

GUIDE FOR TEENAGERS

NEUROFIBROMATOSIS TYPE 1 (Nf1)

By Karen Ferguson

As a teenager with Neurofibromatosis Type 1 (Nf1) you might be asking more questions about your condition than ever before.

If you have Nf1 then it's good to have as much knowledge on it as possible. Knowing about your condition can mean you will worry less, and you can answer most of the questions other people may ask you.

At the end of this booklet there's a sheet where you can fill in any medication you take and a tick sheet for how your Nf1 affects you and what consultants you see regularly. It might be helpful to keep this information handy rather than keep repeating yourself.

So here we go.....

Nf1 is a genetic condition that happens when a certain gene changes.

Genes are what tell our bodies how to grow and develop and there is a gene called the Nf1 gene. Everyone has two copies of the gene – you receive one copy from mum and one copy from dad - even people who don't have Nf1. Sometimes when the gene is copying over it is changed – a bit like a spelling mistake – and when it is changed it doesn't work as it should – resulting in Nf1.

When a person with Nf1 becomes a parent they then pass a copy of the gene to their baby. Since they have a "good" copy of the gene and a "faulty" copy then they have a fifty-fifty chance of passing on the gene with Nf1. Half of all babies born with Nf1 inherit it from a parent with the condition.

The other half of babies born with Nf1 are the result of a brand-new change in the gene as it's being "copied over" from their parents (this is a new mutation). It's no one's fault.

If you have Nf1 you will have a fifty-fifty chance of passing it onto any children you may have. This is the same whether you inherited it from one of your parents or whether you were a new "mutation".

Nf1 does not skip a generation and it affects males and females equally. It doesn't matter your race, skin colour, social status or where in the world you live - the same rules apply. Nf1 doesn't make any allowances.

FEATURES OF Nf1

To be given the diagnosis of Nf1 you must have two features from the list below:

- Six or more café au lait spots
- Freckles in the armpit or groin
- Lisch nodules
- Two or more neurofibromas or a plexiform neurofibroma
- An optic glioma
- A specific bone problem – either sphenoid or tibial dysplasia
- A parent, brother or sister or child with Nf1

It's worth noting that most people with Nf1 never develop all the features. The good news is that most of the features are cosmetic and don't cause any medical problems and they certainly aren't life threatening. On occasion, some of the above may require medical attention and this will mean hospital visits and discussions with your specific consultants. There is nothing to be embarrassed about when discussing your condition. Nf1 is a part of you and it's important you look after your health.

CAFÉ AU LAIT SPOTS

The most common feature seen in Nf1 is the flat, tan birthmarks (CALs) that you probably have on your skin. These won't cause you any medical problems. Some people may have just six and other may have many more. Having lots of CALs doesn't mean your Nf1 will be worse than someone who has less.

FRECKLING

Most people with Nf1 develop freckling in their armpits or groin. These freckles are helpful in diagnosis Nf1 but don't cause any medical problems and are certainly nothing to worry or be embarrassed about.

LISCH NODULES

These are freckles on your iris – the coloured part of your eye. They don't affect your eyesight in any way but again are important for diagnosing Nf1 and nearly everyone with Nf1 will develop these by the time they are adults.

NEUROFIBROMAS

The word “Neuro” refers to the nerves within your body. If you have Nf1 you may develop bumps on the nerves called neurofibromas. These look like lumps under or on the skin. They can grow anywhere there are nerves. They often start to show up during puberty. Sometimes fibromas grow for a while but will eventually stop. No one can predict how many anyone might develop. They don’t usually hurt but sometimes they develop in places where your clothes might rub against them making them a bit tender and sometimes they might itch.

PLEXIFORM NEUROFIBROMAS

These are usually bigger than the ones we just mentioned. They can feel like a lot of small lumps bunched together.

OPTIC GLIOMA

This is a tumour that can develop on the nerve which connects your brain to your eye, also known as the optic nerve. Medical treatment and management are required here to avoid damage to your eyesight. When you were younger you may have had an MRI to look for these as they are something doctors look for in young children. It would be very unusual for a teenager to develop one that would cause any problems.

BONE DYSPLASIA

These are present at birth or just after and are not very common. So if you haven’t been diagnosed by now then you have nothing to worry about.

FAMILY HISTORY

Not everyone with Nf1 has someone else in the family with the condition.

WHAT ELSE CAN HAPPEN AS A TEENAGER?

You probably already have freckles and café au lait spots.

You might see a few lumps and bumps (neurofibromas) start to appear during your teenage years. They grow slowly and may look like a pimple at first. You won’t wake up one morning covered in neurofibromas.

If you have a neurofibroma that is painful or if it's in an awkward place – maybe rubbing on clothing or catching on things – or you feel self-conscious because of it, then it's possible you can have it removed. There is never a guarantee that it won't grow back, or another won't develop near the same spot. It's best to speak to your genetics doctor about this and then be referred to a plastic surgeon who has experience in dealing with Nf1.

STATURE

You may think you are a little shorter than your friends – Nf1 can make you a little shorter.

LEARNING

Every single person in the entire world learns differently, and that is why teachers must undergo continual training. Some people learn better by watching, others through doing and others through discussion. A person with Nf1 will often learn at a different pace than someone without. This isn't a reflection on intelligence but is in fact to do with how your brain best processes information.

It doesn't mean you are dumb or can't learn – it does mean that they might need some additional support in school or at college/university. Even if you struggle in school it doesn't mean you can't go on to college or university with the right help.

FREQUENTLY ASKED QUESTIONS

Q. Does having Nf1 mean I am different?

A: The short answer is NO – other than that we are all different to each other. Most people won't ever know you have Nf1 unless you chose to tell them.

Q. How many neurofibromas will I get?

A: There is absolutely no way to predict how many you may develop throughout your lifetime, but you can have most of them safely removed.

Q. How can I stop neurofibromas from growing?

A. At the moment there is no medication to do this but there are medical trials taking place. You might find that you develop neurofibromas during times of hormone change i.e. puberty and pregnancy.

Q. Will Nf1 stop me from doing things?

A. Unless you have a complication and your consultant/geneticist has advised you not to do something then the answer is no. People with Nf1 can generally do what their friends do – play sport, learn to drive, go to college, find a job, get married, have children.

Q. Should I tell my friends I have Nf1?

A. You may want to tell everyone you have Nf1 or you may prefer to tell only your very closest friends and family. It's a personal decision and one you might want to discuss with a parent/support worker or close friend (if that person already knows). Remember – it's your condition and your decision whether to share that information. Having Nf1 is nothing to be ashamed of but if you do tell people be prepared for lots of questions. Be ready to explain how Nf1 affects you. This booklet should help.

Q. Can I have the HPV Vaccine?

A. Yes

Q. Is Nf1 affected by the sun?

A: No - but everyone should protect their skin with high factor sun cream whether they have Nf1 or not.

Q. Is Nf1 contagious?

A: Absolutely not. Nf1 is caused by a faulty gene in your body. The only person you could pass it onto would be your own child.

Q. Can Nf1 be cured?

A. Currently there is no cure for Nf1 but there are treatments available for the complications.

Q. Will I die because I have Nf1?

A. Very unlikely. Most people who have Nf1 live full and normal lives.

Q. Can I get any help at school/college?

A. Yes, you can. If you are struggling with coursework then you need to have a chat with your teacher/ tutor. Explain that you have Nf1. They may already know but not be aware how it affects you.

Q. Should I tell an employer I have Nf1?

A. If you have Nf1 then it is advisable to let your manager know before you start your job. Your manager is not allowed to tell anyone else without your permission and they will only use this information to help you do your job better. If you choose not to tell your employer about your Nf1 they may not understand if you need to take time off for hospital appointments etc. or need a little more training than others. Think ahead when you are applying for jobs even – part time ones.

Q. Does Nf1 make you itch?

A. It can. If this happens speak to your GP about a prescription for Telfast. These are antihistamines which do not make you drowsy but will help stop the itching. Try not to use perfumed soap, shower gels etc. and make sure those showers and baths are not too hot.

Q. Can Nf1 cause me to be depressed?

A. It's not unusual to be down or depressed if you have Nf1. The condition itself isn't causing it but the impact it's having on you can contribute if you feel depressed. Talk to someone – mum, dad, siblings, aunts, cousins, friends – anyone you feel you can confide in. If you feel you can't discuss your feeling with your family or friends then maybe your GP?

If you don't feel that talking to a close one is an option then there is a list of websites where you can find help at the end of this booklet. The main thing is to speak to *someone*. Either face to face, on the phone or even online. Sharing your feelings really will help you feel better. It may be you need some medication to help you over this period, or it may be you just need someone who understands and who can encourage you through this time. Depression is nothing to be ashamed of – it can happen to anyone at any time. Sometimes the smiling happy person you envy is a person hiding depression.

USEFUL WEBSITES

www.funnylumps.org

www.youngminds.org.uk

www.nopanic.org.uk

www.childline.org.uk

www.themix.org.uk

www.samaritans.org

www.breathingspace.scot

www.ocdaction.org.uk

HOW Nf1 AFFECTS ME

ASD	Learning difficulty
ADHD/ADD	Low self esteem
anxiety	Lacking in coNfidence
Brain tumour	Leg length discrepancy
Behavioural problems	Microcephaly
Café au lait spots	Neurofibromas (tumours)
Chronic pain	Noonan's syndrome
Chronic constipation	Overgrowth gene
Chronic diarrhoea	Optic glioma
Developmental delay	Puberty (early)
Delayed speech	Puberty (delayed)
Delayed walking	Pseudoarthrosis
Difficulty with fine motor skills	Plexiform
Difficulty with gross motor skills	Spinal cord tumour
Epilepsy	Social difficulties
Eating difficulties	Sleeping difficulties
Freckling in groin or armpits	Seizures
High blood pressure	Scoliosis
Headaches	Short stature
Hearing impairment	Sphenoid dysplasia
Hypotonia	Separation anxiety
Hypermobility	Sensory processing disorder
Hydrocephalus	Tiredness
Itching	Visual impairment
Lisch nodules	Vomiting
Lack of energy	

MEDICATION I TAKE

NAME OF MEDICATION	WHO PRESCRIBED	HOW MUCH I TAKE

I HAVE AN ALLERGIC REACTION TO

SUBSTANCE/FOOD	HOW I REACT

DOCTORS AND THERAPISTS I SEE

DEPARTMENT	DOCTOR	HOSPITAL	PHONE NO
Audiology			
Cardiology			
CAHMS			
Dermatology			
Dietician			
ENT			
Endocrinology			
Genetics			
Gastroenterology			
Maxo Facial			
Neurology			
Ophthalmology			
Orthopaedics			
O.T.			
Oncology			
Physio Therapy			
Podiatry			
Pain Management			
Physiotherapy			
Plastics			
Paediatrics			
Psychiatry			
Rheumatology			
Renal			
Speech & Lang.			
Urology			

Funny Lumps
Registered charity SC039910
www.funnylumps.org



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