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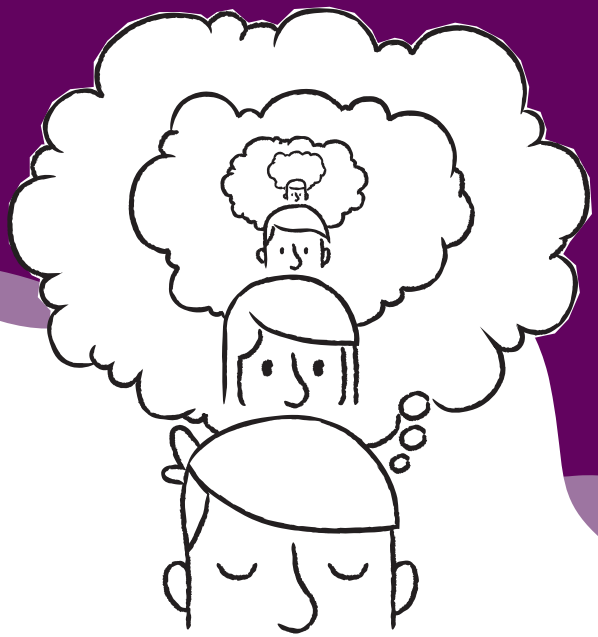
NERVE TUMOURS UK

"Know that it's OK to find NF hard: it is hard. Take a day at a time. Don't be too hard on yourself..."

Emily, NF2 patient



Neurofibromatosis Type 2 (NF2)

Some helpful information and resources

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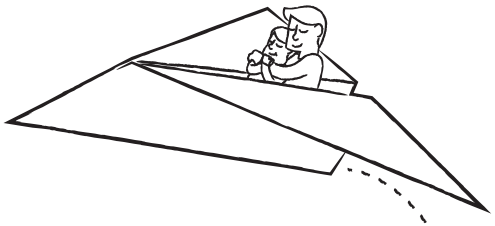


Sue, NF2 patient

Introduction

Neurofibromatosis Type 2 is indiscriminate; it affects all races, age and genders equally. It is a dominant genetic condition, which means that it can be passed on in families from one generation to the next through genetic inheritance. About half of NF2 cases recorded have no previous family history of the condition. This is called a "spontaneous gene mutation".

NF2 is an incredibly variable condition, and might vary from one person to another even within the same family. Some people may be affected very mildly and have very few health complications. Others may have many more complications, or more serious complications that can seriously impact daily life, and restrict what they can do.

With a condition that varies so much, it is important to learn some basic facts about NF2, and to understand when you need to seek help from your doctor.

This is a large part of why Nerve Tumours UK exists; to support people with nerve tumours, and to help you manage your condition and live your best life.

> **You are unique and everyone's NF2 story is so, so different.**
>
> Sue, NF2 patient

Emily's Story

Emily Owen is an author, speaker and workshop facilitator. Her inspiring and motivational story began when, as an energetic teenager, athlete and skilled musician, she was diagnosed as suffering from Neurofibromatosis Type 2 (NF2).

Since then Emily has survived many complicated surgical procedures, been close to death, and seen the gradual erosion of abilities that defined her personality, including the total loss of her hearing.

Despite this she has a remarkable sense of humour and has coped with everything she has been through due to her indomitable personality, strong Christian faith and her supportive family and friends. She's open and honest in telling her story to others and uses her experiences to motivate and inspire people of all ages, backgrounds, professions and cultures.

She says: "For me, there are things I can't do because of NF2. Listen to music, ride a bike... I find I need to accept these limitations (which is not the same as saying I like them), so that I am not fighting things I can't do every day. Do what I can, don't do what I can't. I try and find joy in every day. Little things as well as big. Chocolate, flowers, anything that makes me feel brighter."

> **"Know that it's OK to find NF2 hard: it is hard. Take a day at a time. Don't be too hard on yourself."**

Neurofibromatosis Type 2 (NF2) is a rare genetic condition caused by a 'misspelling' on chromosome 22. NF2 occurs in 1 in 30,000 of the population.

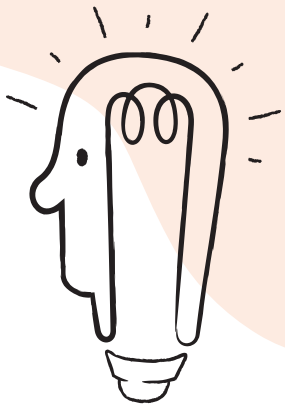
What is NF2?

Neurofibromatosis Type 2 (NF2) a rare genetic condition caused by a "misspelling" on chromosome 22. NF2 occurs in 1 in 30,000 of the population.

NF2 patients often need operations on their brain or spinal cord, due to the tumours that typically grow their as a result of the condition. The hallmark of NF2 is the development of benign tumours called vestibular schwannomas which grow on both hearing nerves. These tumours are can cause hearing loss, deafness, and mobility problems due to the pressure exerted on key nerves. Benign tumours may also develop on the nerve roots as they leave the spine and on the coverings of the brain. Some people with NF2 have a few benign tumours on the skin nerves but, in contrast to NF1, have few, if any, café au lait spots.

NF2 can be passed on from parent to child at the time of conception, a person who has NF2 has a 50% (or 1 in 2) chance of passing on the condition to his/her children.

How is it diagnosed?



NF2 is a genetic disorder that is caused by a misprint in a single gene on chromosome 22. The misprinted gene will be present at birth but signs of the condition do not usually appear until the teenage years, twenties or later.

The doctor or GP who first talks to you about Neurofibromatosis Type 2 may not be a specialist in the condition itself. They may suggest that you should have an appointment with other specialist doctors to confirm what is suspected. One of these specialist services is the genetics department. Genetics and genetic counselling is an NHS service based in regional hospitals.

Clinical genetics services help to make an accurate diagnosis. They can offer you information about NF2 and explain what the diagnosis means for other members of the family. The doctor (geneticist) can answer questions about how the condition has occurred, what are the possible problems that can arise, and how best to manage these. They can discuss the choices you can make if you are planning to have children and the risks of passing an inherited condition on.

How does it affect me?

Most NF2 tumours are slow growing and may cause minimal problems for years. Although they are not malignant (not cancerous) their position may produce significant symptoms.

For the majority of people, the most common first symptoms of NF2 are:

- **Gradual hearing loss**
- **Tinnitus (ringing or roaring in the ears)**
- **Unsteadiness, particularly when walking on uneven ground or in the dark.**

These symptoms are caused by tumours on the hearing nerves (vestibular schwannomas). Other symptoms may relate directly to the pressure caused by tumours on the spine or on the lining of the brain. For example:

- **Headaches**
- **Change in vision**
- **Change in sensation, pain or weakness of an arm or leg.**

What support can I get?

NF2 Service Centre

The NF2 service is coordinated through the NF2 clinics in Manchester, Cambridge, Guys and St Thomas' in London and Oxford.

Each designated centre is supported by experts in facial nerve reconstruction, audiologists and hearing therapists, physiotherapists and psychologists – all professionals who understand NF2. There is also a team of NF2 nurse specialists, who will coordinate patient care and act as the link between the centres and the local services. Some of the Nurses will be based in the other centres with existing NF2 clinics. They also perform NF2 surgeries funded through the service are vestibular schwannoma removal, brain stem and cochlear implants. The other key service is for radiation based treatments for NF2.

Treatments

It is very important to have a detailed discussion with a specialist who is familiar with NF2, to find out about the various options that may be available and to consider the benefits and risks of any operation or treatment. Treatments are based mainly on the symptoms the patient describes, alongside results of physical examinations, scans and hearing tests.

The treatment of vestibular Schwannomas will depend on:

- **the size of the tumours – whether they are pressing on the brain as well as on the hearing nerve**
- **how rapidly tumours are growing**
- **how much hearing loss the tumours have caused**

Some people choose to have X-ray treatment rather than an operation. The gamma knife or stereotactic surgery shrinks the vestibular Schwannomas. This treatment does have risks and should only be undertaken after discussion with doctors who are familiar with NF2 and its management.

Most people who become deaf through NF2 learn to lip read very well. Some people can now be offered an auditory brainstem implant (ABI) to help with hearing after surgery. The ABI does not give a return of hearing but gives an awareness of certain environmental sounds and is an aid to lipreading. This new treatment is only available in Specialist NF2 Centres.

Nerve Tumours UK Specialist Support

Nerve Tumours UK helps fund a team of Support Specialist in a number of regions across the United Kingdom. These specialists work to improve the lives of those affected by Neurofibromatosis and provide crucial support to patients and families.

National Helpline

Another service we work to maintain is out national helpline. **Open Mondays and Wednesdays 9am – 5pm**, if you need someone to talk to or some help getting to the right place. Call **07939 046 030** or email **helpline@nervetumours.org.uk**

Nerve Tumours UK Website

Head over to our website site to find out more information on Neurofibromatosis Type 2 (NF2) and where you can find more help.

nervetumours.org.uk

NF2

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Do you have nerve tumours? We're here to help.

nervetumours.org.uk
info@nervetumours.org.uk
020 8439 1234

/NerveTumoursUK **@NerveTumoursUK** **@NerveTumoursUK**

Our Nerve Tumours UK Specialists are available to offer support to anyone living with nerve tumours.

Call our helpline on 07939 046 030
Monday and Wednesday 9am-5pm

Nerve Tumours UK
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Never Tumours UK is the trading name of the Neurofibromatosis Association.
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