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 then can spread to other areas of the body. To learn more about cancer and how it starts and spreads, see What Is Cancer?1

Gastrointestinal stromal tumors (GISTs) are uncommon cancers that 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 (GI) tract works**

The GI tract processes food for energy and rids the body of solid waste. After food is chewed and swallowed, it goes through the esophagus, a tube that carries food down the throat 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 (stool) goes into the rectum, where it is stored until it passes out of the body through the anus.
Gastrointestinal stromal tumors

Gastrointestinal stromal tumors (GISTs) start in very early forms of special cells in the wall of the GI tract called the **interstitial cells of Cajal** (ICCs). 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 thin lining over the organs and walls inside the abdomen).

Some GISTs seem to be much more likely than others to grow into other areas or spread to other parts of the body. Doctors look at certain factors to help tell whether a
GIST is likely to grow and spread quickly, such as:

- The size of the tumor
- Where it’s located in the GI tract
- How fast the tumor cells are dividing (its **mitotic rate**, described in Tests for Gastrointestinal Stromal Tumors\(^2\))

**Other GI tract cancers**

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 cancers that start in the GI tract, including most esophagus cancers, stomach cancers, and colon and rectum cancers, 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 also have 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\(^3\)** 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\(^4\)**, 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 types of 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.
Hyperlinks


References


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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 cancers.

Current estimates for the total number of GIST cases diagnosed 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 over the age of 50. These tumors are rare in people younger than 40, but they can develop in people of any age.

Survival statistics for people with GIST tumors are discussed in Survival Rates for Gastrointestinal Stromal Tumors¹.

Hyperlinks


References


What’s New in Gastrointestinal Stromal Tumor Research?

Research on gastrointestinal stromal tumors (GISTs) is being done in many medical centers and other institutions around the world. Scientists are learning more about what causes these tumors and how best to treat them. There has been a great deal of progress in recent years, especially in treating GISTs.

Targeted therapy drugs

As researchers have come to understand more about the genetic changes\(^1\) that cause these tumors, they’ve been able to use newer targeted treatments (sometimes called precision medicines) to attack cancer cells with these changes. For example, mutations in the \(KIT\) or \(PDGFRA\) genes are present in the cells of most GISTs.

**Targeted therapy drugs**\(^2\) like imatinib (Gleevec), sunitinib (Sutent), and regorafenib (Stivarga) can affect cells with these gene changes and are often helpful in treating GISTs, but they tend to stop working over time. Doctors 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\(^3\) 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\(^4\).
Other drugs that target the KIT or PDGFRA proteins are also being studied for use against GISTs. Some of these drugs have been shown to help some patients in early studies:

- Sorafenib (Nexavar)
- Nilotinib (Tasigna)
- Dasatinib (Sprycel)
- Pazopanib (Votrient)
- Ponatinib (Iclusig)

Other, newer drugs that target these proteins are also being studied, such as:

- Crenolanib
- (Avipritinib) BLU-285
- Ripretinib (DCC-2618)
- Binimetinib (Mektovi)

Drugs that target different proteins involved in GIST 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.

**Immune checkpoint inhibitors**

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.

**Monoclonal antibodies**
Monoclonal antibodies are man-made versions of immune system proteins created to attack specific parts of cancer cells. An example now being studied for use against GISTs is XmAb18087. This antibody attaches to cells that have the somatostatin receptor 2 (SSTR2) protein, and brings them into contact with immune cells called T cells. The goal is to activate the person’s immune system to attack the cancer cells.

Other types of immunotherapy are now being explored as well.

People with GISTs that are no longer responding to standard treatments may want to ask their doctor about clinical trials\textsuperscript{5} studying these newer types of treatments.

**Hyperlinks**


**References**


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