

ACOUSTIC NEUROMAS

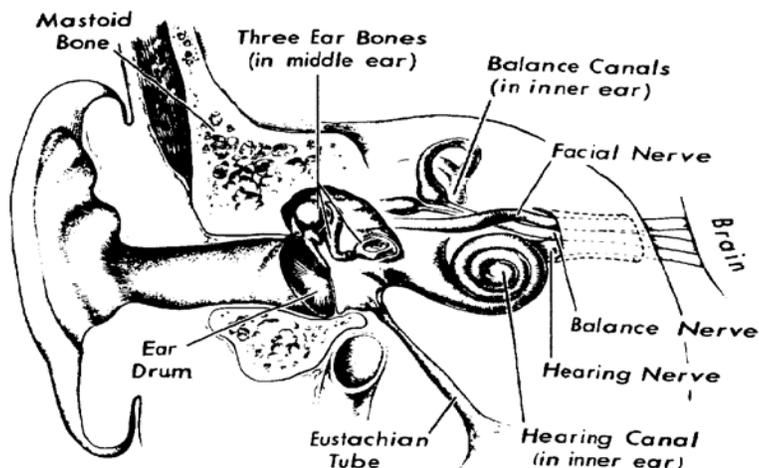
University of Florida ENT Clinic
Patrick J. Antonelli, MD
Matthew R. O'Malley, MD

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ACOUSTIC TUMORS

Acoustic tumors are non-malignant fibrous growths, originating from the balance (vestibular) or hearing (cochlear) nerve, that do not spread (metastasize) to other parts of the body. They constitute six to ten percent of all brain tumors.

These growths are located deep inside the skull and are adjacent to vital brain centers. The first signs or symptoms one notices usually are related to ear function and include noise in the ear or head, hearing loss, and imbalance or vertigo. As the tumors enlarge, they involve other surrounding nerves (eg, the nerve that provides sensation to the face) and brain centers that control vital bodily functions, such as breathing and pumping of blood.



Eventually, tumors may get large enough to cause increased pressure on the brain. If allowed to continue over a long period of time, this pressure on the brain may be fatal.

Thus, the patient with an acoustic tumor has a serious, potentially life-threatening problem. Fortunately, most acoustic tumors grow very slowly before serious problems develop. Some tumors grow more rapidly, so close follow-up with scans (eg, MRI) and examinations by a physician are critical.

Great care is given before, during and after treatment in order to preserve life and bodily function. The preservation of life is the most important objective of treatment in these most difficult cases. A secondary objective of treatment is to preserve as many vital structures as possible. Most patients lead a completely normal life following treatment.

TREATMENT OPTIONS

Surgery

Surgical removal of these tumors is a commonly employed treatment. Surgery can be performed through the inner ear (translabyrinthine), behind the inner ear (suboccipital or retrosigmoid), above the inner ear (middle fossa), or a combination of these approaches. Each may be helpful for acoustic tumors of certain size, locations, and with varying levels of hearing. Hearing is generally lost in translabyrinthine procedures. Useful hearing may be preserved in up to one-half of specially selected patients treated with the suboccipital or middle fossa approaches. Hearing lost before surgery will generally not be recovered by removing the tumor. To accomplish the preservation of life with a minimum of future physical disturbance, this surgery with pre and post operative care is performed and assisted by a team. This team includes an internist, an audiologist, an anesthesiologist, a specially

trained surgical nurse, a neurosurgeon and an otologist (ear surgeon).

Advantages

- The tumor can usually be completely removed, eliminating the need for further treatment.
- Does not expose the whole brain to radiation.
- Results in a stable balance deficit that allows the brain to compensate.
- May be used for any size tumor

Disadvantages

- Greater potential for complications around the brain (eg, swelling, bleeding, and infection) and other nerves (eg, the facial nerve).
- Requires a week in the hospital and up to six weeks of recovery

Radiation

Small to medium size tumors (eg, less than one inch) or recurrent tumors may be treated with special types of radiation. These include stereotactic radiosurgery and the gamma knife. These radiation treatments involve the use of computer guidance to deliver small doses of radiation all over the head, focused on the tumor. Treatment can usually be accomplished in one day, but occasionally, treatment will be administered over several days. These treatments do not remove or completely eliminate the acoustic tumor, but they usually injure the tumor so that it no longer grows. Tumors which remain small do not generally cause any serious problems.

Advantages

- Tumor growth can controlled in more than 90% of patients, eliminating the need for further treatment.
- May be performed as an outpatient
- Less potential for complications around the brain (eg, swelling, bleeding, and infection) and surrounding nerves.

Disadvantages

- Exposes the whole brain to radiation, which may result in subtle changes in thinking processes.
- The balance deficit may continue to change, making it somewhat more difficult for the brain to compensate.
- Should not be used for tumors larger than one inch in diameter.
- May cause cancerous brain tumors in 1 out of 400 patients.

Medical Therapy

Patients with tumors in both ears or medical issues may be candidates for treatment with a medication that may slow or stop tumor growth. The currently available medication, bevacizumab (Avastin) has potentially serious side effects and limited follow-up, so long effectiveness and side effects are not fully understood.

Observation

Some studies suggest that up to half of acoustic neuromas have stopped growing by the time of diagnosis. Thus, in selected patients watching for tumor growth with repeated scans may be an option, particularly if the tumor is not compressing the brain, and if it is not established that the tumor is growing. If unacceptable growth rates are observed, treatment may be performed.

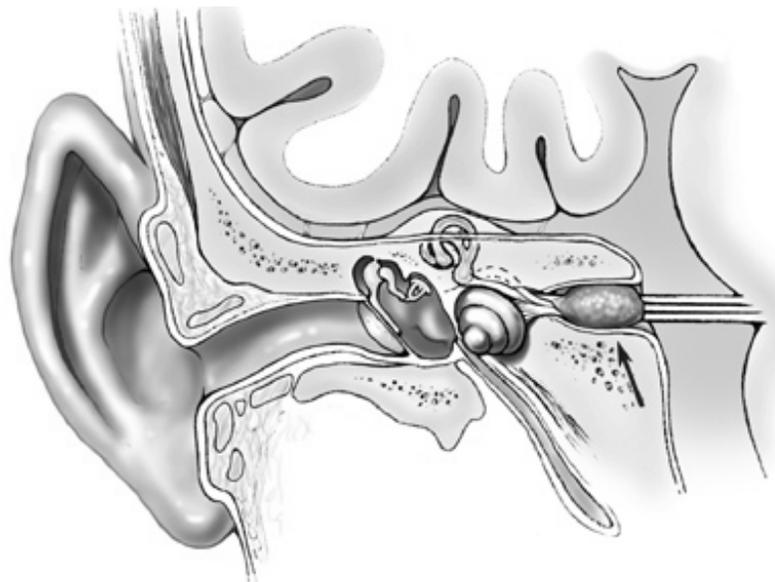
TUMOR SIZE

We classify tumors as large, medium or small. Risks and complications of acoustic tumor surgery vary with the size of the tumor: The larger the tumor, the more serious the complications, and more likelihood of complications. Removal of an acoustic tumor whether large or small, is major surgical procedure, with possibilities of serious complication, including death. The risk involved in the removal of these tumors must never be minimized.

SMALL TUMOR

A small acoustic tumor (< 1 cm) is still confined within the bony canal that extends from the inner ear to the brain. Though this canal pass the hearing, balance and facial nerves and the blood vessels which supply the inner ear.

The operation for removal of a small tumor is performed under general anesthesia using the operating microscope. The surgical approach may be through an incision in front of and above the ear (middle fossa approach) or behind the ear (suboccipital or retrosigmoid, or translabyrinthine). The tumor is gently separated from the hearing and facial nerves. Bone cement and titanium rivets may be used to repair the opening in the bone.



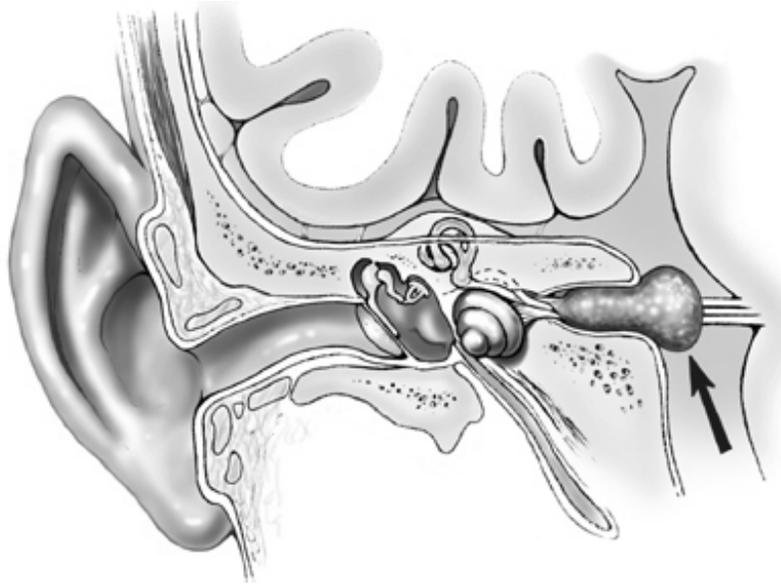
Complete tumor removal is achieved in most cases. On rare occasions only partial removal can be accomplished. Every effort is made to preserve the hearing and still remove the tumor. In about 50% of cases the tumor involves the hearing nerve or the artery leading to the inner ear and total loss of hearing results in the operated ear. If the hearing is poor before surgery, an approach may be used that does not allow for hearing to be preserved (translabyrinthine approach, see below). Normal or near normal preservation of facial nerve function is achieved (long-term) in over 95% of cases.

MEDIUM TUMOR

A medium sized acoustic tumor (1 - 3 cm) extends from the bony canal into the brain cavity but has not yet produced pressure on the brain itself.

The operation for a medium sized tumor is performed under general anesthesia using an operating microscope. The surgical approach (suboccipital or translabyrinthine) is made through an incision behind the ear. The mastoid and the inner ear structures may be removed to expose the tumor (translabyrinthine). The cerebellum (lower portion of the brain) is gently retracted to improve exposure to the more internal extent of the tumor. The tumor is then removed totally by gentle dissecting the tumor off of the underlying facial and hearing nerves. Occasionally only partial removal is accomplished. The bone defect is closed with fat taken from the abdomen or thigh. Bone cement and titanium rivets may be used to repair the opening in the bone.

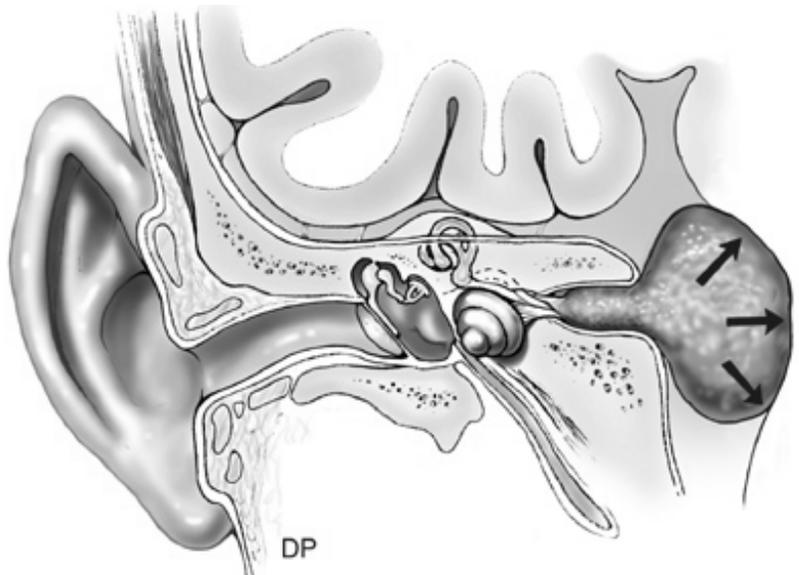
The translabyrinthine approach sacrifices the hearing and balance mechanism of the inner ear. Preservation of the hearing nerve is difficult with the suboccipital approach for tumors larger than 1 ½ cm. Consequently, the ear is usually made permanently deaf. Although the balance mechanism has been removed on the operated ear, the balance mechanism of the opposite ear usually provides stabilization for the patient in one to four months. Preservation of facial nerve normal or near normal facial function is achieved (long-term) in approximately 95% of cases. Temporary weakness of the face is usually mild and resolves over a period of weeks to months.



LARGE TUMOR

A large acoustic tumor (> 3 cm) is one which has extended out of the bony canal into the brain cavity and is sufficiently large to produce pressure on the brain and disturb the vital brain centers.

Operations for large acoustic tumors may require more extensive removal of bone to properly expose the tumor and control the large



blood vessels which obstruct access to the tumor. For this reason special studies may be required along with other procedures necessary to prepare for tumor removal.

The operation for a large tumor is performed under general anesthesia using an operating microscope. The surgical approach (translabyrinthine or suboccipital) is through an incision behind the ear overlying the mastoid bone. The mastoid, inner ear structures and a portion of the skull are removed to expose the tumor (translabyrinthine). Alternatively, bone overlying the cerebellum is removed (suboccipital). The tumor is then totally removed unless vital sign changes occur. If there are changes in the blood pressure, pulse rate or respiration rate the surgery must be terminated before the tumor is totally removed. (In this case a second operation to complete the tumor removal is usually necessary). The bone defect is closed with fat taken from the abdomen or thigh. Bone cement and titanium rivets may be used to repair the opening in the bone.

Removal of a larger tumor sacrifices the hearing and balance nerves and inner ear structures. Consequently, the ear is made permanently deaf. Although the balance mechanism has been removed from the operated ear the balance mechanism of the opposite ear usually provides stabilization for the patient in one to four months. Preservation of facial nerve normal or near normal facial function is achieved (long-term) in 85 - 90% of cases. Temporary weakness of the face is usually mild and resolves over a period of weeks to months.

PARTIAL VS. TOTAL REMOVAL OF AN ACOUSTIC TUMOR

Total removal of an acoustic tumor, without complications, is the goal of the management of these tumors. Partial tumor removal, regardless of its size, may be necessary if the patient's responses during surgery indicate disturbance of the vital brain centers that control breathing, blood pressure, or heart function. If signs of vital brain center disturbance develop during surgery, it is sometimes necessary to terminate the operation before the tumor can be totally removed. This will often allow these vital brain center functions to be restored. Once they are disturbed, however, they sometimes do not recover.

If premature termination of the operation is necessary in the judgement of the operating surgeons, the remaining portion of the tumor may gradually enlarge to again produce symptoms. In that event, a subsequent operation might be necessary. This subsequent operation can often be accomplished without significant changes in vital signs. If large, the bulk of the tumor may be removed at the initial surgery.

In the event your tumor is partially removed, you will be so informed. Usually the first operation reduces the size of the tumor sufficiently so that it has a chance to separate away from the vital brain centers. It can usually be successfully removed at a later date. In most cases we wait four to six months and then electively operate again for tumor removal.

In other cases, a course of continued observation is recommend. In this instance the tumor will be evaluated from time to time for possible regrowth with MRI scans and accordingly a decision made regarding its removal. Alternatively, small tumor remnants may be treated with stereotactic radiation.

RISKS AND COMPLICATIONS OF ACOUSTIC SURGERY

It is not possible to list every complication that might occur before, during and following a surgical procedure. The following discussion is included to indicate some of the risks and complications peculiar to acoustic tumor surgery. In general, the smaller the tumor at the time of surgery, the less chance of complications. As the tumor enlarges, the incidence of complication becomes increasingly greater.

Hearing Loss

In small tumors (ie, less than 1 cm or ½ inch) it is sometimes possible to save the hearing by removing the tumor. Many tumors are larger, however, and the hearing is lost in the involved ear as a result of the surgical procedure. Therefore, following surgery the patient hears only with the remaining good ear.

People with one deaf ear will have trouble with location of the direction of sound, hearing a person on the deaf side and, understanding speech in difficult situations. The patient must learn to watch a speaker carefully in listening situations, using his eyes to help the brain understand words which may sound very much the same, but appear different on the lips (example: pope, coke, soap, dope, cope).

A number of devices are available to assist hearing with one sided deafness. A CROS hearing aid (contralateral routing of sound) is an instrument that receives sound on the deaf side, amplifies it, then routes it to the good ear by either a small wire or use of an FM (wireless) transmitter and receiver. A small device is worn on each ear. Although not everyone will find this type of amplification system helpful, with sufficient need and motivation, the patient usually will realize improved hearing performance with a CROS aid. Alternatively, a bone anchored hearing aid (BAHA) may be used. This involves the surgical placement of a titanium implant behind the deaf ear. The sounds that it receives are transmitted into the skull, which in turn stimulates the unoperated ear. Both the CROS and BAHA devices have advantages and disadvantages. The CROS aid may be tried before proceeding to the BAHA. A BAHA may also be placed at the time that the acoustic tumor is removed.

Tinnitus

Tinnitus (ear noise) remains the same as before surgery in most cases. In 10% of the patients the tinnitus may be more noticeable. In some cases, particularly where the hearing nerve is removed, the tinnitus may be reduced.

Dizziness and Balance Disturbances

In acoustic tumor surgery it is necessary to remove part or all of the balance nerve and the inner ear balance mechanism. Because the balance nerve usually has been damaged by the tumor, its removal frequently results in improvement in any preoperative unsteadiness. Dizziness may occur, nonetheless, following surgery and may be severe for days or a few weeks. Imbalance or unsteadiness with head motion is prolonged in 30% of the patients until the normal balance mechanism in the opposite ear compensates for the loss in the operated ear. Some patients notice unsteadiness when fatigued for several years.

At times the blood supply to the portion of the brain responsible for coordination (cerebellum) is decreased by the tumor or the removal of the tumor. Difficulty in coordination with arm and leg movements (ataxia) may result.

Facial Paralysis

Acoustic tumors are in the intimate contact with the facial nerve, the nerve which controls movement of the facial muscles (eg, eyelids and mouth). Temporary paralysis of the face and muscles which close the eyelids is common following removal of an acoustic tumor. Weakness may persist for six to twelve months and there may be permanent residual weakness.

Facial paralysis may result from nerve swelling or nerve damage. The facial nerve is usually compressed and distorted by the tumor in the internal auditory canal. Careful tumor removal, with the help of an operating microscope, usually results in preservation of the nerve but nerve stretching may result in swelling of the nerve with subsequent temporary paralysis. In these instances facial function is observed for a period of months following surgery. If it becomes certain that facial nerve function will not recover, a second operation may be performed to connect the facial nerve to a nerve in the neck (facial-hypoglossal anastomosis).

In 5% of cases the facial nerve passes through the interior of the acoustic tumor. On occasions the tumor may even originate from the facial nerve (facial neuroma). In either instance it may be necessary to remove all or a portion of the nerve to accomplish tumor removal. When this is necessary it may be possible to immediately reconnect the facial nerve using a skin sensation nerve from the upper portion of the neck. If this is not possible a second operation may be performed to help reanimate the face.

Eye Complications

Should facial paralysis develop the eye may become dry and unprotected. Care by an eye specialist may be indicated. It may be necessary to apply artificial tears or tape the eye shut. When prolonged facial nerve paralysis is expected, implantation of a gold weight or spring into the eyelid helps keep the eye moistened as well as providing comfort and improved appearance.

Taste Disturbances and Mouth Dryness

Taste disturbances and mouth dryness are not uncommon for a few weeks following surgery. The taste nerve is a branch of the hearing nerve. In 5% of patients this disturbance is prolonged.

Other Nerve Weaknesses

Acoustic tumors may contact the nerves which supply the eye muscles, the face, the mouth and throat. These areas may be injured with resultant double vision, numbness of the throat, weakness of the face and tongue, weakness of the shoulder, weakness of the voice and difficulty swallowing. These problems may be permanent.

Headache

Any surgery to remove acoustic tumor can cause headaches. These usually involve spasm of the muscles in the neck or jaw. Others can be caused by inflammation of the lining around the brain. These may require the use of prescription pain medicines. Fortunately, most resolve within weeks to months, and seldom last more than a year. If they do occur, you must be under the care of your surgeon and medical doctor.

Brain Complications and Death

Acoustic tumors are located adjacent to vital brain centers that control breathing, blood pressure and heart function. As the tumor enlarges it may become attached to these brain centers and usually becomes intertwined with the blood vessels supplying these areas of the brain.

Careful tumor dissection, with the help of an operating microscope, usually avoids complications. If the blood supply to vital brain center is disturbed, serious complications may result: loss of muscle control, paralysis, even death. In our experience death occurs rarely as the result of the removal of small acoustic tumors and occurs in less than 1% of the larger tumors.

Postoperative Spinal Fluid Leak

Acoustic tumor surgery results in a temporary leak of cerebrospinal fluid (fluid surrounding the brain). This leak is closed prior to the completion of surgery with fat removed from the abdomen or thigh. Occasionally this leak reopens and further surgery or spinal drainage may be necessary to close it. Leakage of spinal fluid predisposes to infection.

Postoperative Infection

Infection occurs in less than 5% of the patients following surgery. This infection is usually in the form of meningitis, an infection of the fluid and tissue surrounding the brain. When this complication occurs, hospitalization is prolonged. Treatment with high doses of antibiotics is often indicated. These antibiotics can cause allergic reactions, may suppress the body's blood-forming tissues or may produce hearing loss in the good ear. Fortunately these antibiotic complications are rare.

Postoperative Bleeding and Brain Swelling

Bleeding and brain swelling may develop after acoustic tumor surgery. If this occurs a subsequent operation may be necessary to open the wound to arrest bleeding and allow the brain to expand. This complication can result in paralysis or death.

Transfusion reaction

On rare occasions, it may be necessary to administer blood transfusions during or after acoustic tumor surgery. Immediate adverse reactions to transfusions are uncommon. A late complication of transfusion is viral infection of the liver (hepatitis). This occurs in less than 5% of transfused patients. When this complication occurs, medical treatment is necessary, at times requiring rehospitalization. Many patients will consider banking their own blood in advance of surgery. Please ask your doctor for details of such "auto-donations".

CONCLUDING REMARKS

The statements made in this booklet are based on our personal experiences in managing a large series of acoustic tumor cases. If you have any questions about yourself and a possible acoustic tumor, please discuss them with your otologist or neurosurgeon. Also, please refer to the services provided by the **ACOUSTIC NEUROMA ASSOCIATION**. The Acoustic Neuroma Association was established to provide support and information to acoustic tumor patients and to offer therapeutic support to patients with tumor related disabilities.

Acoustic Neuroma Association, Inc.,
P.O. Box 398
Carlisle, Pennsylvania 17013

<http://anausa.org/>

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