



**NeuroEndocrine  
Cancer** Australia

# **Glossary**

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Understanding the terms

# Glossary

## What does that word mean?

Use this glossary to find the meanings of important terms that are highlighted in this brochure.

**Adrenal gland:** A gland near the top of the kidney that creates hormones, like adrenaline and cortisol. Your body has 2 adrenal glands.

**Adrenocorticotrophic hormone (ACTH):** is a hormone produced in the front of the pituitary gland in the brain. Its function is to regulate levels of steroid hormone cortisol which is released from the adrenal gland

**Aggressive:** A term used to describe the speed or extent to which a tumour grows and spreads.

**Atrophic gastritis:** Chronic inflammation of the stomach mucous membrane that leads to decreased acid secretion, increased gastrin levels and enlargement of enterochromaffin-like (type of neuroendocrine cell) cells resulting in increased CgA levels and histamine production.

**Biomarkers:** a measurable indicator of the severity or presence of some disease state.

**Biopsy:** A procedure where a small amount of tissue is removed from the body and examined under a microscope or using other tests to find out if there is a tumour, its type and extent.

**Calcitonin:** A 32-amino acid peptide hormone produced by thyroid C cells that modulates blood calcium levels by opposing the effects of parathyroid hormone, which causes release of calcium from the bone. Calcitonin functions to lower blood calcium levels by inhibiting osteoclast limiting bone cell activity and reducing absorption and reabsorption of calcium in the intestine and kidney, respectively. Calcitonin is also secreted by medullary carcinomas of the thyroid and occasionally by other neuroendocrine tumours.

**Carcinoid:** Has the same meaning as NET or GEP-NET (see below). The words may be used in place of one another. The term carcinoid is an older term used before NET or GEP-NET

**Carcinoid crisis:** A potentially life-threatening heart-lung condition caused by sudden release into the systemic circulation of hormones from a NET. It may occur as a result of an event (e.g food, alcohol, exercise, or a drug administration) or during a diagnostic test. Liver directed therapies may increase the risk of carcinoid crisis. An intravenous somatostatin analogue (such as Octreotide)

and appropriate heart and blood support is often needed to address dangerous changes in heart and blood function.

**Carcinoid syndrome:** A set of symptoms that occur when a functioning NET releases the hormone serotonin. The symptoms may be wheezing, flushing, diarrhea etc and can be sudden or severe.

**Carcinoma:** Cancer that starts in the skin or in tissues that line or cover internal organs.

**Chemotherapy:** The use of drugs to destroy cancer cells.

**Chromogranin A (CgA):** Detection of elevated plasma levels of CgA has been shown to be a sensitive biomarker for neuroendocrine tumours.

**Chronic atrophic gastritis type A:** Chronic inflammation of the stomach usually caused by an autoimmune response associated with loss of parietal cells of foci of endocrine cell (ECL) enlargement

**Cushing syndrome:** Excess secretion of cortisol from the adrenal cortex, which may be secondary to hypersecretion of ACTH from the pituitary, resulting in rapid weight gain, particularly of the trunk and face with sparing of the limbs (central obesity), growth of fat pads along the collar bone and back of the neck (buffalo hump) and a round face often referred to as a 'moon face'. Other symptoms include excess sweating, thinning of the skin and bruising, fatigue, osteoporosis, and diabetes. Cushing was a pioneer in the management of pituitary tumours.

**Differentiation/differentiated cells:** In tumour cells, differentiation refers to how developed the cells are. Differentiated

tumour cells look more like normal cells. Undifferentiated or poorly differentiated tumour cells don't have the structure of normal cells, and don't work the way normal cells do. Poorly differentiated tumour cells usually have a greater chance of being faster growing and more likely to spread

**Duodenum:** The first part of the small intestine, connected to the stomach. The duodenum gets enzymes from the pancreas and chemicals from the liver and the gallbladder to help with digestion.

**Endocrine system:** A group of glands and organs that control different body functions by producing and releasing hormones.

**Functional NET:** A NET that releases hormones and may cause many different symptoms. Also called a secretory NET.

**Gastrin:** A hormone released by the pancreas that tells the stomach to produce digestive acids and enzymes.

**Gastrinoma:** A neuroendocrine tumour coming from gastric mucosa, mainly (60%) occurring the duodenum, that overproduces and secretes gastrin, thereby giving rise to the respective gastrinoma. The typical symptoms of a gastrinoma include peptic ulceration and excessive acid production (e.g. indigestion acid reflux symptoms, abdominal pain, gastrointestinal bleeding, and duodenal perforation). Secretory diarrhoea is also caused due to high gastric output in the duodenum that neutralises the pancreatic enzymes.

**Gastroenteropancreatic NET (GEP-NET):** A NET that most often starts in the gastrointestinal tract or pancreas

**Gastrointestinal (GI) tract:** Another name for the digestive system. It includes the mouth, throat, oesophagus, stomach, small intestine, large intestine, rectum and anus.

**Glucagon:** A hormone released by the pancreas that raises glucose (sugar) levels in the blood.

**Glucagonoma:** A pancreatic NET derived predominantly from alpha cells of the pancreas that secrete glucagon. The inappropriate glucagon secretion by the tumour cells can cause diabetes, mellitus, cachexia, anaemia, venous thrombosis, migratory necrolytic erythema, diarrhoea, and neuropsychiatric symptoms.

**Grade:** A system of classifying tumour cells. The cells are graded based on how they look under a microscope and how quickly the tumour is likely to grow and spread. Low-grade tumours (grades 1 and 2) look like the tissue around them. They are less aggressive. High-grade tumours (grades 3 and 4) do not look like the tissue around them. They are more aggressive.

**Hormone:** A substance, usually a protein, that is released and travels through the bloodstream to different organs. Hormones help control how some of the organs in the body work.

**Hyperglycaemia:** Having high levels of glucose (sugar) in the blood. Symptoms include dry mouth, thirst, frequent urination (including at night) blurry vision, and dry, itchy skin.

**Hypoclorhydria:** Having low levels of hydrochloric acid in the stomach. Symptoms include halitosis (bad breath), heartburn, bloating or belching, gas right after eating, and indigestion.

**Hypoglycaemia:** Low levels of glucose (sugar) in the blood. Symptoms include dizziness, headache, tiredness and confusion.

**Hypokalemia:** Low levels of potassium in your blood. It may be caused by diarrhoea. Low levels can cause muscle weakness, cramp, twitch and if severe paralysis. It can also cause abnormal heart rhythms.

**Insulin:** A hormone released by the pancreas that lowers glucose (sugar) levels in the blood.

**Insulinoma:** A pancreatic NET derived predominantly from beta cells of the pancreas that is benign in 90% of cases. The tumour oversecretes insulin, causing intermittent symptoms of hypoglycaemia (i.e., visual disturbance, irritability, bizarre behaviour, sweating, headache, tachycardia, anxiety, somnolence, paraesthesia, etc.). In more severe cases, seizures, stupor, coma and even permanent brain damage may occur.

**Lesions:** Areas of abnormal tissue that may or may not be cancerous.

**Metastasis:** To spread from one part of the body to another. The words 'localised', 'regional', and 'distant' are sometimes used to describe how much a NET has spread.

**Multiple Endocrine Neoplasia syndrome type 1:** MEN1 is a hereditary condition associated with tumours of the endocrine (hormone producing) glands. It is associated with an increased risk of developing multiple cancerous and non-cancerous tumours in glands such as the parathyroid, pituitary, and pancreas. This disorder affects approximately 1 in 30000 people.

**Non-functional NET:** A NET that doesn't release hormones. This type of NET may only cause symptoms when it grows. Also called a non-secretory NET.

**Pancreas:** An organ that produces hormones and enzymes that help your body digest food.

**Pellagra:** A condition caused by low levels of niacin (a B vitamin) in the blood. Symptoms include rash, dark pigmentation on skin, swollen mouth and bright red tongue, vomiting and diarrhoea, headache, fatigue, depression, disorientation or confusion, memory loss.

**Proteins:** The basic building blocks of tissue and other structures in the body. An enzyme is a kind of protein that causes chemical changes in the body.

**Radiation:** A form of therapy used to kill cancer cells by damaging their DNA. Radiation can damage normal cells too, so treatment should be carefully planned to decrease side effects.

**Radiology:** The use of radiation to treat or diagnose disease.

**Radiotherapy:** The use of high-energy radiation to destroy cancer cells and shrink tumours. The radiation may come from a machine outside the body or from radioactive material that is placed in the body near cancer cells.

**Serotonin:** A hormone made by certain types of cells in the body, mostly in the gastrointestinal tract. Serotonin helps with various functions, including digestion.

**Somatostatin:** A hormone that stops the release of other hormones, including gastrin, insulin, glucagon, and serotonin (see definitions for these hormones in this glossary).

**Somatostatin analogues:** Synthetic analogues of the peptide hormone somatostatin that have a longer half-life in circulation and can be used for imaging or as therapeutic agents.

**Specialised cells:** Cells that have specific jobs in the body. They start as unspecialised cells, also known as stem cells, which are present in babies still in the womb. Unspecialised cells can turn into any kind of cell. The DNA in the cell determines the kind of cells they will become. The cells then grow and change shape, becoming specialised cells.

**Syndrome:** A set of symptoms that occur together. A syndrome may be a sign of a certain disease or it may mean there's an increased chance of developing the disease. For example hypoglycaemia syndrome may be caused by a type of NET called an insulinoma, and Zollinger Ellison syndrome may be caused by a type of NET called gastrinoma.

**Thymus:** An organ near the base of the neck that produces infection-fighting cells.

**Thyroid:** A small gland in the neck, just under the skin below the Adam's apple. It produces thyroid hormones which control metabolism.

**Tumour:** An abnormal growth or mass in the body caused when cells grow out of control or don't die when they are supposed to. A tumour may be non-cancerous (benign) or cancerous (malignant).

**Tumour burden:** The number of cancer cells, size of a tumour, or the amount of cancer in a person's body.

**Ulcer:** A round sore on an external or internal surface of the body, caused by breakdown in the skin or mucous membrane which fails to heal.

**Unspecialised cells:** Also known as stem cells. These cells are present in babies still in the womb. They can turn into any kind of cell. The DNA in the cell determines the kind of cells they will become. The cells then grow and change shape, becoming specialised cells with specific jobs in the body.

**Vasoactive intestinal peptide (VIP):** A substance released by the pancreas that causes watery diarrhoea if levels are too high.

**Von Hippel-Lindau syndrome:** An inherited genetic disorder associated with renal angioma, renal cell carcinoma and pheochromocytoma (a neuroendocrine tumour of the medulla of the adrenal glands). The disorder is caused by mutations of VHL tumour suppressor gene on the short arm of chromosome 3.

**Wheezing:** A whistling sound made during breathing that happens when airways become partially blocked. Some tumours can squeeze an airway and cause the blocking. It can also be caused by the action of some hormones

**Zollinger Ellison Syndrome:** A disorder caused by excess secretion of gastrin from a duodenal or pancreatic neuroendocrine tumour resulting in excessive secretory diarrhoea and intractable peptic ulcers.





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