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# About Pancreatic Neuroendocrine Tumors

## Overview and Types

If you have been diagnosed with a pancreatic neuroendocrine tumor or are worried about it, you likely have a lot of questions. Learning some basics is a good place to start.

- [What Is a Pancreatic Neuroendocrine Tumor?](#)

## Research and Statistics

See the latest estimates for new cases of pancreatic neuroendocrine tumors and deaths in the US and what research is currently being done.

- [Key Statistics for Pancreatic Neuroendocrine Tumor](#)
- [What's New in Pancreatic Neuroendocrine Tumor Research?](#)

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# What Is a Pancreatic Neuroendocrine Tumor?

Pancreatic neuroendocrine tumors (NETs), or islet cell tumors, are a type of cancer that starts in the pancreas. (Cancer starts when cells in the body begin to grow out of

control. To learn more about how cancers start and spread, see [What Is Cancer?](#)<sup>1)</sup>

Pancreatic NETs are a less common type of pancreatic cancer. They make up less than 2% of pancreatic cancers, but tend to have a better outlook (prognosis) than [the more common type](#)<sup>2</sup>.

## Where pancreatic neuroendocrine tumors start

Pancreatic neuroendocrine tumors start in neuroendocrine cells, a special kind of cell found in the pancreas. Neuroendocrine cells are also found in other areas of the body, but only cancers that form from neuroendocrine cells in the pancreas are called pancreatic neuroendocrine tumors.

### The neuroendocrine system

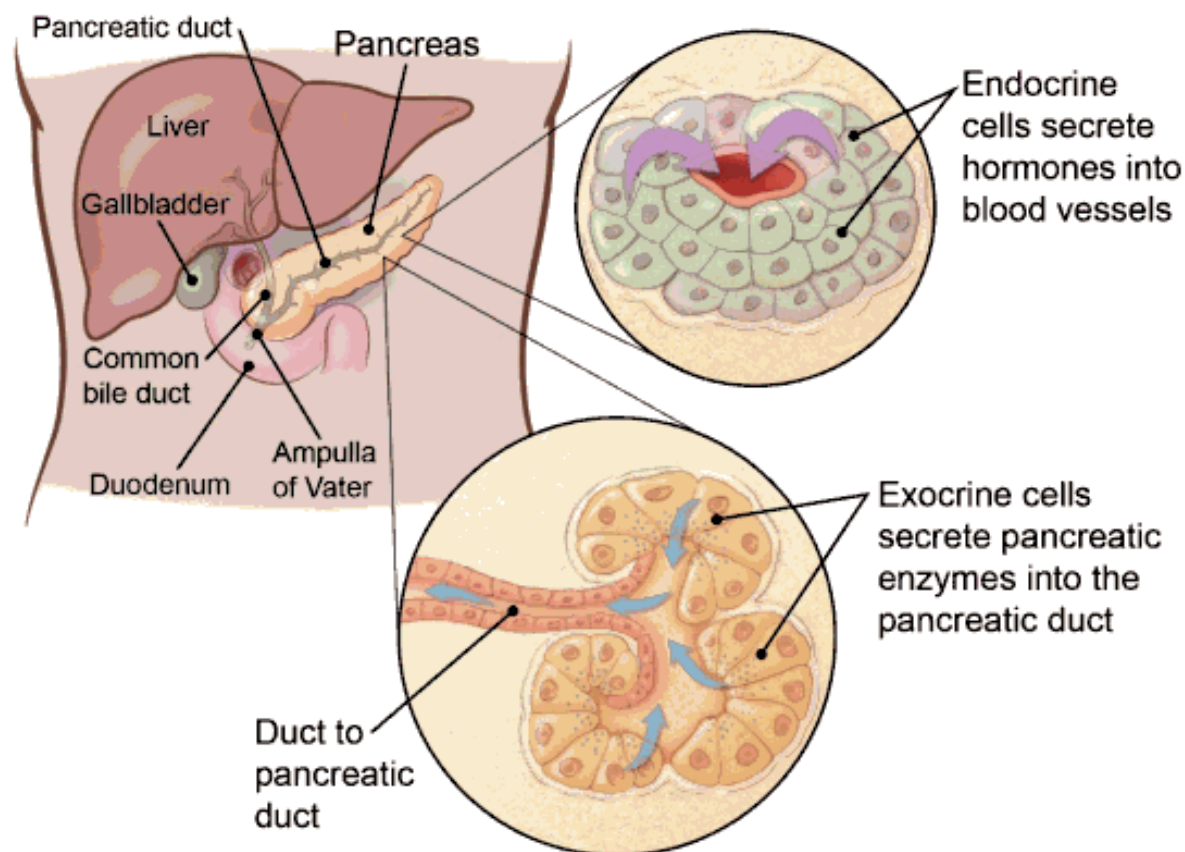
Neuroendocrine cells are like nerve cells in some ways and like hormone-making endocrine cells in other ways. Cells in this system don't form actual organs. Instead, they are scattered throughout other organs like the esophagus, stomach, pancreas, intestines, and lungs.

Neuroendocrine cells (sometimes just called *endocrine cells*) in the pancreas are found in small clusters called *islets* (or *islets of Langerhans*). These islets make important hormones like insulin and glucagon (which help control blood sugar levels), and release them directly into the blood.

### The pancreas

The pancreas is an organ that sits behind the stomach. It's shaped a bit like a fish with a wide head, a tapering body, and a narrow, pointed tail. In adults it's about 6 inches (15 centimeters) long but less than 2 inches (5 centimeters) wide.

- The head of the pancreas is on the right side of the abdomen (belly), behind where the stomach meets the duodenum (the first part of the small intestine).
- The body of the pancreas is behind the stomach.
- The tail of the pancreas is on the left side of the abdomen next to the spleen.



Neuroendocrine tumors start in the *endocrine* cells of the pancreas. But most of the pancreas is actually made up of another type of cell called *exocrine* cells. These cells form the exocrine glands and ducts. The exocrine glands make pancreatic enzymes that are released into the intestines to help you digest foods (especially fats). The most common type of pancreatic cancer, adenocarcinoma of the pancreas, starts from exocrine cells. See [Pancreatic Cancer](#)<sup>3</sup> for more about this type.

**If you are diagnosed with pancreatic cancer, it's very important to know if it's an *exocrine* cancer (see [Pancreatic Cancer](#)<sup>4</sup>) or *endocrine* cancer (discussed here). They have distinct risk factors and causes, have different signs and symptoms, are diagnosed with different tests, are treated in different ways, and have different outlooks.**

## Types of pancreatic neuroendocrine tumors

### Tumor grade

Pancreatic neuroendocrine tumors (NETs) are classified by tumor grade, which

describes how quickly the cancer is likely to grow and spread.

- **Grade 1** (also called *low-grade or well-differentiated*) neuroendocrine tumors have cells that look more like normal cells and are not multiplying quickly.
- **Grade 2** (also called *intermediate-grade or moderately differentiated*) tumors have features in between those of low- and high-grade (see below) tumors.
- **Grade 3** (also called *high-grade or poorly differentiated*) neuroendocrine tumors have cells that look very abnormal and are multiplying faster.

Cancers that are grade 1 or 2 are called **pancreatic neuroendocrine tumors**. These cancers tend to grow slowly and can possibly spread to other parts of the body.

Cancers that are grade 3 are called **pancreatic neuroendocrine carcinomas (NECs)**. These cancers tend to grow and spread quickly and can spread to other parts of the body.

Another important part of grading is measuring how many of the cells are in the process of dividing into new cells. This is described in more detail in [Pancreatic Neuroendocrine Tumor Stages](#)<sup>5</sup>.

## Tumor function

Pancreatic NETs are also named based on whether they are *functioning* (making hormones that cause symptoms) or *non-functioning* (not making hormones).

**Functioning NETs:** About half of pancreatic NETs make hormones that are released into the blood and cause [symptoms](#)<sup>6</sup>. These are called functioning NETs. Each one is named for the [type of hormone](#)<sup>7</sup> the tumor cells make.

- **Gastrinomas** come from cells that make gastrin.
- **Insulinomas** come from cells that make insulin.
- **Glucagonomas** come from cells that make glucagon.
- **Somatostatinomas** come from cells that make somatostatin.
- **VIPomas** come from cells that make vasoactive intestinal peptide (VIP).

Most functioning NETs are gastrinomas or insulinomas. The other types are rare.

**Non-functioning NETs:** These tumors don't make enough excess hormones to cause symptoms. Because they don't make excess hormones that cause symptoms, they can often grow quite large before they're found. Symptoms that may occur when they grow

to a large size include abdominal (belly) pain, lack of appetite, and weight loss.

**Carcinoid tumors:** These NETs are much more common in other parts of the [digestive system](#)<sup>8</sup>, although rarely they can start in the pancreas. These tumors often make serotonin.

The treatment and outlook for pancreatic NETs depend on the specific tumor type and the [stage \(extent\) of the tumor](#)<sup>9</sup>, but the outlook is generally better than for pancreatic exocrine cancers.

## Hyperlinks

1. [www.cancer.org/cancer/cancer-basics/what-is-cancer.html](http://www.cancer.org/cancer/cancer-basics/what-is-cancer.html)
2. [www.cancer.org/cancer/pancreatic-cancer.html](http://www.cancer.org/cancer/pancreatic-cancer.html)
3. [www.cancer.org/cancer/pancreatic-cancer.html](http://www.cancer.org/cancer/pancreatic-cancer.html)
4. [www.cancer.org/cancer/pancreatic-cancer.html](http://www.cancer.org/cancer/pancreatic-cancer.html)
5. [www.cancer.org/cancer/pancreatic-neuroendocrine-tumor/detection-diagnosis-staging/net-staging.html](http://www.cancer.org/cancer/pancreatic-neuroendocrine-tumor/detection-diagnosis-staging/net-staging.html)
6. [www.cancer.org/cancer/pancreatic-neuroendocrine-tumor/detection-diagnosis-staging/signs-and-symptoms.html](http://www.cancer.org/cancer/pancreatic-neuroendocrine-tumor/detection-diagnosis-staging/signs-and-symptoms.html)
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8. [www.cancer.org/cancer/gastrointestinal-carcinoid-tumor.html](http://www.cancer.org/cancer/gastrointestinal-carcinoid-tumor.html)
9. [/content/cancer/en/cancer/pancreatic-neuroendocrine-tumor/detection-diagnosis-staging/staging.html](http://content.cancer/en/cancer/pancreatic-neuroendocrine-tumor/detection-diagnosis-staging/staging.html)

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## Key Statistics for Pancreatic Neuroendocrine Tumor

Pancreatic neuroendocrine tumors (NETs) are rare and account for less than 2% of all cancers that occur in the pancreas. The American Cancer Society's estimates predict that about 3900 people in the United States will be diagnosed with pancreatic NET in 2019.

The occurrence of pancreatic NETs seems to be rising over the years. This is thought to be partly because they are more often found incidentally, when imaging tests such as CT or MRI scans are done for other reasons. Also, the ability to distinguish these tumors from other types of cancers in the lab has improved.

Most people with pancreatic NETs are older, with the average age of diagnosis being 60. They are slightly more common in men than women.

People with pancreatic NETs that are grade 1 or 2 tend to live longer than those with grade 3 pancreatic NETs. For more statistics related to survival, see [Pancreatic Neuroendocrine Tumor Survival Rates by Stage](#).<sup>1</sup>

Visit our [Cancer Statistics Center](#)<sup>2</sup> for more key statistics.

### Hyperlinks

1. [www.cancer.org/cancer/pancreatic-neuroendocrine-tumor/detection-diagnosis-staging/survival-rates.html](http://www.cancer.org/cancer/pancreatic-neuroendocrine-tumor/detection-diagnosis-staging/survival-rates.html)
2. <https://cancerstatisticscenter.cancer.org>

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# What's New in Pancreatic Neuroendocrine Tumor Research?

Research into the causes, diagnosis, and treatment of pancreatic neuroendocrine tumor (NET) is under way in many medical centers throughout the world.

## Genetics and early detection

Researchers are looking for the causes of pancreatic NETs in the hope that this knowledge can be used to help prevent or treat them in the future. A great deal of progress has been made in recent years. For example, scientists have found that

changes in the *MEN1* gene (the gene that causes multiple endocrine neoplasia, type 1) are seen in many people with pancreatic NETs. Other genetic changes that seem to make tumors more aggressive are now being explored as well.

## Treatment

[Surgery](#)<sup>1</sup> is the main treatment option for pancreatic NETs when possible. But better approaches are needed when surgery can't remove all the tumors. New chemotherapy drugs and combinations of drugs are being studied as well as targeted therapy.

### Chemotherapy

Temozolomide is known to be helpful in people with advanced pancreatic NET. New research shows temozolomide works better on tumors that are deficient in a certain DNA-repairing protein called *O*<sup>6</sup>-methylguanine-methyltransferase (*MGMT*). New studies have also shown that using another chemotherapy drug called capecitabine along with temozolomide helped people with advanced pancreatic NET live longer than people treated with temozolomide by itself.

### Targeted therapies

[Targeted drugs](#)<sup>2</sup> work differently from standard chemo drugs in that they attack only specific targets on cancer cells (or nearby cells). Targeted therapies may prove to be useful along with, or instead of, current treatments. They have different side effects than traditional chemo drugs. Looking for new targets to attack is an active area of cancer research.

**Kinase inhibitors:** Sunitinib and everolimus have shown to be helpful in pancreatic NETs. Another kinase inhibitor, cabozantinib, also looks promising and more research is being done. Other kinase inhibitors, such as axitinib, nintedanib, pazopanib, and sulfatinib, are being studied as well.

**Anti-angiogenesis factors:** All cancers depend on new blood vessels to nourish their growth. To block the growth of these vessels and thereby starve the tumor, scientists have developed anti-angiogenesis drugs. These are being studied in [clinical trials](#)<sup>3</sup> for patients with pancreatic NETs.

## Hyperlinks

1. [www.cancer.org/cancer/pancreatic-neuroendocrine-tumor/treating/surgery.html](http://www.cancer.org/cancer/pancreatic-neuroendocrine-tumor/treating/surgery.html)



2. [www.cancer.org/treatment/treatments-and-side-effects/treatment-types/targeted-therapy.html](http://www.cancer.org/treatment/treatments-and-side-effects/treatment-types/targeted-therapy.html)
3. [www.cancer.org/treatment/treatments-and-side-effects/clinical-trials.html](http://www.cancer.org/treatment/treatments-and-side-effects/clinical-trials.html)

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# Pancreatic Neuroendocrine Tumors Causes, Risk Factors, and Prevention

## Risk Factors

A risk factor is anything that affects your chance of getting a disease such as cancer. Learn more about the risk factors for pancreatic neuroendocrine tumors.

- [Pancreatic Neuroendocrine Tumor Risk Factors](#)
- [What Causes Pancreatic Neuroendocrine Tumor?](#)

## Prevention

There is no way to prevent all pancreatic neuroendocrine tumors. But there are things you can do that might lower your risk. Learn more.

- [Can Pancreatic Neuroendocrine Tumor Be Prevented?](#)
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# Pancreatic Neuroendocrine Tumor Risk Factors

A risk factor is anything that increases your chance of getting a disease such as cancer. Different cancers have different risk factors. Some risk factors, like smoking, can be changed. Others, like a person's age or family history, can't be changed.

But having a risk factor, or even many risk factors, does not mean that you will get the disease. And some people who get the disease may have few or no known risk factors.

Several factors can affect a person's chance of getting a neuroendocrine tumor (NET) of the pancreas.

## **Risk factors that can be changed**

### **Smoking**

[Smoking](#)<sup>1</sup> is a risk factor for pancreatic NETs. Most research shows that heavy smoking increases risk, but some studies show that any history of smoking could put you at risk.

### **Alcohol**

Some studies have shown a link between [heavy alcohol use](#)<sup>2</sup> and pancreatic NETs. This link appears to be mostly related to functioning pancreatic NETs rather than nonfunctioning pancreatic NETs. Heavy alcohol use can also lead to conditions such as chronic pancreatitis, which may increase pancreatic NET risk.

## **Risk factors that can't be changed**

### **Family history**

Pancreatic NETs seem to run in some families. In some of these families, the high risk is due to an inherited syndrome (explained below). In other families, the gene causing the increased risk is not known. If family history is a risk factor, it usually involves a first degree relative (parent, sibling, child), a family history of pancreatic NET, or a family history of any cancer.

### **Inherited genetic syndromes**

Inherited gene changes (mutations) can be passed from parent to child. Sometimes these changes result in syndromes that include increased risks of other cancers (or other health problems).

Pancreatic neuroendocrine tumors and cancers can also be caused by genetic syndromes, such as:

- **Neurofibromatosis, type 1**, which is caused by mutations in the *NF1* gene. This syndrome leads to an increased risk of many tumors, including somatostatinomas.
- **Multiple endocrine neoplasia, type I (MEN1)**, caused by mutations in the *MEN1* gene. This syndrome leads to an increased risk of tumors of the parathyroid gland, the pituitary gland, and the islet cells of the pancreas.
- **Von Hippel-Lindau (VHL) syndrome**, which is caused by mutations in the *VHL* gene. This syndrome leads to an increased risk of many tumors, including pancreatic NETs.

Changes in the genes that cause some of these syndromes can be found by genetic testing. For more information on genetic testing, see [Can Pancreatic Neuroendocrine Tumor Be Found Early?](#)<sup>3</sup>

## Diabetes

Pancreatic NETs are more common in people with diabetes. The reason for this is not known. Most of the risk is found in people with type 2 diabetes. This type of diabetes most often starts in adulthood and is often related to being overweight or obese. It's not clear if people with type 1 (juvenile) diabetes have a higher risk.

## Chronic pancreatitis

Chronic pancreatitis, a long-term inflammation of the pancreas, is linked with an increased risk of pancreatic NETs. If chronic pancreatitis is because of heavy alcohol use, then stopping alcohol may help decrease the risk of pancreatic NETs.

## Factors with unclear effect on risk

### Being overweight or obese

[Being overweight](#)<sup>4</sup> or obese could be a risk factor for pancreatic NET. Studies so far are inconclusive.

### Coffee

Some older studies have suggested that drinking coffee might increase the risk of pancreatic NET, but more recent studies have not confirmed this.

## Hyperlinks

1. [www.cancer.org/cancer/cancer-causes/tobacco-and-cancer.html](http://www.cancer.org/cancer/cancer-causes/tobacco-and-cancer.html)
2. [www.cancer.org/healthy/eat-healthy-get-active/acs-guidelines-nutrition-physical-activity-cancer-prevention/summary.html](http://www.cancer.org/healthy/eat-healthy-get-active/acs-guidelines-nutrition-physical-activity-cancer-prevention/summary.html)
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4. <https://www.cancer.org/cancer/cancer-causes/diet-physical-activity/body-weight-and-cancer-risk.html>

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## What Causes Pancreatic Neuroendocrine Tumor?

Scientists don't know exactly what causes most pancreatic neuroendocrine tumors (NETs), but they have found several [risk factors](#) that can make a person more likely to get this disease. Some of these risk factors affect the DNA of cells in the neuroendocrine system in the pancreas, which can result in abnormal cell growth and may cause cancers to form.

DNA is the chemical in our cells that carries our genes, which control how our cells function. We look like our parents because they are the source of our DNA. But DNA affects more than just how we look.

Some [genes](#)<sup>1</sup> control when our cells grow, divide into new cells, and die:

- Genes that help cells grow, divide, and stay alive are called *oncogenes*.
- Genes that help keep cell division under control, repair mistakes in DNA, or cause cells to die at the right time are called *tumor suppressor genes*.

Cancers can be caused by DNA changes (mutations) that turn on oncogenes or turn off tumor suppressor genes.

### Inherited gene mutations

Although 90% of PNETs are sporadic (random), some people [inherit gene changes](#)<sup>2</sup> from their parents that raise their risk of pancreatic NET. Sometimes these gene changes are part of syndromes that include increased risks of other health problems as well.

Syndromes related to changes in three tumor suppressor genes are responsible for many inherited cases of pancreatic NETs:

- **Multiple Endocrine Neoplasia Type 1 (MEN1) syndrome:** Most inherited cases

of PNETs are due to changes in the MEN1 gene. This syndrome can cause cancer in the pancreas, parathyroid glands, and [pituitary glands](#)<sup>3</sup>. These tumors usually happen at younger ages and tend to be non-functioning. Screening people with the MEN1 gene or their family members can sometimes help find pancreatic NET before symptoms appear.

- **Von Hippel-Lindau (VHL) syndrome:** Changes in the *VHL* gene cause a small number of pancreatic NETs, usually developing at earlier ages (sometimes as early as the 20s). These tumors tend to be non-functioning and slow growing.
- **Neurofibromatosis type 1 (NF1) syndrome:** A small number of pancreatic NETs (usually somatostatinomas) are caused by changes in the *NF1* gene. Other cancers are also associated with this syndrome, including brain tumors or benign tumors that form in nerves under the skin (neurofibromas),

The treatment for a pancreatic NET that's caused by a genetic syndrome might be slightly different compared to treatment for a pancreatic NET in someone without a gene mutation.

## Acquired gene mutations

Most gene mutations related to neuroendocrine tumors of the pancreas are caused by random changes. These random mutations are called *acquired* if they occur after a person is born, rather than having been inherited. These acquired gene mutations sometimes result from exposure to cancer-causing chemicals (like those found in [tobacco smoke](#)<sup>4</sup>). But often what causes these changes is not known.

## Hyperlinks

1. [www.cancer.org/cancer/cancer-causes/genetics.html](http://www.cancer.org/cancer/cancer-causes/genetics.html)
2. [www.cancer.org/cancer/cancer-causes/genetics.html](http://www.cancer.org/cancer/cancer-causes/genetics.html)
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## Can Pancreatic Neuroendocrine Tumor

# Be Prevented?

There is no sure way to prevent pancreatic neuroendocrine tumors (NETs). Some **risk factors** such as family history can't be controlled. But there are things you can do that might lower your risk.

## Don't smoke

Smoking is an avoidable risk factor for pancreatic NET. [Quitting smoking](#)<sup>1</sup> may help lower risk. If you smoke and want help quitting, please talk to your health care provider or call us at 1-800-227-2345.

## Limit alcohol use

Heavy alcohol use has been tied to pancreatic NETs in some studies but not in others. This link is still not certain, but heavy alcohol use can lead to conditions such as chronic pancreatitis, which has been associated with an increased risk of pancreatic NET.

## Hyperlinks

1. [www.cancer.org/healthy/stay-away-from-tobacco.html](http://www.cancer.org/healthy/stay-away-from-tobacco.html)

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# Pancreatic Neuroendocrine Tumors

## Early Detection, Diagnosis, and Staging

### Detection and Diagnosis

Catching cancer early often allows for more treatment options. Some early cancers may have signs and symptoms that can be noticed, but that is not always the case.

- [Can Pancreatic Neuroendocrine Tumor Be Found Early?](#)
- [Signs and Symptoms of Pancreatic Neuroendocrine Tumor](#)
- [Tests for Pancreatic Neuroendocrine Tumor](#)

### Stages and Outlook (Prognosis)

After a cancer diagnosis, staging provides important information about the extent of cancer in the body and anticipated response to treatment.

- [Pancreatic Neuroendocrine Tumor Stages](#)
- [Survival Rates for Pancreatic Neuroendocrine Tumor](#)

### Questions to Ask About Pancreatic Cancer

Here are some questions you can ask your cancer care team to help you better understand your cancer diagnosis and treatment options.

- [Questions to Ask About Pancreatic Neuroendocrine Tumor](#)

# Can Pancreatic Neuroendocrine Tumor Be Found Early?

Pancreatic neuroendocrine tumors (NETs) are hard to find early. The pancreas is deep inside the body, so small tumors can't be seen or felt by health care providers during routine physical exams. People usually have no [symptoms](#) until the cancer has already spread to other organs.

At this time, no major professional groups recommend routine screening for pancreatic NET in people who are at average risk. This is because no screening test has been shown to lower the risk of dying from this cancer. (*Screening* means testing for a disease in people who have no symptoms or history of the disease.)

## Genetic testing for people who might be at increased risk

Some people might be at increased risk of pancreatic NET because of a family history of the disease (or a family history of certain other cancers). Sometimes this increased risk is due to a specific [genetic syndrome](#)<sup>1</sup>.

Some of the [gene changes](#)<sup>2</sup> that increase pancreatic NET risk can be tested for. Knowing if you are at increased risk can help you and your doctor decide if you should have tests to look for pancreatic NET early, when it might be easier to treat. But determining whether you might be at increased risk is not simple. Talking to someone with experience in hereditary cancer syndromes such as a genetic counselor, geneticist, or an oncologist (doctor who treats people with cancer) is often helpful.

The American Cancer Society strongly recommends that anyone thinking about [genetic testing](#)<sup>3</sup> talk with a genetic counselor, nurse, or doctor qualified to interpret and explain the test results before they proceed with testing. Before deciding to be tested, it's important to understand what the tests can and can't tell you, and what any results might mean.

Genetic tests look for mutations in your genes that cause inherited conditions. The tests are used to look for these inherited conditions, not the cancer itself. Your risk may be increased if you have one of these conditions, but it doesn't mean that you have or definitely will get pancreatic NET.

## Testing for pancreatic neuroendocrine tumor in people at high risk

For people in families at high risk of pancreatic NET, such as MEN1 syndrome, tests for detecting cancer early may help. Although definitive screening guidelines for people with the MEN1 gene or their family members are not available, doctors have been able to find early, treatable pancreatic NETs in some members of high-risk families with these tests. Some [tests](#) that might be considered include:

- An endoscopic ultrasound of the pancreas every few years.
- A MRI of the pancreas every few years.
- Checking blood levels of certain hormones such as insulin, prolactin, gastrin, and calcium every few years. (Sometimes, changes in hormones can occur 10 years before the tumor is found by clinical symptoms.)
- An Octreoscan on a regular basis.

## Hyperlinks

1. [www.cancer.org/cancer/cancer-causes/genetics/family-cancer-syndromes.html](http://www.cancer.org/cancer/cancer-causes/genetics/family-cancer-syndromes.html)
2. [www.cancer.org/cancer/cancer-causes/genetics/genes-and-cancer.html](http://www.cancer.org/cancer/cancer-causes/genetics/genes-and-cancer.html)
3. [www.cancer.org/cancer/cancer-causes/genetics.html](http://www.cancer.org/cancer/cancer-causes/genetics.html)

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# Signs and Symptoms of Pancreatic Neuroendocrine Tumor

Having one or more of the symptoms below does not mean you have a pancreatic neuroendocrine tumor (NET). In fact, many of these symptoms are more likely to be caused by other conditions. Still, if you have any of these symptoms, it's important to have them checked by a doctor so that the cause can be found and treated, if needed.

Pancreatic NETs often release excess hormones into the bloodstream. Different types of tumors make different hormones, which can lead to different symptoms.

## Gastrinomas

These tumors make gastrin, a hormone that tells the stomach to make more acid. Too much gastrin causes a condition known as *Zollinger-Ellison syndrome*, in which the stomach makes too much acid. This leads to stomach ulcers, which can cause **pain, nausea, and loss of appetite**. Severe ulcers can bleed. Even if the bleeding is mild, it can lead to anemia (too few red blood cells), which can cause symptoms like **feeling tired** and **being short of breath**. If the bleeding is more severe, it can make **stool black and tarry**. Severe bleeding can itself be life-threatening.

If the stomach acid reaches the small intestine, it can damage the cells of the intestinal lining and break down digestive enzymes before they have a chance to digest food. This can cause **diarrhea** and **weight loss**.

## Glucagonomas

These tumors make glucagon, a hormone that increases glucose (sugar) levels in the blood. Most of the symptoms that can be caused by a glucagonoma are mild and are more often caused by something else.

Excess glucagon can raise blood sugar, sometimes leading to diabetes. This can cause symptoms such as **feeling thirsty and hungry**, and **having to urinate often**.

People with these tumors can also have problems with **diarrhea, weight loss, and malnutrition**. The nutrition problems can lead to symptoms like **irritation of the tongue and the corners of the mouth**.

The symptom that brings most people with glucagonomas to their doctor is a rash called

*necrolytic migratory erythema*. This is a **red rash with swelling and blisters that often travels from place to place on the skin.**

## Insulinomas

These tumors make insulin, which lowers blood glucose levels. Too much insulin leads to low blood sugar, which can cause symptoms like **weakness, confusion, sweating, and rapid heartbeat**. When blood sugar gets very low, it can lead to a person **passing out or even going into a coma and having seizures**.

## Somatostatinomas

These tumors make somatostatin, which helps regulate other hormones. Symptoms of this type of tumor can include **belly pain, nausea, poor appetite, weight loss, diarrhea, symptoms of diabetes (feeling thirsty and hungry, and having to urinate often), and jaundice (yellowing of the skin and eyes)**.

The early symptoms of a somatostatinoma tend to be mild and are more often caused by other things, so these tumors tend to be diagnosed at an advanced stage. Often, they are not found until they spread to the liver, when they cause problems like jaundice and pain.

## VIPomas

These tumors make a substance called *vasoactive intestinal peptide* (VIP). Too much VIP can lead to problems with **diarrhea**. This may be mild at first, but gets worse over time. By the time they are diagnosed, most people have severe, watery diarrhea.

Other symptoms can include **nausea, vomiting, muscle cramps, feeling weak or tired, and flushing (redness and warmth in the face or neck)**.

People with these tumors also tend to have low levels of acid in their stomachs, which can lead to **problems digesting food**.

## Carcinoid tumors

These tumors often make serotonin or its precursor, 5-HTP. Carcinoid tumors often don't cause symptoms until they spread outside the pancreas. When these tumors do spread, it is most often to the liver. There, the cancer cells can release hormones



directly into the blood. This can cause the carcinoid syndrome, with symptoms including **flushing (redness and warmth in the face or neck), diarrhea, wheezing, and a rapid heart rate.** These symptoms often occur in episodes, between which the person may feel fine.

Over a long time, the hormone-like substances released by these tumors can damage heart valves, causing **shortness of breath, weakness, and a heart murmur (an abnormal heart sound).**

## Non-functioning neuroendocrine tumors

These tumors don't make excess hormones, so they don't cause symptoms in early stages and often grow quite large before they are found. Most of these start to cause problems as they get larger or spread outside the pancreas. Symptoms can be like those from **exocrine pancreas cancers, including jaundice (yellowing of the eyes and skin), belly pain, and weight loss.** Sometimes, as a pancreatic NET grows, it can go from making too little of a hormone (nonfunctioning) to making too much of a specific hormone (functioning) which causes symptoms.

## Symptoms caused by the cancer spreading

When pancreatic NETs spread, most often they go to the liver. This can enlarge the liver, which can cause **pain and loss of appetite.** It can also affect liver function, sometimes leading to **jaundice (yellowing of the skin and eyes) and abnormal blood tests.**

These cancers can also spread to other organs and tissues. The symptoms depend on where the cancer is growing. For example, cancer spread to the lungs can cause **shortness of breath or a cough.** Spread to bones can cause **pain** in those areas.

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## Tests for Pancreatic Neuroendocrine Tumor

Certain [signs and symptoms](#) might suggest that a person could have a pancreatic neuroendocrine tumor (NET), but tests are needed to confirm the diagnosis.

### Medical history and physical exam

In taking your medical history, the doctor will ask you questions about your general health, lifestyle habits, symptoms, and risk factors. The doctor will also probably ask about symptoms related to excess hormone production such as diarrhea, abdominal (belly) pain, or rash.

Your doctor will also examine you to look for signs of pancreatic NET or other health problems. The exam will probably focus mostly on your belly. Pancreatic NETs can sometimes cause the liver or gallbladder to swell, which the doctor might be able to feel during the exam.

If the results of the exam are abnormal, your doctor will probably order tests, such as imaging, labs, or other procedures, to help find the problem. You might also be referred

to a gastroenterologist (a doctor who treats digestive system diseases) for further tests and treatment.

## Imaging tests

### Computed tomography (CT) scan

A [CT scan](#)<sup>1</sup> uses x-rays taken from different angles, which are combined by a computer to make detailed pictures of the organs. This test is most often used to look at the chest and/or belly (abdomen) to see the pancreas clearly and if the pancreatic NET has spread to nearby lymph nodes or other organs such as the liver. It can also be used to guide a biopsy needle into an area of concern.

### Magnetic resonance imaging (MRI)

Like CT scans, [MRI scans](#)<sup>2</sup> show detailed images of soft tissues in the body. But MRI scans use radio waves and strong magnets instead of x-rays. A dye called gadolinium may be injected into a vein before the scan to see details better. A MRI scan sometimes can see cancer spread to the liver better than a CT scan.

**MR cholangiopancreatography (MRCP)**, is a special type of MRI scan, which can be used to look at the pancreatic and bile ducts, and is described below in the section on cholangiopancreatography.

### Ultrasound

[Ultrasound](#)<sup>3</sup> tests use sound waves to create images of organs such as the pancreas.

**Abdominal ultrasound:** For this test, a wand-shaped probe is moved over the skin of the abdomen. It gives off sound waves and detects the echoes as they bounce off organs. If it's not clear what might be causing a person's abdominal symptoms, this might be the first test done because it is easy to do and it doesn't expose a person to radiation.

**Endoscopic ultrasound (EUS):** This test uses an endoscope with a small ultrasound probe on the end. The scope is then passed through your mouth or nose, down through the stomach, and into the first part of the small intestine. It is then pointed toward the pancreas, which is next to the small intestine. The probe on the tip of the endoscope can get very close to the pancreas, so this is a very good way to look at it. If a tumor is seen, a small, hollow needle can be passed down the endoscope to get biopsy samples

of it.

## **Cholangiopancreatography**

This is an imaging test that looks at the pancreatic ducts and bile ducts to see if they are blocked, narrowed, or dilated. These tests can help show if someone might have a pancreatic neuroendocrine tumor that is blocking a duct. They can also be used to help plan surgery. The test can be done in different ways, each of which has pros and cons.

**Endoscopic retrograde cholangiopancreatography (ERCP):** For this test, an endoscope (a thin, flexible tube with a tiny video camera on the end) is passed down the throat, through the esophagus and stomach, and into the first part of the small intestine. This is usually done while you are sedated (given medicine to make you sleepy).

The doctor can see through the endoscope to find the ampulla of Vater (where the common bile duct empties into the small intestine). The doctor guides a catheter (a very small tube) through the tip of the endoscope and into the common bile duct. A small amount of dye is then injected into the common bile duct, and x-rays are taken. This dye outlines the bile and pancreatic ducts. The x-rays can show narrowing or blockage in these ducts that might be due to pancreatic neuroendocrine tumor. The doctor doing this test can also put a small brush through the tube to remove cells for a biopsy (see below).

ERCP can also be used to place a stent (small tube) into a bile or pancreatic duct to keep it open if a nearby tumor is pressing on it.

**Magnetic resonance cholangiopancreatography (MRCP):** This is a non-invasive way to look at the pancreatic and bile ducts using the same type of machine used for standard MRI scans. Unlike ERCP, it does not require an injection of a dye. Because this test is non-invasive, doctors often use MRCP if the purpose is just to look at the pancreatic and bile ducts. But this test can't be used to get biopsy samples of tumors or to place stents in ducts.

**Percutaneous transhepatic cholangiography (PTC):** In this procedure, the doctor puts a thin, hollow needle through the skin of the belly and into a bile duct within the liver. A dye is then injected through the needle, and x-rays are taken as it passes through the bile and pancreatic ducts. As with ERCP, this approach can also be used to take fluid or tissue samples or to place a stent into a duct to help keep it open. Because it is more invasive (and might cause more pain), PTC is not usually used unless ERCP has already been tried or can't be done for some reason.

## Radionuclide scans

Scans using small amounts of radioactivity and special cameras can be helpful in looking for pancreatic NETs. They can help find tumors or look for areas of cancer spread if doctors aren't sure where it is in the body.

**Positron emission tomography (PET) scan:** For most types of cancer, [PET scans](#)<sup>4</sup> use a form of radioactive glucose (sugar) to find tumors. This type of PET scan is useful in finding poorly differentiated pancreatic neuroendocrine carcinomas (NECs), but a newer type of PET scan, called a Gallium-68 PET/CT Dotatate scan is being used for pancreatic NETs. It uses the radioactive agent gallium-68 dotatate which attaches to the somatostatin protein on neuroendocrine tumor cells. A special camera can detect the radioactivity. This Gallium-68 PET/CT scan is slowly becoming more widely available since it was approved by the FDA in 2016 and is able to find neuroendocrine tumors better than an OctreoScan (described below).

**Somatostatin receptor scintigraphy (SRS or OctreoScan):** This test can be very helpful in finding pancreatic NETs. It uses a drug called octreotide joined to radioactive *indium-111*. *Octreotide* is a hormone-like substance that attaches to pancreatic NET cells. A small amount of the octreotide-radioactive substance is injected into a vein and travels through the blood where it attaches to the tumor types of many types of pancreatic NET cells (although it is less helpful for insulinomas). A few hours after the injection, a special camera can be used to show where the radioactivity has collected in the body. More scans may be done over the next few days as well. Along with showing where tumors are, this test can also tell whether treatment with certain drugs such as octreotide and lanreotide is likely to be helpful.

## Blood and urine tests

Several types of blood and urine tests can be used to help diagnose pancreatic NET or to help determine treatment options if it is found.

Blood tests looking at the levels of certain pancreatic hormones can often help diagnose pancreatic NETs. Tests might be done to check blood levels of:

- Hormones made by different types of pancreatic NET cells, such as insulin, gastrin, glucagon, somatostatin, pancreatic polypeptide, and VIP (vasoactive intestinal peptide)
- Chromogranin A (CgA)
- Glucose and C-peptide (for insulinomas)

**Carcinoid tumors:** For carcinoids, a blood test may be done to look for serotonin, which is made by many of these tumors. The urine might also be tested for serotonin and for related chemicals such as 5-HIAA and 5-HTP.

Other common tests to look for carcinoids include blood tests for chromogranin A (CgA), neuron-specific enolase (NSE), substance P, and gastrin.

Depending on where the tumor might be located and the patient's symptoms, doctors might do other blood tests as well.

Some of these tests can also be used to show how well [treatment](#)<sup>5</sup> is working, since the levels of these substances tend to go down as tumors shrink.

## Biopsy

In many cases, the only way to know for sure if a person has some type of pancreatic NET is to remove cells from the tumor and look at them in the lab. This procedure is called a *biopsy*. Biopsies can be done in different ways.

**Percutaneous (through the skin) biopsy:** For this test, a doctor inserts a thin, hollow needle through the skin over the abdomen and into the pancreas to remove a small piece of a tumor. This is known as a *fine needle aspiration* (FNA). The doctor guides the needle into place using images from ultrasound or CT scans.

**Endoscopic biopsy:** Doctors can also biopsy a tumor during an endoscopy. The doctor passes an endoscope (a thin, flexible, tube with a small video camera on the end) into the throat, down the esophagus, and into the small intestine near the pancreas. At this point, the doctor can either use endoscopic ultrasound (EUS) to pass a needle into the tumor or endoscopic retrograde cholangiopancreatography (ERCP) to remove cells from the bile or pancreatic ducts. These tests are described in more detail above.

**Surgical biopsy:** In rare cases, an endoscopic biopsy or a CT-guided needle biopsy will not be able to get enough tissue to identify the type of tumor. In such cases, surgery may be needed to remove a tissue sample. Surgical biopsies are done much less often now than in the past since PNETs are mostly diagnosed using imaging (CT or MRI scans), somatostatin receptor-based imaging, EUS biopsy, and checking for excessive levels of hormones.

### Some people might not need a biopsy

Rarely, the doctor might not do a biopsy on someone who has a neuroendocrine tumor in the pancreas if imaging tests, blood tests, and somatostatin receptor-based imaging show the tumor is very likely to be cancer and if it looks like surgery can remove all of it. Instead, the doctor will proceed with surgery, at which time the tumor cells can be looked at in the lab to confirm the diagnosis. During surgery, if the doctor finds that the cancer has spread too far to be removed completely, only a sample of the cancer may be removed to confirm the diagnosis, and the rest of the planned operation may be stopped.

See [Testing Biopsy and Cytology Specimens for Cancer](#)<sup>6</sup> to learn more about different types of biopsies, how the biopsy samples are tested in the lab, and what the results will tell you.

## Hyperlinks

1. [www.cancer.org/treatment/understanding-your-diagnosis/tests/ct-scan-for-cancer.html](http://www.cancer.org/treatment/understanding-your-diagnosis/tests/ct-scan-for-cancer.html)
2. [www.cancer.org/treatment/understanding-your-diagnosis/tests/mri-for-cancer.html](http://www.cancer.org/treatment/understanding-your-diagnosis/tests/mri-for-cancer.html)
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## Pancreatic Neuroendocrine Tumor Stages

After someone is diagnosed with a pancreatic neuroendocrine tumor (NET), doctors will try to figure out if it has spread, and if so, how far. This process is called *staging*. The stage of a cancer describes how much cancer is in the body. It helps determine how serious the cancer is and how best to [treat](#)<sup>1</sup> it. Doctors also use a cancer's stage when talking about survival statistics.

The stages of pancreatic NET range from I (1) through IV (4). As a rule, the lower the stage, the less the cancer has spread. A higher number, such as stage IV, means cancer has spread more. Although each person's cancer experience is unique, cancers with similar stages tend to have a similar outlook and are often treated in much the same way.

### How is the stage determined?

The staging system most often used for pancreatic NETs is the American Joint Committee on Cancer (AJCC) **TNM** system, which is based on 3 key pieces of information:

- The size and extent of the main **tumor (T)**: How large is the tumor? Has it grown



into nearby structures or organs?

- The spread to nearby lymph **nodes (N)**: Has the cancer spread to nearby lymph nodes?
- The spread (**metastasis**) to distant sites (**M**): Has the cancer spread to distant organs? (The most common site of spread is the liver.)

Numbers or letters after T, N, and M provide more details about each of these factors. Higher numbers mean the cancer is more advanced. Once a person's T, N, and M categories have been determined, this information is combined in a process called *stage grouping* to assign an overall stage. For more information, see [Cancer Staging](#)<sup>2</sup>.

The system described below is the most recent version of the AJCC system, effective as of January 2018. It is used to stage well-differentiated pancreatic NETs, but not high-grade pancreatic NETs (known as *neuroendocrine carcinomas*) or other types of pancreatic cancer, which have [their own staging system](#)<sup>3</sup>.

Pancreatic NETs are typically given a **clinical stage** based on the results of a physical exam, biopsy, and imaging tests (as described in [Tests for Pancreatic Neuroendocrine Tumor](#)). If surgery is done, the **pathologic stage** (also called the *surgical stage*) is determined by examining tissue removed during the operation.

Staging for pancreatic NETs can be complex. If you have any questions about the stage of your cancer or what it means, please ask your doctor to explain it to you in a way you understand.

## Stages of pancreatic neuroendocrine tumors

AJCC Stage	Stage grouping	Stage description*
I	<b>T1 N0 M0</b>	The tumor is less than 2 centimeters (cm) across and is still just in the pancreas (T1). The cancer has not spread to nearby lymph nodes (N0) or to distant parts of the body (M0).
II	<b>T2 N0 M0</b>	The tumor is at least 2 cm across but no more than 4 cm across, and it is still just in the pancreas (T2). The cancer has not spread to nearby lymph nodes (N0) or to distant parts of the body (M0).
	OR	
	<b>T3 N0 M0</b>	The tumor is more than 4 cm across and is still just in the

		pancreas, OR the tumor has grown into the duodenum (the first part of the small intestine) or the common bile duct (T3). The cancer has not spread to nearby lymph nodes (N0) or to distant parts of the body (M0).
III	<b>T4 N0 M0</b>	The tumor has grown into nearby organs (such as the stomach, spleen, colon, or adrenal gland) or it has grown into nearby large blood vessels (T4). The cancer has not spread to nearby lymph nodes (N0) or to distant parts of the body (M0).
	OR	
	<b>Any T N1 M0</b>	The tumor can be any size and might or might not have grown outside of the pancreas (any T). It has spread to nearby lymph nodes (N1), but not to distant parts of the body (M0).
IV	<b>Any T Any N M1</b>	The tumor can be any size and might or might not have grown outside of the pancreas (any T). It might or might not have spread to nearby lymph nodes (any N). The cancer has spread to distant parts of the body (M1).

\* The following additional categories are not listed in the table above:

- **TX:** The main tumor cannot be assessed due to lack of information.
- **T0:** There is no evidence of a main tumor.
- **NX:** Nearby lymph nodes cannot be assessed due to lack of information.

## Other prognostic factors

Although not formally part of the TNM system, other factors can also be important in determining a person's prognosis (outlook).

### Tumor grade

The grade describes how quickly the cancer is likely to grow and spread. For pancreatic NETs, an important part of grading is measuring how many of the cells are in the process of dividing into new cells. This is determined by:

- The **mitotic count**, which is the number of cells seen under a microscope that are in the process of splitting into two new cells (mitosis).
- The **Ki-67 index**, which is a measure of the portion of cells that are almost ready to

start splitting.

Based on these tests, NETs are divided into 2 main groups:

- **Well-differentiated tumors** (which include low-grade [G1] and intermediate-grade [G2] tumors) have 20 or fewer mitoses and a Ki-67 index of 20% or lower.
- **Poorly differentiated tumors** (high-grade [G3] tumors) have more than 20 mitoses or a Ki-67 index of more than 20%. These are also called **neuroendocrine carcinomas (NECs)**, and they often grow and spread quickly.

### Tumor functionality

The outlook for pancreatic NETs can be affected by whether the tumor is [functioning \(making hormones\) or non-functioning](#)<sup>4</sup>. For functioning tumors, the type of hormone can also be important. For example, insulinomas (NETs that make insulin) tend to have a lower risk of spreading than other types of NETs.

### Hyperlinks

1. [www.cancer.org/cancer/pancreatic-neuroendocrine-tumor/treating/pnets-by-extent.html](http://www.cancer.org/cancer/pancreatic-neuroendocrine-tumor/treating/pnets-by-extent.html)
2. [www.cancer.org/treatment/understanding-your-diagnosis/staging.html](http://www.cancer.org/treatment/understanding-your-diagnosis/staging.html)
3. [www.cancer.org/cancer/pancreatic-cancer/detection-diagnosis-staging/staging.html](http://www.cancer.org/cancer/pancreatic-cancer/detection-diagnosis-staging/staging.html)
4. [www.cancer.org/cancer/pancreatic-neuroendocrine-tumor/about/what-is-pnet.html](http://www.cancer.org/cancer/pancreatic-neuroendocrine-tumor/about/what-is-pnet.html)

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# Survival Rates for Pancreatic Neuroendocrine Tumor

Survival rates can give you an idea of what percentage of people with the same type and stage of cancer are still alive a certain amount of time (usually 5 years) after they were diagnosed. They can't tell you how long you will live, but they may help give you a better understanding of how likely it is that your treatment will be successful.

**Keep in mind that survival rates are estimates and are often based on previous outcomes of large numbers of people who had a specific cancer, but they can't predict what will happen in any particular person's case. These statistics can be confusing and may lead you to have more questions. Talk with your doctor about how these numbers may apply to you, as he or she is familiar with your situation.**

## What is a 5-year relative survival rate?

A **relative survival rate** compares people with the same type and stage of pancreatic neuroendocrine tumor (NET) to people in the overall population. For example, if the **5-year relative survival rate** for a specific type and stage of pancreatic NET is 90%, it means that people who have that cancer are, on average, about 90% as likely as people who don't have that cancer to live for at least 5 years after being diagnosed.

## Where do these numbers come from?

The American Cancer Society relies on information from the SEER\* database, maintained by the National Cancer Institute (NCI), to provide survival statistics for different types of cancer.

The SEER database tracks 5-year relative survival rates for pancreatic NET in the United States, based on how far the cancer has spread. The SEER database, however, does not group cancers by [AJCC TNM stages](#) (stage 1, stage 2, stage 3, etc.). Instead, it groups cancers into localized, regional, and distant stages:

- **Localized:** There is no sign the cancer has grown outside of the pancreas. This includes stage I and some stage II cancers.
- **Regional:** The cancer has grown outside the pancreas into nearby tissues or has spread to nearby lymph nodes. This includes mainly stage III cancers and some stage II.
- **Distant:** The cancer has spread to distant parts of the body such as the lungs, liver

or bones. For pancreatic NET, this includes stage IV cancers.

### 5-year relative survival rates for pancreatic NET

(Based on people diagnosed with pancreatic NET between 2008 and 2014.)

SEER Stage	5-year Relative Survival Rate
Localized	94%
Regional	76%
Distant	27%
All SEER stages combined	54%

### Understanding the numbers

- **These numbers apply only to the stage of the cancer when it is first diagnosed.** They do not apply later on if the cancer grows, spreads, or comes back after treatment.
- **These numbers don't take everything into account.** Survival rates are grouped based on how far the cancer has spread, but your age, overall health, how well the cancer responds to treatment, [tumor grade, tumor function](#)<sup>1</sup>, and other factors can also affect your outlook.
- **People now being diagnosed with pancreatic NET may have a better outlook than these numbers show.** Treatments improve over time, and these numbers are based on people who were diagnosed and treated at least five years earlier.

\*SEER= Surveillance, Epidemiology, and End Results

### Hyperlinks

1. [www.cancer.org/cancer/pancreatic-neuroendocrine-tumor/about/what-is-pnet.html](http://www.cancer.org/cancer/pancreatic-neuroendocrine-tumor/about/what-is-pnet.html)

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submission, posted to the SEER web site, April 2018.

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## Questions to Ask About Pancreatic Neuroendocrine Tumor

It's important to have honest, open discussions with your cancer care team. They want to answer all your questions, so that you can make informed treatment and life decisions. For instance, consider these questions:

### When you're told you have a pancreatic neuroendocrine tumor

- What [kind of pancreatic neuroendocrine tumor](#)<sup>1</sup> do I have?
- Has my cancer spread beyond where it started?
- What is the [stage](#)<sup>2</sup> of my cancer and what does that mean?
- Is my cancer resectable (removable by surgery)?
- Are my symptoms because the cancer is making too many hormones?
- Will I need any other tests before we can decide on treatment?
- Will I need to see other doctors or health care professionals?
- If I'm concerned about the costs and insurance coverage for my diagnosis and treatment, who can help me?

### When deciding on a treatment plan

- How much experience do you have treating this type of cancer?
- What are my [treatment options](#)<sup>3</sup>?
- What do you recommend and why?
- What is the goal of the treatment?
- Should I get a [second opinion](#)<sup>4</sup>? How do I do that? Can you recommend a doctor or cancer center?
- How is treatment likely to help me?

- What risks or side effects might I expect? Are there things I can do to reduce these side effects?
- Should I think about taking part in a [clinical trial](#)<sup>5</sup>?
- How quickly do I need to decide on treatment?
- What should I do to be ready for treatment?
- How long will treatment last? What will it be like? Where will it be done?
- What are the chances the cancer will [recur](#)<sup>6</sup> (come back) with these treatment plans?
- Will treatment affect my daily activities? Can I still work full time?
- What would my options be if the treatment doesn't work or if the cancer comes back?
- What if I have [transportation problems](#)<sup>7</sup> getting to and from treatment?

## During treatment

Once treatment begins, you'll need to know what to expect and what to look for. Not all of these questions may apply to you, but getting answers to the ones that do may be helpful.

- How will we know if the treatment is working?
- Is there anything I can do to help manage [side effects](#)<sup>8</sup>?
- What symptoms or side effects should I tell you about right away?
- How can I reach you on nights, holidays, or weekends?
- Do I need to change what I eat during treatment?
- Are there any limits on what I can do?
- Can I exercise during treatment? If so, what kind should I do, and how often?
- Can you suggest a mental health professional I can see if I start to feel [overwhelmed, depressed, or distressed](#)<sup>9</sup>?

## After treatment

- Are there any limits on what I can do?
- Do I need a special diet after treatment?
- What symptoms should I watch for?
- What kind of exercise should I do now?
- What type of [follow-up](#)<sup>10</sup> will I need after treatment?

- How often will I need to have follow-up exams and tests? Will I need any blood tests?
- How will we know if the cancer has come back? What should I watch for?
- What will my options be if the cancer comes back?

Along with these sample questions, be sure to write down some of your own. Keep in mind that doctors aren't the only ones who can give you information. Other health care professionals, such as nurses and social workers, can answer some of your questions. To find out more about speaking with your health care team, see [The Doctor-Patient Relationship](#)<sup>11</sup>.

## Hyperlinks

1. [www.cancer.org/cancer/pancreatic-neuroendocrine-tumor/about/what-is-pnet.html](http://www.cancer.org/cancer/pancreatic-neuroendocrine-tumor/about/what-is-pnet.html)
2. [/content/cancer/en/cancer/pancreatic-neuroendocrine-tumor/detection-diagnosis-staging/staging.html](http://content.cancer/en/cancer/pancreatic-neuroendocrine-tumor/detection-diagnosis-staging/staging.html)
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10. [www.cancer.org/cancer/pancreatic-neuroendocrine-tumor/after-treatment.html](http://www.cancer.org/cancer/pancreatic-neuroendocrine-tumor/after-treatment.html)
11. [www.cancer.org/treatment/understanding-your-diagnosis/talking-about-cancer/the-doctor-patient-relationship.html](http://www.cancer.org/treatment/understanding-your-diagnosis/talking-about-cancer/the-doctor-patient-relationship.html)

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# Treating Pancreatic Neuroendocrine Tumors

If you've been diagnosed with a pancreatic neuroendocrine tumor (NET), your cancer care team will discuss your treatment options with you. It's important to weigh the benefits of each treatment option against the possible risks and side effects.

## How are pancreatic neuroendocrine tumors treated?

Depending on the type and stage of the cancer and other factors, treatment options for people with pancreatic neuroendocrine tumor (NET) can include:

- [Surgery for Pancreatic Neuroendocrine Tumor](#)
- [Ablation or Embolization Treatments for Pancreatic Neuroendocrine Tumor](#)
- [Radiation Therapy for Pancreatic Neuroendocrine Tumor](#)
- [Chemotherapy for Pancreatic Neuroendocrine Tumor](#)
- [Targeted Therapy for Pancreatic Neuroendocrine Tumor](#)
- [Other Drugs for Pancreatic Neuroendocrine Tumors](#)

## Common treatment approaches

For pancreatic neuroendocrine tumors (NETs), treatment options might include surgery, ablation or embolization treatments, radiation therapy, or different types of medicines.

- [Treating Pancreatic Neuroendocrine Tumor, Based on Extent of the Tumor](#)

## Who treats pancreatic neuroendocrine tumors?

Depending on the resources available in your community, you can have different types

of doctors on your treatment team. The doctors on your cancer treatment team might include:

- A **surgeon**: a doctor who uses surgery to treat cancers or other problems
- An **endocrinologist**: a doctor who specializes in the diagnosis and treatment of diseases involving hormones
- A **radiation oncologist**: a doctor who specializes in treating cancer with radiation
- A **medical oncologist**: a doctor who specializes in treating cancer with chemotherapy, immunotherapy, targeted therapy and other medicines
- A **gastroenterologist**: a doctor who specializes in diagnosing and treating diseases of the digestive system.

Many other specialists may be involved in your care as well, including nurse practitioners, nurses, psychologists, social workers, rehabilitation specialists, and other health professionals.

- [Health Professionals Associated With Cancer Care<sup>1</sup>](#)

## **Making treatment decisions**

It's important to discuss all of your treatment options, including their goals and possible side effects, with your doctors to help make the decision that best fits your needs. Some important things to consider include:

- Your age and expected life span
- Any other serious health conditions you have
- The stage (extent) of your cancer
- Whether or not surgery can remove (resect) the cancer
- The likelihood that treatment will cure the cancer (or help in some other way)
- Your feelings about the possible side effects from treatment

You may feel that you must make a decision quickly, but it's important to give yourself time to absorb the information you have just learned. Ask questions if there is anything you're not sure about.

If time permits, it is often a good idea to seek a second opinion. A second opinion can give you more information and help you feel more confident about the treatment plan you choose.

- [Questions to Ask About Pancreatic Neuroendocrine Tumor<sup>2</sup>](#)
- [Seeking a Second Opinion<sup>3</sup>](#)

### **Thinking about taking part in a clinical trial**

Clinical trials are carefully controlled research studies that are done to get a closer look at promising new treatments or procedures. Clinical trials are one way to get state-of-the-art cancer treatment. In some cases they may be the only way to get access to newer treatments. They are also the best way for doctors to learn better methods to treat cancer. Still, they're not right for everyone.

If you would like to learn more about clinical trials that might be right for you, start by asking your doctor if your clinic or hospital conducts clinical trials.

- [Clinical Trials<sup>4</sup>](#)

### **Considering complementary and alternative methods**

You may hear about alternative or complementary methods that your doctor hasn't mentioned to treat your cancer or relieve symptoms. These methods can include vitamins, herbs, and special diets, or other methods such as acupuncture or massage, to name a few.

Complementary methods refer to treatments that are used along with your regular medical care. Alternative treatments are used instead of a doctor's medical treatment. Although some of these methods might be helpful in relieving symptoms or helping you feel better, many have not been proven to work. Some might even be harmful.

Be sure to talk to your cancer care team about any method you are thinking about using. They can help you learn what is known (or not known) about the method, which can help you make an informed decision.

- [Complementary and Alternative Medicine<sup>5</sup>](#)

### **Help getting through cancer treatment**

Your cancer care team will be your first source of information and support, but there are other resources for help when you need it. Hospital- or clinic-based support services are an important part of your care. These might include nursing or social work services, financial aid, nutritional advice, rehab, or spiritual help.

The American Cancer Society also has programs and services – including rides to treatment, lodging, and more – to help you get through treatment. Call our National Cancer Information Center at 1-800-227-2345 and speak with one of our trained specialists.

- [Find Support Programs and Services in Your Area](#)<sup>6</sup>

### **Choosing to stop treatment or choosing no treatment at all**

For some people, when treatments have been tried and are no longer controlling the cancer, it could be time to weigh the benefits and risks of continuing to try new treatments. Whether or not you continue treatment, there are still things you can do to help maintain or improve your quality of life.

Some people, especially if the cancer is advanced, might not want to be treated at all. There are many reasons you might decide not to get cancer treatment, but it's important to talk to your doctors and you make that decision. Remember that even if you choose not to treat the cancer, you can still get supportive care to help with pain or other symptoms.

- [If Cancer Treatments Stop Working](#)<sup>7</sup>
- [Palliative or Supportive Care](#)<sup>8</sup>

*The treatment information given here is not official policy of the American Cancer Society and is not intended as medical advice to replace the expertise and judgment of your cancer care team. It is intended to help you and your family make informed decisions, together with your doctor. Your doctor may have reasons for suggesting a treatment plan different from these general treatment options. Don't hesitate to ask him or her questions about your treatment options.*

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## **Surgery for Pancreatic Neuroendocrine Tumor**

Two general types of surgery can be used for pancreatic neuroendocrine tumors (NETs):

- **Potentially curative surgery** is used when the results of exams and tests suggest that it's possible to remove (resect) all the cancer.
- **Palliative surgery** may be done if imaging tests show that the cancer is too widespread to be removed completely. This surgery is done to reduce tumor size to relieve symptoms from excess hormone production or to prevent certain complications like a blocked bile duct or intestine. The goal is not to try to cure the cancer.

Before any surgery is done, it is important to treat and control any symptoms caused by too much hormone production. This may be done by starting [somatostatin analog drugs or other medicines](#).

## Laparoscopy to stage the cancer

To determine which type of surgery might be best, it's important to know the [stage](#)<sup>1</sup> (extent) of the cancer. Sometimes it can be hard to stage pancreatic NETs accurately just using [imaging tests](#)<sup>2</sup>. Exploratory laparoscopy may be done first to help determine the extent of the cancer and if it can be resected.

For this procedure, the surgeon makes a few small incisions (cuts) in the abdomen (belly) and inserts long, thin instruments. One of these has a small video camera on the end so the surgeon can see inside the abdomen. The surgeon can look at the pancreas and other organs for tumors and take [biopsy](#)<sup>3</sup> samples of abnormal areas to learn how far the cancer has spread.

## Potentially curative surgery

Pancreatic NETs that have not spread outside the pancreas should be completely removed, if possible, because these tumors are more likely to be cured with surgery. Sometimes, however, after the surgeon starts the operation it becomes clear that the cancer has grown too far to be completely taken out. If this happens, the operation may be stopped, or the surgeon might continue with a smaller operation to help prevent or relieve symptoms. (See "Palliative surgery" below.) This is because the planned operation would be very unlikely to cure the cancer and could still lead to major side effects. It would also make the recovery time longer, which could delay other treatments.

Many of these types of surgery are complex and can be very hard for patients. They can cause complications and can take weeks or months to make a full recovery. If you're thinking about having this type of surgery, it's important to weigh the potential benefits

and risks carefully.

Types of potentially curative surgery include enucleation (removing only the tumor), central pancreatectomy, distal pancreatectomy, the Whipple procedure (pancreaticoduodenectomy), and total pancreatectomy. The type of surgery needed depends on several factors, including the location, size, and specific kind of pancreatic NET (functioning or nonfunctioning).

### **Enucleation (removing just the tumor)**

Sometimes if a pancreatic NET is small, just the tumor itself is removed. This is called *enucleation*. This operation may be done using a laparoscope, so that only a few small cuts on the belly are needed.

This operation may be all that is needed to treat an insulinoma. Small gastrinomas and some other pancreatic NETs may also be treated with enucleation, but sometimes the duodenum (the first part of the small intestine) is removed as well.

The [lymph nodes](#)<sup>4</sup> around the pancreas might also be removed so that they can be checked for cancer cells.

### **Central pancreatectomy**

A central pancreatectomy is used to treat small, low grade tumors. For this operation, the surgeon removes only the neck and part of the body of the pancreas keeping the head and tail intact. This helps maintain most of the function of the pancreas.

### **Distal pancreatectomy**

A distal pancreatectomy is used to treat pancreatic NETs found in the tail and body of the pancreas. In this operation, the surgeon removes only the tail of the pancreas or the tail and a portion of the body of the pancreas. The spleen is usually removed as well.

The spleen helps the body fight infections, so if it's removed you'll be at increased risk of infection with certain bacteria. To help with this, doctors recommend that patients get certain vaccines before this surgery.

### **Whipple procedure (pancreaticoduodenectomy)**

A Whipple procedure is used to treat pancreatic NETs found in the head of the pancreas. During this operation, the surgeon removes the head of the pancreas and

sometimes the body of the pancreas as well. Nearby structures such as part of the small intestine, part of the bile duct, the gallbladder, lymph nodes near the pancreas, and sometimes part of the stomach are also removed. The remaining bile duct and pancreas are then attached to the small intestine so that bile and digestive enzymes can still go into the small intestine. The pieces of the small intestine (or the stomach and small intestine) are then reattached so that food can pass through the digestive tract.

Most often, this operation is done through a large incision (cut) down the middle of the belly. Some doctors at major cancer centers also do the operation laparoscopically, which is sometimes known as *keyhole surgery* (see [What's New in Pancreatic Neuroendocrine Tumor Research?](#)<sup>5</sup>).

This is a very complex operation that requires a surgeon with a lot of skill and experience. It carries a relatively high risk of complications that can be life threatening. When the operation is done in small hospitals or by doctors with less experience, as many as 15% of patients may die as a result of surgical complications. In contrast, when the operation is done in cancer centers by surgeons experienced in the procedure, less than 5% of patients die as a direct result of surgery.

To have the best outcome, it's important to be treated by a surgeon who does many of these operations and to have the surgery at a hospital where many of them are done. In general, people having this type of surgery do better when it's done at a hospital where at least 15 to 20 Whipple procedures are done per year.

Still, even under the best circumstances, many patients have complications from the surgery. These can include:

- Leaking from the various connections between organs that the surgeon has joined
- Infections
- Bleeding
- Trouble with the stomach emptying after eating
- Trouble digesting some foods (which might require taking pancreatic enzymes in pill form to help with digestion)
- Weight loss
- Changes in bowel habits
- Diabetes

### **Total pancreatectomy**

Total pancreatectomy might be an option if the cancer has spread throughout the



pancreas but can still be removed. This operation removes the entire pancreas, as well as the gallbladder, part of the stomach and small intestine, and the spleen. But this type of surgery is used less often than the other operations because there doesn't seem to be a major advantage in removing the whole pancreas, and it can have major side effects.

It's possible to live without a pancreas. But when the entire pancreas is removed, people are left without the cells that make insulin and other hormones that help maintain safe blood sugar levels. These people develop diabetes, which can be hard to manage because they are totally dependent on insulin shots. People who have had this surgery also need to take pancreatic enzyme pills to help them digest certain foods.

Before you have this operation, your doctor will recommend that you get certain vaccines because the spleen will be removed.

## **Palliative surgery**

If the cancer has spread too far to be removed completely, any surgery being considered would be palliative (intended to relieve symptoms). This type of surgery may be considered in some people with pancreatic NETs whose tumor has recurred and is causing local problems or is making too many hormones that are causing symptoms.

Sometimes surgery might be started with the hope it will cure the patient, but once it begins the surgeon discovers this is not possible. In this case, the surgeon might do a less extensive, palliative operation known as *bypass surgery* instead to help prevent or relieve symptoms.

Cancers growing in the head of the pancreas can block the common bile duct as it passes through this part of the pancreas. This can cause pain and digestive problems because bile can't get into the intestine. The bile chemicals will also build up in the body, which can cause jaundice, nausea, vomiting, and other problems.

There are 2 main options for relieving bile duct blockage: stent placement, and bypass surgery.

## **Stent placement**

The most common approach to relieving a blocked bile duct does not involve actual surgery. Instead, a stent (small tube, usually made of metal) is put inside the duct to keep it open. This is usually done through an endoscope (a long, flexible tube) while you are sedated. Often this is part of an endoscopic retrograde

cholangiopancreatography (ERCP). The doctor passes the endoscope down the throat and all the way into the small intestine. The doctor can then insert the stent into the bile duct through the endoscope. The stent can also be put in place through the skin during a percutaneous transhepatic cholangiography (PTC). (These tests are described in [Tests for Pancreatic Neuroendocrine Tumor<sup>6</sup>](#).)

The stent helps keep the bile duct open even if the surrounding cancer presses on it. But after several months, the stent may become clogged and may need to be cleared or replaced. Larger stents can also be used to keep parts of the small intestine open if they are in danger of being blocked by the cancer.

A bile duct stent can also be put in to help relieve jaundice before curative surgery is done (which would typically be a couple of weeks later). This can help lower the risk of complications from surgery.

### **Bypass surgery**

In people who are healthy enough, another option for relieving a blocked bile duct is surgery to reroute the flow of bile from the common bile duct directly into the small intestine, bypassing the pancreas. This typically requires a large incision (cut) in the abdomen, and it can take weeks to recover from this. Sometimes surgery can be done through several small cuts in the abdomen using special long surgical tools. (This is known as *laparoscopic* or *keyhole surgery*.)

Having a stent placed is often easier and the recovery is much shorter, which is why this is done more often than bypass surgery. But this surgery can have some advantages:

- It can often give longer-lasting relief than a stent, which might need to be cleaned out or replaced.
- It might be an option if a stent can't be placed for some reason.
- During surgery, the surgeon may be able to cut some of the nerves around the pancreas or inject them with alcohol. This may reduce or get rid of any pain caused by the cancer.

Sometimes, the end of the stomach is disconnected from the duodenum (the first part of the small intestine) and attached farther down the small intestine during this surgery as well. (This is known as a *gastric bypass*.) This is done because over time the cancer might grow large enough to block the duodenum, which can cause pain and vomiting and often requires urgent surgery. Bypassing the duodenum before this happens can sometimes help avoid this.

Bypass surgery can still be a major operation, so it's important that you are healthy enough to withstand it and that you talk with your doctor about the possible benefits and risks before you have the surgery.

### **Surgery for cancer that has spread**

Surgery may be used to remove metastases if a pancreatic NET has spread to the liver (the most common site of spread) or the lungs. Surgically removing metastases can improve symptoms and help patients with pancreatic NETs live longer. In rare cases, a liver transplant might be used to treat pancreatic NETs that have spread to the liver.

### **More information about Surgery**

For more general information about surgery as a treatment for cancer, see [Cancer Surgery](#)<sup>7</sup>.

To learn about some of the side effects listed here and how to manage them, see [Managing Cancer-related Side Effects](#)<sup>8</sup>.

### **Hyperlinks**

1. </content/cancer/en/cancer/pancreatic-neuroendocrine-tumor/detection-diagnosis-staging/staging.html>
2. [www.cancer.org/cancer/pancreatic-neuroendocrine-tumor/detection-diagnosis-staging/how-diagnosed.html](http://www.cancer.org/cancer/pancreatic-neuroendocrine-tumor/detection-diagnosis-staging/how-diagnosed.html)
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## Ablation or Embolization Treatments for Pancreatic Neuroendocrine Tumor

Ablation and embolization treatments are different ways of destroying tumors, rather than removing them with surgery.

**When might one of these treatments be used?**

Ablation or embolization can sometimes be used to help treat pancreatic neuroendocrine tumor (NET) that has spread to other organs, especially the liver. When pancreatic NETs have spread to other sites, these treatments can often reduce tumor size and improve symptoms. But these treatments are very unlikely to cure cancers on their own. They are more likely to be used to help prevent or relieve symptoms, and are often used along with other types of treatment.

## Ablative treatments (ablation)

Ablation refers to treatments that destroy tumors, usually with extreme heat or cold. They are generally best for tumors no more than about 2 cm (a little less than an inch) across. There are different kinds of ablative treatments:

- **Radiofrequency ablation (RFA)** uses high-energy radio waves. A thin, needle-like probe is put through the skin and into the tumor. Placement of the probe is guided by an ultrasound or CT scan. The tip of the probe releases a high-frequency electric current which heats the tumor and destroys the cancer cells.
- **Microwave thermotherapy** is similar to RFA, except it uses microwaves to heat and destroy the cancer cells.
- **Ethanol (alcohol) ablation** (also known as *percutaneous ethanol injection*) kills the cancer cells with concentrated alcohol injected directly into the tumor. This is usually done using a needle through the skin, guided by ultrasound or CT scans.
- **Cryosurgery** (also known as *cryotherapy* or *cryoablation*) destroys a tumor by freezing it with a thin metal probe. The probe is guided through the skin and into the tumor using an ultrasound. Then very cold gasses are passed through the probe to freeze the tumor, killing the cancer cells. This method may be used to treat larger tumors than the other ablation techniques, but it sometimes requires general anesthesia (where you are asleep).

## Side effects of ablation treatments

Possible side effects after ablation therapy include abdominal pain, infection, and bleeding inside the body. Serious complications are uncommon, but they are possible.

## Embolization

During embolization, substances are injected into an artery to try to block the blood flow to cancer cells, causing them to die. This may be used for larger tumors (up to 5cm or 2

inches across) in the liver.

There are 3 main types of embolization:

- **Arterial embolization** (also known as *trans-arterial embolization* or *TAE*) involves putting a catheter (a thin, flexible tube) into an artery through a small cut in the inner thigh and threading it up into the hepatic artery feeding the tumor. Blood flow is blocked (or reduced) by injecting materials to plug up that artery. Most of the healthy liver cells will not be affected because they get their blood supply from a different blood vessel, the portal vein.
- **Chemoembolization** (also known as *trans-arterial chemoembolization* or *TACE*) combines embolization with [chemotherapy](#). Most often, this is done by using tiny beads that give off a chemotherapy drug during the embolization. TACE can also be done by giving chemotherapy through the catheter directly into the artery, then plugging up the artery.
- **Radioembolization** combines embolization with [radiation therapy](#). In the United States, this is done by injecting small radioactive beads (called *microspheres*) into the hepatic artery. The beads lodge in the blood vessels near the tumor, where they give off small amounts of radiation to the tumor site for several days. Since the radiation travels a very short distance, its effects are limited mainly to the tumor.

### Side effects of embolization

Possible complications after embolization include abdominal [pain](#)<sup>1</sup>, [fever](#)<sup>2</sup>, [nausea](#)<sup>3</sup>, [infection](#)<sup>4</sup>, and blood clots in nearby blood vessels. Serious complications are not common, but they can happen.

### Hyperlinks

1. [www.cancer.org/treatment/treatments-and-side-effects/physical-side-effects/pain.html](http://www.cancer.org/treatment/treatments-and-side-effects/physical-side-effects/pain.html)
2. [www.cancer.org/treatment/treatments-and-side-effects/physical-side-effects/fever.html](http://www.cancer.org/treatment/treatments-and-side-effects/physical-side-effects/fever.html)
3. [www.cancer.org/treatment/treatments-and-side-effects/physical-side-effects/nausea-and-vomiting.html](http://www.cancer.org/treatment/treatments-and-side-effects/physical-side-effects/nausea-and-vomiting.html)
4. [www.cancer.org/treatment/treatments-and-side-effects/physical-side-effects/infections.html](http://www.cancer.org/treatment/treatments-and-side-effects/physical-side-effects/infections.html)

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# Radiation Therapy for Pancreatic Neuroendocrine Tumor

Radiation therapy uses high-energy rays (such as x-rays) or radioactive particles to kill cancer cells.

**Surgery** is the main treatment for most pancreatic neuroendocrine tumors (NETs), but radiation therapy may be an option for those who can't have surgery for some reason. It may also be given after surgery in some cases if there's a chance some of the tumor was not removed and is causing problems. Radiation is sometimes used to treat pancreatic NETs that have spread to the bone and are causing pain. It may also be

used in the form of radioembolization to treat NETs that have spread to the liver. (See [Ablation or Embolization Treatments for Pancreatic Neuroendocrine Tumor](#).)

## External beam radiation therapy

[External beam radiation therapy](#)<sup>1</sup> uses a machine that delivers a beam of radiation to a specific part of the body.

Before your treatment starts, the radiation team will determine the correct angles for aiming the radiation beams and the proper dose of radiation. The treatment is much like getting an x-ray, but the radiation is stronger. The procedure itself is painless. Each treatment lasts only a few minutes, although the setup time – getting you into place for treatment – usually takes longer. Most often, radiation treatments are given 5 days a week for several weeks, but this can vary based on the reason it's being given.

Some common side effects of radiation therapy include:

- Skin changes in areas getting radiation, ranging from redness to blistering and peeling
- Nausea and vomiting
- Diarrhea
- Fatigue
- Loss of appetite
- Weight loss
- Low blood counts, which can increase the risk of serious infection.

Usually these side effects go away within a few weeks after the treatment is complete. Ask your doctor what side effects to expect and how to prevent or relieve them.

## Radioactive drugs

### Radioembolization

Radioembolization combines embolization with radiation therapy and can be used to treat liver metastases. Small beads called *microspheres* are attached to a radioactive element called *yttrium-90* (or *90Y*) and then injected into an artery close to the liver. The beads travel in the liver blood vessels until they get stuck in small blood vessels near the tumor. There they give off radioactivity for a short while, killing nearby tumor cells. The radiation travels a very short distance, so its effects are limited mainly to the tumor.



## Peptide receptor radionuclide therapy (PRRT)

People with somatostatin receptor-positive neuroendocrine tumors may be candidates for PRRT. In PRRT, a radioactive element is linked to a small part (peptide) of a [somatostatin analog](#), and injected into a vein in the arm. The drug travels throughout the body, attaches to the somatostatin receptor (a protein) on the cancer cell, and gives off radiation to kill it. The radiation is delivered directly to the tumor, so there is less effect on healthy tissue. There are several drugs that might be used:

- The radioactive element **Yttrium-90**
- The radioactive element **Lutathera (lutetium or Lu-177 dotatate)**

If you are already taking octerotide or lantreotide, you will most likely need to stop taking these medicines for a certain time before you can be treated with PRRT.

Common side effects of PRRT include low levels of white blood cells, abnormal liver tests, nausea and vomiting, high levels of blood sugar, and pain.

Serious side effects include low levels of blood cells, development of certain blood or bone marrow cancers, kidney damage, liver damage, abnormal levels of hormones in the body, and infertility. Tell your cancer care team if you are pregnant or might become pregnant, because Lu-177 dotatate can harm the baby. There is not enough information regarding Yttrium-90 in pregnant women so you should discuss this with your doctor.

Since these drugs expose you to radiation, people who might come into contact with you need to follow certain radiation safety practices to limit their exposure. See [Systemic Radiation Therapy](#)<sup>2</sup> for more information.

## More information about radiation therapy

To learn more about how radiation is used to treat cancer, see [Radiation Therapy](#)<sup>3</sup>.

To learn about some of the side effects listed here and how to manage them, see [Managing Cancer-related Side Effects](#)<sup>4</sup>.

## Hyperlinks

1. [www.cancer.org/treatment/treatments-and-side-effects/treatment-types/radiation/external-beam-radiation-therapy.html](http://www.cancer.org/treatment/treatments-and-side-effects/treatment-types/radiation/external-beam-radiation-therapy.html)
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## Chemotherapy for Pancreatic Neuroendocrine Tumor

Chemotherapy (chemo) uses anti-cancer drugs injected into a vein or taken by mouth to kill cancer cells. These drugs enter the bloodstream and reach almost all areas of the body, making this treatment useful for some types of cancers that have spread.

Chemo is most often used to treat pancreatic neuroendocrine tumors (NETs) if they:

- Have not responded to other medicines (such as [somatostatin drugs](#) or [targeted therapy](#)),
- Have spread to other organs,
- Are large or growing quickly,
- Are causing severe symptoms, **or**
- Are high grade (grade 3)

The most commonly used drugs for pancreatic NETs include:

- Doxorubicin (Adriamycin)
- Streptozocin
- Fluorouracil (5-FU)
- Dacarbazine (DTIC)
- Temozolomide (Temodar)
- Capecitabine (Xeloda)
- Oxaliplatin (Eloxatin)

Some tumors might be treated with more than one drug. Possible combinations include:

- Doxorubicin plus streptozocin

- 5-FU plus doxorubicin plus streptozocin
- Temozolomide plus capecitabine
- 5-FU plus streptozocin

## How is chemotherapy given?

Chemo drugs are typically given into a vein (IV), either as an injection over a few minutes or as an infusion over a longer period of time. This can be done in a doctor's office, chemotherapy clinic, or in a hospital setting.

Doctors give chemo in cycles, with each period of treatment followed by a rest period to give you time to recover from the effects of the drugs. Cycles are most often 2 or 3 weeks long. The schedule varies depending on the drugs used. For example, with some drugs, the chemo is given only on the first day of the cycle. With others, it is given for a few days in a row, or once a week. Then, at the end of the cycle, the chemo schedule repeats to start the next cycle.

The length of treatment for advanced pancreatic NETs is based on how well it is working and what side effects you have.

## Possible side effects of chemotherapy

Chemo drugs attack cells that are dividing quickly, which is why they work against cancer cells. But other cells in the body, such as those in the bone marrow (where new blood cells are made), the lining of the mouth and intestines, and the hair follicles, also divide quickly. These cells are also likely to be affected by chemo, which can lead to side effects.

The side effects of chemo depend on the type and dose of drugs given and the length of time they are taken. Common side effects can include:

- Nausea and vomiting
- Loss of appetite
- Hair loss
- Mouth sores
- Diarrhea or constipation
- Increased chance of infections (from having too few white blood cells)
- Easy bruising or bleeding (from having too few blood platelets)

- Fatigue (from having too few red blood cells)

Most side effects go away after treatment is finished. Tell your cancer care team about any side effects or changes you notice while getting chemotherapy, so that they can be treated promptly. Often medicines can help prevent or minimize many of the side effects. For example, your doctor can prescribe drugs to help prevent or reduce nausea and vomiting. In some cases, the doses of the chemo drugs might need to be lowered or treatment might need to be delayed or stopped to keep the effects from getting worse.

## More information about chemotherapy

For more general information about how chemotherapy is used to treat cancer, see [Chemotherapy](#)<sup>1</sup>.

To learn about some of the side effects listed here and how to manage them, see [Managing Cancer-related Side Effects](#)<sup>2</sup>.

## Hyperlinks

1. [www.cancer.org/treatment/treatments-and-side-effects/treatment-types/chemotherapy.html](http://www.cancer.org/treatment/treatments-and-side-effects/treatment-types/chemotherapy.html)
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## Targeted Therapy for Pancreatic Neuroendocrine Tumor

Targeted drugs work differently from standard chemo drugs. These drugs target specific parts of cancer cells. They are sometimes helpful when chemotherapy is not and often have different side effects than chemotherapy.

The targeted drugs used to treat pancreatic neuroendocrine tumors (NETs) work by blocking angiogenesis (the growth of new blood vessels that nourish cancers) or important proteins (called *tyrosine kinases*) in cancer cells that help them grow and survive.

### Sunitinib (Sutent)

Sunitinib blocks several tyrosine kinases and attacks new blood vessel growth. It has been shown to help slow tumor growth. This drug is taken as a pill once a day.

The most common side effects are nausea, diarrhea, changes in skin or hair color, mouth sores, weakness, and low blood cell counts. Other possible effects include tiredness, high blood pressure, heart problems, bleeding, hand-foot syndrome (redness, pain, and skin peeling of the palms of the hands and the soles of the feet), and low thyroid hormone levels.

## Everolimus (Afinitor)

Everolimus blocks a protein known as *mTOR*, which normally helps cells grow and divide. It has been shown to help treat advanced pancreatic NETs. Everolimus is a pill taken once a day.

Common side effects of this drug include mouth sores, infections, nausea, loss of appetite, diarrhea, skin rash, feeling tired or weak, fluid buildup (usually in the legs), and increases in blood sugar and cholesterol levels. A less common but serious side effect is damage to the lungs, which can cause shortness of breath or other problems.

## More information about targeted therapy

To learn more about how targeted drugs are used to treat cancer, see [Targeted Cancer Therapy](#)<sup>1</sup>.

To learn about some of the side effects listed here and how to manage them, see [Managing Cancer-related Side Effects](#)<sup>2</sup>.

## Hyperlinks

1. [www.cancer.org/treatment/treatments-and-side-effects/treatment-types/targeted-therapy.html](http://www.cancer.org/treatment/treatments-and-side-effects/treatment-types/targeted-therapy.html)
2. [www.cancer.org/treatment/treatments-and-side-effects/physical-side-effects.html](http://www.cancer.org/treatment/treatments-and-side-effects/physical-side-effects.html)

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## Other Drugs for Pancreatic Neuroendocrine Tumors

For people with advanced pancreatic neuroendocrine tumors (NETs), several medicines can help control symptoms and tumor growth. These drugs are used mainly when the



tumor can't be removed with surgery.

## Somatostatin analogs

Somatostatin analogs are related to somatostatin, a natural hormone in the body. They can help slow the growth of neuroendocrine tumor cells. They can be very helpful for some patients with pancreatic NETs because these drugs stop tumors from releasing hormones into the bloodstream, which can often relieve symptoms and help patients feel better. They also seem to help slow the growth of some tumors, but cannot cure them.

These drugs can help reduce diarrhea in patients with VIPomas, glucagonomas, and somatostatinomas, help the rash of glucagonomas, and lower the levels of insulin in insulinomas. They are very useful in people who have [carcinoid syndrome](#)<sup>1</sup> (facial flushing, diarrhea, wheezing, rapid heart rate), although this syndrome is not as common with NETs in the pancreas as it is with NETs found in other places. The drugs are also helpful for people whose tumors show up on a somatostatin receptor scintigraphy (SRS) scan or gallium-68 Dotatate scans.

- **Octreotide (Sandostatin):** One version of octreotide is short-acting and is injected 2 to 4 times a day under the skin. There is also a long-acting form of the drug (called Sandostatin LAR Depot) that only needs to be given once a month, by injection into a muscle. Depending on the severity of symptoms, some people are given injections every day when first starting treatment. Once symptoms are controlled, the longer-acting monthly injection may then be used. Other times, the long acting drug may be started from the beginning.
- **Lanreotide (Somatuline Depot):** This somatostatin analog is injected under the skin about once a month.

Either drug may be given by your doctor or nurse, or you may learn how to give the injection at home.

### Possible side effects

The main side effects of these drugs are pain at the site of the injection, and rarely, stomach cramps, nausea, vomiting, headaches, dizziness, and fatigue. These drugs can also cause sludge to build up in the gallbladder, which can lead to gallstones. They can also make the body resistant to the action of insulin, which can raise blood sugar levels and make pre-existing diabetes harder to control. As a result, these drugs are only used to treat insulinomas if the tumor has somatostatin receptors as seen by a

positive somatostatin receptor scintigraphy (SRS) or gallium-68 Dotatate scan.

## Other drugs used for specific pancreatic NETs

Somatostatin analogs can be used to treat most pancreatic NETs. But other drugs may be added to treat specific symptoms or problems that are caused by the excess hormone being produced by the cancer.

**Gastrinomas** make too much gastrin, which increases stomach acid levels, and can lead to stomach ulcers. Proton pump inhibitors, for example omeprazole (Prilosec), esomeprazole (Nexium), or lansoprazole (Prevacid), block stomach acid production and may be given to decrease the chance of ulcers forming.

**Insulinomas** make too much insulin which causes very low blood glucose (sugar) levels. If the somatostatin receptor scintigraphy (SRS) or gallium-68 Dotatate scans are negative, showing the cancer does not have the somatostatin protein, then other treatments besides somatostatin analogs are considered to even out glucose levels. Diazoxide, a drug that keeps insulin from being released into the bloodstream, or diet changes (higher carbohydrate intake or more frequent meals) may be started to raise glucose levels.

**Glucagonomas** make too much glucagon, a hormone that increases blood glucose (sugar) levels. It works opposite of insulin. These cancers may be treated with medicines for diabetes if somatostatin analogs alone are not enough to control the high glucose levels.

**VIPomas** make too much vasoactive intestinal peptide (VIP), a hormone that regulates water and mineral (such as potassium and magnesium) levels in the gut. Treatment may involve giving intravenous (IV) fluids to treat the dehydration from diarrhea as well as certain minerals that are low.

## Hyperlinks

1. [www.cancer.org/cancer/gastrointestinal-carcinoid-tumor/detection-diagnosis-staging/signs-symptoms.html](http://www.cancer.org/cancer/gastrointestinal-carcinoid-tumor/detection-diagnosis-staging/signs-symptoms.html)

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## Treating Pancreatic Neuroendocrine Tumor, Based on Extent of the Tumor

Treatment of pancreatic neuroendocrine tumors (NETs) depends to a large extent on whether they can be resected (removed) completely or not. But other factors, such as your overall health, can also affect treatment options. Talk to your doctor if you have any questions about the treatment plan he or she recommends.

Sometimes it can be hard to determine if cancer is resectable – that is, if it can be removed completely – using just [imaging tests](#)<sup>1</sup>. A laparoscopy might be done before surgery to help determine if the tumor can be removed. But even then, cancers sometimes turn out to have spread farther than was first thought.

Pancreatic NETs are more likely to be resectable than [exocrine pancreas cancers](#)<sup>2</sup> (the most common type of pancreatic cancer). Most NETs that have not spread to distant parts of the body are resectable. Even some NETs that have spread might be resectable if they have not spread too far (such as only to a few spots in the liver).

## Treating resectable tumors

If the tumor is resectable, [surgery](#)<sup>3</sup> will be recommended. The procedure used depends on the type of tumor, its size, and its location in the pancreas. Surgery can range from as little as enucleation (removing just the tumor) to as much as a Whipple procedure (pancreaticoduodenectomy). Lymph nodes are often removed to check for tumor spread.

Before any surgery, [medicines](#) are often given to control any symptoms caused by the tumor. For example, drugs to block stomach acid (like proton pump inhibitors) are used for gastrinomas. Often, people with insulinomas are treated with diazoxide to keep blood sugar from getting too low. If the tumor was visible on [somatostatin receptor scintigraphy](#)<sup>4</sup>, a somatostatin analog such as octreotide may be used to control any symptoms.

Surgery alone is all that is needed for many pancreatic NETs, but after surgery, close monitoring is important to look for signs that the cancer may have come back or spread.

## Treating unresectable tumors

Unresectable tumors can't be removed completely with surgery. Pancreatic NETs are often slow growing, so lab and imaging tests are used to monitor the tumor(s) and look for signs of growth.

People with NETs that have spread outside the pancreas often have symptoms like diarrhea or hormone problems. These can often be helped with [medicines like](#)

[octreotide](#), [lanreotide](#), [diazoxide](#), and [proton pump inhibitors](#). Some of these might also slow the growth of the tumor.

If further treatment is needed, [chemotherapy](#) or [targeted drugs](#) (such as sunitinib or everolimus) might be used, but this is usually delayed until a person is having symptoms that can't be controlled with other drugs or has signs of tumor growth on scans. [Surgery](#) or [ablative techniques](#)<sup>5</sup> might also be used to treat cancer spread to the liver.

For people with poorly differentiated tumors (neuroendocrine carcinomas), chemotherapy is typically the first treatment.

For adults with somatostatin (a type of hormone) receptor-positive pancreatic neuroendocrine tumors, a [radiopharmaceutical drug](#), called Lutathera (lutetium Lu 177 dotatate), is also an option for treatment.

If treatment is no longer working at some point, you might want to think about taking part in a [clinical trial](#)<sup>6</sup> testing a newer treatment. While these are not always the best option for every person, they may benefit you as well as future patients.

*The treatment information given here is not official policy of the American Cancer Society and is not intended as medical advice to replace the expertise and judgment of your cancer care team. It is intended to help you and your family make informed decisions, together with your doctor. Your doctor may have reasons for suggesting a treatment plan different from these general treatment options. Don't hesitate to ask him or her questions about your treatment options.*

## Hyperlinks

1. </content/cancer/en/cancer/pancreaticcancer/detailedguide/pancreatic-cancer-diagnosis.html>
2. [www.cancer.org/cancer/pancreatic-cancer.html](http://www.cancer.org/cancer/pancreatic-cancer.html)
3. </content/cancer/en/cancer/pancreaticcancer/detailedguide/pancreatic-cancer-treating-surgery.html>
4. </content/cancer/en/cancer/pancreaticcancer/detailedguide/pancreatic-cancer-diagnosis.html>
5. </content/cancer/en/cancer/pancreaticcancer/detailedguide/pancreatic-cancer-treating-ablative-techniques.html>
6. [www.cancer.org/treatment/treatments-and-side-effects/clinical-trials.html](http://www.cancer.org/treatment/treatments-and-side-effects/clinical-trials.html)

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# After Pancreatic Neuroendocrine Tumor Treatment

## Living as a Cancer Survivor

For many people, cancer treatment often raises questions about next steps as a survivor.

- [Living as a Pancreatic Neuroendocrine Tumor Survivor](#)

## Cancer Concerns After Treatment

Treatment may remove or destroy the cancer, but it's very common to worry about the risk of developing another cancer.

- [Second Cancers after Pancreatic Neuroendocrine Tumors](#)
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# Living as a Pancreatic Neuroendocrine Tumor Survivor

For some people with a pancreatic neuroendocrine tumor (NET), treatment can remove or destroy the cancer. Completing treatment can be both stressful and exciting. You may be relieved to finish treatment, but find it hard not to worry about cancer coming back. This is very common if you've had cancer.



For other people, the cancer might never go away completely, or it might come back in another part of the body. These people may stay on drug therapy or get regular treatments with chemotherapy, radiation therapy, or other therapies to help keep the cancer under control for as long as possible. Learning to live with cancer that does not go away can be difficult and very stressful.

## Follow-up care

When treatment ends, your doctors will still want to watch you closely. It is very important to go to all your follow-up appointments. During these visits, your doctors will ask if you are having any problems, examine you, and order lab tests or x-rays and scans if needed to look for signs of cancer or treatment side effects. Almost any cancer treatment can have [side effects](#)<sup>1</sup>. Some may last for a few weeks to months, but others might last a long time. Some side effects might not even show up until years after you have finished treatment. It's important for all pancreatic NET survivors, to let their health care team know about any new symptoms or problems, because they could be caused by the cancer coming back or by a new disease or [second cancer](#).

## Doctor visits and follow-up tests

Your schedule of doctor visits, exams, and tests will depend on the original extent of your cancer, the specific type of pancreatic NET you had, how it was treated, and other factors.

Doctors often advise most people who have had their pancreatic NET completely removed to return in 6-12 months for a complete physical exam and certain imaging tests to look for any signs of recurrence. Blood and or urine tests may be helpful for some patients. One year after surgery, further visits with labs and imaging may be recommended every 6-12 months to continue for 10 years.

Follow-up visits and imaging tests may be slightly more frequent if your cancer could not be completely removed with surgery, or if the cancer has spread to other organs like the liver or is growing very quickly.

## Ask your doctor for a survivorship care plan

Talk with your doctor about developing a [survivorship care plan](#)<sup>2</sup> for you. This plan might include:

- A suggested schedule for follow-up exams and tests
- A list of possible late- or long-term side effects from your treatment, including what

- to watch for and when you should contact your doctor
- A schedule for other tests you might need, such as [early detection \(screening\) tests](#)<sup>3</sup> for other types of cancer, or tests to look for long-term health effects from your cancer or its treatment
  - Diet and physical activity suggestions that might improve your health, including possibly lowering your chances of the cancer coming back
  - Reminders to keep your appointments with your primary care provider (PCP), who will monitor your general health care

## Keeping health insurance and copies of your medical records

Even after treatment, it's very important to keep [health insurance](#)<sup>4</sup>. Tests and doctor visits cost a lot, and even though no one wants to think of their cancer coming back, this could happen.

At some point after your cancer treatment, you might find yourself seeing a new doctor who doesn't know about your medical history. It's important to keep copies of your medical records to give your new doctor the details of your diagnosis and treatment. Learn more in [Keeping Copies of Important Medical Records](#)<sup>5</sup>.

## Can I lower the risk of my cancer progressing or coming back?

If you have (or have had) pancreatic NET, you probably want to know if there are things you can do that might lower your risk of the cancer growing or coming back, such as exercising, eating a certain type of diet, or taking nutritional supplements. Unfortunately, it's not yet clear if there are things you can do that will help.

Adopting healthy behaviors such as [not smoking](#)<sup>6</sup>, [eating well](#)<sup>7</sup>, [getting regular physical activity](#)<sup>8</sup>, and [staying at a healthy weight](#)<sup>9</sup> is important. We know that these types of changes can have positive effects on your health that can extend beyond your risk of cancer.

### Quitting smoking

Smoking has been linked to pancreas NET, so not smoking may help reduce your risk. We don't know for certain if this will help, but we do know that [quitting smoking](#)<sup>10</sup> can have other health benefits such as improved healing, lowering your risk of some other cancers, as well as improving your outcome (prognosis) from the cancer. If you want to

quit smoking and need help, call the American Cancer Society at 1-800-227-2345.

### **About dietary supplements**

So far, no [dietary supplements](#)<sup>11</sup> (including vitamins, minerals, and herbal products) have been shown to clearly help lower the risk of pancreatic NET progressing or coming back. This doesn't mean that no supplements will help, but it's important to know that none have been proven to do so.

Dietary supplements are not regulated like medicines in the United States – they do not have to be proven effective (or even safe) before being sold, although there are limits on what they're allowed to claim they can do. If you're thinking about taking any type of nutritional supplement, talk to your health care team. They can help you decide which ones you can use safely while avoiding those that might be harmful.

### **If the cancer comes back**

If your cancer does come back at some point, your treatment options will depend on the where the cancer is, what treatments you've had before, and your current health and preferences. Treatment options might include surgery, radiation therapy, chemotherapy, targeted therapy or some combination of these. For more on how recurrent cancer is treated, see [Treatment of Pancreatic Neuroendocrine Tumor, Based on Extent of the Tumor](#)<sup>12</sup>.

For more general information on recurrence, see [Understanding Recurrence](#)<sup>13</sup>.

### **Second cancers after treatment**

People who've had a pancreatic NET might still get other cancers. Learn more in [Second Cancers After Pancreatic Neuroendocrine Tumor](#).

### **Getting emotional support**

Some amount of feeling depressed, anxious, or worried is normal when pancreatic NET is a part of your life. Some people are affected more than others. But everyone can benefit from help and support from other people, whether friends and family, religious groups, support groups, professional counselors, or others. Learn more in [Life After Cancer](#)<sup>14</sup>.

### **Hyperlinks**

1. [www.cancer.org/treatment/treatments-and-side-effects/physical-side-effects.html](http://www.cancer.org/treatment/treatments-and-side-effects/physical-side-effects.html)
2. [www.cancer.org/treatment/survivorship-during-and-after-treatment/survivorship-care-plans.html](http://www.cancer.org/treatment/survivorship-during-and-after-treatment/survivorship-care-plans.html)
3. [www.cancer.org/healthy/find-cancer-early/cancer-screening-guidelines.html](http://www.cancer.org/healthy/find-cancer-early/cancer-screening-guidelines.html)
4. [www.cancer.org/treatment/finding-and-paying-for-treatment/understanding-health-insurance.html](http://www.cancer.org/treatment/finding-and-paying-for-treatment/understanding-health-insurance.html)
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6. [www.cancer.org/healthy/stay-away-from-tobacco.html](http://www.cancer.org/healthy/stay-away-from-tobacco.html)
7. [www.cancer.org/healthy/eat-healthy-get-active/eat-healthy.html](http://www.cancer.org/healthy/eat-healthy-get-active/eat-healthy.html)
8. [www.cancer.org/healthy/eat-healthy-get-active/get-active.html](http://www.cancer.org/healthy/eat-healthy-get-active/get-active.html)
9. [www.cancer.org/healthy/eat-healthy-get-active/take-control-your-weight.html](http://www.cancer.org/healthy/eat-healthy-get-active/take-control-your-weight.html)
10. [www.cancer.org/healthy/stay-away-from-tobacco.html](http://www.cancer.org/healthy/stay-away-from-tobacco.html)
11. [www.cancer.org/treatment/treatments-and-side-effects/complementary-and-alternative-medicine/dietary-supplements.html](http://www.cancer.org/treatment/treatments-and-side-effects/complementary-and-alternative-medicine/dietary-supplements.html)
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## Second Cancers after Pancreatic Neuroendocrine Tumors

Survivors of pancreatic neuroendocrine tumors (NETs) can be affected by a number of health problems, but often their greatest concern is facing another cancer. Cancer that comes back after treatment is called a *recurrence*. But some cancer survivors develop a new, unrelated cancer later. This is called a *second cancer*.

Unfortunately, being treated for one cancer doesn't mean you can't get another. People who have had a pancreatic NET can still get the same types of cancers that other people get. In fact, they might be at higher risk for certain types of cancer.

People who have or had a pancreatic NET can get any type of second cancer, but they have a higher risk than the general population of developing:

- [Prostate cancer](#)<sup>1</sup>
- [Female breast cancer](#)<sup>2</sup>
- [Colon and rectal cancer](#)<sup>3</sup>
- [Lung cancer](#)<sup>4</sup>

### What can you do?

Many people with a pancreatic NET are treated with medicines that keep the disease in

check without curing the disease, so they need to see their doctors regularly. Let your doctor know if you have any new symptoms or problems. They could be from the pancreatic NET getting worse or from a new disease or cancer.

All people with a pancreatic NET should [avoid tobacco smoke](#)<sup>5</sup>. Smoking is linked to an increased risk of many cancers and might further increase the risk of some of the second cancers seen in patients with pancreatic NETs. They should also keep up with [early detection \(screening\) tests](#)<sup>6</sup> for other types of cancer.

To help maintain good health, survivors should also:

- Get to and stay at a [healthy weight](#)<sup>7</sup>
- Adopt a [physically active lifestyle](#)<sup>8</sup>
- Eat a [healthy diet](#)<sup>9</sup>, with an focus on plant foods
- Limit use of [alcohol](#)<sup>10</sup> to no more than 1 drink per day for women or 2 per day for men

These steps may also lower the risk of some cancers.

See [Second Cancers in Adults](#)<sup>11</sup> for more information about causes of second cancers.

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1. [www.cancer.org/cancer/prostate-cancer.html](http://www.cancer.org/cancer/prostate-cancer.html)
2. [www.cancer.org/cancer/breast-cancer.html](http://www.cancer.org/cancer/breast-cancer.html)
3. [www.cancer.org/cancer/colon-rectal-cancer.html](http://www.cancer.org/cancer/colon-rectal-cancer.html)
4. [www.cancer.org/cancer/lung-cancer.html](http://www.cancer.org/cancer/lung-cancer.html)
5. [www.cancer.org/cancer/cancer-causes/tobacco-and-cancer.html](http://www.cancer.org/cancer/cancer-causes/tobacco-and-cancer.html)
6. [www.cancer.org/healthy/find-cancer-early/cancer-screening-guidelines.html](http://www.cancer.org/healthy/find-cancer-early/cancer-screening-guidelines.html)
7. [www.cancer.org/cancer/cancer-causes/diet-physical-activity/body-weight-and-cancer-risk.html](http://www.cancer.org/cancer/cancer-causes/diet-physical-activity/body-weight-and-cancer-risk.html)
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