Gastrinomas

NET Patient Foundation
Gastrinomas are relatively rare neuroendocrine tumours that were first described in 1955. They are mostly found in the duodenum, the first part of the small bowel, and the pancreas. However they are the most common of all ‘functional’ pancreatic neuroendocrine tumours – in other words tumours that not only grow but also secrete large quantities of hormones causing side effects and symptoms that collectively make up a recognised syndrome.

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

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

The main symptoms are:

- Abdominal pain
- Acid reflux
- Diarrhoea

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

How are Gastrinomas diagnosed?

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

There are a number of tests that can be performed to confirm a diagnosis.

Hormone tests

**Fasting gut hormone blood test**

You will be asked to fast overnight or for at least six hours before this blood test. The test will reveal levels of certain hormones produced by the tumour - in this case gastrin - and also neuroendocrine tumour markers called chromogranin A and B. If you are taking medication known as PPIs or proton pump inhibitors (for example omeprazole) you may have to stop taking it for one week before the test. This is because PPIs can interfere with test results and give a false high reading. Some patients will not be able to come off their medication and this will be discussed with you by your doctor. Please do not come off any medication without being told by the medical/nursing team to do so.

**Secretin stimulation test**

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

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

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

**Endoscopic ultrasound (EUS)**

Is a technique that uses a special endoscope that has an ultrasound machine at the tip. *Endoscopy* refers to the procedure of inserting a long flexible tube via the mouth or the rectum to see the digestive tract, whereas *ultrasound* uses high-frequency sound waves to produce images of the organs and structures inside the body such as the liver and pancreas. Using the EUS scope, doctors can place the ultrasound probe in the GI tract, close to the area of interest, so that a very detailed image of the deeper layers of the GI tract, surrounding lymph nodes, blood vessels, and organs can be obtained. Biopsies can also be taken at the same time. EUS is performed under sedation in the endoscopy unit. For detection of the primary tumour this procedure has a high sensitivity and accuracy level.

- **Biopsy** - any tissue samples collected will be sent to a histopathologist for investigation. Under a microscope gastrinomas often appear ‘well differentiated’ which is a term used to indicate that they are slow-growing tumours.

**Endoscopy**

Refers to the procedure of inserting a thin flexible tube with a tiny video camera and light on the end via the mouth into the stomach. The high quality picture from the endoscope is relayed onto a TV monitor and the image gives a very detailed view of the digestive tract and any suspect areas.

**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 tumours, and regular scans are useful to find out more about the rate of tumour growth and how your tumour is responding to treatment. When you arrive at the clinic you will probably be asked to drink a litre of fluid that contains a contrast agent that helps to highlight tumours, and you may also have a cannula inserted through which a special contrast dye is administered during the scan. These both help your specialists to read the scans more clearly as the tumours are highlighted.

**MRI scan**

This is a whole body scan. Magnetic resonance imaging (MRI) scans can help reveal where tumours are positioned. It uses magnetism rather than X rays to take pictures of inside the body. Scans can take up to one hour to complete and you have to stay very still inside the scanner lying on a couch. MRI is often used in conjunction with CT and SRS scans. They have a good value when detecting liver metastases (spread of tumour into the liver).

**SRS Somatostatin Receptor Scintigraphy (Octreotide scan)**

This is a common scan for NET patients. It can help to detect tumours that might be missed on other conventional scans. A high proportion of gastrinomas have special receptors on their surfaces called somatostatin receptors. Octreotide is a somatostatin analogue, a substance that mimics the action of naturally occurring 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 hot spots. This is a useful test to find out more about your tumours, where they are positioned, and also whether you would be suitable for certain treatments that use Octreotide or another chemical called Lanreotide as a carrier agent.
Treatment for Gastrinomas

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

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

Surgery
This is the only treatment that can cure gastrinomas and should be performed by a surgeon experienced in treating these tumours. If the tumour has already spread to other organs or if there are other contraindications to surgery then other treatments may be considered.

Transarterial chemoembolisation
May be considered if there has been spread of disease to the liver. This procedure involves cutting off the blood supply to the tumours with or without the addition of intra-arterial chemotherapy. Occasionally other ablation techniques such as radiofrequency ablation might be used if the tumours in the liver are small and few in number. This involves guiding a special needle electrode to the tumour and then passing a radio frequency current through it to heat the tumour tissue and ablate, or eliminate, it.

Medical treatment with somatostatin analogues
These are injections often prescribed to alleviate some of the problematic symptoms associated with NETs. Daily or monthly injections of somatostatin analogues are available. Somatostatin analogues are synthetic versions of somatostatin, a naturally occurring hormone produced in the brain and digestive tract. It inhibits the release of several other hormones and chemicals from our internal organs. The two common brands are Sandostatin LAR and Lanreotide. These medications are usually given by health care professionals and are available in different doses.

Chemotherapy
With streptozotocin and doxorubicin with or without 5FU may be considered for patients with fast growing advanced tumours. You may be offered different combinations of chemotherapy and your medical team will discuss this with you.

Radionuclide targeted therapy
Also known as magic bullet treatment, is considered for patients who have advanced inoperable tumours that have positive uptake on the Octreotide scan. This treatment carries a radioactive particle e.g. Yttrium-90 or Lutetium-177 attached to Octreotide to wherever there are tumour cells (which have lit up on the Octreotide scan).

New Drugs
Sunitinib and Everolimus are two drugs which have recently been shown to be beneficial to treat pancreatic NETs. These drugs are both given in tablet form. They work by interfering with the development of blood vessels to the tumour and they also disrupt the ability of the tumour cells to grow. It is also possible for these drugs to be used in combination with somatostatin analogues.
Multidisciplinary teams

Gastrinoma care 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 among 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.

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

- Oncologist
- Surgeon
- Endocrinologist
- Radiology staff
- Dietitian
- Nurse Specialist
- Palliative Care Team
- Pain Team
- General Practitioner/Practice Nurse
- Counselling Staff
- Various Technicians
- Clinic Staff
- Hospital Staff
- Hospice Team
- Nuclear Medicine Physician

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 gastrinoma patients.

Clinical Research

Research is a step-by-step process that involves collecting and examining information. Research into gastrinomas is vital to improve our understanding of the disease and how it can be treated.

Research goals include:

- Understanding what causes gastrinomas
- Understanding how gastrinomas form
- Formulating more effective diagnostic scans and tests
- Discovering new treatment options, and ensuring that current treatments are being implemented to provide the best therapeutic benefit

Gastrinomas are a rare form of cancer, and there are small teams of dedicated medical professionals around the world who treat patients every day. It is important that these specialists are allocated the resources to carry out research within their units, so that our understanding of this disease and how to treat it continues to grow.

In clinical trials, patients agree to try new therapies (under careful supervision) in order to help doctors identify the best treatments with the fewest side effects.

If patients want to take part in a clinical trial, they should discuss this with their specialist, who will know whether they are eligible.

All studies are run on strict inclusion and exclusion criteria for the safety of the patients. It can be frustrating for patients to discover that they are ineligible, but no medical professional is able to influence any decisions based on these criteria.

No one should ever include a patient in a clinical trial without his or her knowledge. A doctor, nurse or other researcher will ask for permission, and they cannot enter a patient into the trial unless that patient has given his or her consent.

To help patients decide whether they want to take part, the researchers should tell them all about the study:

- what it is trying to find out
- how they will be treated
- what they will have to do.
Even after consent has been given, a patient may leave the trial without giving a reason at any time. If a patient is having a new treatment as part of a trial and then leaves the trial, he or she may not be able to continue having the new treatment. In this situation, patients would be given the appropriate standard treatment for their type of cancer.

You can find out more about current trials at:

**Current Controlled Trials**
This website allows users to search, register and share information about randomised controlled trials. Covers multiple registers, including England, Scotland and the US.
http://www.controlled-trials.com/

**ClinicalTrials.gov**
This is a register of federally and privately supported clinical trials conducted in the United States and around the world.
clinicaltrials.gov

You can also search for research trials available to patients in the UK and Europe on these websites:
www.macmillan.org.uk
www.cancerhelp.org.uk
www.ctu.mrc.ac.uk (The Medical Research Council Clinical Trials Unit)
www.ncrn.org.uk (National Cancer Research Network)
www.eortc.be/ (European Organisation for Research and Treatment of Cancer)

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**Useful Organisations**

**NET Patient Foundation**
Patient support and advice
0800 434 6476
www.netpatientfoundation.org

**A.M.E.N.D (Association for Multiple Endocrine Neoplasia Disorders)**
Support and advice for people affected by MEN1 and other Multiple Endocrine Neoplasia Disorders
www.amend.org.uk
Further support

NET Patient Foundation

From diagnosis, throughout treatment and beyond, our services are here every step of the way. Here 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 here 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 is 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 also 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 cancer. Our website is here round-the-clock giving you instant access to information when you need it. As well as clinical information, you’ll 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.

Ask the nurse

If you find it difficult to talk about your cancer, we can answer your questions by email instead. Our ‘ask the nurse’ service is available on the website - complete a short form that includes your question and we’ll get back to you with a confidential, personal response.

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, invaluable Question & Answer sessions, as well as an opportunity to meet other NET patients. For information about meetings in your area, please see our website or call our helpline.

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

Macmillan Cancer Support

89 Albert Embankment London SE1 7UQ
General enquiries: 020 7840 7840
Helpline: 0808 808 0000
Website: www.macmillan.org.uk
Textphone: 0808 808 0121 or Text Relay

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. The NET Patient Foundation incorporating Living with Carcinoid was formed at the start of 2006 and has Charity Commission status.

The Foundation has the following aims:

- To provide accurate and up-to-date information for people living with, or affected by, NET cancers
- To provide support for patients and others affected by NET cancers
- To provide education for healthcare professionals in the treatment and care of NET patients
- To raise awareness of NET cancers within the medical community, and amongst the general public
- To improve access to care for NET patients
- To raise funds, which will help to support research around the UK & Ireland

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

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.”

Did you know?

“...a moth.”

plateau window, don’t assume it’s a butterfly.

“...a moth.”

[Image -8x518 to 565x604]
The NET Patient Foundation supports people diagnosed with neuroendocrine tumours and their families.

For further information and to make contact telephone 0800 434 6476 or visit our website: www.netpatientfoundation.org

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