CONDITION

Bilateral acoustic neuroma

Learn how bilateral acoustic neuromas affect hearing and balance, their link to NF2-related schwannomatosis, and how they are diagnosed, treated, and monitored.

WHAT IT IS
Noncancerous Schwann cell tumors on both hearing and balance nerves

BODY SYSTEM
Nervous system

MAIN SYMPTOMS
Hearing loss, tinnitus, poor balance, vertigo

WHAT CAUSES IT
Inherited or new NF2 gene change

HOW IT IS DIAGNOSED
MRI of the brain and internal auditory canals

MAIN TREATMENT
Monitoring, surgery, focused radiation, medicines

CAN IT BE CURED?
Sometimes, surgery removes the entire tumor

IS IT AN EMERGENCY?
Sometimes, severe worsening headache with vomiting

LONG-TERM OUTLOOK
Effects vary widely with possible bilateral hearing loss

Overview

An acoustic neuroma is a noncancerous tumor that grows from Schwann cells, which cover and protect nerves. The medical name is vestibular schwannoma . These tumors usually begin on the balance portion of the vestibulocochlear nerve, also called the eighth cranial nerve. This nerve carries hearing and balance signals between the inner ear and the brain.

Bilateral acoustic neuroma means that vestibular schwannomas are present on both sides. This can affect hearing and balance in both ears. The tumors do not usually spread to distant parts of the body, but they can grow and press on nearby nerves or the brainstem.

Bilateral vestibular schwannomas are strongly associated with NF2-related schwannomatosis , previously called neurofibromatosis type 2. This is a genetic condition that can also cause other nerve tumors, meningiomas around the brain or spinal cord, spinal tumors, eye changes,

and nerve problems. Finding tumors on both sides should therefore lead to an assessment for this condition, even when there is no known family history.[1-3]

The effects vary widely. Some tumors grow slowly and can be monitored for years. Others threaten useful hearing or cause pressure on nearby structures. Because treatment itself can affect hearing, balance, and facial movement, management usually requires careful planning by a team experienced in vestibular schwannomas and NF2-related schwannomatosis.

Symptoms

Symptoms depend on the tumors' size, location, and rate of growth. A small tumor can still affect hearing, while a larger tumor may cause few symptoms until it presses on nearby structures.

Possible symptoms include:

With bilateral tumors, hearing loss may develop at different times in each ear. Changes in speech understanding can be important even when a standard hearing test shows only a modest change in hearing thresholds.

Symptoms elsewhere in the body may come from NF2-related schwannomatosis rather than from the vestibular schwannomas themselves. These can include weakness or numbness in an arm or leg, persistent back pain, muscle wasting, vision changes, or problems related to other brain, spine, or peripheral nerve tumors.

Sudden hearing loss should receive prompt medical assessment, ideally the same day. Sudden hearing loss has several possible causes, and early evaluation may affect treatment options.

Seek emergency care for a new seizure, fainting, severe or rapidly worsening headache with vomiting, confusion, major weakness, or difficulty staying awake. New facial drooping, speech difficulty, or one-sided body weakness can also be signs of a stroke and should not be assumed to come from an acoustic neuroma.

- Gradual hearing loss in one or both ears
- Difficulty understanding speech, especially in noise
- Tinnitus, meaning ringing, buzzing, or another sound without an outside source
- Unsteadiness or poor balance
- Episodes of vertigo, a spinning or moving sensation
- A feeling of fullness in an ear
- Numbness, tingling, pain, or weakness in the face

- Headache, nausea, or coordination problems when a large tumor causes pressure

Causes and risk factors

Most single, one-sided vestibular schwannomas occur without a clear inherited cause. Bilateral vestibular schwannomas are different: they are a key feature of NF2-related schwannomatosis.[1-3]

This condition results from a harmful change in the NF2 gene. The gene normally helps produce merlin, a protein involved in controlling cell growth. When merlin does not work normally, Schwann cells and certain other cells can form tumors.

NF2-related schwannomatosis follows an autosomal dominant inheritance pattern. This means a person with the condition may pass the altered gene to a child. However, many affected people are the first in their family to have the genetic change.

Some people have mosaic NF2-related schwannomatosis . Mosaicism means the gene change occurred after early development began, so it is present in only some of the body's cells. Blood genetic testing can therefore be negative even when the condition is present. Testing tumor tissue may sometimes help identify mosaicism.[2,3]

Age at diagnosis, the number and type of tumors, and family history can provide clues. Symptoms often begin in young adulthood, but the timing and severity vary. A person should not assume they have an inherited disorder based on hearing or balance symptoms alone.

Diagnosis

Evaluation usually begins with a medical and family history, an ear and nerve examination, and formal hearing tests. An audiologist may measure the softest sounds a person can hear and how well they understand spoken words. Balance testing may be useful when dizziness or unsteadiness is a major concern.

Magnetic resonance imaging, or MRI , is the main imaging test. Detailed MRI of the brain and internal auditory canals can show the nerves that connect the inner ears to the brain. A contrast material called gadolinium is often used to make small tumors easier to see. The care team considers kidney function, pregnancy, prior reactions, and other health factors when planning contrast imaging.

If bilateral tumors are found, evaluation may include MRI of the spine, an eye examination, a skin and neurologic examination, and assessment for other tumors linked to NF2-related schwannomatosis. The exact testing plan depends on age, symptoms, previous imaging, and

family history.[3,5]

Genetic counseling can help a person understand what testing can and cannot show. Testing may look for an NF2 gene change in blood or saliva. If those results are negative but suspicion remains, analysis of more than one tumor may help identify a mosaic form. A negative blood test does not always rule out the condition.

Diagnosis should be based on the full clinical picture. International criteria consider bilateral vestibular schwannomas, other characteristic tumors or eye findings, family history, and genetic results.[2]

Treatment and management

Treatment is individualized for each tumor and each ear. Important factors include tumor size and growth, remaining hearing, speech understanding, symptoms, age, other NF2-related tumors, and the person's priorities. Preserving overall function may be more important than treating both tumors in the same way.

Active monitoring may be appropriate for a small or stable tumor that is not causing major problems. This involves scheduled MRI scans, hearing tests, and clinical reviews. Monitoring is an active treatment strategy, not a lack of care. A change in hearing can matter even when imaging shows little growth.

Microsurgery removes all or part of a tumor through an operation. It may be considered for a large or growing tumor, significant brainstem pressure, or symptoms that cannot be managed in another way. The surgical approach depends on tumor size, anatomy, and whether useful hearing remains.

Surgery may control pressure and tumor growth, but it can cause hearing loss, balance problems, facial weakness, numbness, headache, or leakage of the fluid surrounding the brain. An experienced skull-base team can explain the expected benefits and risks for a particular situation.[4]

Stereotactic radiosurgery delivers focused radiation without a traditional open operation. Despite its name, it is a radiation treatment rather than surgery. It may control selected small or medium tumors, but its role requires special consideration in younger people and those with NF2-related schwannomatosis. Tumors can continue to affect hearing after treatment, and long-term follow-up remains necessary.[4,5]

Bevacizumab is a medicine that blocks signals involved in blood-vessel growth. Specialist teams may consider it for some people with NF2-related schwannomatosis who have rapidly growing vestibular schwannomas or worsening hearing. Some patients have tumor shrinkage

or improved hearing, while others do not. It is not approved specifically for this use in all countries and can have important adverse effects, so treatment requires expert monitoring.[3,5]

Hearing support is a central part of care. Options may include conventional hearing aids, devices that route sound from one side to the other, assistive listening technology, captioning, or communication training.

A cochlear implant may help selected people when the cochlear nerve still functions. If that nerve is absent or no longer works, an auditory brainstem implant may be considered. These devices do not restore normal hearing, and results vary. Assessment should occur at a center experienced with implantable hearing devices and NF2-related schwannomatosis.

Vestibular rehabilitation uses guided exercises to improve balance and help the brain adjust to changed inner-ear signals. Physical therapy, fall-prevention planning, psychological support, and workplace or school accommodations may also be useful.

Complications

The most common long-term concern is loss of useful hearing. Bilateral hearing loss can affect communication, education, employment, safety, and social connection. Early access to hearing rehabilitation and communication support can reduce these effects.

Tumor growth may compress nearby cranial nerves. This can cause facial numbness or weakness, swallowing difficulty, voice changes, or reduced coordination. Large tumors may press on the brainstem or block the normal flow of cerebrospinal fluid, leading to hydrocephalus, which is a buildup of fluid and pressure in the brain.

Treatment can also have complications. Surgery may affect hearing, balance, facial movement, or sensation. Radiation can cause temporary swelling and may not stop later hearing decline. Medicines such as bevacizumab require monitoring for treatment-specific adverse effects.

NF2-related schwannomatosis can cause additional tumors and nerve problems beyond the ears. Care therefore needs to address the whole nervous system rather than focusing only on the acoustic neuromas.

Follow up

Follow-up is usually long term. Visits may include MRI, hearing and speech-understanding tests, balance assessment, neurologic examination, and review of new symptoms. People with NF2-related schwannomatosis may also need periodic spinal imaging, eye examinations, and

monitoring for peripheral nerve problems.[3,5]

The timing of scans and tests is not the same for everyone. It depends on age, tumor growth, symptoms, treatment history, and the other findings present. After surgery, radiation, or medicine treatment, the schedule may change.

Contact the care team promptly for a noticeable change in hearing, new persistent facial symptoms, worsening balance, repeated falls, new weakness or numbness, or headaches that are becoming more frequent. These symptoms do not always mean a tumor has grown, but they deserve assessment.

Genetic counseling may also be offered to relatives. Whether a relative needs genetic testing or clinical screening depends on the affected person's test results and family history.

Sources

- National Institute on Deafness and Other Communication Disorders. "Vestibular Schwannoma (Acoustic Neuroma) and Neurofibromatosis." National Institutes of Health.
- Plotkin SR, Messiaen L, Legius E, et al. "Updated diagnostic criteria and nomenclature for neurofibromatosis type 2 and schwannomatosis: An international consensus recommendation." Genetics in Medicine . 2022;24(9):1967-1977.
- Evans DGR. "NF2-Related Schwannomatosis." GeneReviews . University of Washington, Seattle; National Center for Biotechnology Information Bookshelf.
- Goldbrunner R, Weller M, Regis J, et al. "EANO guideline on the diagnosis and treatment of vestibular schwannoma." Neuro-Oncology . 2020;22(1):31-45.
- Halliday D, Emmanouil B, Pretorius P, et al. "ERN GENTURIS clinical practice guidelines for the diagnosis, treatment, management and surveillance of people with NF2-related schwannomatosis." EClinicalMedicine . 2023;63:102107.