

ACOUSTIC NEUROMA

This handout is intended as a general introduction to the topic. As each person is affected differently, speak with your health care professional for individual advice.

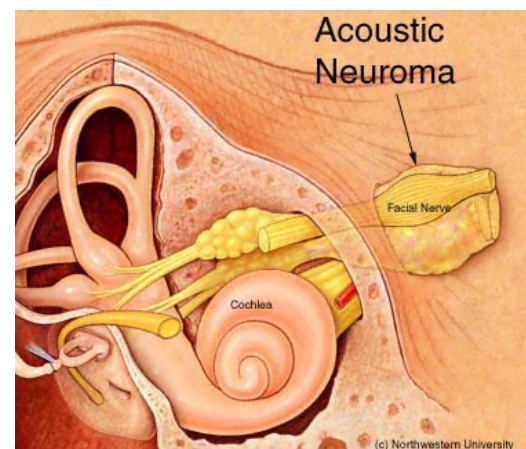


Key points

- A rare benign (not cancerous) tumour on the nerve that runs from the inner ear to the brain (vestibulocochlear nerve).
- Most common in 30- to 60-year-olds but can happen at any age.
- Slow growing, usually.
- Most common first symptom is hearing loss in one ear.
- Other symptoms include tinnitus (ringing in the ear), imbalance, and vertigo (spinning sensation).
- Treatment options include observation, surgical removal, or radiation.
- Certain drugs may be prescribed “off-label” to slow down growth of the tumour.
- Vestibular rehabilitation (an exercise-based therapy) and physiotherapy soon after surgery can help with vertigo and balance problems.
- In rare cases, can be life-threatening if the tumour gets very large and is not treated.
- Hardly ever comes back after treatment.

What is acoustic neuroma?

Acoustic neuroma is a benign (not cancerous) tumour on the vestibulocochlear nerve (8th cranial nerve). It usually grows slowly. The vestibulocochlear nerve carries balance and hearing information between the inner ear and the brain. This means that as the tumour gets bigger, it can cause hearing loss and tinnitus on the affected side, dizziness, and balance problems.



Acoustic neuroma – swelling of vestibulocochlear nerve (8th cranial nerve), just under facial nerve

Acoustic neuroma is also called vestibular schwannoma (shwa-NO-ma) or neurilemmoma (noo-roe-lem-OH-ma).

Acoustic neuroma affects about 1 person in 100,000 each year. It is most common in people aged 30 to 60, but it can happen at any age.

If an acoustic neuroma gets very large, it can interfere with the brainstem and the cerebellum (sar-a-BELL-um). These parts of the brain help to control the body's basic functions, including breathing, heartbeat, sleep, eating, and movement. This means that in rare cases, acoustic neuroma can be life-threatening if it is not treated.

What are the causes?

Acoustic neuroma happens when the cells that wrap around the vestibulocochlear nerve start to grow out of control. These cells are called Schwann cells. They produce a substance called myelin (MAI-uh-luhn), which acts like insulation around an electric wire and helps the nerves to carry information around the body.

Scientists believe that Schwann cells start to grow out of control because of a change in the genetic code (a mutation) on chromosome 22. The affected gene controls the growth of Schwann cells.

In most cases, an acoustic neuroma happens randomly and there is a tumour only in one ear.

In some cases, though, acoustic neuroma happens as part of a syndrome called neurofibromatosis 2 (NF2). If someone has acoustic neuroma as part of NF2, they usually have tumours in both ears, not just one. NF2 makes tumours grow in the brain and on many different nerves, not just the vestibulocochlear. People with NF2 usually start to develop symptoms when they are teenagers or young adults.

About half of people with NF2 inherit it from one of their parents. The other half seem to get it spontaneously, meaning that nobody else in their family has the condition.

What are the symptoms?

Acoustic neuroma usually includes the following symptoms:

- hearing loss in one ear, or asymmetric hearing loss (more in one ear than the other), which can be either sudden or progressive in nature
- tinnitus (ringing in the affected ear)
- dizziness or vertigo
- loss of coordination
- loss of balance or trouble walking
- visually induced dizziness (sensitivity to visually busy environments)

Many of these symptoms can be caused by other conditions as well, so it may not be easy to diagnose acoustic neuroma, especially in the early stages. But asymmetric signs and symptoms (more on one side than the other) are often caused by an acoustic neuroma, so a healthcare professional will always want to rule out an acoustic neuroma when making a diagnosis.

As the tumour gets bigger, it can start to press on other nerves nearby, such as the facial nerve and the trigeminal nerve. This can cause various problems, including:

- numbness, tingling, weakness, or paralysis of the face
- double vision
- trouble with swallowing or speaking

How is it diagnosed?

Acoustic neuroma is usually diagnosed by a specialist, such as a neurologist or an otolaryngologist (an ear, nose, and throat or ENT doctor).

Your doctor will ask about your symptoms and your medical history. Your doctor will also do a thorough physical and neurological exam, including an ear exam.

You will likely have the following diagnostic tests:

- hearing and balance tests (audiogram, vestibular function tests and auditory brainstem responses)
- imaging (MRI scan)

If someone has asymmetric hearing and balance symptoms (more on one side than the other), a health care professional will always order an imaging test to rule out an acoustic neuroma. An MRI scan with contrast (using gadolinium-based dye) is the gold standard for diagnosing an acoustic neuroma. It can find very small tumours that might not have been noticed without an MRI with contrast.

How is it treated and managed?

Treatment for acoustic neuroma will depend on how large the tumour is, where it is, how fast it is growing, what symptoms it is causing, your age and health, and what you would prefer. Your doctor will discuss the risks and benefits of each treatment option with you.

If the acoustic neuroma happened because of NF2, you would need treatment from a team of health care professionals who have experience with NF2.

Observation

If the acoustic neuroma is small and is not causing symptoms, your doctor may suggest watching it for a while to see if it grows or causes symptoms (“watchful waiting” or the “wait and see” approach). This means you will have regular follow-up appointments and MRIs. If anything changes, your doctor may decide that treatment is needed.

Surgery

If the acoustic neuroma is small, it may be possible to remove it with surgery (an operation). If the surgery goes well, it can preserve hearing and improve other symptoms.

If the acoustic neuroma is larger, it may already have damaged the vestibulocochlear nerve and other nerves nearby. This means that surgery may be more difficult, or it may not help with the symptoms.

Surgery should be done by a neurosurgeon who specializes in removing acoustic neuromas. There are several different surgical options. Which procedure is used will depend on various factors, including:

- how big the acoustic neuroma is
- what symptoms it is causing
- how much it is affecting your hearing
- how experienced your surgeon is with each procedure

Surgery for acoustic neuroma can sometimes make the symptoms worse because it may damage the vestibulocochlear nerve and other nerves nearby. Compared with other treatment options, surgery is more likely to damage your hearing and vestibular function permanently.

Radiation treatment

Radiation treatment involves sending large doses of radiation to a small area of the body. The goal is to kill the tumour cells without damaging the healthy parts of the body nearby. Radiation treatment is often a good option for small, non-malignant tumours like acoustic neuromas. Compared with surgery, it may be a better option for preserving your hearing.

Your doctor may suggest radiation treatment if:

- you would prefer not to have surgery, or surgery is not a good option for you
- the acoustic neuroma is affecting your only hearing ear, or you have acoustic neuromas affecting both ears

There are two options for radiation treatment:

- stereotactic radiosurgery, which is a single, focused dose of radiation
- radiotherapy, which is multiple lower doses of radiation over several days

Vestibular rehabilitation

Surgery for an acoustic neuroma can damage your vestibular nerves. This can cause balance problems and vertigo. Surgery can also damage the nerves that control the muscles in your face, including the facial nerve and the trigeminal nerve. This can cause weakness and trouble with movement. Vestibular rehabilitation and physiotherapy soon after surgery can help with these problems.

Vestibular rehabilitation is a type of exercise therapy. Its goal is to help your brain compensate for the loss of balance function on one side. With vestibular rehabilitation, your brain can relearn how

to balance and how to respond to signals from the visual and vestibular systems. In many patients, this compensation happens naturally, so they do not need vestibular rehabilitation. Other patients, however, may continue to have symptoms, and therefore they may benefit from vestibular exercises. One study found that patients who followed a customized vestibular rehabilitation program after surgery for an acoustic neuroma did better on balance tests than patients who were only given general instructions. A vestibular therapist can help design an appropriate program for you.

Vestibular rehabilitation after surgery for acoustic neuroma may include:

- moving your head while you look at an object that is standing still
- exercises to help you balance better while you are standing or walking

It is very important to start exercises gradually and increase them slowly and steadily. If you try to do too much, too soon, your dizziness may get worse.

Facial retraining

In uncommon cases, acoustic neuroma removal surgery damages the facial nerve leading from the affected side to the brain. Paralysis or drooping on one side of your face is the result. Facial retraining, sometimes called facial neuromuscular re-education (NMR), uses your brain's ability to change and adapt as a result of experience (neuroplasticity). Active and persistent patient participation is needed to retrain the brain as the nerve regrows. The goals of facial retraining are a more symmetrical face, regained control of the muscles used in facial expression, and increased facial range of movement. A physiotherapist or occupational therapist specializing in facial retraining, also known as can help design an appropriate program for you.

Facial retraining for acoustic neuroma might include:

- exercises to improve muscular control and decrease unwanted movement

- mime therapy using a mirror to “trick” your brain into thinking that the paralyzed side of your face is moving as you look at the other side in a mirror
- manual stretching and self-massage techniques to decrease muscle tightness

It is important to practice the exercises regularly at home for change and recovery to happen.

Acoustic neuroma removal surgery sometimes damages your facial leading from the affected side to the brain.

New treatments for acoustic neuroma

Recent research suggests that certain drugs may slow the growth of acoustic neuroma, including:

- bevacizumab (Avastin[®], Mvasi[®], Zirabev[®]), cancer chemotherapy drugs
- mifepristone (Mifegymiso[®]), used to treat Cushing's syndrome and for medical termination of pregnancy

Right now, these treatments are not approved for treating acoustic neuroma. But your doctor may decide to prescribe one of them anyway (off-label use), or you may decide to join a clinical trial of one of these drugs.

What to expect in the future

If you have surgery or radiation treatment, you will need regular follow-up appointments, including MRI scans and hearing tests, to make sure that nothing has changed. It is very rare for an acoustic neuroma to come back after treatment.

Many people still have hearing loss, tinnitus, dizziness, or other symptoms after treatment:

- One study found that 8 years after treatment for acoustic neuroma, 3 in 4 patients were deaf in the affected ear. People who already had some hearing loss before they were treated were more likely to lose their hearing completely.
- Another study found that eight years after treatment, about 1 in 2 patients still had some dizziness symptoms. People who had larger

tumours or symptoms of dizziness, headache, or migraine before they were treated were more likely to have dizziness later on. Vestibular rehabilitation can help to reduce dizziness.

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View this and other articles about vestibular disorders – www.balance&dizziness.org.

In addition, find information about how the balance system works, the journey from diagnosis to treatment, building a wellness toolkit, and more.

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