

November 10th is Worldwide NET Cancer Awareness Day

Raising awareness of NETs among decision makers, health professionals and the general public.

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

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Your guide to Neuroendocrine Tumours

What are Neuroendocrine Tumours?

How does it feel to be diagnosed with a Neuroendocrine tumour?

Fearful, concerned and perhaps a bit confused?



One person, recently diagnosed with a NET stated: *"Suddenly I found myself facing a whole collection of scans and blood tests, many of which I had never heard of. It was so frightening and I had nothing to read that seemed to relate to what I was experiencing"*.

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.

Around 3,000 new cases are diagnosed every year in the UK, but it is thought that a larger number of

people have a NET, but remain undiagnosed.

This is because NETs can be difficult to diagnose as the symptoms may be vague and attributed to more common problems such as irritable bowel syndrome (IBS), Crohn's disease, peptic ulcer disease, gastritis, asthma, blood glucose problems, menopause and blood pressure problems.

NETs have a wide spectrum of clinical behaviours meaning that they can behave in different ways.

Some can behave in quite an indolent way behaving like 'cancer in slow motion' whilst some can behave more aggressively acting more like some of the more common cancers.

However the fact that some of the lower grade NETs can have an associated syndrome for example 'carcinoid syndrome', can make them quite difficult to manage.

NETs can affect people of all ages and often affect people during their working lives, and are often misdiagnosed leading to patients being diagnosed once the NET has had a chance to spread (metastasise).

Most NETs occur without any known genetic link at present, but there are cases where NETs are present as part of a familial endocrine syndrome for example, Multiple Endocrine Neoplasia Type 1 (MEN1), neurofibromatosis type 1 or Von Hippel Lindau.

There is ongoing research into the genetics of neuroendocrine tumours.

Types of Neuroendocrine Tumours

There are a number of different types of NET.

They all have a slightly different way of presenting themselves, both in terms of symptoms and how they look under a microscope.

Examples of NETs

- Gastrointestinal NETs: gastric, duodenal, pancreatic, small intestine, appendiceal, colon, rectal,
- Lung NETs
Thymic NET
- Medullary Thyroid Cancer (MTC)
- Pancreatic NETs (non-functioning)
- Pancreatic NETs (functioning):
Gastrinoma, VIPoma, Insulinoma, Somatostatinoma, Glucagonoma
- MEN (Multiple Endocrine Neoplasias)
- Pheochromocytoma and Paraganglioma
- Goblet Cell Carcinoma
Merkel Cell NET
NETs of unknown primary

The place where the NET first appears is called the 'primary' site.

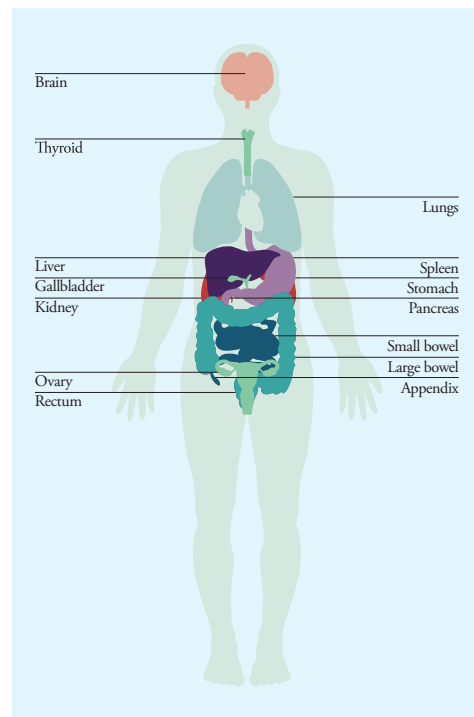
However, the NET may spread and be found in other parts of the body e.g. the liver, and if this occurs your doctor may refer to these as a 'secondary' tumour or metastases.

Example of where NETs may occur

Some NETs produce abnormally large amounts of hormones.

These NETs have a related syndrome which means that the hormones secreted cause noticeable symptoms like flushing, diarrhoea, cramps, wheezing, heart valve damage and skin changes.

An example of a syndrome is 'carcinoid syndrome'



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.

The group of tumours that arise in the pancreas can be classified into two different groups; functioning and non-functioning.

The functioning group will produce a number of clinical syndromes that are related to where they originate, for example, an insulinoma will over-secrete insulin and gastrinomas are gastrin-secreting tumours.

The non-functioning group which accounts for around 30-40% of pancreatic tumours, may secrete certain hormones and peptides like other NETs, but the release of these chemicals does not cause an identifiable 'syndrome' or collection of symptoms.

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

For prophylaxis:

IV Octreotide 600 mcg in 60ml Normal Saline syringe (infusion rate: 5ml/h) started 12h pre-op and continued for 24-48h post-op.

For unexpected carcinoid crisis:

Bolus IV Octreotide 100-500mcg, followed by the infusion above.

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Classification, Stage and Grade

The World Health Organisation (WHO) classification scheme puts neuroendocrine tumours into three categories:

- Well-differentiated neuroendocrine tumours, further subdivided into tumours with benign and those with uncertain behaviours
- Well-differentiated (low grade) neuroendocrine tumours with low-grade malignant behaviour
- Poorly-differentiated (high grade) neuroendocrine carcinomas

Stage: the extent of disease

Stage is a measure of how far a tumour has spread. Health organisations around the world use slightly different classification systems for NET staging. The WHO classification uses four stages (I through IV), with a higher number indicating more widespread disease.

The following example shows the stage descriptions for a NET of the small intestine:

- **Stage I** tumour is less than 1 cm in size and has not spread to the lymph nodes or other parts of the body.
- **Stage II** tumour is greater than 1 cm in size and has started to spread beyond the original location, but has not spread to the lymph nodes or other parts of the body.
- **Stage III** is any size tumour that has spread to nearby areas of the body and also to one lymph node.
- **Stage IV** is any size tumour that has spread to one or more lymph nodes and has also spread to other, more distant areas of the body (for example, to the liver).

Please note that the stage descriptions may also vary depending on the type of NET you have.

For example, stages for a NET of the lung can differ from stages for a NET of the pancreas.

Grade

The "grade" of a NET is an estimation of how fast the cells are dividing to form new cells.

The grade of a NET is related to its differentiation.

There are several systems for grading NETs, including a new WHO system introduced in 2010, which is shown here.

Grade	Differentiation
NET Grade 1	Well-differentiated tumour with a low number of cells actively dividing
NET Grade 2	Well-differentiated tumour, but with a higher number of cells actively dividing
Neuroendocrine carcinoma Grade 3 (NEC)	Poorly differentiated, malignant carcinoma (most aggressive form of NET)

By using information about stage, differentiation, and grade together, your doctor can decide how best to manage your NET.

Although your doctor may not talk about grade and differentiation with you, he or she may talk about how serious or aggressive your NET is, which refers to how quickly or how much it may grow or spread.

Be sure to ask questions if you are unsure about something or want to better understand your disease.

Different NETs affect people differently

Different NETs affect people in different ways in terms of how the tumour grows, the symptoms produced, whether or not they spread and how they spread.

Gaining the histology (what the tumours look like under a microscope) is very important in order to classify the cancer into a type, and the NET team can then work with you to plan the most appropriate treatment.

Although NETs share similar characteristics, the diagnosis and the way your NET may behave may be different.

The most important aspect of caring for a person with a NET is that the care should be tailored to suit the individual and provided by a specialist in the field of NETs. Your quality of life is paramount and so teamwork is essential to provide a solid plan of treatment and follow-up.

There has been much research work done by specialist healthcare professionals, and progress has been made in terms of understanding these tumours. It is important to ensure that you are seen by these specialists in order to access all the knowledge available.

NETs can be complex. For you the journey can encompass not only a whole host of emotions, but also a range of investigations, treatments and healthcare professionals.

Often there is more than one treatment option available, and so there has to be a collaboration amongst all key healthcare professional groups who are making clinical decisions for you.

This collaboration is called a multidisciplinary team or MDT. The MDT management approach is now being used across the world in the care of people with NETs.



You may potentially see the following healthcare professionals:

Healthcare professionals who may be involved in your care

- Gastroenterologist
- Oncologist
- Surgeon
- Endocrinologist
- Radiologist
- Nuclear Medicine Physician
- Nurse Specialist
- Dietitian
- Palliative Care Team
- Pain Team
- General Practitioner / Practice Nurse
- Counselling Staff
- Various Clinic Staff
- Hospital/Ward Staff
- Various Technicians
- Hospice Team
- Advocacy/patient charity



Diagnosis

NETs are difficult to diagnose.

The onset of symptoms takes an average of three to five years, and even then symptoms are often non-specific and vague.

Early diagnosis can potentially mean a better prognosis for patients, but at present however, most NETs are diagnosed at a later stage, when they have already spread to other parts of the body.

In these cases, they can rarely be cured, although the symptoms can often be managed successfully for a number of years.

The symptoms may be mild or non-specific, and are often similar to those of other, more common conditions.

Many doctors are unfamiliar with NETs and are therefore less likely to suspect a NET in their initial investigations.

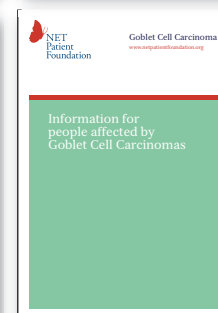
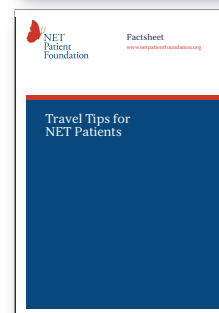
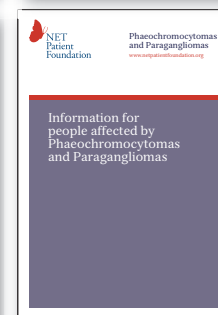
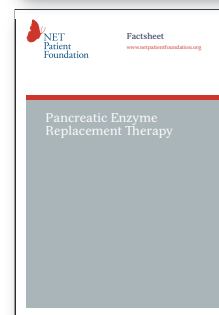
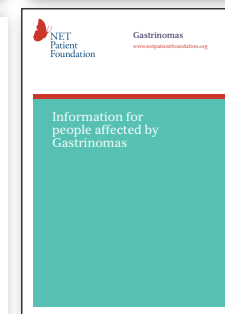
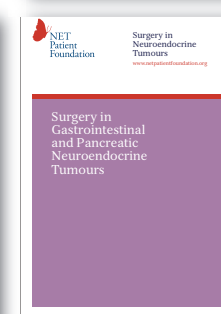
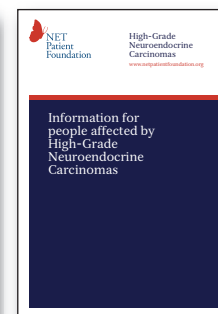
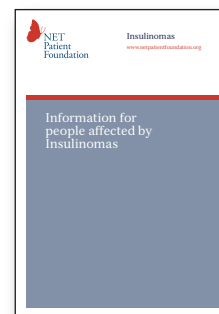
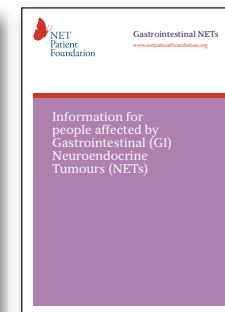
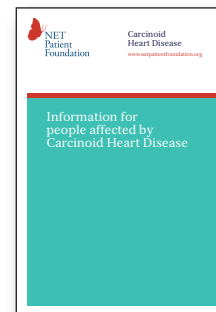
NETs are difficult to detect for a number of reasons:

Reasons why NETs are difficult to detect

- **Size:** they are often small; some NET cancers can be less than a centimetre in size
- **Location:** NET cancers can occur almost anywhere in the body
- **Symptoms:** these can vary enormously, and some patients have no symptoms at all
- **Testing:** there are many different types of NET cancer, and a series of specialised tests is necessary for a definitive diagnosis



There are some helpful tools that you can share with your doctor – for example, booklets about your diagnosis or maybe a GP toolkit. Please get in touch to request your copy.



Your Guide to Neuroendocrine Tumours

We have produced a video about NETs and their treatment, with perspectives from consultants, specialist nurses and patients.

You can view the video on our website: www.netpatientfoundation.org



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.

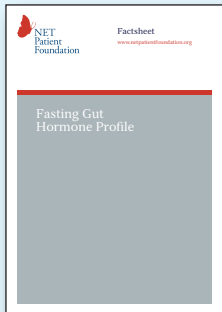
Biopsies are very important in medicine. It is virtually impossible to diagnose some types of cancer any other way. Often, the only way to be sure of the diagnosis is to actually look for cancer cells under the microscope.

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.

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)
- Pro-BNP (a blood test to check for carcinoid heart disease)



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

Urine tests When certain hormones are broken down by the liver, they are excreted as a substance called 5-hydroxyindoleacetic acid (5HIAA) in the urine.

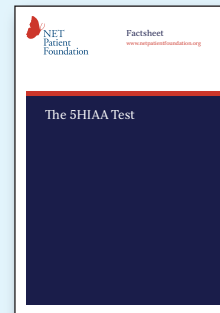
Higher than normal levels of serotonin produced by NET patients show up as raised levels of 5HIAA in their urine.

You may be asked to do a urine collection over a 24 hour period so that your 5HIAA levels can be checked.

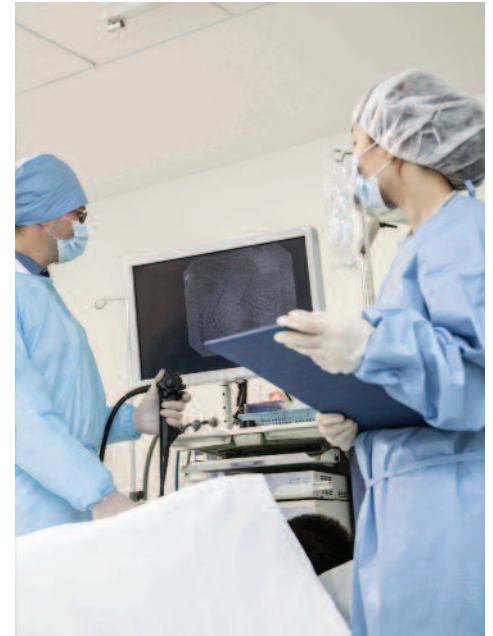
You will be asked to avoid certain foods prior to and during the test including chocolate, olives, bananas, pineapple and its juice, all tomato products, plums, aubergine, avocado, kiwi fruit, walnuts, brazil nuts, cashew nuts, tea, coffee and alcohol.

You will also be asked to avoid certain cough, cold and flu remedies three to seven 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.



For further information see the NET Patient Foundation Factsheet on The 5H1AA Test.



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 (the size of the large vitamin pill) capsule, which contains a colour camera, battery, light source and transmitter. The camera takes six 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'.

MIBG scan This is a similar type of scan to the OctreoScan, except a different radioactive labelled substance is used.

A radioactive MIBG will show up those tumours which have receptors for meta iodo benzyl guanidine (MIBG) on the surface.

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

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

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.

Bronchoscopy A bronchoscopy is an examination of the major air passages of your lungs.

There are two types of bronchoscopes:

- A flexible bronchoscope
- A rigid bronchoscope

During a bronchoscopy your doctor may take samples.

They are sent to the laboratory for analysis and are used by your doctor to make a diagnosis

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.

Reasons for different scans

- Screening
- Detecting the primary tumour in a person suspected of having a NET
- Assessing the extent of disease
- Assessing the response to treatment
- Follow-up after an initial diagnosis

There is no standard group of tests.

Each patient may need to undergo a different combination of tests.

If you have any questions about these tests, please do ask your medical team.

Treatments

The treatment of a NET cancer depends on the size and location of the tumour, whether the cancer has spread, and your overall health.

These are a complex group of cancers to manage, and ideally, a MDT will work with you to determine the best treatment plan. The MDT will always have several goals in mind as they formulate your treatment plan.

Not all of the treatments described here are suitable for all people with NETs. Treatment plans should always be tailored to the individual.

Treatment Goals

- Remove the tumour by surgery; however, if the tumour has spread, this may not be possible
- Alleviate symptoms
- Control the tumour growth
- Maintain a good quality of life for you

Some of the treatments that are used to reduce or stabilise tumour size and alleviate symptoms are discussed below.

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.

Potentially 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.

Symptom Control 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, by removing it all future problems can be prevented by removing disease from key areas before the vital structure has been damaged or blocked.

AT RISK - Bowel Obstruction

A bowel obstruction is a blockage in the bowel. Your bowel may become completely or partly blocked.

The symptoms of a bowel obstruction are a combination of:

- | | |
|----------------------------|--------------------------|
| 1 Feeling bloated and full | 4 Vomiting large amounts |
| 2 Severe Tummy Pain | 5 Constipation |
| 3 Feeling sick | |

If you are worried you must contact your team during working hours. Weekends and out of hours you need to go to the A&E

Please seek medical advice immediately if you suspect that you have a bowel obstruction.

Download a Bowel Obstruction Factsheet at www.netpatientfoundation.org

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Cyto-reductive or Debulking surgery

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

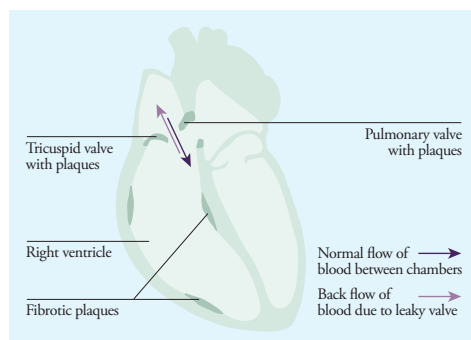
This means reducing the number of living tumour cells inside the patient.

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.

Cardiac surgery This involves valve replacement and may be required for patients with carcinoid heart disease.

Most planned surgery for NET cancers should be carried out in specialist units where the surgeons work as part of a team including oncologists, gastroenterologists, nurses, radiologists and many other doctors all of whom have particular expertise in managing NET cancers.

These are rare and complex tumours so it is best to have a dedicated team of experts jointly involved in deciding which treatment is best.



Somatostatin Analogues Somatostatin is a substance produced naturally in many parts of the body. It can stop the over-production of hormones that cause symptoms such as diarrhoea, flushing and wheezing.

Lanreotide and octreotide are somatostatin analogues i.e. drugs that copy or mimic the action of somatostatin.

Some NETs produce hormones that can cause other symptoms, for example, patients with a carcinoid tumour may have diarrhoea, flushing, and wheezing. You may have different symptoms depending on the type of tumour that you have.

These symptoms can be distressing and often affect your quality of life.

The aim of this treatment is to block the release of the extra hormones your body is producing and therefore improve your symptoms.

Lanreotide Lanreotide can be given as an injection every 7-14 days or as a long-acting injection every 28 days. The long-acting injection can be administered by a nurse, either in hospital or by a practice nurse.

For some patients who are stabilised on their treatment with lanreotide, it may be possible for the patient, or a relative or friend, to be taught how to give the injection themselves. The injection is given in the upper, outer quadrant of the buttock or, if you are self-injecting, into the upper, outer thigh.

If you are using lanreotide at home it should be kept in the refrigerator, in its original package, at a temperature between 2°C and 8°C; it should not be frozen. Your doctor can arrange with Ipsen for lanreotide to be delivered direct to your home or GP surgery, or for trained nurses who can provide guidance and support.

Octreotide Octreotide can be given as a short-acting injection two to three times a day, or as a long-acting injection administered by a healthcare professional every 28 days. The short-acting form is injected into the tissue under the skin, either in the upper arm, thigh or stomach. The long-acting form is injected in the large muscle in the buttock.

Octreotide should be stored between 2°C and 8°C; it should not be frozen.

Side effects

There may be some side effects, but it is important to remember that the following are only possible side effects and do not affect all patients:

- Loss of appetite and problems in the gastrointestinal tract (gut) such as nausea, vomiting, abdominal pain and bloating, wind, and upset bowels
- Occasional discomfort at the site of the injection
- After a period of time some patients may develop gallstones as a result of the treatment but your centre monitors this when you have your regular scans
- Short and longer-acting injections can affect blood sugar levels

People with diabetes should discuss this with their nurse or doctor.

If you experience any problems (or feel you may have developed another side effect) please tell your doctor or nurse.

Further information There are DVDs available for you and the person giving the injections, and a host of other information so please get in touch with the NET Patient Foundation who will be able to provide the information to you.

There are also resources for your GP and practice nurse which provide in-depth information about these injections.

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.

Interferon Interferon alpha is a man made copy of a substance that the body makes naturally. The body makes interferon as part of the immune response. This is when the body reacts to anything it recognises as 'foreign' or abnormal, for example infection or cancer cells.

Interferon works in several ways. It directly interferes with how cells grow and multiply, and it stimulates the immune system by encouraging immune system cells, such as killer T cells, to attack cancer cells.

It also encourages cancer cells to produce chemicals that attract the immune system cells. It is sometimes referred to as biological therapy or immunotherapy and is used to treat some patients with NETs.

In some patients it is given on its own, but can also be given as a combination therapy with a somatostatin analogue.

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).

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 NETs.

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

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.

Psychological aspects

A diagnosis of cancer is one of the greatest challenges you can face. It is normal to feel anxious, frightened, and worried about the future.

When you're dealing with a NET, there's no need to "go it alone."



There are so many things to think about, you may feel overwhelmed.

Even though you and your healthcare team will be focused on managing your physical well-being, it is also important to take care of your emotional and social well-being.

Many patients diagnosed with cancer suffer from anxiety and depression. This can be a vicious cycle because if you are anxious or depressed, it may affect your ability to keep doctors' appointments, take medication, or even interact with your friends and family.

If you think anxiety or depression is making a negative impact on your life, talk to your doctor, nurse, friend or family about this.

One way to deal with the emotional toll of cancer is to connect with others in the same situation. The easiest way to do this is to get involved in a support group.

By talking to others facing the same challenges, you'll realise you're not alone... and you may even find some helpful advice on how to cope with certain situations.

Surround yourself with a personal support group — anyone and everyone who can be a positive force in your life and help keep you motivated when times get tough.

Living with a NET opens the door to many challenges and questions about what will happen next.

Of course, you cannot control the future, but you can make a difference in what you do today.

No matter what type of NET you have, you do have a life and a future.

Questions to ask in clinic

Many patients with NETs will see their doctors and other healthcare professionals on a regular basis.

To make the most of each visit, do the following.

Prepare for each visit

- Bring records of previous visits/tests with you
- Write down any questions or concerns before you go

Know your healthcare team

- Know what each specialist's role/focus is
- Ask why certain referrals, consultations, or tests are being requested

Talk openly with your doctors and nurses

- Ask questions
- Take notes

Keep track of and write down all your symptoms, even if they seem minor or unrelated to your NET. This includes:

- Changes in frequency or severity
- New symptoms
- Changes in your eating habits or daily routine

Have a friend or caregiver go with you for emotional support and to take notes so that you can keep track of everything that was discussed.



Suggested questions to ask the doctor

An important part of managing your care is knowing what questions to ask of your doctor. Every person's needs are unique, and your questions will change over time.

Here are some examples of the types of questions you may want to ask your doctor, and room to record the answers.

General information

- What type of cancer do I have?
.....
.....
- Where exactly is it located?
.....
.....
- What are the risk factors for this disease?
.....
.....
- Is this type of cancer caused by genetic factors? Are other members of my family at risk?
.....
.....
- How many people are diagnosed with this type of cancer each year?
.....
.....
- What lifestyle changes (diet, exercise, rest) do you recommend I make to stay as healthy as possible before, during, and after treatment?
.....
.....
- Where can I find more information about my cancer?
.....
.....



Symptoms

- What are some common symptoms or side effects of this type of cancer?
.....
.....
- How can I avoid these and/or manage them with my daily activities?
.....
.....
- Is there anything that can be done to make my symptoms or side effects better?
.....
.....

- Are there activities that may make them worse?
.....
.....
- If new symptoms or side effects arise or existing ones worsen, what should I do?
.....
.....

Diagnosis

- What diagnostic tests or procedures are necessary? How often?
.....
.....
- What information will these tests tell us?
.....
.....
- How can I prepare myself for each test or procedure?
.....
.....
- Where do I need to go to have this test?
.....
.....
- When will I get the results? How will I get the results (over the phone, at the next appointment, etc.)?
.....
.....
- Can you explain my pathology report (laboratory test results) to me?
.....
.....
- If I seek a second opinion, will I have to repeat any tests or procedures?
.....
.....

- How much information about my diagnosis should I share, and at what time, with my friends and loved ones?
.....
.....

Staging

- What is the stage of my cancer? What does this mean?
.....
.....
- Has cancer spread to my lymph nodes or anywhere else?
.....
.....
- How is staging used to find out cancer treatment?
.....
.....
- What is my prognosis (chance of recovery)?
.....
.....

Treatment

- What are my treatment options?
.....
.....
- Which treatments, or combination of treatments, do you recommend? Why?
.....
.....
- What is the goal of the treatment you are recommending?
.....
.....

- What clinical trials (research studies involving people) are open to me?
.....
.....
- Who will be part of my treatment team, and what does each member do?
.....
.....
- How much experience do you (or the treatment team) have treating this type of cancer?
.....
.....
- Will I need to be hospitalised for treatment, or is this treatment done in an outpatient clinic?
.....
.....
- What is the expected timeline for my treatment plan? Do I need to be treated right away?
.....
.....
- How will this treatment affect my daily life? Will I be able to work, exercise, and perform my usual activities?
.....
.....
- What are the short-term side effects of this treatment?
.....
.....
- What long-term side effects may be associated with this cancer treatment?
.....
.....

- Will this treatment affect my fertility (ability to become pregnant or father children)?
.....
.....
- Besides treating cancer, what can be done to treat my symptoms?
.....
.....
- How can I keep myself as healthy as possible during treatment?
.....
.....

Clinical trials

- What are clinical trials?
.....
.....
- How do clinical trials help people with cancer?
.....
.....
- Is this a treatment option for me?
.....
.....
- What happens during a clinical trial?
.....
.....
- What are the benefits and risks of participating in a clinical trial?
.....
.....
- How will I be monitored while participating in a clinical trial?
.....
.....

- What are my responsibilities during the clinical trial?
.....
.....
- Are there any costs associated with my participation in a clinical trial?
.....
.....
- Where can I learn more about clinical trials?
.....
.....

Support

- What support services are available to me? To my family?
.....
.....
- Whom should I call with questions or concerns during out of hours?
.....
.....
- May I contact you or the nurse to talk about additional information I find?
.....
.....

- Can you recommend a social worker to help locate support services?
.....
.....
- Where can I find resources for children? For teenagers? For young adults? For older adults?
.....
.....
- If I'm worried about managing the costs related to my cancer care, who can help me with these concerns?
.....
.....

Follow-up care

- What follow-up tests do I need, and how often will I need them?
.....
.....
- Is there anything else I should be asking?
.....
.....



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.

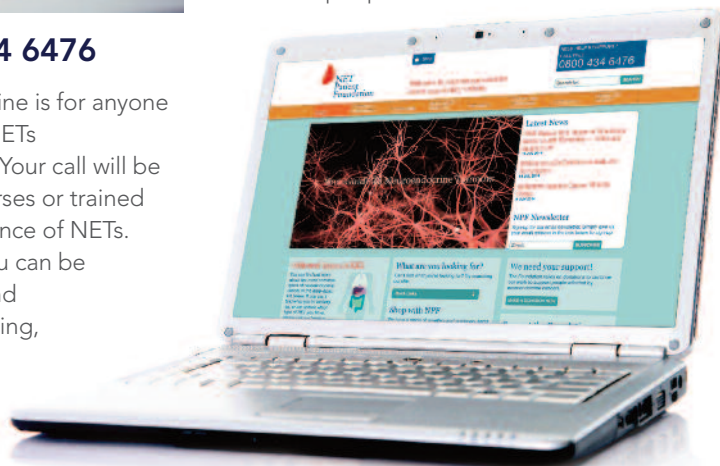
We operate a call-back service for those who wish to leave a message out of hours.

Website – www.netpatientfoundation.org

We know how important it is to understand as much as possible about your NETs.

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 NETs 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.



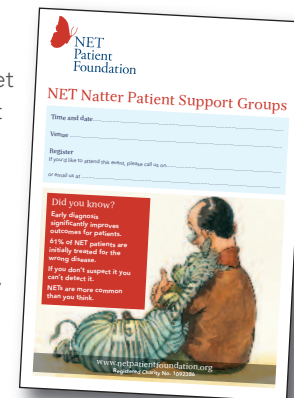
NET Natter Groups

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.

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.



Further support



AMEND

Supporting patients with multiple endocrine neoplasia and associated endocrine growths

AMEND Admin Office

The Warehouse, No 1 Draper Street,
Southborough, Tunbridge Wells,
Kent TN4 0PG

Telephone: 01892 516076

www.amend.org.uk



Cancer Research UK

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www.cancerresearchuk.org

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CANCER SUPPORT**

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
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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