Neuroendocrine Tumors

SANFORD
CANCER CENTER

What Are Neuroendocrine Cells?

Neuroendocrine cells line your internal organs and help with healing after injury or infection. Neuroendocrine cells are like a nerve cell and a hormone-making cell.

What Are Neuroendocrine Tumors?

Neuroendocrine tumors can form anywhere neuroendocrine cells are in the body. Most commonly, they are in the digestive tract (stomach, appendix, small intestine, colon or rectum) or the lungs. Neuroendocrine tumors can also develop in the pancreas (pancreatic neuroendocrine tumors or PNETs). A separate guide is available for PNETs.

Carcinoid is a word to describe certain types of neuroendocrine tumors, usually of the digestive tract and the lung.

Hormones made by neuroendocrine tumors are usually destroyed by liver enzymes in the blood. If the tumor has spread to the liver, and the liver enzymes cannot destroy the extra hormones, high amounts staying in the body can cause carcinoid syndrome. This can also happen if tumor cells enter the blood. Signs and symptoms of neuroendocrine syndrome include:

- Redness or a feeling of warmth in the face and neck
- Pain in the abdomen (stomach-area)
- Feeling bloated
- Diarrhea
- Wheezing or other trouble breathing
- Fast heartbeat

All of these symptoms can be caused by other problems. It is important to report any of these to your healthcare provider.

Many people with neuroendocrine syndrome find that stress, heavy exercise, and drinking alcohol can bring on these symptoms or make them worse.

Over a long time, these hormone-like substances can damage heart valves, causing:

- Shortness of breath
- Weakness
- Heart murmur (an abnormal heart sound)

What Are the Risk Factors for Gastrointestinal Neuroendocrine Tumors?

Anything that increases the chance of getting a disease is called a risk factor. Having a risk factor does not mean that you will get cancer; not having risk factors does not mean that you will not get cancer. Risk factors for Gastrointestinal (GI) neuroendocrine tumors include:

- Having a family history of multiple endocrine neoplasia type 1 (MEN1) syndrome or neurofibromatosis type 1 (NF1) syndrome
- Having certain conditions that affect the stomach's ability to make stomach acid, such as atrophic gastritis, pernicious anemia, or Zollinger-Ellison syndrome

What Are the Signs and Symptoms of Gastrointestinal Neuroendocrine Tumors?

Some gastrointestinal neuroendocrine tumors have no signs or symptoms in the early stages. Signs and symptoms may be caused by the growth of the tumor and/or the hormones the tumor makes. Tumors of the stomach or appendix may not cause signs or symptoms. Neuroendocrine tumors are often found during tests or treatments for other conditions.

What Are the Risk Factors for Thymus (A Gland Which Produces Immune Cells) And Lung Neuroendocrine Tumors?

Anything that increases the chance of getting a disease is called a risk factor. Having a risk factor does not mean that you will get cancer; not having risk factors does not mean that you will not get cancer. Risk factors for thymus and lung neuroendocrine tumors include:

- Lung neuroendocrine tumors may occur at a younger age (45-55), more often in women than men, and are more common in whites than African Americans
- Multiple endocrine neoplasia type 1 (MEN1, an inherited syndrome

What Are the Signs and Symptoms of Thymus and Lung Neuroendocrine Tumors?

Signs and symptoms depend upon the location of the tumor, whether or not they are blocking airways, and if symptoms are caused by hormones from the tumor. These could include:

- Cough, which can sometimes be bloody
- Wheezing
- Shortness of breath
- Chest pain, especially when taking deep breaths

Carcinoid syndrome: Rarely, lung neuroendocrine tumors release enough hormone-like substances into the bloodstream to cause symptoms. This causes carcinoid syndrome. Symptoms can include:

- Facial flushing (redness and warm feeling)
- Diarrhea
- Wheezing
- Fast heartbeat

Many people with carcinoid syndrome find that stress, heavy exercise, and drinking alcohol can bring on these symptoms or make them worse. Over a long time, these hormone-like substances can damage heart valves, causing:

- Shortness of breath
- Weakness
- Heart murmur (an abnormal heart sound)

Cushing syndrome: In rare cases, lung neuroendocrine tumors may make a hormone called ACTH. This causes the adrenal glands to make too much cortisol (a steroid hormone) and other hormones. This can lead to:

- Weight gain
- Easy bruising
- Weakness
- Drowsiness
- High blood sugar (or even diabetes)
- High blood pressure
- Increased body and facial hair

What Tests Are Used To Detect (Find) And Diagnose Neuroendocrine Tumors?

The following tests and procedures may be used:

Physical exam and history

Blood chemistry studies may include hormone levels

Tumor marker test: A procedure in which a sample of blood, urine, or tissue is checked to measure the amounts of certain substances, such as chromogranin A, made by organs, tissues, or tumor cells in the body. Chromogranin A is a tumor marker. It has been linked to neuroendocrine tumors when found in increased levels in the body.

Twenty-four-hour urine test: A test in which urine is collected for 24 hours to measure the amounts of certain substances, such as 5-HIAA or serotonin (hormone). An unusual (higher or lower than normal) amount of a substance can be a sign of disease in the organ or tissue that makes it. This test is used to help diagnose neuroendocrine tumors.

Octreotide scan: A type of scan used to find neuroendocrine and other types of tumors. Radioactive octreotide, a drug similar to somatostatin, is injected into a vein and travels through the bloodstream. The radioactive octreotide attaches to tumor cells that have receptors for somatostatin. A radiation-measuring device detects the radioactive octreotide, and makes pictures showing where the tumor cells are in the body.

CT scan (CAT scan): A procedure that makes a series of detailed pictures of areas inside the body, taken from different angles. The pictures are made by a computer linked to an x-ray machine. A dye may be injected into a vein or swallowed to help the organs or tissues show up more clearly.

MRI (magnetic resonance imaging): A procedure that uses a magnet, radio waves, and a computer to make a series of detailed pictures of areas inside the body. This procedure is also called nuclear magnetic resonance imaging.

PET scan (positron emission tomography scan): A procedure to find malignant tumor cells in the body. A small amount of radioactive glucose (sugar) is injected into a vein. The PET scanner rotates around the body and makes a picture of where glucose is being used in the body. Malignant tumor cells show up brighter in the picture because they are more active and take up more glucose than normal cells.

- A Gallium-68 PET/CT Dotatate scan is being used for low (grade 1) or intermediate-grade (grade 2) GI carcinoid tumors. It uses the radioactive agent gallium-68 dotatate which attaches to the somatostatin protein on carcinoid cells.

Endoscopic ultrasound (EUS): A procedure in which an endoscope is inserted into the body, usually through the mouth or rectum. An endoscope is a thin, tube-like instrument with a light and a lens for viewing. A probe at the end of the endoscope is used to bounce high-energy sound waves (ultrasound) off internal tissues or organs, such as the stomach, small intestine, colon, or rectum, and make echoes. The echoes form a picture of body tissues called a sonogram.

Upper endoscopy: A procedure to look at organs and tissues inside the body to check for abnormal areas. An endoscope is inserted through the mouth and passed through the esophagus into the stomach. Sometimes the endoscope also is passed from the stomach into the small intestine. An endoscope is a thin, tube-like instrument with a light and a lens for viewing. It may also have a tool to remove tissue or lymph node samples, which are checked under a microscope for signs of disease.

Colonoscopy: A procedure to look inside the rectum and colon for polyps, abnormal areas, or cancer. A colonoscope is inserted through the rectum into the colon. A colonoscope is a thin, tube-like instrument with a light and a lens for viewing. It may also have a tool to remove polyps or tissue samples, which are checked under a microscope for signs of cancer.

Capsule endoscopy: A procedure used to see all of the small intestine. The patient swallows a capsule that contains a tiny camera. As the capsule moves through the gastrointestinal tract, the camera takes pictures and sends them to a receiver worn on the outside of the body.

Biopsy: The removal of cells or tissues so they can be viewed under a microscope to check for signs of cancer. Tissue samples may be taken during endoscopy and colonoscopy.

What Determines How Neuroendocrine Tumors Are Treated and Prognosis?

The prognosis (chance of recovery) and treatment options depend on the following:

- Where the tumor is located
- The size of the tumor
- The "grade" of the tumor which is how fast the abnormal cells grow and spread. This may be called low grade (grow more slowly) or high grade (grow more quickly).
- Whether the cancer has spread
- Whether the patient has neuroendocrine syndrome or has neuroendocrine heart syndrome
- Whether the cancer can be completely removed by surgery
- Whether the cancer is newly diagnosed or has recurred

Staging is the process used to find out how far the cancer has spread. The information gathered from the staging process determines the stage of the disease. The results of tests and procedures used to diagnose neuroendocrine tumors may also be used for staging. A bone scan may be done to check if there are rapidly dividing cells, such as cancer cells, in the bone. A very small amount of radioactive material is injected into a vein and travels through the bloodstream. The radioactive material collects in the bones with cancer and is detected by a scanner.

When cancer spreads to another part of the body, it is called metastasis. The metastatic tumor is the same type of tumor as the primary tumor. For example, if a gastrointestinal (GI) neuroendocrine tumor spreads to the liver, the tumor cells in the liver are actually GI neuroendocrine tumor cells. The disease is metastatic GI neuroendocrine tumor, not liver cancer.

How Are Neuroendocrine Tumors Treated?

Treatment depends on the type of tumor, where it is located, whether it is producing hormones, how fast it is growing, and if it has spread to other parts of the body.

Surgery

Treatment of GI neuroendocrine tumors usually includes surgery. One of the following surgical procedures may be used:

Endoscopic resection: Surgery to remove a small tumor that is on the inside lining of the GI tract. An endoscope is inserted through the mouth and passed through the esophagus to the stomach and sometimes, the duodenum. An endoscope is a thin, tube-like instrument with a light, a lens for viewing, and a tool for removing tumor tissue.

Local excision: Surgery to remove the tumor and a small amount of normal tissue around it.

Resection: Surgery to remove part or all of the organ that contains cancer. Nearby lymph nodes may also be removed.

Radiofrequency ablation: The use of a special probe with tiny electrodes that release high-energy radio waves (similar to microwaves) that kill cancer cells. The probe may be inserted through the skin or through an incision (cut) in the abdomen.

Treatment of neuroendocrine tumors of the lung and thymus usually include surgery. The type of surgery will depend upon the stage, location, size, and your overall health.

Radiation Therapy

Radiation therapy is a cancer treatment that uses high-energy x-rays or other types of radiation to kill cancer cells or keep them from growing.

Chemotherapy

Chemotherapy is a cancer treatment that uses drugs to stop the growth of cancer cells, either by killing the cells or by stopping them from dividing.

Hormone Therapy

Hormone therapy is a treatment that stops extra hormones from being made. These include hormones to lessen flushing and diarrhea. Some may also help to slow tumor growth.

Treatment for Carcinoid Syndrome May Also Be Needed:

- Hormone therapy to help lessen flushing and diarrhea
- Medicine for diarrhea
- Skin rash medicine
- Medicines to breathe easier
- Taking medicine before having anesthesia for a medical procedure
- Avoiding things that cause flushing or difficulty breathing such as alcohol, nuts, certain cheeses and foods with capsaicin, such as chili peppers
- Avoiding stressful situations and certain types of physical activity can also help treat neuroendocrine syndrome
- For some patients with carcinoid heart syndrome, a heart valve replacement may be done

Follow-up Tests May Be Needed

Some of the tests that were done to diagnose the cancer or to find out the stage of the cancer may be repeated. Some tests will be repeated in order to see how well the treatment is working. Decisions about whether to continue, change, or stop treatment may be based on the results of these tests.

Support is available for coping with changes that may have happened because of cancer treatment. Your healthcare team can offer ideas as well as a plan of care for long-term follow-up.

Clinical Trials

Clinical trials are done to find out if new cancer treatments are safe and effective or better than the standard treatment.

People who take part in a clinical trial may receive:

- The standard drugs alone or
- The standard drugs plus the new treatment being studied

Taking part in a clinical trial helps improve the way cancer will be treated in the future. Even when clinical trials do not lead to effective new treatments, they often answer important questions and help move research forward.

Some clinical trials only include people who have not yet received treatment. Other trials test treatments for those whose cancer has not gotten better. There are also clinical trials that test new ways to stop cancer from coming back or reduce the side effects of cancer treatment.

Many of today's standard treatments for cancer are based on earlier clinical trials.

Ask if there is a clinical trial right for you.

My Health Care Team	Contact Information
Surgeon:	
Medical Oncologist:	
Radiation Oncologist:	
Primary Care Doctor:	
Navigator:	
Nurse:	
Registered Dietitian Nutritionist:	
Other	

Common Questions

What does the pathology report say?

What is the stage of my cancer?

What are my goals for treatment?

What are my treatment choices?

What kind of support services are available for me about finances, emotions, spiritual questions, etc.?

To Learn More About Neuroendocrine Tumors:

American Cancer Society
https://www.cancer.org/

National Cancer Institute
https://www.cancer.gov/

National Comprehensive Cancer Network Guidelines for Patients
https://www.nccn.org/patients/guidelines/cancers.aspx

MedlinePlus
https://medlineplus.gov/

Adapted Adapted from: PDQ Carcinoid Tumors Treatment.

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