

A FEW BASICS



**YOU DON'T
NEED TO GO IT
ALONE**

HELLO AND WELCOME...

... to your personal guide to living with neuroendocrine tumours, or NETs. No doubt you'd rather you weren't reading this - we would too. But as you are, we hope it'll give you a lot of useful and straightforward information about a subject it can be hard to find out about.

This isn't a book that's intended to be read cover to cover. Instead, we hope you'll dip in and out when you want more information about tests, time in hospital, talking about cancer, living with a NET or whatever else is on your mind.

We've designed it so you can pull out pages and take them to appointments, and you can store extra information in the flap at the back, too. So, carry parts of the guide or the whole thing with you; write in it; stick reminders in it and get people to read it, especially if you get tired of explaining what NETs are. We'd love to see copies of this handbook looking dog-eared and well used.

We're here for you

At the NET Patient Foundation, we've been supporting people who are diagnosed with NETs since 2006. We have put this guide together based on what people have told us; to include what will be most useful - after diagnosis, through treatment and living with NETs.

You'll find plenty of medical information - because our charity is run by a variety of medical specialists in NETs - explaining what's happening inside your body in clear, simple language is a big part of what we do.

But this guide goes well beyond that, because a cancer diagnosis doesn't only affect your body. It can have a big impact on your emotions, your relationships, your job, your bank balance and many other parts of your life. So we cover all of that and more too - guided by people who've been in your position and told us what they wished they'd known.

We really hope you'll find a lot that's useful here. Getting diagnosed with a NET can leave you feeling confused and alone, but we've written this book to help cut through the confusion and to make it very clear that you're certainly not on your own.

Cathy Bouvier
CEO NET Patient Foundation

HEARING THE NEWS

People react in all kinds of ways when they are told they have cancer.

Maybe you fell silent or couldn't stop asking questions. Maybe you wanted to be alone or wanted your whole family around you. Maybe you felt numb or overwhelmed by emotion. Maybe you even felt relief.

However you reacted is completely normal. There's no wrong or right way. The situation with NETs can be even more complicated because these types of tumours are often misdiagnosed initially or can take a long time to be correctly identified.

It's not unusual at all to feel unwell for years before a NET is diagnosed, or to be wrongly told that you have another condition, like irritable bowel syndrome, Crohn's disease, gastritis, asthma or problems linked to the menopause or blood pressure.

On the other hand you may not have felt unwell at all and have received your diagnosis as a result of a routine health check.

Not all NETs produce symptoms.

When the diagnosis is made, you may find it difficult to get your head around the fact that not only do you have cancer, but you have a type of cancer you, and those around you, may never have heard of.

You don't need to go it alone

The main thing to try and remember at this stage is that you really aren't alone. It's easy to feel isolated when you find out you have an uncommon condition, but getting the correct diagnosis means you should now have access to the right expert care team who understand your condition and can help.

So if you have questions, ask them and keep asking until you understand the answer. We hope a lot of issues that might come up are answered in this book, but don't be afraid to ask your care team anything that isn't - or to give us a call. There really is no such thing as a stupid question.

And while we know it can often be difficult, talking to family and friends can help - though it may be hard to decide how and when. Bottling things up can often make things seem worse.

Many people find our online forums and support groups are a helpful way to talk to others, who really do understand what being diagnosed and living with NETs is all about. You can find out more about these on our website. Our contact details are below:

Helpline number
0800 434 6476

Office number
01926 883487

nikie@netpatientfoundation.org
www.netpatientfoundation.org

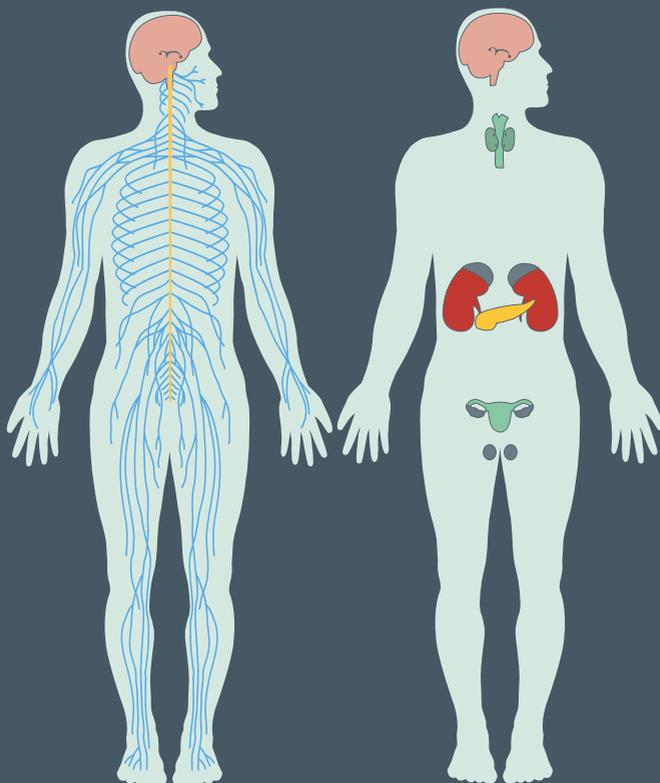
SO WHAT ARE NETs?

It can be hard enough coming to terms with the fact you have cancer, let alone trying to understand what the neuroendocrine system is or how neuroendocrine cells can cause a tumour.

So if you're struggling to get your head around this, hopefully this section will make things a little clearer.

The Neuroendocrine System

- the endocrine and nervous systems working together



Neuroendocrine cells help our bodies to function

Our bodies are made up of billions of cells - from blood cells to brain cells. **Neuroendocrine cells** are found in many parts of the body, throughout a network called the **neuroendocrine system**.

These specialised cells produce hormones (endocrine action), usually in response to signals from other cells, or the nervous system (neurological stimulus).

Neurological stimulus for an **endocrine function** = Neuroendocrine. These cells have different functions depending on where they are, but they all release hormones and other chemicals into the blood to help our bodies work normally. For example:

- Neuroendocrine cells in the **digestive system** produce hormones that move food through your intestines and help to break down food in your gut
- Neuroendocrine cells in the **respiratory system** produce hormones that help with the development of the lungs and regulate breathing

- Neuroendocrine cells in your brain produce hormones including oxytocin, which plays a role in everything from social bonding to breastfeeding
- Other neuroendocrine cells help to control things like **blood pressure** and **heart rate**.

You can think of the neuroendocrine system as a bit like a bicycle inner tube. It's essential to keep things running smoothly - but you're hardly aware it's there.

All neuroendocrine cells release hormones and other chemicals that help our bodies work normally.

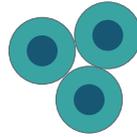
Cancer is caused by cells acting strangely

When neuroendocrine cells work well, our bodies work well. But, as in all cancers, problems start when abnormalities occur and the cells start growing and behaving abnormally.

A normal cell:

- Exists in a specific place in the body
- Divides and replicates itself only when necessary
- Has a life cycle, so does its job then dies off
- Repairs or destroys itself if it gets damaged
- Doesn't cause damage to neighbouring cells by growing too large or invading them.

Normal cell

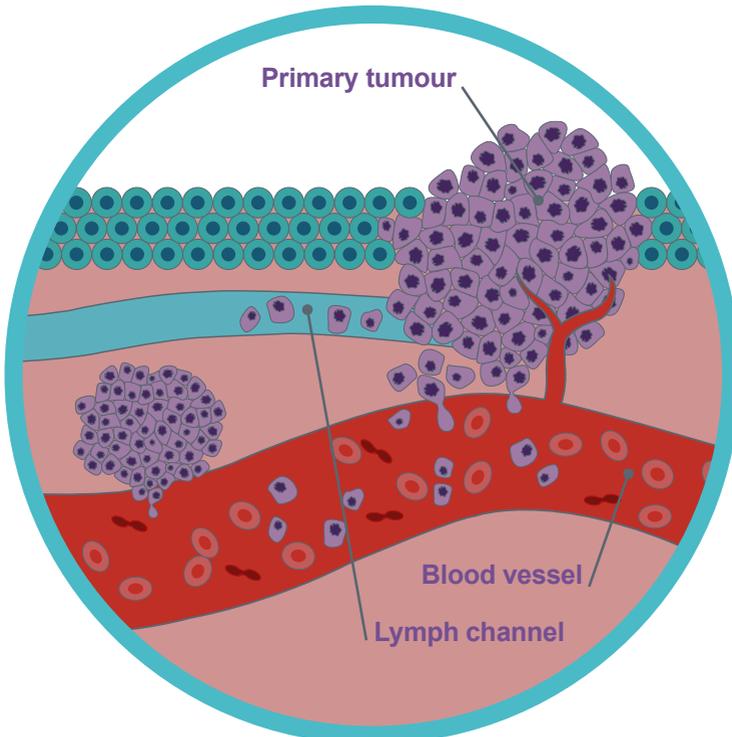


Cancer cell



A cancer cell:

- Can detach from the tumour (collection of cancer cells) and travel to other parts of the body
- Keeps dividing and growing - as it doesn't know to stop
- Often grows abnormally, so can't perform a function properly
- Doesn't destroy itself if it gets damaged
- Ignores warnings from neighbouring cells to stop growing and can invade them.



So how do NETs differ from other cancers ?

In more common cancers like carcinomas (which includes most lung, breast, prostate and bowel cancers), between 20% and 100% of cells in a tumour will be actively replicating and growing.

In NETs, however, the percentage of cells that are active can be lower. This can be described using a grading system:

- In Grade 1 NETs, fewer than 3% of tumour cells are actively dividing
- In Grade 2 NETs, between 3% and 20% of tumour cells are actively dividing
- In Grade 3 NETs (known as NECs - neuroendocrine carcinomas), more than 20% of tumour cells are actively dividing.

In lower grade NETs, the relatively small percentage of cells dividing means that tumours grow slowly, which is one of the reasons it can take years to diagnose them. Grade 1 NETs are sometimes described as being like 'cancer in slow motion'.

Higher grade NETs (NECs) often grow more rapidly and can behave more like the more common cancers. The highest-grade NETs are actually called neuroendocrine carcinomas (NECs), rather than neuroendocrine tumours. You can read more about this in section 2.

Around 4,500 new NETs are diagnosed in England each year.

[Based on 2013-4 PHE data]

For NETs explained turn to section 2, page 13.

Malignant vs benign NETs (cancer vs not cancer)

Not all NETs are malignant, a small percentage are truly benign.

Your tumour may be described as benign but it is important to be absolutely clear as to whether it is truly benign or just behaving benignly (that is very slowly growing).

This guide is for those diagnosed with a malignant NET - whatever the rate of growth.

THE NETs MYTHBUSTER

To help overcome common misconceptions related to NETs, we asked people we support about the issues that confused or concerned them after diagnosis. These are some of the myths they wanted us to put right...

Myth #1

Incurable and terminal are the same thing

Not true

If a condition is incurable it can't be cured, which is not the same as being life-shortening. A terminal illness is when it can reasonably be expected to shorten a person's life to a limited amount of time (for example less than 6 months). But a condition that's incurable isn't necessarily terminal - diabetes, asthma, NETs.

That's an important distinction, because at the time of diagnosis NETs may well be incurable* but are rarely terminal. Many people can live well for years with a NET.

Myth #2

You don't have cancer

Not true

This is one that can really upset or confuse people, and rightly so. Because some NETs develop much more slowly than other cancers, you might meet people - including medical staff - who downplay their significance, or mistakenly refer to them as benign - which is massively unhelpful. Cancerous NETs may behave differently to some cancers, and may grow significantly slower - but they are, unfortunately, most definitely still a form of cancer.

Myth #3

NETs always grow slowly

Not true

Most low-grade NETs do grow slowly, but other moderate to high-grade NETs can grow as quickly as more common cancers. The rate of growth of even low-grade tumours can change over time, too - their behaviour can be unpredictable. If someone tells you that NETs are always slow to develop, they don't understand NETs.

*Just to be totally clear, a small number of NETs may be considered cured if they're caught early enough and have been completely removed by surgery - before they've spread or infiltrated nearby cells. Even in that situation, close follow-up care may still be needed.

Myth #4

NETs are always in the bowel

Not true

Strange one this, but it's certainly out there. Bowel NETs are more common (more than 40% of all NETs start within the small or large bowel), so it can sometimes be hard to find information on any of the other primary sites.

If you read section 2 of this book you'll see the many parts of the body in which NETs might start.

Myth #5

The stage of your tumour defines how serious it is

Not true

This is another myth it's important to tackle upfront, because NETs are not only defined by the stage they are at. The stage of a tumour can give important information about the primary site, and whether it has spread. However, other factors such as how active it is (grading), what the tumour cell looks like (differentiation) and your overall health, alongside tumour staging, will determine how serious it is.

So please make sure you always chat to your care team to get the full picture about your condition.

We explain more about how NETs are classified in section 2

Myth #6

Treatment is something that happens to you

Though it may at times feel like it - not true

This one isn't specific to NETs, but it is something vitally important to be clear on. In short, treatment shouldn't be something that just happens to you - it should be something you're constantly involved in.

Treatment cannot happen without your consent - your informed consent. If you don't feel you have received enough information to make a decision - ask for more, and more, again and again if need be, until you feel certain and confident in making a decision that's right for you.

Myth #7

Everyone responds to treatment in the same way

Not true

Absolutely not. You are unique - you were unique before you were diagnosed with a NET, and that's still true now. So don't let your experience be filtered through other people's, there will be similarities and shared experiences, however please remember that your condition and your reaction and response to treatment will never be exactly the same as anyone else's.

Not only are you unique, so is your NET, so even if someone else has the same type - your responses may be different.

GETTING THE RIGHT CARE

As soon as you're diagnosed with a NET, it's vital to get the best possible advice and treatment from the right team of people - and across the UK there are procedures in place to ensure this happens.

If you're diagnosed with a NET, a specialist, accredited, Multi-Disciplinary NET Team (MDT) should review your case on an ongoing basis.

That team will be made up of a range of clinical experts who specialise in NETs. We talk more about MDTs in section 3.

Getting referred to a specialist NET team

Referrals to specialist NET services work slightly differently across the UK. They can also vary slightly depending on who refers you - for example your GP, A&E staff or a specialist in another area of medicine. NB most NET specialist centres have specific NET referral forms that can be used.

This doesn't necessarily mean that you need to travel to and keep travelling to your nearest specialist centre, but it does mean that information about your situation, including the results of any scans and other tests, should be looked at by the NET MDT. This team can also advise on or arrange the most appropriate tests and treatment for you.

So wherever you are and whoever is caring for you, it is vital to get support from a specialist NET MDT as soon as possible.

You should be referred to one automatically, but if that doesn't happen you can ask for a referral to be made.

At the NET Patient Foundation we work with all of the UK's specialist NET services either directly or through UKI NETS. UKI NETS - The UK and Ireland Neuroendocrine Tumour Society - is a professional organisation made up of the UK and Ireland's experts and other clinicians and nurses who work in the field of NETs. The society maintains an up-to-date list of NET experts, clinics and accredited Centres of Excellence - along with a current list of research projects and clinical trials.

For a current list of experts, clinics and Centres of Excellence please visit the clinical practice page on the UKI NETS website: www.ukinets.org/net-clinics-research

TREAD CAREFULLY ONLINE

It's natural to want to have more information about your condition, and understanding what's happening definitely puts you in a position of power. But do remember to click carefully, because...



The 4am surf is not your friend

We've all been there. Can't sleep. Can't get comfortable. Thinking the worst. So you reach for your phone, tablet, laptop or pc . . . and half an hour later you're still reading something that's only making you more worried.

Trust us - the 4am surf is usually a bad idea. So if you can't sleep and think of a question you want answered, write it down - with or without some of the concerns you have, then try to rest. You can revisit it in the morning instead when things may seem clearer - or when you can, pick up the phone and talk it through with a friend, your GP or a member of your care team.

Balance matters

It's easy to be affected more by negative stories or by articles that seem to promise miraculous cures. So it's always worth checking how reliable that information is, for example, for any information on treatments - has it been published and reviewed in a reputable medical journal such as the British Medical Journal or New England Journal of Medicine? Are the claims supported by evidence and references?

It's also worth preparing yourself for reading things that you might not like, so try to look for a balance of information if you do hunt for answers online.

Some sites are more reliable than others

Your care team will be able to recommend websites you can trust to find out more about your condition.

Apart from our own pages, we cannot comment on or guarantee the content of websites, however you may find the following useful:

www.netpatientfoundation.org

Our online hub, where you can find further information including our support groups, events and online forums for people who have NETs

www.taect.scot

The website of Scottish NETs charity the Ann Edgar Charitable Trust

www.amend.org.uk

The Association for Multiple Endocrine Neoplasia Disorders

www.ukinets.org

The website of the UK and Ireland Neuroendocrine Tumour Society

www.enets.org

The website of the European Neuroendocrine Tumour Society

www.cancer52.org.uk

An alliance of 80 organisations that support people who have rare cancers

www.cancerresearch.org

For general information on cancer and cancer research

www.netcancerday.org

Celebrating the global day to raise awareness of NETs

www.nhs.uk

The NHS website

There are also online forums - these tend to be self-regulated or rarely censored - and so you may occasionally find some of the information contradictory or questionable - however they can be incredibly good for sharing experiences, discussions, information and making supportive friendships.

www.netpatientfoundation.org/forum-net-community/forum

Our online forum

www.facebook.com/netpatientfoundation

Our Facebook page

www.facebook.com/groups/27046513943

Friends of the NET Patient Foundation Private Facebook Page

www.facebook.com/groups/lovablelungnoids

A Facebook page focused on lung NETs

www.facebook.com/groups/583107718470111

A Facebook page all about high-grade NETs (NECs)

www.facebook.com/groups/insulinoma

An Insulinoma Awareness and Support Community

www.facebook.com/groups/NINETpatients

A Facebook page for people living with NETs in Northern Ireland

www.planetscharity.org

Southampton Hospital Charity fundraising for pancreatic, liver and neuroendocrine tumours

www.livingwithnets.com

IPSEN patient website

THE POWER OF BEING PREPARED

Whether you want to know every detail of what a NET can mean or would rather your care team just got on with it, there's a lot to be said for being prepared.

Everyone deals with illness differently. Some people want to know exactly what's happening, others prefer doctors to keep things as general as possible - and it's completely up to you how you approach your life with a NET.

However, trying to prepare for what is to come can help. This doesn't mean you suddenly need to spend hours reading medical textbooks or scouring websites about the neuroendocrine system. But it does mean that getting your head around some of the facts and potential implications of a NET diagnosis can put you in a stronger and maybe less stressful position.

Being prepared for treatments, for instance, can help you know what to expect, what is usual and what's not - for example what side effects could happen and how to deal with them, if they occur.

Being prepared for appointments and conversations with consultants can help you to think of the questions you may want to ask - that may help you to get the information you need.

Being aware of the effect your diagnosis may have on certain friendships and relationships may help to prepare you for those who might act differently around you or begin to drift away. Others will carry on as normal and there will be those who may pleasantly surprise you.

We talk more about the emotional impact and relationships in section 6.

Any type of cancer can cause your life to change in all kinds of ways, and NETs are no different. So being prepared for those changes, and spending a little time thinking about how you want to respond, is usually a very good idea - and it's one we'll keep coming back to throughout this book...