

What is (Primary) Neuroendocrine Cancer?

What is cancer?

Cancer is a condition where cells within the body start to grow and reproduce uncontrollably. These cells can invade and destroy surrounding healthy tissue, including organs and some can spread to other parts of the body (metastasise).

Uncontrolled growth may be quick, moderate or slow.

The difference between normal cells and cancer cells

Normal cells:

- Develop and grow in a controlled manner
- Do what they are supposed to do, then die off as they are replaced by new cells
- When they become damaged - try to repair themselves or will die and be replaced if they can't
- Do not spread to other parts of the body
- Do not infiltrate (grow into) surrounding tissue - if they grow too big they tend to push against rather than through

Cancer cells:

- Have an uncontrolled development and growth rate
- Do not always do what they are supposed to, do not die off but continue to grow alongside new abnormal cells
- Do not try to repair themselves or die, but continue to grow producing further abnormal cells
- Can spread to other parts of the body, by either growing into surrounding tissue or entering the blood or lymphatic system
- Will grow into surrounding tissue rather than push against it when they grow too big



What are neuroendocrine cells?

Neuroendocrine cells exist throughout the body and are there to produce hormones and other chemicals to ensure our bodies function normally. These cells make up a coordinated system – the neuroendocrine system – a network of communication to control and regulate that hormone and chemical release.

What is Neuroendocrine Cancer ?

Neuroendocrine Cancer is cancer that starts in neuroendocrine cells.

World Health Organisation (WHO) recommends the clinical term **Neuroendocrine Neoplasms (NENs)**.

NENs (Neuroendocrine Cancers) are grouped according to their appearance and growth rate (when examined under a microscope) - also by site and symptoms.

There are 2 main types:

- Neuroendocrine Tumours (NET)
- Neuroendocrine Carcinoma (NEC)

Cancerous neuroendocrine cells **look abnormal** and the degree of abnormality can be described by the word **differentiation**.

Growth rate, often referred to as **grading**, is how many of the cells within a cancer are developing and dividing. Understanding this, helps to determine how slowly or rapidly a neuroendocrine cancer might grow.

In terms of cancer development, normal cells have a set of rules that regulate growth and behaviour, in cancer the control signals go wrong and the rules are forgotten!

The 2 main types of Neuroendocrine Cancer (NENs) by appearance:

Well-differentiated (WD)

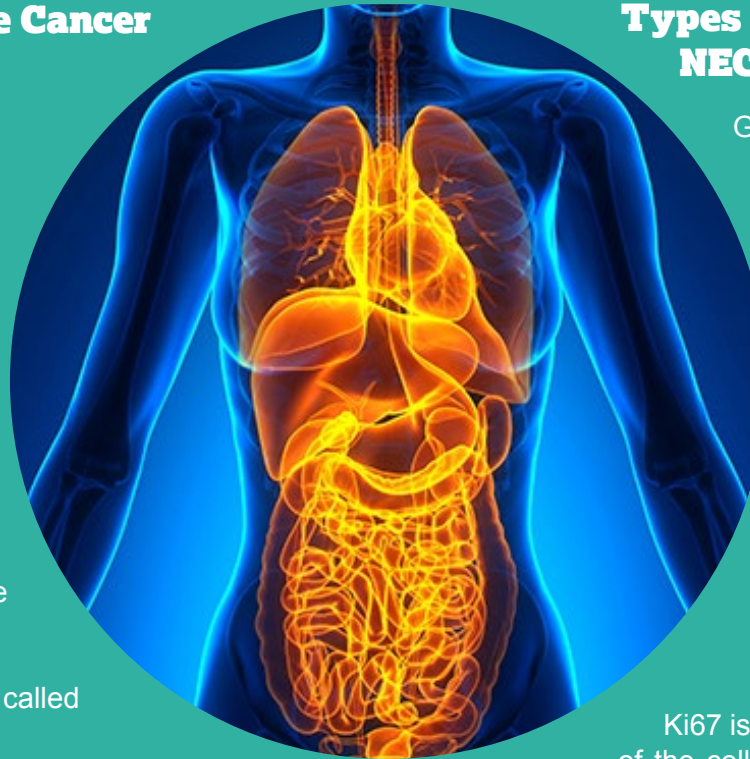
Neuroendocrine cancer cells still have some features of the original cell but have started to change shape and size. They tend to have a slow or moderate rate of growth, though some may grow rapidly.

Well-differentiated Neuroendocrine Cancers are called Neuroendocrine Tumours or NETs.

Poorly-differentiated (PD)

Neuroendocrine cancer cells that have lost almost all features of the original cell, have changed size and shape. They tend to grow rapidly.

Poorly-differentiated Neuroendocrine Cancers are called Neuroendocrine Carcinomas or NECs.



Types of Neuroendocrine Cancer (NET or NEC) by growth rate:

Growth rate or Grading - is a measurement of how many abnormal cells with a tumour or carcinoma are actively dividing and growing.

Most Neuroendocrine Cancers use Ki67 as a measurement - however there are exceptions which may use Mitotic Index.

Ki67 is expressed as a percentage - of cells actively dividing and growing:

- Grade 1 = less than 3% - and cells appear well-differentiated
- Grade 2 = between 3-20% - and cells appear well-differentiated
- Grade 3 = more than 20% - cells can be either well or poorly differentiated.

Ki67 is a protein that is present during all of the active stages of the cell cycle - a useful marker of proliferation (cell division and growth) - often expressed as a percentage (%).

Mitotic Index is a measure of the number of cells dividing and growing. It is defined as the percentage of cells undergoing division in a given number of cells.

Where Neuroendocrine Cancers may grow - site & stage?

Neuroendocrine cells are found throughout the body so can start almost anywhere. Where they start is called the Primary site. Most common primary sites include the lungs, pancreas and small bowel. Cancer that has spread elsewhere - to other sites - is called Secondary or Metastatic disease.

Staging tells us whether the cancer is limited to the primary site or has spread elsewhere. There are usually 4 stages:

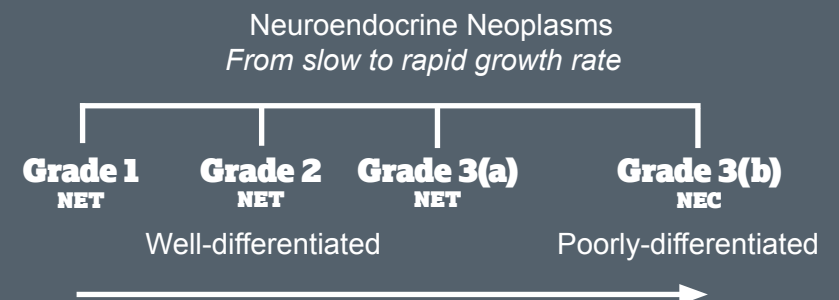
1 = confined to primary site with no evidence of spread

4 = has spread to a part of the body away from the primary site.

Stages 2 - 3 may vary in definition depending on primary site.

The Grading Spectrum of NENs

Accurate, expert histopathology review is vital to confirming diagnosis to help guide best care.



Symptoms & Diagnosis

As mentioned, neuroendocrine cells exist throughout the body and are there to produce hormones to ensure our bodies function normally.

Symptoms may be due to either the presence of the cancer itself - for example, pain, or to the abnormal excess release of hormone the cell is normally responsible for. For example, diarrhoea & flushing due to too much serotonin.

Terms you may hear include Functioning or Non-functioning.

Most Neuroendocrine Cancers are Non-functioning - so do not release excess amounts of the hormone the cell is usually responsible for.

Diagnosis can be difficult in Neuroendocrine Cancer - as not only is it rare - meaning it may not be something all healthcare professionals are aware of, but its symptoms may mimic other cancers or more common health conditions - such as IBS, asthma and menopause.



Diagnosis & Treatment

Not everyone will have symptoms (especially at early stage) and diagnosis may then only occur as a result of tests for something else or as part of a health screening programme.

More than half of those diagnosed will already have secondary spread that is Stage 4 disease at the time of diagnosis. However, this may not significantly alter life-expectancy and well-being - as both are as dependent on grading and symptom control, as it is on staging, perhaps more so. How fast or slow the cancer is growing and how well symptoms are managed, can be as important in Neuroendocrine Cancer care, as where it is growing.

Treatment can vary and may not follow a 'traditional cancer' route, for example surgery with or without chemotherapy. This is because Neuroendocrine Cancer can behave very differently to other more common cancers and therefore needs targeting in different ways. What is important, is that you have input from Neuroendocrine Cancer specialists who have the experience and knowledge of how to best

Factors that may influence treatment:

- Whether you have a NET or a NEC - and Grade
- Primary Site and Stage
- Your overall health and any other health conditions you may have
- Your personal choice and informed decision

Diagnosis, Treatment & Follow on care

There is global consensus that everyone with Neuroendocrine Cancer should be reviewed by a Neuroendocrine Cancer specialist multidisciplinary team (MDT) to promote and ensure best care.

Expert guidelines can be used and adapted to personalise your treatment to promote well-being and lifelong care.

Neuroendocrine Cancers vary - not just by appearance, grading, stage and function - but also between individuals. Further information on all of the areas covered in this factsheet are available on our website.



Neuroendocrine Cancer UK

Neuroendocrine Cancer UK exists to support and inform patients and families from diagnosis, enabling access to the best care and treatment, whilst stimulating Neuroendocrine Cancer research, increasing national awareness and influencing improvements in outcomes.

www.neuroendocrinecancer.org.uk
hello@nc-uk.org

NCUK Nurse Helpline: 0800 434 6476
NCUK Office: 01926 883 487

Registered charity number: 1092386



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