CONDITION

Islet cell cancer

Islet cell cancer is a pancreatic cancer that begins in hormone-making neuroendocrine cells.

WHAT IT IS
Cancer arising from hormone-making cells of the pancreas

BODY SYSTEM
Digestive and hormone systems

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

WHAT CAUSES IT
Usually unknown; sometimes linked to inherited gene changes

HOW IT IS DIAGNOSED
Imaging and biopsy with laboratory testing

MAIN TREATMENT
Surgery, hormone control, targeted or radioligand therapies

CAN IT BE CURED?
Sometimes - surgery can remove localized cancer

IS IT AN EMERGENCY?
Sometimes - severe low blood sugar needs immediate treatment

LONG-TERM OUTLOOK
Often slow-growing, but grade and spread strongly matter

Overview

Islet cell cancer begins in neuroendocrine cells in the pancreas. These cells help make hormones, which are chemical messengers that control processes such as blood sugar and digestion.

The condition is now usually called a pancreatic neuroendocrine tumor , or pancreatic NET. "Islet cell cancer" is an older name. It is different from the much more common type of pancreatic cancer, called pancreatic ductal adenocarcinoma. The two cancers behave differently and are treated differently.

Some pancreatic NETs release extra hormones. These are called functioning tumors . Their symptoms depend on the hormone produced. Other tumors do not release enough hormone to cause a clear syndrome. These are called nonfunctioning tumors and may not cause

symptoms until they grow or spread.

Two features are especially important:

Most pancreatic NETs are well-differentiated, meaning the cancer cells still resemble normal neuroendocrine cells. Poorly differentiated neuroendocrine carcinomas look more abnormal and usually grow faster. This distinction strongly affects treatment and outlook.

- Stage describes the tumor's size and whether it has spread
- Grade describes how abnormal the cells look and how quickly they are dividing

Symptoms

A small pancreatic NET may cause no symptoms. When symptoms occur, they may come from the tumor pressing on nearby organs, from cancer that has spread, or from excess hormone production.

Possible general symptoms include:

Functioning tumors can cause more specific patterns.

An insulinoma makes excess insulin, which lowers blood sugar. Symptoms can include sweating, shaking, hunger, weakness, blurred vision, confusion or fainting. Symptoms may improve after eating.

A gastrinoma makes excess gastrin, which causes the stomach to produce too much acid. This may lead to repeated stomach or small-intestine ulcers, burning abdominal pain, heartburn or diarrhea.

A glucagonoma makes excess glucagon. It may cause high blood sugar, weight loss and a distinctive red, blistering or crusted rash that moves between areas of the body.

A VIPoma releases vasoactive intestinal peptide, or VIP. It can cause large amounts of watery diarrhea, dehydration and low levels of potassium, an important body salt.

A somatostatinoma can interfere with digestion and blood sugar control. Possible effects include diabetes, greasy or difficult-to-flush stools, gallstones and weight loss.

These symptoms can have many causes other than cancer. Testing is needed to find the cause.

- Pain or discomfort in the upper abdomen or back
- Unexplained weight loss or reduced appetite
- Nausea or vomiting

- A lump or feeling of fullness in the abdomen
- Yellow skin or eyes if a tumor blocks a bile duct

Causes and risk factors

Most pancreatic NETs do not have a clearly identifiable cause. They develop after changes in a cell's DNA allow it to grow and survive when it normally would not.

A small proportion occur as part of an inherited condition. These include:

Having one of these conditions does not mean that you will develop islet cell cancer. However, it may lead to planned screening or genetic assessment.

A family history of pancreatic NETs or an inherited endocrine tumor syndrome may also increase concern. Genetic counseling can help you understand whether testing is appropriate and what a result could mean for relatives.

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

Diagnosis

Diagnosis usually starts with a medical history, examination and imaging. Blood tests may check blood sugar, liver function and general health. If symptoms suggest excess hormone production, blood or urine tests can measure specific hormones or related substances.

Imaging may include CT or MRI scans of the abdomen. An endoscopic ultrasound uses a thin, flexible tube passed through the mouth into the stomach and small intestine. An ultrasound probe on the tube creates detailed pictures of the pancreas and can guide a biopsy.

Some tumors have proteins called somatostatin receptors on their surface. Somatostatin receptor imaging uses a small amount of radioactive tracer to locate cells carrying these receptors. This can show whether the cancer has spread and whether receptor-targeted treatment may work.

A biopsy removes a small tissue sample for examination under a microscope. It can confirm that the tumor is neuroendocrine and help determine its grade. The pathology report may include a Ki-67 index, which estimates how many tumor cells are dividing.

Further scans and sometimes surgery are used to establish the stage. The care team will also determine whether the tumor is functioning, well-differentiated or poorly differentiated.

Treatment and management

Treatment is based on the tumor's location, size, grade, hormone activity and spread. Your general health and treatment preferences also matter. Care is often coordinated by specialists in pancreatic surgery, cancer medicine, hormone conditions, radiology and nuclear medicine.

Surgery offers the main chance of cure when the cancer can be removed completely. The type of operation depends on where the tumor is in the pancreas and whether nearby structures are involved. Some operations remove only the tumor or part of the pancreas. Others also remove the spleen, gallbladder, nearby lymph nodes, or part of the stomach or small intestine.

Small, slow-growing tumors may sometimes be monitored with planned scans instead of being treated immediately. This approach is only suitable in selected situations.

Somatostatin analogues are medicines that act like the natural hormone somatostatin. They can reduce symptoms caused by hormone release and may slow the growth of some well-differentiated tumors.

Other medicines can control a specific hormone syndrome. For example, treatment may help prevent low blood sugar or reduce excess stomach acid. Fluids, salts and nutritional support may be needed when diarrhea is severe.

For cancer that cannot be removed or has spread, options may include:

Poorly differentiated neuroendocrine carcinoma is generally treated differently from a well-differentiated pancreatic NET and often requires chemotherapy.

Clinical trials may provide access to treatments being studied. Palliative care can be given at any stage to manage pain, nausea, diarrhea, anxiety, nutrition problems or other effects of the condition and its treatment.

- Targeted medicines that block signals used by cancer cells
- Peptide receptor radionuclide therapy, or PRRT, which delivers radiation to somatostatin receptor-positive cells
- Chemotherapy, particularly for faster-growing or higher-grade cancers
- Liver-directed procedures when much of the cancer is in the liver
- Radiotherapy for selected tumors or areas causing symptoms

Complications

Islet cell cancer can spread to lymph nodes, the liver, bones or other organs. Spread to the liver may affect liver function or increase hormone-related symptoms.

Hormone-producing tumors can cause specific complications. Repeated low blood sugar can lead to falls, seizures or loss of consciousness. Severe diarrhea can cause dehydration, kidney problems and abnormal body-salt levels. Gastrinomas can cause bleeding or a hole in the stomach or intestine because of severe ulcers.

A tumor may block a bile duct or pancreatic duct. This can cause jaundice, infection, pancreatitis or problems digesting food.

Treatment also has possible long-term effects. Surgery may reduce insulin or digestive enzyme production. Some people develop diabetes or need pancreatic enzyme capsules to help absorb nutrients. Removing the spleen increases vulnerability to certain infections, so additional vaccinations may be advised.

Prevention

Most islet cell cancers cannot be prevented because their cause is unknown. There is no established lifestyle change that guarantees prevention.

If you have an inherited syndrome associated with pancreatic NETs, the inherited gene change itself cannot be prevented. Regular surveillance may find a tumor earlier. The timing and type of surveillance are based on the specific syndrome.

Genetic counseling may be helpful if you have a strong family history, several hormone-related tumors, or a known gene change in your family. It can explain the benefits and limits of genetic testing without assuming that cancer is present.

Living with it and what to expect

The course can differ greatly between people. A small, well-differentiated tumor may grow slowly over years. A high-grade or poorly differentiated cancer may progress more quickly. Whether the cancer can be removed and whether it has spread are also important.

After surgery, follow-up usually continues for years because pancreatic NETs can return after a long interval. Follow-up may include appointments, blood tests and scans. Hormone tests may be repeated if the original tumor produced a measurable hormone.

Advanced pancreatic NET is often managed as a long-term condition. Treatment may keep it stable for a period, followed by a change in treatment if scans, symptoms or laboratory results show progression. Stable scan results do not always mean that hormone symptoms are fully controlled, so both need attention.

Eating may be more difficult after pancreatic surgery or during treatment. A specialist dietitian can help with weight loss, diarrhea, blood sugar changes or signs that food is not being absorbed. Pancreatic enzyme treatment may help if stools become pale, oily, loose or difficult to flush.

Many people continue usual activities during monitoring or treatment, although appointments, fatigue and digestive symptoms may require adjustments. Emotional reactions can also change over time. Support from family, counseling, cancer support groups or specialist nurses may help.

When to get medical help

Arrange a routine medical appointment if you have persistent or recurring symptoms such as unexplained abdominal pain, weight loss, repeated diarrhea, frequent ulcers, a new unusual rash or episodes that may represent low blood sugar.

If you have already been diagnosed, contact your care team promptly about worsening hormone symptoms, difficulty eating, ongoing vomiting, new jaundice, increasing pain or treatment side effects. These concerns usually need timely assessment but are not always emergencies.

Seek urgent or emergency help for:

If low blood sugar is suspected and the person is awake and able to swallow safely, follow their established emergency plan. Do not give food or drink to someone who is unconscious or having a seizure.

- Confusion, fainting, a seizure or inability to wake, especially with possible low blood sugar
- Severe weakness or dizziness with ongoing watery diarrhea
- Vomiting blood, black stools or sudden severe abdominal pain
- Signs of severe dehydration, such as very little urine or marked drowsiness
- Fever with jaundice or severe abdominal pain
- New chest pain, severe breathlessness or stroke-like symptoms

Common questions

Is islet cell cancer the same as pancreatic cancer?

It is a type of pancreatic cancer, but it is not the usual pancreatic ductal cancer. Islet cell cancer starts in neuroendocrine cells and often grows and responds to treatment differently. The exact pathology is important when comparing treatments or survival information.

Are all islet cell tumors cancer?

The term "islet cell tumor" has sometimes been used for both benign and malignant growths. Modern classification recognizes that pancreatic neuroendocrine tumors can have malignant potential, even when they look slow-growing. "Islet cell cancer" specifically refers to a malignant tumor.

Can islet cell cancer be cured?

Sometimes. Surgery may cure a localized tumor that can be removed completely. Cure is less likely after the cancer has spread, but treatment may control growth, reduce hormone symptoms and maintain quality of life for a meaningful period.

How fast does islet cell cancer grow?

Many well-differentiated pancreatic NETs grow slowly, but some are more active. Grade, Ki-67 index, changes on repeat scans and whether the tumor is poorly differentiated give more useful information than the name alone.

Does islet cell cancer run in families?

Most cases are not known to be inherited. Some occur with inherited conditions such as MEN1 or von Hippel-Lindau disease. Genetic counseling may be appropriate when there is a relevant personal or family history.

Do I need a special diet?

There is no single diet that treats the cancer. Dietary needs may change if you have diarrhea, low blood sugar, diabetes, weight loss or reduced pancreatic enzyme production. Advice should be matched to the specific symptoms and treatment.

Sources

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