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TREATMENT UPDATE

# Gastrointestinal Stromal Tumors

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TREATMENT UPDATE

# Gastrointestinal Stromal Tumors

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## Editor


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# *Advances in our understanding of GIST have led to newer treatment options.*

**Gastrointestinal stromal tumor (GIST) is a form of sarcoma that arises in the digestive tract.** Sarcomas are cancers of the connective tissues that hold organs together. About 6,000 to 7,000 people are diagnosed with GIST each year. Most people diagnosed with GIST are older than 50, but GISTs can occur in people at any age. They are slightly more common in men.

GISTs occur mostly in the stomach, but they also can arise in the small intestine and, rarely, in the colon or esophagus (the tube leading from the mouth to the stomach). Not all GISTs are cancerous. Often, GISTs are detected by doctors because they cause bleeding into the digestive tract. Brisk bleeding from the lining of the stomach or small intestine can make bowel movements black and tarry. Brisk bleeding from the lining of the large intestine can produce bowel movements with obvious red blood in the stool. Bleeding also can cause a person to vomit blood. Other signs and symptoms of GIST include abdominal pain or fullness, constipation, fatigue, weight loss and—with a large tumor—the development of a lump or mass in the abdomen.

Although there are no blood tests that can tell if you have a GIST, your doctor may order some nevertheless. The results of these tests may alert your doctor to the possibility of a tumor and tell him or her about whether you carry certain gene mutations (changes) that increase the risk of developing a GIST.

# Diagnosing GIST

**To confirm the presence of a GIST, your doctor may order an x-ray examination, computed tomography, magnetic resonance imaging, positron emission tomography, endoscopy or colonoscopy.**

**Computed tomography (CT)** uses a machine to make a series of detailed x-ray pictures of areas inside the body, taken from different angles. The pictures are made by a computer linked to an x-ray scanner. CT scans take longer than regular x-rays and require you to hold still in certain positions.

**Magnetic resonance imaging (MRI)** uses a large magnet, radio waves and a computer to make a series of detailed pictures of areas inside the body.

**Positron emission tomography (PET)** uses a radioactive drug (tracer) injected into the body to detect cancers and

make them stand out on x-rays. PET is particularly useful in identifying GISTs, since most of them are quite small.

**Endoscopy** uses a thin, flexible tube with a light and a lens to look inside the digestive tract. The instrument, called an endoscope, is gently inserted through the mouth and lowered into the esophagus, stomach and upper part of the small intestine to look for the source of the bleeding. Endoscopy also may be used to remove a small piece of tissue (biopsy) for examination under a microscope. The procedure usually is painless but may be somewhat uncomfortable.

**Colonoscopy** uses a thin, flexible, lighted tube with a tiny video camera on the end to search for a GIST in the large intestine (colon). During the procedure, a small instrument can be passed through the scope to remove any suspicious growths for microscopic examination. Patients usually are given a sedative or other medicine to help them relax and sleep through the procedure, so they don't feel anything.

Because of recent advances in our knowledge about GIST and improvements in its diagnosis and treatment, some 50,000 to 70,000 Americans diagnosed with GIST are able to manage their diagnosis and go about their daily lives today.

# Treatment Options

**Once you have been diagnosed with GIST, your health care team will discuss with you the best way to proceed with your treatment. Factors that influence the choice of treatment plan include the size of the tumor, its location, how quickly it is growing, whether it has spread to other parts of the body and your overall health.**

## Surgery

Surgery is the top treatment choice for GISTs that have not spread from the place they started. If the tumor is small, it often can be removed completely using a procedure called laparoscopic surgery. This is done by making one or more small cuts (incisions) above the abdomen. A thin lighted tube called a laparoscope is then inserted into one of the incisions to locate the tumor. Several small surgical instruments are then passed through the other cuts to remove the tumor and some of the surrounding tissue. For a larger GIST, patients may need to have a more standard-sized surgical incision to allow the tumor to be removed. Unlike other cancers, GIST rarely spreads to the lymph nodes, so the surgeon usually does not need to remove the nearby lymph nodes.

If the tumor is large or lies in a difficult location, your doctor may prescribe a short course of targeted treatment (see page 6) before the surgery. This is done to shrink the size of the tumor and to make its removal easier.

After surgery, the tumor needs to be analyzed by an experienced pathologist to determine the risk of GIST returning. Besides examining the tumor under a microscope and measuring it, the pathologist will perform certain tests. These tests include determining the tumor's mitotic index

to establish how quickly the tumor's cells are dividing and growing. The larger the tumor is and the higher its mitotic index, the more likely it will come back after surgery. The pathologist's report also helps doctors to decide whether drug treatment may be needed after surgery to reduce the chances a GIST will return.

For patients with GISTs that have a high risk of recurrence (coming back) after surgery, doctors now recommend that they take imatinib (Gleevec) for up to three years as a preventive measure. Such treatment is not needed if the removed tumor has a low risk of recurrence. After surgery, you should discuss your risk of tumor recurrence with your oncologist and whether or not you should take imatinib.

## Radiation and Chemotherapy

Although often overlooked, radiation may be a useful option for certain patients with GIST. This would include patients with severe pain, especially in the bones, and those with excessive bleeding.

Chemotherapy generally is not an option for the treatment of people with GIST. In clinical trials, chemotherapy has been found to be largely ineffective. Besides, the emergence of effective, well-tolerated targeted treatments for GIST has essentially replaced chemotherapy.

## Targeted Treatments

Targeted treatments have been designed to slow and stop the growth of a GIST by blocking the activity of certain gene mutations. Most GISTs have a mutation of the KIT or PDGFRA gene. The altered proteins from these mutant genes can be targeted by certain drugs, causing tumors to stop growing or even to shrink in some cases.

# The Importance of Clinical Trials

**Clinical trials are the standard by which we measure the worth of new treatments and the quality of life of patients as they receive those treatments. For this reason, doctors and researchers urge people with cancer to take part in clinical trials.**

Your doctor can guide you in making a decision about whether a clinical trial is right for you. Here are a few things that you should know:

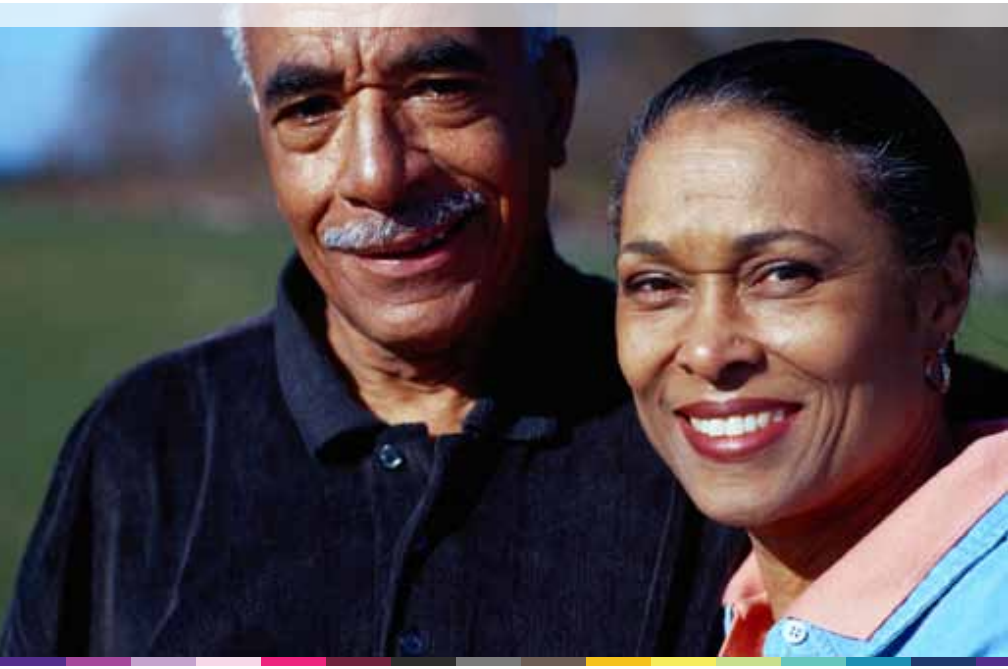
- Often, people who take part in clinical trials gain access to and benefit from new treatments.
- Before you participate in a clinical trial, you will be fully informed as to the risks and benefits of the trial.
- Most clinical trials are designed to test a new treatment against a standard treatment to find out whether the new treatment has any added benefit.
- You can stop taking part in a clinical trial at any time for any reason.

**Imatinib** was the first targeted treatment to be developed for GIST, and it is still the best-tolerated drug for patients to take. However, it has been found that patients with exon 9 mutations may require a higher dose of imatinib. (Exon 9 is a portion of the KIT gene.) Research is being conducted to find out how long imatinib should be given after surgery to help delay or prevent a GIST from coming back. For patients with GISTs that have spread from their original site, indefinite treatment is required. These patients need to be treated until they no longer can safely take imatinib or the tumor becomes resistant to the drug.

**Sunitinib (Sutent)** is very useful in patients with a GIST that has become resistant to imatinib. Sunitinib also may be used when the side effects of imatinib become too difficult for patients to handle. It may be more effective than imatinib for patients whose GIST lacks a mutation in the KIT or PDGFRA gene ("wild-type" GIST).

**Regorafenib (Stivarga)** was approved in 2013 for people with later-stage GIST that cannot be surgically removed and when both imatinib and sunitinib do not work.

For those patients who no longer respond to imatinib, sunitinib or regorafenib or cannot tolerate these medicines, the best treatment option is not known. Patients who find themselves in this situation should talk to their doctor about available clinical trials (see sidebar on page 7) or the use of other targeted treatments, such as sorafenib (Nexavar), pazopanib (Votrient) or ponatinib (Iclusig).



# Working With Your Health Care Team

**It is very important to select a team of health care professionals who can help steer you through your GIST experience and choose the best treatment for you.**

Here are some things to consider:

- Ask your doctors how many patients with GIST they have treated and how familiar they are with GIST. The average oncologist sees perhaps no more than one or two patients with GIST per year. In contrast, comprehensive cancer centers see hundreds of GIST patients yearly and conduct research into its treatment. A full list of comprehensive cancer centers is available on the National Comprehensive Cancer Network's website (www.nccn.org/members/network.aspx).
- Through online patient advocacy resources, patients with GIST and their families can find lists of cancer centers and doctors who specialize in GIST and/or are recommended by other patients with GIST. One of the best of these resources is the Life Raft Group (see page 10).
- When it is time to select a doctor and a treatment center to do your surgery, remember that experience counts.

Once you have a health care team to manage your cancer, it's important to communicate with them. Your doctors and nurses can help you cope with side effects, answer questions about your treatment and monitor your health, particularly if GIST returns. When you meet with your health care team, here are some questions you may want to ask them:

- **How often will I receive treatment?** Try to establish a schedule with your health care team that is the least disruptive to your daily life, if possible.

# The Life Raft Group

**The Life Raft Group supports and provides research, patient support and education and advocacy (supporting a cause) to people with GIST.**

The Life Raft Group website (https://liferaftgroup.org) offers information on the latest treatments of GIST, managing side effects, ways to obtain financial support, finding a support group and a newsletter. Membership is free, and the organization encourages members to become involved. Since the Life Raft Group is a non-profit organization, it seeks volunteers to assist in its projects and activities, such as advocacy, fundraising, community relations, events, patient support and administration.

The Life Raft Group website includes a growing list of more than 160 GIST specialists throughout the United States (https://liferaftgroup.org/gist-specialists/) and contact information for over 40 patient support groups in most states (https://liferaftgroup.org/find-a-support-group/).

To advance future research, the organization maintains a patient registry and a tissue sample bank to help track the status of patients with GIST and provide tissue samples to aid researchers.

The Life Raft Group also offers a six-week expert patient training program. After completing the program, patients will be able to provide expert assistance to other patients with GIST who may be recently diagnosed or beginning treatment.

- **What are the possible side effects of treatment?** Find out how your health care team plans to manage them if they do occur. Create a diary or notebook to keep track of any side effects and share it with your health care team.
- **What are the possible benefits and risks of the treatment proposed?** Make sure that you understand the goals of your treatment, and let your health care team know if you have any wishes or preferences in regard to your treatment. Find out how the treatment plan will affect your daily life.
- **Where can I learn more about this treatment or procedure?** Aside from the information your health care team provides, look up the resources listed on page 17 of this booklet.

# Taking Your Medication Every Day

**Many of the new drugs being developed to treat cancers such as GIST are available as pills or capsules. Medicines that help prevent a recurrence of GIST or treat the side effects of other medicines often come in pill form as well. Research shows that these treatments work best when you follow your health care team's instructions carefully.**

Because pills and capsules are taken by mouth, they might not seem as important as the injections or infusions given at the doctor's office. In truth, oral cancer medicines are just as important as other forms of treatment. Because you are responsible for taking them, adhering to the schedule your doctor has prescribed is especially important, whether you are at work or home, with family and friends or on vacation. However, staying on schedule is not always easy to do.

# Your Support Team

**When you are diagnosed with GIST, you're faced with a series of choices that will have a major effect on your life. Your health care team, family members and friends will likely be an invaluable source of support at this time. You can also turn to these resources:**

**Oncology social workers** provide emotional support for people with cancer and their loved ones. These professionals can help you cope with the challenges of a cancer diagnosis and guide you to resources. Cancer*Care*® offers free counseling from professional oncology social workers who understand the challenges faced by people with cancer and their caregivers. Cancer*Care*'s professional oncology social workers can work with you one-on-one to develop strategies for coping with treatment and its side effects.

Oncology social workers also can help you communicate with your doctor and other members of your medical care team about the health care issues that are important to you.

**Support groups** provide a caring environment in which you can share your concerns with others in similar circumstances. Support group members come together to help one another, providing insights and suggestions on ways to cope. At Cancer*Care,* people with cancer and their families can participate in support groups in person, online or on the telephone.

**Financial help** is offered by a number of organizations to assist with cancer-related expenses such as transportation to treatment, child care or home care.

**To learn more about how Cancer*Care* helps, call us at 800-813-HOPE (4673) or visit www.cancercare.org.**

At the very beginning of your treatment, it is important to get instructions in writing. The instructions should explain:

- The size, appearance and dose of the pill or capsule;
- How often to take it and when;
- Whether to take it on an empty stomach or with food; and
- What to do if you miss a dose.

Understanding how your cancer medicine works and why you are taking it will help you feel more in control. Be sure your health care team explains how your medicine will benefit you. Find out what, if any, side effects to expect so that you're not taken by surprise. Be aware of any foods or other medicines that could interfere with your cancer medication. You may find it helpful to create a drug diary.

Sometimes, people stop taking their medicine because of financial concerns. Some newer cancer treatments can be costly and may not be covered by health insurance. Financial help is available for people with cancer who cannot afford their medicines.

The websites of the Cancer Financial Assistance Coalition (www.cancerfac.org), the Partnership for Prescription Assistance (www.pparx.org) and NeedyMeds (www.needymeds.org) are excellent resources for people who have difficulty paying for their treatment.

You can also contact Cancer*Care*® (call 800-813-HOPE [4673] or visit www.cancercare.org) to learn about financial assistance programs and for a referral to other organizations that may be able to help.

# Frequently Asked Questions

**Q. Does the degree of tumor shrinkage after taking a targeted treatment provide any clue as to long-term prognosis?**

A. Actually, what your doctor is looking for is not that the tumor has gotten smaller but that it has stopped growing. Unlike other tumors that are treated medically, GISTs do not always shrink after targeted treatment.

**Q. I have been taking Gleevec [imatinib] for several years after being operated on for a GIST. On my doctor's recommendation, I have had a PET scan every**

**year to make sure that the cancer hasn't returned. Is getting an annual PET scan appropriate or even wise in this situation?**

A. More and more cancer centers have stopped using PET scans for periodic follow-up because PET scanning exposes the patient to much more radiation than computed tomography or an MRI scan. However, your doctor may be asking you to undergo a PET scan for a specific reason. It's best to speak with your doctor about that.

## Q. Is radiation useful if a GIST has stopped growing after medical treatment?

A. Radiation generally is not as effective as removing a GIST surgically. It also can have more long-lasting side effects than surgery. Radiation usually is reserved to relieve bone pain if the GIST has invaded a bone or to stem bleeding caused by a GIST in the rectum.

# Resources

**Cancer*Care*®**
800-813-HOPE (4673)
www.cancercare.org

**American Cancer Society**
800-227-2345
www.cancer.org

**Cancer.Net**
888-651-3038
www.cancer.net

**Cancer Support Community**
888-793-9355
www.cancersupportcommunity.org

**National Cancer Institute**
800-422-6237
www.cancer.gov

**National Comprehensive Cancer Network**
215-690-0300
www.nccn.org/patients

**GIST Support International**
215-340-9374
www.gistsupport.org

**The Life Raft Group**
973-837-9092
www.liferaftgroup.org

## CLINICAL TRIALS WEBSITES

**Coalition of Cancer Cooperative Groups**
215-789-3600
www.cancertrialshelp.org

**EmergingMed**
877-601-8601
www.emergingmed.com

**National Cancer Institute**
800-422-6237
www.cancer.gov/clinicaltrials

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