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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

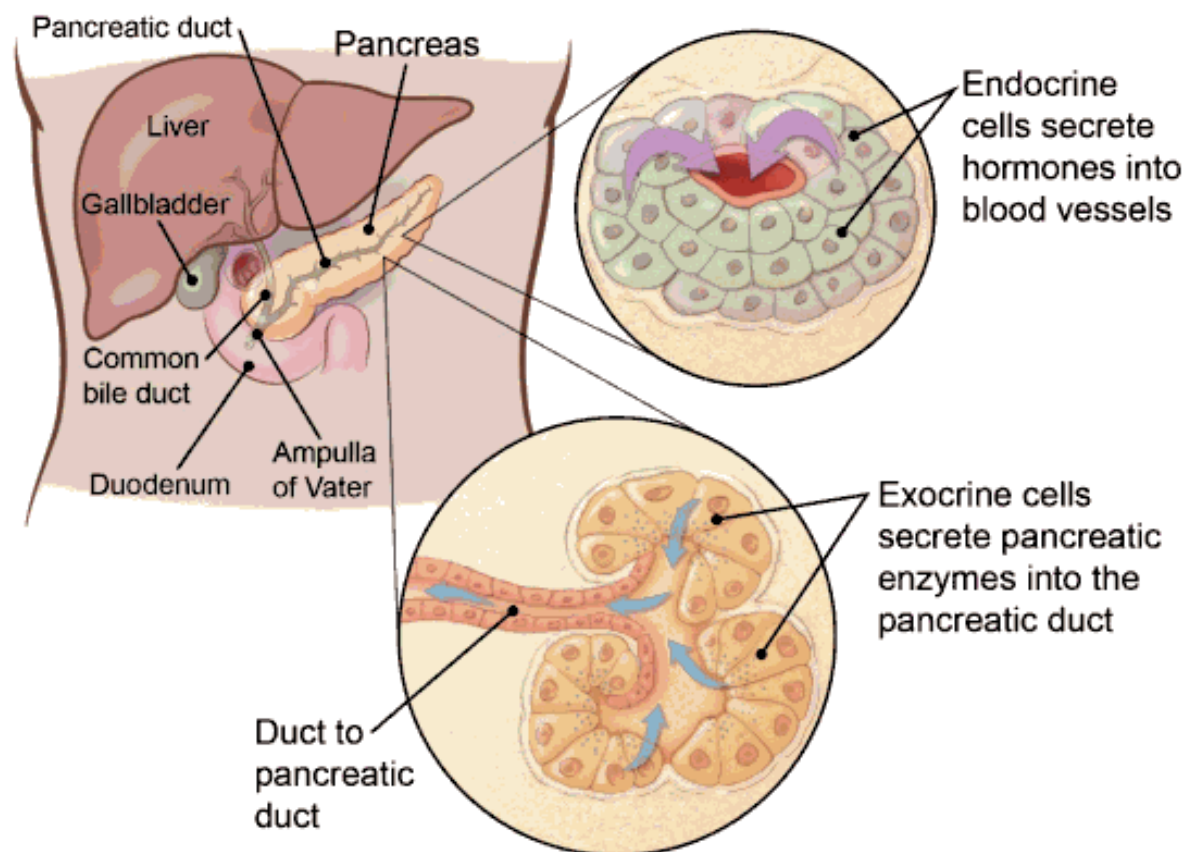
Neuroendocrine cells are like nerve cells in some ways and like hormone-making endocrine cells in other ways. Cells in this system don't form actual organs. Instead, they are scattered throughout other organs like the esophagus, stomach, pancreas, intestines, and lungs.

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.

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

Most (up to 70%) functioning NETs are insulinomas. The other types are much less common.

**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)
7. [www.cancer.org/cancer/pancreatic-neuroendocrine-tumor/treating/other-drugs.html](http://www.cancer.org/cancer/pancreatic-neuroendocrine-tumor/treating/other-drugs.html)
8. [www.cancer.org/cancer/gastrointestinal-carcinoid-tumor.html](http://www.cancer.org/cancer/gastrointestinal-carcinoid-tumor.html)
9. [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)

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## Key Statistics for Pancreatic Neuroendocrine Tumor

Pancreatic neuroendocrine tumors (NETs) are rare and account for about 7% of all cancers that occur in the pancreas. The American Cancer Society's estimates predict that about 4,032 people in the United States will be diagnosed with pancreatic NET in 2020.

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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