CONDITION

Neurofibromatosis 2

Neurofibromatosis 2 is a genetic condition that causes mostly noncancerous tumors on nerves and around the brain and spinal cord.

WHAT IT IS
Genetic condition causing tumors on nerves and brain coverings

BODY SYSTEM
Nervous system, hearing, eyes, and skin

MAIN SYMPTOMS
Hearing loss, tinnitus, balance problems, weakness

WHAT CAUSES IT
Inherited or new NF2 gene change

HOW IT IS DIAGNOSED
Clinical criteria, MRI scans, and NF2 genetic testing

MAIN TREATMENT
Monitoring, surgery, hearing support, and tumor-controlling medicine

CAN IT BE CURED?
No - gene change cannot be removed

IS IT AN EMERGENCY?
Sometimes - sudden weakness or severe new neurological symptoms

LONG-TERM OUTLOOK
Lifelong monitoring; many people remain active with specialist care

Overview

Neurofibromatosis 2 is a genetic condition that causes tumors to grow along nerves and around the brain and spinal cord. Its current medical name is NF2-related schwannomatosis , although many people and services still use "NF2" or "neurofibromatosis type 2."

Most NF2-related tumors are noncancerous. However, a noncancerous tumor can still cause serious problems if it presses on a nerve, the brain, or the spinal cord.

The most characteristic tumors are vestibular schwannomas . These grow on the nerves that carry hearing and balance signals from the inner ears to the brain. They are also called acoustic neuromas. In NF2, they commonly affect both sides.

NF2 may also cause other schwannomas, which arise from the cells covering nerves. Other

possible tumors include meningiomas around the brain or spinal cord and ependymomas within the spinal cord. Eye changes, including cataracts at a young age, can occur.

The condition is lifelong. Regular monitoring is important because tumors may grow without causing obvious symptoms at first. Care usually involves specialists in genetics, neurology, hearing, eye health, neurosurgery, and rehabilitation.

Symptoms

Symptoms differ according to which nerves or structures are affected. They may begin in childhood or adulthood. Some people first develop an eye problem, skin tumor, weakness, or spinal symptom rather than hearing loss.

Vestibular schwannomas may cause:

Tumors on other nerves may cause pain, tingling, numbness, or weakness in an arm or leg. A visible or tender lump may form under the skin.

Tumors affecting the brain or spinal cord can cause headaches, seizures, weakness, walking problems, or changes in bladder and bowel control. These symptoms can have many other causes and do not by themselves mean that someone has NF2.

Eye problems may include clouding of the lens, called a cataract, or changes involving the retina at the back of the eye. These can reduce vision. Some children with NF2 develop cataracts before other features become clear.

- Gradual hearing loss in one or both ears
- Tinnitus, meaning ringing or other sounds without an outside source
- Poor balance, dizziness, or unsteadiness
- A feeling of fullness in the ear
- Numbness or weakness of the face

Causes and risk factors

NF2 is caused by a harmful change in the NF2 gene . This gene normally helps control cell growth. When it does not work properly, cells covering certain nerves can grow into tumors.

The condition follows an autosomal dominant inheritance pattern. This means that a person with an NF2 gene change has a 1 in 2 chance of passing that change to each child. The outcome is separate for each pregnancy.

Some people inherit the gene change from a parent. In others, it begins as a new change during early development.

A new change may be present in only some of the body's cells. This is called mosaic NF2 . Mosaicism can lead to milder or more limited features, although this is not certain for an individual. It may also make the gene change harder to find in a blood sample.

NF2 is not caused by anything a person or parent did. It is separate from neurofibromatosis type 1, which involves a different gene and has different typical features.

Diagnosis

Diagnosis is based on the pattern of tumors, hearing and eye findings, family history, scans, and genetic testing. No single symptom confirms NF2.

MRI scans can show vestibular schwannomas and tumors around the brain or spinal cord. MRI uses a strong magnet rather than X-rays. Hearing assessment may include standard hearing tests and tests of how sound signals travel along the hearing nerve.

An eye examination can identify cataracts or retinal changes. A neurological examination checks strength, sensation, coordination, reflexes, and walking.

A blood or saliva test may find a harmful NF2 gene change. If mosaicism is suspected, testing a stored or removed tumor may provide additional information. A negative blood test does not always rule out NF2, particularly when only some cells carry the change.

Doctors use agreed clinical criteria to decide whether a combination of findings supports the diagnosis. Genetic counselling can help you understand test results, inheritance, and what the result may mean for relatives.

Treatment and management

There is no treatment that removes the underlying gene change. Management focuses on watching tumors, preserving hearing and nerve function, treating symptoms, and dealing with tumors that are growing or causing harm.

Monitoring may include regular:

The timing of these checks is tailored to age, symptoms, previous results, and known tumors.

Surgery may be considered when a tumor is growing, pressing on important structures, causing symptoms, or threatening function. For a vestibular schwannoma, the team weighs tumor control against possible effects on hearing, balance, and the facial nerve.

Radiotherapy uses focused radiation to control tumor growth. It is considered cautiously in NF2 because radiation may create additional long-term risks, including new tumors or malignant change. The decision requires an experienced specialist team.

Bevacizumab is a medicine that blocks a signal involved in blood-vessel growth. In selected people with a growing vestibular schwannoma and worsening hearing, it may shrink the tumor or stabilize or improve hearing. It does not cure NF2, and benefits may not last after treatment stops. Monitoring is needed for effects such as high blood pressure, protein in the urine, bleeding, and delayed wound healing.

Hearing support may include hearing aids, communication technology, cochlear implants, or auditory brainstem implants. The most suitable option depends on the hearing nerve and previous treatment. Lip-reading, sign language, captions, and workplace or school adjustments can also help.

Pain management, physiotherapy, occupational therapy, mobility support, and psychological care may be useful when tumors affect movement or everyday life.

- MRI scans of the brain and spinal cord
- Hearing and balance assessments
- Eye examinations
- Neurological and skin examinations
- Reviews of pain, movement, communication, and daily function

Complications

Hearing loss is one of the main long-term complications. It may be gradual, uneven between the ears, or occasionally sudden. Balance problems can increase the risk of falls.

Large tumors may press on the brainstem or interfere with the normal flow of fluid around the brain. This can cause worsening headaches, vomiting, confusion, or other neurological symptoms.

Spinal tumors may compress the spinal cord or nerve roots. Possible effects include pain, weakness, numbness, walking difficulty, and loss of bladder or bowel control.

Surgery can sometimes affect hearing, facial movement, swallowing, balance, or sensation, depending on the tumor's location. Specialist rehabilitation can help with some of these changes.

Most NF2-related tumors remain noncancerous. Malignant, or cancerous, transformation is

uncommon, but concern is greater after radiation treatment. This is one reason radiotherapy decisions are made carefully.

Living with repeated scans, uncertain tumor growth, hearing changes, or multiple procedures can also affect mental health. Anxiety, low mood, social isolation, and communication difficulties deserve the same attention as physical symptoms.

Prevention

NF2 cannot currently be prevented in someone who carries the gene change. Lifestyle choices do not stop NF2-related tumors from forming.

Monitoring does not prevent the condition, but it can find changes before they cause major loss of function. Keeping planned hearing tests, scans, eye examinations, and specialist appointments is therefore an important part of care.

If an NF2 gene change has been identified, genetic counselling can explain reproductive options. These may include testing during pregnancy or testing embryos created through in vitro fertilisation. These are personal choices, and availability differs by location.

Relatives may be offered genetic assessment when a familial gene change is known. Finding the change does not predict exactly which tumors or symptoms a person will develop.

Living with it and what to expect

NF2 usually changes over years rather than days. A tumor may remain stable for a long time, while another grows. Growth can also occur in periods, so one scan cannot predict the full course.

Some people have relatively limited symptoms, particularly with mosaic NF2. Others develop several tumors and need repeated treatment. Even people in the same family can have different experiences.

Hearing may stay useful for years, decline slowly, or worsen after tumor growth or treatment. Planning communication support early can make later changes less disruptive. Schools, universities, and employers can often provide captions, listening systems, written communication, or other adjustments.

Ongoing specialist care is usually needed throughout life. Follow-up may become more frequent when a tumor changes, then return to a routine schedule if it is stable.

Many people with NF2 study, work, have relationships, travel, and raise families. Some need

increasing hearing, mobility, or personal support. The condition can shorten life when tumors cause major neurological complications, but an individual outlook is shaped by tumor location, growth, available treatment, and response to care.

When to get medical help

Contact your NF2 team or usual healthcare service promptly if you notice a new or persistent change such as:

Sudden hearing loss needs urgent same-day medical assessment. Early treatment may help in some causes of sudden hearing loss, whether or not it is related to NF2.

Seek emergency help for a seizure, fainting with incomplete recovery, sudden severe weakness, new inability to walk, confusion, or a severe headache with repeated vomiting. New loss of bladder or bowel control together with leg weakness or numbness may indicate spinal cord compression and also needs emergency assessment.

After surgery, urgent advice is needed for fever, increasing wound redness or drainage, worsening headache, or a clear watery leak from the nose, ear, or wound.

- Worsening hearing, tinnitus, or balance
- New facial numbness or weakness
- New pain, tingling, or limb weakness
- A changing or painful lump
- Persistent headaches or changes in vision
- New trouble walking or repeated falls

Common questions

Is neurofibromatosis 2 cancer?

NF2 is not itself cancer. Most tumors linked to it are noncancerous, meaning they do not usually spread to distant organs. They can still be harmful by pressing on nerves, the brain, or the spinal cord.

Are NF1 and NF2 the same condition?

No. They are caused by changes in different genes and have different typical features. NF1 more often causes café-au-lait skin patches and neurofibromas. NF2 is more strongly linked with vestibular schwannomas, meningiomas, and hearing loss.

Will everyone with NF2 lose their hearing?

No single outcome applies to everyone. Hearing loss is common, but its timing and severity differ. Monitoring, tumor treatment, hearing devices, implants, and communication support may help preserve hearing or reduce its effect on daily life.

Can my children inherit NF2?

If you carry an NF2 gene change, each child has a 1 in 2 chance of inheriting it. Mosaic NF2 can make the chance harder to estimate, so genetic testing and counselling are important for understanding your situation.

Can I have NF2 if my genetic test is negative?

Yes. A blood or saliva test may miss mosaic NF2 because the gene change is not present in every cell. Doctors may use clinical findings, MRI results, family history, or genetic testing of tumor tissue to clarify the diagnosis.

Why is it now called NF2-related schwannomatosis?

The updated name reflects that schwannomas are a central feature and separates the condition more clearly from NF1. "Neurofibromatosis 2" and "NF2" are still widely recognized.

Sources

- Evans DG. NF2-Related Schwannomatosis. GeneReviews, University of Washington, Seattle; updated 2023.
- Plotkin SR, Messiaen L, Legius E, and colleagues. Updated diagnostic criteria and nomenclature for neurofibromatosis type 2 and schwannomatosis: an international consensus recommendation. Genetics in Medicine. 2022;24(9):1967-1977.
- Goldbrunner R, Weller M, Regis J, and colleagues. EANO guideline on the diagnosis and treatment of vestibular schwannoma. Neuro-Oncology. 2020;22(1):31-45.
- National Cancer Institute. Neurofibromatosis Type 1 and Neurofibromatosis Type 2 Treatment (PDQ), Health Professional Version.
- NHS. Neurofibromatosis type 2. Overview, symptoms, diagnosis, and management information.