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About Pancreatic Neuroendocrine Tumors

Get an overview of pancreatic neuroendocrine tumors and the latest key statistics in the US.

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

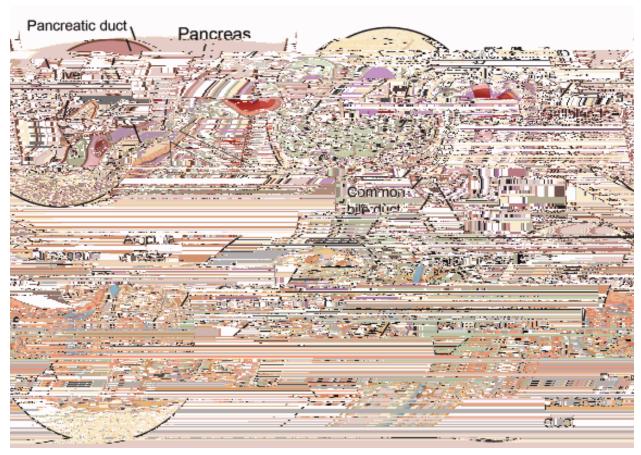
See the latest estimates for new cases of pancreatic neuroendocrine tumors and deaths in the US and what research is currently being done.

- Key Statistics for Pancreatic Neuroendocrine Tumor
- What's New in Pancreatic Neuroendocrine Tumor Research?

What Is a Pancreatic Neuroendocrine Tumor?

- Where pancreatic neuroendocrine tumors start
- Types of pancreatic neuroendocrine tumors

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 ______



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 <u>Pancreatic Cancer</u>³ 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 Cancer⁴) 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.

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 <u>symptoms</u>⁶. These are called functioningNETs. Each one is named for the <u>type of hormone</u>⁷ 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

https://www.cancer.gov/types/pancreatic/patient/pnet-treatment-pdq on October 10, 2018.

Schneider DF, Mazeh H, Lubner SJ, Jaume JC, Chen H. Chapter 71: Cancer of the endocrine system. In: Niederhuber JE, Armitage JO, Dorshow JH, Kastan MB, Tepper JE, eds. *Abeloff's Clinical Oncology*. 5th ed. Philadelphia, Pa. Elsevier: 2014.

Strosberg JR. Classification, epidemiology, clinical presentation, localization, and staging of pancreatic neuroendocrine neoplasms. UpToDate website. https://www.uptodate.com/contents/classification-epidemiology-clinical-presentation-localization-and-staging-of-pancreatic-neuroendocrine-neoplasms. Updated Jan. 23, 2018. Accessed October 10, 2018.

Yao JC, Evans DB. Chapter 85: Pancreatic neuroendocrine tumors. In: DeVita VT, Lawrence TS, Rosenberg SA, eds. *DeVita, Hellman, and Rosenberg's Cancer: Principles and Practice of Oncology.* 10th ed. Philadelphia, Pa: Lippincott Williams & Wilkins; 2015.

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Key Statistics for Pancreatic Neuroendocrine Tumor

Pancreatic neuroendocrine tumors (NETs) are rare. Less than 2% of all cancers found in the pancreas each year are pancreatic NETs.

However, the number of pancreatic NETs diagnosed each year has been rising over time. This is thought to be partly because they are being found more often incidentally, when imaging tests such as CT or MRI scans of the abdomen are done for other reasons. The ability to distinguish these tumors from other types of cancers in the lab has also improved, leading to more of them being diagnosed.

Most people with pancreatic NETs are older, with the average age at diagnosis being 60.

For statistics related to survival, see <u>Pancreatic Neuroendocrine Tumor Survival Rates</u> by Stage.¹

Visit our Cancer Statistics Center² for more key statistics.

Hyperlinks

- 1. <u>www.cancer.org/cancer/types/pancreatic-neuroendocrine-tumor/detection-diagnosis-staging/survival-rates.html</u>
- 2. cancerstatisticscenter.cancer.org

References

American Joint Committee on Cancer. Neuroendocrine Tumors of the Pancreas. *AJCC Cancer Staging Manual*. 8th ed. New York, NY: Springer; 2017.

Das S, Dasari A. Epidemiology, Incidence, and Prevalence of Neuroendocrine Neoplasms: Are There Global Differences? *Curr Oncol Rep.* 2021 Mar 14;23(4):43. doi: 10.1007/s11912-021-01029-7. PMID: 33719003; PMCID: PMC8118193.

National Cancer Institute. Pancreatic Neuroendocrine Tumors (Islet Cell Tumors) Treatment (PDQ®). 8/19/22. Accessed at https://www.cancer.gov/types/pancreatic/hp/pnet-treatment-pdq on August 4, 2024.

Last Revised: August 22, 2024

What's New in Pancreatic Neuroendocrine Tumor Research?

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

- Genetics and early detection
- Treatment

Genetics and early detection Tr opy delookises of pancreatiwh hapoe able fTradiy deallys (MEN1,)Tj 0 g 1 0 48.905

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. Scientists have found changes in the MEN1, VHL, NF1, and TSC genes in many people with pancreatic NETs. Other genetic changes that seem to make tumors more aggressive are now being explored as well.

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Written by

The American Cancer Society medical and editorial content team (https://www.cancer.org/cancer/acs-medical-content-and-news-staff.html)

Our team is made up of doctors and oncology certified nurses with deep knowledge of cancer care as well as editors and translators with extensive experience in medical writing.

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