



*This booklet was
produced with thanks to
the friends and family of
Edwin Rennie.*

**The NET Patient Foundation:
supporting the neuroendocrine cancer community.**

For further information and
to make contact telephone
0800 434 6476 or visit our
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Information for people affected by Carcinoid Heart Disease

What are neuroendocrine tumours (NETs)?

Neuroendocrine tumours (NETs) are a complex group of tumours that develop predominantly in the digestive or respiratory tracts, but can occur in many areas of the body. These tumours arise from cells called neuroendocrine cells.

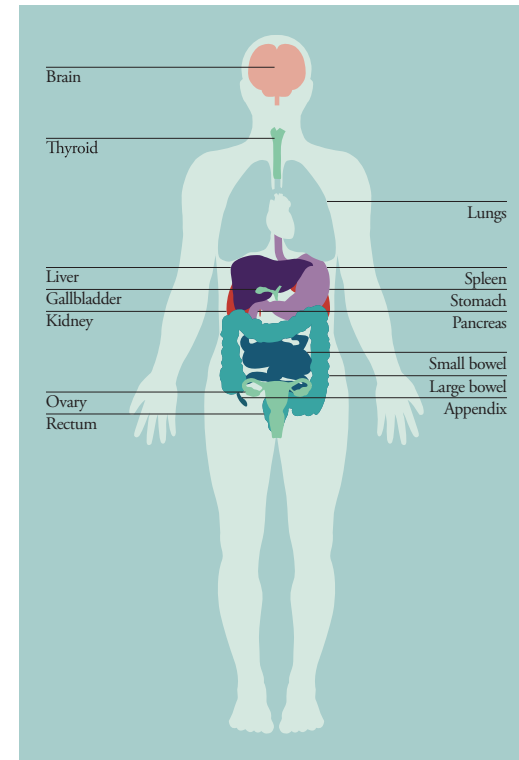
Like all cancers, NETs develop when the specialised cells undergo changes causing them to divide uncontrollably and grow into an abnormal tissue mass (tumour).

Neuroendocrine tumours can be benign or malignant. Back in 1907, when neuroendocrine tumours were first classified, they were named 'carcinoid' (meaning 'cancer-like') as they seemed to grow slowly and were therefore not thought to be truly cancerous. The use of the term 'carcinoid' is being phased out in medical literature, as we now know that these tumours can be malignant. Current terminology is either simply neuroendocrine tumours (or NETs) or your NET will be named dependent on the site of your primary tumour, for example a bronchial (lung) NET, a bowel NET or a pancreatic NET.

Where do neuroendocrine tumours occur?

For information about other types of NETs, please visit our website at www.netpatientfoundation.org

Examples of where NETs can occur as primary sites or as metastases (spread)



What is 'carcinoid syndrome'?

When neuroendocrine tumours spread ('metastasise'), the most common site for metastatic tumours ('secondaries') is the liver. Other areas of spread can, more rarely, include the bones, the lungs and the lymphatic system.

Many NETs have an associated syndrome caused when the neuroendocrine cells produce too much of a particular hormone. The most common of these is 'carcinoid syndrome', which is caused when too many hormones such as serotonin, histamine, somatostatin and chromogranin A are produced. The symptoms of carcinoid syndrome vary and can often be highly individual.

Typical symptoms include:

- flushing
- diarrhoea
- wheezing
- abdominal pain
- fatigue
- skin changes

It is by no means certain that you will experience 'carcinoid syndrome'. Not everyone with NETs will have this collection of symptoms, even if their disease has spread.

Complications

Carcinoid crisis

Sometimes patients may suffer a particularly bad episode of carcinoid syndrome triggered by stress, general anaesthetic or certain treatments.

Symptoms include intense flushing, diarrhoea, abdominal pain, wheezing, palpitations, low or high blood pressure, an altered mental state and, in extreme cases, coma.

Without treatment the complications can be life threatening, but if you are having any procedures your NET specialist will ensure you are monitored and may give you an infusion of a somatostatin analogue as a preventative measure. Your NET specialist will also liaise with any other team, for example a surgical team, and pass on the guidelines that are available as a preventative measure for patients at risk.

Carcinoid Crisis cards are available from the NET Patient Foundation.

AT RISK - Carcinoid Crisis

I have a neuroendocrine cancer with 'carcinoid syndrome'. I am at risk of a carcinoid crisis if I have an invasive procedure.

I will need perioperative prophylactic treatment of intravenous octreotide for acute surgery. I may need additional intravenous octreotide if I still have symptoms. A lower dose can be given subcutaneously for more minor procedures.

ALL DOSAGES GIVEN ON THE REVERSE OF THIS CARD

www.netpatientfoundation.org

What is Carcinoid Heart Disease?

Carcinoid heart disease develops in around 60-80% of patients with 'carcinoid syndrome', although it is clinically significant in a much smaller percentage. Some NETs release chemicals called serotonin and tachykinins into the blood stream that travel to the heart and can affect the cardiac valves.

Fibrotic plaques are deposited onto the valves and heart muscle, and these can eventually impair the function of the heart and cause leaky valves. Heart specialists do not yet fully understand why they build up, but know that they do in some NET patients, and that they can cause damage to the heart.

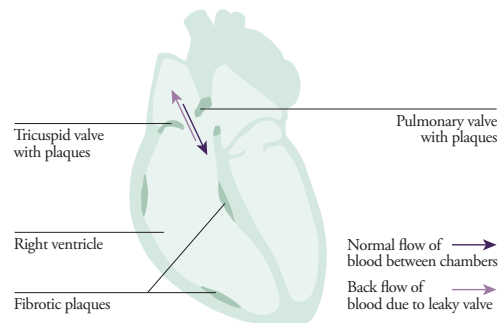
What are the symptoms of Carcinoid Heart Disease?

The main symptoms are breathlessness and swollen ankles that can progressively worsen if left untreated. However some patients have few or no symptoms at all.

What should I do if I notice symptoms?

Report them to your GP or your specialist so that you can have further tests.

Carcinoid Heart Disease



What tests should I expect?

To screen for carcinoid heart disease in patients without symptoms a blood test called pro-BNP is a very good screening test.

However, for anyone with symptoms an echocardiogram should be performed.

Routine BNP screening every year for patients without carcinoid heart disease is adopted by some NET specialists.

Doctors will assess your symptoms, examine your heart and organise the following investigations:

Echocardiogram, or ultrasound, of the heart: a procedure that is usually performed by a cardiac technician. At the same time a 'contrast echo' will sometimes be performed. This involves injecting a small amount of sterile water into a vein that is pictured as it passes around the heart.

Blood and urine tests to measure the concentration of chemicals and hormones released by the tumour, for example: pro-BNP, 5HIAA.

Depending on the results you may need two further investigations:

Transoesophageal echocardiogram – you may need to be admitted as a day patient for this procedure and you can be sedated if you wish. The whole procedure takes up to 30 minutes. You will have a small plastic tube inserted into a vein in your arm so that medication such as sedation can be administered, if required. This procedure allows better visualisation of the heart valves and is done by passing a small camera down the throat and into the oesophagus (food pipe) so that it sits behind the heart and sends pictures onto a screen that can be viewed by a cardiologist (a consultant heart specialist).

Cardiac catheterisation – this is also a day case procedure and the actual procedure takes about 30-60 minutes, although patients will need to stay in hospital for two to four hours afterwards. The procedure involves passing a small tube into an artery in the groin under local anaesthetic. The tube passes through the artery to the heart and a special dye is then passed down it. This dye can be picked up by special x-rays showing the cardiologist the vessels of your heart and allowing him or her to determine how severely your heart valves are affected.

Treatments

The decision about treatment is determined by your own particular disease and needs, so the treatments described below provide an outline of possible options but your specialist will discuss your own individual treatment plan with you before you start treatment.

Patients should ideally be treated within a specialist multidisciplinary team (MDT). Treatment is tailored to the individual patient and is determined by the severity of symptoms, the extent of the tumours and previous treatments, and any other existing medical conditions that could make a surgical option more difficult.

For some patients, the initial treatment plan may be to monitor the heart with a yearly echocardiogram, and one or more of the tests listed above.

Some patients may require surgical valve replacement. This is major cardiac surgery on a par with heart by-pass operation and involves replacing the damaged valves with metallic or tissue valves.

Your cardiologist will discuss which is the best option for you. Metallic valve replacement requires long-term use of blood thinning agents, for example the drug warfarin.

Multidisciplinary teams

The care of NETs can be complex, and for the patient the journey can encompass not only a whole host of emotions, but also a whole range of investigations, treatments and healthcare professionals. The very fact that there is often not just one treatment option at diagnosis and throughout the patient journey, means that there has to be a collaboration between all key healthcare professional groups, who are making clinical decisions for individual patients.

This collaboration has been termed an MDT (multidisciplinary team). This is a formula that is now being used across the world in the care of cancer patients.

An MDT will usually be formed by a particular specialist with expertise and interest in NETs and other adrenal tumours. This specialist is commonly an endocrinologist or oncologist.

A patient may see some of or all of the following people:

Gastroenterologist

Oncologist

Surgeon

Endocrinologist

Cardiologist

Radiology staff

Nuclear medicine physician

Dietician

Nurse Specialist

Palliative Care Team

Pain Team

General practitioner/ Practice Nurse

Counselling Staff

Various Technicians

Clinic Staff

Hospital Staff

Hospice Team

Patients can feel more confident in the knowledge that all aspects of their care have been discussed and that the best possible treatment plan will be formulated. A well coordinated and disciplined MDT is a very important aspect for care when striving to achieve the best quality of life and the best outcome for NET patients.

Being diagnosed with cancer can be a confusing and frightening time for you and your loved ones. Although your healthcare team will do their best to support you, medical appointments can be stressful and it is worthwhile to be reminded of ways to get the most out of each appointment.

Multidisciplinary teams (*continued*)

A few things to keep in mind:

- The most important member of the healthcare team is YOU
- YOU know more about YOU than anyone else does
- Your doctor can help you more if you are an active partner in your treatment
- If you are dissatisfied with the care that you receive, and discussion with your doctor does not resolve the situation, you can ask for a second opinion – how you feel about the care you receive is the most important thing.

Tips:

Bring a trusted friend or family member with you to the appointment if you can. If you are feeling anxious, you may not hear everything that's said, or ask all the questions you wanted to. It helps to have additional ears there to listen, and your guest may help to make sure your concerns are brought up.

Try to get into the practice of taking notes on how you are feeling, and take these notes into your appointments.

Write down a list of questions you want to ask. If you don't understand the answers to any of your questions, don't be embarrassed to ask for the answer to be repeated or rephrased. Make sure you know who your point of contact is and how to get hold of them in case you have questions later.

Deciding on a treatment strategy can be difficult so choosing to get a second opinion is quite common. Physicians are usually very open to this because they appreciate the fact that making an effective treatment choice can be very challenging.

If getting a second opinion through your specialist is not possible, you can get a referral from any of your physicians, including your general practitioner.

You have the right to know as much about your own prognosis as you wish and you have the right to know the overall treatment strategy, including what options are available to you if initial treatments are not successful in stabilising your disease.

You have the right to make decisions for yourself, even if the decision is against having medical treatment or to end medical treatment.

Further Support

NET Patient Foundation

From diagnosis and throughout treatment and beyond our services are here every step of the way. Below is an overview of all the services we offer to people living with and beyond NETs.

Helpline – 0800 434 6476

Our free, confidential helpline is for anyone who has questions about NETs (neuroendocrine tumours). Your call will be answered by one of our nurses or trained staff members with experience of NETs. Whatever your concern, you can be confident we will understand the issues you might be facing, and that the information you receive will be clear and up-to-date. We will also let you know where else you can go for further support. The helpline is open 10am-2pm Monday to Friday. We operate a call-back service for those who wish to leave a message out of hours.

Website – www.netpatientfoundation.org

We know how important it is to understand as much as possible about your NET. Our website is here round-the-clock giving you instant access to information when you need it. As well as clinical information, you will find real life experiences and access to the largest online NET community in the UK, so you can share your questions or concerns with other people in a similar situation.

Discussion forums

Through our discussion forums you can exchange tips on coping with the side effects of treatment, ask questions, share experiences and talk through concerns online. Our dedicated areas for popular topics should make it easy for you to find the information you're looking for. The discussion forums are easy-to-use. If you're feeling anxious or just need to hear from someone else who's been there, they offer a way to gain support and reassurance from others in a similar situation to you.

Information and support sessions

We run information and support sessions for people living with NETs. These meetings include talks from some of the country's top NET specialists plus invaluable Question & Answer sessions and an opportunity to meet other NET patients. For information about meetings in your area, please see our website or call our helpline.

NET Natter groups

These are informal groups which meet locally on a regular basis. To find out if there is a NET Natter group in your area, or if you'd like to set one up, please visit our website.

Further Support *(continued)*

Information resources

Our free information resources are for anyone affected by NETs. They are here to answer your questions, help you make informed decisions and ensure you know what to expect. All of our information is written and reviewed regularly by healthcare professionals and people affected by NETs, so you can trust the information is up-to-date, clear and accurate. You can order our publications by sending us an email or calling the helpline. All our publications can also be downloaded from our website.

Other organisations

NHS Choices

Information on clinical trials and treatment centres, including reviews and ratings.

www.nhs.uk

Cancer Research UK

Trusted information on all cancer types.

www.cancerresearchuk.org

Macmillan Cancer Support

Macmillan Cancer Support provides practical, medical, emotional and financial support to people living with cancer and their carers and families. Over the phone, its cancer support specialists can answer questions about cancer types and treatments, provide practical and financial support to help people live with cancer, and are there if someone just wants to talk.

Its website features expert, high-quality information on cancer types and treatments, emotional, financial and practical help, and an online community where people can share information and support. Macmillan also funds expert health and social care professionals such as nurses, doctors and benefits advisers.

www.macmillan.org.uk

General enquiries: 020 7840 7840

Helpline: 0808 808 0000

Textphone: 0808 808 0121 or Text Relay



You could make a difference to the lives of people living with neuroendocrine cancers...



PLEASE DONATE NOW



We Support:

- Vital clinical research in the UK
- Education for healthcare professionals

We Fund:

- The provision of supportive care for patients and carers living with neuroendocrine cancers
- Research into patient focused issues related to patient experience and equality of care

To find out more or to make a donation please call us on **01564 785577** or visit our website www.netpatientfoundation.org

Did you know?

- Over 50% of patients are diagnosed when the cancer has already spread
- Early diagnosis significantly improves outcomes for patients
- There are dedicated NET clinics around the UK. Find out where at www.netpatientfoundation.org

Why the moth?

We have chosen the moth as our logo to symbolise this 'camouflaged' condition. We aim to encourage medical professionals to consider an uncommon alternative when symptoms persist (i.e. a NET rather than IBS).

“When you see an insect beating its brightly patterned wings against your window, don't assume it's a butterfly. It could be a moth.”

About us

The Foundation has the following aims:

- To provide support, education and information to anyone affected by neuroendocrine cancers
- To advocate for neuroendocrine cancer patients so that they may achieve the best possible outcomes
- To encourage standardised care for all NET patients
- To provide community supportive care to patients and their carers or family members
- To raise awareness of NETs throughout the UK
- To raise funds for clinical research projects.

If you found this booklet useful and would like to make a donation to the NET Patient Foundation, please contact us.