CONDITION

Pancreatic neuroendocrine tumors

Pancreatic neuroendocrine tumors are growths that begin in the pancreas's hormone-producing cells.

WHAT IT IS
Tumors of hormone-producing pancreatic cells

BODY SYSTEM
Digestive and hormone systems

MAIN SYMPTOMS
Abdominal pain, weight loss, diarrhea, low blood sugar

WHAT CAUSES IT
Usually unknown, sometimes inherited

HOW IT IS DIAGNOSED
Biopsy, imaging scans, and hormone blood tests

MAIN TREATMENT
Surgery, medicines, targeted radiation, or monitoring

CAN IT BE CURED?
Sometimes - localized tumors can be removed

IS IT AN EMERGENCY?
Sometimes - severe low blood sugar causes confusion or fainting

LONG-TERM OUTLOOK
Often slow-growing, shaped by grade and spread

Overview

Pancreatic neuroendocrine tumors, often shortened to pancreatic NETs or pNETs, are growths that begin in the neuroendocrine cells of the pancreas. These cells make hormones that help control digestion and blood sugar.

Pancreatic NETs are different from the more common type of pancreatic cancer, called pancreatic ductal adenocarcinoma. They start in different cells, behave differently, and are treated differently. Many pancreatic NETs grow slowly, although some are more aggressive.

A pancreatic NET may be functional , meaning it releases enough hormone to cause symptoms. A nonfunctional tumor does not cause a clear hormone-related syndrome. Nonfunctional tumors can still make small amounts of hormones, but not enough to produce typical symptoms.

Doctors describe these tumors by their size, location, grade, and stage. The grade reflects how abnormal the cells look and how quickly they are dividing. The stage describes the tumor's size and whether it has spread. These details help guide treatment and give a clearer picture of what to expect.

Symptoms

Some pancreatic NETs are found during a scan performed for another reason. Others cause symptoms because of their size, spread, or hormone production.

Nonfunctional tumors may cause:

Functional tumors cause different symptoms depending on the hormone involved.

An insulinoma releases insulin, which lowers blood sugar. Symptoms can include sweating, shaking, hunger, weakness, confusion, blurred vision, or fainting. Symptoms may occur after fasting or exercise and may improve after eating.

A gastrinoma releases gastrin, which increases stomach acid. It can cause repeated stomach or small-intestine ulcers, burning abdominal pain, acid reflux, or ongoing diarrhea.

A glucagonoma may cause high blood sugar, weight loss, blood clots, and a distinctive red, blistering, or scaly rash. A VIPoma can cause frequent watery diarrhea, dehydration, and low potassium. A somatostatinoma may be linked with diabetes, diarrhea, fatty stools, and gallstones.

These symptom patterns can also have more common causes. Having one or more of them does not mean you have a pancreatic NET.

- Ongoing pain or pressure in the upper abdomen
- Unexplained weight loss or reduced appetite
- Nausea, vomiting, or feeling full quickly
- Yellow skin or eyes if a bile duct becomes blocked

Causes and risk factors

For most people, the exact cause is unknown. A tumor develops when changes in a neuroendocrine cell's DNA allow that cell to grow and divide without normal control.

A small proportion of pancreatic NETs are related to inherited genetic conditions. These include:

An inherited condition may be considered if you develop several neuroendocrine tumors, are diagnosed at a young age, or have relatives with related tumors. Genetic counseling can help you understand what testing could mean for you and your family.

Most pancreatic NETs occur in people without an inherited syndrome or a clear avoidable risk factor.

- Multiple endocrine neoplasia type 1, or MEN1
- Von Hippel-Lindau syndrome
- Neurofibromatosis type 1
- Tuberous sclerosis complex

Diagnosis

Diagnosis usually begins with a review of your symptoms, medical history, and family history. Blood tests may check blood sugar, liver function, and general health.

If symptoms suggest a functional tumor, blood or urine tests may measure specific hormones or substances related to them. These tests often need careful preparation because food, medicines, and other health conditions can affect the results.

Imaging shows where the tumor is and whether it has spread. Tests may include CT or MRI scans. An endoscopic ultrasound uses a flexible tube passed through the mouth into the stomach and small intestine. An ultrasound probe on the tube creates detailed images of the nearby pancreas.

Some tumors have proteins called somatostatin receptors on their surface. A somatostatin receptor PET scan can detect these proteins using a small amount of radioactive tracer. The result can also help determine whether receptor-targeted treatments may work.

A biopsy removes a small sample of tissue for examination. A pathologist looks at the cells and measures how actively they are dividing. One measure, called the Ki-67 index , helps determine the tumor's grade.

Well-differentiated tumors resemble normal neuroendocrine cells and often grow more slowly. Poorly differentiated neuroendocrine carcinomas look very abnormal and usually behave more aggressively. This distinction affects treatment.

Treatment and management

Treatment is planned around the tumor's size, location, grade, stage, hormone production, and somatostatin receptor status. Your general health and treatment preferences also matter.

Surgery offers the best chance of cure when a tumor can be removed completely. Depending on its position, surgery may remove only the tumor, the left part of the pancreas, or the head of the pancreas and nearby structures. Pancreatic surgery can affect digestion or blood sugar, so its possible benefits and risks need careful discussion.

For some small, low-grade tumors that are not causing symptoms, active monitoring may be considered. This means having planned scans and appointments rather than beginning treatment immediately. Monitoring is an active treatment strategy, not a lack of care.

Treatment for tumors that cannot be fully removed may include:

Medicines may also control the effects of excess hormones. For example, treatment may focus on preventing low blood sugar, reducing stomach acid, or replacing fluids lost through diarrhea.

Treatment may change over time if the tumor grows, symptoms change, or side effects become difficult. Care often involves specialists in cancer treatment, hormone conditions, surgery, radiology, nutrition, and symptom control.

- Somatostatin analogue medicines to slow growth or control hormone symptoms
- Targeted medicines that interfere with specific tumor growth pathways
- Peptide receptor radionuclide therapy, which delivers radiation to receptor-positive cells
- Chemotherapy, especially for faster-growing or higher-volume tumors
- Liver-directed procedures when much of the tumor burden is in the liver

Complications

A pancreatic NET can spread to nearby lymph nodes or other organs. The liver is a common site of spread. Tumors that spread can still sometimes be controlled for long periods, especially when they are well differentiated and slow-growing.

Hormone-producing tumors can cause complications even when they are small. Severe low blood sugar can affect the brain and lead to seizures or loss of consciousness. Persistent diarrhea can cause dehydration, kidney problems, or abnormal salt levels in the blood. Excess stomach acid can lead to bleeding or a hole in the stomach or intestine.

Other possible complications include blood clots, poor nutrition, blockage of the bile duct or intestine, and pain from growing tumors.

Pancreatic surgery can reduce the production of digestive enzymes or insulin. Some people need enzyme capsules with food, treatment for diabetes, or both after surgery.

Prevention

Most pancreatic NETs cannot be prevented because their cause is usually unknown. There is no established lifestyle change or screening test that prevents a sporadic pancreatic NET.

Inherited genetic conditions also cannot be prevented. If one runs in your family, genetic counseling and planned surveillance may help find related tumors earlier. This is early detection rather than prevention.

Avoiding tobacco and maintaining general health remain worthwhile, but they have not been shown to guarantee prevention of pancreatic NETs.

Living with it and what to expect

The course over months and years differs widely. A small, low-grade tumor may remain stable for a long time. A high-grade tumor may change more quickly and need prompt treatment. Stage, grade, tumor biology, and response to treatment are more informative than the name alone.

After surgery, you will usually have follow-up for possible recurrence. Follow-up may include scans, blood tests, and review of hormone-related symptoms. Because some tumors return years later, monitoring may continue long term.

If the tumor cannot be cured, treatment often aims to slow its growth, control hormone symptoms, and preserve day-to-day function. Some people have several different treatments over the course of their condition.

You may need practical adjustments. Regular meals can be important for people prone to low blood sugar. Persistent diarrhea may affect hydration, travel, and work. Pancreatic enzyme problems can lead to oily stools, weight loss, and difficulty absorbing nutrients, but replacement enzymes may help.

Most people benefit from knowing the tumor's exact type, grade, stage, and hormone status. These details make general survival statistics less misleading and help explain why another person's experience may be very different from yours.

When to get medical help

Arrange a routine medical appointment if you have persistent upper abdominal pain, unexplained weight loss, repeated ulcers, ongoing diarrhea, or episodes of sweating and shaking that improve after eating. These symptoms often have other causes, but they deserve assessment.

Contact your care team promptly if you have a diagnosed pancreatic NET and develop worsening diarrhea, new jaundice, increasing pain, repeated low blood sugar episodes, or difficulty eating and drinking.

Seek emergency help for:

Do not drive yourself if you are confused, faint, or think your blood sugar is severely low. If you already have an emergency plan for hormone-related symptoms, follow that plan while help is arranged.

- Confusion, a seizure, fainting, or unconsciousness linked with low blood sugar
- Vomiting blood or passing black, tar-like stools
- Severe weakness or fainting with ongoing watery diarrhea
- Sudden shortness of breath, chest pain, or one-sided leg swelling

Common questions

Is a pancreatic neuroendocrine tumor the same as pancreatic cancer?

It is a type of pancreatic tumor, but it is not the same as pancreatic ductal adenocarcinoma, the most common pancreatic cancer. Pancreatic NETs begin in hormone-producing cells and often have different treatments and a slower course.

Can a pancreatic neuroendocrine tumor be benign?

Some remain localized and cause few problems for many years. However, pancreatic NETs are generally considered to have some potential to spread. Grade, size, invasion into nearby tissue, and evidence of spread give more useful information than the word "benign."

How fast do pancreatic neuroendocrine tumors grow?

Many well-differentiated, low-grade tumors grow slowly. High-grade tumors and poorly differentiated neuroendocrine carcinomas can grow much faster. Repeat scans and the Ki-67 index help estimate the tumor's behavior.

Can a blood test detect a pancreatic neuroendocrine tumor?

Blood tests can identify excess hormones from some functional tumors, but they cannot

reliably detect or rule out every pancreatic NET. Imaging and, in many cases, a biopsy are needed to confirm and classify the tumor.

Can pancreatic neuroendocrine tumors be cured?

Sometimes. Surgery may cure a localized tumor that can be removed completely. If it has spread, treatment is more often aimed at long-term control, symptom relief, and slowing further growth.

What does Ki-67 mean in my pathology report?

Ki-67 is a marker of how many tumor cells are actively dividing. A lower percentage usually suggests slower growth, while a higher percentage suggests faster growth. It is interpreted together with the tumor's appearance, stage, and scan results.

Sources

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