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[Neurofibromatosis Type 2](#) [1]

What is neurofibromatosis type 2?

Neurofibromatosis type 2 (NF2) is a hereditary condition most commonly associated with bilateral vestibular schwannomas, also known as acoustic neuromas. These are benign (noncancerous) tumors that occur on the nerves for balance leading to the inner ear. Although these tumors are benign, they can cause hearing and balance problems.

People with NF2 also have an increased risk of other tumors of the nervous system. These tumors are generally not cancerous, but they may still lead to significant medical problems, especially if there are multiple tumors in or next to the brain. Other nervous system tumor types include:

- Schwannomas of other nerves
- [Meningiomas](#) [2]
- Gliomas, including [ependymomas](#) [3] of the brain or spinal cord

People with NF2 are at an increased risk of developing cataracts in the eyes and benign skin tumors. They may have café-au-lait spots, which are light brown pigmentation, the color of “coffee with milk”, like individuals with [Neurofibromatosis Type 1](#) [4], but people with NF2 usually have fewer café-au-lait spots than people with NF1.

Signs of NF2 usually develop in the late teenage years or early 20s. Multiple features have been

associated with NF2, but the overall cancer risk is low. The number of features present and the severity of symptoms can vary among people with NF2, even within the same family.

What causes NF2?

NF2 is a genetic condition. This means that the cancer risk and other features of NF2 can be passed from generation to generation in a family. The gene associated with NF2 is also called *NF2*. A mutation (alteration) in the *NF2* gene, which is a “tumor suppressor,” gives a person an increased risk of developing cancerous and benign tumors and other symptoms of NF2. Most people with NF2 have a mutation in the *NF2* gene. Research is ongoing to learn more about the causes of NF2.

How is NF2 inherited?

Normally, every cell has two copies of each gene: one inherited from the mother and one inherited from the father. NF2 follows an autosomal dominant inheritance pattern, in which a mutation happens in only one copy of the gene. This means that a parent with a gene mutation may pass along a copy of their normal gene or a copy of the gene with the mutation. Therefore, a child who has a parent with a mutation has a 50% chance of inheriting that mutation. A brother, sister, or parent of a person who has a mutation also has a 50% chance of having the same mutation.

Options exist for couples interested in having a child when they know that one of them carries a gene mutation that increases the risk for this hereditary cancer syndrome. Preimplantation genetic diagnosis (PGD) is a medical procedure done in conjunction with in-vitro fertilization (IVF). It allows people who carry a specific known genetic mutation to have children who do not carry the mutation. A woman’s eggs are removed and fertilized in a laboratory. When the embryos reach a certain size, one cell is removed and is tested for the hereditary condition in question. The parents can then choose to transfer embryos that do not have the mutation. PGD has been in use for over a decade, and more recently has been used for several hereditary cancer predisposition syndromes. However, this is a complex procedure with financial, physical, and emotional factors for couples to consider before starting. For more information, talk with an assisted reproduction specialist at a fertility clinic.

How common is NF2?

It is estimated that about one in 40,000 people has NF2. About 50% of people with NF2 do not have any family history of the condition. They have a *de novo* (new) mutation in the *NF2* gene.

How is NF2 diagnosed?

Many sets of diagnostic guidelines have been proposed for NF2, and future revisions are likely. The features are very specific, and it is important to have a careful evaluation by a doctor who has experience with NF2 to make the diagnosis. In general, someone is suspected of having NF2

if they have:

- Bilateral, meaning both sides, vestibular schwannomas, which are tumors that grow from the nerve that controls hearing and balance
- A parent, sibling, or child with NF2 and either of the following:
 - A single vestibular schwannoma diagnosed before age 30, or
 - Any two of the following: a meningioma, glioma, schwannoma, or cataract
- A single vestibular schwannoma diagnosed before age 30 and a meningioma, glioma, schwannoma, or cataract
- Multiple meningiomas and a unilateral, meaning affecting only one side, vestibular schwannoma diagnosed under age 30, a glioma, schwannoma, or cataract

Genetic testing for mutations in the *NF2* gene is available for people diagnosed with NF2.

How are tumors related to NF2 treated?

Although the primary method of treating tumors associated with NF2 is surgery, many people with this disease have tumors that grow slowly or not at all. Such patients can be closely monitored in an approach called watchful waiting, watch and wait, or active surveillance. In this approach, active treatment would begin if there are signs that the tumors could cause neurological problems or the pattern of growth threatens brain or spinal cord function.

Specifically regarding bilateral acoustic neuromas, a major goal of treatment is to preserve the patient's hearing for as long as possible, even though it is known that hearing will eventually be lost at some point. [Surgery](#) [5] is usually done either in patients who can no longer hear or in those with a small tumor where a meaningful attempt can be made at preserving the nerve that makes hearing possible.

In addition, focused [radiation therapy](#) [6] is sometimes used for vestibular schwannomas. Like surgery, it carries a risk that the treatment will hurt the patient's hearing in order to control the tumor. The other tumors seen in this disease are usually meningiomas and ependymomas and are typically removed only if they grow to an extent that they put enough pressure on the nearby brain or spinal cord to affect its functioning.

Reports of using bevacizumab (Avastin), a drug that interferes with blood vessel formation to stop tumor growth, for treating vestibular schwannomas associated with NF2, have been encouraging. Although only a few patients have been treated with this drug, the results include both measurable shrinkage of tumor and partial restoration of hearing loss in some, but not all, patients. This is an experimental treatment and needs more testing, but it holds promise that other drugs may be proven effective as well. Talk with your doctor about [clinical trials](#) [7], which are research studies, for new treatments.

What are the estimated cancer risks associated with NF2?

The lifetime risk of cancer in a person with NF2 is considered to be low.

What are the screening options for NF2?

Suggested screenings for people with NF2 or at risk for NF2 include:

- A yearly [magnetic resonance imaging](#) [8] (MRI), beginning in the teenage years
- Hearing evaluations, including audiometry and a brain stem auditory evoked response (BAER) test, which is a test that detects electrical activity in the cochlea and auditory pathways

Screening recommendations may change over time as new technologies are developed and more is learned about NF2. It is important to talk with your doctor about appropriate screening tests. In some areas, comprehensive NF2 clinics may be available to help with coordination of medical care.

Learn more about [what to expect when having common tests, procedures, and scans](#) [9].

Questions to ask the doctor

If you are concerned about your risk of cancerous and benign tumors, talk with your doctor. Consider asking the following questions of your doctor:

- What is my risk of developing these types of tumors?
- What can I do to reduce my risk of this?
- What are my options for screening?

If you are concerned about your family history and think you or your other family members may have NF2, consider asking the following questions:

- Does my family history increase my risk of developing cancer?
- Does it suggest the need for a cancer risk assessment?
- Will you refer me to a genetic counselor or other genetics specialist?
- Should I consider [genetic testing](#) [10]?

More Information

[The Genetics of Cancer](#) [11]

[Genetic Testing](#) [10]

[What to Expect When You Meet With a Genetic Counselor](#) [12]

[Collecting Your Family Cancer History](#) [13]

[Sharing Genetic Test Results with Your Family](#) [14]

Additional Resources

Neurofibromatosis, Inc

www.nfinc.org [15]

Children's Tumor Foundation

www.ctf.org [16]

National Institute of Neurological Disorders and Stroke

<http://www.ninds.nih.gov/disorders/neurofibromatosis/neurofibromatosis.htm> [17]

Acoustic Neuroma Association

www.anausa.org [18]

National Cancer Institute

www.cancer.gov [19]

To find a genetic counselor in your area, ask your doctor or visit these websites:

National Society of Genetic Counselors

www.nsgc.org [20]

National Cancer Institute: Cancer Genetics Services Directory

www.cancer.gov/cancertopics/genetics/directory [21]

Links

[1] <http://www.cancer.net/cancer-types/neurofibromatosis-type-2>

[2] <http://www.cancer.net/node/31264>

[3] <http://www.cancer.net/node/31311>

[4] <http://www.cancer.net/node/19450>

[5] <http://www.cancer.net/navigating-cancer-care/how-cancer-treated/surgery/what-expect-when-having-surgery>

[6] <http://www.cancer.net/node/24661>

[7] <http://www.cancer.net/node/24863>

[8] <http://www.cancer.net/node/24578>

[9] <http://www.cancer.net/node/24959>

[10] <http://www.cancer.net/node/24895>

[11] <http://www.cancer.net/node/24897>

[12] <http://www.cancer.net/node/24907>

[13] <http://www.cancer.net/node/30761>

[14] <http://www.cancer.net/node/24906>

[15] <http://www.nfinc.org/>

[16] <http://www.ctf.org/>

[17] <http://www.ninds.nih.gov/disorders/neurofibromatosis/neurofibromatosis.htm>

[18] <http://www.anausa.org/>

[19] <http://www.cancer.gov/>

[20] <http://www.nsgc.org/>

[21] <http://www.cancer.gov/cancertopics/genetics/directory>