# Acoustic Neuroma
# Basic Overview

**IMPORTANT POINTS TO KNOW ABOUT AN ACOUSTIC NEUROMA:**

- An acoustic neuroma, also called a vestibular schwannoma, is a rare benign tumor of the hearing and balance nerves.
- It is usually slow growing and expands at its site of origin (1.5mm/yr.).
- The most common first symptom is hearing loss in the tumor ear.
- The cause is generally unknown, although may be genetic in some cases.
- If an acoustic tumor becomes large it will push on the surface of the brainstem but not grow into brain tissue.
- Continued tumor growth may threaten neurological function and even life.
- The treatment options are observation, surgical removal or radiation.

## WHAT IS AN ACOUSTIC NEUROMA?

An acoustic neuroma, known as a vestibular schwannoma, is a benign (non-cancerous) growth that arises on the eighth cranial nerve leading from the brain to the inner ear. This nerve has two distinct parts, one part associated with transmitting sound and the other with sending balance information to the brain from the inner ear. The eighth nerve, along with the facial or seventh cranial nerve, lie adjacent to each other as they pass through a bony canal called the internal auditory canal. This canal is approximately 2 cm (0.8 inches) long and it is generally here that acoustic neuromas originate from the sheath surrounding the eighth nerve. The seventh or facial nerve provides motion to the muscles of facial expression.

Acoustic neuromas usually grow slowly over a period of years. They expand in size at their site of origin and when large can displace normal brain tissue. The brain is not invaded by the tumor, but the tumor pushes the brain as it enlarges. The slowly enlarging tumor protrudes from the internal auditory canal into an area behind the temporal bone called the cerebellopontine angle. The tumor now assumes a pear shape with the small end in the internal auditory canal. Larger tumors can press on another nerve in the area (the trigeminal nerve) which is the nerve of facial sensation. Vital functions to sustain life can be threatened when large tumors cause severe pressure on the brainstem and cerebellum. Tumors are typically described as small (less than 1.5 cm), medium (1.5 cm to 2.5 cm) or large (more than 2.5 cm).

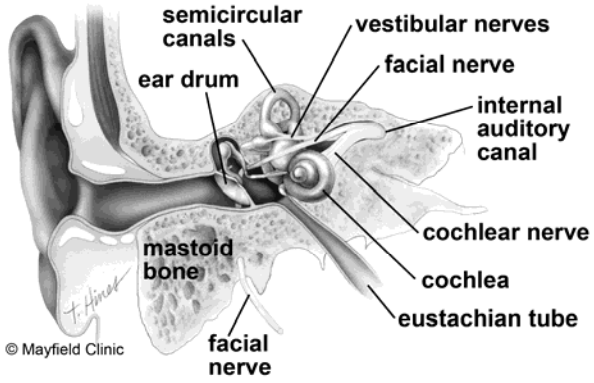


**Figure 1**. The normal anatomy of the ear. The outer ear funnels sound to the eardrum, which vibrates three tiny bones called ossicles (malleus, incus and stapes). The spiral-shaped cochlea is filled with liquid, which moves in response to vibrations. As the fluid moves, thousands of hair cells are stimulated, sending signals along the cochlear nerve (responsible for hearing) to the brain. Attached to the cochlea are three semicircular canals positioned at right angles to each other. The three canals are able to sense head position and posture. Electrical signals from the semicircular canals are carried to the brain by the superior and inferior vestibular nerves (responsible for balance). The cochlear and vestibular nerves form a bundle inside the bony internal auditory canal. Inside the canal, the vestibulocochlear nerve lies next to the facial nerve (responsible for facial movement). *(Printed with permission of the Mayfield Clinic – www.mayfieldclinic.com)*

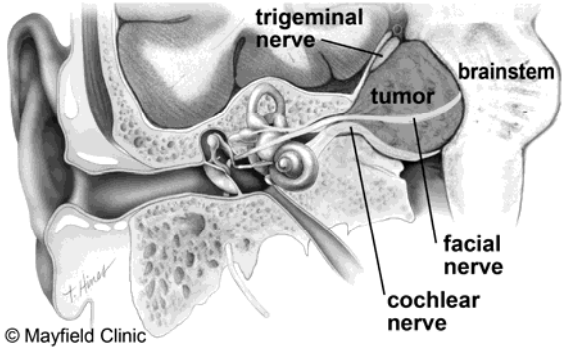


**Figure 2.** An acoustic neuroma expands out of the internal auditory canal displacing the cochlear, facial and trigeminal nerves located in the cerebellopontine angle. Eventually the tumor can compress the brainstem. *(Printed with permission of the Mayfield Clinic – www.mayfieldclinic.com)*

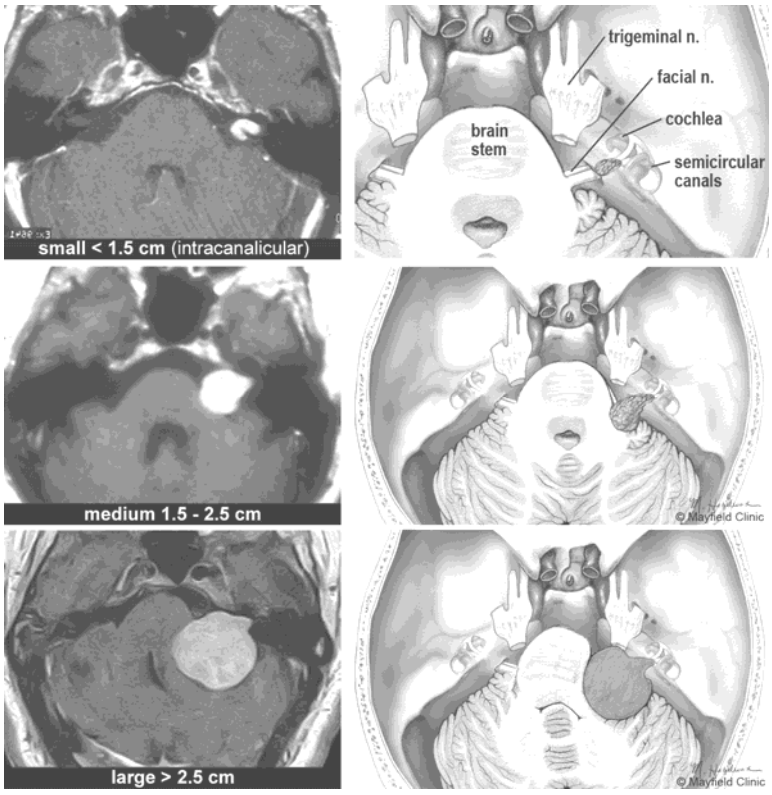


**Figure 3.** Acoustic neuromas are classified according to their size. MRI scans and correlative illustrations of small (intracanalicular), medium and large acoustic neuromas. *(Printed with permission of the Mayfield Clinic – www.mayfieldclinic.com)*

## ARE ACOUSTIC NEUROMAS HEREDITARY?

Although there is a heritable condition called Neurofibromatosis Type 2 (NF2) which can lead to acoustic neuroma formation, most acoustic neuromas occur spontaneously without any evidence of family history (95%).

## HOW OFTEN DO ACOUSTIC NEUROMAS OCCUR?

Most recent publications suggest that the incidence of acoustic neuromas is rising. This is because of advances in MRI scanning both on incidental scans and for patients experiencing symptoms. Studies in Denmark published in 2004 show the incidence is 17.4 per million or close to 2 persons per 100,000. Most acoustic neuromas are diagnosed in patients between the ages of 30 and 60.

## SYMPTOMS OF ACOUSTIC NEUROMA

Early symptoms are easily overlooked, thus making diagnosis a challenge. There usually are symptoms, however, indicating the possibility of an acoustic neuroma. The first symptom in 90% of those with a tumor is a reduction in hearing in one ear, often accompanied by ringing in the ear called tinnitus. The loss of hearing is usually subtle and worsens slowly, although occasionally a sudden loss of hearing can occur. There may be a feeling of fullness in the affected ear. These early symptoms are sometimes mistaken for normal changes of aging, or attributed to noise exposure earlier in life and therefore the diagnosis is often delayed.

Since the balance portion of the eighth nerve is where the tumor arises, unsteadiness and balance problems or even vertigo (the feeling like the world is spinning), may occur during the growth of the tumor. The remainder of the balance system sometimes compensates for this loss, and, in some cases, no imbalance will be noticed. Larger tumors can press on the trigeminal nerve, causing facial numbness and tingling—constantly or intermittently. Tumor related increase of intracranial pressure may cause headaches, clumsy gait and mental confusion. This can be a life-threatening complication requiring urgent treatment.

Even though the facial nerve (the nerve that moves the face) may be compressed by the tumor, it is unusual for patients to experience weakness or paralysis of the face from acoustic neuromas—although this may occasionally occur.

## TYPICAL SYMPTOMS RECAP

1. **Unilateral Hearing Loss** (on one side only) - This can occur suddenly (sudden hearing loss) but it can be very gradual, over months or years. In most acoustic neuroma patients, the loss is more pronounced in the higher frequencies. Unilateral hearing loss is usually the first symptom that leads to discovery of this benign brain tumor.
2. **Tinnitus** (ringing or buzzing in the ears) - Not all patients with tinnitus have a brain tumor and not all acoustic neuroma patients have tinnitus. Most of them do however, both before and after treatment.
3. **Feeling of fullness in the ears** - Acoustic neuroma patients sometimes complain of a feeling that their ear is plugged or "full."
4. **Balance problems, vertigo** - Acoustic neuroma patients often experience balance issues before diagnosis. It can occur very gradually and may go unnoticed as the body has many compensating mechanisms.
5. **Headaches** - Acoustic neuroma patients sometimes recall, after diagnosis, that they had unexplained headaches.
6. **Facial pain, numbness, paralysis** - Acoustic neuromas are usually discovered before they cause facial symptoms. However, if they are large or impacting one of the facial nerves, they can cause numbness or even facial paralysis.

## IDENTIFYING THE TUMOR

Advances in medicine have made possible the identification of small acoustic neuromas (those still confined to the internal auditory canal). Routine auditory tests may reveal a loss of hearing and speech discrimination (the patient may hear sounds in that ear, but cannot comprehend what is being said). An audiogram should be performed to effectively evaluate hearing in both ears. A loss in one ear should prompt an MRI.

Magnetic resonance imaging (MRI) is the preferred diagnostic test for identifying acoustic neuromas. Gadolinium, an enhancing contrast material, is often used during the scan to reveal the tumor. The image formed clearly defines an acoustic neuroma if it is present. Currently MRI is the "gold standard" by which the diagnosis is confirmed. This technique can identify tumors measuring only a few millimeters in diameter.

An auditory brainstem response test (a.k.a. ABR, BAER, or BSER) may be done in some cases. This test provides information on the passage of an electrical impulse along the circuit from the inner ear to the brainstem pathways. An acoustic neuroma can interfere with the passage of this electrical impulse through the hearing nerve at the site of tumor growth in the internal auditory canal, even when the hearing is still essentially normal. This implies the possible diagnosis of an acoustic neuroma when the test result is abnormal. An abnormal auditory brainstem response test should be followed by an MRI.

When an MRI is not available or cannot be performed, a computerized tomography scan (CT scan) with contrast is suggested for patients in whom an acoustic neuroma is suspected. The combination of CT scan and audiogram approach the reliability of MRI in making the diagnosis of acoustic neuroma.

## TREATMENT OPTIONS FOR ACOUSTIC NEUROMA

There are three treatment options available to a patient: 1) Observation, 2) Microsurgical Removal and 3) Radiation ("radiosurgery" or "radiotherapy"). Choosing the best treatment is a decision that must be made by both the patient and the physician after careful review of the size of the tumor, the location of it, the patient's age and physical health. An open discussion should be encouraged about symptoms such as headaches, hearing loss, dizziness and facial numbness and the experience and treatment philosophy of the physicians involved in the patient's care. ANA recommends treatment from a medical team with substantial acoustic neuroma experience.

### 1. Observation – Watch & Wait:

Acoustic neuromas may be discovered incidentally, typically when an MRI is performed to evaluate another condition. Also, the tumor may be discovered when it is very small and there are very few symptoms. Since acoustic neuromas are benign tumors and produce symptoms by slowly applying pressure on surrounding nerves, careful observation over a period of time may be appropriate

for some patients. When a small tumor is discovered in an older patient, observation to determine the growth rate of the tumor may be indicated if serious symptoms are not present. There is now good evidence from large observational studies that suggest many small tumors in older individuals do not grow, thus allowing tumors with no growth to be observed successfully. If it appears that the tumor will not need to be treated during the patient's normal life expectancy, treatment and its potential complications may be avoided. In this case, MRI scans are performed periodically and if the tumor does not show growth, observation is continued. If the tumor grows, treatment may become necessary.

Another example of a group of patients for whom observation may be indicated includes patients with a tumor in their only hearing or better hearing ear, particularly when the tumor is of a size that hearing preservation with treatment would be unlikely. In this group, MRI is used to follow the growth pattern. Treatment is recommended if either hearing is lost or the tumor size becomes life-threatening, thus allowing the patient to retain hearing for as long as possible.

**2. Microsurgical Tumor Removal:**

**A. Subtotal Removal:**

Subtotal removal is indicated when anything further risks life of neurological function. In these cases the residual tumor should be followed for risk of growth (approximately 35%). If the residual grows further, treatment will likely be required. Periodic MRI studies are important to follow the potential growth rate of any tumor.

Older patients with large tumors causing a threat to life may elect to have their surgeon sub-totally remove their tumor. Partial tumor removal has also been advocated in some patients who have large tumors in their only hearing ear. This surgical management will reduce the tumor in size, so that it may cause no threat to the patient's health during his or her life expectancy. This approach may reduce the probability of facial nerve dysfunction as a result of the surgery.

**B. Near Total Tumor Removal:**

This approach is used by experienced centers when small areas of the tumor are so adherent to the facial nerve that total removal would result in facial weakness. The piece left is generally less than 1% of the original and poses a risk of regrowth of approximately 3%. Periodic MRI studies are important to follow the potential growth rate of any tumor.

**C. Total Tumor Removal:**

Many tumors can be entirely removed by surgery. Microsurgical techniques and instruments, along with the operating microscope, have greatly reduced the surgical risks of total tumor removal. Preservation of the facial nerve to prevent permanent facial paralysis is the primary task for the experienced acoustic neuroma surgeon. Preservation of hearing is an important goal for patients who present with functional hearing.

Both facial nerve function and hearing is electrically monitored during surgery. This is a valuable aid for the surgeon while the tumor is being removed.

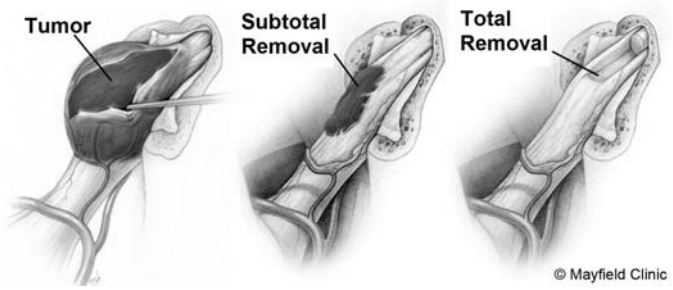


**Figure 4.** Comparison of partial and total tumor removal. Every effort is made to remove the tumor without damaging the adjacent nerves or vital brainstem functions. Sometimes it may be best to leave small pieces of tumor capsule attached to critical structures rather than risk damage. If over time the tumor remnant grows, futher treatment is warranted. *(Printed with permission of the Mayfield Clinic – www.mayfieldclinic.com)*

**D. Surgical Procedures:**

There are three main surgical approaches for the removal of an acoustic neuroma: translabyrinthine, retrosigmoid/sub-occipital and middle fossa. The approach used for each individual patient is based on several factors such as tumor size, location, skill and experience of the surgeon, and whether hearing preservation is a goal. The surgeon and the patient should thoroughly discuss the reasons for a selected appoach. Each of the surgical approaches has advantages and disadvantages, and excellent results have ben achieved using all three of the techniques.

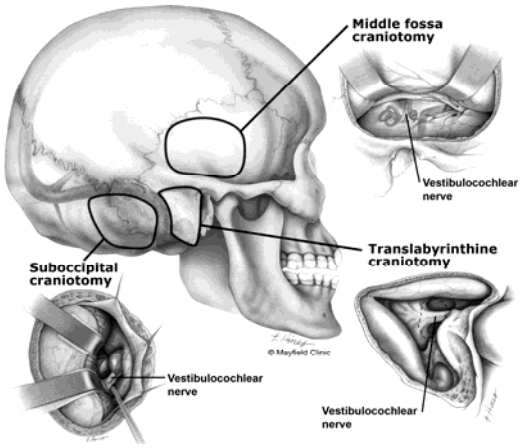


**Figure 5**. Three surgical approaches to an acoustic neuroma: retrosigmoid/sub-occipital, translabyrinthine and middle fossa. *(Printed with permission Mayfield Clinic – www.mayfieldclinic.com)*

• **Translabyrinthine Approach:** The translabyrinthine approach may be preferred by the surgical team when the patient has no useful hearing, or when an attempt to preserve hearing would be impractical. The incision for this

approach is located behind the ear. It involves removing the mastoid bone (the bone behind the ear) and the bone of the inner ear, allowing excellent exposure of the internal auditory canal and tumor. This also results in permanent, and complete hearing loss in that ear. This approach facilitates the identification of the facial nerve in the temporal bone prior to any removal of the tumor. The surgeon has the advantage of knowing the location of the facial nerve prior to tumor dissection and removal. Any size tumor can be removed with this approach. A second, small incision typically is made in the abdomen to harvest fat. This fat is added during surgery and is the substance that is used to prevent a cerebral spinal fluid (CSF) leak after the tumor is removed. This approach affords the least likelihood of long-term postoperative headaches.

• **Retrosigmoid/sub-occipital Approach:** The incision for this approach is located in a slightly different location. This approach creates an opening in the skull behind the mastoid part of the ear, near the back of the head on the side of the tumor. The surgeon exposes the tumor from its posterior (back) surface, thereby getting a very good view of the tumor in relation to the brainstem. When removing large tumors through this approach, the facial nerve can be exposed by early opening of the internal auditory canal. Any size tumor can be removed with this approach. One of the main advantages of the retrosigmoid approach is the possibility of preserving hearing. For small tumors, a disadvantage lies in the risk of long-term postoperative headache.

• **Middle Fossa Approach:** This approach is in a slightly different incision location and is utilized primarily for the purpose of hearing preservation in patients with small tumors, typically confined to the internal auditory canal. A small window of bone is removed above the ear canal to allow exposure of the tumor from the upper surface of the internal auditory canal, preserving the inner ear structures.

**3. Radiation:**

Another treatment option for an acoustic neuroma is radiation. Stereotactic radiation can be delivered as single fraction stereotactic radiosurgery (SRS) or as multi-session fractionated stereotactic radiotherapy (FSR). Both techniques are performed in the outpatient setting, not requiring general anesthesia or a hospital stay. The purpose of these techniques is to arrest the growth of the tumor causing the tumor to die, which is called necrosis.

In single dose treatments, many hundreds of small beams of radiation are aimed at the tumor. This results in a high dose of radiation to the tumor and very little to any surrounding brain structures. Many patients have been treated this way with high success rates. Facial weakness or numbness, in the hands of experienced radiation physicians, occurs in only a small percent of cases. Hearing can be preserved in some cases, with a slightly greater opportunity with FSR.

The multi-dose treatment, FSR, delivers smaller doses of radiation over a period of time, requiring the patient to return to the treatment location on a daily basis, from 3 to 30 times, generally over several weeks. Each visit lasts a few minutes and most patients are free to go about their daily business before and after each treatment session. Early data indicates that FSR may result in better hearing preservation when compared to single-session SRS.

The treatment team should consist of a neurosurgeon, and/or a neurotologist (ear and skull base surgeon), a radiation oncologist and a physicist. Follow-up after SRS and FSR typically involves a MRI scan and audiogram at six months, one year, then yearly for several years, then every second or third year indefinitely to make sure the tumor does not start to grow again.

Patients should understand that all types of radiation therapy for acoustic neuromas may result in "tumor control" in which the tumor cells die and necrosis occurs. Tumor control means that the tumor growth may slow or stop and, in some cases, the tumor may shrink in size. In almost no cases have acoustic neuroma tumors been completely eliminated by radiation treatments. In other words, radiation does not remove the tumor like microsurgery can. Furthermore, radiated patients require lifetime follow-up with MRI scans. Tumors under 2.5 – 3.0 cm, without significant involvement of the brainstem, are more favorable for radiation treatment. Side effects can occur when the brainstem is irradiated and in cases of large tumors, radiation is contraindicated. Patients should understand there have been rare reports of malignant degeneration (a benign tumor becoming malignant) after radiotherapy.

In some cases, the tumor does not die and continues to grow. In those instances, another treatment is necessary – either microsurgery or another dose of radiation. Retreatment must be done as always, in the hands of experienced physicians.

Several types of machines deliver focused radiation treatment suitable for treating acoustic neuromas, such as Gamma Knife® and linear accelerator (LINAC), such as CyberKnife®, Novalis® and Trilogy®. The underlying premise is to treat the tumor with a high dose of radiation while sparing the nerves and brain tissues. Much of the long term data comes from the Gamma Knife literature since this was one of the earliest techniques used to radiate acoustic neuromas on a large scale.

The Gamma Knife uses 195-201 fixed Cobalt-60 radiation sources that are "collimated" to intersect at the site of the tumor and is a single dose treatment. In this way, each individual beam of radiation has very little effect, but where they all intersect produces a maximum effect on the tumor. Very similar results can be obtained using a linear accelerator (LINAC) as the radiation source, such as with the Novalis or CyberKnife with multi-dose treatment.

Studies are beginning to appear for the other modalities. All of the techniques use computers to create three dimensional models of the tumor and surrounding neural structures. Radiation physicists then create dosimetry maps showing the level of radiation to be received by the tumor and the normal tissues. Surgeons, radiation therapists and physicists then modify the dosimetry to maximize tumor doses and minimize radiation toxicity to surrounding normal tissues. The head is stabilized with a metal frame pinned to the head (Gamma Knife) or a fitted mask shield (CyberKnife, linear accelerator, fractionated XRT). Treatments generally last 30-60 minutes. Just like for surgery, the experience of the team in treating acoustic neuromas with all modalities (surgery and radiation) can affect outcomes.

There are a multitude of studies supporting short-term (<5 yrs.) and longer-term (over 10 yrs) tumor control with radiation. Unfortunately, as is the case with microsurgical studies, most have inconsistent follow-up to draw definitive conclusions.

## CAUSE OR ETIOLOGY OF ACOUSTIC NEUROMA

There is a growing body of evidence that sporadic defects in tumor suppressor genes may give rise to these tumors in some individuals. Other studies have hinted at exposure to loud noise on a consistent basis. One study has shown a relationship of acoustic neuromas to prior exposure to head and neck radiation, and a concomitant history of having had a parathyroid adenoma (tumor found in proximity to the thyroid gland controlling calcium metabolism). There are even controversies on hand held cellular phones. Whether or not the radiofrequency radiation has anything to do with acoustic neuroma formation, remains to be seen. To date, no environmental factor (such as cell phones and diet) has been scientifically proven to cause these tumors. ANA does recommend that frequent cellular phone users use a hands free device to enable separation of the device from the head.

## NEUROFIBROMATOSIS (NF2)

NF2, a genetic disorder, occurs with a frequency of 1 in 30,000 to 1 in 50,000 births. The hallmark of this disorder is bilateral acoustic neuromas (an acoustic neuroma on both sides). This creates the perplexing problem of the possibility of complete deafness if the tumors are left to grow unchecked. Preventing or treating the complete deafness that may befall individuals with NF2 requires complex decision making. The trend at most academic U.S. medical centers is to recommend treatment for the smallest tumor which has the best chance of preserving hearing. If this goal is successful, then treatment can also be offered for the remaining tumor. If hearing is not preserved at the initial treatment, then usually the second tumor, in the only-hearing ear, is just observed. If it shows continued growth and becomes life-threatening, or if the hearing is lost over time as the tumor grows, then treatment is undertaken. This strategy has the highest chance of perserving hearing for the longest time possible.

There are now several options to try to rehabilitate deafness in NF2 patients. Implanting the hearing part of the brainstem (Auditory Brainstem Implant) can help restore some sound perception to these patients. Also cochlear implants can be used if the cochlear nerve is preserved following surgery. Radiosurgery may be an option although stereotactic radiosurgery may not have the effect on the NF2 patient as in patients with unilateral sporadic tumors.There are some centers using radiation therapy for NF2 with mixed results. The risk of malignant transformation after radiation is higher in this group. Recent studies have shown that these individuals may have more tumors that are resistant to radiation, due to the cell type. These cases should be handled in centers with very experienced skull base teams.

## AFTER TREATMENT

Surgical removal of an acoustic neuroma is a complex and delicate process. In general, the smaller the tumor at the time of surgery, the fewer the complications. The hospital stay after microsurgery is getting shorter, generally three to five days on average, with approximately four to six to twelve weeks suggested for recovery. Longer stays may be necessary for patients with large tumors. Patients routinely spend at least one night after surgery in the intensive care unit for close monitoring. The time after surgery can be filled with days or perhaps weeks of new sensations. There is usually head discomfort and fatigue. Some patients may experience emotional lows after major surgery, and those lows are believed to be a part of the natural healing process. Some patients require a period of either outpatient or inpatient rehabilitation for balance issues. Other potential postoperative complications (which are discussed preoperatively between patient and surgeon) may prolong hospitalization.

Even when tumor removal has been accomplished, there is a small chance of tumor recurrence. Therefore, a follow-up MRI after tumor removal should be performed within one to five years.

There are few acute side effects following SRS or FSR. Hearing loss, if it is going to occur, usually happens several weeks or months after treatment and can continue to progress. In radiosurgery patients, tumor cell growth is not arrested immediately. Some tumor cells die in a matter of weeks, but others do so more gradually, generally 6 to 18 months after treatment. While this treatment usually arrests the growth of the tumor and some tumors will shrink in size, the tumor does not disappear. Follow-up scans are important because some tumors will continue to grow after this treatment or at some time in the future. It is not possible to determine which tumors will continue to grow larger after radiation. Therefore, periodic MRI's are necessary throughout life.

It is helpful to remain active after treatment as this is the best way to combat the dizziness. Often the expertise of a qualified physical therapist can hasten recovery.

## RESIDUAL PROBLEMS AFTER TREATMENT

A minority of patients experience short and long-term issues after treatment. These include hearing loss, dizziness, facial weakness and headaches. Care and solutions for these issues should be organized by your treatment team. Cerebrospinal fluid leak (CSF), clear fluid from the nose or headache with fever need to be addressed immediately. Besides hearing loss, the most common problems are excessive eye dryness, balance difficulties, tinnitus, facial weakness and headaches. Separate booklets are available from ANA on these topics. (See an order form at the back of this booklet.) Postoperatively, a small percentage of patients experience cerebrospinal fluid leak (CSF) through the incision or nose, and this occurrence should be reported to the surgeon promptly.

Patients with large tumors are likely to have significant hearing loss and are in a situation where preservation of hearing is unrealistic or impossible. In most cases, the best chance to preseve hearing with either surgery or radiation is when the tumor is small.

Patients with partially preserved hearing may benefit from a hearing aid. If there is total single-sided hearing loss, the patient might want to try one of the many hearing devices available. Some use a CROS (Contralateral Routing Of Sound) hearing aid system, whereby a microphone type of hearing aid on the non-hearing ear routes the sound to the normal hearing ear, providing some hearing from the deaf side. The sound may also be conducted from the tumor side to the hearing side via the bone with an implanted bone conduction device. These devices have gained popularity in habilitating single-sided deafness related to acoustic neuromas because of the excellent sound quality and the need for only one device which is worn behind, not in, the ear.

Tinnitus or "ear noise" is common in acoustic neuroma patients, and preservation of hearing may not eliminate the tinnitus. Similarly, removal of the hearing nerve with the tumor generally does not eliminate the tinnitus.

Within up to two years following treatment, the tumor can swell due to inflammation from the radiation and increase symptoms. Typically this is short-term and improves within time. Patients can develop facial numbness, facial weakness or deafness on the side of treatment. These deficits typically occur between 6 to 18 months after treatment.

**NOTE:** In a continuing effort to advance the understanding of acoustic neuromas, ANA occasionally conducts patient surveys. The 2007-2008 patient survey results are available online on our website at www.ANAUSA.org. The results provide information regarding symptoms, diagnosis, treatment and post-treatment issues experienced by acoustic neuroma patients.

## SUMMARY

### TREATMENT OPTION COMPARISON

**Typical Advantages of Microsurgery over Radiation**

1. Surgery removes the tumor for those that want it "out of their body."
2. Some patients have a fear of the very rare long-term (many years post-treatment) effects of radiation, such as induced malignancy.
3. Size and/or position of the tumor may make radiation unadvisable, due to post-treatment swelling. Tumors larger than 2.5 to 3 cm in size are not recommended for radiation.
4. Younger age is generally another determining factor for choosing surgery.
5. Subtotal tumor removal may make surgery the best option, followed by radiation.
6. Some physicians do not recommend radiosurgery for large tumors if there has been prior radiation treatment in the same area.

**Microsurgical Options - 3 Approaches - Pros and Cons**

**Translabyrinthine**

***Pros:***
- Oldest approach – longest history.
- An option when there is no useful hearing to preserve.
- Approach facilitates identification of facial nerve for preservation.
- Any size tumor can be removed with this approach.

***Con:***
- Results are permanent with complete hearing loss in the AN ear.

**Retrosigmoid/Sub-occipital**

***Pros:***
- Possible preservation of hearing - 50% chance of this when the tumor is small to medium size.
- Approach provides a good view of the AN in relation to brainstem.
- Possible preservation of facial nerve.
- Any size tumor can be removed with this approach.

***Cons:***
- Hearing preservation decreases if the tumor is large.
- Headaches are a more prevalent post-op side effect.

**Middle Fossa**

***Pros:***
- Possible preservation of hearing with small tumors in the right location, typically confined to the internal auditory canal.

***Con:***
- Most often used only with small tumors, typically confined to the internal auditory canal.

**Typical Advantages of Radiation over Microsurgery**

1. Good option for patients in their mid-50's and older or with health issues.
2. Radiation is typically an outpatient procedure, though some patients may stay in the hospital overnight. The radiation session itself is relatively quick. Some procedures are done in one session and others take several sessions.

3. There is usually no need to take time off from work. Some people are treated on their way to or from work when having multiple sessions.
4. There is no recuperation or convalescence time immediately after treatment.
5. There are usually no immediate complications. In the medium term there may occasionally be complications, as radiation takes time to fully present symptoms.
6. The tumor can swell for up to 18 months to two years. Symptoms can be worse during this time and may include acute side effects following radiation treatment. Some of these may be short term and while others may last longer and become long term.

**Radiation Treatment Options**
**Gamma Knife**

- Early use for AN began in late 1980's and early 1990's
- Use of head frame attached by pins attached to the patient's skull
- Local anesthetic is used
- Stereotactic radiosurgery
- Always single session

**Linear Accelerator (LINAC)**

- Generally for ANs used for FSR (multiple sessions)
- Cyber-Knife is a well-known variation of the LINAC machine which uses a robotic arm guided by X-ray imaging

**Typical Advantages of "Watch & Wait" over Microsurgery or Radiation Treatment**

1. Good option for small tumors, especially in older individuals; AN may not grow and may not require treatment.
2. Hearing may be preserved longer in cases where the tumor presents on the only hearing side.
3. All medical treatments, surgical or radiation, carry some risks. As ANs are benign and grow very slowly, many physicians will recommend having a second MRI at least 6 months after the first, to establish the growth rate. If the tumor is not growing, avoiding treatment altogether is a possibility.
4. In time, safer treatments for acoustic neuromas, other than surgery or radiation, may be found.

**When to Seek Microsurgical or Radiation Treatment**

- If there is tumor growth
- If the tumor grows to 2 cm or more, treatment should be considered
- An increase in symptoms may indicate that the tumor is growing. Symptoms include increased hearing loss, tinnitus, increased balance issues and numbness in the face.

**ACOUSTIC NEUROMA DECISION TREE**
**A Patients Guide to Discovery, Diagnosis and Treatment**

Begin to experience the symptoms – loss of hearing on one side only, tinnitus, fullness in the ear, problems with balance, vertigo

See physician (a neurologist specialist) if possible.

Hearing test (audiogram) and MRI with contrast should be performed to determine diagnosis

Acoustic neuroma diagnosis

**Patient should:**

1. Learn as much as possible about AN at www.ANAUSA.org
2. Order free ANA information packet which includes the Willing to Talk list
3. Contact others who have had an AN
4. Review all treatment options available based on the size of the tumor, its location, your age, symptoms, status of hearing and overall health
5. After MRI, get copy on disk to share with other physicians when seeking additional opinions
6. Consult with a minimum of two to four physicians, including a neurosurgeon, neurologist and radiation oncologist
7. Consider physician who has substantial experience in treating acoustic neuroma, even if not in your home town
8. Investigate health insurance coverage and determine which physicians/medical center will take your insurance

**Patient works with medical team to choose best course of action:**
Watch and Wait
Microsurgery
Radiosurgery

## ACOUSTIC NEUROMA KEYWORDS

ACOUSTIC: Pertaining to hearing.

ACOUSTIC NEUROMA: Benign tumor of the eighth cranial nerve.

AUDIOGRAM: A chart of hearing acuity recorded during hearing tests.

BENIGN: Not malignant, non-cancerous. Does not invade surrounding tissue or spread to other parts of the body.

BILATERAL: Pertaining to both sides of the body.

BRAINSTEM: Connects the upper brain to the spinal cord - less than three inches (7.6 cm) long (pons is the middle part of the brainstem which may be compressed by acoustic neuromas).

CENTIMETER (cm): .394 inch (2.54 cm equals one inch) - Ten millimeters equal one centimeter.

CEREBELLOPONTINE ANGLE: Space bounded by the petrous bone, brainstem, and cerebellum, and through which cranial nerves six (VI), seven (VII), eight (VIII), nine (IX), ten (X) and eleven (XI) pass.

CEREBELLUM: Located behind the brainstem, extending from the brainstem out toward each mastoid bone. It carries 11% of the brain's weight and controls muscular coordination.

CRANIAL NERVES: The cranial nerves control the sensory and muscle functions around the eyes, face and throat. There are two sets each of twelve cranial nerves. Each set involves one side of the body.

CROS HEARING AID: Contralateral Routing of Sound, a CROS hearing aid system, can be used with single-sided deafness. It receives sound on the deaf side, amplifies it, and carries it to the good ear.

CSF (cerebrospinal fluid): A watery fluid, continuously being produced and absorbed, which flows in the ventricles (cavities) within the brain and around the surface of the brain and spinal cord.

CT SCAN (Computerized tomography): A special X-ray test which creates a cross-sectional picture of any part of the body. This X-ray can distinguish among tissue, fluid, fat, and bone, and, after intravenous injection of a dye, will show an acoustic neuroma unless the tumor is very small.

CYBERKNIFE: A robotic radiosurgery system that delivers multiple beams of radiation, used to treat benign tumors and cancers and other medical conditions located anywhere in the body. It consists of a linear accelerator and a robotic arm, delivered in multiple sessions.

FSR (Fractionated Stereotactic Radiation): Fractionated stereotactic radiation refers to any focused radiation treatment that requires more than one treatment delivery session. Several different dosing schedules are available based on the type of radiation equipment.

GADOLINIUM: A contrast material given at the time of MRI which concentrates in the tumor and makes it more visible.

GAMMA KNIFE: The Gamma Knife is a radiosurgical machine that contains 195 to 201 separate radioactive cobalt sources. The radiation beams from each source (gamma rays) are focused together at the tumor, delivered in one session.

LINAC: Linear accelerator is a radiosurgical machine that produces radiation electronically. These radiation beams are called X-rays. LINACs are also commonly used for conventional radiation treatment of tumors elsewhere in the body.

MAGNETIC RESONANCE IMAGING (MRI): A body imaging system employing a magnet which surrounds the patient. A magnetic field causes small harmless movement of the atoms in the area of the body being studied. A low energy radio wave is then passed through the same area and the small change this imparts to the atoms in the magnetic field causes signals to be emitted which are picked up and analyzed by modern computer technology. An image of the tissue is produced in clear detail. Copies of MRI's are available to patients.

MIDDLE FOSSA: Surgical approach (from above the ear) to an acoustic neuroma primarily used for the purpose of hearing preservation.

NEUROFIBROMATOSIS: A familial condition characterized by developmental changes in the nervous system, muscles, bones, and skin—the central form (Neurofibromatosis Type 2 – NF2) may produce bilateral acoustic neuromas.

NEUROMA: Benign growth originating on a nerve.

PEACOCK RADIATION: The Peacock device is an attachment to the LINAC radiosurgery system that allows complex radiation beam shapes to be generated.

PONS: The middle part of the brainstem located at the base of the brain in front of the cerebellum. This section of the cranium is a mass of nerve tissue which coordinates the activities of the various lobes of the brain.

POSTERIOR FOSSA: The cavity in the back part of the skull which contains the cerebellum, brainstem and cranial nerves 5-12.

PROTON RADIATION: Proton radiation differs from the photon radiation produced by both the Gamma Knife and LINAC systems. Proton beams have a unique physical property that allows them to be sharply focused within a tumor. They are theoretically advantageous for radiosurgery treatment.

RADIOSURGERY (STEREOTACTIC RADIOSURGERY): A technique based on the principle that a single or fractionated dose of radiation delivered precisely to a small area will arrest or kill the tumor, while minimizing injury to surrounding nerves and brain tissue and/or function. Typically this is performed in a single session.

RADIOTHERAPY: Radiation treatment typically delivered over an extended period of time with multiple doses.

RETROSIGMOID/SUB-OCCIPITAL: Surgical approach to an acoustic neuroma creating an opening in the cranium behind the mastoid part of the ear.

SENSORINEURAL HEARING LOSS: Deafness caused by failure of the acoustic nerve.

SHUNT: A tube implanted in the cranium to balance the flow of cerebrospinal fluid and used in the treatment of hydrocephalus.

TINNITUS: A noise produced in the inner ear, such as ringing, buzzing, roaring, clicking, etc.

TRANSLABYRINTHINE: Surgical approach to an acoustic neuroma through the mastoid bone and inner ear (labyrinth).

UNILATERAL: Involving only one side.

VERTIGO: Spinning dizziness - a symptom sometimes caused by an acoustic neuroma.

VESTIBULAR: Associated with the balance system.

**WHAT IS THE ACOUSTIC NEUROMA ASSOCIATION (ANA)?**
Acoustic Neuroma Association was founded in Carlisle, Pennsylvania, in 1981 by a recovered patient, Virginia Fickel Ehr. She found no patient information or patient support available when she had surgery for the removal of an acoustic neuroma in 1977. She resolved that future acoustic neuroma patients should have easy-to-read medical material about their condition, and support and comfort from each other. With the help of her physician, she contacted eight other patients and formed the organization.

The association is incorporated and is a 501(c)(3) non-profit organization. The patient-focused, member organization now serves close to 5,000 members, is governed by an all-patient Board of Directors and is operated by a small staff in metropolitan Atlanta, GA.

ANA membership benefits include receipt of a quarterly newsletter, patient information booklets, access to a network of local support groups, access to a list of acoustic neuroma patients willing to talk about their experience throughout the country, our website Member Section and an invitation to a biennial symposium on acoustic neuroma. Our exclusive website Member Section includes published medical journal articles on acoustic neuroma and all of our patient information booklets and newsletters and many symposium presentations. ANA also maintains an interactive website at www.ANAUSA.org with an ANA Discussion Forum.

ANA is patient-founded, patient-focused and patient-funded. ANA recommends treatment from a medical team with substantial acoustic neuroma experience. Although the association cannot recommend specific doctors, medical centers or medical procedures, guidelines for selecting a qualified medical professional can be found at the ANA website, www.ANAUSA.org. Now available on our website is a listing of medical resources. The physicians and organizations listed have self-reported data to meet criteria established by ANA for having substantial experience in treating acoustic neuromas. The listings should NOT in any way be construed as an endorsement or recommendation by ANA. It is every individual's responsibility to verify the qualifications, education and experience of any healthcare professional.

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