF2 **[20,55-61]** since the first case report in 1999 **[60].** In these patients CI is mainly proposed when the cochlear nerve remains intact, as in patients with a stable tumor or after nerve preserving surgery **[24-26, 28, 55- 65].**

A favourable outcome (in squelch effect and sound localization) was also reported in patients with sporadic VS in the only or better hearing ear, with similar results to those patients without VS treated with CI **[55,64].**

From the present literature review it emerged that the role of hearing rehabilitation (CI) can improve audiological results for VS patients treated with GKRS. Improvements were described ranging from changes in awareness of sounds, lipreading to open set words, and sentence recognition under noise competition. A general analysis of data reported in the included studies, showed that after a mean follow up of 24.42 months from CI, 56.8% of patients improved audiologically, 36.4% were unchanged and only

6.8% deteriorated. However, due to differences in audiological test items (phonemes, words, sentences) and procedures (i.e. speech in quiet vs noise, lipreading vs open set speech perception), and due to the small numbers of patients implanted, it was not possible to perform a statistical analysis.

More in depth, the case series included reported 45.5% of PTA hearing improvement. Most of these subjects were NF2 patients with bilateral audiological deterioration. In these patients the CI has the main role of rehabilitating one-side hearing prior to the loss of contralateral hearing. Speech perception outcomes showed an improvement in 45.5% of cases, while only thirteen (13) reports have described an improvement of speech perception in noise. 4 patients had serviceable hearing in the controlateral ear, (one of them had bilateral CI) and in this case could have been analysed as quasi-SSD. In case of bilateral profound deafness, CI has the role of restoring speech perception, and improving patients' communication. A descriptive analysis of the pool of patients without audiological improvement (36.4% unchanged and 6.8% deteriorated: 43.2% of the total) after a mean follow up of 24.42 months from CI implant showed that the most relevant influencing factors of negative outcome were: severity of NF2 phenotype, long history of ipsilateral profound deafness before implantation, progressive tumor growth after SRS (necessitating surgical resection with resultant cochlear nerve sacrifice and CI removal) and high radiation dose (20 and 40 Gy). Although interesting, since this pool of patients were NF2, these data may not be relevant for sporadic cases.

CI in SSD patients (4 patients: P23,27,28,29) has the role of restoring binaural hearing; 3/4 subjects showed an audiological improvement. The only patient that remained unchanged (in Carlos. 2016 series **[29]**) presented a long history of profound hearing loss before implant (Preoperative Word score and sentences score: 0%), and the intraoperative NRT was absented. It is interesting to note, how in none of these patients the role of CI in binaural hearing has been assessed.

Summarizing the present findings it might be inferred that CI could represent a potential option to recover hearing loss both in sporadic cases, and in NF2 VS patients treated with GKRS. CI should be considered in patients with an anatomically preserved cochlear nerve, when BKB sentence scores drop below 50% in best-aided conditions.

In the early stage after VS radiosurgery, if the nerve remains functionally intact with preservation of its anatomy and blood supply (noted by minimal residual hearing at audiogram test), then CI should represent a valid alternative to other implants. When the cochlear nerve remains anatomically intact after SRS, the CI can still be partially effective also in the presence of acoustic nerve atrophy. As a matter of fact, 81.25% of subjects tested intraoperatively showed a neural response to electric stimulation through the implant. The interest in preserving the integrity of the cochlear nerve is therefore increasing in VS treatment, and SRS is a valid option also in the perspective of future hearing rehabilitation.

Finally, one more issue must be considered, which is the need of frequent MRI scans during the first 5 years of follow-up. Because of the magnetic components present in the CI receiver placed under the skin, image artefacts occur during the acquisition of the MRI images, limiting the diagnostic validity.

The 1.5 T MRI is required at 6 months after the first treatment and then every year for 5 years. After 5 years, MRI can be performed every 2 years. Considering the progression of hearing deterioration (23-53% at 3 years, 36-72% at 5 years and 55–77% at 10 years) **[9,44, 51-53]**, most patients may benefit from a CI in the first 2 years after radiosurgery. Removable magnet CI and a low Tesla MRI should facilitate post CI imaging. From a technical point of view, a more horizontal and posterior position of the magnet has been purposed to allow better visualization of the IAC and the labyrinth, making cerebellopontine angle visualization possible **[68]**. Finally, in selected cases CT scan with contrast enhancement could be a valid option.

Implant dislocation, pain or demagnetization of the CI magnet can also occur, further disadvantaging patients. Usually MRI scans up to 1.5T are safe in patients carrying CIs with non-removable magnets if strict guidelines are followed **[66-67].** More recent devices are approved for 3T MRI with no risks of pain for patients. Magnet removal remains an important issue for devices implanted before 2016 **[69].** Although magnet removal can be easily performed under local anaesthesia via a small incision behind the ear, the procedure may lead to additional discomfort for the patient and an increased risk of possible complications (scars, adhesions, infections).

There are numerous limitations inherent within our study. The study is a descriptive review of the literature and the included cases lacked validated outcome measures. The patients' population is heterogeneous and the radiosurgical treatment modality and parameters are not reported in all papers. The number of collected patients is limited. The lack of a consistent cohort of sporadic VS patient did not allow us to establish a control group for comparison.

CONCLUSION

The negative impact on a patient's quality-of-life from hearing loss after Gamma Knife for VS is still an underestimated problem, and the role of hearing rehabilitation is an under investigated topic. Nevertheless, data concerning CI outcomes in VS patients (sporadic and NF2) treated with SRS are promising. Therefore, radiosurgery followed by CI when indicated, may represent a potential emerging option in the management of these patients, aimed to improve their quality of life.

Parameters like a patient's age, Gardner-Robertson hearing class before irradiation, tumor size, tumor shape, median cochlear dose and marginal radiation dose seem to be related to hearing outcomes after radiosurgery.

A possible relation between factors influencing hearing drops after SRS, and the implantation timing has not yet been comprehensively investigated.

To date the time of implantation depends on the level of sentence recognition reduction (50%) related to the level of stimuli (in dB), and hearing deterioration can happen both in the early stages after treatment or at long-term follow up.

Radiological follow up with periodic 1.5 T MRI may represent a disadvantage for implanted patients, but the related problems have been investigated and partly resolved. An important open question is related to the radiological follow-up in patients with early hearing loss, but related data in the literature is still poor.

Therefore according to these promising results, and to the need for improving the quality of life in patients suffering from SSD or deafness after SRS for VS, a plan for a more evidence-based approach to this complex scenario is advisable.

CONFLICT OF INTEREST

The authors have no personal financial or institutional interest in any of the drugs, materials, or devices described in this article. The Authors declares that there is no conflict of interest

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