

NET Patient Foundation

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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

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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

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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

The Pancreas

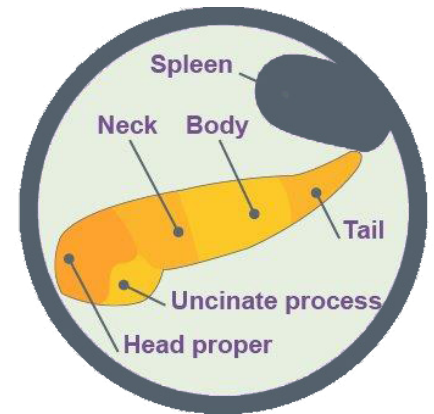
The pancreas is responsible for producing substances (enzymes, peptides/hormones) that play an essential role in converting the food we eat into fuel for the body's cells – as well as regulating our blood sugars.

Exocrine function – enzyme production, released into ducts within the pancreas which then flow into the duodenum via the main pancreatic duct. Endocrine function – hormone production, such as insulin and glucagon, released directly into the bloodstream.

Most pNENs occur incidentally, that is without a family history, but up to 30% may occur on the background of a genetic syndrome/inherited condition e.g. MEN1, VHL, NF1 and Tuberous Sclerosis.

Signs and symptoms of non function pNENs tend to produce symptoms related to tumour position / size, however, raised hormone levels may still be seen in blood results but hormone related symptoms may not occur:

- Nausea,
- Epigastric (the upper abdominal area just beneath the ribs) discomfort or pain – which may radiate through to the back
- Loss of appetite and / or weight
- Rarely jaundice (yellowing of the skin and whites of the eyes).



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)
- Gut hormone profile (as baseline)
- Urinary 5-HIAA

- If inherited disorder present/suspected : screen as per disorder e.g. MEN1, VHL, TS, NF1

Endoscopy

- Endoscopic Ultrasound (EUS)

Scans

- Contrast CT / MRI
- *Gallium-Dotatate PET/CT (SRS SPECT/CT if Dotatate PET n/a)
- FDG-PET – if High Grade / rapidly progressing disease

Pathology

- Differentiation and cellular morphology
- Synaptophysin
- Chromogranin
- Ki67

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

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

For further information about underlying genetic conditions, for example, MEN1 visit www.amend.org

Follow-up for Pancreas - Non-Functioning

As per national and international guidelines nb local policy may differ

Follow up is dependent on grading and completeness of resection (if undertaken):

- G1 : R0 resection / no lymph nodes : post operative check and evaluation.
- G1 (R1/node positive) & G2 : see at 3, 6 and 12 months then annually if stable : labs & CT/MRI.
- EUS +/- Functional imaging (at 1yr then 3 yearly unless recurrence/ progression is suspected - optional).
- G3 : review 2 - 4monthly if on ongoing (or may fit criteria) for treatment. FDG-PET as preferred mode of functional imaging in high grade disease.

If inherited disorder present/suspected : follow up as per disorder e.g. MEN1, VHL, TS, NF1.

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

REFERENCES

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