CONDITION

# Carcinoid apudoma

Learn what the historical term carcinoid apudoma means, including symptoms, diagnosis, treatment, complications, and follow-up for neuroendocrine tumors.

**WHAT IT IS**
Hormone-producing neuroendocrine tumor

**BODY SYSTEM**
Hormone system

**MAIN SYMPTOMS**
Abdominal pain, skin flushing, watery diarrhea, wheezing

**WHAT CAUSES IT**
Usually unknown, sometimes inherited

**HOW IT IS DIAGNOSED**
Biopsy and microscopic examination

**MAIN TREATMENT**
Surgery or tumor-directed therapy

**CAN IT BE CURED?**
Sometimes - surgery can cure localized tumors

**IS IT AN EMERGENCY?**
Sometimes - severe breathing difficulty during carcinoid crisis

**LONG-TERM OUTLOOK**
Often slow-growing, but some invade or spread

## Overview

"Carcinoid apudoma" is an older term for a tumor arising from certain hormone-producing cells. Today, doctors generally call these growths neuroendocrine tumors , or NETs. The word "carcinoid" may still be used, especially for well-differentiated NETs of the digestive tract or lungs.

"APUD" stands for "amine precursor uptake and decarboxylation." This phrase described how some neuroendocrine cells handle chemicals used to make hormones. "Apudoma" was used for tumors thought to develop from these cells. Medical understanding and tumor classification have changed, so pathology reports now describe the tumor by its location, cell appearance, grade, and stage rather than relying on the term apudoma.

Neuroendocrine cells receive signals from the nervous system and release hormones into the

blood. They are found throughout the body. NETs most often begin in the gastrointestinal tract, pancreas, or lungs, but they can arise elsewhere.

Many NETs grow more slowly than common cancers in the same organ. However, their behavior varies. Some remain limited to their original site for years, while others invade nearby tissue or spread to lymph nodes, the liver, bones, or other organs. Even a slow-growing tumor can cause problems because of its location or because it releases hormones.

Current terminology matters because treatment depends on the tumor's exact type. A modern diagnosis usually includes the organ where the tumor started, whether the cells are well differentiated, the tumor grade, and how far it has spread.

## Symptoms

Some neuroendocrine tumors cause no symptoms at first. They may be found during imaging, endoscopy, or surgery performed for another reason.

Symptoms from the tumor itself depend on its location and size. They may include:

Some NETs release active hormones or hormone-like chemicals. These are called functional tumors . The symptoms depend on which substance is released.

Carcinoid syndrome usually occurs when a NET releases serotonin and other chemicals into the circulation. It is most often associated with a gastrointestinal NET that has spread to the liver, although there are exceptions. Possible symptoms include repeated episodes of skin flushing, watery diarrhea, abdominal cramping, wheezing, and a fast heartbeat.

These symptoms have many possible causes and do not confirm that a person has a NET.

A rare, severe release of tumor chemicals is called a carcinoid crisis . Sudden major changes in blood pressure, severe breathing difficulty, confusion, fainting, or a very fast or irregular heartbeat require emergency medical care. A crisis can occur around surgery or another procedure, but it may also happen without a clear trigger.

- Abdominal pain, bloating, nausea, or changes in bowel habits
- Unexplained weight loss or loss of appetite
- Bleeding in the digestive tract, which may cause black stools or anemia
- Cough, wheezing, chest discomfort, or repeated lung infections when a tumor is in the airway
- Yellowing of the skin or eyes if a pancreatic or nearby tumor blocks a bile duct
- Pain or blockage caused by narrowing of the intestine

## Causes and risk factors

The cause of most neuroendocrine tumors is unknown. They result from changes in a cell's DNA that allow the cell to grow and survive when it normally would not. In many people, these changes develop during life rather than being inherited.

A small proportion of NETs are related to inherited conditions. These include multiple endocrine neoplasia type 1, von Hippel-Lindau disease, neurofibromatosis type 1, and tuberous sclerosis complex. The tumor types and risks differ among these conditions, and pancreatic NETs are more strongly linked to some of them than intestinal carcinoid tumors are.

Other associations depend on where the tumor begins. For example, some stomach NETs develop in people with chronic atrophic gastritis, a condition that damages the stomach lining and alters hormone signaling.

Having a possible risk factor does not mean that a NET will develop. Many people diagnosed with a NET have no known inherited syndrome or identifiable exposure. Genetic counseling may be considered when there is a strong family history, multiple endocrine tumors, a diagnosis at an unusually young age, or other features suggesting an inherited condition.

## Diagnosis

Evaluation usually begins with a medical history and physical examination. The clinician may ask about flushing, diarrhea, breathing symptoms, abdominal discomfort, weight changes, medicines, family history, and previous tumors.

Blood and urine tests can look for substances released by a functional NET. For suspected carcinoid syndrome, testing may include 5-HIAA , a breakdown product of serotonin, in urine or blood. Certain foods, medicines, and health conditions can affect results, so patients should follow the laboratory's preparation instructions.

Chromogranin A is sometimes measured, but it is not a stand-alone screening or diagnostic test. Acid-suppressing medicines, kidney disease, inflammation, and other conditions can raise the level. A normal result also does not rule out a NET.

Imaging helps locate the tumor and check whether it has spread. Depending on the situation, this may include:

A biopsy , in which a tissue sample is examined under a microscope, is usually needed for a definite diagnosis. The pathologist evaluates how closely the cells resemble normal neuroendocrine cells. A well-differentiated tumor retains more of the structure of its original cells. A poorly differentiated neuroendocrine carcinoma is a different, generally more

aggressive disease and is treated differently.

The pathology report may include the Ki-67 index and the number of dividing cells. These measurements help determine the tumor's grade, which estimates how quickly it is growing. Stage describes the tumor's size, involvement of nearby structures or lymph nodes, and spread to distant organs.

If carcinoid syndrome is present, clinicians may assess the heart because long-term exposure to tumor chemicals can damage heart valves. Testing may include blood markers and an echocardiogram, an ultrasound examination of the heart.

- CT or MRI scans of the chest, abdomen, or pelvis
- Endoscopy or endoscopic ultrasound to examine the digestive tract and nearby structures
- Somatostatin receptor PET imaging, which looks for receptors found on many well-differentiated NET cells
- Other imaging selected for a specific organ or symptom

## Treatment and management

Treatment is based on the tumor's original site, grade, stage, hormone activity, growth rate, and somatostatin receptor status. A multidisciplinary team may include medical and surgical oncologists, endocrinologists, gastroenterologists, nuclear medicine specialists, radiologists, pathologists, cardiologists, and dietitians.

Surgery may be the main treatment for a localized tumor and can sometimes be curative. The operation depends on where the NET began and whether nearby lymph nodes are involved. Surgery may also be considered in selected people with limited metastatic disease or complications such as bowel blockage.

Some small, low-risk tumors can be monitored rather than treated immediately. Active surveillance involves planned examinations, imaging, and sometimes laboratory testing. It does not mean that symptoms or tumor changes are being ignored.

Somatostatin analogs , including octreotide and lanreotide, act like a natural hormone that limits the release of several other hormones. They can reduce flushing and diarrhea from carcinoid syndrome. They may also slow the growth of some well-differentiated NETs.

If diarrhea from carcinoid syndrome continues despite somatostatin-analog treatment, telotristat ethyl may be considered. It reduces serotonin production. Diarrhea can also result from surgery, pancreatic enzyme deficiency, infection, medicines, or other conditions, so its

cause should be assessed before treatment is changed.

Peptide receptor radionuclide therapy , or PRRT, delivers radiation directly to tumor cells through a molecule that binds to somatostatin receptors. Lutetium Lu 177 dotatate is one form of PRRT. It may be used for certain advanced, receptor-positive NETs. Treatment requires monitoring because it can affect blood cells, the kidneys, or, less commonly, bone marrow function over time.

Other options for advanced disease may include targeted medicines such as everolimus, chemotherapy for selected tumor types, or treatments directed at liver metastases. Liver-directed approaches include embolization, which limits blood flow to tumors, and thermal ablation, which uses heat to destroy tumor tissue. Radiation therapy may help control specific tumors or relieve symptoms in certain locations.

People with carcinoid syndrome may notice that alcohol, large meals, stress, or particular foods trigger flushing or diarrhea. Triggers differ between individuals. A symptom diary may help identify patterns without creating unnecessary dietary restrictions. Persistent diarrhea should be discussed with the care team because it can cause dehydration, electrolyte changes, weight loss, and vitamin deficiencies.

Before surgery or an invasive procedure, the clinical team should know about any functional NET or carcinoid syndrome. Special planning may be needed to reduce the risk of a hormone-related crisis.

## Complications

A NET can press on or grow into nearby organs. Intestinal NETs may cause scarring in the tissue that supports the bowel, leading to pain, reduced blood flow, or blockage. Tumors may also spread, most commonly to lymph nodes or the liver.

Long-standing carcinoid syndrome can cause carcinoid heart disease . Fibrous tissue forms on heart valves, particularly on the right side of the heart. This can lead to leg swelling, abdominal swelling, breathlessness, fatigue, or heart failure.

Frequent diarrhea can cause dehydration, low electrolyte levels, and poor nutrition. Excess serotonin production may contribute to niacin deficiency because the body uses some of the same building blocks to produce serotonin and niacin.

Treatment can also have lasting effects. These may include changes in digestion after gastrointestinal or pancreatic surgery, reduced blood counts, kidney effects, or hormone deficiencies. The care team can monitor for these problems and provide supportive treatment.

## Follow up

Follow-up is individualized according to the tumor's site, grade, stage, treatment, and pattern of growth. Routine follow-up may include symptom review, physical examinations, CT or MRI scans, and selected hormone or tumor-marker tests. The same imaging method is often used over time so that changes can be compared accurately.

Patients should report new or steadily worsening flushing, diarrhea, abdominal pain, wheezing, weight loss, swelling, or reduced ability to exercise. These symptoms usually call for prompt contact with the treating team, but they are not automatically emergencies.

Seek urgent or emergency care for severe breathing difficulty, fainting, confusion, sudden major blood-pressure symptoms, vomiting with inability to keep fluids down, heavy gastrointestinal bleeding, or intense abdominal pain with swelling and inability to pass stool or gas.

Long-term care may also include heart monitoring for carcinoid syndrome, nutrition support, management of treatment effects, and genetic counseling when an inherited condition is possible. Keeping copies of the pathology report, operative notes, imaging reports, and treatment history can help when care involves several specialists.

## Sources

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