

## Information for people affected by Gastrinomas

**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)

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**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.

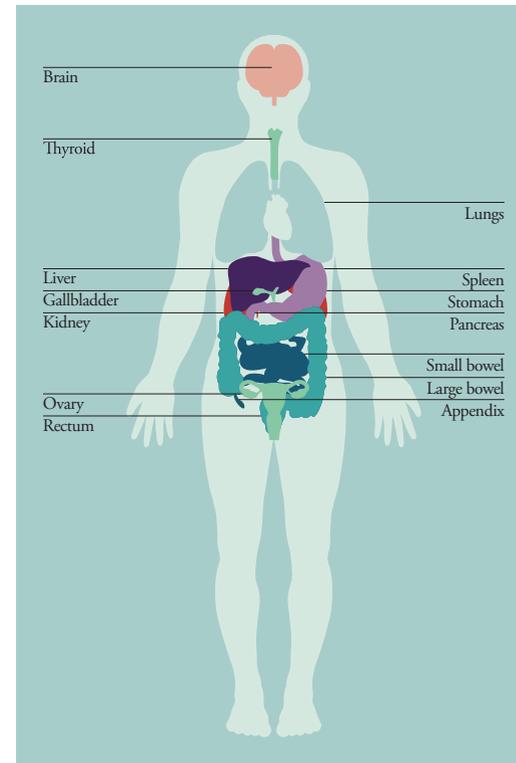
Like all cancers, NETs develop when the specialised cells undergo changes causing them to divide uncontrollably and grow into an abnormal tissue mass (tumour).

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)



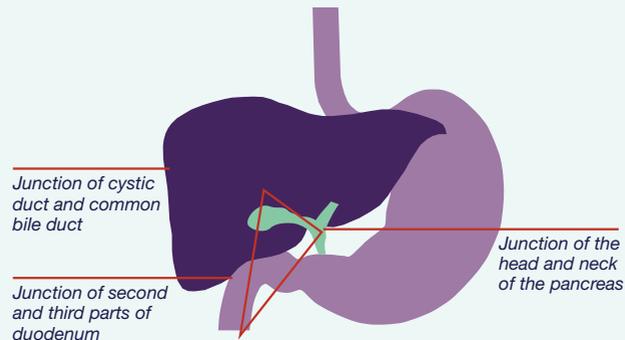
# Gastrinomas

Gastrinomas are relatively rare neuroendocrine tumours that were first described in 1955. They are mostly found in the duodenum, the first part of the small bowel, and the pancreas.

However they are the most common of all 'functional' pancreatic neuroendocrine tumours – in other words tumours that not only grow but also secrete large quantities of hormones causing side effects and symptoms that collectively make up a recognised syndrome.

The tumours develop from cells that produce gastrin, a hormone that regulates acid secretion in the stomach, an important part of the digestive process. When the cells become malignant and develop into tumours, the amount of gastrin rises dramatically leading to some severe abdominal symptoms and pain. They are the cause of 1% of peptic ulcer disease.

## Tumours occur in the so called gastrinoma triangle



## What is known about these tumours?

In 25% of cases there is an association with a genetic, inherited syndrome called Multiple Endocrine Neoplasia Type 1 (MEN1).

Like other neuroendocrine tumours (cancers that develop in cells that play a part in the hormonal system) gastrinomas are most usually slow growing, although in a quarter of cases can be aggressive.

The tumours are often very small; perhaps less than 1cm, but even these tiny tumours can cause severe symptoms. The key to a successful diagnosis is for a doctor to suspect the condition, as symptoms can be very general and may be mistaken for straightforward peptic ulcer disease. Gastrinomas can be found singly, or as multiple, small tumours. Tumours in the pancreas may commonly measure 3 or 4cm while those in the duodenum can be less than 1cm. The usual age for diagnosis is the 50s or 60s, although patients with MEN1 are often diagnosed earlier.

Between 5 and 15% of patients with advanced metastatic disease may develop a second hormonal condition known as Cushing's Disease, which is caused by the tumours secreting the adrenocorticotrophic hormone (ACTH) that in turn stimulates the secretion of cortisol (often known as the 'stress hormone' as it is involved in the response to stress). An over production of cortisol can cause an array of symptoms. For further information about Cushing's Disease speak to your NET specialist.

## What are the symptoms of Gastrinomas?

The main symptoms are:

- Abdominal pain
- Acid reflux
- Diarrhoea

Collectively they are a set of symptoms known as the Zollinger-Ellison syndrome after the scientists who first recognised them. The symptoms are caused by over production of the hormone gastrin by the tumours. This over production causes a marked increase in the secretion of gastric acid in the stomach. The excess acid triggers ulcers in the stomach or duodenum and acid reflux and these can cause pain, nausea, vomiting and bleeding.

## How are Gastrinomas 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.

Peptic ulcers that are recurrent and/or resistant to medical treatment, or the collection of symptoms described above should lead your specialist to consider further tests for gastrinoma.

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)

### Secretin stimulation test

Patients with gastrinomas are likely to have very high levels of gastrin (perhaps ten times the normal level) circulating in the blood stream.

However, if gastrin levels are not very high, but a gastrinoma is still suspected, there is a further test that can be performed.

This is called a secretin stimulation test and involves an injection of the hormone secretin via a cannula into the blood stream to provoke a rise in blood gastrin levels. The test will almost always produce a positive response in patients with gastrinomas but seldom in people without them. The infusion that you will be given will last about 180 minutes and the levels of gastrin will be checked at regular intervals.

**Endoscopy** This is a way of examining your gastrointestinal tract using a flexible fibre optic tube called an endoscope. The tube can either be inserted down the back of the throat, a procedure called a gastroscopy, or into the colon via the rectum (back passage), a procedure known as a colonoscopy. You will be offered sedation before either of the procedures, and the doctor may remove samples of any suspect areas of tissue for analysis.

**Wireless Capsule Endoscopy** This involves swallowing a small capsule (the size of a large vitamin pill), which contains a colour camera, battery, light source and transmitter.

**Capsule Endoscopy** involves swallowing a small (the size of the large vitamin pill) capsule, which contains a colour camera, battery, light source and transmitter. The camera takes two pictures every second for eight hours, transmitting images to a data recorder about the size of a portable CD player that patients wear around the waist.

**Endoscopic ultrasound** This is usually carried out under sedation and involves looking at the gastrointestinal tract with a flexible camera. The test can help pick up small tumours that might not be clearly visible on a scan.

**Octreotide scan** This is a useful diagnostic test that can help reveal the site of tumours. Some NETs have special receptors on their surfaces called somatostatin receptors. Octreotide is a somatostatin analogue, a substance that copies or mimics the action of somatostatin. When octreotide is combined with a mildly radioactive agent and then injected via a vein in the arm, it sticks to somatostatin receptors on the tumour surface and the tumours light up on the screen as radioactive 'hot spots'.

**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.

## How are Gastrinomas diagnosed? (continued)

**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.

Another type of PET scan is the GA-DOTA-octreotate PET scan, which has been shown

to have a higher sensitivity (can see more) for NETs compared to the octreotide scan. Not only is this a more sensitive scan but it is also much quicker than the octreotide scan and results are available in a shorter time. This is new technology and an advancement in NET scanning but at present there are only a few available in the UK.

**Bone scan** You will be given a small injection of radioactive tracer which over time will be absorbed into your skeleton. It takes about three hours for this to occur. Pictures of your skeleton are taken in order to help your doctor determine whether there are any problems associated with it. This procedure takes about ten minutes, and you will be asked to return for the second set of pictures three hours later. For this second set of pictures you will again be asked to lie or sit in front of the gamma camera. The scan will take approximately 30-45 minutes.

**Ultrasound scan** Ultrasound imaging, also called ultrasound scanning or sonography, involves exposing part of the body to high-frequency sound waves to produce pictures of the inside of the body. Ultrasound exams do not use ionizing radiation (as used in X-rays). Because ultrasound images are captured in real-time, they can show the structure and movement of the body's internal organs, as well as blood flowing through blood vessels.

## Treatment for Gastrinomas

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

If the tumour is contained in one area (localised), or if there has been only limited spread, surgery is usually the first choice of treatment. If it is possible to remove the tumour completely, no other treatment may be necessary.

If the tumour has spread to other parts of the body (metastatic), surgery may still be possible to remove the part of the tumour that is producing too many hormones. This is often referred to as tumour debulking.

### Curative surgery

This is when the cancer has not spread outside the organ or area where it first started. If the tumour can be removed whole and intact with a surrounding margin of clear, healthy tissue then the surgery is potentially curative and no other treatment may be necessary. A follow-up plan will need to be put into place after surgery.

### Palliative surgery

When the tumour or tumours have already spread or become too large to remove completely, then surgery may be considered. If the tumours are causing symptoms by pressing on other organs or by releasing hormones then surgery may be performed to reduce the symptoms by removing or bypassing the part of the disease that is causing the problems.

### Pre-emptive surgery

This is when an area of tumour is critically placed close to an important structure such as a blood vessel or the bile duct or bowel. Even if the disease cannot be cured, removing the tumour may prevent problems in the future by removing disease from key areas before the vital structure has been damaged or blocked.

### Cyto-reductive surgery

All of the many medical treatments for NET cancers aim to reduce symptoms and prolong survival by cyto reduction.

This means reducing the number of living tumour cells inside the patient. Operations can also be used to achieve this when the disease has already spread, by surgically removing bulky areas of tumour to reduce the total number of cancer cells inside the patient. Cyto-reduction often involves other treatments used in combination with surgery.

## Treatment for Gastrinomas (*continued*)

### Proton pump inhibitors (PPIs)

For example omeprazole and lansoprazole. These drugs are potent inhibitors of gastric acid and are the first line medical treatment of choice. Much higher doses than given in standard treatment are often needed to control symptoms.

### H2 Antagonists

High doses of drugs known as H2 receptor antagonists, such as ranitidine (Zantac) may also reduce gastric acid secretion and relieve symptoms. These treatments work by inhibiting the action of histamine.

### Somatostatin Analogues

Daily or monthly injections of somatostatin analogues are available to control some of the unpleasant symptoms caused by the tumours. Somatostatin analogues are synthetic versions of somatostatin, a naturally occurring hormone produced in the brain and digestive tract that inhibits the release of several other hormones and chemicals from our internal organs.

Injections of these analogues can stop the overproduction of hormones that cause symptoms such as flushing and diarrhoea.

Recent clinical studies have shown that somatostatin analogues can also be used to help stabilise (stop the growth for a period of time) some low grade NETs, and further studies are ongoing to see how well these drugs work on different types of NET.

### Targeted Peptide Receptor Radionuclide Therapy PRRT

These treatments are sometimes referred to as 'magic bullet' therapies. The treatments are based on using different radiolabelled peptides which target receptors on the tumour surface. In PRRT for neuroendocrine tumours, the radionuclides commonly used are indium-111, lutetium-177 and yttrium-90. The basis for which treatment is suitable for which patient is the diagnostic tracer imaging. To decide whether this therapy is suitable, you will be asked to take an Octreotide, PET or MIBG scan as described in the diagnosis section. The results of these scans will indicate whether or not you have the correct receptors present and whether this sort of therapy would be an option.

### SIRT

SIRT stands for Selective Internal Radiation Therapy. It is a new way of using radiotherapy to treat liver metastases which cannot be removed with surgery.

The doctor inserts a thin tube called a catheter into the hepatic artery. This is the main artery which supplies blood to the liver. They then send tiny beads called microspheres down the catheter and these get stuck in the small blood vessels around the tumour. The microspheres contain a radioactive substance which give a dose of radiation to the tumour. The microspheres are also called SIR-spheres and the radioactive substance is called yttrium 90.

### Embolisation

If the tumour has spread to the liver, you may be offered hepatic artery embolisation (HAE). In this procedure, a catheter is placed in the groin and then threaded up to the hepatic artery that supplies blood to the tumours in the liver. Tiny particles called embospheres (or microspheres) are injected through the catheter into the artery.

These particles swell and block the blood supply to the tumour, which can cause the tumour to shrink or even die. This treatment can also be combined with systemic treatments in some patients where the tumour has spread. It is a procedure that would be carried out by a specialist called an interventional radiologist. The patient would be sedated for the treatment.

Sometimes this embolisation process is combined with chemotherapy (called HACE (Hepatic Artery Chemoembolisation) or TACE (Transcatheter Arterial Chemoembolisation) or radiotherapy (RMT or SIRT).

## Treatment for Gastrinomas (*continued*)

### Radiofrequency Ablation (RFA)

This may be used when a patient has relatively few secondary tumours (metastases) in the liver. A needle is inserted into the centre of the tumour and a current is applied to generate heat which kills the tumour.

### 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.

### Sutent (Sunitinib)

Sutent is a medication that comes in capsule form. It is mainly used in patients with pancreatic neuroendocrine tumours.

It works mainly by blocking a process called angiogenesis. Angiogenesis is the process of making new blood vessels.

Tumours need a good blood supply to grow and Sutent helps stop that process. The drug comes under an umbrella group of drugs known as tyrosine kinase inhibitors.

### Affinitor (Everolimus)

Affinitor is another medication for patients with pancreatic neuroendocrine tumours.

It also comes in a capsule form and is a type of drug that interferes with the process of new cancer growth. The drug comes under an umbrella group of drugs known as mTOR inhibitors.

Clinical trials are running to look at the effectiveness of this medication in other NET types either alone or in combination with other NET medications.

### 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/)

### No treatment

No treatment or watchful waiting may be suitable for you if your NET is not currently causing you any symptoms or problems and the tumour(s) are stable. You may also have other health conditions that would make the NET treatment inadvisable.

### 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.

### 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.

### 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)

#### A.M.E.N.D

Association for Multiple Endocrine Neoplasia Disorders

[www.amend.org.uk](http://www.amend.org.uk)

#### 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

### 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.**