Acoustic Neuromas: Current Treatment Options and Hearing Preservation Results

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Advancements in the treatment of acoustic neuromas have resulted in several options for therapy that offer relatively high rates of hearing preservation and facial nerve function. Patients are now offered a treatment strategy that includes observation with serial scanning, microsurgical removal, or stereotactic radiosurgery alone or in some combination. This article outlines the rationale for these strategies and their indications. The author's personal series of acoustic neuroma patients managed surgically over a 5 year period will be presented and compared to current results obtained with stereotactic radiosurgery utilizing the Gamma Knife. A suggested treatment algorithm is proposed based upon this data.

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Introduction. Acoustic neuromas, also known as vestibular schwannomas, constitute approximately 6 percent of all brain tumors. These tumors occur in all races of people and have a very slight predilection for women over men. In the United States, approximately 10 people per 1 million per year are diagnosed with an acoustic neuroma. This translates to roughly 2,500 newly diagnosed acoustic neuromas per year in the United States based upon a population of around 250 million.[1]

Acoustic neuromas are typically benign fibrous growths that arise from one of the vestibular divisions of the eighth cranial nerve or vestibulocochlear nerve. Malignant tumors account for less than 1 percent of acoustic neuromas. The location of these tumors is in the cerebellopontine angle, which is the region between the brain stem and the temporal bone. As the tumors enlarge, they involve surrounding structures which have to do with vital functions, such as swallowing, coordinated movement, hearing, balance, facial movement and sensation. In the majority of cases, these tumors grow slowly over a period of years. In other cases, the growth rate is more rapid and patients develop symptoms at a faster pace. Usually, the symptoms are mild and many patients are not diagnosed until some time after their tumor has developed. Many patients also exhibit no tumor growth over a number of years when followed by yearly MRI scans.

Acoustic neuromas occur in 2 forms: sporadic and those associated with Neurofibromatosis Type II (NF II). Approximately 95 percent of all acoustic neuromas are sporadic cases and are unilateral. In contrast, those tumors associated with NF II are bilateral and account for approximately 5 percent of acoustic neuroma patients. Patients with sporadic acoustic neuromas tend to begin having symptoms in middle age with the average being around 50 years old at diagnosis. Patients with NF II present at a younger age averaging around 30 years old when they first develop symptoms.[2] There is a high degree of variability, however, and patients may begin having symptoms and be diagnosed with an acoustic neuroma during childhood or young adult life, as well as when elderly.

Several treatment modalities are currently used for the treatment of acoustic neuromas. Until the past decade, surgical removal of the tumor was the standard form of therapy. Patients now also have the option of undergoing a noninvasive radiation treatment, called stereotactic radiosurgery or Gamma Knife surgery, to halt the growth of the tumor. Some patients might also be candidates for a combination of these therapies.

Treatment Options. The goal of therapy of any benign brain tumor is to eradicate or control further growth of the tumor while preserving neurologic function. There are many factors which come to bear in terms of the success of treatment for these tumors. Acoustic neuromas, because of their location in proximity to the brain stem and cranial nerves, are a complicated treatment problem. Every patient has 3 basic treatment options: observation with serial imaging, microsurgery, and stereotactic radiosurgery or Gamma Knife surgery. Observation may be appropriate for patients older than age 65 who have few symptoms considering the known natural history of these tumors in older patients. A number of studies have demonstrated that approximately two-thirds of patients in this age group will not experience growth of the tumor when followed for 3 to 5 years. This is in contrast to younger patients who tend to experience tumor growth or worsening of symptoms at a much higher rate. Therefore, older patients tend to be better advised to wait and have repeat imaging at 6 to 12 month intervals if they are relatively asymptomatic or have hearing loss only. Otherwise, treatment is generally recommended to improve the chance of an optimal outcome from either microsurgery or Gamma Knife surgery.

Surgery Options. Surgery for acoustic neuromas has been performed since the early 1900's. The initial successes were few and far between by the early pioneering neurosurgeons that treated this problem. The past 20 years have witnessed an astounding improvement in our abilities to successfully deal with these tumors while preserving the neurological function of the patient. The choice of surgical approach depends upon the size of the tumor and the level of residual hearing detected on the audiogram. Again, the larger the tumor the lower the chances of saving hearing. The 3 most common surgical approaches for acoustic neuromas are the retromastoid suboccipital, translabyrinthine, and middle fossa approaches.

Retromastoid Suboccipital Approach.[3, 4] An incision is made behind the ear and an opening in the skull is made behind the mastoid bone. The portion of the brain called the cerebellum is retracted away in order to expose the tumor. In most cases the tumor can be completely removed. Every effort is made in this approach to preserve hearing and still completely remove the acoustic neuroma. In some cases, because of invasion of the auditory nerve by the tumor, it is necessary to sacrifice hearing in order to completely remove the neuroma. The success of hearing preservation in these cases is largely dependent upon the size of the tumor and the condition of the auditory nerve in relation to the tumor. [4, 5]

Translabyrinthine Approach.[6] The translabyrinthine approach requires removal of the mastoid bone and the balance canal structures of the inner ear in order to expose the tumor. One of the main advantages in this approach is that there is little or no retraction of the cerebellum required to

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provide excellent exposure of the tumor. Another advantage is early and direct localization of the facial nerve which facilitates separation of the nerve from tumor, optimizing facial nerve outcome. After completion of tumor removal, the opening in the mastoid bone is closed with a fat graft, which is taken from the abdomen, and reconstructed with bone cement to obtain a good cosmetic result. This approach sacrifices the hearing and balance mechanism of the inner ear. As a consequence, the ear is made permanently deaf. Although the balance mechanism is removed on the operated ear, the balance mechanism in the opposite ear provides stabilization for the patient.

Middle Fossa Approach. [7, 8] This approach is used for small tumors and is utilized in cases when hearing is to be conserved. An incision is made beginning just in front of the ear and extends upward in a curved fashion. A small opening in the bone is made above the ear, and the membrane that covers the brain is elevated away from the bone and gently held away from the bony floor of the skull. Bone is then removed over the top of the internal auditory canal to expose the tumor. Every effort is made to preserve hearing and still completely remove the tumor.

Stereotactic Radiosurgery (Gamma Knife Surgery). Stereotactic radiosurgery, or Gamma Knife surgery, was first developed in Sweden in the 1960's by a neurosurgeon at the Karolinska Institute in Stockholm, Lars Leksell. Stereotactic surgery refers to a neurosurgical procedure done in such a way as to precisely localize a point in space inside the brain. The head and brain are considered as a volume in space with each point in the volume described by a set of 3 points in reference to the 3 axes of 3 planes. Neurosurgeons utilize stereotactic methods to navigate to deep structures, to plan surgery, to precisely take biopsies or resect tumors in a minimally invasive fashion, and to precisely deliver doses of radiation to the brain and spine. In stereotactic radiosurgery, radiation is delivered to a target volume by aiming low energy beams from many different angles to converge on the target volume, thereby delivering a very high dose without the surrounding tissue receiving a harmful dose. This technique has been applied to a number of differing tumor types, including acoustic neuromas.[9]

Surgical Treatment. This author has treated 249 patients microsurgically over the past 5 years via the full spectrum of surgical approaches. The Koos Grading System *(Table 1)* is used to divide the tumors into four size-based categories; grade 1=1-10mm, grade 2=11-20mm, grade 3 = 21-30 mm, and grade 4=greater than 31 mm with deformation of the brain stem.[5] The results are presented in *Tables 2-5* and *Figures 1-3.* Considering tumors of all sizes patients had a good facial nerve outcome of 85 percent, defining "good" as House-Brackmann (H-B) grades 1 and 2. Hearing was in Gardner-Robertson class I or II in 106 patients in the series and was therefore attempted to be preserved by either a middle fossa or retrosigmoid approach. Otherwise no attempt was made to preserve hearing, no

Table 1. Koos Grading Scale for Acoustic Neuromas

GRADE	TUMOR SIZE
1	0-10 mm, intracanalicular
2	10-20 mm total, 0-10 mm extrameatal
3	Up to 30 mm total
4	>30mm, brain stem deformation

Table 2. Overall facial nerve and hearing preservation outcomes in the author's personal series of 249 patients with acoustic neuromas treated via microsurgery.

Table 3. Facial nerve and hearing preservation results separated according to Koos Grade of the tumor.

Table 4. Facial nerve function and hearing preservation results in 80 patients undergoing a middle fossa approach.

Table 2.

H-B Facial Nerve Grade	I	2	3	4	5	6	Hearing +	Hearing -
N	186	28	16	3	6	10	62	37
%	76%	11%	6%	1%	2%	4%	63%	37%

Table 3.

	H-B I	2	3	4	5	6	Hearing +	Hearing -
Koos I	62 (86%)	3 (4)	3 (4)	I (1.4)	I (1.4)	2 (2.8)	47 (80%)	12 (20)
2	69 (77.5)	10 (11)	5 (6)	-	I (1)	4 (4.5)	13 (48)	14 (52)
3	36 (72)	10 (20)	2 (4)	I (2)	I (2)	-	2 (15)	11 (85)
4	19 (49)	6 (15)	6 (15)	I (3)	3 (8)	4 (10)	I (25)	3 (75)

Table 4.

	H-B I	2	3	4	5	6	Hearing +	Hearing -
Koos I	52	5	I	I	2	-	47	14
%	85.3	8.2	1.6	1.6	3.3	-	77	23
Koos 2	11	5	I	2	-	-	9	10
%	58	27	5	10	-	-	47	53
Total	63	10	2	3	2	-	56	24
%	79	12	2.5	4	2.5	-	70	30

matter the approach. Hearing was preserved in 63 patients (59 percent) out of 106 attempts. Hearing "preservation" is defined as maintaining Gardner-Robertson (G-R) class I or II hearing (ie, at least 50 percent speech discrimination and 50 dB pure tone average on audiometry).

When broken down by Koos Grade *(Figure 1, Table 3)* the percentage of good facial nerve outcome (H-B 1 and 2) was 90 percent for grade 1, 88.5 percent for grade 2, 92 percent for grade 3, and 64 percent in grade 4 patients. Hearing preservation dropped from 77 percent in grade 1 patients to 41 percent, 25 percent, and 25 percent in grades 2-4 respectively.

Eighty patients were treated via the middle fossa approach *(Figure 2, Table 4)*. Patients with the smallest tumors (Koos grade 1) experienced a good facial nerve outcome in 93.5 percent and hearing preservation in 77 percent. This contrasts to the patients with Koos grade 2 tumors who had 85 percent good facial nerve outcome and 47 percent hearing preservation in G-R classes I or II. Overall, 9 percent of patients experienced a poor outcome in facial nerve function, mostly in patients with grade 2 size tumors.

Twenty-eight patients were treated via the retromastoid suboccipital approach with an attempt at hearing preservation, all Koos grades 2 through 4 *(Figure 3, Table 5)*. All patients with grade 2 tumors (10/10) had a good facial nerve outcome but only 30 percent had hearing preserved. Ninety-two percent of patients with grade 3 tumors had good facial nerve function and 25 percent had hearing preserved. Grade 4 tumor patients did not fare as well with 60 percent

Table 5. Facial nerve function and hearing preservation outcome for patients treated via the Retromastoid Suboccipital approach. (N= 28)

Table 6. Treatment recommendation paradigm for acoustic neuromas based upon tumor size grade.

Table 5.

	H-BI	2	3	4	5	6	Hearing +	Hearing -
Koos I	-	-	-	-	-	-	-	-
%								
Koos 2	9	I					3	7
%	90	10	-	-	-	-	30	70
Koos 3	12	-	I	-	-	-	3	9
%	92	-	8	-	-	-	25	75
Koos 4	3	-	I	-	I	-	I	3
%	60	-	20	-	20	-	25	75
Total	24	I	2	-	I	-	7	19
%	86	3.5	7	-	3.5	-	32	68

having good facial nerve function (3/5) and only 1 of 4 with hearing preservation.

Discussion. The contemporary patient with an acoustic neuroma is faced with a difficult decision in terms of treatment strategy. Patients with tumors larger than 3 centimeters in diameter are mostly relegated to the microsurgical group, however, some patients are now being treated with such large tumors using Gamma Knife surgery[10] or a combination of microsurgery and Gamma Knife surgery. All other patients face an interesting volume of information and sources upon which to try and make an informed decision. Based upon a comparison of the author's personal surgical series with contemporary Gamma Knife surgery data a treatment algorithm can be distilled based upon evidence based criteria.

Stereotactic Radiosurgery or Gamma Knife Surgery. Radiation therapy, in its various forms, has been applied to the treatment of acoustic neuromas for a number of years. Historically this was done since the results of surgery in the past (prior to the 1970's) were actually quite dismal in most cases. However, with improvements in microsurgical technique and surgical approach, very few patients until the past 10 years, have undergone any form of radiation therapy for their acoustic neuroma. Currently stereotactic radiosurgery is utilized more and more for the treatment of these tumors. Though the goal of this therapy is tumor growth control, rather than removal of the tumor, complication rates are extremely low. The low chance of complications, and the outpatient nature of the technique, has led to its gaining wider popularity. In fact, it was estimated that in the year 2000, the number of patients treated with radiosurgery equaled the number of patients treated with microsurgery for acoustic neuroma in the United States. Furthermore, the number of patients choosing radiosurgery is greater than those choosing microsurgery since that time.[1]

Stereotactic radiosurgery, or Gamma Knife surgery, is a method of delivering a radiation dose in such a way as to minimize the affects of the radiation on the surrounding normal tissues while delivering a

Table 6. Treatment Recommendations

Koos I	Gamma Knife surgery or microsurgery
Koos 2	Gamma Knife surgery
Koos 3	Gamma Knife surgery
Koos 4	Microsurgery

Figure 1.

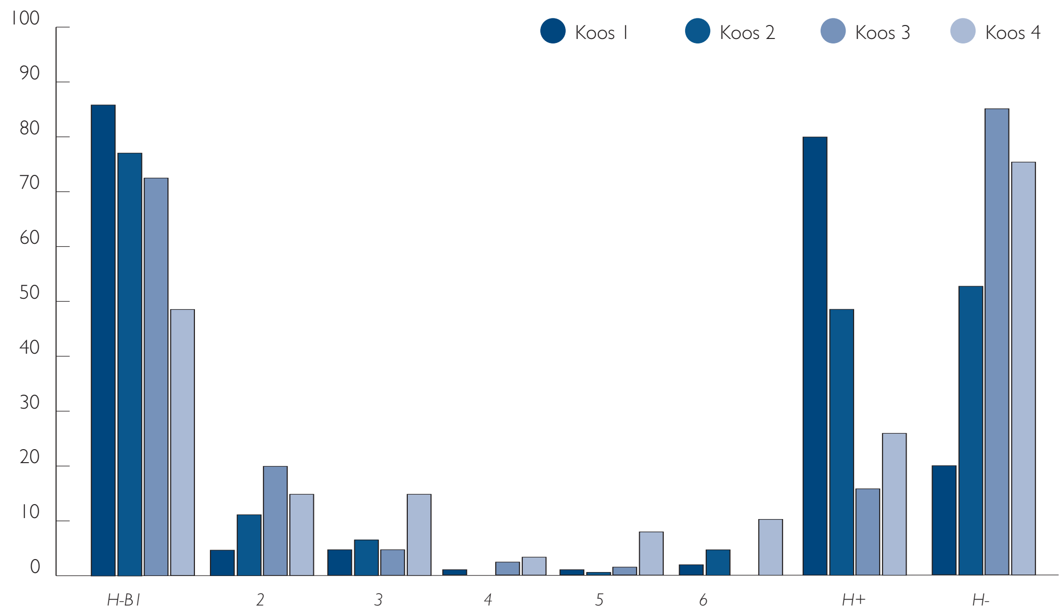


Figure 1.
Histogram comparing the results of *Table 3* for facial nerve function and hearing preservation outcome demonstrating the influence of tumor size.

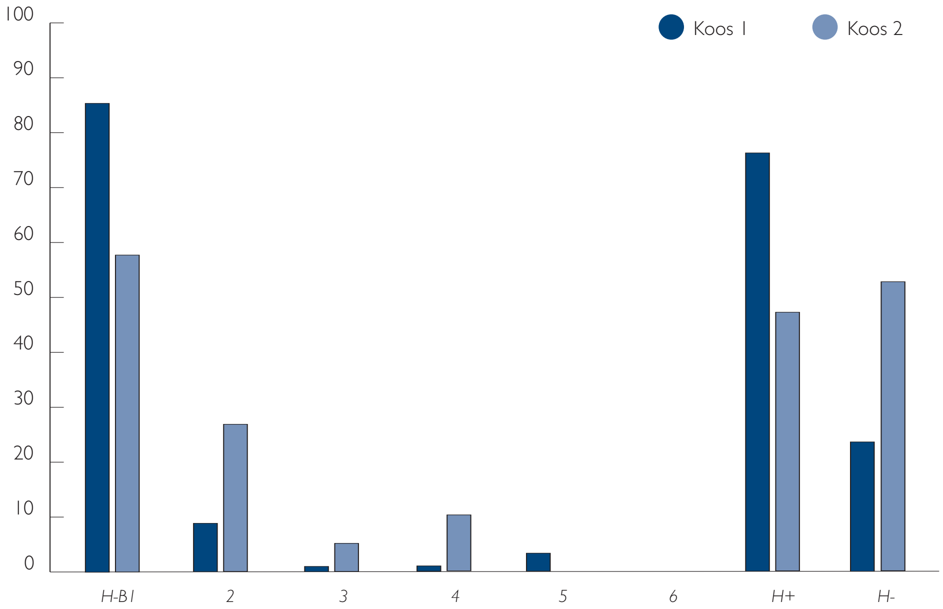
very high dose to the tumor. Low dose radiation beams are aimed from many different directions to converge on the tumor and, thereby, deliver a very high radiation dose. This type of treatment comes in several different forms. These methods are variously named the Gamma Knife, LINAC, proton beam therapy, and fractionated stereotactic radiotherapy (FSR). Questions regarding the long-term results of this therapy have been answered in part by studies published in the past few years. These procedures are attractive to many patients because of the promise of low complication rates and minimal hospital stay. The best results to date, and the highest patient treatment volume, has been with Gamma Knife surgery which will be the focus of comparison with microsurgical outcomes.

Patients treated with Gamma Knife surgery until the mid-1990's were treated with higher doses than in contemporary series. Nevertheless, even at high doses hearing preservation and facial nerve function rates were acceptable. In the University of Pittsburgh series published in 1998 in the New England Journal of Medicine, 51 percent of patients had no change in hearing at long-term follow-up of 4 to 10 years. Seventy-nine percent had grade 1 facial nerve function, however, 26 percent of patients in the series were previously treated with microsurgery and had preexisting weakness before Gamma Knife surgery. Tumor control in the series was 98 percent with 62 percent of tumors shrinking, 33 percent unchanged, and 6 percent enlarged at late follow-up.[9]

A large series treated with the low doses currently in use now was published in 2000 by Prasad and Steiner describing the outcome of Gamma Knife surgery in 153 patients with acoustic neuromas.[10] Ninety six of these patients were treated primarily with Gamma Knife surgery, the other 57 being after failed microsurgery. Patients were followed an average of 4.3 years with a range of 1 to 10 years. In patients treated primarily

Figure 2.
Histogram demonstrating the influence of tumor size on facial nerve and hearing outcomes with the middle fossa approach.

Figure 2.



with Gamma Knife surgery 65 percent decreased in size, 25 percent were stable, and 11 percent increased in size. Only one patient had temporary facial nerve weakness after Gamma Knife surgery for a risk of facial weakness of <0.2 percent. In patients with hearing prior to Gamma Knife surgery, 58 percent had no change after treatment, 35 percent were worse, but 6 percent actually improved. This series reported results with tinnitus as 88 percent unchanged (23/26) after Gamma Knife surgery.

Very favorable results regarding hearing preservation have been reported from the Karolinska Institute in Stockholm utilizing current dosing regimens. In 2001 Kihlstrom reported a series of 92 patients with tumors less than 2.5cm treated by Gamma Knife surgery with a minimum of 5 years follow-up. Gardner-Robertson class I or II hearing was preserved in 70 percent of these patients. Tumor volume was decreased in 56 percent, unchanged in 43 percent, and increased in only 1 percent. Not a single patient suffered a facial nerve paresis, temporary or permanent. [11] The latter 2 series represent what is the general experience at present with Gamma Knife surgery for acoustic neuromas.

Other complications, such as cerebrospinal fluid leaks, infections, meningitis, and medical complications have not been reported to any significant degree with gamma knife surgery. [9, 10] Malignant degeneration has been mentioned as a potential complication of radiosurgery that would result in death. However, it is estimated that the risk of such a complication after Gamma Knife surgery is less than 0.1 percent.[12] Overall, therefore, Gamma Knife surgery for acoustic neuromas has proven extremely safe.

Surgical Results. The surgical results of the author compare well to the major series published in the literature to date with respect to facial nerve functional preservation and hearing conservation in properly selected patients.[5, 13, 14] A common theme in surgical results is the influence of tumor size on the

Figure 3.

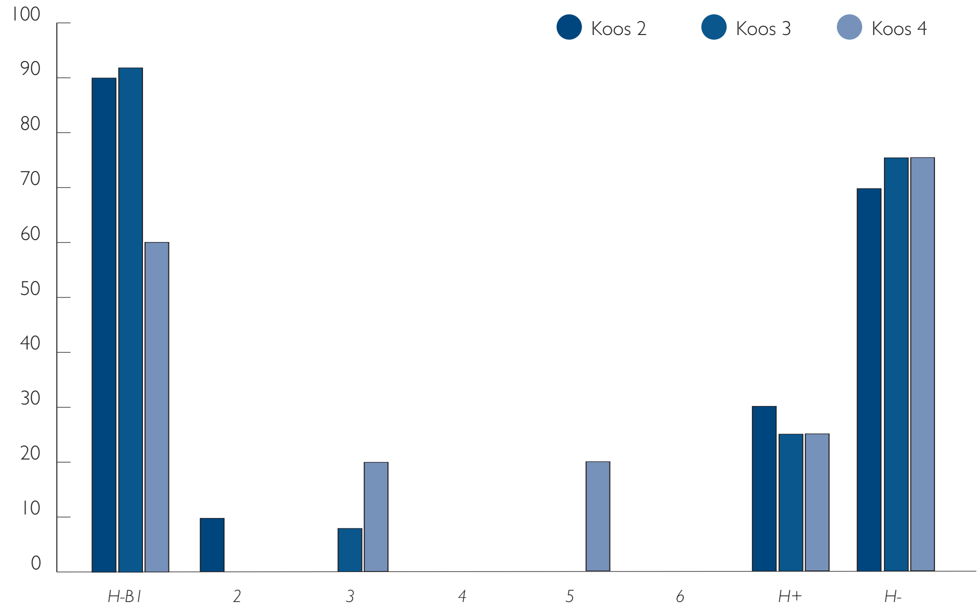


Figure 3.
Facial and Hearing results for the Retromastoid Suboccipital approach.

results both for facial nerve and hearing function. This is easily demonstrated in Tables 3 and 4. Overall, facial nerve results are significantly poorer when the tumor reaches size grade 3 and especially 4. It is clearly more difficult to save facial nerve function in very large tumors (those over 3 cm in diameter). The difference in outcome is especially prominent when utilizing the middle fossa approach in the author's series when tumors extend outside the internal auditory canal, thus becoming Koos grade 2 tumors. The percentage of patients with perfect facial nerve outcome drops from 85.3 percent to 58 percent. The difference in hearing conservation success is just as stark, dropping from 77 percent in the smallest tumors to only 47 percent in the larger using this approach. The hearing results obtained using the retromastoid suboccipital approach would not argue for its employment in grade 2 tumors, but the facial nerve results might. The numbers are small, however, 9 of 10 patients with grade 2 tumors operated retromastoid had a H-B grade 1 facial nerve result. Other complications encountered in the surgical series were delayed facial nerve palsies (14 percent) and cerebrospinal fluid leakage (5 percent). There was one death in the series resulting in a surgical mortality of 0.4 percent.

Current Treatment Paradigm. Based upon an analysis of the results of the author's surgical series and the available data for Gamma Knife surgery for acoustic neuromas a treatment paradigm *(Table 6)* has been developed and is used to advise patients with these tumors. Comparing the author's results by tumor size to the best available Gamma Knife surgery data it is clear that patients with grade 2 and 3 tumors have a higher chance of a better outcome with Gamma Knife surgery. Good facial nerve outcome is 88.5 percent and 94 percent respectively for grade 2 and 3 tumors surgically, versus less than 0.2 percent risk (ie, 99.8 percent good outcome) for Gamma Knife surgery. Hearing preservation is at best 37 percent in these patients versus 70 percent for Gamma Knife surgery. One could argue that for patients with grade 1, ie, intracanalicular, tumors that microsurgery and Gamma Knife surgery are roughly equivalent considering the 77 percent hearing preservation rate with microsurgery versus 70 percent for Gamma Knife surgery. This, however, comes at the expense of a 10 percent risk of poor facial nerve outcome in H-B grades 3-6. For many patients this may not be a satisfactory trade-off. Further, a series of patients with grade 1 tumors treated with a contemporary radiation dose with Gamma Knife surgery at the University of Pittsburgh have achieved 100 percent hearing preservation rates with limited follow-up of less than 3 years.[15] It is likely that these results will remain with little change over time considering the durability of Gamma Knife surgery results. Patients with the largest tumors are relegated to microsurgical treatment at the present time. However, it may be that a less aggressive microsurgical approach in terms of attempting total removal is indicated in order to ensure good facial nerve function. The patient could then be treated by Gamma Knife surgery to control residual tumor. This strategy is being employed by various neurosurgeons but no data is available to date to validate such a strategy.

Conclusions. Patients with acoustic neuromas are now treated employing 1 of 3 strategies; observation with serial imaging, microsurgical removal, and stereotactic radiosurgery. Stereotactic radiosurgery or Gamma Knife surgery has emerged as a primary treatment option for the majority of patients diagnosed with these tumors. Based upon a comparison of results between microsurgery and Gamma Knife surgery focusing on facial nerve function and hearing preservation a treatment recommendation paradigm can be constructed. This paradigm results in recommending Gamma Knife surgery for patients with tumors measuring less than 3 centimeters in diameter (Koos grades 1-3) and microsurgery for those with larger, grade 4 tumors.

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