Acoustic Neuroma

Diagnosis

Advances in medicine, especially imaging technology have made the identification of small Acoustic Neuromas (AN) possible. After routine auditory tests reveal loss of hearing and speech discrimination (i.e. "I can hear sound in that ear, but can't understand what's being said") a special test for hearing which records responses from the brain-stem called the auditory brainstem response test (ABR, BAER, BSER) maybe done. The results of this test detect the cause of a poorly functioning 8th nerve. If an abnormality in the ABR test suggests an AN, imaging is done to confirm the diagnosis. I do not perform the ABR test in all patients to diagnose an acoustic neuroma because imaging techniques (MRI/CT scans) are the gold standard for diagnosis. CT scan has proven to be a powerful tool in locating AN's. The only drawback is that small tumors confined to the internal auditory canal (IAC) may not show on plain CT scan. Such cases require air or contrast materials to be introduced into the body in order to enhance the tumor. Therefore, the MRI, a more recently developed diagnostic test, has become the gold standard for diagnosis of AN. Gadolinium is the contrast material used to define & enhance the tumor.

Small tumors

A small tumor is also called intracanalicular because it is confined within the bony internal auditory canal (figure). A patient with such a tumor may have hearing loss, ringing in the ear or ear noise, and vertigo or dizziness.

Medium tumors

A medium sized acoustic neuroma is one that has extended from the bony canal into the brain cavity, but has not yet produced pressure on the brain itself (figure). Patients with such tumors have worsening of their hearing, difficulty in balance, in addition to dizziness, and occasionally, the onset of headaches due to irritation of the lining of the brain called dura. Some patients may experience numbness of the mid-face or diminished sensation in the eye during the later stages.

Large tumors

A large tumor is one that is extended out of the internal auditory canal into the brain cavity and is sufficiently large to produce pressure on the brain and disturb vital centers in the brain (figure). During this stage, all previous
symptoms worsen; facial twitch and weakness may occur, and finally patient may develop hydrocephalus due to the blockage of the cavity which contains CSF—the resultant symptoms are headache, visual loss and double vision.

Microscopy

The AN usually arises within the nerve trunk of the vestibular part of the 8th nerve. It gradually grows out of the nerve as it increases in size and assumes a peripheral position. The AN's usually arise halfway along the length of the vestibular nerve, which corresponds to the transition zone of the nerve structure. The typical microscopic appearance of AN's has two distinct features of arrangement of the cells—either tightly packed (Antoni A) or loosely packed (Antoni B) fibers. The distinction of these two cell types is of no clinical importance. Indeed, regions of Antoni A and B may coexist in the same tumor. As the tumor grows, it follows the direction of least resistance, usually towards the brain (cerebellopontine angle) and may reach considerable size. Thus, most tumors consist of 2 parts: a stalk or stem within the internal auditory canal (IAC) and another portion near the brain region. Microscopic investigations into the effect of AN's on adjacent facial or 7th nerve have shown tumor involvement in some cases. This involvement may not be recognized by the surgeon during removal of the AN.

This picture shows the microscopic appearance of a normal vestibular (8th) nerve passing through the internal auditory canal (IAC) to supply the organ of balance. The facial (7th) nerve runs along with the 8th nerve in the IAC. The organ of hearing (cochlea) is also seen in this picture.

This picture shows an AN tumor arising from the 8th nerve, within the IAC.
Origin and Cause

What is an acoustic neuroma?
An acoustic neuroma (sometimes also termed a neurinoma or vestibular schwannoma) is a benign or non-cancerous growth that arises from the 8th or vestibulo-cochlear nerve. The 8th nerve is actually 2 separate nerves, the vestibular nerve and the cochlear nerve. The vestibular nerve is responsible for balance while the cochlear nerve is responsible for hearing. The vestibular nerve has 2 parts—the superior vestibular nerve (SVN) and the inferior vestibular nerve (IVN).
These nerves lie adjacent to each other as they pass through a bony canal, from the inner ear to the brainstem. This bony canal is called the internal auditory canal (IAC) and it varies in length from 0.4 to 1.2 cm. We have two figures of a temporal bone (that part of the skull which has the ear in it) dissection to the right.

The first figure is a view from the top showing the middle ear and the internal auditory canal (IAC) with the nerves passing through it. The organ of hearing (cochlea) and the dura lining the IAC can be seen clearly.

The second figure is a magnification of the IAC region showing the different nerves passing through it. This figure also demonstrates clearly, the cochlear nerve supplying the cochlea. Acoustic neuromas usually arise from the cells of the VIII nerve within the internal auditory canal (third figure).

The third figure is a schematic drawing showing an acoustic neuroma arising from the vestibular nerve within the IAC. The facial or 7th nerve that is responsible for facial movement, along with important blood vessels, also passes with the 8th nerve in the canal (figures).

The cause of acoustic neuroma is unknown. A small percentage of individuals have a hereditary condition called neurofibromatosis type 2 (NF-2). These patients may have an acoustic neuroma on both sides with an aggressive growth pattern and often involve adjacent nerves.

What is the growth pattern?
Acoustic neuromas usually grow very slowly over a period of many years. Once the tumor fully occupies the internal auditory canal, it often begins to erode the walls of the canal and enlarges it. This bony erosion however, does not always occur. They typically remain within their capsule or lining and displace the surrounding nerves and brain tissue very slowly. This is why the body has ample time to accommodate the abnormal growth. The tumor first distorts the 8th nerve, and then presses on the adjacent 7th nerve. The 7th nerve is gradually stretched into a ribbon like structure over the enlarging tumor (figure; cross section of the 7th nerve is shown in the right half of the figure). As the tumor slowly enlarges towards the brain, it protrudes from the internal auditory canal into an area of the skull called cerebello-pontine angle. The tumor is now pear or mushroom shaped with the smaller end within the canal and the larger part towards the brain (figure). It is at this stage that the tumor presses adjacent nerves like the trigeminal or 5th nerve responsible for facial sensation. Ultimately, with increasing tumor size, it can press on the brainstem which can be life threatening.
How often do acoustic neuromas occur?
Acoustic neuromas have been known to occur in all areas of the world without any predilection for individuals of any ethnic background. Small AN's without any symptoms, have been found on autopsy in 2.5% of the general population. Estimates of symptomatic AN range from 1 in every 3,500 to 5 in every million people. It appears that women are more affected than men and most AN's are diagnosed between the ages of 30 & 60 years.
For more information, you may visit the Acoustic Neuroma Association Web site

Symptoms

Early symptoms of AN can occur in other conditions of the ear that can be easily overlooked. Early diagnosis of AN is quite challenging because there is no typical pattern. However, there are symptoms that act as indicators to the possibility if an AN. Patients with "inner ear" problems should be completely evaluated to rule out AN as a cause of these symptoms. It is possible that Meniere's disease or hardening of the bone of the middle ear (otosclerosis) could be causing these symptoms. Patients with AN may present the following symptoms:

- Hearing loss
- Ringing in the ears (tinnitus)
- Dizziness (vertigo)
- Difficulty in balance (imbalance or dysequilibrium)
- Fullness or pressure in the ears
- Facial numbness or paralysis (for very large tumors)

HEARING LOSS
In over 90 percent of the patients with AN, the first symptom is a reduction in hearing in one ear due to involvement of the VIII nerve. This is usually accompanied by ringing in the ears or ear noise-also called "tinnitus". The hearing loss is usually subtle and worsens very slowly over a period of time. In some cases, the hearing loss may be sudden. Some patients may experience a sense of fullness in the affected ear. Unfortunately, since hearing loss is often mild and there is no pain, patients tend to ignore the change in hearing and merely shift the phone to the opposite ear or make other compromises for the one-sided hearing loss rather than seek medical attention.

VERTIGO & IMBALANCE
The tumor usually arises from the vestibular or balance nerve. As a result, unsteadiness or balance problems may be one of the earlier symptoms in the growth of the tumor. Since the remainder of the balance system compensates for this loss, balance problems may be forgotten after some time.

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If the tumor grows larger in size it may start to press on other nerves, mainly the trigeminal nerve, causing facial sensation to become affected. Patients may then experience constant or intermittent numbness and facial tingling. Patients may also have facial tics or spasms. If the tumor grows larger and presses on the brainstem raised intracranial pressure may cause headaches, facial weakness, vertigo and an unsteady gait to ensue.

**Treatment**

**There are 3 treatment options available for AN**

1) Observation

2) Microsurgical removal (partial or total)

3) Stereotactic radiation therapy (radiosurgery)

**Observation**

AN are occasionally discovered incidentally while evaluating another problem or when the tumor is very small with subtle symptoms. Since AN are benign tumors and produce symptoms due to pressure on surrounding structures, careful observation over a period of time may be appropriate for some patients. For instance, a small tumor diagnosed in an elderly patient may only require observation to study the growth rate of the tumor if acute symptoms are not present. If it appears that the tumor will not need to be treated during the patient’s normal life expectancy, treatment and its potential risks and complications maybe avoided. In these patients, MRI is performed periodically to monitor growth of the tumor. If there is no growth, observation is continued. On the other hand, if the tumor shows increase in size, treatment may become necessary. Another group of patients for whom observation is preferred is in patients who have a tumor in their only or better hearing ear, particularly if it is a size where hearing preservation is unlikely. In such cases, periodic MRI is done to monitor growth and surgery is considered only if the hearing is lost or the tumor size becomes life threatening.

**Microsurgical removal**

At the present time, the only treatment that can cure the patient is removal of the tumor by surgery. Within the last 2 decades, microsurgical techniques have been pioneered and refined. Use of the operating microscope, finely scaled surgical instruments, alternate cutting & tumor reducing tools, and better anesthesia, have reduced the death rate extremely. In addition, results have improved as surgeons have gained experience in the delicate removal process of the tumor.

Three main surgical approaches are used depending upon the location, tumor size and hearing level of the patient. They are- middle fossa (MF), sub-occipital (SO), and the trans-labyrinthine (TL) approach. Surgery for AN’s is done under general anesthesia using an operating microscope. Postoperatively, one to several days may be spent in the intensive care with careful monitoring. Problems that may develop in the immediate postoperative period including headache, dizziness, imbalance, vomiting and decreased mental alertness due to the development of a blood clot causing obstruction to the flow of cerebrospinal fluid (CSF).

Other early complications may include cerebrospinal fluid leak and meningitis, an infection controlled with antibiotics that will require a longer hospitalization. Some patients and their surgeons prefer incomplete removal of an AN in order to reduce the risk of complications, realizing that further surgery maybe needed in the future. Occasionally in cases with large tumors, disturbances in the vital brain centers during surgery require ending the surgery prior to complete tumor removal. In these cases, the tumor which was left behind is followed with MRI scans and if tumor growth is demonstrated, further surgery maybe necessary to remove the growing tumor. On the other hand, if the tumor shows no growth, observation is continued. Partial tumor removal maybe also be required in a patient with an only hearing ear such as a Neurofibromatosis-2 (NF 2) patient. Unfortunately, partial removal may result in substantial hearing loss in these patients and this risk must be considered.
Small tumor

If the hearing is still preserved in such tumors, a middle fossa approach, incision for which is in front of the ear (figure) may be considered. A small square piece of bone from the side of the skull is then removed (blue shaded area in the figure). The tumor is removed completely in most cases. On rare occasions, partial removal is possible. This approach attempts to preserve the hearing in all cases while removing the tumor. In about half of the patients, the tumor involves the hearing nerve or the artery supplying the inner ear and in such cases, total loss of hearing occurs in the operated ear. In addition, the risk to the facial nerve is far greater in this approach.

Medium tumor

The operation for medium sized tumors is performed by the sub-occipital and/or the trans-labyrinthine approach. The incision for these approaches is behind the ear, overlying the mastoid, the bony projection felt behind the ear (figures). The mastoid and the inner ear structures are removed to expose the tumor, and remove it completely. The opening created in the mastoid bone is closed with fat taken from the abdomen. The translabyrinthine approach sacrifices the hearing and balance mechanism since the inner ear is entered. Consequently, the ear is made permanently deaf. In such cases, the balance mechanism of the opposite ear compensates for the non-functioning operated ear and provides stabilization for the patient within few weeks to months.

Large tumor

Surgery for large tumors requires extensive removal of bone to properly expose the tumor and control the large blood vessels that make access to the tumor difficult. For this reason, special studies of the arteries (angiograms) may be required in addition to the other investigations, in order to diagnose and establish the size of the acoustic tumor. The operation for large tumors is performed by the TL-SO approach as described for medium tumors. The figure to the right shows the area of the skull approached via the TL and the SO approaches. In these patients, total removal is attempted unless changes in vital signs occur. If there are changes in blood pressure, pulse rate, or respiratory rate, the surgery must be terminated even if the tumor has not been totally removed. The opening in the mastoid is closed with abdominal fat. For large tumors, it is often necessary to monitor the patient’s general status by inserting a small tube (arterial line) into an artery in the arm or leg. In these cases, occasionally a blood clot may form in the artery following surgery. In case this complication occurs, further surgery maybe required to remove the blood clot. A very rare complication of this arterial line monitoring is the loss of a finger, toe, or even a hand or a foot.

Stereotactic Radiation Therapy (Radiosurgery):

This is a technique based on the principle that a single relatively high dose of radiation delivered precisely to a small area will arrest or kill the tumor while minimizing injury to the surrounding nerves & brain tissue. The source of radiation is from either radioactive cobalt (called gamma ray) or a linear accelerator (LINAC). The treatment team consists of a neurosurgeon, radiophysicist and a radiation oncologist working together to develop a treatment plan based on the size & shape of the tumor. Radiation, even at relatively high doses such as those used in radiosurgery, does not kill or injure cells immediately. Some tumor cells die in weeks while others die more gradually over 6-18 months after radiation. This treatment usually arrests growth of the tumor and some tumors shrink, but they rarely disappear.

Follow-up of these patients is important because approximately 20% of tumors continue to grow after radiosurgery or at some time in the future. A tumor that has been irradiated and grows may be more difficult to remove than an un-radiated tumor. Symptoms such as dizziness & disturbances in balance typically improve earlier after microsurgical tumor removal than after radiosurgery. This is because effects of radiosurgery may require up to 18 months. Residual dizziness & imbalance may be less after microsurgical treatment. The side effects of radiosurgery may be headache, dizziness, nausea, facial numbness, or rarely, cranial nerve paralysis. In the long term requires follow-up MRI's over the years and there is a potential for additional treatment in

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cases of continued growth or later re-growth.

Microsurgery requires follow-up MRI's suggested at perhaps 1 and 5 years if the tumor has been completely removed. Radiosurgery may be considered in selected patients in whom the risk of surgery is excessive because of advanced age or pre-existing health problems, patients having small to moderate sized tumors or patients with tumors on both sides, or in the only hearing ear.

Postoperative

Microsurgery of an AN is a complex and delicate procedure. The smaller the tumor at the time of surgery, the fewer the chances are for complications. As the tumor size increases, the chances of complications become greater. Thus, there may be problems with the cranial nerves affected by the tumor (like facial paralysis or hearing loss) following surgery that may or may not have been present before tumor removal. Here is a list of some of the more common post-operative issues and problems encountered.

Residual problems

This period is the days or perhaps weeks following surgery. There is a possibility of fatigue or tiredness and increased drowsiness, although some patients may experience "survival euphoria" and a renewed sense of energy and vigor. A period of emotional lows is common as the patient adjusts to physical changes. One symptom that may occur after discharge is a nasal drip of clear colorless fluid, which is particularly noticeable when bending over. This may indicate a cerebrospinal fluid leak and should be reported to the surgeon right away due to the risk of infection.

Follow-up period :After being discharged from the hospital, patients operated for an AN are followed up regularly (every 2-3 months for the first year, every 6 months for the 2nd year, and every year thereafter). These follow up visits are important to monitor the hearing (in patients operated by the MF or SO approach), facial nerve paralysis if any and for recurrence of tumor.

HEARING LOSS

With small tumors, it may be possible to save hearing. In larger tumors, especially those that have extended into the brain cavity, the hearing has usually been partially or totally lost and cannot be restored. This loss means the patient will continue having problems locating sound, hearing on the deaf side and understanding speech over high background noise. Consultation with an audiologist is required for these patients for amplification options like traditional hearing aids or a CROS hearing aid (a device which crosses sound over from the operated ear to the opposite ear) or a BAHA.

TINNITUS

Ear noises usually remain the same as before surgery, though in a few cases noises may increase or begin after surgery. A masking device may help some people affected by tinnitus.

FACIAL WEAKNESS OR PARALYSIS

Since the facial nerve which controls muscles of facial expression is in close proximity with the AN, it is usually necessary to manipulate and at times remove the portion of the nerve. In some cases however, even though the nerve is intact after surgery, nerve damage or swelling may cause temporary or in some cases permanent facial paralysis. Regrowth of the nerve is a slow process that may take up to a year for recovery to be noticeable. If recovery is not observed by 1 year, a second operation may be required to connect the healthy portion of the facial nerve to a nerve in the neck usually the one supplying one side of the tongue. This procedure is called the hypoglossal-facial nerve anastamosis and can restore some but not all facial movement. Spontaneous movements like laughing are asymmetric. There may be loss of tongue function. There are some other procedures that adapt available muscles and nerves to help in toning or reanimating the sagging face. If it becomes necessary to remove a portion of the facial nerve during surgery, the facial nerve may be reconnected directly or by inserting a nerve graft. Usually, the result is asymmetric but will provide some spontaneous movement.
EYE PROBLEMS

Studies have shown that at least half of those who have had an acoustic neuroma removed develop long term eye discomfort and other eye problems, particularly if the tumor was medium or large. Loss of eyelid function and/or altered tear production can cause irritation and scratchiness in the eye because it is dry & unprotected. To deal with this problem, there are various surgical procedures that can be done to protect the cornea. They include canthoplasty (bringing together tendons in either or both corners of the eye), a spring implantation in the upper lid, an elastic prosthesis secured around the upper and lower lids, a gold weight implant in the upper lid; and a tarsorraphy (sewing the lids together). Artificial tears or eye lubricants maybe needed for a short time or permanently. Taping part of the lids together, using protective glasses and moisture chamber, using bandage contact lenses and avoiding eye irritants may be helpful. In a few patients, double vision may be present due to pressure on the 6th cranial nerve that controls the muscles that move the eyes.

TASTE DISTURBANCE AND MOUTH DRYNESS OR EXCESSIVE SALIVATION

There maybe some changes in taste and amount of saliva secretion for a short time following surgery. In some cases this may be prolonged. In the others, increased salivation occurs while chewing or there maybe increased tearing while eating. The appetite maybe affected for some time.

SWALLOWING, THROAT AND VOICE PROBLEMS

In a small number of patients, AN surgery affects the nerves which control the throat, swallowing and voice production leading to hoarseness & difficulty in swallowing. These symptoms usually improve slowly over time.

BALANCE PROBLEMS

The vestibular portion of the VIII nerve is almost always removed during surgery. Usually this part of the nerve is non-functional and has already been destroyed because of the AN. Dizziness is common following surgery and maybe severe for a time. After a while, the balance apparatus of the opposite or normal ear compensates for this loss, and balance improves. This compensation may not be perfect, particularly in darkness, when the patient is fatigued, when there is a sudden change in body position, or while walking on uneven surfaces. Maintaining a good general physical health through proper diet and moderate exercise, can improve balance & general vitality to a great extent.

FATIGUE

Fatigue sometimes remains a prolonged problem for some patients after some of the other symptoms have subsided. It is important in such patients to adjust their pace of life in harmony with their energy level.

HEADACHE

Headaches can be a problem for some patients while still in the hospital. This maybe related to tension from holding the head rigidly, changes in intracranial pressure, muscle spasm, or anxiety. Headaches are almost never related to tumor recurrence. Treatment is with analgesics & muscle relaxation. If severe headaches persist after hospital discharge, medical help should be sought.

DENTAL CARE

If the patient has facial paralysis, food tends to get lost in the mouth on the affected side and can lead to dental problems. Washing and rinsing the mouth is therefore necessary, as well as brushing & flossing the teeth several times a day is important.

PROTECTING THE OTHER EAR

It is important to provide sensible protection to the opposite or good ear that has the remaining hearing apparatus. This is done by avoiding extreme or sudden exposure to loud noises like firearms or some cordless phones near the good ear. Some physicians suggest follow-up MRI scans and/or audiograms for some time following AN removal.
PSYCHOLOGICAL COPING
For some patients, adjustment to a new self after AN removal can be a challenging task. This is because in addition to changes in hearing, the appearance may now be altered along with the presence of other impairments. Return to normal activity may be slow. Concentrating on strengths rather than on weaknesses will help such patients to return to all former activities and also expand their abilities in new areas.