CONDITION

Carcinoid syndrome

Learn how carcinoid syndrome causes flushing, diarrhea, breathing symptoms, and heart problems, and how doctors diagnose, treat, and monitor it.

WHAT IT IS
Group of symptoms caused by neuroendocrine tumors

BODY SYSTEM
Hormone and digestive systems

MAIN SYMPTOMS
Skin flushing, diarrhea, breathing difficulty, fast heartbeat

WHAT CAUSES IT
Hormone-releasing neuroendocrine tumor

HOW IT IS DIAGNOSED
5-HIAA testing, imaging, tumor biopsy

MAIN TREATMENT
Hormone-blocking and tumor-directed treatment

CAN IT BE CURED?
Sometimes - localized tumor can be removed

IS IT AN EMERGENCY?
Sometimes - severe breathing difficulty or fainting

LONG-TERM OUTLOOK
Often controllable, but heart valve damage may develop

Overview

Carcinoid syndrome is a group of symptoms caused by certain neuroendocrine tumors. Neuroendocrine tumors, often shortened to NETs, grow from cells that use hormones and nerve signals to help control body functions.

Some NETs release serotonin and other active chemicals into the bloodstream. When enough of these chemicals reach the rest of the body, they can cause skin flushing, diarrhea, breathing symptoms, and changes in heart valves.

The term "carcinoid" is an older name that is still widely used. A person can have a neuroendocrine tumor without having carcinoid syndrome. The syndrome develops only when a tumor releases substances in amounts that produce symptoms.

Many NETs linked to carcinoid syndrome begin in the small intestine or another part of the digestive tract. Blood from these organs normally passes through the liver, which can break down many tumor-produced chemicals. For this reason, carcinoid syndrome from an intestinal NET often appears after the tumor has spread to the liver. However, this is not always the case. Tumors in the lungs, ovaries, or other locations may release chemicals directly into the general circulation.

Carcinoid syndrome matters because its symptoms can affect nutrition, hydration, sleep, work, and quality of life. Long-term exposure to tumor-produced chemicals may also scar certain heart valves. Treatment can control hormone-related symptoms, address the tumor, and reduce the risk or impact of complications.

Symptoms

Symptoms vary between people and may come and go. Their severity does not always show how large or widespread a tumor is.

Skin flushing is a common feature. The face, neck, or upper chest may suddenly feel warm and become red or darker than usual. An episode can last from a few minutes to longer. Some people also notice a fast heartbeat, itching, or swelling around the face.

Diarrhea may be frequent, watery, and urgent. It can occur with or without flushing. Ongoing diarrhea can lead to dehydration, loss of minerals called electrolytes, weight loss, and difficulty absorbing nutrients.

Other possible symptoms include:

Alcohol, large meals, emotional stress, strenuous activity, or certain foods may trigger symptoms in some people. Triggers are not the same for everyone, and some episodes have no clear cause.

Carcinoid heart disease may develop gradually. Early on, it may cause no obvious symptoms. As heart valves become damaged, a person may develop breathlessness, leg swelling, abdominal swelling, unusual tiredness, or reduced exercise tolerance.

A rare but serious worsening called carcinoid crisis can cause major blood pressure changes, severe flushing, breathing difficulty, confusion, or an abnormal heart rate. It may occur during surgery, anesthesia, a tumor procedure, or severe physical stress. The medical team should know about diagnosed or suspected carcinoid syndrome before any procedure.

Seek emergency care for severe trouble breathing, fainting, chest pain, confusion, bluish or gray lips, or a sudden episode with extreme weakness or signs of dangerously low blood pressure. These symptoms can have several causes and need prompt assessment.

- Abdominal pain or cramping
- Wheezing, coughing, or shortness of breath
- A fast or pounding heartbeat
- Fatigue or weakness
- Loss of appetite or unplanned weight loss
- Swelling in the legs or abdomen
- Reduced ability to exercise

Causes and risk factors

Carcinoid syndrome is caused by an underlying neuroendocrine tumor that releases serotonin or other biologically active substances. Serotonin helps regulate processes such as intestinal movement. In excess, it can contribute to diarrhea and tissue scarring. Other tumor-produced substances can widen blood vessels, causing flushing, or narrow the airways, causing wheezing.

Factors that influence whether a NET causes the syndrome include:

These are not lifestyle choices, and having carcinoid syndrome is not the result of something a person did.

Most NETs occur without a clear inherited cause. A minority are associated with inherited conditions that increase the likelihood of certain tumors, such as multiple endocrine neoplasia type 1. A clinician may discuss genetic counseling when the tumor type, age at diagnosis, personal history, or family history suggests an inherited condition.

Food, stress, and alcohol do not cause carcinoid syndrome. They may only trigger or worsen symptoms in someone whose tumor is already releasing active chemicals.

- Where the tumor began
- Whether it has reached the liver or another organ
- How much hormone the tumor releases
- The number and size of tumors
- How well the body breaks down the released substances

Diagnosis

Diagnosis usually combines a symptom history, laboratory tests, imaging, and examination of tumor tissue. No single test answers every question.

A clinician will ask about flushing, stool frequency, breathing symptoms, weight changes, and possible triggers. It can help to record when episodes happen, how long they last, and whether flushing and diarrhea occur together. Photographs of flushing may also be useful if the skin has returned to normal before an appointment.

A common test measures 5-HIAA , a substance produced when the body breaks down serotonin. It may be measured in urine collected over 24 hours or, in some settings, in blood. Certain foods, medicines, and collection problems can affect results. Patients should follow the laboratory's preparation instructions and should not stop a prescription or nonprescription medicine unless the healthcare team says it is safe.

Blood tests may assess blood counts, kidney and liver function, electrolytes, and nutrition. Chromogranin A is sometimes measured as a tumor marker, but it is not specific to NETs. Acid-suppressing medicines, kidney disease, and other conditions can raise it.

Imaging may include CT or MRI scans to locate tumors and look at the liver and other organs. Somatostatin receptor imaging, often performed with a specialized PET scan, can show whether tumor cells carry receptors that bind the hormone somatostatin. This information may help guide treatment.

A biopsy removes a small tumor sample for examination. The pathology report confirms the tumor type and describes its grade, meaning how quickly its cells appear likely to grow. The report may include a Ki-67 index, which measures how many tumor cells are actively dividing.

Because carcinoid syndrome can affect heart valves, evaluation may include an echocardiogram, an ultrasound of the heart. A blood test called NT-proBNP may help identify heart strain, but it does not replace a full heart assessment.

Treatment and management

Care is usually coordinated by clinicians experienced with neuroendocrine tumors. The team may include medical oncologists, endocrinologists, gastroenterologists, surgeons, radiologists, nuclear medicine specialists, cardiologists, dietitians, and specialist nurses.

Treatment has two connected goals: reduce hormone-related symptoms and control the tumor.

Somatostatin analogs, such as octreotide or lanreotide, act like the natural hormone somatostatin. They can reduce the release of serotonin and other substances, improving flushing and diarrhea for many patients. They may also slow the growth of some NETs. The

formulation, schedule, monitoring, and management of side effects are individualized.

If diarrhea remains poorly controlled despite somatostatin analog treatment, telotristat ethyl may be considered. It reduces serotonin production and is used together with a somatostatin analog in appropriate patients. Other causes of diarrhea-such as infection, medication effects, pancreatic enzyme problems, bile acid diarrhea, or bowel surgery-may need separate treatment.

Tumor-directed options depend on where the NET began, its grade, receptor status, extent, growth rate, and the person's overall health. Options may include surgery, treatment directed at tumors in the liver, peptide receptor radionuclide therapy, targeted medicines, or chemotherapy. Peptide receptor radionuclide therapy delivers radiation to tumor cells that carry somatostatin receptors.

Surgery may remove a localized tumor. In selected people with more extensive disease, surgery or a liver-directed procedure may reduce the amount of hormone-producing tumor even when complete removal is not possible.

Carcinoid heart disease requires joint care from NET and heart specialists. Medicines may help manage fluid buildup, but they cannot reverse a scarred valve. Some people may be evaluated for heart valve surgery.

Practical management may include:

People with prolonged diarrhea can develop niacin, or vitamin B3, deficiency because the tumor uses tryptophan to make serotonin instead of niacin. Supplements should be discussed with the healthcare team rather than started as a substitute for medical assessment.

- Replacing fluids and electrolytes lost through diarrhea
- Working with a dietitian if weight or nutrient intake is affected
- Recording symptoms and possible triggers
- Using prescribed symptom medicines as directed
- Telling every procedure and anesthesia team about carcinoid syndrome

Complications

Carcinoid heart disease is one of the most important long-term complications. Fibrous, scar-like tissue usually affects valves on the right side of the heart, especially the tricuspid and pulmonary valves. Damaged valves can eventually lead to right-sided heart failure, causing leg swelling, abdominal fluid buildup, liver congestion, and breathlessness.

Persistent diarrhea can cause dehydration, kidney strain, low potassium or other electrolyte problems, skin irritation, and poor nutrition. Weight loss may result from reduced intake, rapid movement of food through the intestine, or the effects of the tumor itself.

Niacin deficiency can cause inflamed skin, digestive symptoms, and changes in thinking or mood. Severe deficiency is called pellagra, although it is uncommon when nutritional problems are recognized and treated.

Some small-intestinal NETs cause scar tissue within the abdomen. This may pull on the intestines or nearby blood vessels and can contribute to pain, reduced blood flow, or bowel blockage.

Carcinoid crisis is an uncommon emergency involving a sudden, intense release of tumor chemicals. Planning before surgery or tumor procedures is important because the medical team may use preventive and emergency medicines based on the individual situation.

Follow up

Routine follow-up monitors both the tumor and the effects of hormone release. The schedule depends on the tumor's location, grade, treatment, disease extent, symptoms, and previous test results.

Visits may include a symptom review, physical examination, blood or urine testing, and CT, MRI, or receptor-based imaging. Heart monitoring may include NT-proBNP testing and repeat echocardiograms when clinically appropriate. There is no single follow-up schedule that fits everyone.

Contact the care team between routine visits for a clear increase in diarrhea or flushing, new wheezing, ongoing vomiting, reduced urine output, unplanned weight loss, new leg swelling, increasing abdominal size, or declining ability to exercise. These changes may require earlier testing or a treatment adjustment, but they are not always an emergency.

Seek urgent or emergency care for fainting, severe breathing difficulty, chest pain, confusion, signs of severe dehydration, rapidly worsening swelling with breathlessness, or a sudden severe episode involving major blood pressure or heart-rate changes.

Sources

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