

## Information for people affected by Goblet Cell Carcinomas

The NET Patient Foundation:  
supporting the neuroendocrine cancer community.

For further information and  
to make contact telephone  
**0800 434 6476** or visit our  
website: [www.netpatientfoundation.org](http://www.netpatientfoundation.org)

 [@netpatientfound](https://twitter.com/netpatientfound)

 [www.facebook.com/NetPatientFoundation](https://www.facebook.com/NetPatientFoundation)

Proud to be part of the International Neuroendocrine  
Cancer Community (INCA)

[www.netcancerday.org](http://www.netcancerday.org)

## 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.

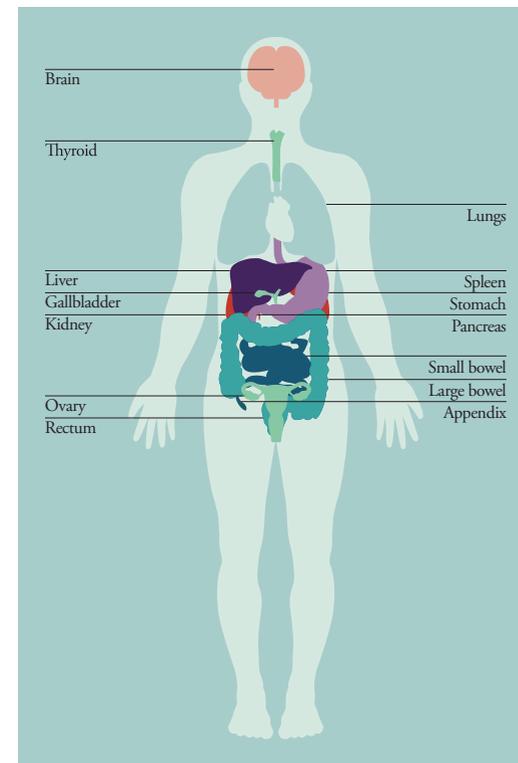
Like all cancers, NETs develop when the specialised cells undergo changes causing them to divide uncontrollably and grow into an abnormal tissue mass (tumour).

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](http://www.netpatientfoundation.org)

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



## Goblet Cell Carcinoma

These tumours start in the appendix and display features of both a neuroendocrine tumour (NET) and a more aggressive form of cancer called an adenocarcinoma.

One person in every 1,500 who have surgery for acute appendicitis is found to have the tumour.

Neuroendocrine tumours originate from cells called enterochromaffin cells that play a part in the hormonal system of the body, while adenocarcinomas are cancers that start in cells that line the inside of organs and secrete substances, in this case mucus.

Under a microscope, goblet cell tumours are found to contain features of both carcinoid tumours, the most common form of neuroendocrine tumour, and cells originating from the lining of the appendix, called epithelial cells.

The name goblet cell tumour refers to the shape of the cells as they appear under a microscope. The epithelial cells in this case are shaped like miniature wine goblets.

### What is known about these tumours?

Around a quarter of all carcinoid tumours originate in the appendix, although the vast majority start elsewhere in the intestinal system. Goblet cell tumours form a rare subgroup of carcinoid tumours found in the appendix. They were first described and recognised as a different condition as recently as 1969.

Some 20% of patients have a family history of colon cancer.

The tumours are most often found in people over 50, and usually at the time of an operation for another condition, or the removal of the appendix. They usually start in the tip or base of the appendix. In some cases they can cause a bowel obstruction as the tumour infiltrates into the small or large intestine from the appendix.

The current evidence suggests that these tumours behave in a more aggressive way than ordinary carcinoid tumours and are more likely to spread (metastasise) beyond the appendix into the abdomen. If the tumour does spread the most common area affected is the space inside the abdomen (the peritoneal cavity). In women the tumour can spread to the ovaries and the womb, and in some cases this type of cancer can be misdiagnosed as classic ovarian cancer.

### What are the symptoms?

Appendicitis, or complications from a burst appendix, are the most common first symptoms.

However, you may also suffer other problems such as diarrhoea, severe lower abdominal pain, or chronic vague abdominal pain.

## How are Goblet Cell Carcinomas diagnosed?

Like all NET patients you can expect to face numerous tests and scans that will provide your doctor with information about the disease, its spread and the rate of growth.

Some of the tests you might undergo include:

**Biopsy** This involves taking a piece of tissue from the suspect tumour and having it analysed in the laboratory by a specialist called a histopathologist. Biopsies are usually taken during medical tests (an endoscopy for example) or operations. The biopsy sample is sent to the laboratory and the cells are looked at very closely under the microscope to see if they are normal or cancer cells. NET cells look quite different to normal cells. Doctors can sometimes tell from biopsies where in the body a cancer has started.

Biopsies are very important in medicine. It is virtually impossible to diagnose some types of cancer any other way. Often, the only way to be sure of the diagnosis is to actually look for cancer cells under the microscope.

**Blood tests** You will be asked to have a fasting gut hormone blood test, and blood will also be collected for a range of other tests.

Doctors will be looking for certain NET markers, particularly chromogranin A and B, and for evidence of a rise in certain peptides and hormones in the blood.

For further information see the NET Patient Foundation Factsheet on *Fasting Gut Hormone Profile*.

### Other blood tests may include:

- Full blood count
- Kidney function test (urea and electrolytes)
- Liver function tests
- Thyroid function tests
- Pituitary hormone screen e.g. adrenocorticotrophic hormone (ACTH), prolactin, growth hormones and cortisol
- Serum calcium, parathyroid hormone levels (as a simple screening test for MEN-1 syndrome)
- CEA, AFP, CA19-9 , CA15-3 and CA125

## How are Goblet Cell Carcinomas diagnosed? *(continued)*

**CT scan** A computerised tomography (CT) scan provides a three dimensional picture of the inside of the body. It can be used to determine the position and size of neuroendocrine tumours and regular scans are useful to find out the rate of tumour growth.

**MRI scan** This 'whole body' magnetic resonance imaging (MRI) scan can help reveal where the tumours are positioned. It uses magnetism rather than X-rays to take pictures of the inside of the body. For some parts of the body and for some types of tumour, it can produce clearer results than a CT scan. For other situations, the CT scan is better. MRI is better than CT at showing how deeply the tumour has grown into body tissues. Your own doctor will know which is the best type of scan for you.

**PET scan** There are different types of Positron emission tomography (PET) scan that may be used for NET patients. PET scanners are used in many cancer types. The scan looks for the activity of the cell tissue and the growing speed of the cells.

Like the octreotide scan a special tracer (most commonly called FDG [18F]-fluorodeoxyglucose) is injected intravenously and the PET camera then produces a 3-dimensional image, highlighting any abnormal activity.

More recently a PET scan is used in combination with a CT camera which is a more advanced diagnostic tool.

A PET scan is not required for everyone but more likely to be used in more aggressive disease.

## How is it treated?

Patients should ideally be treated within a specialist multidisciplinary team (MDT).

Each patient will have an individualised treatment plan: there are a number of options available, depending on the type and location of the tumour, and the general well-being of the patient.

### **Surgery**

Because this type of tumour is more aggressive than the usual carcinoid tumours found in the appendix, you will also be asked to consider a bigger operation to remove part of your large intestine to reduce the risk of the disease spreading. This procedure is called a hemicolectomy.

If the spread is to the ovaries then you may be advised to have removal of one or both of your ovaries. Occasionally an operation may be recommended by some doctors even if there is no spread as a preventative measure.

In the case of a spread to the abdomen you may be advised to have a right hemicolectomy, or other 'debulking' surgery to remove as much tumour spread as possible.

Occasionally some people have to adjust the type of foods they eat after a hemicolectomy in order to keep their bowels regular.

Please talk to one of the nurses or doctors about this if you are in doubt.

### **Curative surgery Chemotherapy**

This may be an option for NET patients especially those with pancreatic, bronchial or high grade NETs. Not all NETs respond equally to chemotherapy, therefore careful selection of patients is imperative so as to maximise the chance of response and avoid unnecessary toxicity. Many chemotherapy treatments involve intravenous drugs, however there are now also oral chemotherapy agents and your NET doctor will discuss the most appropriate option with you. The histology of the tumour i.e. how it looks down the microscope after biopsy or operation, may help determine the type of treatment you receive.

Chemotherapy may sometimes be recommended after surgery (adjuvant therapy). You may be asked to be involved with clinical trials currently underway which are looking into the different combinations of chemotherapy agents that are most appropriate for different types of NET cancer.

**For further information about  
Chemotherapy see the NET Patient  
Foundation booklet on High-Grade  
Neuroendocrine Tumours**

## How is it treated? *(continued)*

### Clinical Trials

Clinical trials are medical research trials involving patients. They are carried out to try to find new and better treatments.

Carrying out clinical trials is the only sure way to find out if a new approach to cancer care is better than the standard treatments currently available.

You can find out more about current NET trials at: [www.netpatientfoundation.org/category/patient-resources/research](http://www.netpatientfoundation.org/category/patient-resources/research)

### Follow up

The follow-up intervals are highly variable depending on the clinical situation, functionality (whether any syndrome is present), stage, grade and time since diagnosis. Intervals may vary, but generally follow-up should be tailored to individual needs and is usually every 3-6 months initially.

## 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](http://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](http://www.nhs.uk)

#### Cancer Research UK

Trusted information on all cancer types.

[www.cancerresearchuk.org](http://www.cancerresearchuk.org)

#### Cure Goblet Cell Carcinoid Cancer

[www.cgccc.co.uk](http://www.cgccc.co.uk)

Facebook page:

[www.facebook.com/](http://www.facebook.com/CureGobletCellCarcinoidCancer)

[CureGobletCellCarcinoidCancer](https://www.facebook.com/CureGobletCellCarcinoidCancer)

#### 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](http://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](http://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](http://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.**