



Understanding GIST

sarcoma.org.uk



Sarcoma UK
The bone & soft tissue
cancer charity

About this booklet

This booklet is aimed at 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 also has information on where you can go for support. It is your personal guide with space available to keep all your diagnosis information and the details of your next hospital appointments.

You can also include contact details of your doctor, sarcoma clinical nurse specialist or other health professionals.

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 questions about anything you read in this booklet please contact Sarcoma UK.

We offer information, support and a signposting service to other organisations that can help.

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

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What is a gastrointestinal stromal tumour (GIST)

What are the different types of 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 caused by a mutation of the KIT or PDGFRA gene. A mutation is a change in cells that causes 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 KIT or PDGFRA 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

An extremely rare inherited form of GIST

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:

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

GIST is the most common type of sarcoma.

How is GIST diagnosed?



What does a GIST diagnosis involve?

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 other medical conditions or even 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.

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

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

“The endoscope is sometimes used to take biopsies which will then be sent to a laboratory to be tested.”

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

Understanding your diagnosis



What is the risk category of my GIST?

Your doctor will use the results of your scans and tests to advise you on treatment options. The treatment for you is based on the risk category you are put in for your GIST coming back. Your risk category could be high, intermediate or low.

This is worked out by looking at:

Size of the tumour

Large tumours are more likely to behave aggressively than smaller tumours.

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, which shows how aggressive the tumour is.

Location

Tumours in the small bowel and back passage are more aggressive than those in the stomach.

Tumour rupture

Sometimes the surface of the tumour can be damaged during the operation to remove it. In other cases the tumour may not be intact at the time of the surgery. In these cases there is a chance that tumour cells may have escaped into the abdomen, which can put you at a higher risk of your GIST coming back.

Mutational analysis

When your tumour is removed it should be sent for mutational analysis. This test finds out what mutation has caused the GIST.

There are two main sites of mutation in GIST:

- In a gene called KIT
- In a gene called PDGFRA

KIT is more common. However if your GIST is caused by a PDGFRA mutation it may affect your treatment options. This is because the drugs used to treat GISTs may not work properly on GISTs caused by a PDGFRA mutation.

Who will treat me?

Anyone with GIST 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. Your sarcoma team will work 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, GI surgeon and other healthcare professionals involved in your care.

"Your sarcoma team will work alongside a specialist gastrointestinal team."

What treatment is available?

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

The type of treatment you receive depends on what part of the GI tract it is in and your risk category of your GIST coming back. 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 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 dieticians can give you advice and support on making changes to your diet.

Some tumours cannot be removed surgically. Other treatment options are considered if this is the case for you.

Targeted drugs

If your tumour is too large to be removed safely, or your GIST has already spread to other parts of the body, it can be treated using targeted drugs. Most people with GIST have a change in the cells

“Some tumours cannot be removed surgically. Other treatment options are considered if this is the case for you.”

called a mutation. This mutation tells the GIST cells to grow and multiply. Targeted drugs work by blocking the growth signals, causing the GIST to stop growing and shrink. The three targeted drugs that are used for GIST are Imatinib, Sunitinib, and Regorafenib.

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.

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.

Treatment with these drugs may harm your fertility. Please speak to your doctor or clinical nurse specialist about your options for preserving fertility before treatment.

It is not advised to become pregnant, breastfeed or father a child when on these treatments.

“Please speak to your doctor or clinical nurse specialist about your options for preserving fertility before treatment.”

“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.”

Treatment before surgery

If the tumour was too large to be removed at the time of diagnosis, it may be treated by a targeted drug like imatinib. If enough shrinkage has occurred after 6–12 months, it may be possible to do an operation more safely.

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.

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. Ask your doctor for more information on clinical trials, or take a look at sarcoma.org.uk/clinical-trials-hub.

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.



What side effects should I look out for?

"If your symptoms continue or are getting worse, please speak to your sarcoma clinical nurse specialist or doctor."

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 getting worse, please speak to your sarcoma clinical nurse specialist or doctor.

What happens after I have had my treatment?

After your treatment is finished you will have follow-up appointments for several years. This is where your doctor can look for signs of your GIST coming back. At follow up appointment you will have:

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

If your GIST does come back a new treatment plan will be put in place for you.

What if my cancer spreads?

GISTs can spread to other parts of the body. This is called metastasis or secondary cancer. Secondary cancers in GIST patients can appear anywhere but are more common in the abdomen and liver. You will be treated with the targeted drugs imatinib and sunitinib. During treatment you will be monitored to see how your cancer is responding to these drugs. You should get support from your sarcoma clinical nurse specialist on managing symptoms and side effects.

"Secondary cancers in GIST patients can appear anywhere but are more common in the abdomen and liver."

What support is available?



What rehabilitation services are available?

Find out more about rehabilitation services at sarcoma.org.uk/rehabilitation

There are many services which can support you throughout your treatment and recovery. More information on physical support and rehabilitation can be found in our Understanding Rehabilitation and Life After Treatment booklet.

Rehabilitation for GIST can involve the services of:

- Speech and language therapists
- Dieticians
- Psychological or counselling services
- Clinical nurse specialists

If you have any questions about any aspect of your physical recovery, you can ask your Clinical Nurse Specialist who will be able to provide you with more information.

Emotional and psychological support

- Sarcoma UK run a **Support Line**, which offers independent confidential advice and support. Our Support Line is available to anyone who has been affected by sarcoma. You can contact us by phone on 0808 801 0401 or by email at supportline@sarcoma.org.uk
- There are a number of **Sarcoma Support Groups** across the country. These provide support and information to patients, carers and family members, and provide an opportunity to meet locally and informally. www.sarcoma.org.uk
- **Maggie's Centres** provide support and advice through a team of cancer support specialists,

benefits advisors, nutritionists, psychologists and therapists. They have 21 centres across the UK, and can offer support online at: www.maggiescentres.org.uk

- **Macmillan Cancer Support** offer support services within hospital cancer centres and across the community. More information can be found at: www.macmillan.org.uk
- **The Professional Standards Authority** provide information on all accredited counselling and psychotherapy services in the UK. More information can be found on their website: www.professionalstandards.org.uk

GIST specific support

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. www.gistsupportuk.com

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

Sarcoma Support Line
You can contact us by phone on 0808 801 0401
supportline@sarcoma.org.uk

What if I am struggling to get the help I need?



What services are available for further help?

If you feel that you are not getting the right support or you are not getting access to the support you need, there are a number of services that can help.

- **The Patient Advice and Liaison Service (PALS)** offers confidential advice and information about healthcare related issues. PALS can help resolve concerns or provide information about the NHS complaints procedure. You can find PALS at your local NHS hospital.
- If you have any challenges getting support, the **Citizens Advice Bureau** can provide free and impartial advice. You can visit their website for information and to find the closest centre to you.
- **Advocacy services** can provide help to ensure your voice is heard. These services may give advice about ways to have more say in your care. There are many different advocacy services available, including Support Empower Advocate Promote www.seap.org.uk and POhWER www.pohwer.net

Getting financial support

Taking time away from work for treatment and recovery can affect your finances. There might also be additional expenses to factor in, such as childcare or travel. There are a number of services which aim to help with this.

Grants and Benefit entitlement

Your sarcoma **Clinical Nurse Specialist** should be able to advise you on the types of benefits you can claim, or on any special funding you are eligible to 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.

Free prescriptions

Patients who are being treated for cancer in England 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.

“Patients who are being treated for cancer in England can apply for a medical exemption (MedEx) certificate...”

My diagnosis details

Date of diagnosis

Details of diagnosis (type of sarcoma)

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

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 dietician, psychologist or social worker.

Name	Contact details and notes

Name	Contact details and notes

Name	Contact details and notes

Appointment diary

You can keep track of your hospital appointments by recording them in the space below.

Date	Hospital	Appointment / Treatment

Date	Hospital	Appointment / Treatment

Date	Hospital	Appointment / Treatment

Date	Hospital	Appointment / Treatment

Date	Hospital	Appointment / Treatment

This booklet has been produced by the Information and Support Team at Sarcoma UK. It has been reviewed by Sarcoma UK's Information Review Panel which includes healthcare professionals and people affected by sarcoma.

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Sarcoma UK makes every reasonable effort to ensure that the information we provide is up-to-date, accurate and unbiased. We hope this booklet adds to the medical advice you have received and helps you make informed decisions about your care and treatment. Please speak to a member of your care team if you are worried about any medical issues. Sarcoma UK does not necessarily endorse the services provided by the organisations listed in our publications.



SarcomaUK

The bone & soft tissue
cancer charity

**Sarcoma UK is a national
charity that funds vital
research, offers support
for anyone affected by
sarcoma cancer and
campaigns for better
treatments**

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