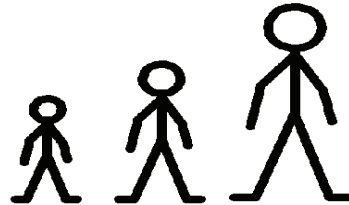


Personal Health Record



Neurofibromatosis Type 2

Contents

Neurofibromatosis Type 2 (NF2) Personal Health Record	3
Personal Details and Useful Contacts	4
Specialist Doctor Details	5-6
Information for You and Your Family	7-8
Information for Health Care Professionals	9-10
Functional Summary	11
Hearing and Vestibular Schwannoma Record	12
Other NF2 Issues	13-14
Scan History	15
Review Appointments	16
Details of Other Hospital Visits and Appointments	17
Sources of Information	19
My Notes	20

Neurofibromatosis Type 2 Personal Health Record

You have been given this booklet because you have a condition called **Neurofibromatosis Type 2 (NF2)**. People with NF2, may see a number of doctors and specialists, and this booklet will help you and your health care team ensure you receive the correct care and appropriate check ups. Please bring it with you whenever you visit your NF2 clinic or any of the following and ask them to complete the relevant pages:

- Your GP *(ask them to complete page 17)*
- Hospital Outpatients Clinic or Emergency Departments *(ask them to complete pages 5 and 17)*
- Your Dentist *(ask them to complete page 17)*
- Your Optician *(ask them to complete page 17)*
- Any other health care appointment *(ask them to complete page 17)*

You may also wish to use the plastic pockets in the front and back of this folder to store appointment letters, letters from your GP and hospital doctors, or anything else related to your NF2 that you want to keep safe.

Spare pages for this booklet are available from the Regional Genetics Service.

Important Note: This record does not replace your hospital, GP or other medical records which will continue to exist alongside this one.

This is the Personal Health Record of:

Name	<input type="text"/>	GM No.	<input type="text"/>
Address	<input type="text"/>	Date of Birth	<input type="text"/>
		Telephone	<input type="text"/>

Useful Contacts:





Main NF Doctor	<input type="text" value="NAME"/>	<input type="text"/>
GP	<input type="text" value="NAME"/>	<input type="text"/>
Optician	<input type="text" value="NAME"/>	<input type="text"/>
Dentist	<input type="text" value="NAME"/>	<input type="text"/>
School (if applicable)	<input type="text" value="NAME"/>	<input type="text"/>
NF Special- ist	<input type="text" value="NAME"/>	<input type="text"/>

NF2 CLINIC DETAILS
Genetics Service should attach a printed sticker here with clinic address and contact details.

Specialist Doctor Details

Sheet Number





People with NF2 are often looked after by more than one specialist, and this page is a record of all the doctors who are involved in caring for you. If you visit a specialist doctor who isn't listed, ask them to add their details below.

Name	Title/Specialty	Address/Phone Number
		
		
		
		

Specialist Doctor Details...continued

Sheet Number

People with NF2 are often looked after by more than one specialist, and this page is a record of all the doctors who are involved in caring for you. If you visit a specialist doctor who isn't listed, ask them to add their details below.

Name	Title/Specialty	Address/Phone Number
		
		
		
		

Information for You and Your Family (part 1)

What is Neurofibromatosis Type 2?

Neurofibromatosis Type 2 (NF2) is a rare genetic disorder. It varies from person to person—some people will have many health problems, others will follow a milder course.

Almost everybody who has NF2 will develop benign (not cancerous) tumours on their nerves.

What Causes NF2?

It is caused by a fault in one of your genes. Genes are responsible for individual characteristics such as eye and hair colour. People with NF2 have a fault in their NF2 gene.

Half the people who have NF2 are the first person in their family to be affected. Others may have inherited NF2 from an affected parent. If you have NF2, usually there is a 1 in 2 chance of passing this on to each child. If planning a family, it may be helpful to discuss this area with a geneticist to explore current options and testing issues.

If you have NF2, genetic testing of your children may be possible. Discussion about this usually takes place around ten years of age.

What are the Features of NF2?

Almost all people with NF2 will have one or all of the following:

- Vestibular Schwannomas (VS)—tumours that grow on the hearing nerves that pass from the brain to the ear
- Spinal Schwannomas— tumours that grow on the nerves leaving the spinal canal
- Meningiomas— tumours that grow on the covering of the brain or spinal chord
- Peripheral Schwannomas— lumps that grown on the nerves supplying the skin and muscles
- Cataracts— these may be present and rarely cause visual problems.

Tumours can cause problems because they are growing in confined spaces and may press on nerves and interfere with their function. When this affects the vestibular nerve, this can lead to symptoms such as tinnitus, balance difficulties and hearing loss. Spinal tumours may cause numbness, tingling and weakness in an arm or leg.

Information for You and Your Family (part 2)

Management of NF2

When a diagnosis of NF2 is made, a full health assessment should be undertaken to work out how NF2 affects you, and to enable doctors to plan effectively for the future. This assessment will include your account of any symptoms you have noticed, a neurological examination (testing your balance, strength, and sensation), and scans (usually Magnetic Resonance Imaging), hearing tests and eye checks.

When this assessment has been made, the doctors caring for you will discuss with you what steps, if any, need to be taken.

You will receive regular checkups. Regular scans and hearing tests will be arranged so that your doctors can monitor the location and size of any tumours and take appropriate action.

Treatment

NF2 is a complex condition and the treatment options need to be considered carefully. Doctors familiar with NF2 will not offer treatment unless they consider it is essential. Identifying tumours on scans does not automatically mean they must be removed. Surgery may be possible to prevent health deterioration due to the pressure from a tumour. However this can carry some risk because tumours may be located close to important nerves.

A second option is to treat small VS tumours with a special type of radiation treatment called stereotactic radiosurgery or gamma knife. This technique involves aiming a very accurately focused beam of radiation at the tumour. This treatment is relatively new and is controversial in NF2.

In some circumstances, the surgeons may recommend monitoring the tumours for growth and delaying intervention. All these approaches will be discussed with you before a decision is made, outlining the benefits and potential risks.



It is important to be alert to any significant changes in your normal state of health. You should seek help from your doctors/specialists if you notice any unusual symptoms. It is important that you attend scan and other hospital appointments that are arranged, and that you alert your doctors if a planned scan is overdue. A record of your scans is kept on page 13.

Information for Health Care Professionals (part 1)

Diagnosis

Neurofibromatosis Type 2 (NF2) is an autosomal dominant condition occurring in approximately 1 in 35,000. Half of individuals inherit the condition from an affected parent, In the other half it results from a spontaneous gene mutation.

The gene responsible for NF2 is known, and a genetic test is available, but diagnosis is usually made on clinical grounds.

- Bilateral Vestibular Schwannomas (VS)
- 1st degree family relative with NF2 and unilateral VS or any 2 of: Meningioma, schwannoma, glioma, neurofibroma, posterior subcapsular lenticular opacities
- Unilateral VS **and** any two of: Meningioma, schwannoma, glioma, neurofibroma, posterior subcapsular lenticular opacities
- Multiple meningiomas (two or more) and unilateral VS or any two of: Schwannoma, glioma, neurofibromas, cataract

Often the first sign of NF2 may be hearing problems: hearing loss, tinnitus and balance difficulties. Rarely NF2 may be diagnosed in a young child presenting with facial weakness or muscle wasting in a limb, or a spinal tumour.

Management of NF2

This is a rare condition, and patients are usually managed in specialist centres where there is extensive experience of VS surgery. A multi-disciplinary team approach enables shared decision making, regular monitoring for health changes, serial scanning and links to ancillary health support services.

Decisions relating to surgery are often complex and need careful consideration, taking account of the patients preference. The aim is to ensure conservation of function and quality of life. The presence of a tumour does not indicate need to remove it. Experienced NF2 surgeons will weigh up the possible benefits of tumour removal and the likely outcome.

Deafness, facial weakness post VS surgery, loss of the blink reflex and mobility problems can be a consequence of surgery, however not proceeding with surgery may also carry risk. Practical and psychological support services should be considered for patients adjusting to the profound health changes that occur with NF2.

Auditory rehabilitation by cochlear implant (CI) or auditory brain stem implant (ABI) may be offered by some centres. Referral to the LINK centre (www.linkcentre.org) may be helpful for those patients likely to lose their hearing.

NF2 is a completely different condition to NF1

Information for Health Care Professionals (part 2)

The Role of the Regional Genetics Service

The clinical geneticist is usually involved in the diagnosis of NF2. They also have a role to play in ensuring patients receive appropriate follow up including genetic counselling, together with information for patients and their families. The clinical geneticist may also act as a co-ordinator where a number of different doctors and specialities are involved.

All NF2 patients and their families should have access to genetic testing. This is because early detection of tumours is important in improving health outcomes. In most NF2 families it is possible to test for the mutation. The question of testing for children will be raised as part of the genetic counselling discussion. Consideration for pre-symptomatic testing should begin at about 10 years. At this age, audiology screening is offered with baseline MRI scan at 12-14 years if a definitive test is not available.

Advice and Further Information

Clinical Management guidelines for NF2 are available on request from the Regional Genetics Service or can be downloaded from our website www.mangen.co.uk

If you are still unsure about any aspect of caring for someone with NF2, you can telephone the Regional Genetics Service or visit our website for specialist advice.

NF2 CLINIC DETAILS

Genetics Service should attach a printed sticker here with clinic address and contact details.



Patients with NF2 may have hearing loss or be deafened. They are likely to use lip-reading as part of their communication skills so bear this in mind when talking to them.

Functional Summary

This page is a summary of how you are affected by your NF2. Your doctor will fill this page in with you.

Date that this page was completed

Hearing and Communication

I have: Normal Hearing Unilateral Deafness? Right Left Total Deafness?

If Deafened I Use: Lip Reading Yes No Sign Language Yes No

Cochlear Implants

I have an effective ABI/Cochlear Implant Yes No Date Implant Fitted

Implant Performance

Balance and Mobility

I am fully mobile

I walk with difficulty

I use a wheelchair

Hearing & Vestibular Schwannoma Record

Your NF2 specialist will complete this page. The information will be updated at each appointment.

LOCATION	DATE									
RIGHT SIDE	Size									
	Hearing Loss									
LEFT SIDE	Size									
	Hearing Loss									

Other NF2 Issues (Page 1)

Your NF2 specialist should complete this page and update it when you attend your clinic appointments.

Features	Description	Active/Static	Surgery Dates and Times
Meningioma			
Spinal Tumours			

Other NF2 Issues (Page 2)

Your NF2 specialist will complete this page. The page will be updated when you attend your review appointments.

Features	Description	Investigations/Management
Ocular Involvement		
Peripheral Nerve Involvement (e.g Schwannomas, neuropathy)		
Other Features		

Scan History

Sheet Number

You and your doctors will use this page to keep a record of when and where scans take place and when they are due.

IF A SCAN BECOMES OVERDUE FOR ANY REASON LET YOUR DOCTOR KNOW IMMEDIATELY.

Organised By	Date Required	Specify Scan Site (✓ type)	Hospital Location	Date Scan Took Place	Date Scan Next Due
		MRI <input type="checkbox"/> CT <input type="checkbox"/> OTHER <input type="checkbox"/>			
		MRI <input type="checkbox"/> CT <input type="checkbox"/> OTHER <input type="checkbox"/>			
		MRI <input type="checkbox"/> CT <input type="checkbox"/> OTHER <input type="checkbox"/>			
		MRI <input type="checkbox"/> CT <input type="checkbox"/> OTHER <input type="checkbox"/>			
		MRI <input type="checkbox"/> CT <input type="checkbox"/> OTHER <input type="checkbox"/>			
		MRI <input type="checkbox"/> CT <input type="checkbox"/> OTHER <input type="checkbox"/>			

NF2 Review Appointments

Sheet Number

This page will be completed when you visit your specialist for a review. At this appointment, the doctor will carry out an examination and review any symptoms.

Date

Doctor

Clinic Location

Next Review Due

Doctors Comments

Date

Doctor

Clinic Location

Next Review Due

Doctors Comments

Details of Other Hospital Visits and Appointments

Sheet Number

1

People with NF2 may be under the care of several specialist doctors and health care professionals. This page provides space for them to record brief details about the appointment and will be a record of who you saw and why.

When (Date)	Where (Hospital/GP etc)	With Who (Name)	Doctors Comments

Details of Other Hospital Visits and Appointments...cont

Sheet Number

People with NF2 may be under the care of several specialist doctors and health care professionals. This page provides space for them to record brief details about the appointment and will be a record of who you saw and why.

When (Date)	Where (Hospital/GP etc)	With Who (Name)	Doctors Comments

Sources of Information

Neurofibromatosis Association

The Neurofibromatosis Association, Quayside House, 38 High Street, Kingston on Thames, KT1 1HL

Helpline: 0845 602 4173 Website: www.nfauk.org

The NF Association is a registered charity that provides help, support and advice to those affected by either form of NF, their families and the professionals working with them. To do this they employ a team of hospital based professional NF Specialist Advisors around the UK. They provide relevant, up to date information on both forms of NF for professionals, families and individuals and also fund research. The Association provides activity holidays and breaks for children and adults, and encourages the establishment of local groups and volunteers throughout the UK.

Contact A Family

Helpline: 0808 808 3555 Website: www.cafamily.org.uk

The Contact a Family website is for families who have a disabled child and those who work with them or are interested to find out more about their needs. Contact a Family is the only UK charity providing support and advice to parents whatever the medical condition of their child. They have information on over 1,000 rare syndromes and rare disorders and can often put families in touch with each other.

LINK Centre

Telephone: 01323 638230 Website: www.linkcentre.org

LINK provide a variety of services for deafened people, their families, and the professionals who work with them. They cater specifically for people who have grown up with normal or near-normal hearing, then for one reason or another lost all or most of it.

Internet

The internet contains a wealth of information about medical conditions and treatments and users need to be confident that the information they read on the internet is reliable and accurate. The Judge Health website **www.judgehealth.org.uk** has guidelines that help you make informed decisions about Web sites and gives advice on how to search the internet for health information.

My Notes

You can use this page for any other information you wish to remember or record. For example, use it to write down any questions you want to ask at your next appointment. Ask your Doctor for more sheets if you need them.

A large, empty rectangular box with a thin black border, intended for the user to write their notes. It occupies most of the page below the introductory text.

My Notes (continued)

A large, empty rectangular box with a thin black border, intended for the user to write their notes. It occupies the majority of the page's vertical space below the header.

The development of this document has been a truly collaborative process. We would like to thank everyone who has contributed by sharing his or her experiences and knowledge especially:

*NF Association
North West Regional Genetics Service Nf2 Consensus Group*

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