

Understanding GIST: Gastrointestinal Stromal Tumours



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The bone & soft tissue cancer charity

About this booklet

This booklet is for anyone who has been diagnosed with a gastrointestinal stromal tumour (GIST). It explains what GIST is, how it is diagnosed and the treatment options available to you. It is your personal guide with space available to keep all your diagnosis information and details of your next hospital appointments. You can also include contact details for your doctor, sarcoma clinical nurse specialist and other healthcare professionals caring for you.

You may find it useful to share the information in this booklet with your partner or family members to help them understand about GIST. If you have any questions about anything you read in this booklet please contact Sarcoma UK. We offer information, support and a signposting service to organisations that can help.

The information provided in this booklet is mainly for adult patients although it may be of some relevance to children and young adults diagnosed with GIST.

Treatment may vary depending on which area of the UK you live in. Please ask your doctor or sarcoma clinical nurse specialist (CNS) about the treatment options available in your area.

Contents

What is GIST?	2
What are the signs and symptoms of GIST?	3
How is GIST diagnosed?	3
Types of diagnostic scans and tests	4
Understanding your diagnosis	5
Who will treat me?	7
What treatment is available?	7
Will there be any side effects of my treatment?	10
What happens after I have had my treatment?	11
What if my cancer has spread to another part of my body?...	12
What support is available?	12

This booklet was produced by **Sarcoma UK**, the only charity in the UK focusing on all types of sarcoma cancer.

Our mission is to amplify awareness, inspire involvement, and fund ground-breaking research to transform the lives of everyone affected by sarcoma.



sarcoma.org.uk



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Online sarcoma.org.uk

By cheque payable to 'Sarcoma UK' and send to: Sarcoma UK, 49-51 East Road, London, N1 6AH

By phone 020 7250 8271

Symbols used in this booklet



Look on our website for more information: sarcoma.org.uk



Questions you might consider asking your doctor or sarcoma clinical nurse specialist?

What is GIST?

GIST is a type of sarcoma, rare cancers that develop in the connective tissues: muscle, bone, nerves, cartilage, tendons, blood vessels and the fatty and fibrous tissues. GIST is the most common type of sarcoma; it develops in the gastrointestinal (GI) tract, a long tube running through the body from the oesophagus (gullet) to the anus (back passage) and includes the stomach and intestines. Most GISTs are found in the stomach and small bowel but can occur anywhere along the GI tract.

Most GISTs occur because a change in the cells (mutation) tells the GIST cells to grow and multiply.

There are also a number of rarer types of GIST:

- **Wild-type GIST**

A type of GIST that is not caused by a known cell mutation.

- **Paediatric GIST**

A GIST affecting children and young adults. Paediatric GIST is very rare

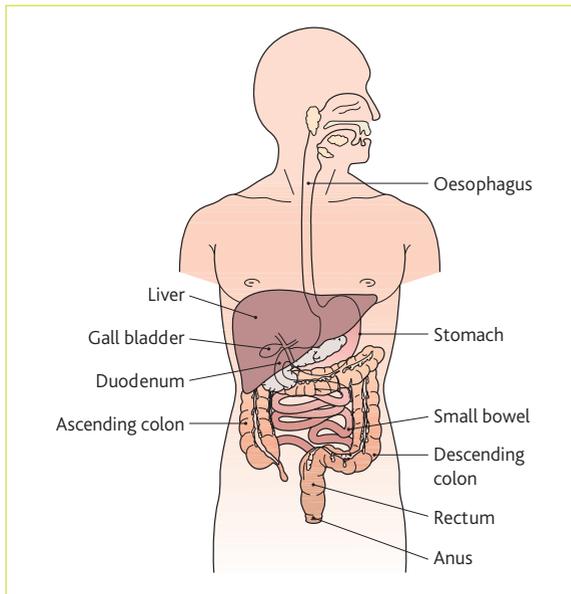
- **Syndromic GIST**

A type of GIST linked to Carney's Triad Syndrome, Carney-Stratakis Syndrome and Neurofibromatosis

- **Familial GIST**

A rare inherited form of GIST

"GIST is the most common type of sarcoma."



What are the signs and symptoms of GIST?

Symptoms of GIST can vary depending on the size and location of the tumour. They may include:

- Fatigue
- Fever and sweating at night
- Discomfort or pain in the abdomen or around organs
- Painless lump in the abdomen
- Feeling sick and vomiting
- Weight loss
- Blood in the stools or vomit
- Anaemia (low level red blood cells)

How is GIST diagnosed?

A diagnosis of GIST may start with a visit to your GP who will examine you and then refer you to a specialist doctor. Some GISTs are discovered through investigations for another medical condition or are diagnosed after surgery. A specialist doctor will diagnose GIST through a series of tests. These may include:

- Clinical examination – looking at or feeling any lump
- A scan – taking pictures of the inside of the body
- A biopsy – taking and testing a sample of tissue

A clear diagnosis will be made after a pathologist with experience in GIST has examined a tissue sample.

"A diagnosis of GIST may start with a visit to your GP."

Types of diagnostic scans and tests

CT

The Computerised Tomography (CT) scan takes a number of x-rays to make a 3D image of an affected area.

Ultrasound scan

Ultrasound uses sound waves to create images of the organs inside the body.

Endoscopy

A thin, flexible telescope called an endoscope is passed through the mouth to the stomach and small bowel. The tip of the endoscope contains a light and a tiny video camera so the doctor can see any abnormalities. The endoscope is sometimes used to take biopsies which will then be sent to a laboratory to be tested.

EUS

The EUS uses an endoscope with an ultrasound scanner attached to investigate tumours like GIST below the lining of the stomach or upper small bowel.

PET

The Positron Emission Tomography (PET) scan shows up changes in tissues that use glucose as their main source of energy. It involves an injection of a small amount of radioactive glucose into the body. The drug travels to places where glucose is used for energy and shows up cancers because they use glucose in a different way from normal tissue.

MRI

Magnetic Resonance Imaging (MRI) uses magnets to create an image of the tissues of the body.

Histopathology

Examination of a tissue sample by a pathologist under a microscope to identify disease.

Blood test

Laboratory analysis of a blood sample. Your doctor may check for anaemia which could be a sign of bleeding in the GI tract.

Understanding your diagnosis

Your doctor will use the results of your scans and tests to diagnose you with a GIST and advise you on treatment options. The best course of action for you is based on the risk category you are put in for your GIST coming back, this is called a recurrence.

The features that are currently used to determine the risk of the cancer coming back in patients with GIST or of the disease spreading to another part of the body are:

Size of the tumour

Large tumours are more likely to behave aggressively than smaller tumours. A small GIST removed when it is less than 2cm in size may not cause problems in the future.

Mitotic count

Mitotic count is the number of actively dividing cells seen under a microscope within a certain area of the tumour. This tells the doctors the rate at which the cancer cells are multiplying and is an indicator of how aggressive the tumour is.

“The specialist doctor will diagnose GIST through a series of tests.”



What sort of tests will I have?

How long do I have to wait for these tests to be carried out?

When will I get my results?

How will I get my results?

“A small GIST removed when it is less than 2cm in size may not cause problems in the future.”

Location

Tumours in the small bowel and rectum appear to be more aggressive than those occurring in the stomach.

Tumour rupture

If the surface of the tumour was damaged during the operation to remove it, or if the tumour was not intact at the time of the surgery, there is a chance that tumour cells may have escaped into the abdomen. Sometimes this alone will define the tumour as high risk.

The risk category you are in will affect your treatment and the number of follow-up appointments you have after treatment.

Mutational analysis

Your tumour should be sent for mutational analysis. There are two main sites of mutation, usually in a gene called KIT, and less commonly in a gene called PDGFRA. While the type of mutation does not routinely influence your treatment options, a mutation in PDGFRA may make the tumour resistant to the standard drugs used to treat GIST. It is important to find out if your GIST is resistant to these drugs if drug treatment after surgery is being considered for you. Fortunately, tumours with a PDGFRA mutation are usually less aggressive than the more common ones with KIT mutation.

Some KIT mutant tumours (exon 9) respond better to larger doses of imatinib but treating exon 9 KIT mutations in this way has not been approved for use in England and Wales.

Unfortunately, some people are diagnosed with GIST when it has already spread to other parts of the body. You can find out more about this in the **What if my cancer has spread to another part of my body?** section on page 12 of this booklet.



Find out more about mutational analysis at www.sarcoma.org.uk/GIST



What risk category am I in?
How will my risk category affect my treatment?

Who will treat me?

In England and Wales the National Institute for Health and Care Excellence (NICE) recommends that anyone with sarcoma should be referred to a specialist sarcoma team for diagnosis and treatment. The care of GIST patients should be supervised by cancer specialists with experience in GIST. You may be treated by a specialist sarcoma team working alongside a specialist gastrointestinal team. Your case will be managed by a team of experts from a wide range of health care professions called a multidisciplinary team (MDT). Your MDT will include your key worker or sarcoma clinical nurse specialist, surgeon and other healthcare professionals involved in your care. See page 16 for more details.

What treatment is available?

The type of treatment you receive depends on what part of the GI tract it is in and your risk category of recurrence. Your MDT will discuss your case and your doctor or nurse will talk you through your options so you are included in deciding what treatment is best for you.

Surgery

Surgery is usually the first treatment method used for GIST. The surgeon will remove the tumour and will aim to take out an area of normal tissue too; this is known as taking a margin. It allows cancer cells that are not visible to the naked eye to be removed along with the tumour. This can reduce the risk of the cancer coming back.

GIST in your small bowel

You may have an operation to remove part of the small

“Surgery is usually the first treatment method used for GIST.”

? What are the possible side effects of my surgery?
What support services are available to me?

bowel. This doesn't usually have any long-lasting side effects.

GIST in your stomach

You may need to have part or most of your stomach removed. This will affect how you eat. Specialist dietitians can give you advice and support on making changes to your diet.

Some tumours cannot be removed surgically. Sometimes the surgery itself can be dangerous, sometimes removing the tumour can damage vital organs. Other treatment options are considered if this is the case for you.

Oral anticancer targeted drugs

Patients with tumours that are too large to be removed safely, or those that have already spread to other parts of the body, can be treated using targeted drugs.

85% of people with GIST have a change in the cells called a mutation. This mutation tells the GIST cells to grow and multiply and is essential to their survival. Targeted agents work by blocking the growth signals within the cancer cells generated by the mutation. The three targeted drugs that are used for GIST are Imatinib (Glivec®), Sunitinib (Sutent®) and Regorafenib (Stivarga).

Imatinib is the first treatment choice for GIST patients. It is effective in 80% of patients and on average will control the disease for about two years. Sometimes the tumour develops a resistance to imatinib and hence over time it stops working. If this happens sunitinib is used as a second treatment option.

The drug Regorafenib (Stivarga) is used to treat people who have GIST that cannot be operated on or has spread to

another part of the body. If you have had treatment with Imatinib and Sunitinib that has not worked or has caused bad side effects then Regorafenib is an alternative treatment.

The All Wales Medicines Strategy Group (AWMSG) has approved regorafenib to treat people with a GIST in Wales.

The Scottish Medicines Consortium have approved regorafenib for treating GIST in Scotland.

NICE has approved Regorafenib to treat people with a GIST in England.

Treatment before surgery

If the tumour was too large to be removed at the time of diagnosis, it may be treated initially by a targeted drug like imatinib. If sufficient shrinkage has occurred after 6-12 months, it may be possible to do an operation both more safely and without the need to remove so much of an organ, like the stomach.

Treatment after surgery

Some people who have had their tumour removed but are at a high risk of the cancer coming back may have a treatment called adjuvant therapy. Adjuvant therapy is an additional treatment after the first treatment which reduces the risk of the cancer returning.

Clinical trials suggest that if it is to be used, giving it for three years is likely to give the best results. Using imatinib as an adjuvant therapy has been approved for use in certain circumstances in Scotland by The Scottish Medicines Consortium (SMC). Adjuvant imatinib is currently available in England and Wales via the Cancer Drugs Fund.



Find out more about clinical trials: sarcoma.org.uk/support-information/clinical-trials

Clinical trials

You may be offered the opportunity to take part in a study to investigate new diagnosis methods, drugs and treatments. Some studies also look at the care and well-being of patients. Your doctor or nurse can give you more information on opportunities for you to take part in a clinical trial.

A number of clinical trials are underway looking at new drugs that may overcome some of the limitations of the existing drugs used to treat GIST. Some clinical trials are trying to find better ways of using the drugs already available. Your oncologist should be able to advise you concerning the availability of a clinical trial for your particular situation.



There is more information on accessing dietary services at sarcoma.org.uk/about-sarcoma/rehabilitation

Will there be any side effects of my treatment?

You may experience side effects from your treatment. If you have had surgery for GIST in your stomach and have had part or most of your stomach removed it may have some lasting impact on your eating habits. Specialist dieticians can give you advice and support on making changes to your diet. Support and information on diet should be available to all patients who have had major abdominal surgery.

Imatinib, sunitinib and regorafenib are strong drugs that can often cause side effects.

Side effects of imatinib can include:

- Tiredness
- Feeling sick
- Diarrhoea
- Swollen ankles

- Puffy eyes
- Itchy rash

Side effects of sunitinib can include:

- Skin rash and soreness
- Tiredness
- Mouth ulcers
- High blood pressure

Side effects of regorafenib can include:

- Tiredness
- Loss of appetite
- Diarrhoea
- Weight loss
- Pain

These side effects can be treated by other medicines and your sarcoma clinical nurse specialist can give you advice and support on managing the effects of taking imatinib and sunitinib. If your symptoms continue or are troublesome please speak to your sarcoma clinical nurse specialist or doctor.

What happens after I have had my treatment?

After treatment you will have regular follow-up appointments for several years. You should receive a follow-up schedule from your sarcoma clinical nurse specialist. The usual practice will include:

- A chance to discuss symptoms
- An examination to look for any signs of GIST returning such as a CT or MRI scan

“After treatment you will have regular follow-up appointments for several years.”

What if my cancer spreads to another part of my body?

A recurrence of GIST may be accompanied by cancer in other parts of the body. This is called metastasis or secondary cancer. Secondary cancers in GIST patients can appear anywhere but they most commonly occur in the abdomen and liver. They are treated with the targeted drugs imatinib and sunitinib. You will be monitored to see how your cancer is responding to these drugs and your follow up schedule will continue throughout your treatment.

What support is available?

Sarcoma UK Support Line

Our Support Line is here for everyone affected by sarcoma.



- Our Support Line is confidential
- We believe no question is a silly question
- We lend a listening ear
- We can point you in the right direction

Emotional support

Talking about it

A diagnosis of cancer can be frightening. There is no right or wrong way to feel or react to diagnosis; you may feel angry, sad or anxious about the future. You may also have concerns for how the news will affect your loved ones. You may find comfort in talking about your concerns with family members or friends.

"It may help to talk to other people."

Talking to others affected by GIST

If you do not feel comfortable discussing your concerns with family or friends, it may help to talk to other people who have been affected by sarcoma.

There are a number of **sarcoma support groups** supported by Sarcoma UK around the country. These offer valuable support and information to patients, carers and family members, and provide the opportunity to meet with other people in the same situation.

Most groups are run by patients and carers working together with local sarcoma clinical nurse specialists or doctors. A typical meeting may include a talk from an invited speaker on a related topic, discussion and questions, informal chat with other group members, and refreshments.

There is also **online support** available for people affected by all types of sarcoma. Online support provides members with the opportunity to get in touch with other sarcoma patients or carers to discuss their concerns over a new diagnosis, treatment options or worries about the future.

GIST Support UK offers support to GIST patients and their carers through a telephone helpline and email forum. They also host two patient/carer meetings a year. Their sub-group **PAWS GIST Group** offers support to patients with paediatric, adolescent, wild type and syndromic-GISTs. **www.gistsupportuk.com • 0300 400 0000**

The PAWS-GIST national alliance aims to raise awareness, improve treatments, stimulate research and understand the



causes and mechanisms of GIST in children, young people and those with wild-type GIST.

www.pawsgistclinic.org.uk

Talking to a professional

You may find it helpful to talk to your sarcoma clinical nurse specialist or doctor about your diagnosis; they will be able to answer any questions you may have about your condition. They may also be able to put you in touch with a counsellor for additional support, providing you with a safe, confidential place to talk about your concerns. Your GP will have access to local counselling services who can provide support to people with cancer.

Practical support

Free prescriptions

Patients who are being treated for cancer can apply for a medical exemption (MedEx) certificate which will allow you to get free prescriptions. You can get an application form from your doctor and it will need to be countersigned by your GP or consultant. Prescriptions are free for all patients in Scotland, Wales and Northern Ireland.

Benefit entitlement

Your sarcoma clinical nurse specialist should be able to advise you on the types of benefits you can claim or any special funding you can apply for. Your local Citizens Advice Bureau can also give you benefits information and many branches can help you fill out application forms. Macmillan Cancer Support have a number of benefit advisors who offer financial advice and support to cancer patients including advice about returning to work following cancer treatment.

? What counselling services are available to me?

“Macmillan Cancer Support offer financial advice to cancer patients.”

My diagnosis details

Date of diagnosis

Details of diagnosis

Treatment plan

My sarcoma multi-disciplinary team (MDT)

You can use this space to keep the contact details of your team members who will be involved in your treatment. You may not see all the healthcare professionals listed below.

Key Worker/Clinical Nurse Specialist (CNS)

Offers support and advice, and acts as an important point of contact for patients when they have a concern.

Name

Direct telephone

Email

Notes

Oncologist

A doctor who specialises in cancer treatments other than surgery, for example, chemotherapy or radiotherapy.

Name

Direct telephone

Email

Notes

Gastroenterologist

A doctor who specialises in the treatment of conditions affecting the intestines.

Name

Direct telephone

Email

Notes

Surgeon

Treats cancer through the removal of tumours.

Name

Direct telephone

Email

Notes

Physiotherapist

Advises on exercises to help with rehabilitation before, during and after treatment with surgery, radiotherapy and chemotherapy.

Name

Direct telephone

Email

Notes

Occupational Therapist

Advises on activities of daily life and equipment to assist recovery and independent living. Also works with local social services to ensure patients are properly supported once they leave hospital.

Name

Direct telephone

Email

Notes

Dietician

Advises on nutrition. They can provide advice and support on any changes you may have to make to your diet.

Name

Direct telephone

Email

Notes

Radiologist

A doctor who specialises in diagnosing medical conditions through images, for example, x-rays.

Notes

Radiographer

A clinical technician who takes images, such as x-rays, and may also give radiotherapy as a treatment.

Notes

Pathologist

A doctor who identifies diseases by studying tissue samples.

Notes

Other professionals

You can record the contact details of other professionals you come into contact with in the space below. These may include a psychologist or social worker.

Name

Contact details and notes

Name	Contact details and notes
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The
Information
Standard

Certified member

This organisation has been certified as a producer of reliable health and social care information.

www.theinformationstandard.org

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