

Cancer of the Unknown Primary Factsheet

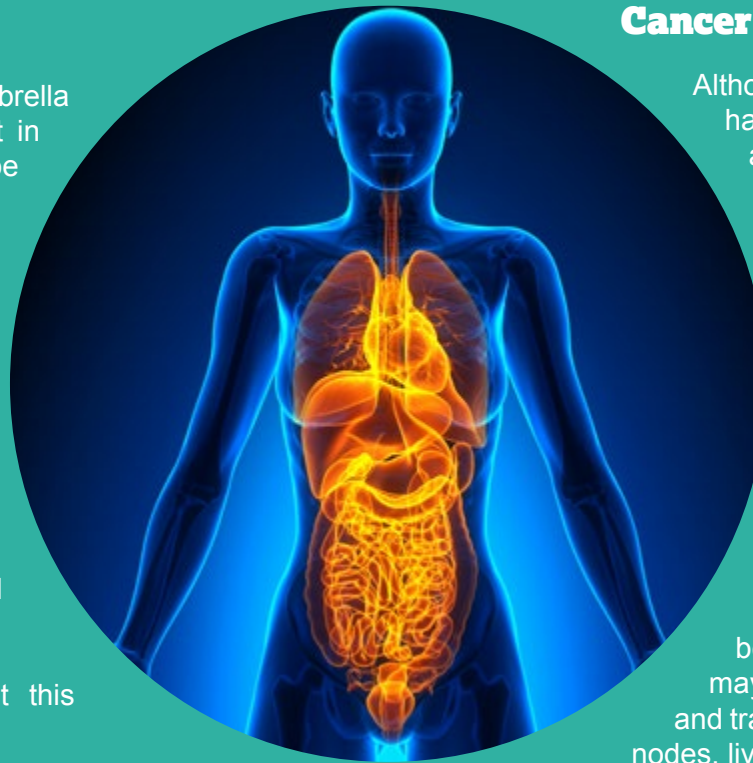
The Facts

Neuroendocrine Neoplasm (NENs) is an umbrella term used to cover a group of cancers that start in neuroendocrine cells. These cancers may also be referred to as NETs, NECs, or even Carcinoids. NENs occur when neuroendocrine cells stop working normally and start to grow or behave abnormally.

There are 2 key types of NEN:

1. NETs (neuroendocrine tumours) are called 'well-differentiated' and tend to have a slow to moderate growth pattern.
2. NECs (neuroendocrine carcinomas) - are called 'poorly differentiated' and tend to grow rapidly.

To simplify, we use the term NENs throughout this Factsheet.



Cancer of the Unknown Primary (CUP)

Although, it is usually possible to work out where a cancer has started by assessing symptoms and reviewing tests and scans, sometimes confirmation of the primary site is not always possible.

This may be due to the way NENs develop, they start at a size similar to a grain of sand and may take many months, possibly years to develop into the size that may be detectable by scans.

Despite the small size (sometimes simply a few cells) NENs may still spread to other organs (metastasise) – this does not necessarily indicate an aggressive nature, it may occur as a result of the cancer being present in your body for a long time. This means that the original collection of cells have been there so long, that over time, one or more cells may have had time to enter the blood of the lymph system and travel elsewhere in the body - most commonly the lymph nodes, liver, lungs or bone.

Your medical team may classify your cancer as being of unknown origin – or CUP (Cancer of Unknown Primary).

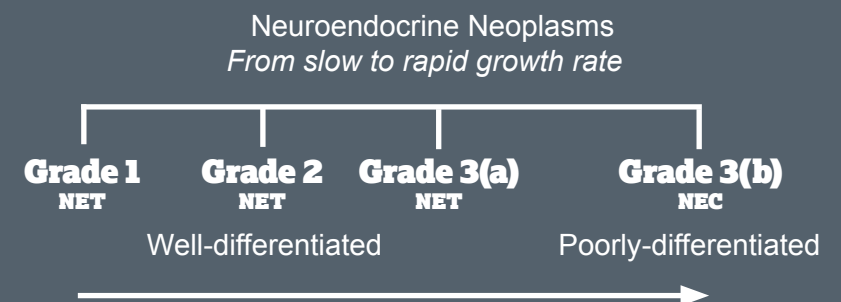
Grading

Not all cancers are the same or behave in the same way. Some may grow very slowly, may not have invaded nearby structures, or spread to other parts of the body at the time of diagnosis - but they have the potential to. However some may have already spread at the time of diagnosis.

The grade is the rate at which the NEN grows. You might see this referred to as 'Ki67' or 'Mitotic Index' mentioned in your clinic letters or medical reports. Grade 1 is slow-growing through to Grade 3, a more aggressive, rapidly growing disease.

The Grading Spectrum of NENs

Accurate, expert histopathology review is vital to confirming diagnosis to help guide best care.



Causes and / or risk factors

We do not know exactly what causes NENs, however research is ongoing, to truly understand both the pathology and the biology with the hope that this will lead to a much better understanding of the causes.

Most NENs do not run in families; however, there are a number of rare conditions that may increase your risk. Therefore, if other members of the family have been diagnosed with a particular cancer (especially aged 50 years or less) or cancer-associated genetic condition, it is important that you tell your specialist team about not only your medical history, but also any family medical illnesses or conditions.

So how is it treated?

For all patients, there are many things to consider in planning treatments. Your treatment will be personalised to you and the type of NEN you have.

Even if you have a diagnosis that sounds the same as another patient, your treatment and follow up plan may be different.



Your care team will discuss your treatment options with you - giving you both written and verbal information - to help you make an informed choice. Together you can agree on the most appropriate treatment for you. There is consensus agreement that all NEN patients should be reviewed by a specialist MDT.

Treatment options will depend on the type, position and size of your NEN, and whether (or to where) it has spread. It will also depend on whether you have any other health concerns and / or illnesses and your general health and fitness. A big part of meeting with your doctors or specialist nurse/ team, is to make sure you get the information you need to understand your condition.

Talking with your team about treatments

A big part of meeting with your doctors, or specialist nurse, is to make sure you get the information you need to understand what's happening, so that you can make an informed choice about your care.

Suggestions that may help when talking to your team:

- Prepare a list of questions that are important to you
- Ask for simple explanations - do not be worried about asking your nurse or doctor to repeat what they have said
- Take someone with you or ask if you can record the conversation.
- If you have a nurse specialist - keep in touch. They can be a great source of information and support for you.

Questions to ask at your appointment:

- What treatments are most suitable for me and my NEN? Do I have to have treatment?
- What is the aim of treatment?
- What are the possible side effects?
- How could the treatments affect my everyday life?
- What do you hope the impact of treatment will be?
- Will I be able to keep working during treatment? Or do I need to arrange time-off (e.g. for surgery and recovery)?
- How will I be involved about decisions about my treatment?
- Can I choose where I go for treatment?
- Can I get a second opinion about my treatment?

The key aim of treatment, should be to help you have the best possible care and quality of life, by ensuring access to appropriate treatment, management of symptoms and addressing what's most important to you. One or more of the approaches below may be suggested:

- Removal of all or part of your NEN
- Control of your disease, by slowing or stopping the growth of your NEN
- Palliation, or easing of, your symptoms.

Treatment options include:

- Surgery
- Somatostatin analogues (SSAs)
- Chemotherapy
- Targeted Molecular Therapies
- Peptide receptor radionuclide therapy (PRRT)
- Radiotherapy
- Endoscopic procedure
- Interventional radiology
- Irreversible electroporation (IRE also known as Nanoknife)
- Clinical Trials
- Symptom control
- Surveillance - *can be used to assess how well treatment is working or in periods between treatments (which may be months/years).*

Visit our website for explanations on all treatments available in the UK.



Physical well-being

Staying active during and after cancer treatment makes a lot of sense and there is evidence to support this:

- It can boost your energy levels and your mood
- It can help your bones, muscles and heart to stay strong
- It can help you to stay at a healthy weight.
- It can help you to sleep better and improve your appetite
- And it can increase your range of movement and control side effects like constipation.

Your care team can advise you on how much exercise or physical activity is right for you. Keeping a record of your progress can help, and so can reminding yourself of the benefits if you start to struggle. And make sure you choose activities you enjoy, otherwise it can be much harder to stick with them.

You may want to consider downloading the 'My NETs' mobile App from your App store. It can be used for day to day information gathering such as symptom tracking, managing and storing health details, (such as blood and other test results, diet, symptoms, medications and appointments).

What can I do to help myself?

Living well with NENs is possible, but may require adjusting expectations of what life following diagnosis may be like - many describe it as a "New Normal" : a way to live that supports both your physical and mental well-being.

Finding out more about your diagnosis, the tests and treatments that may be involved, can help in both dealing with the effects of diagnosis and decision-making. It can help to restore a sense of control. But everyone takes in information in different ways : it can sometimes feel as though you are being bombarded with information - whilst at other times you may feel there's not enough.

What can we do to help you?

- A dedicated nurse freephone helpline: **0800 434 6476**
- Our 'Natter' Support Groups are online or local, community based meetings that are available to offer an opportunity to meet others who are also affected
- Our Facebook (closed) groups provide a safe place for those affected to talk and help each other by sharing experiences and knowledge, to both give and receive advice, support and friendship
- A free, confidential counselling service for patients and supporters, family members or friends.

Visit our website for further details of the services listed above.



Neuroendocrine Cancer UK exists to support and inform patients and families from diagnosis, enabling access to the best care and treatment, whilst stimulating Neuroendocrine Cancer research, increasing national awareness and influencing improvements in outcomes.

Visit our website to read more about our work and for more in-depth information, additional resources and links related to everything that we have talked about in this leaflet.

Neuroendocrine Cancer UK
www.neuroendocrinecancer.org.uk

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Twitter: [@ncukcharity](https://twitter.com/ncukcharity)

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