

### NET Patient Foundation

NET Patient Foundation (NPF) is a UK wide charity solely dedicated to providing support and information to those affected by Neuroendocrine Cancer.

### The Neuroendocrine System

The Neuroendocrine System is made up of specific cells, found throughout the body, that help regulate normal bodily functions such as breathing and digestion.

### Neuroendocrine Cancer

Neuroendocrine Cancer is a term used to cover a group of cancers that start in neuroendocrine cells. These cancers may also be referred to as NETs, NECs, **NENs (Neuroendocrine Neoplasms)** or even Carcinoids. Neuroendocrine Cancer occurs when neuroendocrine cells stop working normally and start to grow or behave abnormally.

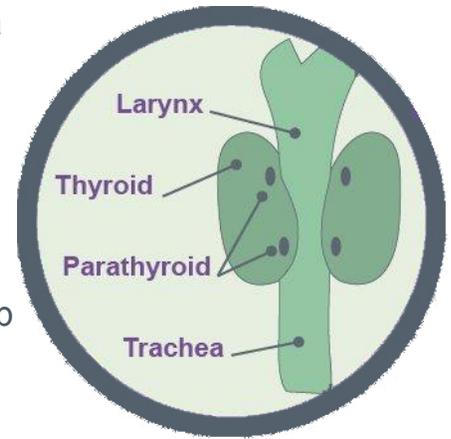
Further information about Neuroendocrine Cancer, including videos and support services can be found at [www.netpatientfoundation.org](http://www.netpatientfoundation.org)

## The Thyroid

The thyroid is a butterfly shaped gland, that sits just below your Adam's apple, lie either side of your windpipe : its 2 lobes are joined together by a bridge (the isthmus).

The thyroid is made up of 2 main types of cell:

- Follicular cells which produce thyroid hormones (T4- thyroxine & T3 triiodothyronine) – that help regulate metabolism
- Parafollicular cells (also called C cells) which produce a hormone called calcitonin - that helps regulate levels of calcium and phosphate.



In the cells and tissues of the body the T4 is converted to T3 and T3 regulates the speed with which your body cells work (metabolism).

Medullary Thyroid Cancer (MTC) is the third most common type of thyroid cancer (<5%) and originates in the C cells or parafollicular cells of the thyroid, therefore, MTCs rarely produce hypo- or hyper-thyroid symptoms.

Hypothyroidism (underactive thyroid): tiredness, feeling cold, weight gain, poor concentration, depression.

Hyperthyroidism (overactive thyroid): weight loss, heat intolerance, anxiety, and, sometimes, sore and gritty eyes. Sometimes there are very few symptoms.

Symptoms, if they do occur, may include a tender/painful lump in the thyroid and/ or diarrhoea (usually associated with raised calcitonin levels)

Most MTCs are sporadic, that is, non-familial (not genetic), however, up to 33% are associated with a family history - in particular, with around 25%, are those with MEN (Multiple Endocrine Neoplasia).

**NET or Neuroendocrine Tumour** is neuroendocrine cancer with well-differentiated cells that usually has a slow to moderate growth rate.

**NEC or Neuroendocrine Carcinoma** is neuroendocrine cancer with poorly-differentiated cells that grow more rapidly.

## Blood / Urine Tests

- Full blood count
  - (B12 + serum Iron)
  - Liver and kidney function
  - Biochemical :
  - Chromogranin A (and B)
  - Calcitonin
  - Calcium
  - Thyroid function tests
  - PTH
  - CEA
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- Cushing syndrome check - if clinically indicated
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- Genetic screening is recommended – especially if familial / genetic disease present/suspected eg family history of MTC and / or MEN.
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- RET (Proto-oncogene tyrosine-protein kinase receptor) proto-oncogene analysis should be performed after surgery: even in the absence of a familial history
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- Provocative pentagastrin or calcium test

## Pathology

- Neck USS
- Chest CT
- CT / MRI Liver
- Bone scintigraphy -a s clinically indicated / suspected
- FDG-PET – if High Grade / rapidly progressing disease - nb frequently negative.

## Scans

- Differentiation and cellular morphology
- Synaptophysin
- Chromogranin
- Ki67
- Calcitonin, CEA
- C-cell hyperplasia

For all patients, there are many things to consider in planning treatments. Your treatment will be personalised to you and the type of NEN you have.

Even if you have a diagnosis that sounds the same as another patient, your treatment and follow up plan may be different.

Your care team will discuss your treatment options with you - giving you both written and verbal information - to help you make an informed choice. Together you can agree on the most appropriate treatment for you.

Information about the treatments that are used in NET and NEC can be found in the NPF Handbook - Your Guide to Living with Neuroendocrine Cancer - [www.netpatientfoundation.org](http://www.netpatientfoundation.org)

There is consensus agreement that all Neuroendocrine Cancer patients should be reviewed by a Specialist Neuroendocrine Cancer MDT.

Follow up as dictated by histology, staging and presence/absence of inherited disorder:

At 3months - biomarkers, physical exam +/- neck USS  
Then 3 - 6monthly for first 2 years.

Further follow dictated by stratified risk - ongoing surveillance recommended at least annually and further investigation to be expedited in presence of rising markers and / or clinical evidence of recurrence/progressive disease.

Lifelong follow-up is recommended for all patients with MTC. Screening should include calcitonin and CEA. Thyroid-stimulating hormone suppression is not necessary. Rising calcitonin levels should trigger investigations to identify potentially treatable metastatic disease.

Advanced disease: follow up as per guidelines – nb should be guided by prognosis, expected treatment efficacy and treatment related toxicity (performance status and clinical indication for active intervention).

A big part of meeting with your doctors, or specialist nurse, is to make sure you get the information you need to understand what's happening, so that you can make an informed choice about your care. Asking questions can be difficult, especially if you're feeling nervous, confused, frightened or struggling to understand what you are being told. You might want to know as much as possible straight away or prefer to take things in small amounts at your own pace.

### **Suggestions that may help:**

- Prepare a list of questions that are important to you
- Ask for simple explanations - do not be worried about asking your nurse or doctor to repeat what they have said
- Take someone with you or ask if you can record the conversation. Many mobile phones have a record function or an app you can download
- Ask for a copy of any letters sent to your GP and/or other care team(s)
- If you have a nurse specialist - keep in touch. They can be a great source of information and support for you.

### **Example questions:**

- Who can I call if I have any questions? Who is my main point of contact?
- Who will be involved in my care?
- What are the treatment options for me? How might they affect me ?
- How often will I need to have scans and tests?
- Are there any flags or warning signs I need to look out for?

Further information about making the most of your consultations can be found in our handbook: [www.netpatientfoundation.org](http://www.netpatientfoundation.org)

#### REFERENCES

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