

# Acoustic Neurinoma and Neurofibromatosis

## What is an acoustic neurinoma?

An acoustic neurinoma is a benign tumor that may develop on the hearing and balance nerves near the inner ear. The tumor results from an overproduction of Schwann cells—small sheet-like cells that normally wrap around nerve fibers like onion skin and help support the nerves. When growth is abnormally excessive, Schwann cells bunch together, pressing against the hearing and balance nerves, often causing gradual hearing loss, tinnitus or ringing in the ears, and dizziness. If the tumor becomes large, it can interfere with the facial nerve, causing partial paralysis, and eventually press against nearby brain structures, becoming life-threatening.

Early diagnosis of an acoustic neurinoma is key to preventing its serious consequences.

## How is an acoustic neurinoma diagnosed?

Early diagnosis of an acoustic neurinoma is key to preventing its serious consequences. Unfortunately, early detection of the tumor is sometimes difficult because the symptoms may be subtle and may not appear in the beginning stages of growth. Also, hearing loss, dizziness, and tinnitus are common symptoms of many middle and inner ear problems. Therefore, once the symptoms appear, a thorough ear examination and hearing test are essential for proper diagnosis. Computerized tomography (CT) scans and magnetic resonance imaging (MRI) are helpful in determining the location and size of a tumor and also in planning its microsurgical removal.

## How is an acoustic neurinoma treated?

If an acoustic neurinoma is surgically removed when it is still very small, hearing may be preserved and accompanying symptoms may go away. As the tumor grows larger, surgical removal is often more complicated because the tumor may become firmly attached to the nerves that control facial movement, hearing, and balance.

The removal of tumors attached to hearing, balance, or facial nerves can make the patient's symptoms worse because sections of these nerves must also be removed with the tumor. As an alternative to conventional surgical techniques, radiosurgery may be used to reduce the size or limit the growth of the tumor. Radiosurgery, utilizing carefully focused radiation, is sometimes performed on the elderly, on patients with tumors on both hearing nerves, or on patients with a tumor growing on the nerve of their only hearing ear. If the tumor is not removed, MRI is used to carefully monitor its growth.

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## What are the two types of acoustic neurinoma?

There are two types of acoustic neurinoma: unilateral and bilateral. Unilateral acoustic neurinomas affect only one ear. They account for approximately 8 percent of all tumors inside the skull. Symptoms may develop at any age but usually occur between the ages of 30 and 60 years.

Bilateral acoustic neurinomas, which affect both ears, are hereditary. Inherited from one's parents, this tumor results from a genetic disorder known as neurofibromatosis-2 (NF2). Affected individuals have a 50 percent chance of passing this disorder on to their children. Unlike those with a unilateral acoustic neurinoma, individuals with NF2 usually develop symptoms in their teens or early adulthood. Because NF2 patients usually have multiple tumors, the surgical procedure is more complicated than the removal of a unilateral acoustic neurinoma. Further research is needed to determine the best approach in these circumstances.

In addition to tumors arising from the hearing and balance nerves, NF2 patients may develop tumors on other cranial nerves associated with swallowing, speech, eye and facial movement, and facial sensation. NF2 patients may also develop tumors within the spinal cord and on the brain's thin covering.

Scientists believe that both types of acoustic neurinoma form following a loss of the function of a gene on chromosome 22. A gene is a small section of DNA responsible for a particular trait like hair color or skin tone. Scientists believe that this particular gene on chromosome 22 suppresses the growth of Schwann cells. When this gene malfunctions, Schwann cells can grow out of control. Scientists also think that this gene may help suppress other types of tumor growth. In NF2 patients, the faulty gene on chromosome 22 is inherited. For individuals with unilateral acoustic neurinoma, however, some scientists hypothesize that this gene somehow loses its ability to function properly as a result of environmental factors.

## What is being done about acoustic neurinoma?

Once the gene that suppresses Schwann cell growth is "mapped" or located, scientists can begin to develop gene therapy to control the overproduction of these cells in individuals with acoustic neurinoma. Also, learning more about the way genes help suppress acoustic neurinoma may help prevent brain tumors and lead to a treatment for cancer.

## Where can I get additional information?

### Acoustic Neuroma Association (ANA)

P.O. Box 12402  
Atlanta, GA 30355  
(404) 237-8023 (Voice)  
anusa@aol.com (E-mail)  
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### American Academy of Otolaryngology-Head and Neck Surgery (AAO-HNS)

One Prince Street  
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