



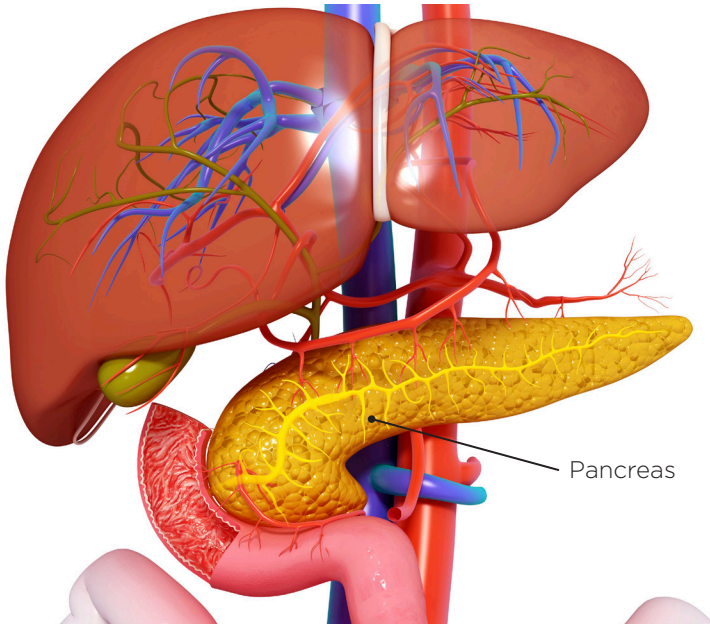
# **Pancreatic Neuroendocrine Tumors**

(NETs)



# About the Pancreas

The pancreas is a gland about 6 inches long that is shaped like a thin pear lying on its side. The wider end of the pancreas is called the head, the middle section is called the body, and the narrow end is called the tail. The pancreas lies between the stomach and the spine.



## The Pancreas Has Two Main Jobs in the Body

- To make juices that help digest (break down) food.
- To make hormones, such as insulin and glucagon, that help control blood sugar levels. Both of these hormones help the body use and store the energy it gets from food.

## What Are Different Types of Pancreatic Tumors?

One type, pancreatic neuroendocrine tumors (NETs), begins in the endocrine cells which make some hormones. NETs may be benign (not cancer) or malignant (cancer).

Pancreatic NETs are much less common than pancreatic exocrine tumors and have a better prognosis. Endocrine pancreas cells:

- Make several kinds of hormones (chemicals that control the actions of certain cells or organs in the body), such as insulin to control blood sugar.
- Cluster together in many small groups (islets) throughout the pancreas. These are called islet cells or islets of Langerhans.

Another type of pancreatic tumor begins in the exocrine cells that make digestive juices. Pancreatic cancers that begin in the exocrine cells occur far more often than NETs. The most common type of pancreatic cancer is adenocarcinoma.

## Pancreatic NETs May or May Not Cause Signs or Symptoms

Pancreatic NETs may be functional or nonfunctional:

- Functional tumors make extra amounts of hormones, such as gastrin, insulin, and glucagon, that cause signs and symptoms. The signs and symptoms of a functional pancreatic NET depend on the type of hormone being made.
- Nonfunctional tumors do not make extra amounts of hormones. Signs and symptoms are caused by the tumor as it spreads and grows. Most nonfunctional tumors are malignant (cancer).

## There Are Different Kinds of Functional Pancreatic NETs

Pancreatic NETs make different kinds of hormones such as gastrin, insulin, and glucagon. Functional pancreatic NETs include the following:

- **Gastrinoma:** A tumor that forms in cells that make gastrin. Gastrin is a hormone that causes the stomach to release an acid that helps digest food. Both gastrin and stomach acid are increased by gastrinomas. When increased stomach acid, stomach ulcers, and diarrhea are caused by a tumor that makes gastrin, it is called Zollinger-Ellison syndrome. A gastrinoma usually forms in the head of the pancreas and sometimes forms in the small intestine. Most gastrinomas are malignant (cancer).
- **Insulinoma:** A tumor that forms in cells that make insulin. Insulin is a hormone that controls the amount of glucose (sugar) in the blood. It moves glucose into the cells, where it can be used by the body for energy. Insulinomas are usually slow-growing tumors that rarely spread. An insulinoma forms in the head, body, or tail of the pancreas. Insulinomas are usually benign (not cancer).
- **Glucagonoma:** A tumor that forms in cells that make glucagon. Glucagon is a hormone that increases the amount of glucose in the blood. It causes the liver to break down glycogen. Too much glucagon causes hyperglycemia (high blood sugar). A glucagonoma usually forms in the tail of the pancreas. Most glucagonomas are malignant (cancer).
- **Other types of tumors:** There are other rare types of functional pancreatic NETs that make hormones, including hormones that control the balance of sugar, salt, and water in the body. These tumors include:
  - Vipomas, which make vasoactive intestinal peptide. Vipoma may also be called Verner-Morrison syndrome.
  - Somatostatinomas, which make somatostatin.

## Having Certain Syndromes Can Increase the Risk of Pancreatic NETs

Anything that increases your risk 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 doesn't mean that you will not get cancer. Talk with your doctor if you think you may be at risk.

Multiple endocrine neoplasia type 1 (MEN1) syndrome is a risk factor for pancreatic NETs.

### Screening

Approved screening tests help find a pre-cancer condition or cancer at an early stage. There is no approved screening test for cancer of the pancreas or PNETs.

## Different Types of Pancreatic NETs Have Different Signs and Symptoms

Signs or symptoms can be caused by the growth of the tumor and/or by hormones the tumor makes or by other conditions. Some tumors may not cause signs or symptoms. Check with your doctor if you have any of these problems.

## Signs and Symptoms of a Non-Functional Pancreatic NET

A non-functional pancreatic NET may grow for a long time without causing signs or symptoms. It may grow large or spread to other parts of the body before it causes signs or symptoms, such as:

- Diarrhea
- Indigestion
- A lump in the abdomen
- Yellowing of the skin and whites of the eyes

## Signs and Symptoms of a Functional Pancreatic NET

The signs and symptoms of a functional pancreatic NET depend on the type of hormone being made.

**Too much gastrin may cause:**

- Stomach ulcers that keep coming back
- Pain in the abdomen, which may spread to the back. The pain may come and go and it may go away after taking an antacid
- The flow of stomach contents back into the esophagus (gastroesophageal reflux)
- Diarrhea

**Too much insulin may cause:**

- Low blood sugar: This can cause blurred vision, headache, and feeling lightheaded, tired, weak, shaky, nervous, irritable, sweaty, confused, or hungry
- Fast heartbeat

**Too much glucagon may cause:**

- Skin rash on the face, stomach, or legs
- High blood sugar. This can cause headaches, frequent urination, dry skin and mouth, or feeling hungry, thirsty, tired, or weak
- Blood clots. Blood clots in the lung can cause shortness of breath, cough, or pain in the chest. Blood clots in the arm or leg can cause pain, swelling, warmth, or redness of the arm or leg
- Diarrhea
- Weight loss for no known reason
- Sore tongue or sores at the corners of the mouth

### Too much vasoactive intestinal peptide (VIP) may cause:

- Very large amounts of watery diarrhea
- Dehydration. This can cause feeling thirsty, making less urine, dry skin and mouth, headaches, dizziness, or feeling tired
- Low potassium level in the blood. This can cause muscle weakness, aching, or cramps, numbness and tingling, frequent urination, fast heartbeat, and feeling confused or thirsty
- Cramps or pain in the abdomen
- Weight loss for no known reason

### Too much somatostatin may cause:

- High blood sugar. This can cause headaches, frequent urination, dry skin and mouth, or feeling hungry, thirsty, tired, or weak
- Diarrhea
- Steatorrhea (very foul-smelling stool that floats)
- Gallstones
- Yellowing of the skin and whites of the eyes
- Weight loss for no known reason

## Tests to Find, Diagnose, and Monitor Pancreatic Disorders

Not every person needs every test or procedure.

### Physical exam and history.

**Blood chemistry studies:** A blood sample is checked to measure substances such as bilirubin. A higher or lower amount of a substance can be a sign of disease.

- Chromogranin A: A higher than normal amount of Chromogranin A in the blood and normal amounts of hormones can be a sign of a non-functional pancreatic NET.
- Other kinds of lab tests: used to check for the specific type of pancreatic NETs.

**Endoscopic ultrasound (EUS):** A procedure in which a lighted scope is inserted into the body, usually through the mouth. A probe at the end of the scope is used to bounce sound waves (ultrasound) to see structures near the pancreas.

**Biopsy:** The removal of cells or tissues so they can be viewed under a microscope by a pathologist to check for signs of cancer.

**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.

**Octreotide scan (somatostatin receptor scintigraphy):** A type of scan that may be used to find small pancreatic NETs. A hormone with a radioactive agent can be given in a vein and scanning provides a picture of where tumors may be in the body.

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

**PET scan (positron emission tomography scan):** 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. Cancer cells show up brighter in the picture because they are more active and take up more glucose than normal cells do.

**Abdominal ultrasound:** An ultrasound exam uses sound waves to make pictures of the inside of the abdomen.

**Endoscopic retrograde cholangiopancreatography (ERCP):**  
A procedure used to x-ray the ducts (tubes) that carry bile from the liver to the gallbladder and from the gallbladder to the small intestine. Sometimes pancreatic cancer causes these ducts to narrow and block or slow the flow of bile, causing jaundice. A lighted scope is passed through the mouth, esophagus, and stomach into the first part of the small intestine. If jaundice is caused by a tumor, a fine tube may be inserted into the duct to unblock it. This tube (or stent) may be left in place to keep the duct open.

**Percutaneous transhepatic cholangiography (PTC):** PTC is a way to x-ray the liver and bile ducts. A thin needle is inserted through the skin below the ribs and into the liver. Dye is injected into the liver or bile ducts and an x-ray is taken. If a blockage is found, a thin, flexible tube called a stent is sometimes left in the liver to drain bile into the small intestine or a collection bag outside the body. This test is done only if ERCP cannot be done.

**Laparoscopy:** A surgery using a lighted scope to look at the organs inside the abdomen to check for signs of disease.

**Bone scan:** A procedure 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 blood. The radioactive material collects in bones where cancer cells have spread and is detected by a scanner.

## Certain Factors Affect Prognosis (Chance of Recovery) and Treatment Options

Pancreatic NETs can often be cured. The prognosis (chance of recovery) and treatment options depend on the following:

- The type of cancer cell
- Where the tumor is found in the pancreas
- Whether the tumor has spread to more than one place in the pancreas or to other parts of the body
- Whether the patient has MEN1 syndrome
- The patient's age and general health
- Whether the cancer has just been diagnosed or has recurred (come back)

# How Cancer Spreads

The process used to find out if a cancer has spread is called staging. After cancer has been diagnosed, tests are done to find out if cancer cells have spread within the area or to other parts of the body.

There are 3 ways that cancer spreads in the body. Cancer can spread through tissue, the lymph system, and the blood:

- **Tissue:** The cancer spreads from where it began by growing into nearby areas.
- **Lymph system:** The cancer spreads from where it began by getting into the lymph system. The cancer travels through the lymph vessels to other parts of the body.
- **Blood:** The cancer spreads from where it began by getting into the blood. The cancer travels through the blood vessels to other parts of the body.

The process used to find out where the cancer is located is called staging. PNETs may be found in 1 place in the pancreas, in more than 1 place, or to lymph nodes near the pancreas or to other parts of the body such as the liver, lung, peritoneum, or bone.

The distant spread of cancer, called the metastatic tumor, is the same type of cancer as the primary tumor. For example, if lung cancer spreads to the liver, the cancer cells in the liver are actually lung cancer cells. The disease is metastatic lung cancer, not liver cancer.

## Recurrent Pancreatic Neuroendocrine Tumors

Recurrent pancreatic neuroendocrine tumors are those that come back after treatment. The tumors may come back in the pancreas or other parts of the body.

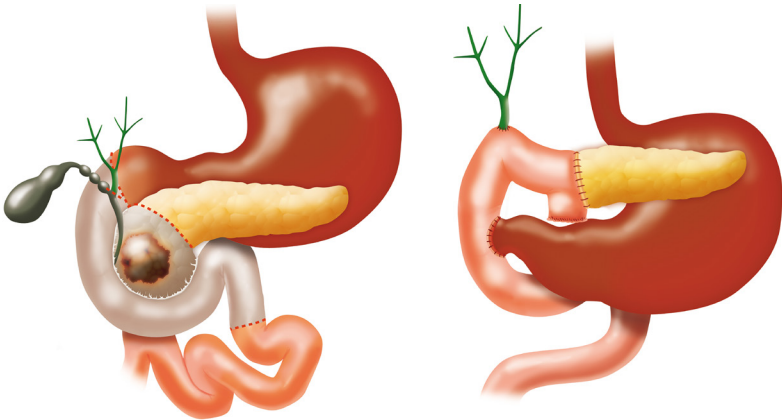
# Treatment Option Overview

Different types of treatments are available for patients with pancreatic neuroendocrine tumors (NETs).

## Surgery

One of the following types of surgery may be used to take out the tumor:

- **Enucleation:** Surgery to remove the tumor only.
- **Whipple procedure:** A surgical procedure in which the head of the pancreas, the gallbladder, part of the stomach, part of the small intestine, and the bile duct are removed. Enough of the pancreas is left to produce digestive juices and insulin.



- **Distal pancreatectomy:** The body and the tail of the pancreas and usually the spleen are removed.
- **Total gastrectomy:** Surgery to remove the whole stomach.
- **Parietal cell vagotomy:** Surgery to cut the nerve that causes stomach cells to make acid.
- **Liver Resection:** Surgery to remove part or all of the liver.
- **Radiofrequency ablation:** The use of a special probe with tiny electrodes that kill cancer cells.

## 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. When chemotherapy is taken by mouth or injected into a vein or muscle, the drugs enter the bloodstream and can reach cancer cells throughout the body (systemic chemotherapy). The way the chemotherapy is given depends on the type and stage of the cancer being treated.

## Hormone Therapy

Hormone therapy is a cancer treatment that removes hormones or blocks their action and stops cancer cells from growing. Hormones are substances made by glands in the body and circulated in the bloodstream. Some hormones can cause certain cancers to grow. If tests show that the cancer cells have places where hormones can attach (receptors), drugs, surgery, or radiation therapy is used to reduce the production of hormones or block them from working.

## Hepatic Arterial Occlusion or Chemoembolization

Hepatic arterial occlusion uses drugs, small particles, or other agents to block or reduce the flow of blood to the liver through the hepatic artery (the major blood vessel that carries blood to the liver). This is done to kill cancer cells growing in the liver. Chemotherapy delivered during hepatic arterial occlusion is called chemoembolization.

## Targeted Therapy

Targeted therapy is a type of treatment that uses drugs or other substances to identify and attack specific cancer cells without harming normal cells.

## Supportive Care

Supportive care is given to lessen the problems caused by the disease or its treatment. Supportive care for pancreatic NETs may include treatment for the following:

- Stomach ulcers may be treated with drug therapy.
- Diarrhea may be treated with intravenous (IV) fluids or medicines.
- Blood sugars may be managed by diet or drug therapy.

## 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.**

# As Treatment Progresses

Some tests may be repeated to see how well your treatment is working. Decisions about whether to continue, change, or stop treatment may be based on the results of these tests.

A plan for your long term care will be discussed with your treatment team and shared with your primary care provider.

## Learn More About Pancreatic NETs

### **American Cancer Society**

[www.cancer.org](http://www.cancer.org)

### **National Cancer Institute**

[www.cancer.gov](http://www.cancer.gov)

### **National Comprehensive Cancer Network Guidelines for Patients**

[www.nccn.org/patients/guidelines/cancers.aspx](http://www.nccn.org/patients/guidelines/cancers.aspx)

### **MedlinePlus**

[medlineplus.gov](http://medlineplus.gov)

### **Pancreatic Cancer Action Network**

<https://www.pancan.org/>

# Common Questions

What do my test results tell me?

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.?

My Health Care Team	Contact Information
Gastroenterologist:	
Navigator:	
Pharmacy:	
Medical Oncologist:	
Endocrinologist:	
Dietitian/Nutritionist:	
Surgeon:	
Primary Care Doctor:	
Counselor/Therapist:	
Other:	

## Notes

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