

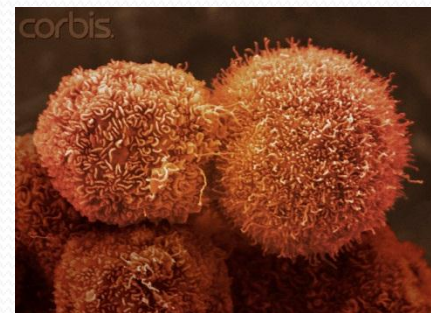
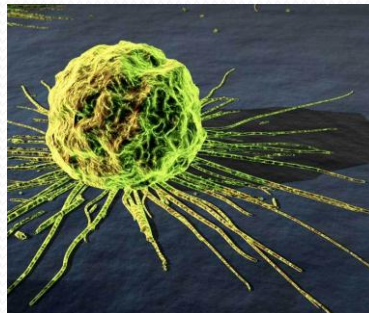
An Introduction to Gastrointestinal Stromal Tumors (GIST)



By The Life Raft Group

What is cancer?

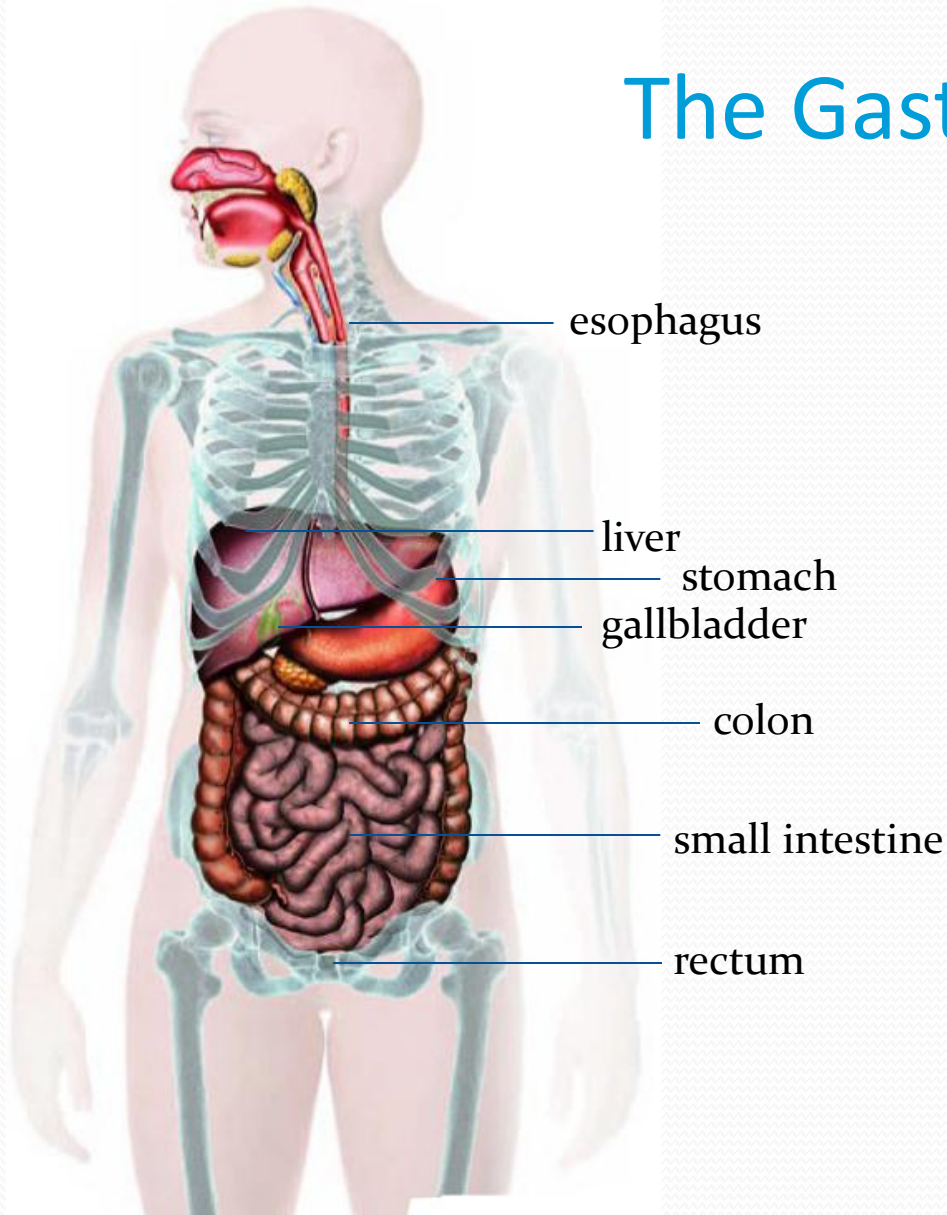
- Cancer consists of over 100 diseases.
- Cancer begins when there is an **abnormal growth** of cells that cannot be controlled.
- Because different types of cancers behave differently, it is important to get treated according to the specific **diagnosis**.



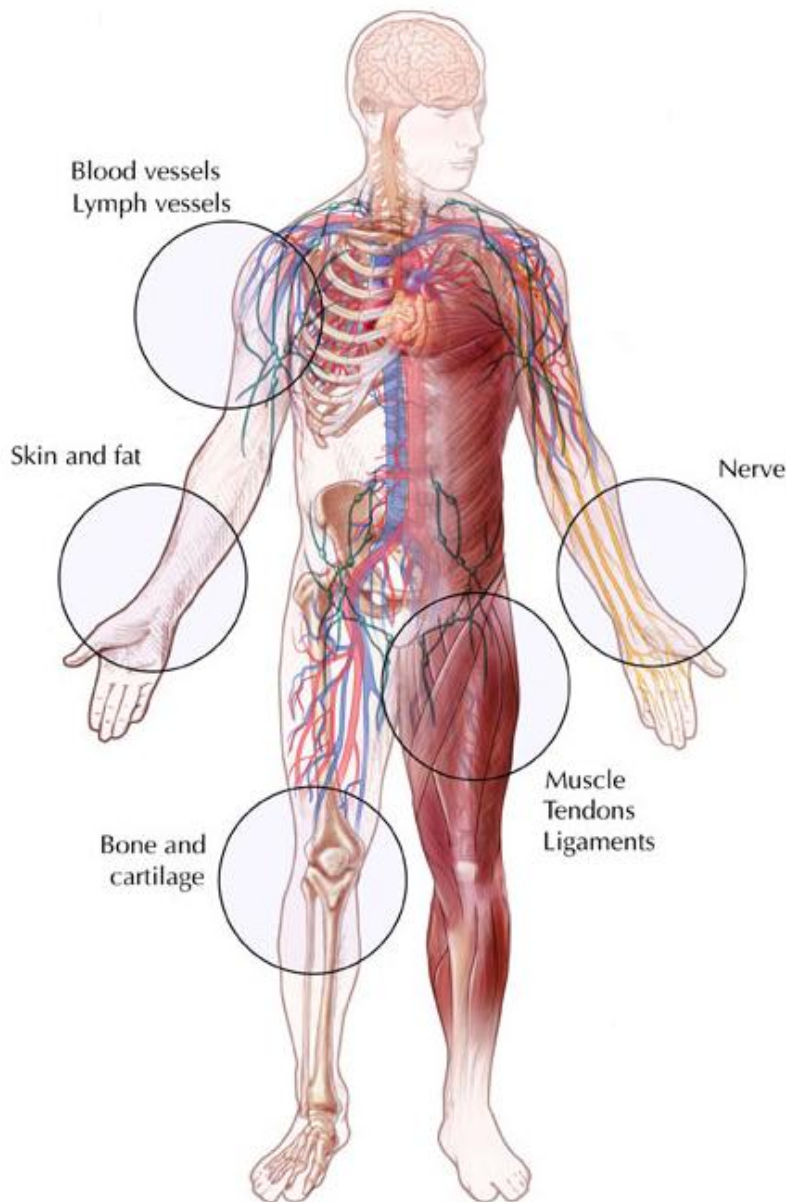
Diagnosis: a medical decision that determines the nature of a disease.

**What are gastrointestinal
stromal tumors or GISTs?**

The Gastrointestinal System



- Also known as the **GI tract** or the **digestive system**.
- Processes food for energy and rids the body of solid waste.

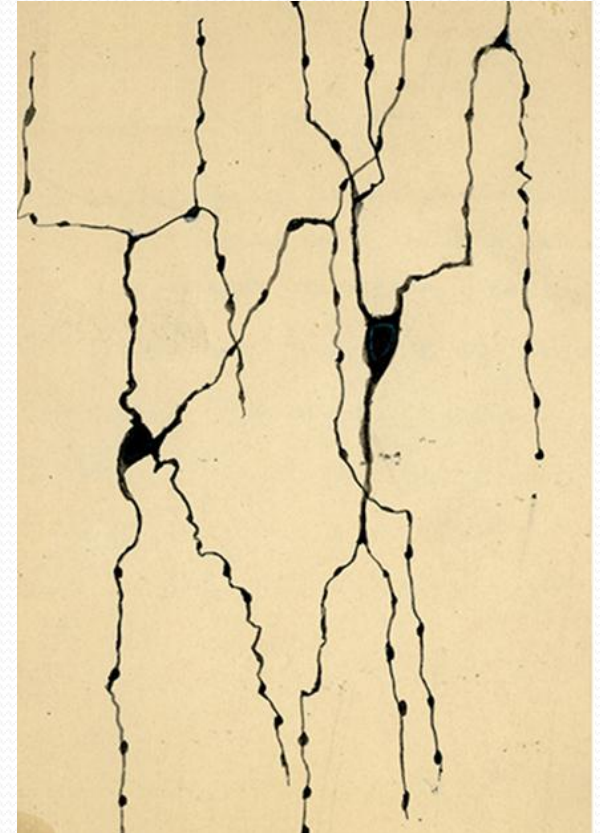
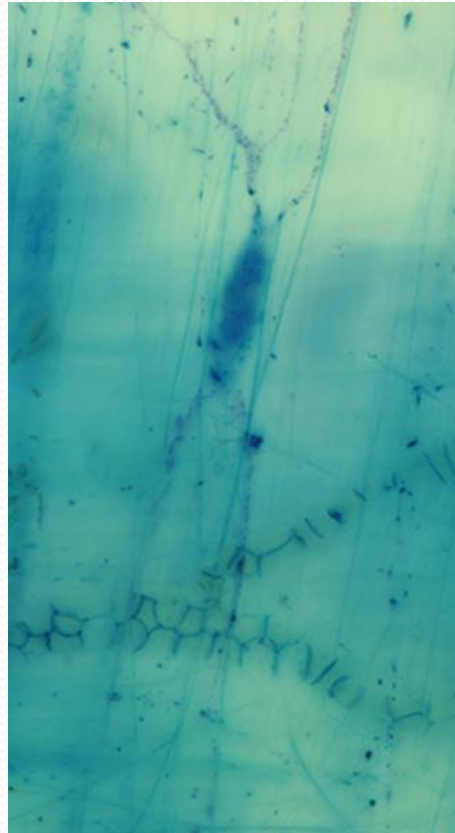


What is GIST?

- Gastrointestinal stromal tumors (GISTs) belong to a group of cancers called **sarcomas**.
- **Sarcomas** are a rare type of cancer that can occur in bones, muscles, fat, nerves, blood vessels, connective tissues, and cartilage.

Interstitial Cells of Cajal or ICCs

ICCs are sometimes called the “pacemakers” because they tell the muscles in the digestive system to move food and liquid through the GI tract.



GIST continued

- Even within GIST, there are different GIST subtypes, like **wild type, pediatric, and familial GIST**. More information about these subtypes can be found on the LRG website.
- GIST is unlike other GI cancers because it starts in different types of cells so the treatment and outlook of GISTs are quite different.
- It is very important for the patient and doctor to understand what type of cancer he or she has, to receive the right treatment.

**What are key GIST
statistics?**

Statistics

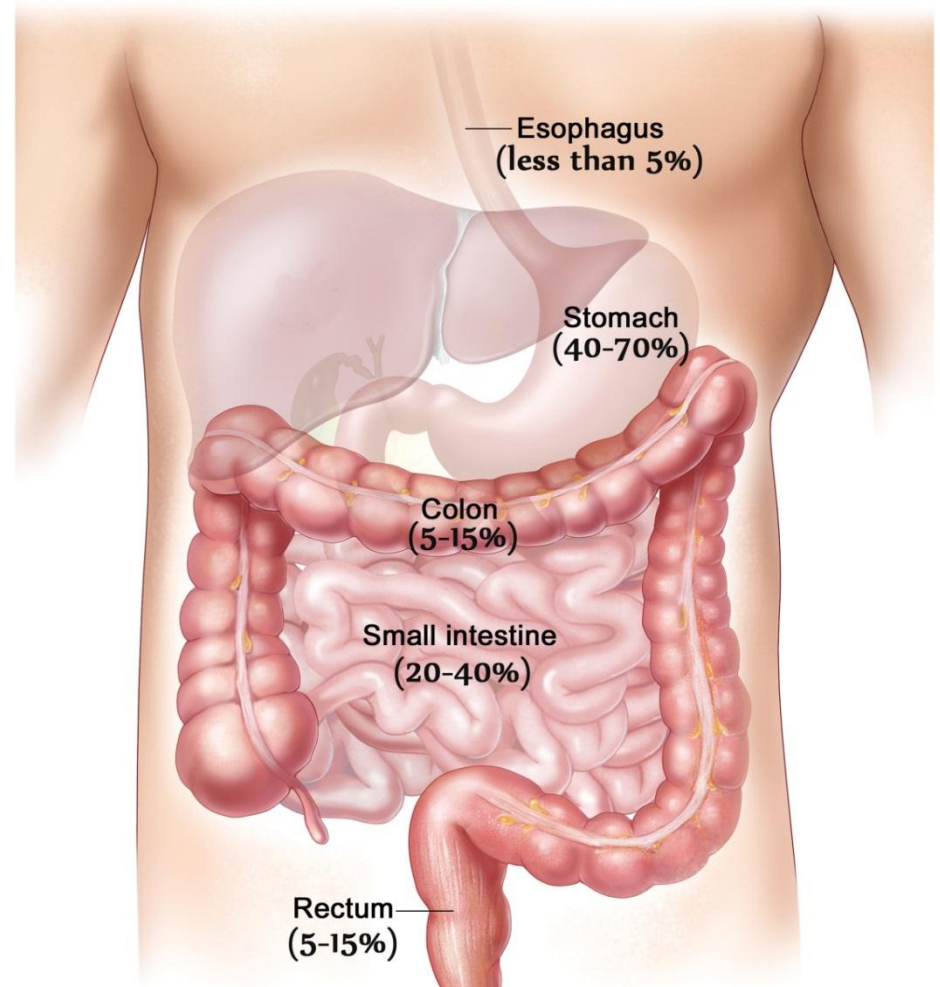
- The exact number of people diagnosed with GIST every year is **unknown**.



- In the United States, it is estimated that there are about **4,000 to 5,000** new cases of GIST each year.
- Most people diagnosed with GIST are **older than 50**, but can occur in any age.

Where is GIST found?

- 40-70% of GISTs arise from the **stomach**
- 20-40% arise from the **small intestine**
- 5-15% arise from the **colon and rectum**
- Less than 5% can also be found in the **esophagus** or elsewhere in the GI tract



Survival Rate

- With the advent of a drug called Gleevec and other treatments the survival rate has increased dramatically.
- “Many patients with **metastatic** GIST survive more than 5 years, some even past 10 years with treatment.” – Jonathan Trent, MD, PhD GIST Specialist

Metastases: The spread of a tumor to a distant location from the original tumor.

**What are some
risk factors of GIST?**

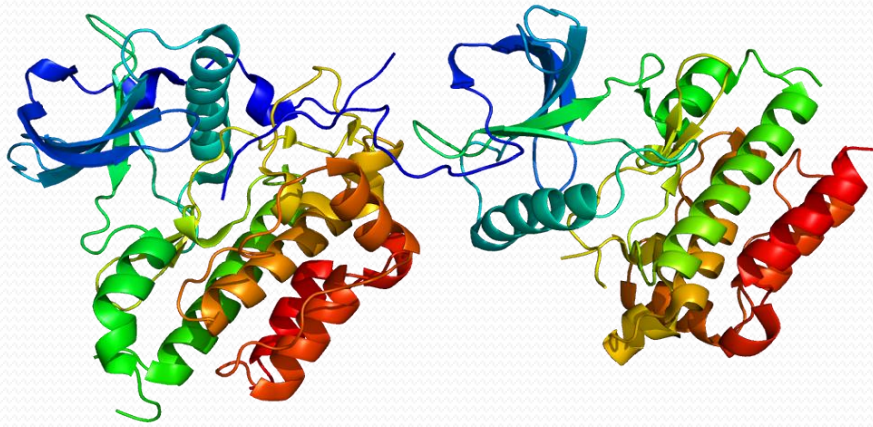


Most GISTs are not inherited and have no clear cause, but rare cases have found members of the same family to have GIST – called **familial** or **hereditary GIST**.

**What are some
causes of GIST?**

C-Kit

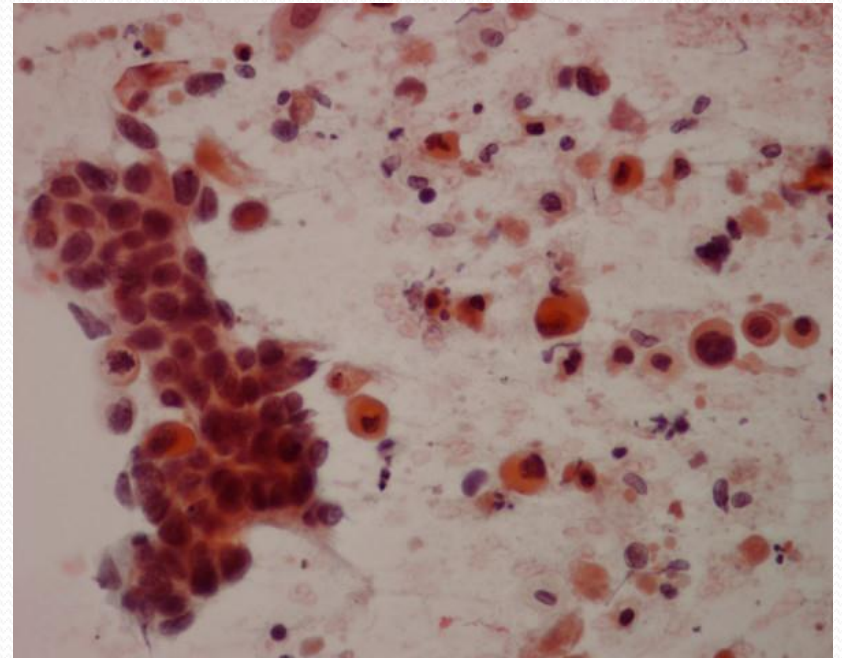
(tyrosine-protein kinase Kit or CD117)



- We still do not know the exact causes of GIST.
- Most GIST cancer cells have a change in a gene that encourages cell growth and division called **c-kit** also called **KIT**.
- The KIT gene is found in all cells of the body and directs a few cells to grow and divide.
- In most GISTs, the KIT is mutated and is always active.
- Cancer cells are always growing and dividing.

Metastasis

- **Metastasis** is when the cells from the original tumor spread through the body to other locations, and create new tumors.
- When a tumor metastasizes, it still acts like the original tumor, but it may be more difficult to treat.



Metastases: The spread of a tumor to a distant location from the original tumor.

**Can someone get
screened for GIST?**

Screening

- At this time, no effective screening tests are available for GIST
- Some GISTs are found incidentally during an exam or surgery for another problem
- Most are found because of symptoms or signs
- Not all tumors may cause symptoms

Possible GIST Symptoms and Signs

- Black and tarry stool
- Abdominal/belly discomfort
- Mass/swelling in the abdomen
- Nausea
- Vomiting (with or without blood)
- Loss of appetite
- Weight loss
- Feeling full after eating small amount of food



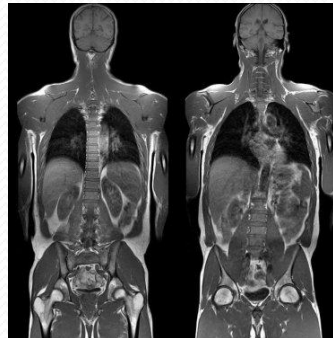
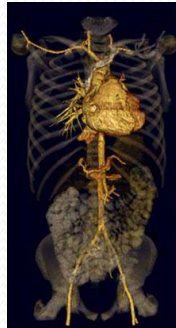
How is GIST diagnosed?

Diagnosis: a medical decision that determines the nature of a disease.

Imaging Test

Imaging tests create pictures of the inside of the body and they include:

- X-rays
- Computed tomography or CT scans
- Magnetic resonance imaging or MRI scans
- Positron emissions topography or PET scans



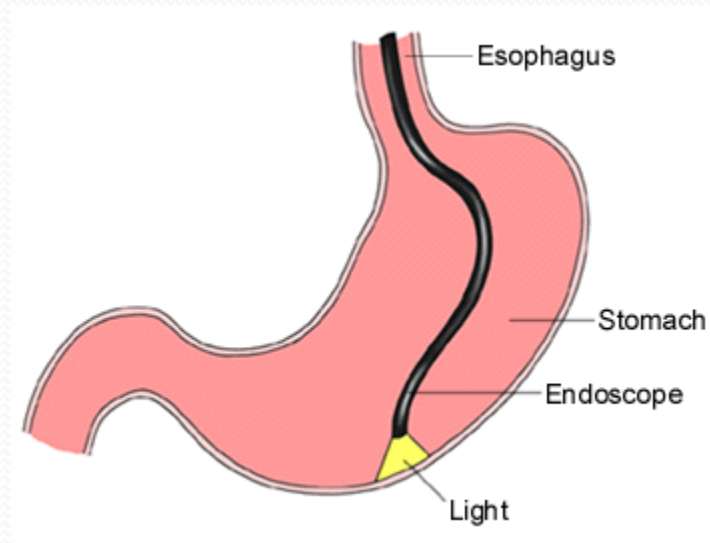
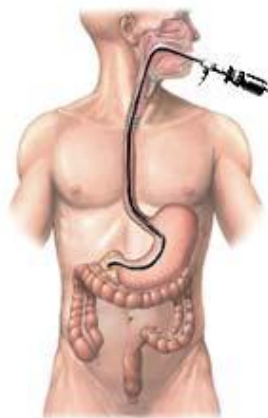
Endoscopy

- Endoscopy uses a slim, flexible tube, called an endoscope
- An endoscope has a tiny camera attached on the end to take photos of any masses inside the body

Lower endoscopy



Upper endoscopy



Biopsy



- If abnormal masses are found, small pieces can be removed by surgery, endoscopy, or needle biopsy to be looked at under the microscope. This process is called a **biopsy**.
- A **biopsy** is the only procedure to ensure proper diagnosis.

**How are GISTs
assessed?**

Assessment

- GIST is assessed by its **risk of recurrence**, sometimes referred to as **staging** in other cancers.
- **Staging** is commonly used in other cancers but it typically is not used with GIST.
- Four important criteria in **risk of recurrence**:
 1. Tumor size (measured in centimeters)
 2. Mitotic index (or mitotic count, mitotic rate)
 3. Tumor location
 4. Whether or not the tumor has ruptured

Risk of recurrence: the chances of GIST returning after surgery.

Staging: the process of determining how far a cancer has spread in the body, often helping to select treatment options and predicting the patient's outlook.

Mitotic Index

- **Mitotic index** is the measurement of how quickly the cancer cells are growing and dividing (a low mitotic index predicts a better outcome).
- Based on tumor size and location, mitotic index and whether or not the tumor has ruptured, the doctor determines the **risk of recurrence** of the cancer.

Risk of recurrence: the chances of GIST returning after surgery.

**Can GIST tumors be
removed?**

Removal of a tumor depends on:



- Size and location of the tumor.
- Whether or not the tumor has spread or **metastasized**.
- Whether the patient is healthy enough for surgery.

Metastases: The spread of a tumor to a distant location from the original tumor.

Removal of a Tumor

- If a tumor can be removed by surgery, it is called **resectable**.
- If a tumor cannot be removed completely, it is called **unresectable**.
- If a tumor is unresectable, a physician may suggest reevaluating the tumor to assess other options.

What kind of treatments are available once GIST is found?

Preparing for Treatment

- Once a GIST is found and assessed, a team of specialists will discuss **treatment options**, depending on the severity of the disease.
- It is very important to **discuss** all treatment options and their side effects to fit their needs.
- Ideally, a patient's treatment should be managed by a **team**.

Initial Treatment



Initial treatment will depend on several factors including:

- Whether GIST has spread or **metastasized**
- The expected difficulty of the surgery
- The size of the original tumor
- The general health of the patient

Metastases: The spread of a tumor to a distant location from the original tumor.

Surgery



- Surgery is typically the first treatment for GIST to remove resectable tumors.
- The goal is to remove the tumor entirely.
- If the tumor cannot be removed, the doctor may treat the patient with medication first to shrink the tumor enough to remove it.

Gleevec



- Also known as “imatinib” can help stop the spread of cancer cells.
- **Gleevec** is a drug taken daily as a **pill or tablet**.
- The standard treatment for GIST tumors that have spread or metastasized.
- About **2 out of 3** tumors shrink by at least half when treated with Gleevec.

Gleevec continued

- Gleevec is sometimes given before surgery with the goal of shrinking the size of the tumor(s) to make surgery easier. This is called **neoadjuvant**
- Gleevec is given as **adjuvant** treatment in hopes of preventing or delaying the tumor from returning
- Gleevec is strongly recommended to patients whose tumors are unresectable or are unable to be removed by surgery

Neoadjuvant: treatment given before surgery

Adjuvant: additional treatment given after usually surgery.

Possible Gleevec Side Effects



- Nausea
- Edema
- Fatigue
- Diarrhea
- Eye puffiness
- Cramping
- Reflux
- Pain
- Weight change
- Eye blurriness
- Skin problems
(such as rashes)

NOTE: In the beginning of treatment, patients may feel side effects but over time many patients get better

For more information, visit the **Side Effects** section on the LRG website

Edema: abnormal accumulation of fluid beneath the skin that produces swelling.

Fatigue: a feeling of tiredness, exhaustion, or lack of energy.

Reflux: regurgitated gastric content.

**Can GIST return after
treatment?**

Recurrence



- If GIST returns after surgery removes the primary tumor it is called a **recurrence**
- **Recurrence** can occur at or near the original location, or new tumors can appear in other locations in the body

Recurrence

- **Recurrences** that occur while a patient is taking Gleevec are typically resistant to Gleevec
- If a patient stopped taking Gleevec before the **recurrence**, there would be a good chance that they would still be responsive to Gleevec
- **Sutent** is an FDA-approved medicine to treat GIST patients who are resistant to or cannot tolerate Gleevec

Recurrence: when GIST returns after surgery.

Sutent: medicine that was approved by the FDA for the treatment of Gleevec-resistant GIST.

Sutent



Sutent is known by several different names:

- Sutent
- Sunitinib malate
- SU11248 (sometimes written as SU011248)

Sutent is taken as a pill and its most common side effects are:

- Diarrhea
- Mouth irritation
- Skin color changes
- High blood pressure
- Increased risk of bleeding
- Swelling

The Benefits of Consultations

1. The National Comprehensive Cancer Network (NCCN) suggests all GIST patients to be evaluated by a multidisciplinary team of GIST experts at a sarcoma center
2. GIST experts will be able to identify any new problems
3. **Consultations** can confirm whether or not patients have a **recurrence**

Recurrence: when GIST returns after surgery removes the primary tumor.

Consultation: a meeting with physicians to evaluate a patient's case and treatment.

Clinical Trial



- Clinical trials are **carefully conducted research studies** about the drug reactions and effects of new and promising drugs and therapy methods, for patients who **volunteer** for them
- If a patient would like to volunteer, he/she should look into facilities doing trials **specifically for GIST**
- Patients must meet **requirements** to qualify for each clinical trial

The LRG Clinical Trial Database

The website offers information about all trials ongoing or completed relevant to GIST

To access the database:

visit the **Clinical Trials** section on the LRG website

www.liferaftgroup.org

**What happens after
treatment?**

Regular Visits to the Doctor

- Make regular **follow-up appointments** with doctors to keep a close watch on any new growth or side effects
- It is recommended that patients get **CT scans every 3 to 6 months** for at least several years after treatment
- Patients should never hesitate to **contact their doctors** about any questions or concerns

Support Groups

- There are many **local groups** across the **U.S.** and **globally**, hosted by members of the Life Raft Group
- Groups are open to **all GIST patients, their caregivers, and family members**
- Great way to meet other patients and advocates coping with GIST and learn more about GIST
- For more information, visit the **Join Support Group** section on the LRG website

www.liferaftgroup.org

Lifestyle Changes



Patients may want to consider making lifestyle changes to help them stay healthy and feel the best they can:

- Healthy diet
- Exercise
- Cutting down on alcohol
- Giving up tobacco



The Life Raft Group

The Life Raft Group (LRG) recognizes that coping with GIST can be very stressful and no one should be without the right support and educational resources.

The mission of the Life Raft Group is to **ensure the survival of GIST patients** while maintaining the quality of their lives. To accomplish this mission, the Life Raft Group devotes its efforts to **research, treatment surveillance, information and support, patient outreach and assistance, and advocacy.**



For more information, visit:
<http://www.liferaftgroup.org>

or email us at:
liferaft@liferaftgroup.org

or call us at:
973.837.9092



10-year GIST survivors at Life Fest 2010