

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.

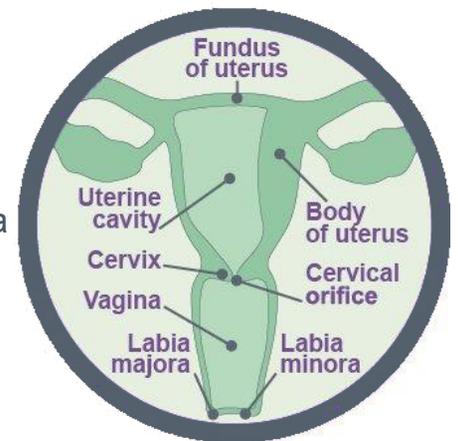
Neuroendocrine Cancer

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

Gynaecological NENs

The ovaries produce the female hormones oestrogen and progesterone, they are also responsible for the production and release of eggs into the Fallopian tubes at the mid-point of each menstrual cycle. The uterus is the shape of an upside down pear and sits within the pelvis, between the bladder and the bowel. It is where a foetus develops and grows during normal pregnancy. The cervix is a cylinder-shaped neck of tissue that connects the vagina and uterus (womb). The vagina allows for menstrual blood to flow out of the body from the uterus, it is also the 'birth canal'. The vulva is the external female genitalia - it includes the clitoris and openings of both the vagina and urethra (the opening through which urine from the bladder is passed). There is a network of blood vessels and nerve pathways linking the uterus to the external genitalia, that play a role in sexual response and orgasm. NENs of the gynaecological organs are uncommon to incredibly rare, depending on site of origin.



Ovarian NENs are rarely primary, they are usually secondary deposits from the bowel or appendix. Primary ovarian NEN is diagnosed is approximately 30 women a year in the UK. Primary NENs of the cervix and uterus are even rarer, with vaginal and vulval NENs the most rare - only a handful of women, across the world diagnosed. Symptoms, if present, mimic ovarian, uterine, cervical, vaginal and vulval adenocarcinoma - diagnosis is usually made by histology review, following surgery or a biopsy. Some uterine or ovarian NENs may also mimic small bowel symptoms, and a small percentage of ovarian NENs do experience Carcinoid Syndrome.

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
- Calcium
- Thyroid function tests
- LDH, Ca125, CEA
- Urinary 5HiAA
- NT-Pro-BNP

- Assess for presence of Cushing Syndrome, SIADH, Hypoglycaemia, Carcinoid syndrome and hypercalcaemia

- PAP smear

- Echocardiogram : as a baseline in the presence of carcinoid syndrome / raised U5HiAA and / or elevated NT-Pro-BNP +/- clinical signs of heart valve impairment/R sided heart failure.

Scans

- USS / Transvaginal Ultrasound (TVUSS)
- CT chest/abdomen/pelvis
- MRI Liver
- Functional imaging : Gallium-Dotatate PET/CT (SRS SPECT/CT if Dotatate PET n/a)
- FDG-PET – if High Grade / rapidly progressing disease
- Bone scintigraphy - if bone disease present/ suspected.

Pathology

- Differentiation and cellular morphology
- Synaptophysin
- Chromogranin
- Ki67
- HPV18
- CD56, PYY - optional
- Exclude Merkel cell in vulval disease.

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.

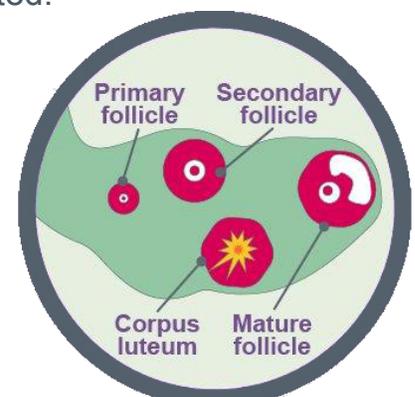
Information about the treatments that are used in NET and NEC can be found in the NPF Handbook - Your Guide to Living with Neuroendocrine Cancer - www.netpatientfoundation.org

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

No definitive agreement exists on the best post-treatment surveillance and should be guided by histology grading and staging, and potential for treatment:

- Gynecological examination including PAP smear is usually performed every 3 months for the first 2 years, every 6 months for the next 3 years, and yearly thereafter
- Biomarkers as indicated / if elevated at diagnosis +/- if recurrence, metastatic disease develops
- CT or PET/CT scan should be performed as clinically indicated.

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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