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Pancreatic Cancer

Endocrine Pancreatic Cancer

Definitions

Endocrine: Relating to the endocrine glands and the hormones, such as insulin, which are secreted by those glands.

Islet cells: Pancreatic cells that produce insulin and other hormones.

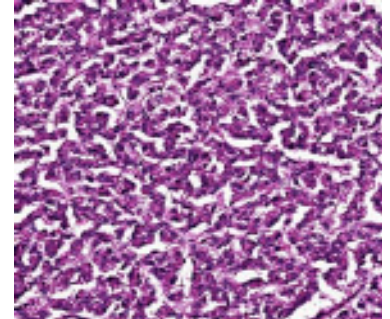
Malignant: Cancerous and capable of spreading.

Pathologist: A physician who examines tissues and fluids to diagnose disease in order to assist in making treatment decisions.

What is endocrine pancreatic cancer?

Endocrine pancreatic cancer, also known as islet cell cancer, is a rare cancer. Only about 5 percent of the more than 43,000 annual cases of pancreatic cancers begin in the islet cells. When islet cells become **neoplastic**, they may make too much insulin and other types of hormones. Different types of endocrine pancreatic cancer have different names. The three most common types are:

- **Gastrinoma**, which makes large amounts of the gastrin hormone, causing increased stomach acid and leading to ulcers.
- **Insulinoma**, which makes too much insulin, causing the body to store sugar rather than burn it for energy. Insulinoma can cause **hypoglycemia** (low blood sugar levels).
- **Glucagonoma**, which produces too much of the glucagons hormone, causing too much blood sugar, resulting in a condition called **hyperglycemia**.



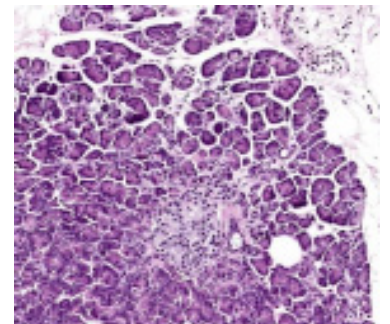
Insulinomas are composed of cube-like cells often arranged in a ribbon-like pattern.

Who is most likely to have endocrine pancreatic cancer?

Endocrine pancreatic cancer is more common among men and African-Americans. Risk factors include smoking, long-standing diabetes, chronic pancreatitis, and certain hereditary conditions including hereditary pancreatitis, multiple endocrine neoplasia type 1 syndrome, hereditary nonpolyposis colon cancer (HNPCC; Lynch syndrome), von Hippel-Lindau syndrome, ataxia-telangiectasia, and the familial atypical multiple mole melanoma syndrome. Other risk factors include a family history of pancreatic cancer, older age, and obesity. Exposure to certain pesticides, dyes, or chemicals related to gasoline is another risk.

What characterizes endocrine pancreatic cancer?

Endocrine pancreatic cancer is difficult to detect early because its symptoms are similar to other illnesses and because the pancreas is hidden behind other organs. Often, this type of cancer is not detected until it has spread. Symptoms include abdominal pain, diarrhea, chronic fatigue, fainting, or weight gain without eating too much. These symptoms, coupled with the risk factors described earlier, warrant a trip to see a doctor as soon as possible.



Normal pancreas cells.

How does the pathologist diagnose endocrine pancreatic cancer?

Your primary care physician will order **blood and urine tests** for the pathologist to examine for the amounts of endocrine hormones in the body. Your medical team also may order imaging tests such as **chest x-rays; CT, PET or MRI scans; or endoscopic ultrasound (EUS)** to view inside the body.

Laparoscopy allows physicians to view inside the body using a thin, lighted tube inserted through small incisions in the abdominal wall. **Endoscopic retrograde cholangiopancreatography (ERCP)** examines the bile ducts for narrowing or blockages sometimes associated with pancreatic cancer. **Biopsy specimens**, or tissue samples, can be gathered during these procedures.

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What kinds of questions should I ask my doctors?

Ask any question you want. There are no questions you should be reluctant to ask. Here are a few to consider:

- Please describe the type of cancer I have and what treatment options are available.
- What is the stage of my cancer?
- What are the chances for full remission?
- What treatment options do you recommend? Why do you believe these are the best treatments?
- What are the pros and cons of these treatment options?
- What are the side effects?
- Should I receive a second opinion?
- Is your medical team experienced in treating the type of cancer I have?
- Can you provide me with information about the physicians and others on the medical team?

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What else does the pathologist look for?

The pathologist reviews the biopsy specimens and the results of all tests to make a diagnosis. If cancer is found, the pathologist will determine the **stage** of the cancer. Stage 1 endocrine pancreatic cancers are small and confined to the pancreas, and stage 4 tumors have spread beyond areas near the pancreas. Stages 2 and 3 describe conditions in between these two extremes. The chance of recovery depends on the type of endocrine pancreatic cancer, how far the cancer has spread, and your overall health.

How do doctors determine what treatment will be necessary?

The pathologist consults with your primary care physician or specialist after reviewing the test results and determining the stage of the cancer. Together, using their combined experience and knowledge, they determine treatment options most appropriate for your condition. It's important to learn as much as you can about your treatment options and make the decision that's right for you.

What kinds of treatments are available for endocrine pancreatic cancer?

Endocrine pancreatic cancer can be treated with **surgery, chemotherapy, hormone therapy**, or a combination of these methods. In addition, patients may have treatment to control pain or ease emotional or practical problems.

Surgery, the most common treatment, removes the cancerous tumor, part or all of the pancreas, and sometimes other tissues or organs. Surgical options include:

- The **Whipple procedure**, in which the head of the pancreas, the gallbladder, the bile duct, and parts of the stomach and small intestine are removed
- **Total pancreatectomy**, which removes the entire pancreas, the common bile duct, the gallbladder, the spleen, and parts of the stomach and small intestine
- **Distal pancreatectomy**, which involves removing the body and tail of the pancreas and usually the spleen

If the cancer has spread and cannot be removed, other surgical procedures may be used to relieve pain and other symptoms.

If the cancer has spread outside the pancreas—or if there is a chance it has—your doctor may recommend chemotherapy. This treatment delivers drugs throughout the body, and may slow the cancer's progression and reduce pain. **Systemic chemotherapy** delivers drugs throughout the body, while **regional chemotherapy** directs the drugs into a particular organ or area of the body, such as the pancreas or abdomen.

The most commonly used chemotherapy drug is **5-fluorouracil (5-FU)**. Recent studies, however, show **gemcitabine** to be more effective. Further studies comparing the effectiveness of these two drugs in treating various types of cancers are in progress, as well as other studies combining these two drugs with each other or other chemotherapy drugs.

Hormone therapy may stop cancer cells from growing and relieve pain and other symptoms caused by excessive hormone production. **Octreotide** is the most commonly used hormone-related medication. Scientists continue to investigate the effectiveness of other treatment approaches.

Clinical trials of new treatments for endocrine pancreatic cancer may be found at www.cancer.gov/clinicaltrials. These treatments are highly experimental in nature but may be a potential option for advanced cancers. Some trials may involve biologic therapy, which uses the natural defenses of the immune system to fight cancer.

For more information, go to www.cancer.gov (National Cancer Institute) or www.cancer.org (American Cancer Society). Type the keywords **pancreatic cancer** or **islet cell cancer** or into the search box.