

NEWLY DIAGNOSED WITH NF2:
A GUIDE TO THE BASICS

CHILDREN'S
TUMOR
FOUNDATION
ENDING NF
THROUGH RESEARCH

ctf.org
1-800-323-7938



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NEWLY DIAGNOSED? You Are Not Alone

At the Children's Tumor Foundation (CTF) we know that a diagnosis of neurofibromatosis type 2 (NF2) can be overwhelming. There's a lot to digest all at once, and everyone deals with difficult news in a different way. Some like to take in the information little by little, taking time to absorb each new fact or bit of advice. Others prefer to dive in immediately and get as much information as they can. Both approaches are perfectly normal.

People also have different emotional reactions to being diagnosed with neurofibromatosis. Some may become anxious, depressed, or feel a sense of shock from the unexpected news. Instead of trying to take control of the unpredictable, one of the most helpful strategies can be to try to accept the diagnosis. Once acceptance begins, it becomes much easier for negative feelings to lessen and to cope effectively.

What is most important is to understand that you are not alone. Approximately 10,000 Americans have NF2. The Children's Tumor Foundation has many resources for you, all over the United States, and we would like you to think of us as your safe haven. These resources include local CTF representatives you can speak to, and many events that you can attend to help you meet more members of the NF community around you.



Dealing with a diagnosis of a genetic disorder such as neurofibromatosis can be hard. There isn't a cure yet, and the course of NF2 can be difficult to foresee, especially if you are the first person in your family to develop it. When the disorder runs in families, it tends to be more predictable. Having NF2 does create challenges, and you may feel overwhelmed at times. The Children's Tumor Foundation can, however, help you take some steps to make things feel more manageable.

- **Get the facts** – Read the “NF Basics” page in this booklet, or visit the Foundation’s website at ctf.org to learn more about the three types of NF.
- **Find a doctor that knows about NF2** – We understand how difficult it is to find healthcare professionals who have experience diagnosing and caring for NF2 patients. We are constantly working to expand our clinic network and add to the list of NF2 professionals available on our website.
- **Connect** – The NF Forum is a national patient and family gathering that allows those living with NF and their families to connect, support, and learn from one another while attending seminars on relevant topics pertaining to neurofibromatosis. Find out more at ctf.org.
- **Sign up for research** – Join the NF Registry at nfregistry.org to learn about, and participate in, advanced scientific research for all forms of NF.
- **Get in touch** – Reach out to your regional contact on CTF’s staff, listed on the Foundation’s website at ctf.org.
- **Get involved** – National programs like the NF Walk, NF Endurance, NF Camp, and Racing4Research are great ways to empower yourself and your community in the fight for treatments and a cure for NF. It is also a way to meet others in your local NF community.
- **Stay in touch** – Visit the CTF website, “like” our Facebook page, and follow us on Twitter and Instagram for all the latest news and info. You can also join social media groups specifically for NF2 patients and families.
 - Web: ctf.org
 - Facebook: facebook.com/childrenstumor
 - Twitter: twitter.com/childrenstumor
 - Instagram: instagram.com/childrenstumor
 - YouTube: youtube.com/childrenstumor



The Children's Tumor Foundation

Founded in 1978, the Children's Tumor Foundation (CTF) began as the first grassroots organization solely dedicated to the goal of finding treatments for NF. Today, CTF is a highly recognized national nonprofit foundation, the leading force in the fight to end NF, and a model for other innovative research endeavors.

The Mission of the Children's Tumor Foundation:

Drive research, expand knowledge, and advance care for the NF community.

Our Vision: End NF.

Please feel free to call the Children's Tumor Foundation for information at 1-800-323-7938, or email info@ctf.org.

NF Basics

Neurofibromatosis (NF) is caused by a genetic change that makes people more likely to develop benign (non-cancerous) tumors around nerves. NF is a lifelong condition that affects all populations equally, regardless of gender or ethnicity. Neurofibromatosis has been classified into three distinct types: NF1, NF2, and schwannomatosis. Each type is linked to changes in a different gene. One type cannot turn into another type. While NF1 is caused by a change, or mutation, in a gene located on chromosome 17, both NF2 and schwannomatosis are caused by mutations in genes located on chromosome 22 (although other genes not yet identified may also be associated with schwannomatosis).

Neurofibromatosis type 1 (NF1), formerly known as von Recklinghausen NF or peripheral NF, is the most common of the three types of neurofibromatosis and is also one of the most common inherited neurological disorders, affecting about 1 in 3,000 people throughout the world. The disorder is characterized by multiple café au lait (light brown) skin spots and neurofibromas (small benign tumors) on or under the skin. Some people with NF1 develop large, potentially disfiguring neurofibromas. About 50% of people with NF1 also have learning challenges. Softening and curving of bones and curvature of the spine (scoliosis) occur in some patients with NF1. Occasionally, tumors may develop in the brain, or arise from cranial or spinal nerves. While NF tumors are not cancerous, they may cause health problems by pressing on nearby body tissues. Sometimes a benign tumor may become malignant (cancerous), but 85 to 90% of people with NF1 will never develop a malignant tumor related to neurofibromatosis. NF1 is usually diagnosed in childhood.

Neurofibromatosis type 2 (NF2) is less common than NF1, affecting about 1 in 25,000 people worldwide. The disorder is characterized by the development of benign tumors called vestibular schwannomas (previously called acoustic neuromas) on the nerve that carries sound and balance information from the inner ear to the brain (the eighth cranial nerve). These tumors often affect both ears, and may lead to partial or complete hearing loss. NF2 can also cause schwannomas to develop on other cranial or peripheral nerves. People with NF2 can also develop other tumors such as meningiomas (tumors of the membranes surrounding the brain and spinal cord) and ependymomas (tumors that develop from cells lining the ventricles of the brain and center of the spinal cord). The disorder can also cause the development of juvenile cataracts, compromising vision. Most people with NF2 develop symptoms in the late teen and early adult years, although about 10% of people develop symptoms during late childhood. The most common symptoms of NF2 include ringing in the ears (tinnitus), gradual hearing loss, and balance problems.

Schwannomatosis is a rare form of neurofibromatosis that has only recently been identified. The genetic disorder affects about 1 in 40,000 people and causes the development of benign tumors—called schwannomas—usually on spinal and peripheral nerves. These tumors develop when Schwann cells, which form the insulating cover around nerve fibers, grow abnormally. These tumors may cause pain that can be hard to manage. Rarely people with schwannomatosis may develop meningiomas or a single vestibular schwannoma. Schwannomatosis is usually diagnosed in adulthood, and is sometimes mistaken as NF2.

How Does Someone Get NF2?

Anyone who has any form of NF, including NF2, was born with it. Sometimes, a person can inherit NF2 from a parent; other cases may arise from a spontaneous mutation, and the person may be the first in the family to have NF2. It is important to know that NF2 does not result from anything a parent did or did not do during pregnancy, and it is not contagious in any way. Because one affected gene is enough to cause the disorder, NF2 can be passed from one generation to the next when only one parent has the gene.

NF2 (like NF1) is an autosomal dominant disorder.

Autosomal means that the gene in question is located on one of the numbered, or nonsex, chromosomes. The NF2 gene is located on the long arm of chromosome 22, at a position designated as 22q12.2.

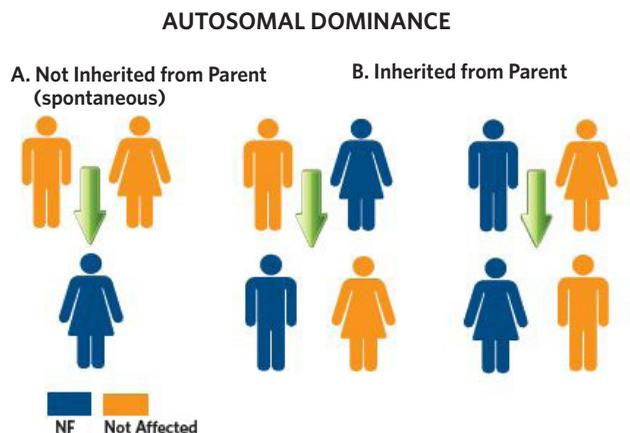
Dominant means that a single copy of the disease-associated mutation is enough to cause the disease.

The altered gene that causes NF2 affects men and women equally, and it can be passed down by either parent to a son or daughter. In just under half the cases, a person born with NF2 inherits it from an affected parent. In the other cases, for reasons that we do not understand, the change that causes NF2 occurs spontaneously. Spontaneous NF2 may affect all of the cells in a person's body, if the mutation occurred in the parent's sperm or egg.

If only some of the tissues and organs are affected, this is called *mosaic NF2*, and the mutation occurred in one of the embryo's cells while it developed. As many as one-third of people who have spontaneous NF2 have the mosaic version of the disorder. People with mosaic NF2 often, but not always, have a milder form of the disorder; for example, they may develop hearing problems only in one ear. They may also develop fewer tumors in other areas of the body.

If a couple includes one parent with NF2 and one without, each of their children has a 50/50 chance of receiving the *NF2*-affected gene. If a parent is proven to have mosaic NF2, the risk will be less than 50% and will depend on the proportion of egg or sperm cells affected by the genetic alteration.

If the change is spontaneous—which means that neither parent carries the *NF2*-affected gene—the couple is extremely unlikely to have other children with NF2.



NF2 in Depth

We talk about an *NF2* gene. It is an odd fact of genetic research that genes are often named for what they don't do properly. That is, the gene is often identified by investigating what happens when the gene is damaged or mutated. That's the case with the *NF2* gene: we all have two *NF2* genes, inheriting one from our father and one from our mother. In general, NF2 refers to the condition and *NF2* (in italics) refers to the gene.

The gene is a package of instructions for making a protein (the "building blocks," "gears," and "levers" of the body). The *NF2* gene carries the instructions for making a relatively large protein, called merlin (from **Moesin-Ezrin-Radixin-Like Protein**; it's also called schwannomin and occasionally neurofibromin 2, though this last term is not generally used by specialists in NF2 research). Merlin appears to go into action when cells come into contact with other cells and helps prevent them from continuing to divide and multiply.

When the *NF2* gene is changed or damaged, the merlin protein may not always do its job properly, and some cells, especially those surrounding certain nerves, may continue to divide and grow. This causes tumors; though these are benign (non-cancerous), some of them can compress or damage nearby nerves. So it is a damaged, mutated, or affected *NF2* gene that appears to be at the root of neurofibromatosis type 2.

As of 2014, researchers had found more than 300 different variations of the *NF2* gene that seem to be associated with neurofibromatosis type 2. Researchers are actively working to understand exactly how specific *NF2* alterations affect the way the condition develops. (That's one of the reasons why joining the NF Registry at nfregistry.org is so important.)

Origins

Unfortunately, thanks to generations of science fiction movies and books, *mutation* is a scary word. But it simply means a change in a gene's DNA sequence. We now believe that almost everyone's genes contain many mutations, best referred to as genetic variants. Most variants are neutral: they have no effect on a person's health or appearance. Some are beneficial: they may improve a person's ability to survive. Some, like the variants associated with NF2, can be harmful: they impair an essential biological process. And a surprising number are both beneficial and harmful: in some circumstances, they may help a person resist disease, for example, while at the same time making the carrier prone to other maladies.

Think of a gene as a very long string of letters (the *NF2* gene is about 95,000 characters long) and as being monotonous (there are only four letters used: A, C, G, and T). Overall,



“You will find a way to do what you want to do, and what you need to do.”

— **DERRICK HELTON,**

Wheelchair Rugby National Champion who lives with NF2, pictured here with Connor, who also lives with NF2.

the genetic code for a human being is about 3 billion characters long; embedded in it are the instructions for the roughly 23,000 proteins that are the basis of our bodies. (The actual number is still uncertain; it could be as low as 19,000 or as high as 25,000.) As the body develops, these three-billion-letter instructions are copied and recopied millions of times as cells divide and grow.

Imagine that someone gives you a sheet of paper listing one thousand As, Cs, Gs, and Ts in a seemingly random sequence, and tells you to copy the list by hand onto another sheet of paper. Almost everyone will make some errors—skipping some letters, possibly, or adding some, or mistaking a G for a C.

The body, too, makes errors as it copies genes. The cell does have molecular “proofreaders” to find and correct the errors, and they do a very good job...but not a perfect job. Sometimes, the error sneaks past them, becoming a source text for the next round of copying. That’s how most mutations occur. (While chemicals or radiation can damage DNA and cause mutations, copying and proofreading errors are much more common causes.) This is why it is important to recognize that genetic conditions like NF2 are not anybody’s fault. They’re a product of the way life works, and, indeed, a price all living creatures pay for the ability to change and adapt to new circumstances and challenges.

How Is the Diagnosis Made?

NF2 is diagnosed in individuals with **one of the following** collections of signs and symptoms (defined on pg. 11 in “What Are the Possible Symptoms of NF2?”). Accurate diagnosis of NF2 can only be made by a physician with expertise in the diagnosis and treatment of neurofibromatosis, and often relies on MRI images of the brain, spine, or body, and a detailed eye exam. Specific clinical and molecular diagnostic criteria have been established based on a consensus of experts.

The diagnostic criteria for NF2 include:

Bilateral vestibular schwannomas (affecting both ears)

OR a first degree relative with NF2, PLUS:

- Unilateral (affecting one ear) vestibular schwannoma OR
- Any two of the following:
 - meningioma
 - glioma (for example, spinal ependymoma)
 - neurofibroma
 - schwannoma
 - juvenile cataracts

OR unilateral vestibular schwannoma PLUS any two of the following:

- meningioma
- glioma
- neurofibroma
- schwannoma
- juvenile cataracts

OR multiple meningiomas (two or more) PLUS

- Unilateral vestibular schwannoma, OR
- Any two of the following:
 - glioma
 - neurofibroma
 - schwannoma
- juvenile cataracts



Genetic testing

NF2 is caused by a genetic change. This change can sometimes be detected by a specialized blood test. If you have family members with NF2, the genetic test is very accurate because doctors can identify the gene change in an affected parent and then determine if any offspring have inherited it. Occasionally, the genetic change cannot be identified in the parent, most often because they are mosaic for NF2 (defined on pg. 6 in “How Does Someone Get NF?”). Generally, if an individual inherits NF2, the severity of their disorder (age of onset and progression of symptoms) will be similar to that of the parent. However, if the parent has a mosaic form of NF2, the severity of symptoms in the child may be different from that in the parent.

If you are the first person in your family to develop NF2, genetic screening and predicting the course of the disorder becomes much harder. Right now, doctors can't look at the early symptoms of spontaneous NF2, or even a detailed genetic sequence, and predict precisely how the condition will, or will not, progress, although the type and position of the gene change can provide a reasonable guide to how mildly or severely someone may be affected. You may hear this referred to as the genotype/phenotype correlation; *genotype* refers to the genes responsible for a trait, and *phenotype* refers to the expression of the trait itself. Researchers are further studying genotype-phenotype relationships in NF2 in order to be able to better predict the severity of symptoms.

Genetic testing (“DNA testing”) for NF2 may be done in certain circumstances. In some instances, genetic testing can help to confirm the diagnosis if it is uncertain. For the most part, genetic testing does not very accurately predict the severity or specific complications of NF2. Results of genetic testing are mainly used for the testing of other family members or for deciding about reproductive options, such as starting a family. The decision to have genetic testing is a personal one. A genetics healthcare provider or a genetic counselor can help you with this decision. Some of the laboratories that perform genetic tests for NF2 can be found at ctf.org.

What Are the Possible Symptoms of NF2?

NF2's most common first symptoms (such as hearing and balance problems) can be traced to slow-growing, nonmalignant tumors on both nerves leading to the ears (also called the eighth cranial nerves). These nerves branch into the acoustic (cochlear) nerves (which carry information about sound to the brain) and vestibular nerves (which carry balance information from the inner ear to the brain).

The hallmark tumors seen in NF2 are vestibular schwannomas, formerly known as acoustic neuromas. Vestibular schwannomas are benign tumors made up of abnormal Schwann cells, which are the cells that give the nerves the lining and insulation needed to conduct information. Vestibular schwannomas can cause hearing loss in one or both ears, depending on whether the tumors are unilateral (on one side of the head) or bilateral (on both sides).

When vestibular schwannomas are the result of NF2, they tend to have a higher complication rate than vestibular schwannomas that are the result of some other disorder. This is one reason it is so important to be seen by a specialist familiar with NF2, and why is it important to ask for a second opinion before treatment.

Each nerve in the body is associated with a different function; therefore damage can affect senses, muscles, and even organs. Nerve damage for individuals with NF2 may result in some, but not all, of the following:

- Ringing in the ears (tinnitus)
- Hearing loss
- Problems with balance
- Facial weakness
- Brain and cranial nerve damage
- Swallowing difficulties
- Seizures
- Vision loss
- Loss of of balance and mobility, owing to
 - Spinal tumors
 - Peripheral neuropathy
 - Muscle wasting
 - Drop foot/foot drop
 - Pain

Meningiomas: These are encapsulated, or contained, benign tumors found on the lining of the brain and spinal cord. They will occur in about 80% of people with NF2 in their lifetime, but may not cause noticeable symptoms (such as headaches, seizures, blurred vision, weakness, or numbness) until they are large in size. Many meningiomas in people with NF2 will never require treatment.

Ependymomas: These are tumors that develop from cells lining the ventricles of the brain and center of the spinal cord. Ependymomas will occur inside the spine in about 20% of people with NF2.

Eye/vision issues: A type of juvenile cataract known as posterior subcapsular opacities is common in people with NF2, and may or may not cause vision problems associated with cataracts. In rare cases, people with NF2 may encounter eye problems due to damage to the cranial (head), optic (eye), trigeminal (face and eye), and facial (eyelid muscle) nerves.



“Focus on what you can, not what you can’t. For me, I can’t listen to a live band, you know, at a bar; I’m not able to really pick up anything that’s happening because of all the noise. So I could really get down about that. But I can hear my kids say, ‘I love you, Dad.’ And I’ll take that over a bar band any day. And the ABI gave me that.”

— **MATT HAY**, Children’s Tumor Foundation Board Member who lives with NF2, on his ABI (auditory brainstem implant) during his presentation entitled, “Coping with Hearing Loss” at the 2015 NF Forum

Medical Management of NF2

Because NF2 involves many different systems of the body, doctors and staff from many different specialties may be involved in caring for people with NF2. Current NF2 treatment works best when the providers work as a team. And, as one parent of a child with NF2 says, "The team needs a quarterback, someone to call the plays and hand off the ball on every play."

An NF clinic is the facility most likely to be that quarterback; it can offer the full range of necessary services. You can find a list of NF specialty clinics on the CTF website. Look particularly for facilities that specialize in NF2, which means that they see many patients with this disorder.

Many NF clinics offer coordinated care, so members of the specialist team are in regular communication with one another...and it may be possible for several team members to see you on the same visit.

If you do not have access to a specialty NF2 clinic, you may need to consult specialists one at a time. It is important to see providers who know about NF2. For example, seek out an otolaryngologist (ear, nose, and throat specialist) who knows what changes to look for in NF2 patients and the right methods to check for hearing loss and bilateral vestibular schwannomas. Your doctor may refer you to healthcare professionals who are experts in different NF2-related issues.



"Sure, NF2 is hard, we all know that. It's really hard. And the resultant disabilities throw up roadblocks everywhere you turn. But it also teaches you the stuff you are made of. You are smart; you are strong; you are resourceful. So, focus on the things you can do. Get out there and have fun. Don't put it off."

— **MICHIE O'DAY**, CTF Honorary Board Member and artist who lives with NF2



“Find the experts, the people who see lots of NF2 patients. If you look at the NF Clinic portion of the CTF website, you will see the number of NF2 patients seen at each center. I think that’s a great place to start.”

— **TRACY TULLOH GALLOWAY**, Children’s Tumor Foundation Board Member, speaking to NF2 patients at the NF Forum

In the course of treatment, a person with NF2 may see specialists in:

- **Neurology:** Neurologists are trained in the treatment and diagnosis of conditions affecting the brain and nerves.
- **Otolaryngology (ENT):** These are specialists trained to diagnose and treat disorders of the ear, nose, throat, and vestibular (inner-ear balance) systems.
- **Ophthalmology:** These specialists diagnose, monitor, and treat (through medicine or surgery) conditions affecting the eyes and vision.
- **Neuro-ophthalmology:** These specialists take care of visual problems that do not come from the eyes themselves but are related to the nervous system.
- **Oncology:** These doctors focus on diagnosing and treating benign and malignant tumors.
- **Pediatric Medicine:** Pediatricians are doctors who care for children and adolescents.
- **Genetics:** Medical geneticists are physicians with expertise in the diagnosing and managing of genetic disorders, including NF. Genetic counselors are healthcare professionals with training in medical genetics and personal counseling. Their job is to explain how genes affect a medical condition and outline the choices that can be made to deal with that condition.
- **Neurosurgeons:** These surgeons specialize in operating on the brain and spinal cord.

Empower Yourself as a Patient

Because NF2 is rare, patients and families often play a big role in procuring proper treatment. Here are some tips on the process from people with NF2 who have been there:

Empower yourself as a patient (or as a parent, if your child is affected). You may very well find yourself spending a lot of time looking for doctors and other medical resources. Remember that all medical centers do not always necessarily have experience in NF2. You may want to look beyond your local area for specialists. (If you do, remember that there are groups like Angel Flight that provide transportation to the help you need. For more information, see ctf.org.)

Look for knowledgeable doctors who attend conferences on NF2 and stay current with clinical trials and the latest surgical techniques. Ask doctors questions to understand how much experience they have had with NF2. You may want to start a notebook to keep track of all the information you will collect from doctor visits and treatments.

Don't be afraid to ask for second and third opinions; you may find that the best doctors encourage you to see others. Ask for a digital copy of the scan after every MRI procedure; this makes things easier when you see specialists or seek additional opinions. It is appropriate to remind your doctor that NF2 patients are missing a tumor suppressor gene, so to limit the amount of X-ray radiation received (this includes dental X-rays). Specialists may prefer to use MRI scans when possible, to reduce the number of X-rays and CAT scans.

If you can manage it, it may be helpful to go to a local symposium or a national meeting, such as the annual NF Forum organized by the Children's Tumor Foundation. It is a unique combination of a scientific/medical conference and a meeting of NF2 patients and families. It can be a useful place to keep up with the latest medical knowledge, build a support group, and make friends who understand your situation.

Sharing the News

Who to tell and what to tell them are some of the more difficult challenges of dealing with a new diagnosis of NF2. Although we all have our own strategies and personal preferences for what and how to tell others, here are a few suggestions that might help make the process a little bit easier.

SAMPLE MESSAGE TO FRIENDS AND FAMILY

Dear _____,

I want to share with you that _____ has been diagnosed with neurofibromatosis type 2.

Neurofibromatosis type 2, or NF2, is a disorder that causes benign (non-cancerous) tumors to grow on the lining and nerves of the brain and spine. _____ will likely require special medical care and lifelong monitoring.

The most obvious signs of NF2 may be hearing loss, facial weakness, or balance issues. NF2 is something that people are born with, although it may not be diagnosed until later in life. It is caused by a genetic change, occurs in about 1 in 25,000 births, and is not contagious. There is not yet a cure, but scientists and doctors are working toward understanding and treating NF2.

If you would like to learn more about NF2, the Children's Tumor Foundation has free, online information about the condition at www.ctf.org.

We appreciate all of your love and support.

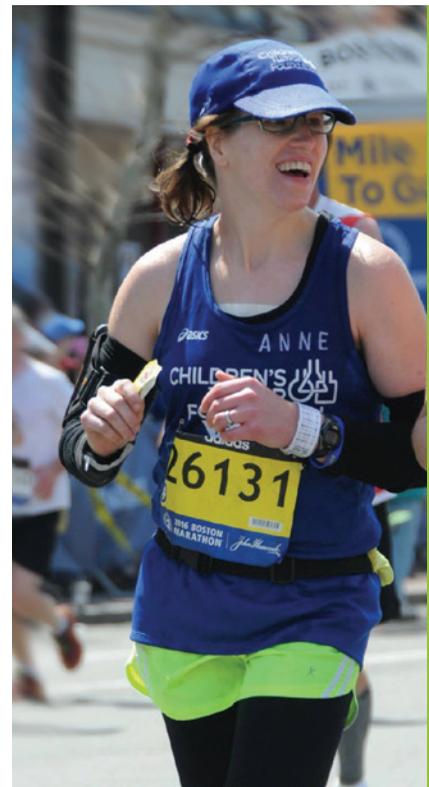
Sincerely,

Coping in the First Months After Diagnosis

Parents and patients might have difficulty coping during the first days, weeks, and months after diagnosis. Remember, living with NF2 is a marathon, not a sprint. The medical challenges of NF2 tend to develop over years, even decades, and you will have time to get information and adapt to the situation. You don't need to know everything or do anything immediately.

"Unless there's a real emergency," says the parent of a teen diagnosed with NF2, "take a deep breath and slow down."

An NF2 diagnosis can be a shock that might trigger a reaction that is out of character. Don't be surprised if you or those close to you have conflicting feelings, are more irritable than usual, or quieter than usual. Your partner or loved ones may react and cope differently than you do. It is part of a process. The process may follow a succession of stages, or it may be disorderly and unpredictable. You might be prepared to encounter reactions very much like the five stages of grief described in popular literature: denial ("This isn't really happening"), anger (at the world, family, fate, oneself, or even the patient), bargaining ("If I will just get better then I'll..." [fill in the blank]), depression ("What's the point?"), and acceptance ("It is what it is and I'll make the best of it"). Keep in mind that these emotions vary from one person to the next, and may not occur in exactly this order. It's possible, too, that they may occur again and again as new challenges arise.



Taking Care of Yourself

People with NF2, and loved ones who have been through the process, say that it can be difficult but that they got through it.

“Take care of yourself” is their common-sense advice. Talk to friends. Get exercise. Get enough sleep. Eat properly. Get involved with helping to treat the disease, but don’t make it all-consuming: make time to stay involved with family, friends, and activities you enjoy. Seek counseling from clergy or a therapist if you feel overwhelmed. Learn to be flexible and to adapt.

It may be helpful to reach out to others who have experience dealing with NF2 in their own families. “No matter what you are thinking and feeling,” says CTF Board Member Tracy Tulloh Galloway, “we have had those same thoughts and feelings and we can all completely and empathetically relate.” You can find other NF2 patients and families through friends, and friends of friends, through the Children’s Tumor Foundation, through online support groups, and through gatherings like the Foundation’s annual NF Forum for patient and family support. Check the Children’s Tumor Foundation website for details on these meetings, events, and resources.

As you reach out, especially on the Internet, remember that NF2 takes on many different forms and affects people in many different situations. At this stage, you may not yet have the background to evaluate the information that comes flooding in. If you cast a wide net, you will probably get a broad range of information and a lot of different advice, ranging from the very encouraging to the frightening. Much of it will be conflicting, and some will be flat-out wrong. In the early stages it may be helpful to take everything with a healthy grain of salt, as information that might—or might not—apply in your case.

And as we said earlier, this is a marathon, not a sprint. In most cases, you don’t have to be in a hurry to make up your mind.

Anne Noble, who lives with NF2, competes with the Children’s Tumor Foundation NF Endurance Team. Anne has completed dozens of races of every distance, from 5K to marathon to triathlon.



Advice for Parents: Taking Care of Children and Teens

Adults trying to help children with NF2 should be particularly aware of the child's stage of development, and make an effort to understand what the child is emotionally and intellectually equipped to handle. Understand that the child may be going through the same kind of process you are.

Encourage younger children to express their feelings and concerns. They may very well misunderstand what you tell them at first or make up explanations to fill in what they don't know. Listen for these misconceptions. Younger children need simple facts and short explanations.

Teens grow and change quickly, and it may be difficult to know just how much information they are ready to receive. It may be helpful to talk to a counselor for advice. You might also gain insight from other families in the NF Community who can help you understand what is typical behavior for teens, so that you can be more sensitive to variations in their thinking or behavior. Again, take time to listen, and look for opportunities to talk, but don't force it.



Advice for Teens and Young Adults: Taking Care of Yourself

The teenage years are tough enough on their own. You've already been going through many changes as you become an adult. If you've just learned you have NF2, it may seem overwhelming. You may wonder what the future holds, what your friends will think, and how this will affect dating.

Because NF2 is rare, you may find that the easiest way to talk to others with the disorder is to visit online communities offering information and support. The Children's Tumor Foundation is another place you can turn to for guidance. The Foundation offers resources for teens as well as popular programs, including the NF Summer Camp.

JT lives with NF2 and attended the CTF's annual NF Camp



Insurance Coverage for Neurofibromatosis

Billing and payment for healthcare services in the United States are based on procedure and diagnosis codes. These codes are used to determine medical necessity for health services, but the billing and coding processes are quite complex. It is not uncommon for healthcare providers to make unintentional errors that can result in services not being covered.

It is important to know the correct diagnosis and procedure codes for NF. Being proactive and working closely with providers and insurance companies can eliminate most of the hassle and frustration in this process.

Current ICD-10 codes for neurofibromatosis are as follows:

DIAGNOSIS	DIAGNOSIS CODE
NF, type 1	Q85.01
NF, type 2	Q85.02
Schwannomatosis	Q85.03
NF, unspecified	Q85.00

If an insurance company does deny a claim, check with your doctor's office to make sure the claim was fully documented. A lack of information may be the reason why the claim was denied. Make sure that the claim filed includes an NF diagnosis code, the reason for the procedure (e.g., pain, loss of function), and the correct procedural code.

If you do not have health insurance, contact your state's Medical Assistance Administration for additional information.

RESOURCES

WEBSITES

Children's Tumor Foundation's website (ctf.org) The website of the oldest and largest national foundation for NF in the United States. Here you can find information on NF, support groups, NF specialists, research news, educational events, family events, and many other ways to support the NF community.

NF Registry (nfrepository.org) A patient-centered resource for sharing your experiences to help guide NF research. You can find clinical trials that you may want to consider participating in, see how your NF experiences compare to those of others, and be part of an international NF awareness effort. There is strength in numbers, and the NF Registry needs your participation to make a difference.

BOOKS

Neurofibromatosis: A Handbook for Patients, Families, and Healthcare Professionals, 2nd edition. By Bruce R. Korf and Allan E. Rubenstein. New York: Thieme, 2005. Information about the natural history and genetics of NF, as well as management and research.



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