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NERVE TUMOURS UK

"Remember, even though NF2 is a rare condition there are so many people out there to help."

Sue, mother of Ben, both NF2 patients

NF2-related Schwannomatosis (NF2)

Some helpful information and resources for parents

Contents

"I have high hopes that one day there will be a cure. Maybe not in my life time but I am convinced there will be a cure for my son."

Sue, mother to Ben

Sue and Ben's Story

I did not know I had NF2 when I had my children but once diagnosed (at the age of 29) my geneticist told me about the 50/50 chance that my sons would inherit NF2 from me. When they were a bit older they were tested – my eldest, Ryan, tested negative, but Ben tested positive.

This was very hard to take, even harder than accepting that I had NF2 – at the time Ben was too young to be told but now he's a teenager and has yearly tests and meetings like me.

Ben has just started the drug Bevacizumib (Avastin), which is a drug that can stop tumour growth over time. Hopefully it can save Ben's hearing and from having future surgery. He has remarkably embraced the situation. He makes my husband, Ian, and me so proud of him. In fact, Ben is more concerned with the cannula being inserted than the actual infusion itself. But he does enjoy the free dinner he gets while he's there (gourmet burger and chips) so not all bad.

I have high hopes that one day there will be a cure. Maybe not in my life time but I am convinced there will be a cure for my son.

You are unique and everyone's NF2 story is so so different. Remember, even though this is a rare condition there are so many people out there to help.

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

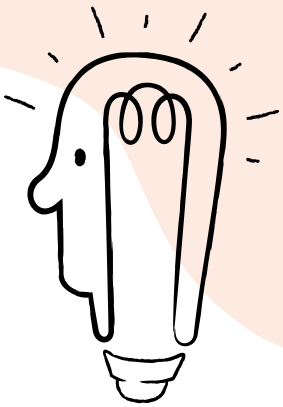
What is NF2?

NF2-related Schwannomatosis (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 there 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 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 NF2 may not be a specialist in the condition itself. They may suggest that you should have an appointment with other specialist doctors to confirm that your child has the condition.

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 your child and 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 more children and the risks of passing an inherited condition on.

How does it affect my child?

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.

> "You are unique and everyone's NF2 story is so, so different."
>
> Sue, mother of Ben

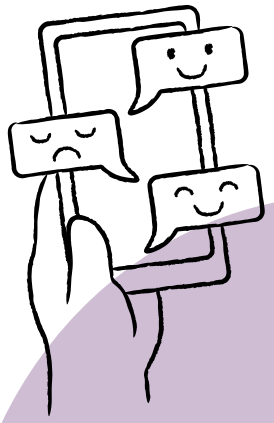
Ben, NF2 patient

Bullying

Some of the symptoms of NF2, such as tumours, deafness and a different way of speaking, can often make children targets of bullying.

A parent knows their child best and will know if something is wrong. Some behaviours to look out for if you suspect your child may be being bullied include:

- reluctance to go to school
- being mysteriously 'ill' each morning, or skipping school
- belongings getting 'lost' or damaged
- being nervous, losing confidence, or becoming distressed and withdrawn



Social Media & Cyber Bullying

Social media, and the internet have become an integral part of modern life, although these tools can be useful and fun, it can be difficult to protect your children online.

Signs of cyber bullying include:

- being withdrawn or upset after texting or being online
- being unwilling to talk about what they're doing online or on their phone
- spending much more or much less time texting or online
- many new phone numbers, texts or email addresses show up on their phone, laptop or tablet

How to help

Knowing or suspecting that your child is being bullied can be very upsetting, but there's lots you can do to help tackle the problem.

After talking to your child about the issues they are facing, it is important to notify your child's school. All schools are required by law to have an anti-bullying policy, and you should discuss the best plan of action to tackle the bullying with your child's teachers.

Why does my child have NF2?

NF2 may be passed on from parent to child at the time of conception, or it may start in a family with no previous history of the disorder. A person who has NF2 has a 50% (or 1 in 2) chance of passing on the condition to each of his/her children.

However, if your child is the first person in the family to be affected, the risk can be lower than that. When people are first diagnosed, they are referred to a Clinical Geneticist or Genetic Counsellor to discuss the genetic risk.

Sue, mother of Ben, both NF2 patients

What should I tell my child?

Once you think the time is right, when you are comfortable and calm, you can start the conversation. You know the words your child will understand. The first conversation is just a starting point, a bit like the first step on a ladder. Don't plan too much as your child may ask questions that take you by surprise and so throw you off track.

Allow your child to ask questions... these may be immediate or come some days later. Let your child take the lead in where the conversation goes. Don't try to cram too much into the first session or overload your child with too much information. Keep language simple with short sentences.

Examples of how you can start the conversation are as follows:

"Tomorrow we're going to see the doctor to make sure you're doing well. Do you know why we have these check-ups?"

"Next week we're going to have your hearing tested at the hospital. This is to check to see if your hearing nerve is working properly."

Encourage your child to ask questions. Keep your comments open so that you can reassure your child to express what they are thinking to help you share their feelings. It helps if you ask open questions that don't just lead to a yes or no reply. For example you might say: "Tell me about..." or "What do you think about...."

If your child does ask you a question, make sure you answer the question they ask and don't go off at a tangent. If you don't know the answer, then say so but offer to try to find out.

Finally, it is important to reassure your child that there will always be people who love them and will care for them. That having nerve tumours does not change the person that they are. NF2 is just a part of who they are.

Here are a few other things you can discuss about NF2-related Schwannomatosis (NF2).

Hearing Loss

Hearing loss in NF2 can be gradual or sudden and each brings its own difficulties. Adjustment to this loss is never easy and takes time. Friends and family are there to help with this big change in your and their lives. Professionals can supply the information and support to make the change more manageable. Learning to lipread is important. For some, learning sign language opens up new opportunities.

Facial nerve damage

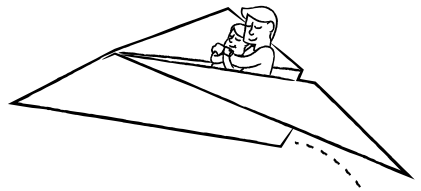
Help to remember that even the most beautiful models usually have some part of their body they don't like – we're all the same under the skin! They shouldn't try to cover the facial nerve damage with their hand or hair as this only distances them further from people and makes it harder to communicate.

Balance difficulties

Explain about the balance difficulty so that they feel safe and confident. They can take a friend along for support. They may prefer to use a walking stick which also alerts others to their needs without them needing to spell it out. If possible, it is always good to check out new venues before visiting e.g. steps, parking, lighting, noise levels.

Tiredness

Recognise and accept their limitations. Plan ahead for important events. Explain to others so that they understand the issue. Make sure they get enough rest.



What support can we get?

NF2 Service Centre

The NF2 service is coordinated through the NF2 clinics in Manchester, Cambridge, Guy's 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: 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 Specialists in a number of regions across the United Kingdom. These Specialists work to improve the lives of those affected by NF, the medical group name for Neurofibromatosis Type 1 (NF1), NF2-related-Schwannomatosis (NF2) and Schwannomatosis (SWN), and provide crucial support to patients and families.

National Helpline

The Nerve Tumours UK national helpline offers advice and support. Open **Monday, Wednesday** and **Friday 9am – 5pm**. Call **07939 046 030/ Freephone 0300 102 17 22** or email **helpline@nervetumours.org.uk**

Nerve Tumours UK Website

Visit **nervetumours.org.uk** for in-depth information about NF2-related Schwannomatosis (NF2) and the charity's support services.

NF2

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

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

Our Nerve Tumours UK Specialists are available to offer support to anyone living with NF, the medical group name for NF1, NF2 and SWN, all of which cause tumours to grow on nerve endings.
Call our Helpline on 07939 046 030/ Freephone 0300 102 17 22 or email helpline@nervetumours.org.uk
Monday, Wednesday and Friday 9am-5pm

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