Radiosurgery Practice Guideline Initiative

Stereotactic Radiosurgery for Patients with

Vestibular Schwannomas

Radiosurgery Practice Guideline Report #4-06

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This practice guideline, together with a report on "Vestibular Schwannoma Management" is an original guideline approved by the IRSA® (International RadioSurgery Association) Board of Directors and issued in May 2006.

Preface

Summary
The IRSA® (International RadioSurgery Association) Radiosurgery Practice Guideline Initiative aims to improve outcomes for vestibular schwannoma radiosurgery by assisting physicians and clinicians in applying research evidence to clinical decisions while promoting the responsible use of health care resources.

Disclaimer
This guideline is not intended as a substitute for professional medical advice and does not address specific procedures or conditions for any patient. Those consulting this guideline are to seek qualified consultation utilizing information specific to their medical situation. Further, IRSA does not warrant any instrument or equipment nor make any representations concerning its fitness for use in any particular instance nor any other warranties whatsoever.

KEY WORDS **• vestibular schwannomas • acoustic tumor • acoustic neuroma • stereotactic radiosurgery • Gamma Knife® • linear accelerator • proton beam • image guided • robotic • irradiation**

Consensus Statement

Objective

To develop a consensus-based radiosurgery practice guideline for vestibular schwannoma management to be used by medical and public health professionals.

Participants

The working group included physicians from the staff of major medical centers that provide radiosurgery.

Evidence

The first authors (AN/LDL) conducted a literature search in conjunction with the preparation of this document and development of other clinical guidelines. The literature identified was reviewed and opinions were sought from experts in the diagnosis and management of vestibular schwannoma, including members of the working group.

Consensus Process

The initial draft of the consensus statement was a synthesis of research information obtained in the evidence gathering process. Members of the working group provided formal written comments that were incorporated into the preliminary draft of the statement. No significant disagreements existed. The final statement incorporates all relevant evidence obtained by the literature search in conjunction with final consensus recommendations supported by all working group members.

Group Composition

The Radiosurgery Guidelines Committee is comprised of neurosurgeons, neurotologists, and radiation oncologists. Other physician specialties are incorporated as appropriate.

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Conclusions

Specific recommendations are made regarding target population, treatment alternatives, interventions and practices and additional research needs. Appropriate use of radiosurgery for patients with vestibular schwannoma is recommended.

This guideline is intended to provide the scientific foundation and initial framework for the person who has been diagnosed with a vestibular schwannoma. The assessment and recommendations provided herein represent the best professional judgment of the working group at this time, based on research data and expertise currently available. The conclusions and recommendations will be regularly reassessed as new information becomes available.

Stereotactic Radiosurgery

Intracranial **s**tereotactic **r**adiosurgery involves the use of precisely directed closed skull single session radiation to create a desired radiobiologic response within the intracranial target with minimal effects to surrounding structures or tissues. In the case of vestibular schwannoma, highly conformal, precisely focused radiation is delivered to the acoustic tumor in a single session under the direct supervision of a radiosurgery team. At Centers of Excellence, the radiosurgery team includes a neurosurgeon, a radiation oncologist, a physicist and a registered nurse. In some centers, a neurotologist is also a member of the primary team.

Vestibular Schwannoma: Overview and Surgical Management

Background

Vestibular schwannoma (also known as acoustic neuroma, acoustic neurilemoma, or acoustic neurinoma) is a slow-growing, intracranial extra-axial benign tumor that usually develops from the balance (vestibular) nerve or very rarely from the hearing (cochlear) nerve, supplying the inner ear. The tumor originates from an overproduction of Schwann cells, which normally wrap around nerve fibers to support and insulate nerves. As the tumor grows, it presses against the hearing and balance nerves, usually causing unilateral hearing loss, tinnitus (ringing in the ear) and imbalance/dizziness. Further tumor growth can interfere with the trigeminal nerve, resulting in facial numbness. A vestibular schwannoma can also press on the facial nerve giving rise to facial weakness and/or twitching. As these tumors increase in size, they eventually occupy a large portion of the cerebellopontine angle. If the tumor continues to grow, it will eventually press against the brainstem and the cerebellum, and will become life threatening or cause blockage of cerebrospinal fluid flow, a condition known as hydrocephalus.

Unilateral vestibular schwannomas affect only one ear. They account for approximately 8% of all tumors inside the skull; one out of every 100,000 individuals per year develops a vestibular schwannoma.[104] Symptoms may develop at any age but usually occur between the ages of 30 and 60 years. Unilateral vestibular schwannomas are not considered to be hereditary. Both males and females are equally affected by the disease.

Bilateral vestibular schwannomas affect both hearing nerves and are usually associated with a genetic disorder called neurofibromatosis 2 (NF2). Half of affected individuals inherit the disorder from an affected parent and half seem to be the first in their family to have a gene mutation. Each child of an affected parent has a 50% chance of inheriting the disorder. Bilateral vestibular schwannomas are a principal clinical feature of neurofibromatosis 2, although other manifestations, including peripheral neurofibromata, meningioma, glioma, and juvenile posterior subcapsular lens opacities, are often present as well. Spinal cord tumors, mainly ependymomas, frequently affect the patient. Two distinct phenotypes can be described, one severe (Wishart phenotype) and one mild (Gardner phenotype), corresponding respectively to distinct modifications of the gene and of distinct prognostic value. Most patients with NF2 present in late adolescence or early childhood.

Pathophysiology

The vast majority of vestibular schwannomas develop from the Schwann cell covering of the vestibular portion of the vestibulocochlear nerve.[47] The superior and inferior vestibular nerves appear to be the nerves of origin with about equal frequency. Very rarely schwannomas (less than 5%) arise from the cochlear portion of the vestibulocochlear nerve. Because vestibular schwannomas arise from the investing Schwann cell, tumor growth generally compresses vestibular fibers to the surface. Destruction of vestibular fibers is slow and gradual and the reduced vestibular function is compensated for by central cerebral mechanisms. Consequently, many patients experience little or no disequilibrium or imbalance. Once the tumor has grown sufficiently large to fill the internal auditory canal, it may continue to grow either by eroding or expanding the bone and/or by extending out into the cerebellopontine angle. Vestibular schwannomas, like other space-occupying lesions, produce symptoms by any of four recognizable mechanisms: (1) blockage of the cerebrospinal fluid spaces, (2) displacement of the brainstem, (3) compression of vessels or (4) compression of nerves.

Vestibular schwannomas can continue to grow until they reach 3–4 cm in intracranial size before symptoms of major mass effect develop. The facial nerve is quite resistant to the stretching imposed by tumor growth without clinically apparent deterioration of function until the tumor has reached a very large size. The cochlear and vestibular nerves, on the other hand, are much more sensitive to this stretching and compression so that even small tumors confined to the internal auditory canal may produce early symptoms in the form of hearing loss or vestibular disturbance. As the tumor approaches 1.5 cm in intracranial diameter, it generally begins to abut the lateral surface of the brainstem. Further growth can occur only by compressing or displacing the brain stem toward the contralateral side. A 2.0 cm tumor usually extends sufficiently far anteriorly and superiorly to compress the trigeminal nerve and sometimes produces facial hypoesthesia (impairment of sensitivity). Growth over 4.0 cm generally results in progressive effacement of the cerebral aqueduct and fourth ventricle with eventual development of hydrocephalus. However, other mechanisms may be responsible for the occasional development of hydrocephalus in tumors as small as 2.0 cm. A known factor of importance is the increase (up to 10–15 fold) of cerebrospinal fluid protein content in the presence of a vestibular schwannoma.[66,79]

Frequency

Clinically diagnosed acoustic neuromas occur in 0.7–1.0 people per 100,000 population. The true prevalence

of asymptomatic vestibular schwannomas is likely greater (7/10,000).[1] The observed incidence may be greater since an increasing number of small tumors are diagnosed using magnetic resonance imaging (MRI).[1,104]

Etiology

Both unilateral and bilateral vestibular schwannomas may form due to malfunction of a gene on chromosome 22, which produces a protein (schwannomine/merlin) that controls the growth of Schwann cells. In NF2 patients, the faulty gene on chromosome 22 is inherited and is present in all or most somatic cells. However, in individuals with unilateral vestibular schwannoma, for unknown reasons this gene loses its ability to function properly and is present only in the schwannoma cells.[47]

Symptoms

A progressive decline in unilateral hearing is the most common symptom that leads to the diagnosis of a vestibular schwannoma.[41] The tumor can produce hearing loss through either direct progressive injury to the cochlear nerve (slowly progressive sensorineural hearing loss) or interruption of cochlear blood supply (sudden and fluctuating hearing losses).[10] A significant number of individuals with vestibular schwannomas have a reduction in speech discrimination disproportionate to the reduction in the hearing threshold (pure-tone average). This is consistent with direct injury to the cranial nerve. However, many patients with smaller vestibular schwannomas have normal or near-normal hearing based on speech discrimination scores. There is no strict relationship between the size of the tumor and the quality of the residual hearing. Hearing loss associated with vestibular schwannoma can be sudden or fluctuating in 5–15% of patients. Such hearing loss, usually referred to as "sudden deafness", may improve spontaneously or in response to corticosteroid therapy.

Three to five percent of patients with vestibular schwannoma have normal hearing at the time of diagnosis. The presence of unilateral tinnitus alone is a sufficient reason to evaluate an individual for a vestibular schwannoma. Vertigo and disequilibrium are uncommon presenting symptoms among patients with these tumors. Rotational vertigo (the illusion of movement or falling) is more common when tumors are small. On the other hand, dysequilibrium (a sense of unsteadiness or imbalance) appears to be more common in larger tumors. Overall, approximately 40–50% of patients with vestibular schwannomas report some balance disturbance. However, such disturbance is the presenting symptom in less than 10% of patients. The gradual reduction of vestibular function in general is well compensated for by central mechanisms.

Headaches are present in 50–60% of patients at the time of diagnosis, but fewer than 10% of patients have headache as their presenting symptom. Headache appears to become more common as tumor size increases and is a prominent feature in patients who develop hydrocephalus associated with a large tumor.

Facial numbness occurs in about 25% of patients and is more common at the time of presentation than facial weakness (about 10% of patients). A decrease in the corneal reflex generally occurs earlier and more commonly than documented facial hypoesthesia. Even though approximately 50–70% of individuals with large tumors have facial hypoesthesia on neurological examination, they are often unaware of it, and it is rarely the presenting symptom.

As facial weakness is a sufficiently uncommon phenomenon occurring in 5–10% of patients, the development of facial weakness associated with a small or medium-size tumor should raise suspicion that the diagnosis is not compatible with a vestibular schwannoma. Other diagnoses, such as facial neuroma, meningioma, epidermoid tumor, arteriovenous malformation (AVM) or lipoma should be excluded. Larger tumors can obstruct the flow of cerebrospinal fluid through the ventricular system. In the early decades of the 20th century, 75% of patients presented with hydrocephalus.

Clinical study of the nervus intermedius component often shows dysfunction. If specifically asked, the patient will often notice a change in lacrimation with an eye dryness that can be documented by a Schirmer test. Patients may report dysguesia with a feeling of a metallic taste. Examination may show hypoesthesia in the most superficial part of the external auditory canal. In a population of 46 patients tested by a Schirmer test before radiosurgery, Tamura et al found that 41% of patients experienced some lacrimal deficit on the side of the tumor.[103]

Course

Overall, three separate growth patterns can be distinguished for vestibular schwannomas: (1) no or very slow growth, (2) slow growth (i.e., 0.2 cm/year linear growth on imaging studies), and (3) fast growth (i.e., >1.0 cm/year). While most vestibular schwannomas grow slowly, some grow quite quickly and can double in volume within six months to a year.[11] While most tumors adhere to one or another of these growth patterns, a small number of tumors appear to alternate between periods of no or slow growth and rapid growth. Cystic vestibular schwannomas are sometimes capable of relatively rapid expansion because of filling of their cystic component. Spontaneous intratumoral hemorrhage has been rarely described but mainly occurred in cases of large multicystic tumors.[99]

Imaging Studies

The definitive diagnostic test for patients with vestibular schwannomas is high-resolution, thin slice, gadolinium-enhanced MR imaging, supplemented with 3D volume, long TR images through the internal auditory canal. For patients such as those with pacemakers, who cannot have MRI, CT with contrast remains the imaging procedure of choice even though the smallest intracanalicular tumors cannot be diagnosed with this technique.

Management

Early diagnosis of a vestibular schwannoma is key to preventing its serious consequences. There are three primary options for managing a vestibular schwannoma: (1) surgical removal, (2) radiosurgery and (3) observation with serial imaging studies. In addition, conformal fractionated radiation therapy using linear accelerators or proton beam may be considered.

Observation with Serial Imaging

In some cases, usually elderly or medically infirm patients or individuals with very small tumors, it may be reasonable to "watch" the tumor for potential growth.[33] Repeat MRI scans over time are used to carefully monitor the tumor for any growth.[106] The object of serial observation is that treatment is to be considered when there are signs of growth.[33,74]

Surgical Removal
A variety of surgical approaches can be used to remove vestibular schwannomas. Three main approaches in general use are the retrosigmoid, translabyrinthine and middle fossa approaches.[29,30] A variety of different considerations go into deciding which approach should be used for any individual patient. These variables include preoperative hearing levels in both ears, tumor size and location, age of patient, and patient and surgeon preference. All approaches have advantages and disadvantages as indicated below.

Retrosigmoid (Suboccipital) Approach
Advantages

- The retrosigmoid approach provides the best wide-field visualization of the posterior fossa. The time needed for tumor exposure is short. The inferior portions of the cerebellopontine angle and the posterior surface of the temporal bone anterior to the porus acousticus are more clearly observed than in the translabyrinthine approach. Panoramic visualization is especially helpful when displacement of nerves is not predictable, as occurs commonly with meningiomas.

- Hearing conservation surgery can be attempted even for relatively large tumors through the retrosigmoid approach. Destruction of the labyrinth is not required as part of the retrosigmoid approach.

Disadvantages

- The retrosigmoid approach may require cerebellar retraction or resection, which can lead to the development of postoperative edema, hematoma, infarction and bleeding.

- The retrosigmoid approach is associated with a greater likelihood of protracted postoperative headache.

- The highest incidence of tumor recurrence or incomplete resection occurs with retrosigmoid approaches [76,85] due to poor control of the fundus of the internal auditory canal.

- It may be difficult to perform a facial-to-facial nerve graft repair from this approach.

Translabyrinthine Approach
Advantages

- The translabyrinthine approach provides the best view of the lateral brain stem facing the vestibular schwannoma.

- Retraction of the cerebellum is not needed.

- The fundus and lateral end of the internal auditory canal are completely exposed; the facial nerve can be identified at a location where it is undistorted by tumor growth and compressed into the labyrinthine segment, decreasing the risk of delayed postoperative facial nerve palsy.

- Possible opening of the tentorium in case of an extra large tumor that may extend upward.

- Safe control of the lateral surface of the pons.

- Improved surgical comfort.

- No need for a seated or lateral positioning of the patient.

Disadvantages

- Hearing sacrifice is complete and unavoidable.

- The inferior portions of the cerebellopontine angle and cranial nerves are not as well visualized as they are in the retrosigmoid approach. The temporal bone anterior to the porus acousticus is also less well visualized.

- A tissue graft (fat, fascia, homograft or synthetic dura) is required. Some surgeons prefer hydroxyapatite cement.

- The sigmoid sinus may be more vulnerable to injury.

- Need to extend the approach in case of contract mastoid or high jugular bulb position.

- The third portion of the facial nerve may be vulnerable to injury during the approach.

- The approach requires additional time.

Middle Cranial Fossa Approach
Advantages

- It is the only procedure that fully exposes the lateral third of the internal auditory canal without sacrificing hearing.

Disadvantages

- The facial nerve generally courses along the anterior superior portion of the tumor. Depending on whether the tumor is of superior or inferior vestibular nerve origin, the facial nerve may have to be dissected and displaced to obtain access to the tumor. This may make the nerve more vulnerable to injury during tumor removal.

- The risk of dural laceration becomes increasingly more likely as patients become older. The dura mater in elderly patients is thin and more friable. This becomes especially noticeable beyond the seventh decade of life.

- The approach provides only limited exposure of the posterior fossa but can be extended by an additional anterior petrosectomy following the Kawase procedure.

- The operation is technically challenging.

- Some patients incur postoperative trismus related to manipulation and/or injury to the temporalis muscle.

- The temporal lobe must be retracted, which on rare occasion leads to temporal lobe injury.

- The great superficial petrosal nerve may be injured during the elevation of the temporal dura.

Functional Outcome of Microsurgery

Facial Function

Preservation of facial function continues to improve. However, facial nerve outcomes vary according to tumor size and operator experience.[84] When tumors are smaller than 1.5 cm, good facial nerve function can be expected (House-Brackmann grades I–II) in more than 90% of patients at Centers of Excellence. Only 3.2–6.7% of patients with this size tumor have poor outcomes (House-Brackmann grades III–IV). In addition to tumor size, preoperative electrophysiologic testing can help predict postoperative outcome, although this testing is not commonly used.[56] The overall facial nerve preservation rate is 80%.[63] However, facial nerve function (grades I and II) can be preserved in only 40–50% of patients with large (>4 cm diameter) tumors.[110] Injuries of the nervus intermedius are underestimated because this nerve is rarely assessed preoperatively.[39]

Hearing Outcome

The ability to preserve hearing has increased substantially over the last couple of decades. Depending on criteria used for successful hearing conservation, hearing preservation has been reported in 30–80% of patients considered eligible for hearing preservation surgery.[82] Meta-analysis performed by Gardner and Robinson showed an overall average success rate of about 33%.[27] Delayed hearing deterioration may occur after surgery in 30–50% of patients who originally had successful hearing preservation.[12,92] In various studies, serviceable hearing preservation rates of 8–57%[12,57,85] using the retrosigmoid approach and 32–68%[85] using the middle fossa approach have been reported.

Tinnitus

Tinnitus becomes worse in 6–20% of individuals after tumor removal. In the majority of individuals, tinnitus remains unchanged. Approximately 25–60% of patients experience a decrease in tinnitus. Of patients without preoperative tinnitus, 30–50% developed it in the immediate postoperative period.

Complications

Cerebrospinal fluid leakage through either the wound or the Eustachian tube and middle ear occurs in 2–20% of patients.[3,86,90,95] Although in individual published reports the cerebrospinal fluid leak rate appears higher with the retrosigmoid approach (2.9–18%),[85] recent metaanalysis suggests similar rates of cerebrospinal fluid leak for all surgical approaches (10.6% of 2,273 retrosigmoid surgeries; 9.5% of 3,118 translabyrinthine surgeries; and 10.6% of 573 middle fossa surgeries). While this leakage in general ceases spontaneously, it may sometimes necessitate re-operation to seal the source of the leakage, which otherwise may result in meningitis. The adjunctive use of endoscopy may offer some advantages including a lowered risk of CSF leakage.[105] Other perioperative complications include death (0–3%),[20,83] intracranial hematomas (1–2%), subcutaneous hematoma (3%), cerebellar and brainstem edema, hemiparesis, meningitis (1.2%), wound infections (1.2%), VI nerve paresis (1-2%), and other lower cranial nerve paresis.[83,86]

Recurrence/Residual Tumor

Overall, the recurrence rate of 5–10% is quoted in published literature, although a few studies report no long term recurrence after translabyrinthine approach.[89] However, incomplete resection of vestibular schwannomas is associated with a significant risk of recurrent tumor requiring subsequent intervention.[19]

Patients' Perspective on Surgical Outcome

A variety of complications have been reported after vestibular schwannoma surgery.[6,9,17,18,28,35,97] In a study by Bateman et al, patients' subjective condition after surgery for a vestibular schwannoma was classified either as impairment (141, 51%), disability (95, 34%) or handicap (43, 15%).[5] Most of the impairments were related to problems with facial nerve function. The other most common issues were "balance problems" (19/141, 13%) followed by "hearing loss" (17/141, 12%) and "difficulty with background noise" (14/141, 10%). Tinnitus accounted for 5 of 141 responses (4%). Disabilities resulting from facial nerve dysfunction accounted for most of the disabilities reported by patients. A significant number of disabilities were associated with balance problems (e.g. "unable to drive," "problems changing direction," "unable to swim, cycle, run, climb steps, do aerobics," and "problems bending down") and with hearing loss (e.g. "difficulty locating the source of sounds," "difficulty following conversations in a crowd," "unable to hear people to one side" and "unable to hear doorbell/telephone"). With respect to handicap, social isolation emerged as a strong theme. Fifteen out of forty-three responses (35%) were "reluctance to attend large social gatherings." Employment-related problems were also important with 7 of 43 responses (16%). In a retrospective study (541 Acoustic Neuroma Association members), Wiegand & Fickel stressed that eye-related problems were experienced by 84% of respondents. Each respondent was asked to characterize the most difficult aspect of his experience. Thirty percent said that it was the change in appearance, 19% the hearing loss, 16% the loss of independence and 14% the eye problems. Of interest, 38% of patients experienced depression, 18% dental problems, and 10% sexual dysfunction.[108]

Stereotactic Radiosurgery

Vestibular schwannoma stereotactic radiosurgery using the Gamma Knife® was first performed by Leksell in 1969.[49] During the past decade radiosurgery has emerged as the effective alternative to surgical removal of small to moderate-sized vestibular schwannomas. The evolution of radiosurgery has impacted the management algorithm for the treatment of these tumors. The long-term results have established radiosurgery as an important minimally invasive alternative to microsurgery. Advanced dose planning software, MRI-guided dose planning and dose optimization over the past 20 years reflect the evolution of this technology. The recent introduction of robotics (automated positioning systems) as part of the Gamma Knife® has further improved conformality (ability to conform dose to the target) and selectivity (ability to reduce dose to surrounding structures) of dose plans for vestibular schwannomas. Other imaged guided linear accelerator devices (Trilogy®, SynergyS®, Novalis® and CyberKnife®) use robotics to deliver conformal stereotactically-delivered hypofractionated radiation therapy, usually in 3–5 sessions. Proton beam technology is also used to deliver hypofractionated radiation therapy. The goals of vestibular schwannoma radiosurgery are to prevent further tumor growth, preserve cochlear and other cranial nerve function where possible, and maintain or improve the patient's neurological status.

Radiosurgery Technique for Vestibular Schwannomas

Pre-Radiosurgery Evaluation

Patients with vestibular schwannomas are evaluated with high resolution MRI (CT may be substituted in patients who cannot undergo MRI scans) and audiological tests that include pure tone average (PTA) and speech

discrimination score (SDS) measurements. Hearing is graded using the Gardner-Robertson modification of the Silverstein and Norell classification and/or the American Academy of Otolaryngology-Head and Neck Surgery guidelines, and facial nerve function is assessed according to the House-Brackmann grading system. "Serviceable" hearing (Class I and II) is defined as a PTA or speech reception threshold lower than 50 dB and speech discrimination score better than 50%. The Committee on Hearing and Equilibrium of the American Academy of Otolaryngology-Head and Neck Surgery has established guidelines for reporting vestibular schwannoma results. In this classification hearing loss at a higher frequency (3000 Hz) is also included in calculating the PTA. "Serviceable" hearing (Class A and B) is similar to class I and II of Gardner-Robertson hearing classes.

Radiosurgery Technique

Radiosurgery can be performed using the Gamma Knife®, modified LINACs or the proton beam. Techniques of head frame fixation, stereotactic imaging, dose planning and dose delivery are different for these three modalities. In Gamma Knife® radiosurgery the procedure begins with rigid fixation of an MRI compatible Leksell stereotactic frame (model G, Elekta Instruments, Atlanta, GA, USA) to the patient's head. Local anesthetic scalp infiltration (5% Marcaine and 1% Xylocaine) is used, supplemented by mild intravenous sedation as needed. High-resolution images are acquired with a fiducial system attached to the stereotactic frame. For vestibular schwannoma radiosurgery, a 3-D volume acquisition MRI using a gradient pulse sequence (divided into 1 or 1.5 mm thick 28–36 axial slices) is performed in order to cover the entire lesion and surrounding critical structures. A T2 weighted 3-D volume sequence is performed to visualize cranial nerves and delineate inner ear structures (cochlea and semicircular canals). Stereotactic images are transferred via a fiberoptic ethernet to the GammaPlan® dose planning computer where images are first checked for any distortion or inaccuracy. Planning is performed on narrow slice thickness axial MR images with coronal and sagittal reconstructions. Centers using LINAC or proton beam systems may use mask immobilization of the patient's head along with image guidance and typically deliver the radiation dose in three or more stages. CT is used for planning but may be fused to MRI scans.

Radiosurgical Dose Planning

Dose planning is a critical aspect of radiosurgery. Complete coverage of the tumor and preservation of facial, cochlear and trigeminal nerve function is given priority during dose planning. For large tumors, preservation of brainstem function is also a consideration. A conformal radiosurgery plan is necessary for hearing and facial nerve preservation. Highlights of Gamma Knife® vestibular schwannoma radiosurgery planning include outlining of tumor volume, use of multiple isocenters, beam weighting and use of plug patterns. Precise three-dimensional conformality between treatment and tumor volumes is needed to avoid radiation related complications.[50] This degree of conformality can be achieved through complex multiisocenter planning. Vestibular schwannoma planning is usually performed using a combination of small beam diameter (4- and 8-mm) collimators. For large tumors, 14 mm collimators are also used. A series of 4 mm isocenters are used to create a tapered isodose plan to conform to the intracanalicular portion of the tumor. Success of vestibular schwannoma radiosurgery depends upon high conformality to the tumor margin. Because the facial and the cochlear nerve complex generally courses along the anterior margin and anterior-inferior side of the tumor, the dose plan should by highly conformal in this region. LINAC based systems may use robotics and multileaf collimators to deliver the photon radiation.

Dose Prescription

After optimizing the plan, a maximum dose to the target is determined. The treatment isodose, maximum dose and dose to the margin are jointly decided by a neurosurgeon, radiation oncologist, medical physicist and, in some centers, a neurotologist after considering the goal of radiosurgery in an individual patient and the tolerance of the surrounding structures. In Gamma Knife® radiosurgery a dose of 12–13 Gy is typically prescribed to the 50% (or other) isodose line that conforms to the tumor margin. Dose prescription for vestibular schwannomas has changed significantly over the past 10 years. A margin dose of 12–13 Gy is associated with a low complication rate and yet maintains a high rate of tumor control. It is unclear how much, if any, of the radiosurgery dose prescription should be lowered, since the tumor control rate may suffer. Most centers are reluctant to prescribe lower margin doses (such as 12 Gy) for vestibular schwannomas including those that have recurred after apparently successful complete excision because these tumors may be more aggressive and patients usually do not have serviceable hearing. Lower radiosurgery doses may be a better management strategy for patients with bilateral NF2 vestibular schwannomas or patients with contra-lateral deafness from other causes, for whom hearing preservation may be more critical. After prescribing the margin dose, the fall off on cochlea, semicircular canal and brainstem are checked to keep them below tolerance level. For LINAC based treatments using a frameless system the total dose may be divided into 3–5 delivery sessions, typically prescribed to the 80% isodose line, using a total mean dose ranging up to 17 Gy.[37] The higher dose is an issue as the risk of complications is directly related to the dose and treatment volume.

Dose Delivery

Gamma Knife® radiosurgery is performed with a 201 source, cobalt-60 unit (model B or model C, Elekta Instruments, Atlanta, GA) where the patient's head and stereotactic frame are immobilized within the appropriate collimator helmet at a calculated target coordinate. Dose delivery is accomplished in a single session by positioning the head serially for each subsequent isocenter until a fully conformal field encompasses the tumor volume. Radiosurgery using modified LINACs is typically performed with micromultileaf collimators or a cone system. Dedicated LINAC stereotactic radiotherapy treatments use a robotically or automatically directed LINAC. Cyclotron facilities use a specially modulated proton beam to deliver energy to the target.

Postoperative Care

There is no consensus on the use of corticosteroids on the day of radiosurgery. Some do not use steroids at all before, during or after radiosurgery. At some centers patients receive an intravenous dose of 40 mg of methylprednisolone at the onset or conclusion of the procedure. At other centers, 6 mg of dexamethasone is given immediately before dose delivery and is repeated every three hours for the duration of the treatment. The stereotactic frame is removed immediately after radiosurgery. Patients are observed for a few hours in the same day surgery unit and are usually discharged within 24 hours.

Post-Radiosurgery Evaluations

After radiosurgery, all patients are followed up with serial gadolinium-enhanced MRI scans, which are generally requested following a schedule such as 6 months, 12 months, and 2, 4, 8 and 16 years. All patients who have some preserved hearing are advised to obtain audiological tests (PTA and SDS) near the time of their MRI follow-ups.

Gamma Knife Radiosurgery: Clinical Results

Tumor Growth Control

Long-term results of Gamma Knife® radiosurgery for vestibular schwannomas have been documented[14,22,32,42,45,55]. Recent reports suggest a tumor control rate of 93–100% after radiosurgery.[14,16,21-24,31,32,34,36,37,42-45,50-52,54,55,61,67,68] Kondziolka et al studied 5 to 10-year outcomes in 162 vestibular schwannoma patients who had radiosurgery at the University of Pittsburgh.[44] In this study a long-term 98% tumor control rate was reported. Sixty-two percent of tumors became smaller, 33% remained unchanged, and 6% became slightly larger. Some tumors initially enlarged 1–2 mm during the first 6 to 12 months after radiosurgery as they lost their central contrast enhancement. Such tumors generally regressed in volume compared to their pre-radiosurgery size. Only 2% of patients required tumor resection after radiosurgery. Norén, in his 28-year experience with vestibular schwannoma radiosurgery, reported a 95% long-term tumor control rate. Litvack et al reported a 98% tumor control rate at a mean follow-up of 31 months after radiosurgery using a 12 Gy margin dose.[53] Niranjan et al analyzed the outcome of intracanalicular tumor radiosurgery performed at the University of Pittsburgh.[65] All patients (100%) had imaging-documented tumor growth control. Flickinger et al performed an outcome analysis of acoustic neuroma patients treated between August 1992 and August 1997 at the University of Pittsburgh. The actuarial 5-year clinical tumor control rate (no requirement for surgical intervention) was 99.4 $\pm$ 0.6%.[21,22] The long-term (10–15 year) outcome of benign tumor radiosurgery has been evaluated. In a study which included 157 patients with vestibular schwannomas**,** the median follow-up for the patients still living at the time of the study (n=136) was 10.2 years. Serial imaging studies after radiosurgery (n=157) showed a decrease in tumor size in 114 patients (73%), no change in 40 patients (25.5%), and an increase in three patients who later had resection (1.9%).[45] No patient developed a radiation-associated malignant or benign tumor (defined as a histologically confirmed and distinct neoplasm arising in the initial radiation field after at least two years have passed).

Hearing Preservation

Pre-radiosurgery hearing can now be preserved in 60–70% of patients, with higher preservation rates found for smaller tumors. In a long-term (5–10 year follow-up) study conducted at the University of Pittsburgh, 51% of patients had no change in hearing ability.[21,44] All patients (100%) who were treated with a margin dose of 14 Gy or less maintained a serviceable level of hearing after intracanalicular tumor radiosurgery.[65] Among patients treated after 1992, the 5-year actuarial rates of hearing level preservation and speech preservation were 75.2% and 89.2%, respectively, for patients (n=89) treated with a 13 Gy tumor margin dose. The 5-year actuarial rates of hearing level preservation and speech preservation were 68.8% and 86.3%, respectively, for patients (n=103) treated with >14 Gy as the tumor margin dose.[22]

Unlike microsurgery, immediate hearing loss is uncommon after radiosurgery. If hearing impairment is noted, it occurs gradually over 6 to 24 months. Early hearing loss after radiosurgery (within three months) is rare and may result from neural edema or demyelination. The exact mechanism of delayed hearing loss after radiosurgery is still unclear. Perhaps gradual obliteration of microvessels or even direct radiation axonal or cochlear injury is implicated. The effect of radiation on normal microvessels supplying the cochlear nerve or cochlea itself is not known. However, with doses as low as 12–13 Gy (which are sufficient to halt the tumor growth) vascular obliteration of normal vessels seems less likely. This dose probably does not adversely affect the vessels as well as the axons. Although with current imaging techniques the cochlear nerve cannot be well visualized, efforts should be made to achieve high conformality at anterior and inferior margin of the tumor. Conformal dose planning using 4 mm collimators for the intracanalicular portion of the tumor may prevent further injury to the cochlear nerve. It is likewise important to avoid radiation of the cochlea.[70]

Facial Nerve and Trigeminal Nerve Preservation

Facial and trigeminal nerve function can now be preserved in the majority of patients (>95%). In the early experience at University of Pittsburgh normal facial function was preserved in 79% of patients after five years and normal trigeminal nerve function was preserved in 73%. These facial and trigeminal nerve preservation rates reflected the higher tumor margin dose of 18–20 Gy used during the CT based planning era before 1991. In a recent study using MR based dose planning, a 13 Gy tumor margin dose was associated with 0% risk of new facial weakness and 3.1% risk of facial numbness (5-year actuarial rates). A margin dose of >14 Gy was associated with a 2.5% risk of new onset facial weakness and a 3.9% risk of facial numbness (5-year actuarial rates).[22] None of the patients who had radiosurgery for intracanalicular tumors developed new facial or trigeminal neuropathies.

Neurofibromatosis 2

Patients with vestibular schwannomas associated with neurofibromatosis 2 represent a special challenge because of the risk of complete deafness. Unlike the solitary sporadic tumors that tend to displace the cochlear nerve, tumors associated with NF2 tend to form nodular clusters that engulf or even infiltrate the cochlear nerve. Complete resection may not always be possible. Radiosurgery has been performed for patients with NF2. Subach et al studied 40 patients (with 45 tumors) who were treated with radiosurgery for NF2. Serviceable hearing was preserved in 6 of 14 patients (43%), and this rate improved to 67% after modifications made to the technique in 1992. The tumor control rate was 98%.[98] Only one patient showed imaging documented growth. Normal facial nerve function and trigeminal nerve function was preserved in 81% and 94% of patients, respectively. In two recent series,[78,80] serviceable hearing was preserved in only 30%[78] and 40%[80] of cases, respectively. The tumor control rate was respectively 71%[78] and 79%.[80] It now appears that preservation of serviceable hearing in patients with NF2 is an attainable goal with modern radiosurgery technique, and some centers propose this early treatment when the hearing level is still excellent.

Proton Beam Radiosurgery: Clinical Results

Weber et al. evaluated 88 patients with vestibular schwannomas treated with proton beam stereotactic radiosurgery, in which two to four convergent fixed beams

of 160-MeV protons were applied.[107] A median dose of 12 cobalt Gy equivalents was prescribed to the 70–108% isodose lines (median, 70%). The median follow-up period was 38.7 months. The actuarial 2- and 5-year tumor control rates were 95.3% and 93.6%. Serviceable hearing was preserved in 33.3% of patients. Actuarial 5-year normal facial and trigeminal nerve function preservation rates were 91.1% and 89.4%. Harsh et al evaluated 68 patients with vestibular schwannomas who were treated with proton beam radiosurgery using a marginal dose of only 12 Gy.[31] After a mean clinical follow-up of 44 months and imaging follow-up of 34 months, actuarial control rates of 94% at two years and 84% at five years were reported. Cranial neuropathies included persistent facial hypoesthesia (4.7%), intermittent facial paresthesias (9.4%), persistent facial weakness (4.7%) requiring oculoplasty, transient partial facial weakness (9.4%) and synkinesis (9.4%).

LINAC Radiosurgery: Clinical Results

Suh et al. evaluated 29 patients treated with a modified linear accelerator stereotactic radiosurgery system.[100] Median margin dose was 1,600 cGy. The 5-year local disease control rate was 94%. Long-term complications included new or progressive trigeminal and facial nerve deficits with estimated 5-year incidences of 15% and 32%, respectively. Subjective hearing reduction or loss occurred in 14 of the 19 patients (74%) who had useful hearing prior to treatment. Since there was a high risk of cranial nerve neuropathy, these authors did not recommend using only computed tomography-based planning and high prescription doses. Spiegelman et al reported their results of LINAC radiosurgery for 44 patients with vestibular schwannoma.[96] After a mean follow-up period of 32 months (range, 12–60 months), 98% of the tumors were controlled. The actuarial hearing preservation rate was 71%. New transient facial neuropathy developed in 24% of the patients and persisted to a mild degree in 8%.

Stereotactic Radiation Therapy: Clinical Results

Stereotactic radiation therapy (SRT) or fractionated stereotactic radiation therapy (FSRT) refers to the delivery of a standard fractionation scheme of radiation, used with rigidly applied or relocatable stereotactic guiding devices. Some LINAC based radiosurgery centers (driven by the desire to reduce complication rates) have shifted to fractionated stereotactic radiotherapy for vestibular schwannomas.[37,60,69,81,88,94,100,102] Ishihara et al reported 94% tumor control rate at a median follow-up of 31.9 months in a series of 38 patients who had CyberKnife® SRT for vestibular schwannoma. One patient developed transient facial paresis (2.6%) and one developed trigeminal nerve neuropathy (2.6%).[37] Fuss et al described 51 patients with vestibular schwannomas who were treated with SRT.[25] Mean follow-up was 42 months and the actuarial 5-year tumor control rate was 95%. One patient developed transient facial nerve paresis and two noted new trigeminal dysesthesias. Chung et al, using SRT for 25 patients with useful hearing, reported 57% hearing preservation at two years.[13] The mean pre- and post-SRT speech recognition threshold was 20 and 38 dB, respectively. The mean proportion of pre- and post-SRT speech discrimination was 91% and 59%, respectively. Sawamura et al treated 101 patients with vestibular schwannoma using fractionated SRT at a radiation level of 40–50 Gy, administered in 20–25 fractions over a 5- to 6-week period.[88] The median follow-up period was 45 months, and the actuarial 5-year rate of tumor control was 91.4%. The actuarial 5-year rate of useful hearing preservation (Gardner-Robertson Class I or II) was 71%. The complications of fractionated SRT included transient facial nerve palsy (4%), trigeminal neuropathy (14%) and balance disturbance (17%). Eleven patients (11%) who had progressive communicating hydrocephalus after FSRT required a shunt. Meijer et al performed a single-institution trial to study whether fractionated stereotactic radiation therapy is superior to single-session LINAC-based radiosurgery with respect to treatment-related toxicity and local control in patients with vestibular schwannoma.[59] These authors analyzed 129 vestibular schwannoma patients who were treated at a LINAC-based radiosurgery facility. Stereotactic radiation therapy was performed on 80 patients with a relocatable guidance device using 5 x 4 Gy and later 5 x 5 Gy at the 80% isodose. Forty-nine patients had stereotactic radiosurgery of 1 x 10 Gy and later 1 x 12.5 Gy at the 80% isodose using a stereotactic frame. There was no statistically significant difference between the single-fraction group and the fractionated group with respect to mean tumor diameter (2.6 vs. 2.5 cm) or mean follow-up time (both 33 months). Outcome differences between the single-session group and the fractionated treatment group with respect to 5-year local control probability (100% vs. 94%), 5-year facial nerve preservation probability (93% vs. 97%), and 5-year hearing preservation probability (75% vs. 61%) were not statistically significant. The difference in 5-year trigeminal nerve preservation (92% vs. 98%) reached statistical significance ($p = 0.048$). These authors concluded that LINAC-based radiosurgery was as good as LINAC-based fractionated stereotactic radiation therapy in vestibular schwannoma patients, except for a small difference in trigeminal nerve preservation rate in favor of a fractionated schedule. At the present time there are limited data on SRT for vestibular schwannomas.[15] There are no compelling radiobiological principles supporting the use of SRT over single session radiosurgery for achieving an optimal therapeutic response for the slowly proliferating, late-responding tissue of a schwannoma. The long-term results (5–10 years) of SRT are not yet available. For some centers, SRT may be an option for vestibular schwannomas if they have a higher complication rate using LINAC radiosurgery.

Comparison of Radiosurgery and Microsurgery Options

A recent review of English language literature published over 23 years (110 articles) found no level 1 or 2 evidence to support either surgical resection or radiosurgery and highlighted the need for properly designed studies.[64] However, it must be recognized that a randomized clinical trial will probably never be successfully completed to compare surgical resection with radiosurgery for vestibular schwannoma. Several carefully performed retrospective studies have compared the results of microsurgery and stereotactic radiosurgery. Karpinos et al analyzed 96 patients with unilateral acoustic neuromas treated with the Leksell Gamma Knife® or microsurgery and concluded that radiosurgery was associated with a lower rate of immediate and long-term development of facial and trigeminal neuropathy, postoperative complications, and hospital stay. Radiosurgery yielded better measurable hearing preservation than microsurgery and equivalent serviceable hearing preservation rate and tumor growth control.[40] Pollock et al studied 87 patients with unilateral, previously unoperated vestibular schwannomas with an average diameter of less than 3 cm treated at the University of Pittsburgh between 1990 and 1991.[72] Preoperative patient characteristics and average

tumor size were similar between the treatment groups. Microsurgical or radiosurgical techniques were used by experienced surgeons in both treatment groups. The treatment groups were compared based on cranial nerve preservation, tumor control, postoperative complications, patient symptomatology, length of hospital stay, total management charges, effect on employment status and overall patient satisfaction. Stereotactic radiosurgery was more effective in preserving normal postoperative facial function and hearing preservation with less treatment associated morbidity. Effect on preoperative symptoms was similar between the treatment groups. Postoperative functional outcomes and patients' satisfaction were greater after radiosurgery when compared to microsurgery. Patients returned to independent functioning sooner after radiosurgery. Hospital length of stay and total management charges were less in the radiosurgical group.

In a similar study of vestibular schwannoma patients, Régis et al used objective results and questionnaire answers to compare the results of radiosurgery (97 consecutive patients) with a microsurgery group (110 patients who fulfilled the inclusion criteria).[75] Questionnaire answers indicated that 100% of patients who underwent Gamma Knife® radiosurgery compared with 63% of patients who underwent microsurgery had no new facial motor disturbance. Ninety-one percent of patients treated with Gamma Knife® radiosurgery, and 61% in the microsurgery study, had no functional deterioration after treatment. The mean hospitalization stay was three days after Gamma Knife® radiosurgery and 23 days after microsurgery. All working patients who underwent Gamma Knife® radiosurgery kept the same professional activity, compared to 56% in the microsurgery arm. The mean time away from work was seven days for Gamma Knife® radiosurgery compared to 130 days for the microsurgery group. Among patients whose preoperative hearing level was Class 1 according to the Gardner and Robertson scale, 70% preserved functional hearing after Gamma Knife® radiosurgery (Class 1 or 2), compared with only 37.5% in the microsurgery group. Findings after four years of follow up indicated that Gamma Knife® radiosurgery provided better functional outcomes than microsurgery. It was concluded that stereotactic radiosurgery was an effective and less costly management strategy for unilateral vestibular schwannomas less than 3 cm in diameter, and was considered a primary management option.

In a recently published study, Myrseth et al compared the quality of life outcomes for 189 acoustic neuroma patients with tumors less than 30 mm in diameter who were treated with either microsurgery or radiosurgery.[63] The outcome analysis included assessments of tumor control, cranial nerve preservation rates and complications. The results showed that cranial nerve function and overall patient outcomes were better in the radiosurgery group. The results reveal that from the patients' perspective, radiosurgery provides a more desirable outcome than microsurgery.

Radiosurgery after Failed Microsurgery

Pollock et al analyzed patient outcomes to define the role of radiosurgery in patients who had undergone prior microsurgical resection of their vestibular schwannomas.[71] These authors evaluated the pre- and postoperative clinical and neuroimaging characteristics of 76 consecutive patients with 78 vestibular schwannomas who underwent radiosurgery after previous surgical resection. Forty-three patients (55% of tumors) had significant impairment of facial nerve function (House-Brackmann Grades III-VI) after their microsurgical procedure; 50% had trigeminal sensory loss and 96% had poor speech discrimination (< 50%). At a median follow-up of 43 months after radiosurgery, tumor growth control after radiosurgery was achieved in 73 tumors (94%). Roche et al reported on 60 patients who underwent radiosurgery after one or more attempts at surgical resection.[77] The mean interval between surgical removal and radiosurgery was 71.5 months. Technical difficulties during the procedure were observed in the 12 cases, primarily due to problems in identifying the target. The median follow-up was 51.6 months and the tumor control rate was 93%. No patient developed facial and trigeminal nerve deficit. One case developed lower cranial nerve deficit due to bulbopontine radiation injury. Radiosurgery proved to be a safe and effective alternative to additional microsurgery in patients in whom the initial microsurgical removal failed. Stereotactic radiosurgery should be considered for all patients who have regrowth or progression of previously surgically treated vestibular schwannomas.

Microsurgery after Failed Radiosurgery

Microsurgery is rarely needed after vestibular schwannoma radiosurgery (2–5%). The role and scope of microsurgery after radiosurgery is controversial. Some surgeons believe that resection after radiosurgery in these rare patients may be difficult due to the effects of radiation. In order to address this issue, Pollock et al reviewed 13 patients who had delayed microsurgery at a median of 27 months after radiosurgery.[73] Six of the thirteen patients had undergone one or more microsurgical procedures before they underwent radiosurgery. Gross-total resection was achieved in seven patients and near-gross-total resection in four patients. The surgery was described by the responsible surgeon as more difficult than that typically performed for schwannoma in eight patients, no different in four patients, and easier in one patient. At the last follow-up evaluation, three patients had normal or near-normal facial function, three patients had moderate facial dysfunction, and seven had facial palsies. These authors concluded that there was no clear relationship between the use of radiosurgery and the subsequent ease or difficulty of delayed microsurgery. Because some patients have temporary enlargement of their tumor after radiosurgery, the need for surgical resection after radiosurgery should be reviewed with the neurosurgeon who performed the radiosurgery and should be delayed until sustained tumor growth is confirmed. The need for tumor decompression usually arises in patients who had radiosurgery for large vestibular schwannomas. A subtotal tumor resection should be considered for such patients if they require surgical resection of their tumor after radiosurgery. Lee et al performed a retrospective review of four patients who underwent microsurgical resection of vestibular schwannoma after Gamma Knife® radiosurgery or stereotactic radiation therapy. These authors found no significant scarring that could be attributed to the radiation effect.[48] Szeifert et al reviewed 22 patients who underwent surgical resection in a series of 1350 Gamma Knife® radiosurgery patients. These authors studied the histopathological findings and concluded that radiosurgery works by destroying tumor cells directly (with necrosis or inducing apoptosis), as well as by vascular damage. These authors suggested that patients should not undergo craniotomy solely on the basis of radiological progression of the tumor without clinical deterioration.[101]

Delayed Oncogenesis

After radiosurgery, delayed malignant transformation of a histologically "benign" vestibular schwannoma to a more aggressive neoplasm is potentially possible.[4,93] To date, at least five cases of a radiation-related secondary malignant neoplasm have been reported in patients having undergone radiosurgery. One was in a patient treated for a vestibular schwannoma.[2,26,38,58,62,87,91,109] The observed incidence of secondary tumors after radiosurgery is unclear since neither an accurate denominator nor numerator are known. The estimated risk of such oncogenesis over a 5–30 year period (fitting the description of a radiation related cancer) is estimated to be less than 1:1000.[58] This can be compared to the surgical mortality at Centers of Excellence of 0.5% of patients (1/200) in the first post-operative month after microsurgery.

Progression Despite Radiosurgery

Progression after radiosurgery is rare (2–5%).

Complications after Radiosurgery

Further hearing loss, facial numbness, facial pain, facial weakness and temporary unsteadiness.

Repeat Radiosurgery

Since the tumor control rate after radiosurgery is 95–98%, other management options after radiosurgery are rarely needed. Repeat radiosurgery or microsurgery can be considered for patients with documented tumor growth after radiosurgery. Repeat Gamma Knife® radiosurgery can be delivered safely without an increased risk of complications as compared to the initial treatment.[66]

Indications for Radiosurgery

Indications for radiosurgery include newly diagnosed vestibular schwannomas, residual vestibular schwannomas after microsurgery, and recurrent vestibular schwannomas.

Clinical Algorithm

A number of patient related factors are considered in making a recommendation. These factors include:

- Age
- Symptoms
- Hearing status
- Current neurological status
- Medical condition
- Presence or absence of NF2
- Presence or absence of prior procedures
- Concern and risk tolerance for hearing, facial and trigeminal nerve function
- Patient desires
- Patient's decision after informed consent

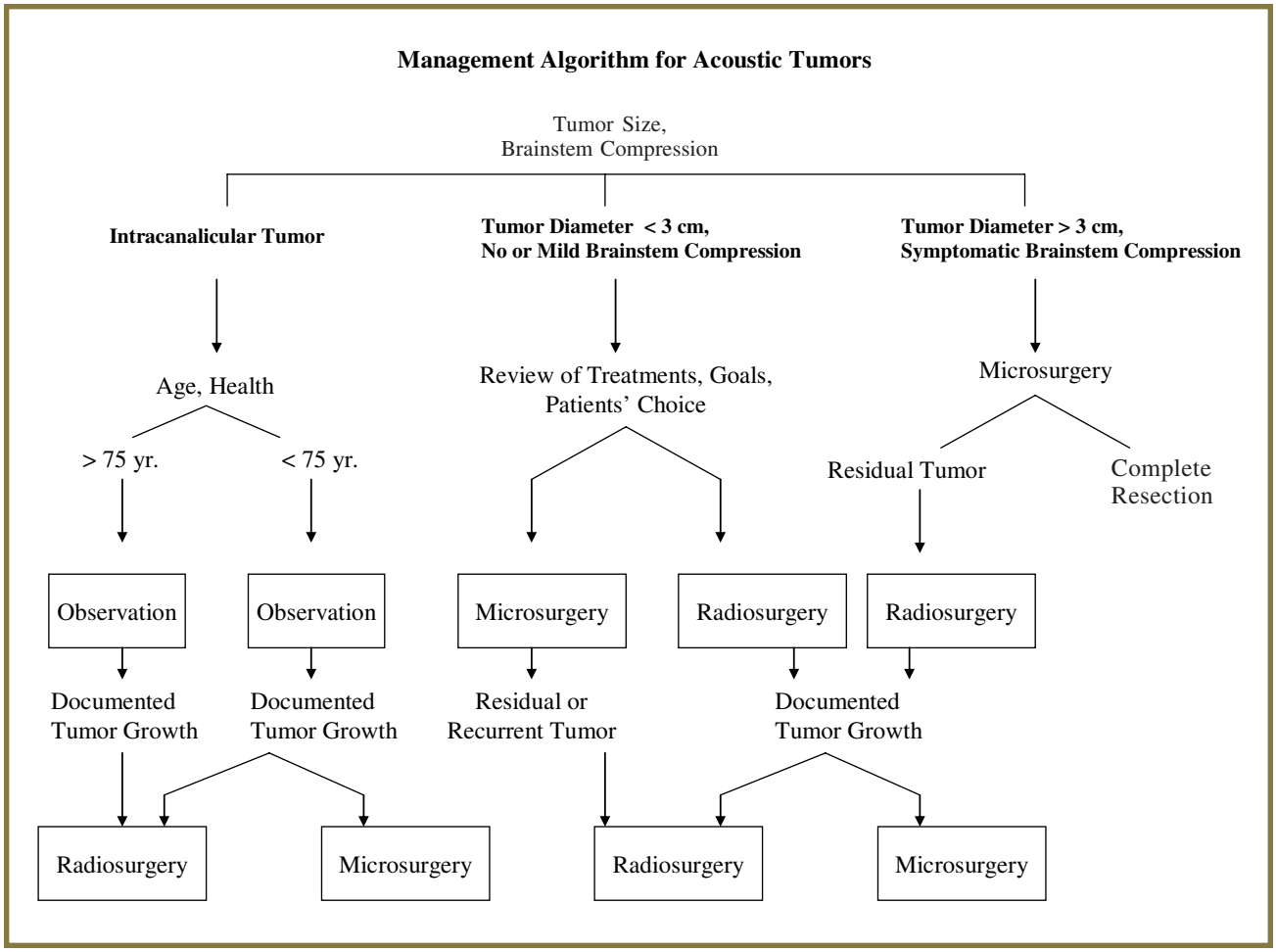
Preoperative Hearing Level
Opinions vary considerably about what constitutes useful hearing. The 50/50 rule is frequently quoted. The rule suggests that individuals with a pure-tone average greater than 50 dB and speech discrimination less than 50% do not have useful hearing.

Tumor Anatomy
Radiosurgery can be performed for intracanalicular tumors and small to medium size tumors without brainstem compression and without signs of hydrocephalus. If hydrocephalus is present in old or infirm patients, a shunting procedure should be considered in addition to radiosurgery. There is no broadly accepted classification of tumor volumes. In addition to tumor diameter, Koos classification[46] is useful because it takes into account the mass effect of the tumor on the brainstem. Koos IV tumors (large tumors with brain shift) with a main diameter less than 3 cm should be offered microsurgery as first management. For intracanalicular tumors, hearing level may influence the decision.[7] Some authors believe that for tumors with a predominant cystic component microsurgery may be more suitable.

Patient Preference
Patients' preference is also considered in selecting a management approach. Some patients prefer tumor removal rather than tumor stabilization. Some patients are willing to sacrifice good hearing if doing so even slightly enhances the possibility of complete tumor removal. For these patients tumor resection is an obvious choice. Many patients prefer effective non-invasive management techniques like radiosurgery. Some patients insist on hearing conservation even when the treating physician is quite convinced that the patient's preoperative hearing is non-serviceable.

Neurofibromatosis 2
Considerations in NF2 patients may be different and additional parameters such as contralateral hearing, previous treatment, lip reading acquisition and additional tumors should be taken into account.

A broad outline of a management algorithm is shown; however, the final recommendation is usually influenced by the recommending physician's experience along with the patient's preference.



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COMPLETE SUMMARY

TITLE:

Stereotactic Radiosurgery for Patients with Vestibular Schwannomas

RELEASE DATE:

May 2006

DEVELOPER AND FUNDING SOURCE:

IRSA (International RadioSurgery Association)

DEVELOPER COMMENT:

IRSA (International RadioSurgery Association) is a non-profit entity dedicated to promoting the development of scientifically relevant practice guidelines for stereotactic radiosurgery. IRSA is a professional organization that works to educate and provide support for physicians, hospitals, insurers and patients.

COMMITTEE:

The IRSA Medical Advisory Board Guidelines Committee and physician representatives in the industry.

GROUP COMPOSITION:

This radiosurgery guidelines group is comprised of neurosurgeons, neurotologists, and radiation oncologists.

Names of Group Members: L. Dade Lunsford, M.D., Neurosurgeon, Chair; Ajay Niranjan, M.B.B.S., M.Ch., Neurosurgeon; Georg Norén, M.D., Neurosurgeon; Jay Loeffler, M.D., Radiation Oncologist; Alain de Lotbinière, M.D., Neurosurgeon; Jordan Grabel, M.D., Neurosurgeon; Douglas Kondziolka, M.D., Neurosurgeon; Jean Régis, M.D., Neurosurgeon; Pierre-Hughes Roche, M.D., Neurosurgeon; Robert Smee, M.D., Radiation Oncologist; Neurosurgeon; Burton Speiser, M.D., Radiation Oncologist; Mark Alden, M.D., Radiation Oncologist; Sandra Vermeulen, M.D., Radiation Oncologist; William F. Regine, M.D., Radiation Oncologist; Barry Hirsch, M.D., Neurotologist; Tonya K. Ledbetter, M.S., M.F.S., Editor; Rebecca L. Emerick, M.S., M.B.A., C.P.A., ex officio.

DISEASE/CONDITION:

Vestibular schwannoma (acoustic neuroma)

NUMBER OF REFERENCES:

110

CATEGORY:

Treatment, proposed surgical management, radiosurgery

CLINICAL SPECIALTY:

Neurological surgery
Neurotology
Radiation oncology

INTENDED USERS:

Physicians
Health Care Providers
Hospitals
Managed Care Organizations
Nurses
Utilization Management

OBJECTIVES:

To develop an evidence and consensus-based stereotactic radiosurgery practice guideline for radiosurgery treatment recommendations to be used by medical and public health professionals following the diagnosis of vestibular schwannoma.

TARGET POPULATION:

Patients diagnosed with vestibular schwannoma (acoustic neuroma).

INTERVENTIONS AND PRACTICES:

The management options for a vestibular schwannoma include serial observation, surgical removal and radiosurgery. Stereotactic radiosurgery is an effective management approach for patients with small to moderate tumor sizes.

Routine follow-up after stereotactic radiosurgery of the vestibular schwannoma is performed using the following schedules:

- Assess MRI scans for tumor response following a long-term schedule (such as six months, one year, two years, four years and eight years).
- Assess hearing levels at each imaging follow-up in patients with serviceable hearing.
- Assess hydrocephalus in patients with large vestibular schwannomas.

OUTCOMES CONSIDERED:

Tumor growth control, hearing, balance symptoms and function, tinnitus, facial nerve function, trigeminal nerve function and overall patient satisfaction.

METHODS TO COLLECT EVIDENCE:

Hand searches of published literature (primary sources); hand searches of published literature (secondary sources); searches of electronic databases

DESCRIPTION OF METHODS TO COLLECT EVIDENCE:

MEDLINE and PUBMED searches were completed for the years 1966 to March 2006. Search terms included vestibular schwannoma, schwannoma, neuroma, neurinoma, stereotactic radiosurgery, stereotactic radiation therapy, Gamma Knife®, CyberKnife®, linear accelerator, irradiation, proton beam, clinical trial, research design, practice guidelines and meta-analysis. Bibliographies from recent published reviews were reviewed and relevant articles were retrieved.

METHODS TO ASSESS THE QUALITY AND STRENGTH OF THE EVIDENCE:

Expert consensus (committee)

METHODS TO ANALYZE EVIDENCE:

Review of published literature

REVIEW METHODS:

External peer review; internal peer review

DESCRIPTION OF REVIEW METHODS:

The recommendations were a synthesis of research obtained in the evidence gathering process by a core group of two members (AN and LDL). These recommendations were mailed to all committee members. Feedback was obtained through this mailed survey consisting of proposed guidelines which asked for comments on the guidelines and whether the recommendation should serve as a practice guideline. No significant disagreements existed. The final statement incorporates all relevant evidence obtained by the literature search in conjunction with the final consensus recommendations supported by all listed working group members.

MAJOR RECOMMENDATIONS:

- Patients with vestibular schwannomas defined by modern neurodiagnostic imaging (CT, MRI) constitute the study group. Such patients typically present with symptoms of hearing loss, tinnitus and/or imbalance. Vestibular schwannomas are considered suitable for various management strategies such as observation with serial imaging, stereotactic radiosurgery, and surgical excision. Stereotactic radiosurgery is typically employed as the first management option in patients with small to medium size tumors (without symptomatic brainstem compression). It is also used to control growth of recurrent or residual tumor after surgical resection. Stereotactic radiosurgery, a minimally invasive, single high-dose, closed skull management strategy, may be especially suitable for patients who desire preservation of neurological function (cochlear, facial nerve) and a high rate of tumor growth control.

- Stereotactic volumetric MR imaging (high resolution) is usually necessary for precisely conformal dose planning. Contrast-enhanced gradient recalled MR images are ideal for radiosurgery dose planning. T2 weighted MR images (3-D volume) are helpful in defining the cranial nerves and the inner ear structures (cochlea and semicircular canals). Sharp fall-off of the radiation dose outside of the target volume is required.

- Current radiation delivery technologies for volumetric conformal stereotactic radiosurgery include Gamma Knife®, proton beam using the Bragg peak effect, and specially modified linear accelerators.

- The optimal single session dose range for volumetric conformal stereotactic vestibular schwannoma radiosurgery has been largely established based on tumor anatomy (proximity of brainstem), hearing status, tumor volume and estimated adverse radiation risks. Minimum doses to the margin of vestibular schwannoma typically range from 12–13 Gy in a single session.

- Depending upon treating physicians' preferences, patients may or may not receive a single stress dose of corticosteroids at the beginning or conclusion of the radiosurgery procedure. Alternatively several doses of steroid at regular intervals (3–4 hourly) can be given on the day of the procedure. Patients can continue to take other medications as recommended by their physicians.

- Post-radiosurgical clinical examinations and MR studies are typically performed at predetermined intervals such as at six months, one year, two years and four years. For patients with preserved serviceable hearing, audiograms are recommended at intervals coinciding with clinical and neuroimaging re-evaluations. Tumors proven to be stable over 4–5 years can subsequently be re-assessed at 2–4 year intervals.

- Patients with large tumors causing symptomatic brainstem compression should be managed with surgical decompression of the tumor. Residual tumor can be treated by radiosurgery.

- Patients with hydrocephalus but without symptoms of brainstem compression can have a shunt inserted prior to radiosurgery, especially if the patient is aged or medically infirm and consequently not a good candidate for resection.

- Causes for failure of stereotactic radiosurgery include inadequate visualization of the tumor, lack of intraoperative stereotactic 3-D (volumetric) imaging, and insufficient dose (due to large tumor volume and proximity to the brainstem) to achieve a growth control response.

STEREOTACTIC RADIOSURGERY FOR VESTIBULAR SCHWANNOMAS:

Stereotactic radiosurgery is defined as a single session, high-dose delivery of focused radiation precisely to the vestibular schwannoma, as identified by stereotactic imaging. In systems requiring head fixation of the stereotactic frame (e.g. Gamma Knife®), radiation delivery occurs under the direct supervision of a medical team consisting of a neurosurgeon, radiation oncologist, registered nurse and medical physicist, at a minimum. At some centers a neurotologist is also part of the radiosurgery team. The neurosurgeon and/or neurotologist are an integral part of the critical decision making steps and the target planning and dose approval within the brain for both LINAC and proton beam based systems (whether single session or stereotactically hypofractionated radiation therapy) regardless of head fixation system. The radiation delivery of the approved targeting and dosing plan (as designed and approved by the neurosurgeon/neurotologist and radiation oncologist together) may occur on subsequent days for LINAC or proton beam based single session or hypofractionated sessions under the direct supervision of a radiation oncologist without the neurosurgeon or neurotologist present. In this case, the minimum delivery team should consist of a certified radiation therapist and medical physicist. Should the original targeting plan require modification during the radiation delivery of the subsequent sessions, the neurosurgeon and/or neurotologist should review/design and approve the new targeting and dosing plan before the continuation of the radiation delivery by the radiation oncologist.

TYPE OF EVIDENCE:

Type III evidence exists in support of stereotactic radiosurgery for vestibular schwannomas.

POTENTIAL BENEFITS:

Minimally invasive approach, and high rates of tumor growth control (95–98%), serviceable hearing preservation (60–70%), facial nerve preservation (>95%) and trigeminal nerve preservation (>95%). The medical literature has documented the cost savings benefit of stereotactic radiosurgery versus open surgical procedures and the lower risk potential of bleeding, anesthesia problems, infections and side effects which may result in transient or permanent disabilities from open surgery.

SUBGROUP(S) MOST LIKELY TO BENEFIT:

Patients diagnosed with small to medium size vestibular schwannomas without symptomatic brainstem compression. Patients with residual or recurrent vestibular schwannoma after resection. Single session radiosurgery conducted by experienced Centers of Excellence with neurosurgeons/neurotologists and radiation oncologists working as a team have the greatest opportunity for long-term benefit and outcomes superior to centers with relatively little experience or without a team approach.

POTENTIAL HARMS:

Major adverse effects of radiosurgery are based on location, volume and dose. These risks can be estimated based on published data and experience. Individual risks are related to the anatomic proximity of vestibular schwannoma to the cochlea, semicircular canals, cochlear nerve, facial nerve, trigeminal nerve and the brainstem. For LINAC and proton beam based fractionated radiation treatments with a frameless system, the total radiation dose is usually higher than with single session radiosurgery. The higher dose is an issue as the risk of complications is directly related to the dose and treatment volume. At this time, little or no long-term data (5–10 years after treatment) on complications or tumor control exist for fractionated stereotactic radiation therapy.

SUBGROUP(S) LIKELY TO BE HARMED:

Patients with large volume vestibular schwannomas causing symptomatic mass effect on the brainstem.

GUIDELINE STATUS:

This is the full current release of the guideline.

GUIDELINE AVAILABILITY:

Electronic copies: available in Portable Document Format (PDF) from www.IRSA.org

Print copies: available from IRSA, P. O. Box 5186, Harrisburg, PA 17110

PATIENT RESOURCES:

Patient resources are available online at www.IRSA.org, by email at office@IRSA.org or by calling +717-260-9808.

See "publications" for patient resources for vestibular schwannoma: www.IRSA.org/publications.html/
Brain Talk® Volume 9, No. 2; ***Another Perspective***® Volume 2, No.1

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