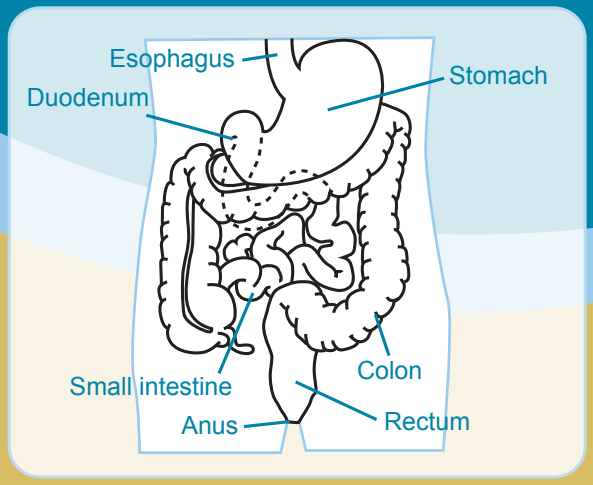
What is GIST?

Gastrointestinal stromal tumor (GIST) is a type of sarcoma. GISTs originate from the connective tissue wall of the digestive tract or nearby membranes. GISTs often grow outward, pushing against other organs. GISTs behave differently than common gastric or intestinal cancers that arise inside the digestive tract from its lining.



GIST can occur anywhere along the length of the digestive tract: stomach (55%), small intestine (30%), esophagus (5%), rectum (5%), and colon (2%). Rare locations include supporting structures of the abdominal organs, liver, pancreas, ovaries, uterus, and prostate.The liver is the most common site of GIST metastasis.

Who gets GIST?

GISTs are diagnosed at a yearly rate of 15 cases per million people. Most GIST patients are over age 50, but some are young adults or rarely children.
Symptoms may be vague (abdominal fullness, digestive discomfort, palpable mass) or alarming (bleeding, vomiting, diarrhea, anemia causing fatigue).

What causes GIST?

Mutations in the genes for growth factor receptors (KIT or PDGFRA) cause uncontrolled tumor growth in adult forms of GIST.Genetic abnormalities in pediatric GISTs are still unknown. Most GISTs are sporadic, with no preventable risk factors. Neurofibromatosis 1 and rare familial syndromes increase the risk for developing GISTs.

How is GIST diagnosed?

A pathologist examines biopsy samples or the surgically removed tumor. Most GISTs test positive for KIT protein (CD117). Mutation testing is advised for CD117-negative GISTs and aids treatment decisions for risky GISTs.
The pathologist evaluates cell division rate and tumor size to estimate the risk of recurrence or metastasis.

How is GIST treated?

Surgery, the mainstay of treatment, is potentially curative for primary tumors. Surgeons specializing in sarcomas are most skilled at removing GIST.
Radio frequency ablation and hepatic artery embolization are used for selected liver tumors.

Molecularly targeted therapies are effective against most GISTs. Imatinib mesylate (Gleevec) shrinks inoperable GISTs for durable disease control. Sometimes drugs are used before surgery to reduce the scope of surgery. Sunitinib malate (Sutent) is available for imatinib-resistant tumors and patients who cannot tolerate imatinib. These drugs have fewer side effects than traditional chemotherapies, which are ineffective against GIST. Because targeted therapies are not curative, the drugs are taken indefinitely.