

## Hearing Preservation in Vestibular Schwannomas – A concise look at evidence based recommendations

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**Abstract:** In today's age of high expectations from surgery despite the nature of the lesion, and affected organ, functionality post surgery sometimes becomes more important than tumor clearance and recurrence. One such lesion is the vestibular Schwannoma which post-surgery usually is associated with hearing loss. In the last decade there has been an explosion of literature regarding hearing preservation in surgery as well as other modalities used to treat this disease such as Radiation therapy and Neuro-observation. We have reviewed at least 20 recent manuscripts and reports in literature and compiled the results together to allow for broad based inferences to be understood.

**Keywords:** Vestibular Schwannoma, Hearing preservation, Retromastoid Craniotomy, Radiation therapy, neuro-observation

### INTRODUCTION

Throughout the years, significant advances have been made from Cushing's original treatise where mortality was 80% and hearing loss obligatory. Apart from surgery, other non invasive modalities such as radiation therapy have also come into the fray with their respective champions propounding their virtues high and low. The greatest surprise was the resurface of the time and tested method of blissful inactivity (neuro-observation) in certain specific cases. The crux of such a multipronged assault on vestibular schwannomas is the preservation of hearing. As mentioned earlier, obligatory hearing loss was no longer acceptable, and slowly hearing preservation became achievable. Technology coupled with differing objectives have produced dependable outcomes.

Evolution of management is such that hearing (functional) outcome is preferable to tumor eradication. Thus, small remnants with preserved hearing is not just socially acceptable, but also neurosurgically and medically suitable with multiple modalities available. The current benchmarks of therapy are tumor control, hearing preservation and facial nerve function. The appropriate balance among these 3 modalities achieves the best result of them all

### THE PROBLEM

The data in literature about long term hearing preservation is conflicting. Various studies with

Disparate methodology, Heterogeneous reporting and unreliable documentation make such reports difficult to depend upon. There is also the very real prospect of not always considering serviceable hearing but merely the presence of any hearing, which is unfortunate. The many different incompatible classification systems for measuring to measure hearing preservation systems lead to discrepancies in correlation. Lastly, baseline data of hearing prior to therapy isn't measured, leading to falsely bad results later after surgery.

Some studies only provide overall prevalence of hearing preservation as opposed to hearing status at specific time points in follow up. Long term post procedure follow up isn't always available, and the Length of audiometric follow up not uniform (or sometimes not mentioned at all). Another significant issue not probably addressed well is the Lack of reporting of hearing status in the contralateral ear (This assists in overall hearing decline especially in the elderly) These are a variety of reasons of why and where much of the available data today falls short. In this examination of reliable literature reports and evidence, we look impartially at radiation, surgery and observation as well as compare and contrast these modalities with certain situations to provide the best answer to the question of hearing preservation.

Hence in this document, we look at evidence regarding Radiation therapy, Surgery and Observation alone and as compared with other modalities and present a consolidated look at the evidence available for Hearing preservation.

### RADIATION THERAPY

The most consistent prognostic features associated with maintenance of serviceable hearing are, good preoperative word recognition, pure tone thresholds with variable cut-points reported, smaller tumor size, marginal tumor dose  $\leq 12$  Gy, and a cochlear dose  $\leq 4$  Gy.

In 2010, Regis et al presented a consecutive series of 47 patients with intracanalicular Vestibular Schwannomas who were managed with conservative observation and 34 patients with intracanalicular tumors who received proactive radiosurgery using a median dose of 12 Gy to the tumor margin. They found that of the 31 patients

with serviceable hearing at the time of observation commencement, 21 (68%) maintained useful hearing.

In 2012, Rasmussen et al<sup>62</sup> compared the outcomes of 42 patients who received fractionated radiation therapy to a historical cohort of 409 control subjects who received observation and were matched by initial hearing levels. They reported that at 2 years after radiation therapy, only 8 of an initial 21 (38%) patients with serviceable hearing maintained GR grade I or II hearing, and at 10 years all had progressed to non serviceable hearing.

In 2013, Breivik et al prospectively compared an observational cohort (n = 124) to a radiosurgical arm (n= 113) receiving 12 Gy to the margin, and all 237 patients had tumors with extracanalicular extension. At a mean follow-up of 55 months, 17 of 71 (24%) conservatively managed patients with serviceable hearing at baseline maintained GR grade I or II hearing, compared to 19 of 53 (36%) who received radiosurgery. It is notable that treatment was not randomized, but followed an institutional algorithm. (larger tumors went into the treatment arm)

Kim et al evaluated a cohort of 41 patients with serviceable pre-treatment hearing who underwent radiosurgery and compared this to a historical cohort of 15 patients who were managed with observation. However, analyses comparing the radiosurgery and observation cohorts were only made for 19 of the radiosurgery patients who experienced acute hearing decline and received glucocorticoid therapy

Thus combining all results and data we find that there is Class III evidence supporting the conclusion that the risk of HL increases with time, well beyond the first 2 years following radiation treatment. When evaluating all patients with serviceable hearing at baseline, approximately 72% will maintain serviceable hearing at 2 years, 63% at 5 years, and 33% at 10 years.

## **SURGERY**

The most consistent prognostic features associated with maintenance of serviceable hearing are, good preoperative word recognition, pure tone thresholds with variable cut-points reported, smaller tumor size, and presence of a distal internal auditory canal cerebrospinal fluid fundal cap.

In 2005, Grayeli et al<sup>115</sup> compared the results of microsurgery and conservative observation in a cohort of 416 unilateral VSs: 114 intracanalicular and 302 with  $\leq 15$  mm in greatest cisternal dimension. The 111 conservatively managed patients consisted of those over 60 years of age and those who had contraindications or refused surgery. Of the 44 patients who presented with serviceable hearing, 25 (57%) maintained AAO-HNS class A or B at last follow-up. The mean follow-up in the microsurgery arm was 18 months. Initially, 183 patients had serviceable hearing at baseline and of these, 145 underwent attempted

hearing preservation via the middle fossa or retrosigmoid approach. Of the latter, 45 (31%) maintained serviceable hearing at one year following surgery.

In 2005, Lin et al published a retrospective study comparing hearing preservation outcomes consisting of a group of 16 patients who received hyper fractionated radiation therapy (50 Gy, 25 fractions over 5 weeks), 113 patients who received retrosigmoid craniotomy for hearing preservation microsurgery, and 51 patients who were managed with conservative observation. With the microsurgical arm, 30 (27%) had serviceable hearing in the immediate postoperative period, and over a mean follow-up of 9.5 years, 18 (16%) maintained long-term useful hearing. The rate of initial hearing preservation following microsurgery for tumors  $< 2$  cm was relatively low; however, it is notable that only 10% of patients progressed to nonserviceable hearing after a follow-up of nearly 10 years if useful hearing was initially preserved.

In 2003, Chee, Nedzelski, and Rowed found that among patients who had serviceable hearing immediately following retrosigmoid tumor resection, 15 of 23 (65%) patients maintained useful hearing at a mean follow-up of 9.5 years following surgery.

In 2010, Sughrue et al evaluated surgical outcomes in patients less than 40 years of age and found that if hearing was initially preserved, no patients progressed to nonserviceable hearing in the operated ear even after 10 years of follow-up.

In 2014, Quist et al reported that 12 of 16 (75%) patients who had hearing initially preserved following middle fossa tumor resection maintained AAO-HNS class A or B hearing after 5 years of follow-up.

In 2014, Yamakami et al reported that 80% (12/15) of patients who initially had hearing preserved following microsurgery maintained useful hearing at a median follow-up of 7 years.

Thus the synthesis of all reports and result present that the greatest risk to hearing with surgery occurs upfront. If hearing is initially preserved following surgery, the results tend to be durable. This is in contrast to conservative observation and radiation where the immediate risk is low, but delayed or protracted loss of serviceable hearing is common.

## **OBSERVATION**

The most consistent prognostic features associated with maintenance of serviceable hearing are good preoperative word recognition, pure tone thresholds with variable cut-points reported, as well as nongrowth of the tumor. Initial tumor size has no bearing on hearing preservation.

In 2010, Stangerup et al evaluated the outcomes of 1144 patients who were initially managed with conservative observation. Within this group, 377

patients had a minimum of 5 years of follow-up, and 102 patients had at least 10 years. Overall, 249 of 455 (55%) patients who presented with AAO-HNS class A or B hearing maintained serviceable hearing at last follow-up, and when only evaluating those who presented with class A hearing, 81% (144/178 patients) maintained serviceable hearing at last follow-up.

In 2008, Ferri et al reported the results of a prospective study where 123 patients with VSs were observed for a mean follow-up of 4.8 years. Of 56 patients who initially presented with serviceable hearing, 41 (73%) maintained useful hearing at last follow-up.

Thus we see that the risk of HL increases with time during conservative management. Similar to radiation therapy, the development of nonserviceable hearing is often protracted, continuing many years beyond diagnosis. The 2 strongest prognostic factors for the development of nonserviceable hearing are tumor growth and poor hearing at the beginning of observation.

Now, we move on to literature reports of comparisons between the modalities listed above.

### RADIATION VS SURGERY

The risk of HL with surgery is upfront; if useful hearing is initially preserved following surgery, the results appear to be durable in many cases, for at least 10 years. This is in contrast to radiation and conservative observation, where the initial risk to hearing is low; however, delayed loss is common and progressive over time. Hence it is only appropriate that we look at hearing preservation with respect to time.

### SHORT TERM VS LONG TERM

Therefore, in the short term, patients are most likely to maintain useful hearing following conservative management or contemporary low-dose radiation therapy. However, if progressive HL continues indefinitely in the latter 2 groups, which could be reasonably inferred from the current data, then the long-term advantage may favour microsurgery.

In 2006, Pollock et al reported the first prospective, nonrandomized study comparing outcomes between 36 patients who received microsurgery and 46 patients who received radiosurgery. Preservation of serviceable hearing was greater for the radiosurgery arm than the microsurgical group at 3 months (77% vs 5%,  $P < .001$ ), 1 year (63% vs 5%,  $P < .001$ ), and last follow-up (63% vs 5%,  $P < .0001$ ). A similar finding was reported when comparing the rate of AAO-HNS class A hearing between groups

In 2009, Myrseth et al reported the second prospective, nonrandomized study comparing outcomes of 63 patients who underwent Gamma Knife radiosurgery and 28 patients who underwent microsurgery. At both the 1- and 2-year time points, the Gamma Knife

radiosurgery cohort had a statistically significantly greater proportion of patients with hearing preservation compared to the microsurgery group.

In 2003, Yamakami et al published a large review comparing outcomes following radiation therapy (9 studies, 1475 patients), microsurgery (16 studies, 5005 patients), and conservative observation (13 studies, 903 patients). In total, 57% of 271 patients who received radiation treatment retained useful hearing following treatment, 36% of 1448 patients who underwent microsurgical resection with intent of hearing preservation, and 63% of 60 patients who were observed.

Maniakas and Saliba published a review comparing long-term hearing and tumor control outcomes between microsurgery and radiation therapy for small (<2 cm) VSs, requiring a minimum of 5 years of follow-up. Eight studies analysing 410 cases were included in the stereotactic radiation population. The mean duration of follow-up was 6.9 years and 70.2% of patients had a useful hearing preservation outcome. This is compared to 7 studies with 77 patients who underwent microsurgery, including 38 who received retrosigmoid craniotomy and 39 who underwent middle fossa craniotomy. There was no statistical difference between surgical approaches, and the overall hearing preservation rate of 50.3% was seen at an average follow-up of 7.1 years.

Kaylie et al also performed a review comparing microsurgery and radiosurgery and found that the prevalence of hearing preservation was identical between modalities. Specifically, at a mean follow-up of 24 months, 44% of 599 patients who received microsurgery and 44% of 219 patients who received radiosurgery retained serviceable hearing following treatment.

Thus, a synthesis of results show that Radiation and observation are better at overall hearing preservation than surgery. However, not all tumors can however be managed by RT and observation alone. Large tumors, recurrent lesions, and syndromic tumors require surgery (where sometimes hearing preservation is often not discussed at all)

### CONCLUSION

Although radiation and Observation are overall better in hearing preservation, not all cases can be treated by them alone. Surgery is needed in many tumors mentioned above. Therefore a patient to patient based approach weighing the pros and cons must be taken before subjecting the patient to therapy.

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