About Gastrointestinal Carcinoid Tumors

Overview and Types

If you have been diagnosed with a gastrointestinal carcinoid 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 Gastrointestinal Carcinoid Tumor?

Research and Statistics

See the latest estimates for new cases of gastrointestinal carcinoid tumor in the US and what research is currently being done.

- Key Statistics About Gastrointestinal Carcinoid Tumors
- What's New in Gastrointestinal Carcinoid Tumor Research?

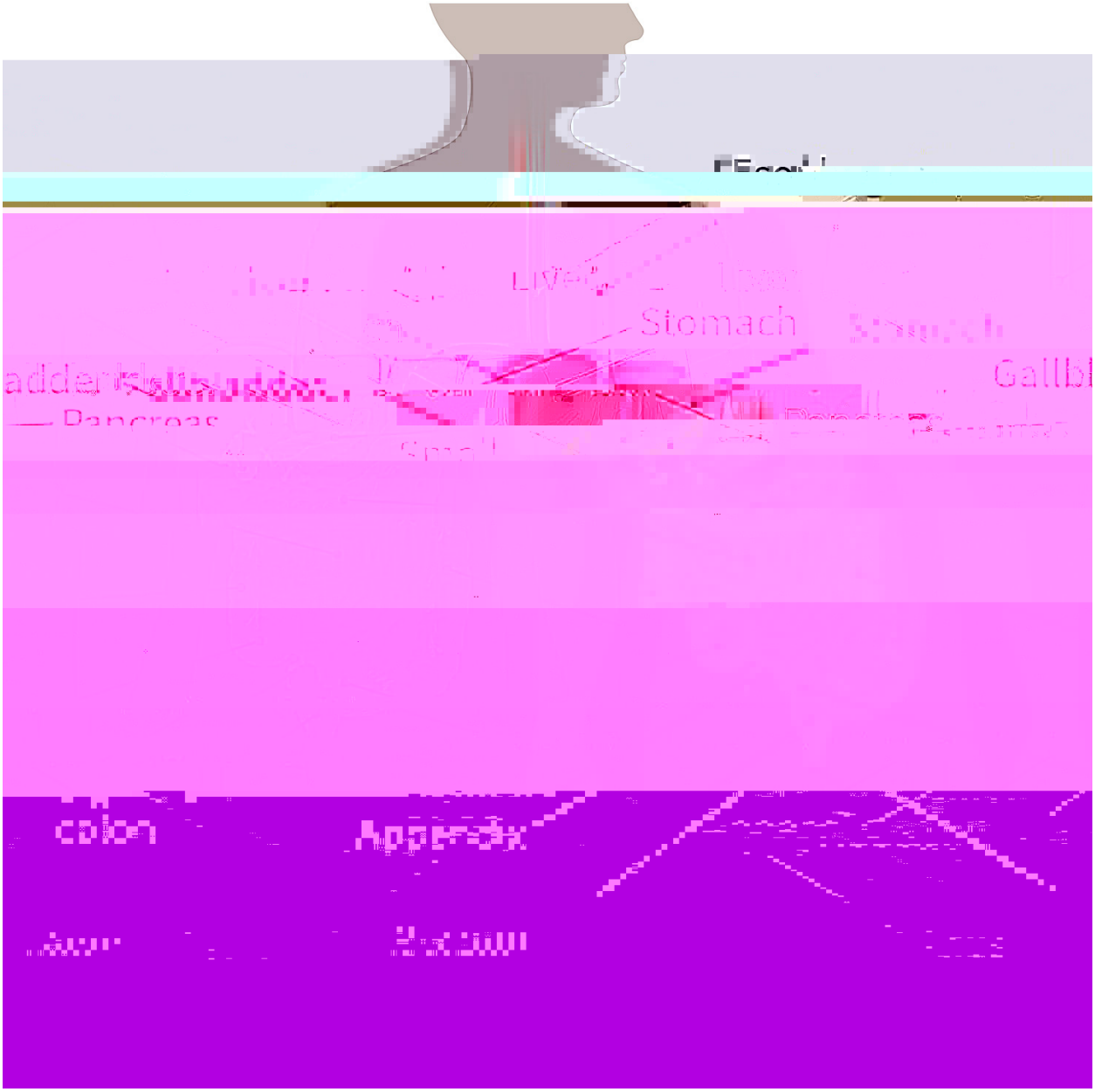
What Is a Gastrointestinal Carcinoid Tumor?

Gastrointestinal carcinoid tumors are a type of cancer that forms in the lining of the gastrointestinal (GI) tract. Cancer starts when cells begin to grow out of control. To learn more about what cancer is and how it can grow and spread, see What Is Cancer?[1]

To understand gastrointestinal carcinoid tumors, it helps to know about the gastrointestinal system, as well as the neuroendocrine system.

The gastrointestinal system

The gastrointestinal (GI) system, also known as the digestive system, processes food for energy and rids the body of solid waste. After food is chewed and swallowed, it enters the esophagus. This tube carries food through the neck and chest to the stomach. The esophagus joins the stomachjust beneath the diaphragm (the breathing muscle under the lungs). The stomach is a sac that holds food and begins the digestive process by secreting gastric juice. The food and gastric juices are mixed into a thick fluid, which then empties into the small intestine.



The small intestine keeps breaking down food and absorbs most of the nutrients. It is the longest section of the gastrointestinal (GI) tract, measuring more than 20 feet (6 meters). The small intestine then joins the colon. This is a wider, muscular tube about 5 feet (1.5 meters) long. The appendix is near the junction of small intestine and colon. The colon absorbs water, minerals, and nutrients from food and serves as a storage place for waste. The waste that is left after this process goes into the rectum. From there it leaves the body through the anus as stool (feces).

The neuroendocrine system

The neuroendocrine system has cells that act like nerve cells in certain ways and like hormone-makingendocrine cells in others. These cells don't form an actual organ like the adrenal or thyroid glands. Instead, they are scattered throughout organs like the esophagus, stomach, pancreas, intestines, appendix, and lungs. The digestive system has more neuroendocrine cells than any other part of the body. This might be why carcinoid tumors most often start there.

Neuroendocrine cells help control the release of digestive juices and how fast food moves in the GI tract. They may also help control the growth of other types of digestive system cells. Like most cells in the body, GI tract neuroendocrine cells sometimes go through certain changes that cause them to grow too much and form cancers. These cancers as a group are called neuroendocrine tumors.

Neuroendocrine (carcinoid) tumors

Neuroendocrine tumors (NETs) are mostly slow growing, but some are not and can possibly spread to other parts of the body

They are classified by tumor grade which describes how quickly the cancer is likely to grow and spread..

- **Grade 1** (low grade) NETs have cells that look more like normal cells and are not multiplying quickly.
- **Grade 2** (intermediate grade) NETs have features in between those of low- and high-grade tumors.
- **Grade 3** (high grade) NETs have cells that look very abnormal and are multiplying faster.

Cancers that are grade 1 or grade 2 are called **GI 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 **GI neuroendocrine carcinomas (NECs).** These cancers tend to grow and spread quickly and can spread to other parts of the body.

The term "carcinoid" is often used to describe grade 1 and grade 2 GI NETs. The term carcinoid will be used here unless referring to NECs specifically. Carcinoid tumors that start in the lungs are not covered here, but you can find more information in Lung Carcinoid Tumor[2].

Other gastrointestinal tumors

Schneider DF, Mazeh H, Lubner SJ, Jaume JC, Chen H. Cancer of the Endocrine System. In: Niederhuber JE, Armitage JO, Doroshow JH, Kastan MB, Tepper JE, eds. Abeloff's Clinical Oncology. 5th ed. Philadelphia, Pa: Elsevier; 2014:1112-1142.

See all references for Gastrointestinal Carcinoid Tumor (www.cancer.org/cancer/gastrointestinal-carcinoid-tumor/references.html)

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Key Statistics About Gastrointestinal Carcinoid Tumors

Although the exact number isn't known, about 8,000 carcinoid tumors and cancers that start in the gastrointestinal tract (the stomach, intestine, appendix, colon, or rectum) are diagnosed each year in the United States. These tumors can also start in the lungs[1] and the pancreas[2], and a small number develop in other organs.

The number of carcinoid tumors diagnosed has been increasing for many years. The reason for this is unknown. Some think it may be the result of more medical tests being done to look for something else and finding carcinoid tumors. Since many carcinoids never cause any symptoms, there are probably many people with carcinoid tumors that are never diagnosed. These tumors might only be seen during an autopsy when a person dies of something else, or when someone has surgery or imaging tests for an unrelated condition.

The most common locations of gastrointestinal (GI) carcinoid tumors are the small intestine and the rectum. Other common sites include , the colon (large intestine), the appendix, and the stomach.

The average age of people diagnosed with GI carcinoid tumors is early 60s . Carcinoid tumors are more common in African Americans than in White people, and are slightly more common in women than men .

Hyperlinks

1. www.cancer.org/cancer/lung-carcinoid-tumor.html

Research is always going on in the field of gastrointestinal (GI) carcinoid tumors. Scientists keep looking for the causes of, and new ways to prevent, diagnose, and treat these tumors.

Genetics

Researchers hope finding the causes of GI carcinoid tumors can be used to help prevent or treat them in the future. For example, the *IPMK* gene (the gene associated with a small intestinal neuroendocrine tumor that runs in families) has now been found in people with GI carcinoid tumors who might not have a family history of small intestinal neuroendocrine tumors. Other genetic changes that seem to make tumors more aggressive are now being explored as well.

Diagnosis and staging

Because the outlook and treatment of GI carcinoid tumors and other cancers of the digestive tract are very different, accurate diagnosis is important. Tests that can detect specific substances found in the cells of carcinoid tumors are being developed. Most of these tests treat tissue samples with special, man-made antibodies. The antibodies are designed to recognize specific parts of proteins that appear only in certain types of tumors.

In the past few years, a new imaging test[1] called a Gallium-68 PET/CT Dotatate scan[2] has been approved to look for GI carcinoid tumors in the body. This scan appears to find carcinoid tumors better than the Octreoscan. Researchers are now looking at other imaging methods to see if they can detect carcinoid tumors early.

Treatment

Surgery is the main treatment for carcinoid tumors that can be removed. Sometimes, removing the bulk of the carcinoid can also reduce the severity of the carcinoid syndrome. But better approaches are needed when surgery can't remove all of the tumors. Chemotherapy has had limited success. New chemotherapy drugs and combinations of drugs are being studied, but true advances are likely to come from other approaches.

Targeted therapy

Several newer types of drugs, known as targeted therapies, are now being studied for use against neuroendocrine tumors. Targeted therapy are drugs or other substances

that identify and attack cancer cells while doing little damage to normal cells. These therapies attack the parts of cancer cells that make them different from normal, healthy cells. Each type of targeted therapy works differently, but all can change the way a cancer cell grows, divides, repairs itself, or interacts with other cells.

Bevacizumab (Avastin®) is a type of targeted drug that attacks a tumor's blood supply. It is already being used against some types of cancer and is being studied for carcinoid tumors.

Other targeted therapies block the molecules that increase the growth of cancer cells. Some of these (such as **erlotinib**, **temsirolimus**, and **sorafenib**) are used in other types of cancer and are now being tested for use against carcinoids.

Netazepide is new drug that blocks the hormone gastrin. In early studies of patients who have carcinoid tumors of the stomach and high gastrin levels, this drug helped the tumors shrink. More studies are planned.

Immunotherapy drugs are showing promise in many cancer types. A new immune checkpoint inhibitor, **RRx-001**, is a next generation immunotherapy drug that affects many parts of the immune system to kill cancer cells and is being studied in people with carcinoid tumors.

The FDA has approved a newer, more improved radionuclide treatment for patients with advanced, worsening GI carcinoid tumors that have the somatostatin protein. This treatment takes **Lu-177-Dotatate** (a radioactive substance) which attaches to carcinoid tumors with the somatostatin protein and then releases small doses of radiation to kill the cancer cells.

Hyperlinks

1. www.cancer.org/cancer/gastrointestinal-carcinoid-tumor/detection-diagnosis-staging/how-diagnosed.html
2. www.cancer.org/cancer/gastrointestinal-carcinoid-tumor/detection-diagnosis-staging/how-diagnosed.html
3. www.fda.gov/newsevents/newsroom/pressannouncements/ucm594043.htm?elqtrackid=3866a921a66f4a658a229f679f663d96

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Strosberg J, ElHaddad G, Wolin E, et al: Phase 3 trial of 177Lu-Dotatate for midgut neuroendocrine tumors. *N Engl J Med.* 2017; 376:125-135.

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See all references for Gastrointestinal Carcinoid Tumor (www.cancer.org/cancer/gastrointestinal-carcinoid-tumor/references.html)

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