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.

- What Are the Key Statistics About Gastrointestinal Carcinoid Tumors?
- What’s New in Gastrointestinal Carcinoid Tumor Research and Treatment?

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. Cells in nearly any part of the body can become cancer. To learn more about what cancer is and how it can grow and spread, see What Is Cancer?

To understand gastrointestinal carcinoid tumors, it helps to know about the gastrointestinal system, as well as the diffuse neuroendocrine 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 stomach just beneath the diaphragm (the breathing muscle under the lungs). The stomach is a sac-like organ 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 continues breaking down food and absorbs most of the nutrients. It is the longest section of the gastrointestinal (GI) tract, measuring more than 20 feet. The small intestine then joins the colon. This is a wider, muscular tube about 5 feet long. The appendix is found near the junction of small intestine and colon. The colon absorbs water and mineral nutrients from food and serves as a storage place for waste. The
waste left after this process goes into the rectum. From there it passes out of the body through the anus as stool (feces).

**The diffuse neuroendocrine system**

Carcinoid tumors start from cells of the diffuse neuroendocrine system. This system consists of cells that are like nerve cells in certain ways and like hormone-making endocrine cells in other ways. These cells don’t form an actual organ like the adrenal or thyroid glands. Instead, they are scattered throughout other organs like the esophagus, stomach, pancreas, intestines, and lungs. The digestive system is large and has more neuroendocrine cells than any other part of the body. This might be why carcinoid tumors most often start in the digestive system.

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.

**Neuroendocrine tumors**

Like most cells in the body, GI tract neuroendocrine cells sometimes go through certain changes that cause them to grow too much and form tumors. These tumors are known as **neuroendocrine tumors** (NETs) and **neuroendocrine cancers**. In the past, most abnormal growths of neuroendocrine cells were called *carcinoids*. But in 2000, the World Health Organization (WHO) reclassified carcinoids as neuroendocrine tumors and neuroendocrine cancers.

Neuroendocrine tumors are growths that look benign but can possibly spread to other parts of the body. Neuroendocrine *cancers* are abnormal growths of neuroendocrine cells which can spread to other parts of the body.

Neuroendocrine cancers (also known as *neuroendocrine carcinomas*) are divided into groups based on the way the cells look under a microscope:

- **Well-differentiated** neuroendocrine cancers have cells that do not look very abnormal and are not multiplying rapidly. These tumors tend to grow and spread slowly. Well-differentiated neuroendocrine cancers can look identical to benign neuroendocrine tumors when seen under the microscope. Sometimes the only way to know for certain that a mass is a neuroendocrine cancer (and not a benign tumor) is when it spreads to other organs or tissues.
Poorly-differentiated cancers have cells that look very abnormal and are multiplying more rapidly. Poorly-differentiated cancers tend to grow and spread quickly.

- Moderately-differentiated cancers have features in between those of well-differentiated and poorly-differentiated cancers.

Neuroendocrine tumors of the pancreas

Neuroendocrine tumors in the pancreas are known as islet cell carcinomas or pancreatic neuroendocrine tumors. Islet cell tumors are not the same as carcinoid tumors. They have a different prognosis (course of disease and outlook) and respond differently to treatment. Neuroendocrine tumors of the pancreas are not covered here, but you can find more information in Pancreatic Cancer.

Carcinoid tumors

Carcinoid is the term used to describe well to moderately-differentiated neuroendocrine tumors in the stomach, intestine, appendix, rectum, and lung. Carcinoid tumors that start in the lungs are not covered here, but you can find more information in Lung Carcinoid Tumor.

Neuroendocrine tumors and cancers act like the cells they come from, often releasing certain hormone-like substances into the bloodstream. In most people with carcinoid tumors, the levels of these hormones are not high enough to cause symptoms. But in about 1 person out of 10 with a carcinoid tumor, the tumor spreads and grows enough to release high amounts of these hormones. This can cause a set of symptoms known as the carcinoid syndrome. Some symptoms of the carcinoid syndrome include flushing (redness of the skin with a feeling of warmth), wheezing, diarrhea, and a fast heartbeat.

Other gastrointestinal tumors

Carcinoids and other neuroendocrine tumors are different from the more common tumors of the GI tract. Most GI tract tumors start from the glandular cells that produce mucus and make up the inner lining of the digestive system. When these tumors are benign, they are called adenomas. When these cells develop into cancer, the tumors are known as adenocarcinomas.

These tumors differ quite a lot from carcinoid tumors in their symptoms, their outlook, and their treatment. For these reasons, it is important to know what type of tumor you have: a neuroendocrine tumor, a neuroendocrine cancer, an adenoma, an adenocarcinoma, or some other type of tumor. Information about adenocarcinomas of
the GI tract can be found in Esophagus Cancer, Stomach Cancer, Small Intestine Cancer, and Colorectal Cancer.

In general, neuroendocrine tumors and neuroendocrine cancers grow slower than other cancers in the GI tract. But how they grow and whether or not they spread to other areas varies widely. This depends to some extent on which part of the body the tumor starts in.

- References
See all references for Gastrointestinal Carcinoid Tumor

What Are the Key Statistics About Gastrointestinal Carcinoid Tumors?

Although the exact number isn’t known, about 8,000 neuroendocrine 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 (see Lung Carcinoid Tumor for more information) and the pancreas, 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 a byproduct of doing more medical tests 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 found. 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 location of gastrointestinal carcinoid tumors is the small intestine, often in the section near the appendix (called the ileum). Other common sites include the rectum, the colon (large intestine), the appendix, and the stomach.
The average age of people diagnosed with carcinoid tumors is in the early 60s. Carcinoid tumors are more common in African Americans than in whites, and are slightly more common in women than men.

- References
See all references for Gastrointestinal Carcinoid Tumor

What’s New in Gastrointestinal Carcinoid Tumor Research and Treatment?

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

Genetics

Researchers are looking for the causes of GI carcinoid tumors in the hope that this knowledge can be used to help prevent or treat them in the future. A great deal of progress has been made in recent years. For example, scientists have found that changes in the \textit{MEN1} gene (the gene that causes multiple endocrine neoplasia, type 1) are seen in many people with GI carcinoids. 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. Researchers have made great progress in developing tests that can detect specific substances found in
the cells of carcinoid tumors. Most of these tests treat tissue samples with special antibodies made in the lab. The antibodies are designed to recognize specific substances that appear only in certain types of tumors.

OctreoScan® is an imaging test commonly used to look for GI carcinoid tumors in the body. Researchers are now looking at other radionuclide methods to see if they can detect carcinoid tumors early.

**Treatment**

Surgery is the main treatment option for carcinoid tumors when possible. 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 uses drugs or other substances to 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 alter the way a cancer cell grows, divides, repairs itself, or interacts with other cells.

Everolimus (Afinitor®) is a targeted drug that works by blocking a cell protein known as mTOR, which normally helps cells grow and divide. It is approved by the FDA to treat advanced pancreatic neuroendocrine tumors. It is also being studied to see if it can help patients with GI carcinoids. In one study, adding everolimus to octreotide (Sandostatin) was better than octreotide alone in halting tumor growth.

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 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.
Radionuclide scans, such as the I-131 MIBG scan, can help find neuroendocrine tumors because they use substances that are attracted to neuroendocrine cells. These substances are attached to slightly radioactive elements so that they can be detected with special cameras. Using higher doses of I-131 MIBG delivers more radiation to the tumor cells and is used in Europe to treat neuroendocrine tumors and cancers. But doctors are now studying the use of a form of octreotide (or a similar drug called edotreotide) that has been attached to a radioactive form of the element yttrium called 90-Y. When injected into the body, the drug homes in on the tumor cells, allowing the radiation from the 90-Y to kill them. So far, results have been promising, but this approach is still only available in the United States as a part of a clinical trial.

- References
  See all references for Gastrointestinal Carcinoid Tumor

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