About Gastrointestinal Stromal Tumor

Overview and Types

If you have been diagnosed with a gastrointestinal stromal tumor or are worried about it, you likely have a lot of questions. Learning some basics is a good place to start.

- What Are Gastrointestinal Stromal Tumors?

Research and Statistics

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

- Key Statistics for Gastrointestinal Stromal Tumors
- What's New in Gastrointestinal Stromal Tumor Research?

What Are Gastrointestinal Stromal Tumors?

Cancer starts when cells in the body begin to grow out of control. Cells in nearly any part of the body can become cancer, and can spread to other areas of the body. To learn more about how cancers start and spread, see What Is Cancer?

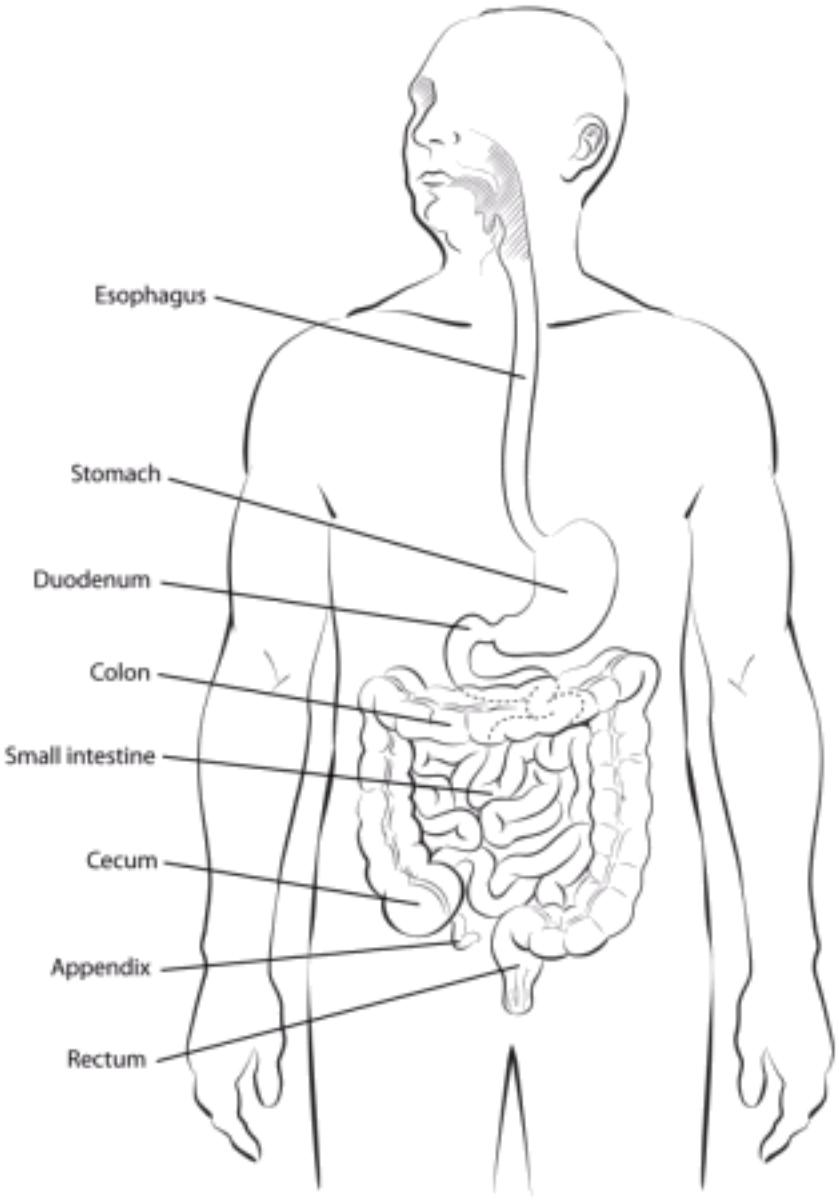
Gastrointestinal stromal tumors (GISTs) start in special cells in the wall of the gastrointestinal (GI) tract, also known as the *digestive tract*. To understand GISTs, it helps to know something about the structure and function of the GI tract.

How the gastrointestinal tract works

The gastrointestinal (GI) tract processes food for energy and rids the body of solid waste. After food is chewed and swallowed, it enters the esophagus, a tube that carries food through the neck and chest to the stomach. The esophagus joins the stomach just beneath the diaphragm (the thin band of muscle below the lungs).

The stomach is a sac-like organ that helps the digestive process by mixing the food with gastric juices. The food and gastric juices are then emptied into the small intestine. The small intestine, which is about 20 feet long, continues breaking down the food and absorbs most of the nutrients into the bloodstream.

The small intestine joins the large intestine, the first part of which is the colon, a muscular tube about 5 feet long. The colon absorbs water and mineral nutrients from the remaining food matter. The waste left after this process goes into the rectum as stool (feces), where it is stored until it passes out of the body through the anus.



Gastrointestinal stromal tumors

Gastrointestinal stromal tumors (GISTs) are uncommon tumors of the GI tract. These tumors start in very early forms of special cells in the wall of the GI tract called the *interstitial cells of Cajal* (ICCs). ICCs are cells of the *autonomic nervous system*, the

part of the nervous system that regulates body processes such as digesting food. ICCs are sometimes called the “pacemakers” of the GI tract because they signal the muscles in the GI tract to contract to move food and liquid along.

More than half of GISTs start in the stomach. Most of the others start in the small intestine, but GISTs can start anywhere along the GI tract. A small number of GISTs start outside the GI tract in nearby areas such as the omentum (an apron-like layer of fatty tissue that hangs over the organs in the abdomen) or the peritoneum (the layer of tissue that lines the organs and walls of the abdomen).

Some GISTs seem to be much more likely to grow into other areas or spread to other parts of the body than others. Doctors look at certain factors to help tell whether a GIST is likely to grow and spread quickly, such as how large the tumor is, where it's located in the GI tract, and how fast the tumor cells are dividing (its mitotic rate, described in Tests for Gastrointestinal Stromal Tumors).

Other GI tract cancers

It's important to understand that GISTs are not the same as other, more common types of GI tract cancers that develop from other types of cells.

Cancers can occur anywhere in the GI tract from the esophagus to the anus. Most GI tract cancers, including those of the esophagus, stomach, colon, and rectum, start in the gland cells that line almost all of the GI tract. The cancers that develop in these cells are called **adenocarcinomas**.

Cancers can also start in squamous cells, which are flat cells that line some parts of the GI tract, like the upper part of the esophagus and the end of the anus. Cancers starting in these cells are called **squamous cell carcinomas**.

The GI tract also has neuroendocrine cells. These cells have some features in common with nerve cells but other features in common with hormone-producing (endocrine) cells. Cancers that develop from these cells are called **neuroendocrine tumors (NETs)**. These cancers are rare in the GI tract. Carcinoid tumors are an example of a neuroendocrine tumor found in the GI tract.

Other rare types of cancer in the GI tract include different types of **soft tissue sarcomas**, such as:

- Leiomyosarcomas: cancers of smooth muscle cells
- Angiosarcomas: cancers of blood vessel cells

- Malignant peripheral nerve sheath tumors (MPNSTs): cancers of cells that support and protect nerves

GISTs are different from these other GI tract cancers. They start in different types of cells, need different types of treatment, and have a different prognosis (outlook). This is why doctors need to figure out whether a person with a tumor in the GI tract has a GIST, some other type of cancer, or a non-cancerous condition.

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Key Statistics for Gastrointestinal Stromal Tumors

Gastrointestinal stromal tumors (GISTs) are not common, and the exact number of people diagnosed with these tumors each year is not known. Until the late 1990s, not

much was known about these tumors (and doctors didn't have good ways of identifying them with lab tests), so many of them ended up being classified as other kinds of GI cancers.

Current estimates for the total number of GIST cases each year in the United States range from about 4,000 to about 6,000.

These tumors can start anywhere in the GI tract, but they occur most often in the stomach (about 60%) or the small intestine (about 35%). Most of the rest are found in the esophagus, colon, and rectum. A small number develop in the abdomen outside the GI tract.

GISTs are most commonly found in people in their 60s. These tumors are rare in people younger than 40, but they can occur in people at any age.

Survival statistics for GIST are discussed in Survival Rates for Gastrointestinal Stromal Tumors.

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What's New in Gastrointestinal Stromal Tumor Research?

Important research on gastrointestinal stromal tumors (GISTs) is going on in many

university hospitals, medical centers, and other institutions around the world. Scientists are learning more about what causes the disease and how best to treat it. There has been a great deal of progress in recent years, especially in treating GISTs.

Targeted therapy drugs

As researchers have come to understand the genetic changes that cause these tumors, they've been able to use newer treatments to target these changes.

Doctors know targeted treatments like imatinib (Gleevec), sunitinib (Sutent), and regorafenib (Stivarga) often work, but they still aren't sure exactly how and when to give them to make them most effective. For example, should these types of drugs be given after surgery to all patients, even those with very small tumors? How long should drug treatment be continued? These and other questions are now being studied in clinical trials.

Other drugs that target the KIT or PDGFRA proteins are also being studied for use against GISTs. Some of these, such as sorafenib (Nexavar), nilotinib (Tasigna), dasatinib (Sprycel), pazopanib (Votrient), and ponatinib (Iclusig), have helped some patients in early studies. Other, newer drugs that target these proteins, such as crenolanib and BLU-285, are also being studied.

Many other drugs that target different proteins involved in tumor cell growth are now being tested as well.

Immunotherapy

Immunotherapy is the use of medicines to boost the body's own immune response to help fight the cancer.

For example, immune system cells normally have proteins that act as checkpoints to keep them from attacking other healthy cells in the body. Cancer cells sometimes take advantage of these checkpoints to avoid being attacked by the immune system.

Newer drugs called **immune checkpoint inhibitors** work by blocking these checkpoint proteins, which can boost the immune response against cancer cells in the body. These drugs have been shown to be helpful against many types of cancer in recent years. Some of these drugs, such as such as nivolumab (Opdivo) and ipilimumab (Yervoy), are now being studied for use against GISTs.

Other types of immunotherapy are now being explored as well.

People with GISTs who are no longer responding to standard treatments may want to ask their doctor about clinical trials of these newer types of treatments.

- References

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