Comparison of management options for patients with acoustic neuromas

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Management options for patients with vestibular schwannomas (acoustic neuromas) include observation, tumor resection, stereotactic radiosurgery, and fractionated radiotherapy. In this report the authors review their 15-year experience with radiosurgery and discuss indications and expectations in relation to the different approaches. They conducted a survey of neurosurgeons to determine management preferences in two different cases of intra- and extracanalicular tumor presentations. Patient decisions must be based on quality information derived from peer-reviewed literature.

KEY WORDS • acoustic neuroma • vestibular schwannoma • radiosurgery • gamma knife

For the treatment of patients with acoustic neuromas there are several options including observation, resection, SRS, and fractionated SRT. Many patients choose between SRS and resection based on their own specific goals and their understanding of possible results. The decision can be difficult and depends on the sources and strengths of information given to them. These include discussions with surgeons and other physicians, written material disseminated by peer-reviewed medical journals, brochures from support groups, internet-based reports (of variable reliability), and discussions among fellow patients. We believe that information provided in the peer-reviewed medical literature is the most reliable for patient education. Nevertheless, some patients become confused by what they perceive as conflicting opinions among physicians. Many such physicians provide only one kind of treatment and may appear to be biased toward their own approach.

Resection is indicated for patients harboring larger tumors that have caused major neurological deficits due to brain compression. Surgeons perform SRS for small- or medium-sized tumors, their goals being to preserve neurological function and prevent tumor growth. The long-term outcomes of SRS, particularly GKS, have proven its role in the primary or adjuvant management of this tumor. Fractionated radiotherapy has been suggested as an alternative in selected patients with larger tumors for whom microsurgery may not be feasible, as well as in some patients in whom preservation of cranial nerve function is being attempted. Most such centers do not offer conformal SRS. Cases of NF2 pose specific challenges, particularly the preservation of hearing and other cranial nerve function.

Primary clinical issues include avoiding tumor-related or treatment-related mortality, prevention of further tumor-induced neurological disability, minimizing treatment risks such as CSF leakage, infections, or cardiopulmonary complications, maintaining regional cranial nerve function (facial, trigeminal, cochlear, and glossopharyngeal/vagal), avoiding hydrocephalus, maintaining quality of life and employment, and reducing cost. In choosing any particular therapy, the team should strive to meet all of these goals.

MANAGEMENT OF ACOUSTIC NEUROMA

Resection of the Lesion

There are three main surgical routes for resection of an acoustic neuroma. These approaches are chosen based on educational bias, surgery-related experience, and the spe-
cific goals of the operation. The suboccipital retrosigmoid approach is the oldest and remains widely used, particularly when hearing preservation is attempted. The translabyrinthine approach destroys hearing but provides direct exposure to the tumor without requiring cerebellar retraction. Even large tumors can be removed through this route. The “middle fossa” approach is performed via a temporal craniotomy but requires elevation of the temporal lobe and drilling of the temporal bone to expose the auditory canal from above. Using this route, hearing preservation can be attempted. It is usually chosen for patients with intracanalicular tumors.

The goals and acceptable clinical outcomes have evolved over time. Many years ago, the goal was simple debulking of the tumor (which was often large) and relief of regional brainstem compression and hydrocephalus. The goal was life saving. Neurological deficits such as hearing loss, facial weakness, or balance disorders were tolerated as simply part of the expected result. In the 1970s, the introduction of the operating microscope facilitated meticulous dissection of the tumor, making possible attempts at cranial nerve preservation. Over the ensuing 20 years, preservation of facial nerve continuity became more common than not. During the 1990s, hearing preservation became an achievable goal in selected cases. At the same time, improvements in anesthesia-related technique and wound closure reduced the risk of cerebellar infarction, meningitis, and CSF leakage. Nevertheless, these problems still continue to exist and CSF remains a significant problem after resection.

Stereotactic Radiosurgery

Stereotactic radiosurgery has become a common therapeutic choice for patients with acoustic tumors (VSs). Experience with radiosurgery now extends more than 30 years. During the late 1980s and early 1990s, patients and their doctors chose radiosurgery or resection based mainly on early outcomes data derived from limited patient series. In 1987 we began a prospective assessment of the response of patients with acoustic tumors to GKS. Both early and later (5–10–year) outcomes were determined by obtaining and evaluating serial neuroimaging or CT scanning between 1987 and 1991. Subsequently MR imaging was the modality of choice after a prospective comparison study confirmed the accuracy of MR imaging-based stereotactic targeting. Multiple isocenters were used to conform the radiation margin to the intra- and extracanalicular tumor components (Fig. 1). The 50% isodose line was used to cover the tumor margin in 88% of patients with solitary tumors. An initial tumor margin dose of 18 to 20 Gy was selected based on the initial experience reported by the Karolinska group. This dose was decreased to 16 to 18 Gy within the first 2 years and by 1992 was decreased further to a margin dose of 14 to 16 Gy. Repeated reevaluations of the cranial nerve response prompted further small decreases in dose to preserve cranial nerve function. The mean dose delivered to the margin in cases of both non-NF2 and NF2 tumors was 14 Gy, although the most commonly prescribed margin dose at the present time is 13 Gy, and this dose has been fairly constant for the past 9 years. Dose selection in individual patients was based on the following factors: tumor volume, history of surgery, hearing status, facial motor function, and patient wishes. After SRS, all patients received a single 40-mg dose of intravenous methylprednisolone and were discharged from hospital the next morning.

Follow-Up Evaluations. Serial imaging studies (MR imaging or CT scanning when MR imaging was contraindicated) were requested every 6 months for the first 2 years, annually for the next 2 years, and then biannually. Serial audiograms were obtained at 6 to 12–month intervals in patients with preserved hearing. Contrast-enhanced imaging studies were conducted to define the tumor response and to identify any peritumoral changes. Before and after SRS, each tumor was measured in five separate dimensions (three extracanalicular and two intracanalicular) by using a method previously reported. A significant imaging–documented change determined using this caliper technique was defined as a difference of ± 2 mm.

The Long-Term Experience. We continue to evaluate a cohort of 162 patients who have undergone follow-up evaluations for at least 10 years since their procedure. This study represented results of our initial techniques. The majority of irradiated acoustic tumors (~ 70%) decreased in size over time (Fig. 1). In nine patients tumors increased in size and all were identified within the first 3 years after SRS. Enlargement of the lesion represented either true neoplastic tumor growth (four cases) or tumor death with an expansion of the tumor margins as the central portion of the tumor became necrotic. In the latter five patients subsequent imaging studies confirmed tumor volume regression. Four patients underwent resection. No
Management options for acoustic neuroma

Fig. 1. Axial serial MR images obtained at SRS (left), 6 months posttreatment (center), and after 9 years of follow up (right). This 44-year-old man was treated with an 11-isocenter gamma knife plan to a margin dose of 14 Gy. He maintains normal facial nerve function and experienced a marked return of his energy within several months of the procedure.

Further increase in tumor volume was identified in any patient at additional follow-up examination. Patients returned to their routine activities immediately. In our 5 to 10-year review, three patients developed hydrocephalus and required placement of a ventriculoperitoneal shunt. All new or worsened post-SRS deficits occurred within 28 months of the procedure, and no patient described a treatment-related problem after the 3rd year.

Current Experience With Solitary Tumors. Refinements in technique followed a continued review of results. In 1991 we began to use MR imaging–based stereotactic planning because CT scanning–based planning did not demonstrate well the intracanalicular portion of the tumor. With the former, we could visualize the tumor and regional neural structures in greater detail. This facilitated the use of multiple small irradiation isocenters for more conformal SRS. With this type of SRS, the rate of cranial nerve morbidity dropped precipitously. Similarly, our analysis of hearing preservation in patients with NF2 showed significant gains.

One hundred ninety-two patients underwent GKS between 1992 and 1997 and were eligible for extended follow-up reviews. The maximum follow-up interval in this cohort was 65 months. The median tumor margin dose was 13 Gy. The actuarial 5-year clinical tumor control rate (defined as no need for additional treatment) was 97%. One patient underwent a resection 6 months after radiosurgery. Five-year actuarial rates of developing any facial weakness, facial numbness, hearing level preservation, and preservation of testable speech discrimination were 1.1 ± 0.8%, 2.6 ± 1.2%, and 71 ± 4.7%, and 91 ± 2.6%, respectively. At a tumor margin dose of 13 Gy or less, the rate of facial neuropathy was 0%, whereas above 13 Gy it was 2.5% (usually mild and transient). The tumor diameter did not significantly affect results. The authors of studies from other centers have reported similar results.

Neurofibromatosis Type 2. Serial neuroimaging studies of 45 tumors during a median 36-month follow up (range 6–120 months) demonstrated that 16 tumors (36%) had regressed, 28 tumors (62%) remained unchanged in size, and one tumor (2%) had progressed. Loss of central contrast within the tumor was observed in some patients and thought to reflect tumor necrosis.

At our last detailed review, the mean clinical follow-up period was 41 months (range 6–120 months). No patients exhibited improvement on their clinical examination after GKS. In 30 patients (67%) clinical status was maintained and in 15 patients (33%) some degree of clinical deterioration was demonstrated. Two patients (4%) died during the follow-up period of unrelated illnesses. The median Karnofsky Performance Scale score after GKS was 80. Thirty-five patients (78%) were able to perform normal daily activities at the time of the last examination (Karnofsky Performance Scale score ≥ 80).

Of the 14 tumors associated with useful hearing (Gardner–Robertson Grade I or II) at the time of SRS, six (43%) demonstrated no change in hearing class during the follow-up period. Eight other patients lost all functional hearing (defined as absent speech discrimination) at a mean of 6 months after SRS (range 3–15 months). The overall rate of hearing preservation was 43%. In 1992, we began to use MR imaging–guided stereotactic planning and increased numbers of smaller isocenters. By specifically dividing the population into patients treated prior to 1992 and those treated after 1992, the difference in hearing preservation again becomes apparent. Prior to 1992, five patients with useful hearing (Grade I or II) were treated. All patients subsequently lost speech discrimination. After 1992, nine patients with useful hearing underwent GKS. Six (67%) of the patients had hearing preservation at the time of last examination. Thirty-one tumors (69%) were associated with intact facial nerve function (House–Brackmann Grade I) at the time of GKS. The overall rate of facial nerve preservation (Grade I) was 81%. Thirty-six of the treated tumors were associated with intact trigeminal nerve function. Three patients (8%) experienced trigeminal distribution sensory loss at a mean of 5 months (range 4–5 months) after radiosurgery. One patient subsequently recovered all trigeminal function, whereas residual deficits developed in two. The overall rate of trigeminal nerve preservation was 94%.

Tumor Resection or SRS? There have been a number of patient-based surveys conducted to study outcomes after treatment for acoustic neuroma. One of the first was a survey of 541 patient members of the Acoustic Neuroma Association who provided data on tumor resection be-
between 1973 and 1983. Of these patients facial weakness was present in 62%, eye-related problems in 84%, depression in 38%, sleep disturbance in 26%, and speech or swallowing difficulties in 16%. More recently a larger survey of 1579 cases in which resections were performed between 1989 and 1994 found improved results that included a 44% rate of facial weakness, an 11% rate of CSF leakage, and persistent balance problems after 1 year in 9%. In approximately 8%, recurrent or residual tumor was revealed on follow-up imaging.37

Better results following resection have been documented since that time. Samii and Matthies32 and Gornley, et al.13 found that complete tumor removal was a frequent outcome. Neurological and systemic morbidity, however, remained present with 1% mortality rates and CSF fistula rates of 9.2 and 15%, respectively. Experienced surgical teams have reported significant reductions in postresection complication rates, although the incidence of specific problems such as CSF leakage have remained unchanged. Brennan, et al., reported that translabyrinthine approach–related leaks had a higher incidence of surgical repair than retrosigmoid approach-related leaks. For patients with large acoustic tumors (> 3 cm in extracanalicular diameter) and those with progressive neurological deficits that require brainstem decompression, surgical resection (total or subtotal) is the preferred option. We believe that a complete resection should be performed in such patients if possible, but not at the expense of sacrificing neurological function. Stereotactic radiosurgery can be considered for patients with intracanalicular, small- or medium-sized acoustic tumors because most such cases do not involve a rapidly progressive neurological syndrome. The initial symptoms caused by most acoustic tumors are not improved by resection.28

In a recent report, Martin, et al., evaluated quality of life in patients after tumor resection. They found a disparity between the patients’ reports and the physicians’ assessments of function, with decreases in physical functioning, general health, and social functioning after surgery. More severe balance functions led to worse social functioning (Table 1).

The long-term effects of both resection and SRS must be documented to assist in physician and patient decision making. Surprisingly little information has been published on long-term imaging-based outcomes after resection. Cerullo, et al., noted a 10% recurrence rate by 10 years following resection. Mazzoni, et al., reported a series of more than 100 patients in whom hearing preservation was attempted. The overall tumor recurrence rate was 8.1%. Although these papers are often criticized for their results, they represent an honest evaluation of longer-term imaging results. All groups should strive to obtain serial imaging studies. Post, et al., found that in four (7%) of 56 patients the resection was incomplete in their attempt at preserving hearing and that in three regrowth developed within 3 years. In the largest series, Samii and Matthies32 reported a complete resection in 98% of patients and documented recurrence in six of 880 without NF2. In our SRS series, 98% of the patients required no further surgery and in 94% there was imaging confirmation of persistent tumor control. Tumors that increased in size in the 1st or 2nd year after GKS did so usually in association with central tumor necrosis, with a small expansion of the tumor capsule. Most such tumors then regressed to a size smaller than that at baseline with longer follow up. Such transient expansion may be associated with transient retroauricular pain, perhaps due to regional dural inflammation. Recurrence or continued tumor growth may follow resection or SRS, and periodic neuroimaging studies should be obtained in all patients.

We believe that all patients with newly diagnosed, residual, or recurrent acoustic tumors (< 3 cm in extracanalicular diameter) are now suitable candidates for SRS.8,17 Larger tumors are not as good candidates because of the dose reduction necessary to reduce the rate of potential radiation-related side effects. Hearing preservation should be attempted in younger patients with good hearing, either with SRS or resection.31 In our first 3-year experience, we accepted elderly patients, those with concomitant medical problems that argued against resection, those with residual or recurrent tumors after resection, and those with preserved hearing function. By 1991 we began to offer radiosurgery to all patients with acoustic tumors regardless of age, surgical history, or symptoms. For older patients (age > 75 years) with small and minimally symptomatic tumors, we continue to advocate observational serial imaging–based evaluation.2,3,9

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<th>Issue</th>
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<td>new facial nerve dysfunction</td>
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* As reported by Martin, et al.
tion and 6 months following SRS. The time interval after irradiation was too short to indicate causation.14 In a second report investigators noted the development of a temporal lobe glioblastoma 7.5 years after radiosurgery for a nearby acoustic neuroma. The temporal lobe had received a low radiation dose.14 In contrast, one of our patients underwent initial management of a frontal lobe astrocytoma and years later developed an acoustic neuroma. Was the development of these tumors related in some oncogenetic way or were they radiation related? We believe the risk of developing a tumor years after radiosurgery is much less than that of mortality immediately after resection, and likely less than the risk of developing another tumor in another body location.

**Fractionated Radiotherapy.** In the last several years, a number of groups have used fractionated radiotherapy to treat patients with acoustic neuromas. This technique was developed when surgeons at several centers that used LINAC-based irradiation technology were not satisfied with the results or accuracy of their device after single-fraction irradiation (SRS). To decrease the cranial nerve deficits, they began to deliver radiation over multiple sessions (fractionation). The goal of this approach is to weaken precisely to the tumor, and the regional brain structures should be spared radiation. This is not the case when administering fractionated doses in which larger volumes of regional tissue are irradiated. We believe that any advantage in fractionation in limiting toxicity only makes sense if the target volume contains normal brain or nerve. Sophisticated stereotactic radiosurgical instruments allow regional brain or nerve to be spared because of frame-based, single-session, image guidance. We do not believe that fractionation provides any useful advantage over radiosurgical techniques that have been used for the last 10 years.

**Survey of Neurosurgeons on Acoustic Neuroma Management.** A survey was mailed to members of the Congress of Neurological Surgeons in July 2002. Six hundred sixty-three surgeons (30%) responded to the survey. There were four questions written on one page. Forty one percent of responders were between the age of 40 and 50 years (Table 2). Eighty percent of neurosurgeons (530) surveyed had either performed radiosurgery in a patient with an acoustic neuroma or had referred a patient for neurosurgery.

**Survey Case One.** Question: You are a 37-year-old neurosurgeon who presents with mild decreased hearing on one side. You have no tinnitus and no balance problems. Facial function is normal. An MR image reveals an intracanalicular acoustic neuroma and serial images have demonstrated a small amount of growth. Which management strategy would you choose for yourself? Observation; resection; SRS; or fractionated radiotherapy? (Fig. 2).

Response: The majority of surgeons (283 [43%]) stated that they would choose SRS for management of their small acoustic tumor. Only 122 surgeons (18%) stated that they would choose resection. Fractionated radiotherapy was chosen by 2% of responders. Interestingly, 240 surgeons (36%) stated that they would continue to observe their tumor rather than undergo any specific treatment at the time. It had been stated in the case presentation that serial images had already demonstrated a small amount of growth. This tumor had been observed and was increasing in volume. Nevertheless, approximately one third of responders continued to choose observation for a 37-year-old patient with a small but growing tumor.

We evaluated the age of the responding surgeon and compared this to the treatment chosen by that surgeon (Table 2). Across the age groups between 30 and 70 years,
at least twice as many neurosurgeons chose SRS for their tumor rather than resection. This is most pronounced in the younger surgeon age group (30–40 years), in which the number of surgeons choosing SRS over resection was fourfold higher. Observation, however, continued to be chosen by many. Although one might think that an older person might choose radiosurgery over resection, simply to avoid the risks of general anesthesia or the surgical exposure, this did not necessarily appear to be true. This case reflected the care of an actual neurosurgeon who had undergone GKS. He remains well 18 months following this procedure, maintaining a full practice. He has experienced no facial weakness or change in hearing.

**Survey Case Two.** Question: You are a 50-year-old neurosurgeon who presents with mild decreased hearing on one side. You have tinnitus but no balance problems. Fac- ial function is normal. An MR image reveals a left acoustic neuroma. Which management strategy would you choose for yourself? Observation; resection; SRS; or fractionated radiotherapy? (Fig. 3).

Response: In this scenario, the neurosurgeon had a medium-sized acoustic tumor that indented the middle cerebellar peduncle but did not compress the fourth ventricle. The tumor measured 22 mm in the maximum diameter. The minority of surgeons (6%) recommended continued observation for a tumor of this size. Resection was recommended by 347 surgeons (52%), whereas SRS was chosen by 261 surgeons (39%). Fractionated radiotherapy was only chosen by 3%. When the results were stratified by age, resection was the most popular choice across the groups between the age of 30 and 60 years. Radiosurgery, however, became more popular with advancing age of the survey group, passing resection as the most popular choice when the neurosurgeon was older than age 60 years. It appears that surgeons chose to undergo resection because of the larger volume of the tumor that indented the lateral surface of the brainstem. This patient was also a real neurosurgeon who had undergone SRS. He remains well 18 months after the procedure and the tumor has decreased in size. Facial function remains normal.

**CONCLUSIONS**

Patients with acoustic neuromas have several treatment options. Large tumors with significant brainstem compression usually require resection. For patients with small- or medium-sized tumors, SRS has become a common treatment, with excellent reported long-term results. Patients must be comfortable with the concept of tumor control rather than tumor removal. Most seem to be satisfied with this concept, if it allows them to avoid brain surgery. Surgeons should strive to educate their patients with information from the peer-reviewed literature. Confusion among patients exists because the information provided by internet sources, newsletters, support groups, and physicians has not always been validated and supported by outcomes data. Although we are asked to provide our opinions, our comments should not be based on myth, conjecture, training bias, or socioeconomic concerns.

**References**


![Fig. 2. Axial MR image obtained in a 37-year-old man with a right intracanalicular acoustic neuroma (Survey Case 1).](image)

![Fig. 3. Axial MR image obtained in a man with a left acoustic neuroma (Survey Case 2).](image)
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