Information for people affected by Ovarian Neuroendocrine Tumours (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

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Proud to be part of the International Neuroendocrine Cancer Community (INCA)
www.netcancerday.org

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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).

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

Where do neuroendocrine tumours occur?

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

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

- Brain
- Thyroid
- Liver
- Gallbladder
- Kidney
- Ovary
- Rectum
- Lung
- Spleen
- Stomach
- Pancreas
- Small bowel
- Large bowel
- Appendix

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Ovarian NETs

What is an Ovarian Neuroendocrine Tumour?
The ovaries are the reproductive organs responsible for producing eggs and female hormones. Ovarian cancer is difficult to detect, as the symptoms are often vague and subtle, similar to those in other non-cancer conditions affecting women.

Ovarian neuroendocrine tumours account for around 5% of all neuroendocrine tumours and only around 0.1% of all ovarian cancers. That is around 30-40 patients seen each year in the UK.

The majority are seen as metastases ('secondary' cancer that has spread from the primary site), where the primary site can be seen in either the small bowel (mid gut) or appendix. Patients with goblet cell carcinomas may also have metastasis in the ovary.

They can vary in stage and grade.

Primary ovarian NETs are extremely rare and there is very limited information on this tumour type. What we do know is that they can be found unexpectedly and incidentally. They are often small and can have two different patterns when looking under the microscope. It is also possible that these NETs can cause the patient to have ‘carcinoid syndrome’.

What is ‘carcinoid syndrome’?
This is 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’.

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

Some of the tests you might undergo include:

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

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

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

Doctors will be looking for certain NET markers, particularly chromogranin A and B, and for evidence of a rise in certain peptides and hormones in the blood, as well as checking how well your kidneys and liver are functioning.

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)

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.

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

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.

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

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’. This investigation is done on a machine called a SPECT CT.

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 for looking at a particular area of your body in more details. 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 3 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.

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 your body’s internal organs, as well as blood flowing through blood vessels.
Treatments

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.

Some of the treatments listed below may only be relevant if the tumour has spread to the liver or other sites and further details can be found in the GI NET booklet.

- Surgery
- Chemotherapy
- Somatostatin Analogues
- Targeted Peptide Receptor Radionuclide Therapy - PRRT
- Liver directed therapies
- Clinical Trials

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.

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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.

Inform aid for people affected by Ovarian NETs

www.netpatientfoundation.org
Multidisciplinary teams (continued)

A few things to keep in mind:

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

Tips:

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

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

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

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

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

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

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

Further Support

NET Patient Foundation

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

Helpline – 0800 434 6476

Our free, confidential helpline is for anyone who has questions about NETs (neuroendocrine tumours). Your call will be answered by one of our nurses or trained staff members with experience of NETs. Whatever your concern, you can be confident we will understand the issues you might be facing, and that the information you receive will be clear and up-to-date.

We will also let you know where else you can go for further support. The helpline is open 10am -2pm Monday to Friday. We operate a call-back service for those who wish to leave a message out of hours.

Website – www.netpatientfoundation.org

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

Discussion forums

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

Information and support sessions

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

NET Natter groups

These are informal groups which meet locally on a regular basis. To find out if there is a NET Natter group in your area, or if you’d like to set one up, please visit our website.
Information resources
Our free information resources are for anyone affected by NETs. They are here to answer your questions, help you make informed decisions and ensure you know what to expect. All of our information is written and reviewed regularly by healthcare professionals and people affected by NETs, so you can trust the information is up-to-date, clear and accurate. You can order our publications by sending us an email or calling the helpline. All our publications can also be downloaded from our website.

Other organisations
NHS Choices
Information on clinical trials and treatment centres, including reviews and ratings.
www.nhs.uk

Cancer Research UK
Trusted information on all cancer types.
www.cancerresearchuk.org

Macmillan Cancer Support
Macmillan Cancer Support provides practical, medical, emotional and financial support to people living with cancer and their carers and families. Over the phone, its cancer support specialists can answer questions about cancer types and treatments, provide practical and financial support to help people live with cancer, and are there if someone just wants to talk. Its website features expert, high-quality information on cancer types and treatments, emotional, financial and practical help, and an online community where people can share information and support. Macmillan also funds expert health and social care professionals such as nurses, doctors and benefits advisers.
www.macmillan.org.uk
General enquiries: 020 7840 7840
Helpline: 0808 808 0000
Textphone: 0808 808 0121 or Text Relay

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

We Support:
• Vital clinical research in the UK
• Education for healthcare professionals

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

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

• Over 50% of patients are diagnosed when the cancer has already spread

• Early diagnosis significantly improves outcomes for patients

• There are dedicated NET clinics around the UK. Find out where at www.netpatientfoundation.org

Why the moth?

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

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

About us

The Foundation has the following aims:

• To provide support, education and information to anyone affected by neuroendocrine cancers

• To advocate for neuroendocrine cancer patients so that they may achieve the best possible outcomes

• To encourage standardised care for all NET patients

• To provide community supportive care to patients and their carers or family members

• To raise awareness of NETs throughout the UK

• To raise funds for clinical research projects.

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