

## Information for people affected by Lung NETs

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

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

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

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

© NET Patient Foundation May 2015

## 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).

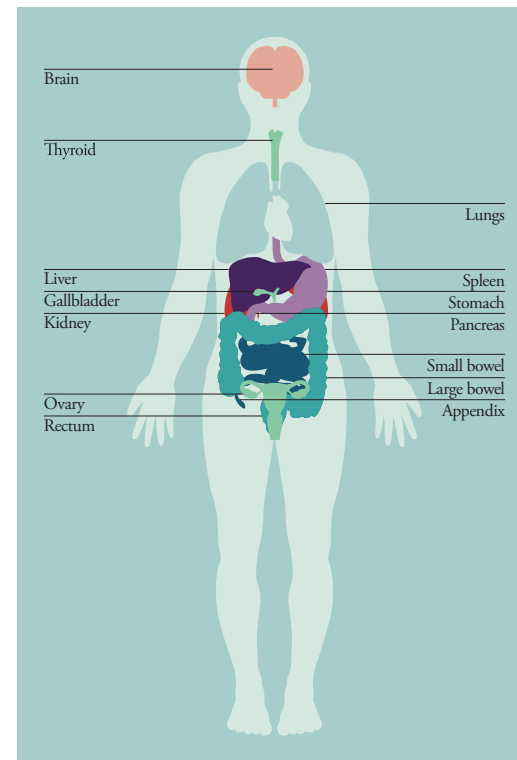
Neuroendocrine tumours can be benign or malignant. Back in 1907, when neuroendocrine tumours were first classified, they were named 'carcinoid' (meaning 'cancer-like') as they seemed to grow slowly and were therefore not thought to be truly cancerous.

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 with the primary location of the tumour added before the word NET, for example Lung NET, Bowel NET, 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)



This booklet will focus on neuroendocrine tumours which arise in the lungs.

## What is a Lung NET?

Lung NETs are an uncommon type of tumour that start in the lungs. These tumours can occur at any age with the average onset in the 4th to 6th decade of life. They tend to grow slower than other types of lung cancers. They are made up of special kinds of cells called neuroendocrine cells. Other terms you may come across are pulmonary NET, bronchial NET or carcinoid or lung NET or carcinoid. 'Pulmonary' and 'bronchial' are both medical terms relating to the lungs.

To understand lung NETs it helps to know something about the normal structure and function of the lungs, as well as the neuroendocrine system.

The lungs are two sponge-like organs in your chest cavity. Your right lung has three sections, called lobes. The left lung has two lobes. It is smaller because the heart takes up more room on that side of the body.

The lungs bring air in and out, taking in oxygen and getting rid of carbon dioxide.

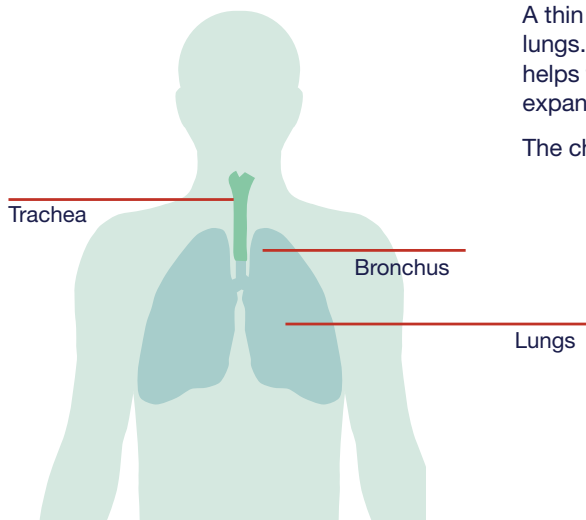
When you breathe in, air enters through your mouth and nose and goes into your lungs through the trachea (windpipe).

The trachea divides into tubes called the bronchi (singular, bronchus), which divide into smaller branches called the bronchioles.

At the end of the bronchioles are tiny air sacs known as alveoli.

A thin lining called the pleura surrounds the lungs. The pleura protects your lungs and helps them slide back and forth as they expand and contract during breathing.

The chest cavity is called the pleural cavity.



## Types of Lung NETs

There has been an increase in the amount of people living with a lung NET over the last 30 years of around 6%.

This is likely to increase with improved awareness and diagnostic tests.

**There are three main types of lung NET:**

- typical carcinoid tumour (TC)
- atypical carcinoid tumour (AC)
- large cell neuroendocrine carcinoma (LCNEC)

Both the Typical and Atypical lung NETs have unique traits that separate them clearly from the poorly differentiated NETs like the LCNECs (Large Cell Neuroendocrine Carcinomas)

Hormone related symptoms are not common for lung NETs, but a small number of patients experience "carcinoid syndrome" – flushing, diarrhoea, palpitations and wheezing. *See page 19.*

Up to 5% of patients with Multiple Endocrine Neoplasia type 1 will develop lung NETs.

### Typical lung NETs (TCs)

These are well differentiated NETs and are low grade malignant tumours. These grow slowly and only rarely spread beyond the lungs. About 9 out of 10 lung carcinoids are typical carcinoids.

There is only a small association with smoking in around 20% of patients.

### Atypical lung NETs (ACs)

These are also well differentiated NETs but are intermediate grade malignant tumours which mean that they may develop or grow faster than the typical lung NET. They are much less common than typical lung NETs.

There is about a 40% association with smoking in this type of NET.

Typical and atypical lung NETs account for around a third of all NETs and around 1-2 % of all lung cancers.

### Large cell neuroendocrine carcinoma

Large cell neuroendocrine carcinoma (LCNEC) is a rare cancer that, except for the size of the cells forming the cancer, is very similar to small cell lung cancer in its prognosis (outlook) and in how patients are treated.

### Diffuse idiopathic pulmonary neuroendocrine cell hyperplasia (DIPNECH)

This is a rare and not well characterised condition in which multiple small tumours, sometimes termed tumourlets, appear as nodules on scans.

They are generally very slow growing and have an excellent prognosis.

# Staging, grading and differentiation

## Why staging is important

Staging is important because it helps your treatment team to know which treatments you need. If a cancer is just in one place, then a local treatment such as surgery or radiotherapy could be enough to get rid of it completely. A local treatment treats only one area of the body.

If a cancer has spread, then local treatment alone will not be enough. You will need a treatment that circulates throughout the whole body. These are called systemic treatments. Chemotherapy, hormone therapy and biological therapies are systemic treatments because they circulate in the bloodstream.

Sometimes doctors aren't sure if a cancer has spread to another part of the body or not. They look at the lymph nodes near to the cancer. If there are cancer cells in these nodes, it is a sign that the cancer has begun to spread. Cancer doctors call this having positive lymph nodes. The cells have broken away from the original cancer and got trapped in the lymph nodes. But it is not always possible to tell if they have gone anywhere else.

If cancer cells are found in the lymph nodes, doctors usually suggest adjuvant treatment. This means treatment alongside the treatment for the main primary tumour (chemotherapy after surgery, for example). The aim is to kill any cancer cells that have broken away from the primary tumour.

## Types of staging systems

There are two main types of staging systems for cancer. These are the TNM system and the number system.

### The systems mean that:

- Doctors have a common language to describe the size and spread of cancers
- Treatment results can be accurately compared between research studies
- Guidelines for treatment can be standardised between different treatment hospitals and clinics

## The TNM staging system

TNM stands for Tumour, Node, Metastasis. This system describes the size of the initial cancer (the primary tumour), whether the cancer has spread to the lymph nodes, and whether it has spread to a different part of the body (metastasised). The system uses numbers to describe the cancer.

- T refers to the size of the cancer and how far it has spread into nearby tissue – it can be 1, 2, 3 or 4, with 1 being small and 4 large
- N refers to whether the cancer has spread to the lymph nodes – it can be between 0 (no lymph nodes containing cancer cells) and 3 (lots of lymph nodes containing cancer cells)
- M refers to whether the cancer has spread to another part of the body – it can either be 0 (the cancer hasn't spread) or 1 (the cancer has spread)

## Number staging systems

Number staging systems usually use the TNM system to divide cancers into stages. Most types of cancer have 4 stages, numbered from 1 to 4. Often doctors write the stage down in Roman numerals. So you may see stage 4 written down as stage IV.

### Here is a brief summary of what the stages mean for most types of cancer.

- **Stage 1** usually means that a cancer is relatively small and contained within the organ it started in.
- **Stage 2** usually means the cancer has not started to spread into surrounding tissue but the tumour is larger than in stage 1. Sometimes stage 2 means that cancer cells have spread into lymph nodes close to the tumour. This depends on the particular type of cancer.
- **Stage 3** usually means the cancer is larger. It may have started to spread into surrounding tissues and there are cancer cells in the lymph nodes in the area.
- **Stage 4** means the cancer has spread from where it started to another body organ. This is also called secondary or metastatic cancer.

## What grading is

You may hear your doctor talk about the grade of your cancer. Tumour grade describes a tumour in terms of how abnormal the tumour cells are compared to normal cells. It also describes how abnormal the tissues look under a microscope.

The grade gives your doctor some idea of how the cancer might behave. A low grade cancer is likely to grow more slowly and be less likely to spread than a high grade one. Doctors can't be certain exactly how the cells will behave. But the grade is a useful indicator.

Tumour grade is sometimes taken into account as part of cancer staging systems. The stage of a cancer describes how big the cancer is and whether it has spread or not.

## Common grading systems

Some types of cancer have their own grading systems but generally there are 3 grades.

### They are described as:

- **Grade 1** – The cancer cells look very similar to normal cells and are growing slowly
- **Grade 2** – The cells look unlike normal cells and are growing more quickly than normal
- **Grade 3** – The cancer cells look very abnormal and are growing quickly

### Some systems have more than 3 grades.

- GX means that the grade can't be assessed. It is also called undetermined grade.

## Differentiation

Another way of describing the cells is by how differentiated they are. Differentiation refers to how well developed the tumour cells are and how they are organised in the tumour tissue. If the cells and tissue structures are very similar to normal the tumour is called well differentiated. These tumours tend to grow and spread slowly.

In poorly differentiated or undifferentiated tumours the cells look very abnormal and the cells are not arranged in the usual way. So the normal structures and tissue patterns are missing. These tumours may be more likely to spread into surrounding tissues or to other parts of the body.

*Information compiled with the help of Cancer Research UK.*

## What are the symptoms of Lung NETs?

Some people with lung NETs may have no symptoms.

### Symptoms may include:

- airway obstruction – sometimes presenting with a hoarse voice
- cough
- recurrent chest infections
- wheezing
- pneumonia
- pleurisy
- difficulty with breathing
- weakness
- nausea
- weight loss
- night sweats
- neuralgia
- Cushing's syndrome. Up to 40% of patients with ectopic Cushing's are found to have a lung NET.

## How are Lung NETs 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 (a bronchoscopy 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

The blood tests you may be required to undergo are often related to the type of bronchial NET you have. If it is a typical or atypical bronchial NET then it is possible you will have some of the following 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, as well as checking how well your kidneys and liver are functioning.

#### Baseline blood tests may include:

- Kidney function test
- Calcium
- Glucose
- Chromogranin A

#### If Cushing Syndrome is suspected then you will have:

- serum cortisol
- 24 hour urine free cortisol
- adrenocorticotrophic hormone (ACTH)

#### In very rare cases and only where clinically indicated you may have:

- serum growth hormone (GH)
- insulin-like growth factor (IGF-1)
- growth hormone-releasing hormone (GHRH)

You may also be asked to give an extra blood sample for use in research studies. You should always be informed of this and asked to sign a consent form.

# How are Lung NETs diagnosed?

*(continued)*

## 24 Hour urine 5-hydroxyindoleacetic acid (5HIAA)

You will only be asked to do a urine test if you have carcinoid syndrome. When serotonin breaks down in the body, it is converted first to 5-HT and then to 5HIAA, which is excreted into the urine. A urine sample is collected, and the level of 5HIAA in the urine is measured. Higher than normal levels of serotonin produced by NET patients show up as raised levels of 5HIAA in their urine. You may be asked to do a urine collection over a 24 hour period so that your 5HIAA levels can be checked.

You will be asked to avoid certain foods prior to and during the test including chocolate, olives, bananas, pineapple and its juice, all tomato products, plums, aubergine, avocado, kiwi fruit, walnuts, brazil nuts, cashew nuts, tea, coffee and alcohol. You will also be asked to avoid certain cough, cold and flu remedies 3 to 7 days prior to the test. This is because they contain substances that might artificially raise your serotonin levels and so give a false test result.

## Chest X-ray

More than 40% of lung NETs may be seen on a standard chest x-ray.

## 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.

A contrast CT scan is considered to be the gold standard for lung NETs.

## Functional respiratory tests

These should always be performed to assess surgical risk and to check for any other respiratory conditions.

## Echocardiography

This is a test that uses sound waves to build up a picture of your heart. It should be used when you are diagnosed, and during follow-up, to check for carcinoid heart disease. *See page 20.*

This test should also be performed before surgery.

## Octreotide scan

This test may be used for patients with typical and atypical lung NETs. It is not useful for patients with LCNEC. It is most useful if looking for bone metastases.

It 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'. This investigation is done on a machine called a SPECT CT.

## SPECT/CT

This is a useful combination of an octreotide scan and CT scan. It is likely to see nearly 80% of primary tumours.

## 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. Your own doctor will know which is the best type of scan for you.

This scan may be used to detect tumours that may have spread to other parts of the body, for example, the bones.

## 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 regular PET scan is not required for everyone but may be used if you have a high grade tumour.

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.



## Treatments

You should ideally be treated within a specialist multidisciplinary team (MDT). You will have an individualised treatment plan: there are a number of options available, depending on the type and location of the tumour, and your general well-being.

### Surgery

Surgical removal is the treatment of choice for lung NETs. The aim is to remove the tumour and to preserve as much lung tissue as possible.

The surgical approach is dependent on the size, location and tissue type.

### Surgery for localised disease

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.

### Peripheral lung tumours

If the tumour is towards the edges of the lungs, the surgery team may attempt to remove it completely (this may be called a 'lobectomy' or 'segmentectomy').

### Central airway tumours

For patients with central airway tumours, there are highly specialised procedures that aim to remove all of the tumour and as little of the lung as possible. These are called 'bronchial sleeve resection' or 'sleeve lobectomy'. If necessary, patients should be referred to a centre of excellence if this expertise is not available locally.

### Local Bronchoscopic Therapy

In some cases non-invasive procedures called laser bronchoscopy or cryotherapy may be used to destroy the tumour cells.

## Treatments for 'carcinoid syndrome', disease that has spread or cannot be removed with surgery

### External radiotherapy

The radiation is aimed at your body from a machine. You have the treatment in the hospital radiotherapy department. Your treatment plan will depend on what type and stage of cancer you have.

### Internal radiotherapy

This is also called brachytherapy or endobronchial therapy. You have the treatment during a bronchoscopy. The radiation source is inside a narrow tube that the doctor puts inside your airway for a few minutes.

Doctors use this type of treatment to shrink a tumour that is blocking or pressing on your airway and making breathing difficult for you.

### Somatostatin Analogues

Around 30% of lung NETs will cause hormone related symptoms or 'syndromes' (see 'Carcinoid Syndrome' on page 19).

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.

There is good evidence now demonstrating that these injections also slow down the rate of growth of tumours. Your doctor may prescribe Somatostatin analogues as a first line treatment if your tumour cannot be resected but you have a low grade tumour and you have an octreotide positive scan.

### Chemotherapy

Systemic chemotherapy may be considered if the tumour cannot be removed with surgery. There are a number of different chemotherapy treatments and trials are in progress looking at trying to find which work best for lung NET patients and how well they work with other treatments. Your specialist will discuss options with you.

### These options may be:

- Temozolomide alone
- Combination of cisplatin and etoposide if the tumour is high grade.

## Treatments *(continued)*

### 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 or Gallium 68 PET 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. This therapy is currently of very limited availability in the UK.

### 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. You 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).

### Radiofrequency Ablation (RFA)

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

### Sutent (Sunitinib)

Sutent is a medication that comes in capsule form. 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 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 lung NETs 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.

There have not been many clinical trials in bronchial NETs previously. This is changing, however, and a number of clinical trials have been running.

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. This is especially true for patients with DIPNECH for whom annual monitoring is the only requirement.

### Follow-up

After curative surgery, patients with TC and AC should be followed at least yearly to begin with. They will need continued, long term follow up to check that the disease has not recurred. A CT patient should expect to be scanned after 3 months, 6 months and then every 12 months for 2 years. After that, you should have a chest x-ray, blood tests and a CT scan every three years.

AC patients may expect to be monitored more closely.

Your follow up programme will be individualised to you.



## Multidisciplinary teams

The care of NET cancers 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 NET cancers and other adrenal tumours.

This specialist is commonly an endocrinologist or oncologist.

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

- Respiratory physician
- Thoracic surgeon
- Oncologist
- Surgeon
- Endocrinologist
- Gastroenterologist
- 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 cancer 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.

## 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 or peptide.

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'.

Only 6-8% of patients with lung NETs will develop carcinoid syndrome, 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 Heart Disease

The hormones released by the tumours into the bloodstream can affect the heart by causing deposits to build up on both the cardiac valves and the muscle. These can eventually impair the function of the heart valves and cause the valves to become leaky, which can cause symptoms such as breathlessness and swollen ankles. Carcinoid heart disease is detected by an echocardiogram.



For further information see the NET Patient Foundation booklet on *Carcinoid Heart Disease*.

## 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 NET cancer.

### Helpline – 0800 434 6476

Our free, confidential helpline is for anyone who has questions about NET cancers (neuroendocrine tumours).

Your call will be answered by one of our nurses or trained staff members with experience of NET cancer.

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 cancer.

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 cancer 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 NET cancer. These meetings include talks from some of the country's top NET specialists plus invaluable Question and 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 NET cancer. 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 NET cancer, 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)

#### Lovable Lungnoids

A facebook group for people affected by bronchial NETs.

[www.facebook.com/groups/lovablelungnoids](https://www.facebook.com/groups/lovablelungnoids)

### Did you know?

- 50% of patients are diagnosed when the cancer has already spread
- Early diagnosis significantly improves outcomes for patients
- Help us to make a difference. Donate now 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 cancer patients
- To provide community supportive care to patients and their carers or family members
- To raise awareness of NET cancers 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.**