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[patient.information@addenbrookes.nhs.uk](mailto:patient.information@addenbrookes.nhs.uk)



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## Clinic 10

### Patient Information

# Neurofibromatosis Type 2 (NF2)

## Document history

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## Overview of NF2

This leaflet is for patients and relatives who attend the neurofibromatosis type 2 (NF2) clinic in Cambridge. NF2 is a disease caused by a misprint of one of our genes and is present from birth, but signs of the disease do not usually appear until several years later. In some cases, you may be the first person in your family to be diagnosed with NF2, or in other cases you may be aware of other people within your family who also have the condition. NF2 is characterised by benign tumours that occur on nerves within the body, in particular on both balance nerves (known as vestibular schwannomas or acoustic neuromas). This can cause deafness and balance issues over time. Other benign tumours may also grow elsewhere in the body, such as on the lining of the brain, the spine, and under the skin. Some people with NF2 also have cataracts or problems with vision from a young age. We are able to provide you with detailed written information about the NF2 service if you would like to know more.

## The NF2 service

Cambridge University Hospitals NHS Foundation Trust has been designated as one of the four specialist centres for the management of NF2 by the National Specialised Commissioning Team (NSCT), the part of the NHS that deals with organizing care for people with rare diseases. This means that people who have been diagnosed with NF2 have access to a team of people with the specialist knowledge and experience necessary to provide the best care.

We hold the NF2 team meeting and outpatient clinic on the second Monday of each month.

## Location

The clinic is situated in Clinic 10, on level 2 of Outpatients.

If you have any special requirements, we will endeavour to accommodate you.

## Privacy and dignity

We are committed to treating all patients with privacy and dignity in a safe, clean and comfortable environment. This means, with a few exceptions, we will care for you in same sex bays in wards with separate sanitary facilities for men and women.

In some areas, due to the nature of the equipment or specialist care involved, we may not be able to care for you in same sex bays. In these cases staff will always do their best to respect your privacy and dignity, eg with the use of curtains or, where possible, moving you next to a patient of the same sex. If you have any concerns, please speak to the ward sister or charge nurse.



**Hearing LINK** is a voluntary organisation that provides support and information to people with a hearing loss and their families. Of particular interest is the week-long intensive rehabilitation course

run for NF2 patients. If you are interested in this course, please inform the NF2 Nurse Practitioner so that she can refer your details to the relevant people.

<http://www.hearinglink.org/>

### **Contact details (England)**

**Telephone** 0300 111 1113

**SMS** 07526 123255

**Fax** 01323 471260

**Email** [enquiries@hearinglink.org](mailto:enquiries@hearinglink.org)

### **Action on Hearing Loss**

Action on Hearing Loss (formerly the RNID) aim to make day to day life better for people who are deaf or who are hard of hearing. Their website includes lots of useful information on communication equipment, your rights and help with benefits and services.

<http://www.actiononhearingloss.org.uk/>

**Information line telephone** 0808 808 0123

**Information line textphone** 0808 808 9000

**Email** [information@hearingloss.org.uk](mailto:information@hearingloss.org.uk)

### **National Association for Deafened People**

The NADP provide advice to those with recent/severe hearing loss, and their family and friends.

<http://www.nadp.org.uk>

Parking is available in the multi-storey car park, with a fixed price for attending outpatients appointments. These concessionary tickets are available from the customer service desk in the main Hospital car park (located at the car park exit). Proof of appointment/s will be required.

### **Meet the team**

We hold a monthly NF2 multi-disciplinary team meeting. This enables all of the team to work together to discuss the best treatment plan before seeing you in clinic later the same day. The multi-disciplinary team involved in the NF2 service are:

- Mr Patrick Axon MD FRCS (ORL): Consultant Skull Base and Hearing Implant Surgeon.
- Mr Neil Donnelly MSc (Hons) FRCS (ORL-NS): Consultant Skull Base and Hearing Implant Surgeon
- Mr Richard Mannion PhD FRCS (SN): Consultant Neurosurgeon.
- Mr Robert Macfarlane MD FRCS: Consultant Neurosurgeon
- Dr Lucy Raymond MA DPhil FRCP: Consultant in Clinical Genetics.
- Dr Amy Taylor PhD MSc: Genetic Counsellor.
- Dr Anke E Hensiek Dr Med PhD MRCP: Consultant Neurologist.
- Juliette Durie-Gair RN BA (Hons): Skull base & NF2 Nurse Practitioner
- Sally Taylor RN: NF2 Nurse Practitioner.
- Richard Knight PhD MSc: Clinical Scientist (Audiology).
- Tamara Lamb and Susan Hobson MSc: Senior Audiologist.
- Zebunnisa Vanat BSc MSc CS: Clinical Lead for the Adult Hearing Implants Programme, Audiological Scientist.

We are also able to arrange appointments with the Plastic Surgery Department and the eye team on the same day as your NF2 appointment, if we know they are needed in advance. It is your choice whether you undergo any treatment offered by the NF2 team or not. We see all NF2 patients in the Outpatient Department at least annually.

## MRI scanning

As part of the NF2 service we undertake an annual MRI scan of your head and, a MRI scan of your spine every three years to monitor for any tumour growth. Unfortunately we cannot always offer the MRI appointment at the same time as your outpatient appointment. This is because it is better for us to have the results of your scans when we discuss you in our team meeting before we meet you. If you have a cochlear implant or an auditory brainstem implant, we will inform the MRI Department that your head will need a bandage and a piece of plastic over the magnet to secure it in place. Local anaesthetic can be given if necessary.

## Genetic counselling

Our team can explain how NF2 is inherited and discuss whether you would like to have genetic testing, which is done by taking a blood sample. Depending on your results, we may also be able to offer genetic testing to your relatives. Alternatively, we may offer them a hearing test or MRI scan of the head to look for any signs of NF2.

We can also talk to you about planning a family, and the options that would be available if you would like to avoid passing on NF2 to future generations. There is no obligation to have genetic tests; decisions about this will always be left up to you.

## Nurse practitioner

As part of the NF2 service in Cambridge, there are two full-time nurse practitioners. They are able to help with time to talk about NF2 and what it means. They offer support and information to those newly diagnosed and in times of acute need. They are also involved in co-ordinating the clinic and appointments. The nurse practitioners are the first point of contact for patients with NF2 should you have any queries about your care or the service. If you would like to see the nurses separately from the medical team after your outpatient appointment, please let one of the clinic nurses know and we will arrange this.



**NF2UK:** this patient support group has been set up by people with NF2 and the following has been written by them to explain how the group works:

*"NF2UK was founded in 2008 and was intended to provide a means for patients to contact each other and share information and experiences*

*etc. We use the same system as the Nf2 Crew, which is email based (not a forum) and people have to join the group before they can access the members, files and messages from the group.*

*We ask that all group emails are to be regarded confidential, which we hope provides the opportunity for an open discussion about anything to do with our community or care. We support each other through our various trials and treatments or anything to with Nf2.*

*We are one of the many resources now available to patients and the growth of social networking sites like MSN Messenger and Facebook has made it much easier for people to find each other and keep in touch. We hope to have our own Facebook site in the future."*

The groups contact details are:

<http://health.groups.yahoo.com/group/Nf2UK/>

Subscribe: [Nf2UK-subscribe@yahoogroups.com](mailto:Nf2UK-subscribe@yahoogroups.com)

List owner: [Nf2UK-owner@yahoogroups.com](mailto:Nf2UK-owner@yahoogroups.com)

## Support from other organisations



The main charity that supports NF2 in the UK is called the **Neuro Foundation**. <http://www.nfauk.org/>

They represent patients with both neurofibromatosis Type 1 (NF1) and NF2. They aim to provide information to people with NF2, provide practical and emotional advice, improve awareness and campaign for change in the care of people with NF, to connect people with NF2 and those involved in their care, so that they receive the best support and seek to raise funds to provide support to those with NF e.g. travel grants, research development and social support.

The NFAUK also has a network of specialist advisors, who work alongside the specialist nurses, based within NHS Trusts around the UK. They provide emotional support and information to parents, families and individuals on the many aspects of NF.

Neuro Association helpline: 0845 602 4173

To see if there is an advisor in your area, please call 0208 439 1234, or ask the nurse practitioner who will be able to advise you.

We can occasionally offer a home visit if you are unable to attend the clinic for any reason, so please discuss this with the nurse practitioners if you feel this would be useful for you.

## Hearing tests

We may offer you hearing tests called a PTA (pure tone audiogram) and SDS (speech discrimination score) on your arrival in clinic 10 prior to seeing you in your outpatient appointment. This enables us to see how your hearing is being affected by NF2, and how you are able to communicate with other people. If appropriate, we may offer you further assessments with our audiologists, or have a discussion with you about hearing implants.

## Hearing aids

Audiology Reception desk: Tel: 01223 217 797

Audiology Fax: 01223 586 912

Audiology Minicom: 01223 274 494

If you are identified with a hearing loss in the NF2 clinic we will discuss your suitability for NHS digital hearing aids and assistive listening devices. We will see you for a full assessment to discuss your individual hearing needs. NHS hearing aids are usually fitted behind the ear, and we will show you what is appropriate for your particular loss. You may be fitted here at Addenbrookes, or at your nearest Audiology Department if you prefer.

## Unilateral hearing loss clinic

A hearing loss which affects primarily or solely one ear tends to bring a particular set of hearing difficulties, typically with difficulty hearing sound from one side but also increased difficulty hearing amidst background noise and impaired ability to tell where sound is coming from. The Audiology Department at Addenbrooke's Hospital has a dedicated clinic for this particular pattern of hearing loss.

## **Tinnitus**

For patients with NF2 who are troubled by tinnitus (an umbrella term for perceiving any sounds which are not genuine external sounds), there are various approaches which may be employed with the aim of reducing the impact that the tinnitus may have. We can arrange appointments, often on the same day as your visit to the NF2 clinic, to explore this.

## **Vestibular assessments**

Some people with NF2 find that they have problems with their balance due to how the disease can affect your hearing, balance function and your eyesight. In order to investigate your balance further, we can arrange for a vestibular assessment in which we conduct a range of tests which thoroughly assess different aspects of your balance.

## **Vestibular rehabilitation**

Any changes to balance function such as those caused by NF2 can result in sensations of imbalance or dizziness on movement. The aim of a vestibular rehabilitation appointment is to design some exercises which retrain the balance system to make the best use of remaining balance function. Strategies for dealing with difficult environments for balance can also be discussed.

## **Surgery – vestibular schwannoma**

Occasionally, it is necessary to remove a growing tumour to prevent serious complications or to maintain the nerve function. The decision to operate is taken very seriously and is made after a lengthy team discussion and only with the agreement of the patient and their family.

The Cambridge NF2 team has a great deal of experience looking after patients with skull base and spinal tumours. It is one of the largest skull base units in Europe and is happy to share its outcome data with you.

## **Quality of life questionnaires**

In order to see how NF2 is affecting you, and to see which services we offer that could help with your symptoms of NF2, we give out questionnaires to every patient we see in clinic. This asks about your hearing and balance, and other common symptoms of NF2. As part of the NHS Specialised Services contract, we have to complete this for every patient.

## **Patient representatives (Patient and Public Involvement/PPI)**

If you have views about how we provide our NF2 service, the information that we give to people with NF2, or the facilities and environment of the hospital then we would welcome your views in person, via post, fax, phone or email. You do not have to be a patient to be a representative for the NF2 service. We are keen to hear what you have to say about the service and where possible wish to involve you in service development so that we can provide the best patient-focused care possible. Please inform the nurse practitioner if you are interested in this role.

## **Transport and help with finances**

You can get help with travel costs for NHS treatment if you receive Income Support, Income-Based Jobseeker's Allowance, Income Related Employment and Support Allowance, Pension Credit Guarantee Credit, or if you are named on an NHS Tax Credit Exemption Certificate. You may be able to get help if you are on a low income by completing an HC1 form (available from your GP, Jobcentres or the transport reception in outpatients). If you would like further information, please ask the nurse practitioner.

## Speech and language therapy

Speech and language therapists assess, manage and support people with communication and swallowing difficulties experienced by people with NF2. These are generally associated with lower cranial nerve damage. At Addenbrooke's we have speech and language therapists that specialise in both of these areas. We can give information to you and your family and provide therapy and support to help with your communication and/or swallowing as needed.

## Contacts/further information

- Clinic 10 Main Reception: 01223 217588
- Secretary to Mr Axon: Hilary Noculak 01223 217471
- Secretary to Mr Donnelly: Bianca Saunders 01223 586638
- NF2 Nurse Practitioner: telephone (01223) 348672  
e-mail: [juliette.durie-gair@addenbrookes.nhs.uk](mailto:juliette.durie-gair@addenbrookes.nhs.uk)  
[sally.taylor@addenbrookes.nhs.uk](mailto:sally.taylor@addenbrookes.nhs.uk)

Please note that we can only discuss your plan of care with you via e-mail if you have signed a consent form to allow us to do so. If you would like to do this, please ask and we can provide you with this form.

## Special requirements

If you need help in communicating with us, such as an interpreter, a sign language interpreter, or specialist equipment, we can arrange this prior to you being seen at your outpatient appointment. If you find Speech-to-Text software useful, we have this available in the NF2 clinic. Please contact us if you feel this is necessary. If you need assistance in your application for disability benefit, or blue badge application, then please contact the nurse practitioner.

## Surgery - cochlear implantation

NF2 often results in progressive bilateral hearing loss. Hearing loss might occur despite no growth in the tumours causing the hearing loss. If this is the case, it might be possible to insert a cochlear implant on the side of the severe deafness. A cochlear implant is an electronic device that is inserted under the skin behind the ear and has an electrode array that is placed inside the cochlea (the inner ear). This procedure has been shown to improve hearing despite a vestibular schwannoma being present on that side.

Occasionally it might be possible to remove a vestibular schwannoma and leave the hearing nerve intact. Although there is no normal hearing on that side because the cochlea is not functioning, a cochlear implant might be useful option for returning some hearing. The decision is usually made after careful testing to make sure the hearing nerve is still functioning.

More information about cochlear implantation can be supplied if required.

## Surgery - auditory brainstem implantation (ABI)

Occasionally patients require the removal of a growing vestibular schwannoma. If that is the case then the hearing nerve is usually divided in order to remove the tumour completely. The only way of restoring some hearing on that side is to insert an ABI. This is an implant that has an electrode array that sits on the cochlear nucleus which is in the brain stem.

The NF2 team might suggest that an ABI is inserted even though you have reasonable or good hearing in the other ear. The reason for this is that the best chance of success is often at the first operation and it is also possible that hearing in the other ear might deteriorate over time.

More information can be supplied by the hearing implant team if an ABI is being considered

## **Management of spinal lesions**

Looking after the spine is a key component to the management of those with NF2 and all our patients undergo surveillance of the spine through clinical assessment and intermittent MRI scanning to monitor the situation over time. Those with spinal problems are assessed in the multidisciplinary clinic by a neurosurgeon and a neurologist with a special interest in NF2 and complex spinal disease. In most cases, spinal tumours do not require surgery but occasionally surgery is required and this is done here at Addenbrooke's.

## **Facial palsy/reanimation**

The facial reconstruction service provides a number of options for the correction of the facial weakness that can sometimes occur with NF2 or following surgery. Depending on how you are affected, you may be offered a number of treatments to help protect your eye, and to reposition the face if it falls. In some cases these treatments will allow you to actively move your face, in others they will simply reposition the face. In either event, we expect to be able to help you with your facial appearance and function.

## **Neurological deficits**

Dr Hensiek is a consultant neurologist with a special interest in inherited neurological diseases. In the NF2 clinic, she conducts neurological assessments in order to formally determine disability and current neurological problems and symptoms. She manages certain symptoms that can occur in NF2, for example pain, spasticity, or epilepsy. The neurologist will involve other members of the neurological team (speech and language therapists, physiotherapists, epilepsy specialist nurse, and the botox clinic) if indicated. Furthermore, her assessments are useful for guiding decisions on treatment options, and for more formally determining individual patients' disease progression.

## **Raised ICP (Intracranial Pressure) and pulsatile tinnitus**

Patients can occasionally experience headaches and a pulsating noise in the ear that is caused by a rise in pressure inside the head. This is often irritating, but not life threatening. We have a team dedicated to the management of these conditions.

## **Bevacizumab (Avastin)**

In 2009, some research was published in the USA about how bevacizumab had been shown to help some people with NF2, by reducing the size of vestibular schwannomas and in some cases improving hearing. The study was only undertaken on a very few patients so further research is needed. However, bevacizumab has currently been funded for treatment by the National Specialised Commissioning Team (NSCT) provided that strict criteria are met. We can discuss this with you in your outpatient appointment if you would like more details.

## **Radiotherapy/radiosurgery**

As part of the NSCT organisation of NF2 care, NF2 patients who may be suitable for radiotherapy to their tumours should be seen in Sheffield, as this is the approved centre for people with NF2. We can arrange for referral to Sheffield for consultation if necessary.

## **Hearing rehabilitation**

Frances Harris, our hearing rehabilitationist, is able to offer advice to hearing implant users on equipment, adjustment to using hearing implants, communication strategies, and support for deafness.