GIST for Beginners

A guide for patients by patients

You do not have to be alone!

GIST support UK
Gastrointestinal Stromal Tumour.
This is the name of a rare form of cancer which one usually never hears about. This Guide is for all GIST patients, their relatives and for others who may be interested. It is a short explanation of GIST and how it is treated.

This booklet has been inspired by a booklet produced for the Stichting Contactgroep GIST Nederland-België. This was written by Ellen van Arem de Haas and she has very kindly translated it into English for us to use.

DISCLAIMER
This booklet has mostly been written by patients, except where otherwise stated.

The information is not intended to replace expert medical advice that you may be given. We do our best to ensure that any information we publish is accurate, but any decisions about your treatment should be made after discussions with your consultant, (preferably a GIST expert).
Introduction........................................................................................................ 1
What is cancer and what is GIST? ................................................................. 2
What causes GIST? ......................................................................................... 4
Possible symptoms of GIST ......................................................................... 5
Diagnostic tests .............................................................................................. 6
Diagnosis of GIST ............................................................................................ 8
Risk of Recurrence ......................................................................................... 9
Mutation analysis ........................................................................................... 10
Special rare types of GIST .......................................................................... 11
Treatment of GIST ......................................................................................... 12
What if imatinib does not work or stops working? .................................. 14
How targeted drugs work. ........................................................................... 17
Side effects ...................................................................................................... 18
Nutrition .......................................................................................................... 18
Guidelines for treating GIST ....................................................................... 21
Compliance ..................................................................................................... 22
Preparing for your next visit to the oncologist ....................................... 22
Life with GIST ............................................................................................... 23
Finding other GIST patients ......................................................................... 24
About GISTSupport UK ............................................................................... 25
PAWS-GIST Group ......................................................................................... 26
Funding .............................................................................................................. 26
Specialist GIST centres in the UK .............................................................. 27
Speakers at recent GSUK Patient Meetings .............................................. 28
Useful Links ................................................................................................... 29
Introduction

I had my first surgery for a “benign leiomyoma” back in 2001, and I still remember the feelings of fear and isolation I felt when I heard that it was a GIST. My excellent GP said that she had not heard of it, and that we would have to learn together. At the time there was nothing written about GIST except for an article in The Lancet, and as you can imagine this was not very patient friendly. This booklet is really the one I wish I had had back then.

Great strides have been made in the treatment of since 2001, and much more information is now available for GIST patients and their families. The Liferaft Group, based in the USA, was the first patient group to be established, and it has a very informative web site. Various other groups have produced information books about GIST, but none of them is aimed particularly at the UK patient group.

My greatest wish after my diagnosis was to meet other GIST patients. Since GIST is rare, the chances of meeting another GIST patient are very small. I remember very well the relief at meeting a dozen or so others at the first meeting of GIST Support UK. I was no longer so alone, and I could talk to others who really understood.

So welcome to the GIST Community!

Judith Robinson
What is cancer and what is GIST?

Our body makes new cells all the time. Each cell contains information which is copied during cell division. The control mechanism for this process can be disturbed and cause uncontrolled cell growth and division. This produces a tumour.

There are many different kinds of cancer.

**Carcinomas** such as breast, colon, lung and prostate cancers grow from epithelial cells. These are cells that line the major cavities of the body.

**Sarcomas** grow in the connective tissues. Soft tissues are generally the tissues beneath the skin, surrounding the organs and bones, and filling the spaces in between. GIST is a soft tissue sarcoma.

The treatment of sarcomas is a speciality. Sarcomas are much less common than carcinomas, so there are relatively few doctors who specialise in sarcomas. GIST Support UK recommends that you are treated in a specialist GIST centre. There are also smaller hospitals or cancer centres that work closely with the specialist centres. In any case, unless you have a very small GIST on the stomach, it is probably a good idea to make sure that you are being treated in a specialist centre, or at a hospital that has links to one. Your GP can arrange for you to be referred to specialist centre for a second opinion. (See the list on page 27)

GISTs can vary greatly in size and in the ease with which they can be treated. They may be very small, 1-2 cms in diameter, or they can be large, up to 20 or more cms in diameter, with their own blood supply. They can also appear in a single easily removable capsule, or be inextricably wrapped round
internal organs. Secondary tumours, called metastases, can also develop in a different place from the original primary tumour, usually in the liver.

It is very important that the primary GIST is found early and treated, to reduce the risk of it metastasizing, or spreading, to the abdomen or the liver. Although large fast-growing GISTs are the most likely to return, even small tumours may metastasize eventually.

GIST is most commonly found in the stomach (40%), followed by the small intestine (30%), the rectum (10%), and colon (5%). GIST is also rarely found in the oesophagus. Occasionally GIST may be discovered outside the gastrointestinal tract. This can be in the omentum (a large fatty structure covering the intestines inside the abdomen), in the peritoneum (a thin membrane that forms the lining of the abdominal cavity), or in the retroperitoneum (the space in the abdominal cavity behind the peritoneum).

When a GIST is found, metastases may already have grown in other organs. Metastases are found in about half of newly-diagnosed GIST patients, usually in the liver. GISTs rarely spread through lymph nodes. Lung and bone metastases are very rare.

If the tumour bleeds into the abdomen or ruptures during surgery, cells may get into the blood stream, and the GIST can then spread into the abdominal cavity, but metastases are most often found in the liver. This is because blood goes directly from the stomach and intestines to the liver, so GIST cells can sometimes implant in the liver. This is not liver cancer, but GIST in the liver.
GISTs in the stomach or liver should be treated quite differently from the much more common stomach or liver cancers.

**What causes GIST?**

GIST arises from the 'interstitial cells of Cajal'. These are small cells in the outer wall of the digestive tract, commonly called the gut. GISTs can grow from this wall into the abdominal cavity, and sometimes grow very large before causing any problems and being discovered.

All the cells in the body have KIT and PDGFRA genes. If either of these genes changes (mutates), there is a risk of GIST developing. In most cases of GIST, it is the KIT gene that mutates. In about 10% of cases it is the PDGFRA gene that mutates. In a further 10% of cases, neither of these genes has mutated, but a GIST develops anyway. These are called "Wild type" GISTs.

Although GIST is diagnosed mainly in people aged between 40 and 60, it can occur in people much younger than this and even (very rarely) in children. In younger people it is usually called paediatric GIST and it differs in some ways from the types of GIST found in older people. About 54% of GIST patients are male, 46% are female.

GIST is hardly ever hereditary, so there is little likelihood of you passing it to your children.

It is estimated that there about 600-1000 new cases of GIST diagnosed per year in the UK. Accurate data is now being collected.
Possible symptoms of GIST

GISTs often show no symptoms for a long time. The first symptoms are often caused by the tumour pressing on some other organ. Sometimes the GIST may bleed into the abdominal cavity or the gut. This causes anaemia. If the GIST is large, the doctor may be able to feel a swelling in the abdomen. GISTs, particularly in the liver, can also cause night-sweats.

**GIST in the oesophagus** can cause difficulty with swallowing (dysphagia).

**GIST in the stomach** can cause pain or discomfort, indigestion, nausea, vomiting, feeling of fullness, bleeding into the gastrointestinal tract causing black coloured stools, or any combination of these.

**GIST in the intestine** can cause bleeding, pain, constipation, diarrhoea, pain, or just vague abdominal discomfort.

All these symptoms can be caused by common problems. However if they haven’t responded to simple treatments, or got better on their own, your GP will then have referred you to a gastroenterologist. Some of the following tests will then be done.
Diagnostic tests

CT scan (Computed Tomography)
This is the commonest diagnostic tool. It uses X-rays to build up a three-dimensional picture of your inside. It is painless and uses a small amount of radiation. You will usually be asked to drink some water and be given an injection of a contrast medium into a vein. If for some reason you cannot take the contrast medium, you will be given a special liquid to drink before you have the scan.

Ultrasound
This is a painless and harmless procedure using no X-rays. It is used all the time for looking at unborn babies, but is also good for finding tumours.

Endoscopy
A tube with a tiny camera is passed into the stomach. You will be sedated for this procedure. GISTs growing on the outside of the stomach cannot be seen. Sometimes ultrasound is done from inside the stomach to give a clearer picture than would be possible with ordinary ultra-sound. This is called endoscopic ultrasound (EUS). This will show tumours both inside and outside the stomach. During an endoscopy, small pieces of the tumour can sometimes be removed for examination. This is called an endoscopic biopsy.

MRI-scan (Magnetic Resonance Imaging)
This is a technique for measuring the activity of cells and their blood supply by using a magnetic field and radio waves. It uses no radiation, is painless, but noisy, and takes quite a long time.
PET scan (Positron Emission Tomography)
This is rather like a CT scan but you have to take a drink containing a kind of glucose which is slightly radio-active. Any very actively growing cells will absorb more of this glucose than other cells and are then highlighted on the scan. A PET scan is usually used either before surgery if the doctor thinks there may be more than one tumour, or to see how your tumour(s) is responding to treatment.

Blood tests
Your blood will be tested to see whether you are anaemic, and to check whether your liver and kidneys are working well.

Histology
This means looking at the cells from your tumour under a microscope. If a biopsy is taken before you have any surgery, this biopsy sample will be examined and this will help to confirm the diagnosis. If your tumour is removed, it will be looked at by the pathologist. The diagnosis of GIST can then be confirmed. The edges of the tissues removed will be very carefully examined to make sure that all the tumour has been removed.

This is what GIST cells look like under the microscope when they have been stained by the pathologist
This diagram shows the steps that you may have been through before it is suspected that you have a GIST.

**Diagnosis of GIST**

Although the site of a tumour may suggest that it is a GIST, GISTs can only be definitely diagnosed after the pathologist has looked at the cells under his microscope. This may be done before any treatment if a tissue sample has been removed by a biopsy. Otherwise a firm diagnosis will be made after the tumour has been removed.
The pathologist will also estimate the rate at which the tumour is growing. This is done by counting the number of cells which are in the process of dividing (undergoing mitosis) in 50 high-power fields of his microscope. This is the “mitotic count per 50 HPF”. The higher this is, the faster the tumour is growing.

**Risk of Recurrence**

The risk of a GIST coming back after surgery depends on three things:

- How big it is
- Its mitotic count (ie how fast the tumour cells are multiplying)
- Where it is growing

<table>
<thead>
<tr>
<th>Risk category</th>
<th>Tumour size (cms)</th>
<th>Mitotic count per 50HPFs</th>
<th>Place of primary tumour</th>
</tr>
</thead>
<tbody>
<tr>
<td>Very low</td>
<td>Less than 2</td>
<td>5 or less</td>
<td>Anywhere</td>
</tr>
<tr>
<td>Low</td>
<td>Between 2 and 5</td>
<td>5 or less</td>
<td>Anywhere</td>
</tr>
<tr>
<td>Intermediate</td>
<td>Between 2 and 5</td>
<td>More than 5</td>
<td>Stomach</td>
</tr>
<tr>
<td></td>
<td>More than 5</td>
<td>Between 6 and 10</td>
<td>Anywhere</td>
</tr>
<tr>
<td></td>
<td>Between 5 and 10</td>
<td>More than 5</td>
<td>Stomach</td>
</tr>
<tr>
<td>High</td>
<td>Any size</td>
<td>Any</td>
<td>The tumour has ruptured before or during surgery</td>
</tr>
<tr>
<td></td>
<td>More than 10</td>
<td>Any</td>
<td>Anywhere</td>
</tr>
<tr>
<td></td>
<td>Any size</td>
<td>More than 10</td>
<td>Anywhere</td>
</tr>
<tr>
<td></td>
<td>More than 5</td>
<td>More than 5</td>
<td>Anywhere</td>
</tr>
<tr>
<td></td>
<td>Between 2 and 5</td>
<td>More than 5</td>
<td>Not in the stomach</td>
</tr>
<tr>
<td></td>
<td>Between 5 and 10</td>
<td>5 or less</td>
<td>Not in the stomach</td>
</tr>
</tbody>
</table>

Note: This table is proposed by a British group of GIST experts, adapted from a table produced by H Joensuu in 2008. It has not yet been validated.
The best GIST to have (apart from none!) is a small one on the stomach, growing very slowly.

**Mutation analysis**

Mutation analysis is probably a key predictor for the course of the disease, and may determine treatment options in the future.

If GIST is diagnosed, there is usually a mutation discovered in the KIT or PDGFRA genes. This change takes place at a particular place in the chromosome, called an exon.

KIT mutations occur in about 80% of GISTs:
- Exon 9: mostly in tumours in the small intestine
- Exon 11: the commonest mutation, which can occur anywhere in the gut
- Exon 13: rare
- Exon 17: rare

PDGFRA mutations occur in about 10% of GISTs:
- Exon 12
- Exon 14 is very rare
- Exon 18 is the most common

We recommend that all patients with GIST should ask to have mutational testing performed, and we are lobbying to make this standard within the UK. Mutational testing is particularly important in the case of tumours in the high risk group.

There is now a Biobank of GIST tissues where fresh tumour or wax blocks of tissue can be stored.
Special rare types of GIST

Wild-type GIST
If none of the normal mutations already described is found, nor any other mutations in KIT or PDGFRA, then the tumour is called “Wild-type”. This can be found in patients of any age but is commonest in younger patients.

Paediatric GIST
This is GIST in children or young people under 25. It is extremely rare.

Syndromic GIST
GIST may occur as part of Carney’s Triad, which is a combination of GIST, lung chondroma and paraganglioma. Two or three of these may occur together. It may also occur in the Carney-Stratakis Syndrome. Both of these syndromes are extremely rare. GISTs may also occur as part of the neurofibromatosis syndrome (NF1).

Familial GIST
In an extremely small number of families in the world, there appears to be an inherited form of GIST.

This is Pippa, one of our younger patients, who was selected to carry the Olympic torch. She is a fantastic ambassador for young GIST patients. Pippa has been living with Paediatric Wild-type GIST since October 2009. After two operations to remove tumours, Pippa enjoys life as a normal 18 year old, but with the uncertainties that go with having such a rare form of cancer.
Treatment of GIST

GIST requires a multi-disciplinary approach, which means that a group of experts should be involved, probably including:

- A surgeon
- A gastroenterologist (a specialist in the gastrointestinal tract ie the gut)
- An oncologist (a doctor who specialises in treating cancers)
- A pathologist
- A radiologist
- A specialist nurse

They form the Multi-Disciplinary Team, (MDT), and decide together on the best treatment for each patient.

It is really important that a relationship of trust is established between you and your doctors. If for any reason you feel uncomfortable about the way you are being treated, do ask for a second opinion. This is particularly important if you are facing major surgery. Your GP can arrange for a second opinion, and you may be able to find suitable GIST experts from our website or from the lists on pages 27 and 28.

What are the options?

1 Surgery

If the tumour is small and easy to get at, and has not spread, surgery is the usual treatment. This may be possible laparoscopically, (by key-hole surgery), but open surgery may be needed. You may like to get a copy of our booklet, “Living after Surgery for GIST”.
2 Adjuvant treatment with imatinib (Glivec®) after surgery
If the tumour has a high risk of recurrence, trial data shows that taking imatinib after surgery statistically increases the time to recurrence. It also increases overall survival, and more data is being collected to confirm these findings. This use of imatinib has not yet been approved in England and Wales, but funding may be obtainable from the Cancer Drugs Fund.

3 Treatment with imatinib before surgery
This is the usual first-line treatment in the UK if there is more than one GIST, or when the hospital team decide that the tumour or tumours are too difficult to remove surgically. Imatinib usually stops the tumour or tumours growing. It may even cause tumours to shrink, occasionally to the point when they can be removed surgically. However funding is not normally available for a primary tumour to be shrunk in this way, unless, as stated above, it is too difficult to be removed surgically.

Imatinib does not have the same side effects as normal chemotherapy, used for many other cancers. Normal chemotherapy affects all growing cells, and causes healthy cells to be destroyed. This leads to a number of difficult side effects like the loss of all body hair. Imatinib works differently and is one of a group of drugs called targeted drugs (see page 17). These act mainly on the GIST cells, so they cause far fewer side effects than the usual drugs used to treat cancer. Targeted drugs are taken every day as a pill. While you are taking imatinib you should have regular blood tests to make sure that you are not anaemic and that your liver and kidneys are working well. You should also have CT scans every three months.
What if imatinib does not work or stops working?

1. **Change to sunitinib (Sutent®)**

   Because of the problem of resistance, other drugs are being developed to use when imatinib stops working. The only one approved in the UK at the time of printing, is sunitinib (Sutent). This drug blocks the signalling pathway in a different place from imatinib, and it also helps to stop new blood vessels from growing. This is a second factor in stopping the GIST from growing, but because it is a general effect, rather than just affecting the GIST cells, sunitinib has more side effects than imatinib in most people.

   Before you are given sunitinib, you should be given the patient booklet published by Pfizer. This gives you hints about how to prepare, how to take the pills and what to do about any side effects you may have. If the normal regime doesn’t suit you it may be possible to change the dose or the way you take the drug. Discuss this with your oncologist if you need to.

2. **Increase the dose of imatinib to 800 mgs a day**

   Funding for this is not generally available under the NHS, but some Primary Care Trusts will fund it from the Cancer Drug Fund. If you know you have an exon 9 mutation, this would probably be the best course to take if you can get the drug, since it has been proved to work better than 400 mgs for exon 9 mutation GISTs.

3. **Enter a clinical trial**

   There are often new drugs becoming available which are currently under trial. Ask your oncologist about these and whether you might be eligible for entry into a trial. Trials
usually have very strict entry criteria which depend on what treatments you have already had, so eligibility will not be a foregone conclusion.

4. **Have more surgery**  
This could be open or laparoscopic.

5. **Less common treatments**  
   - **Radio Frequency Ablation**, where the tumour is effectively cooked by focused microwaves. How appropriate this treatment is depends very much on where the tumour or tumours are, and on the risk to neighbouring organs.
   - **Radiotherapy**. It had been thought that radiotherapy does not work for GIST. However there has recently been some experimental use of radiotherapy on GIST patients who have no other treatment left to try. It has been found to be quite successful in shrinking the tumours, so more work needs to be done to see how radiation can best be used for GIST patients. The Cyberknife can irradiate the tumour from many directions by highly focused X-rays. Currently there are very few Cyberknife installations in the world, and the treatment is still experimental.
   - **Embolisation**, where the blood supply to the tumour is choked off, so that the tumour dies.

If you like diagrams, the chart on the next page shows the possible steps in your treatment. This has been adapted from a chart in “Suggested Guidelines for the Management of GIST 2011”. Shaded boxes show treatments that are not yet approved by NICE (in 2012), but which may nevertheless sometimes be available.
MDT discussion

Metastatic or considered inoperable

Biopsy (preferably endoscopic)

Pathology

GIST confirmed

CT scan (for future comparison)

Treat with imatinib

3-monthly CT scans

Continue treatment until progression of tumour or symptoms

Increase dose if appropriate

Consider sunitinib

Consider imatinib for relief of symptoms

Consider patient for inclusion in trials of new drugs or experimental radiotherapy

Single operable tumour

Surgery to remove, possibly after biopsy

Pathology

Not a GIST: Consult appropriate MDT

Pathology

GIST confirmed

Assess risk

Surgery if GIST becomes operable

Follow-up

High risk
Consider adjuvant imatinib

No

Yes

Low risk
CT at 3 months + clinical follow-up

Very low risk
Primary care follow-up

CT every 3 months

for 2 years, then 6 monthly for 2 years, then annually

Recurrence:
MDT to assess tumour and start imatinib, or carry our further surgery

Adjuvant imatinib
either for 1 year or for 3 years, depending on local funding. CT every 3 months.

Consider imatinib for relief of symptoms

GIST confirmed

Treat with imatinib

3-monthly CT scans

Continue treatment until progression of tumour or symptoms

Increase dose if appropriate

Consider sunitinib

Consider patient for inclusion in trials of new drugs or experimental radiotherapy

GIST confirmed
How targeted drugs work.

At the time of writing this booklet, (2012), the first and second line targeted drugs available in the UK are imatinib (Glivec®) and sunitinib (Sutent®). Clinical trials are being performed across the world investigating other new targeted drugs.

Tumours grow when a cell starts to grow in an uncontrolled way. It does this because there has been a change in the genes in the cell. We do not know why this happens in GIST. The genetic change means that the very complex system, which should tell the cell when to grow and when not to grow, stops working. The cell just goes on dividing, and a tumour grows. What the targeted drugs do is to block one of the essential steps in the growth-signalling system in these particular cells, without affecting the normal cells of the body.

Imatinib was the first targeted drug developed for GIST, and it is still the first choice because it is usually easy for patients to take and gives few side effects. Unfortunately, we now know that GIST cells quite often mutate again, and find a way round the block in the signalling pathway. This means that the tumour can start to grow again. In some patients this happens in a few months, but we know of some patients who

I must be worth my weight in gold! I have been taking a double dose of Glivec since joining the Europe-wide trial in 2001. And I am still alive and well.

David Cook (Founder of GSUK)
have been taking imatinib for 11 years, and it is still working. The average time is about three years. Why there is this difference is a question we cannot yet answer. Another question we don’t know all of the answer to, is why there is a small percentage of patients who do not respond to imatinib at all. We know that Wild type and paediatric GISTs respond less well than other GISTs to imatinib, but there is much more we need to know about this.

**Side effects**

If you read the leaflet which is enclosed with the pills of either imatinib or sunitinib, you could start feeling ill – before you have even taken a single pill! However, remember that the company which makes the pills has to mention every side effect ever known, and most of them are very rare. Some are fairly common, but most of these, at least for imatinib, are quite easy to cope with (see table on page 19). With both drugs, some patients have virtually no side effects, and many people find that the side effects they have at first get less of a problem after time.

**Nutrition**

Taking targeted medicines may cause a temporary loss of taste at first. Food and drink suddenly taste different. This does usually get better in time. In the meantime, try to keep up a healthy weight by eating all kinds of good food, and try to avoid eating a high-fat diet or too much sugar. Eat plenty of fresh fruit and vegetables.

If you feel sick, it may be helpful to eat small amounts of food and keep the fluids up well. If this doesn't help, your doctor may prescribe you a medicine to tackle the nausea.
**The commonest side effects of imatinib noticed by patients** | **Patients’ suggestions for dealing with them**
---|---
Puffy eyelids | Put up with it: we all have this!
Swollen ankles | Put your feet up when you can
Indigestion or feeling sick | Take your pills with your main meal and with a large glass of water. Try peppermints, or if they don’t work try Gaviscon.
Being sick | Ask your GP for help. There are various medicines which can help
Diarrhoea or abdominal pain | Ask your GP for help.
Rash | Ask your GP for help
Cramp, particularly at night. | Make sure your blood chemistry is fine. If it is, some patients find that quinine helps. Tonic Water contains small amounts of quinine, but if this is not enough, ask your GP.
Tiredness | The only answer is to get more rest.

**Note:** Grapefruit (juice) in combination with imatinib (Glivec®), and with sunitinib (Sutent®), is strongly discouraged, as grapefruit will interfere with the working of these drugs.
There are other side effects that may show up when you have your blood tests at your hospital checks, and the hospital doctors will advise you about these. You will also have regular CT scans, usually every three months, to check that the imatinib is working.

There are a few very rare and possibly serious side effects. These include, chest pain, sudden bleeding in the stomach or intestine producing black stools, sudden high fever or very sore throat, rapid swelling and weight gain, and sudden shortness of breath. If you have any of these, go to your nearest A&E department, and make sure the doctors there know that you are a GIST patient and taking imatinib.

If you are one of the very few unlucky people in whom the side effects become bad, and nothing seems to help, it may be sensible to lower the dose. This is something you must talk to your GIST specialist about. (We do know of a patient who remained stable on only 100mg imatinib per day.) It is your life, and only you can decide when something is too bad to live with.

There are some drugs which interact with imatinib, and should not be taken. GIST Support UK publishes a little pocket booklet called “I am a GIST Patient”, which lists the drugs you need to avoid, and in which you can record the details of your own treatment. If you need medicines for something other than GIST, you can always show your pocket booklet to your GP or to the hospital where you are being treated. Always carry the booklet with you. You should also remember that you must avoid some foods, and over-the-counter remedies, like grapefruit and St John’s Wort. It is a good idea to keep a list of everything you
are taking, even vitamins, and show it to your hospital doctor.

**Guidelines for treating GIST**

Teams of doctors have prepared guidelines for the best way of treating GIST. They have looked at all the known evidence from clinical trials. For example, there are the European ESMO guidelines, which are brought up to date as new evidence becomes available. There is also a version of this for use in the UK which takes into account the UK health system and the treatments permitted by NICE*. These Guidelines also recommend the best follow-up regimen for patients with different risks of recurrence. See diagram on page 16

Sometimes, there is no clear answer as to how you should be treated in a particular situation. The way forward should then be a matter for you and your close family to discuss with the oncologist and surgeon looking after you.

First try to establish that the hospital where you are being treated is a specialist GIST centre (see list on page 27). You have the right to be referred elsewhere if you are doubtful about the experience of the clinicians treating you.

---

* NICE: National Institute for Clinical Excellence. This is the government body responsible for deciding which treatments can and cannot be paid for under the NHS in England, Wales and Northern Ireland. In Scotland the Scottish Medical Consortium has this role.
Compliance
This just means doing what your doctor has told you to do! In other words, take the pills. It is important for your care that you do your very best to remember to take them. If you don’t then the tumour cells have a chance to start growing again. If you do miss a dose or two, do tell your doctor. Your care is a joint venture and you do need to have a doctor you trust, and he can only do his best for you if you are honest with him.

Preparing for your next visit to the oncologist
Make a list of all the medications you use, whether regularly or once in a while. Talk about any drugs and vitamins you take, both from the chemist's and from the supermarket. The best thing is to ask your oncologist or your family doctor what you are allowed to use.

Make written notes of all the questions you want to ask. It is very easy to forget something until you are on the way home. The time with your consultant goes very quickly, but the doctor will be happy to answer your questions.

It’s a very good idea to take a family member or a friend with you. They will often remember things from the discussion that you don’t.

Questions you might like to ask your oncologist about your treatment:

1. Where is the primary tumour?
2. Are there any metastases (secondary tumours)?
3. Do you know the mutation (Kit exon, PDGFRA exon, Wild-type etc)?
4. What treatment are you proposing?
5. What are the possible side-effects and risks?
6. What is the chance that the tumour will come back and that there will be metastases
7. Can I participate in a trial, or are there any new approaches to treatment available?
8. How many GIST patients do you treat?
9. I would like my tissue to be stored in the National GIST Biobank. Are you able to organise this for me?

**Life with GIST**

Hearing the news that you have a rare cancer, and having to face up to all the treatments and hospital appointments can put a huge strain on the life of a GIST patient. Feelings of panic and fear are almost inevitable. Your family and friends will be scared too. There may well be times when you feel depressed, and this is very understandable. Since it often takes years before a GIST is finally discovered, you may have experienced a long period of unexplained symptoms and the lonely feeling of not being understood.

Talking about your feelings helps. You may find it helpful to tell your story to the hospital oncology nurse, who may refer you to a specialist for counselling.

Or you can go to your GP and ask him for support. In any case, do not keep these feelings to yourself, but get help somehow.

There are no regional GIST groups in the UK, but there are other more general cancer support groups available locally. Some towns have Sarcoma groups, and GIST is a sarcoma, so these groups would be open to you. There may be Macmillan cancer support facilities near you, or a Maggie’s
Centre. These provide general information about living with cancer, eating well, and offer complementary therapies which may make you feel better. They also offer a good cup of tea and understanding friendship.

**Finding other GIST patients**
Because GIST is so rare, you are very unlikely to find another GIST patient by chance. You can find out more about GIST by going to our web site [www.gistsupportuk.com](http://www.gistsupportuk.com), and if you want to, you can make contact with this group. Then you will be invited to the regular patient support meetings. You will also be invited to join our Listserve email group. This group is only for patients and their carers. You may just want to read the postings from others, or you may want to contribute with your story, or to ask questions about other patients’ experiences or your own treatment. How you use the group is up to you. Many patients get huge support from this new group of friends, who really do understand what you are talking about.
About GIST Support UK

GIST Support UK has been a registered charity since 2008 and is run by a team of Trustees. Our primary aim is to offer many kinds of support to patients and their carers.

We have:

- a web site [www.gistsupportuk.com](http://www.gistsupportuk.com)
- a phone help line 0300 4000000
- a private Listserv group for patients and carers
- two meetings a year for patients and carers

We can give you:

- a simple patient leaflet which is sent to hospitals treating GIST patients
- a pocket-sized booklet for patients “I am a GIST patient”
- a booklet “Living after GI surgery for GIST”
- a booklet “Help and Tips for Patients After Removal of the Stomach (Gastrectomy)”
- GIST ribbons so that GIST patients can recognise each other (eg at a hospital clinic)
- A poster for your hospital to display
- More copies of this booklet for your GP or family

Our second aim is to improve the treatment of GIST, hoping eventually for a real cure. In order to do this we:

- Encourage patients to get their treatment at the hands of doctors who are GIST experts.
• Work with the pharmaceutical industry to support research and good patient information about their drugs.
• Encourage and support clinical trials and other research (see form on page 30)
• Try to keep informed about results of research being done across the world
• Represent patients’ interests at NICE appraisals of new drugs and other treatments

**PAWS-GIST Group**

GSUK has also set up a sub-group (“PAWS-GIST”) with a special interest in **Paediatric, Adolescent, Wild-type** and **Syndromic GISTs**. This group concentrates on raising awareness, and on raising funds for research and to improve treatments for this particular group of patients in the UK. We are working with medical experts to establish a PAWS-GIST clinic in the UK to complement the work currently being undertaken in America and Dublin. Follow our progress via Facebook at [http://www.facebook.com/pawsgist](http://www.facebook.com/pawsgist), via Twitter at [http://twitter.com/#!/pawsgist](http://twitter.com/#!/pawsgist) or via our website [www.gistsupportuk.com](http://www.gistsupportuk.com)

**Funding**

Initially GSUK was supported by funding from Novartis. More recently we have been supported by unrestricted educational grants from Novartis, and specific grants from Pfizer. These were principally in order to help finance our twice-yearly patient meetings and information materials for patients. In spite of these sources of funding, for which we continue to be very grateful, GSUK has always insisted on maintaining its
independence from the pharmaceutical industry, and we are now engaged in substantial fund raising of our own. As well as paying for our core operation we are now able to finance some research.

Donations can be sent to: The Treasurer GSUK, 15 Somersby Avenue, Walton, Chesterfield, Derbyshire S42 7LY. You can give on line via Just Giving or Virgin Giving, and also on a mobile phone by texting "PAWS01 £xxxxx" to 70070. See our website for details. We have tee-shirts and similar items for use at fund-raising events.

Specialist GIST centres in the UK
These are the ones we know of in 2012:

City Hospital, Belfast  
Queen Elizabeth Medical Centre, Birmingham  
Royal Infirmary, Bristol  
Addenbrooke’s Hospital, Cambridge  
University Hospital of Wales, Cardiff  
The Western Infirmary, Glasgow  
St James’s Hospital, Leeds  
The Royal Marsden, London  
University College Hospital, London  
The Christie Hospital, Manchester  
The Radcliffe Infirmary, Oxford  
The Northern Centre for Cancer Care, Newcastle  
Weston Park Hospital, Sheffield
Speakers at recent GSUK Patient Meetings

Oncologists:
Dr Ramesh Bulusu  Consultant Clinical Oncologist, Addenbrookes Hospital
Dr Ian Geh  Consultant Clinical Oncologist, Queen Elizabeth Hospital, Birmingham
Prof Bass Hassan  Professor of Medical Oncology, University of Oxford
Prof Ian Judson  Professor of Cancer Pharmacology, The Royal Marsden
Dr Mike Leahy  Consultant Medical Oncologist, Christie Hospital, Manchester
Dr Michelle Scurr  Consultant Oncologist, The Royal Marsden (Now living in Australia)
Dr Beatrice Seddon  Consultant Clinical Oncologist, University College Hospital, London
Dr Mark Verrill  Consultant Medical Oncologist, Northern Centre for Cancer Care, Newcastle
Prof Penella Woll  Sarcoma Specialist Medical Oncologist, Sheffield

Surgeons:
Mr Evangelos Efthimiou  Consultant Surgeon, The Royal Marsden
Mr Long R Jiao  Consultant Surgeon, Hammersmith Hospital
Mr Satvinder Mudan  Consultant Surgeon, The Royal Marsden
Mr Simon Wood  Consultant Surgeon, Royal Gwent Hospital

Other experts:
Dr Zahir Amin  Consultant Radiologist, University College Hospital
Ms Jane Ballantyne  Macmillan Benefits Advisor
Dr Dominic Bray  Clinical Psychologist, Southport & Aintree Hospitals
Mr Nick Duncan  Principal Pharmacist, Queen Elizabeth Hospital Birmingham
Prof Andy Hall  Director, Northern Institute for Cancer Research
Dr David Hughes  Consultant Histopathologist, Sheffield Teaching Hospitals
Dr Bill Newman  Consultant Clinical Geneticist, Manchester University
Ms Sarah Newton  Senior Dietician, The Royal Marsden
Mrs Cerys Propert-Lewis  Clinical Nurse Specialist, The Royal Marsden
Dr Peter Stephens  Clinical Research Associate, Newcastle Cancer Centre
Dr Philippe Taniere  Consultant Histopathologist, Queen Elizabeth Hospital, Birmingham
Mr Roger Wilson CBE  Founder of Sarcoma UK and expert patient advocate
Useful Links

Sarcoma UK
An organisation dedicated to support and research on all kinds of sarcoma
www.sarcoma-uk.org

UK Clinical Trials
The NHS site giving details of UK trials
http://www.nhs.uk/Conditions/Clinical-trials/Pages/Introduction.aspx

Macmillan Cancer Support
Macmillan has many leaflets on all aspects of cancer including information on statutory UK benefits
www.macmillan.org.uk

Pediatric & Wild-Type GIST Clinic USA
http://www.pediatricgist.org

Maggie’s Centres
These provide emotional and practical support for all cancer patients
http://www.maggiescentres.org

Carney’s Triad Group
A site for Carney’s Triad patients (in Dutch and English)
www.carneytriad.com/?lang=en

GIST Support International
An independent international US-based patient group. They have a Listserve group and have produced an excellent leaflet on understanding your pathology results.
www.gistsupport.org

SPAEN (Sarcoma PAtients EuroNet)
European organisation for support and advocacy on behalf of sarcoma patients and their carers. GSUK is a member.
www.sarcoma-patients.eu/index.php

Life Raft Group U.S.A.
A US patient group supported by Novartis
www.liferaftgroup.org
Consent for use of tissue samples

I ............................................................

Date of Birth ...........................................

Address ..................................................

.......................................................

Do give my consent for any tissue samples removed from my body at any time during my life or after my death, to be used for research, including genetic research.

Signed ....................................................

Date ........................................................

In the presence of a witness, who is not a relative or part of the medical team:

Name ....................................................

Signature .................................................

Address ..................................................

......................................................

When you have completed this form and had it witnessed, please ask your oncologist to have it filed with your medical records. It might also be helpful if you were to attach a copy to your Will.

Reg Charity No
1129219