About Us
The NET Patient Foundation incorporating Living with Carcinoid was formed at the start of 2006 and has Charity Commission status. The collaboration with Living with Carcinoid meant that the charity could spread its wings to include patients with all types of neuroendocrine tumour.

The Foundation has 5 main aims:
1. To provide accurate and up to date information for people living with, or affected by, neuroendocrine tumours. We do this through information and forums provided on the website (and links to other sources of information on the web).
2. To provide support for patients and others affected by neuroendocrine tumours. We offer a support line which is manned by an experienced healthcare professional; and organise patient support groups around the UK and Ireland.
3. To improve the quality of life for patients and their families. We can do this by providing support and information about services and treatments available, as well as by funding and publicising research and existing medical initiatives.
4. To raise funds, which will help to support research around the UK.
5. To raise awareness of neuroendocrine tumours throughout the UK and Ireland, within the medical community and the general public.

There is much campaigning work to be done. We don’t have the funding that breast or bowel cancer attracts. But our patients deserve exactly the same attention and research support.

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

This booklet is part of series of publications for patients about neuroendocrine tumours and related conditions, treatments and tests.

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Bronchial Carcinoid Tumours

Lung carcinoid tumours are an uncommon type of tumour that starts in the lungs. These tumours can occur at any age with the average onset in the 5th 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.

To understand lung carcinoid tumours it helps to know something about the normal structure and function of the lungs, as well as the neuroendocrine system. The lungs are sponge-like organs in your chest cavity. Your right lung has 3 sections, called lobes. The left lung has 2 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 Neuroendocrine Tumours

Like most cells in your body, lung neuroendocrine cells sometimes go through certain changes that cause them to grow too much and form tumours. There are 4 types of neuroendocrine lung tumours:

- typical carcinoid tumour
- atypical carcinoid tumour
- large cell neuroendocrine carcinoma
- small cell lung cancer

Hormone related symptoms are not common for Bronchial carcinoids, but a small number of patients experience “Carcinoid syndrome” – flushing, diarrhoea, palpitations and wheezing.

Typical carcinoid tumours

These grow slowly and only rarely spread beyond the lungs. About 9 out of 10 lung carcinoids are typical carcinoids.

Atypical carcinoid tumours

These grow faster and are more likely to spread to other organs. Seen under a microscope, they have more cells in the process of dividing and look more like a fast growing tumour. They are much less common than typical carcinoids.

Small cell lung cancer

Small cell lung cancer (SCLC) is one of the fastest growing and spreading of all 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 SCLC in its prognosis (outlook) and in how patients are treated.
Treatment For Bronchial Carcinoids

Surgery is the only curative option for this patient group, although surgery may also be used to remove recurrent (returning) disease.

Chemotherapy
Chemotherapy may be an option for patients with bronchial carcinoid tumours. Many chemotherapy regimens involve intravenous drugs, however there are now also oral chemotherapy agents and your oncologist will discuss the most appropriate option with you.

The histology of the tumour i.e. how it looks down the microscope after biopsy or operation, may help determine the type of treatment you receive. Chemotherapy may sometimes be recommended after surgery (adjuvant therapy). You may be asked to be involved with clinical trials currently underway which are looking into the different combinations of chemotherapy agents that are most appropriate for different types of NET cancer.

Somatostatin analogues
If the Octreotide Scan is positive, some patients may be offered somatostatin analogues. These consist of daily, or monthly injections to control some of the unpleasant symptoms caused by carcinoid tumours. Somatostatin analogues (e.g. lanreotide, octreotide) 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.

The octreotide formulation can be given by infusion, self injections daily, or in a monthly injection which is a slow release formula. The lanreotide formula can be given every other week and in a monthly slow release formula. The monthly injections are mostly administered by healthcare professionals, but it is possible to self inject the lanreotide monthly injection.

Injection of these analogues can stop the overproduction of hormones that cause symptoms such as flushing, wheezing and diarrhoea. Under clinical supervision, these treatments may also be used in NET patients with no clinical syndrome. Long-acting octreotide can also control tumour growth.

Investigations Needed

To find out more about the tumour type you have you may be asked to have:

Blood test
Doctors will be looking for neuroendocrine tumour markers such as Chromogranin A alongside other markers.

CT scan
This can help determine the size and position of the tumours and may be taken regularly to help monitor the tumours.

Octreotide Scans (OctreoScan®)
This scan uses a body imaging technique. Cells that receive hormonal messages do so through receptors on the surface of the cells. For reasons that are not understood, many neuroendocrine tumour cells possess especially strong receptors; for example, NETs often have strong receptors for somatostatin, a very common hormone. The OctreoScan® uses a synthetic form of somatostatin, which is chemically bound to a radioactive substance. This is then injected via a vein in the arm and then observed 24 hours later using a radio-sensitive scan. These scans can diagnose and locate around 80-90% of NETs, although further scans, such as PET scans may still be required.

PET scan
Positive Emission Tomography (PET) scan is a nuclear medicine imaging technique which produces a three-dimensional image of functional processes in the body. When FDG-18 and/or Gallium is injected into your body it travels to places where glucose is used for energy. It shows up cancers because they use glucose in a different way from normal tissue. And it will show up changes in tissues that use glucose as their main source of energy - for example, the brain. It is usually used in conjunction with a CT scan to build up a picture of the size, location and status of tumours.

PET scanners are very expensive and a limited number of hospitals have one. This means that a patient may have to travel to another hospital for their scan. Not everybody who has cancer will need to have a PET scan, other types of tests and scans may be more suitable.

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Radiofrequency Ablation (RFA)

Radiofrequency ablation is used when a patient has relatively few secondary tumours (metastases). A needle is inserted into the centre of the tumour and a current is applied to generate heat, which kills the tumour.

Interferon

Interferon is a naturally occurring substance that is produced by the body's immune system. 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 quite often it is given as a combination therapy with a somatostatin analogue.

New Therapies

In recent years there have been new developments with agents that target the enzymes (chemicals) made by tumour cells that cause the tumours to grow. Such new agents include Sunitinib and Everolimus. Additionally the agent Pasireotide which targets a broad range of somatostatin receptors is being used in clinical trials. It is possible that you may be asked to be in a trial involving one of these or other similar new agents.

For further information on any of the treatments please contact us on 0800 43408476 or at www.netpatientfoundation.com

Peptide Receptor radionuclide therapy

Radionuclide therapies are used in some centres. These can include Yttrium 90 DOTA-Octreotate, Lutetium 177 DOTA Octreotate and I 123 MIBG therapy, they do have possible side effects such as bone marrow suppression, renal toxicity and fatigue and these will be closely monitored after each treatment.

Which treatment is appropriate depends upon the uptake in the corresponding nuclear medicine scan i.e. Octreotide scan or Ga68 PET Scan for Yttrium and Lutetium and MIBG scan for MIBG therapy. These treatments can help stabilise disease and improve symptoms.

Other treatment options include:

**Embolisation**

If the tumour has spread to the liver, the patient 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 with liver metastases and metastasis outside of the liver. It is a procedure that would be done 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).

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