Understanding retroperitoneal sarcoma
About this booklet
This booklet is aimed at anyone who has been diagnosed with a retroperitoneal sarcoma. It explains what retroperitoneal sarcoma 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 healthcare professionals.

You may find it useful to share the information in this booklet with your partner or family members to help them understand about retroperitoneal sarcoma. 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.

This booklet was produced by Sarcoma UK, the only charity in the UK focusing on all types of sarcoma. Our mission is to increase knowledge and awareness of sarcoma through ground-breaking programmes that inspire involvement and transform the landscape for everyone affected by sarcoma. We:

• Raise sarcoma awareness to initiate change and improve standards of treatment and care
• Seek answers through research
• Provide support & information for the sarcoma community

We rely solely on your donations to keep producing these booklets. Any donation is welcome:

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

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

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

Sarcoma is a rare cancer that develops in the connective tissues: muscle, bone, nerves, cartilage, tendons, blood vessels and the fatty and fibrous tissues.

Retroperitoneal sarcomas occur in the retroperitoneum. This is an area behind the peritoneum, the lining of the abdominal space that covers the abdominal organs. The retroperitoneum is deep in the abdomen and pelvis, behind the abdominal lining, where organs such as the major blood vessels, kidneys, pancreas and bladder are located.

The main types of sarcoma that occur in the retroperitoneum are:
- Liposarcoma – cancer of the fatty tissues
- Leiomyosarcoma – cancer of the involuntary muscle
- Other less common types in the retroperitoneum include solitary fibrous tumour, pleomorphic sarcoma, malignant nerve sheath tumour, synovial sarcoma and Ewing’s sarcoma.

What are the signs and symptoms of retroperitoneal sarcoma?

Symptoms of retroperitoneal sarcomas can vary depending on the size and location of the tumour. They may include:
- A noticeable lump in the abdomen
- Increase in abdominal girth
- Dull pain in the abdomen or back
- Intense abdominal pain with bleeding

Other rare symptoms include early satiety (meaning feeling full after eating a small amount of food), weight loss, hernia or anaemia.

How is retroperitoneal sarcoma diagnosed?

A diagnosis of retroperitoneal sarcoma may start with a visit to your GP who will then refer you to a specialist doctor. Some retroperitoneal sarcomas are discovered through investigations for another medical condition or are diagnosed after surgery for a different problem.

Your symptoms will be investigated using a series of tests that may identify sarcoma. Tests may include:
- Physical examination – looking at or feeling any lump
- A scan – taking pictures of the inside of the body using ultrasound, x-ray, CT or MRI
- Biopsy – taking and testing a tissue sample

A clear diagnosis will be made after a pathologist with experience of sarcoma has examined a tissue sample.
Types of diagnostic scans and tests

**Ultrasound**
A scan that uses sound waves to create images from within the body. A scanning microphone is moved over the part of the body that is being scanned, gel is placed on the skin beforehand to help the scan work better.

**CT**
The Computer Tomography (CT) scan takes a number of x-rays to make a 3D image of an affected area. A CT scan is the most helpful scan when diagnosing retroperitoneal sarcoma. A CT scan can determine the size and extent of a retroperitoneal sarcoma and the images can be used to plan surgery. It can also be used to look for any secondary cancers in the lungs.

**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.

Understanding your diagnosis

The diagnosis of a retroperitoneal sarcoma is mostly made from a needle biopsy and analysis of tissue by a pathologist. A diagnosis of retroperitoneal sarcoma should be confirmed by a specialist sarcoma pathologist who will identify the type of sarcoma and grade of the tumour. This will help your sarcoma specialist advise you on the best course of treatment for you.

Identifying the stage and grade of a cancer means your doctor can advise on the best course of treatment for you. It also describes the cancer in a common language which is useful when your doctor is discussing your case with other doctors or healthcare professionals. The stage of cancer is measured by how much it has grown or spread. This can be seen on the results of your scans or tests. The results from a biopsy can tell what grade the cancer is.

**Grading**
- **Low-grade** means the cancer cells are slow-growing, look quite similar to normal cells, are less aggressive and are less likely to spread
- **Intermediate-grade** means the cancer cells are growing slightly faster and look more abnormal
- **High-grade** means the cancer cells are fast growing, look very abnormal, are more aggressive and are more likely to spread

**Staging**
Most sarcomas are staged by looking at:
- **The size and grade of the tumour**
- **Whether the tumour is deep in the body or superficial. This means closer to the surface.**
- **Whether it has spread to another part of the body**

A biopsy may not be required for a retroperitoneal liposarcoma. This type of retroperitoneal sarcoma can sometimes be identified from a CT scan. The CT scan must then be reviewed by a specialist sarcoma radiologist to confirm the diagnosis.
Most retroperitoneal sarcomas are more than 5cm in size and deep in the body. This makes the grade of the cancer a more important factor when finding out the stage of your cancer.

- Stage 1 means the cancer is low grade, small (less than 5cm) and has not spread to other parts of the body
- Stage 2 means the cancer is of any grade, usually larger than stage one but has not spread to other parts of the body
- Stage 3 means a high grade cancer that has not spread to other parts of the body
- Stage 4 means a cancer of any grade or size that has spread to any other part of the body

The European Society of Medical Oncology (ESMO) Guidelines for soft tissue sarcoma recommends that all patients with a suspected retroperitoneal sarcoma should be referred to a high-volume sarcoma centre for confirmation of diagnosis and treatment. This means a centre that sees a large number of retroperitoneal sarcoma patients a year.

Who will treat me?

In England and Wales, the National Institute for Health and Care Excellence (NICE) recommends anyone with a sarcoma should be referred to a sarcoma specialist team for diagnosis and treatment. In Northern Ireland, retroperitoneal sarcoma is diagnosed and treated across a number of hospitals in Belfast. People in Scotland should be referred to the Scottish Sarcoma Service for treatment. 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 sarcoma clinical nurse specialist, surgeon and other healthcare professionals involved in your care. See page 14 for more details.

What treatment is available?

The treatment for retroperitoneal sarcoma is highly individualised and depends on the size and location of your tumour. The main types of treatment are:

**Surgery**

Surgery is the main treatment for this type of sarcoma. The surgeon will remove the tumour and will aim to take out an area of normal tissue around it too; this is known as taking a margin. As retroperitoneal sarcomas can sometimes touch or press on surrounding organs, the surgeon will aim to remove the tumour along with any organs next to it ‘en bloc’ meaning, as a whole. This is to ensure they remove all cancer cells including those that are not clearly visible.

Depending on the size and location of your tumour, the surgery you have may take away part of or whole organs such as the kidney, colon (bowel), pancreas, spleen or bladder which can have implications on your quality of life. Your surgeon will explain to you if this is required and the implications of removing the organ attached or

A colostomy is formed during surgery to divert a section of the large intestine (colon) through an opening in the abdomen (tummy). The opening is known as a stoma. A pouch is placed over the stoma to collect waste products that would usually pass through the colon and out of the body through the anus (back passage).
next to your tumour. A small number of people will need a colostomy or urostomy following surgery.

The treatment you receive should take into account your individual situation and your doctor or clinical nurse specialist should talk you through all possible treatment alternatives. You should also have the opportunity to ask any questions you may have regarding the future implications of your surgery.

**Radiotherapy**

This treatment uses high-energy radiation beams to destroy cancer cells.

In some cases, radiotherapy to the tumour can be used before surgery to reduce the chance of the cancer returning. It can also be used after surgery to kill off any local cancer cells. It is not always possible to use radiotherapy to treat retroperitoneal sarcoma. If the tumour is located near to an organ and there is a risk that the organ will be damaged by the radiation, then this treatment is not used.

**Chemotherapy**

This treatment uses anti-cancer drugs to destroy cancer cells. In some sarcomas which are sensitive to chemotherapy (Ewing’s sarcoma, synovial sarcoma and rhabdomyosarcoma) it is used to try and shrink a large tumour so it is safe to be operated on. Sometimes the tumour completely disappears when treated by chemotherapy. In this case, radiotherapy is used to kill off any local cancer cells that remain in the area of the tumour.

Chemotherapy is sometimes used in the treatment of leiomyosarcoma after surgery to prevent the cancer coming back. Chemotherapy is also used to treat leiomyosarcomas that have spread to other parts of the body.

**Liposarcoma** is not sensitive to chemotherapy and therefore it is not used to treat retroperitoneal sarcomas of this sub-type.

**Surgery to remove a retroperitoneal sarcoma may not always be possible. The location of the tumour and the state of your health may make major surgery too risky for your survival. If this is the case then there are other options available. These include, radiotherapy without surgery, and observation. You may also be offered best supportive care. This is care that aims to help you live as well as possible with the effects of sarcoma. It could include social, physiological or spiritual support. As well as access to dietary services, physiotherapy and occupational therapy.**

**Clinical trials**

You may be offered an opportunity to take part in a study to investigate new diagnosis methods, drugs or 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.

**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 MDT at sarcoma.org.uk.
Will my cancer come back?

Retroperitoneal sarcoma can reappear in the same area after treatment for a previous tumour. This is called a local recurrence. Retroperitoneal sarcoma can come back near the site of your first tumour or spread through to the abdomen. If the sarcoma does reappear, it is important to get treated as quickly as possible. It is useful to check for recurrences yourself through self-examination; your doctor or sarcoma clinical nurse specialist can tell you what to look for.

If you are worried about your sarcoma returning contact your doctor or nurse, they may decide to bring forward the date of your follow-up appointment to investigate your concerns.

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

Sometimes a recurrence of sarcoma may be accompanied by cancer spread to other parts of the body. This is called a metastasis or secondary cancer. In retroperitoneal sarcoma patients, secondary cancer may appear in the lungs, which is why a chest x-ray is taken at follow-up sessions.

Treatment for secondary cancer may involve surgery, radiotherapy or chemotherapy as appropriate; your treatment will be assessed on an individual basis.

What support is available?

Emotional support

Talking about it

A diagnosis of retroperitoneal sarcoma 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.

Talking to others affected by retroperitoneal sarcoma

If you do not feel comfortable discussing your concerns with family or friends, you may find it useful to talk to other people who have been affected by retroperitoneal sarcoma. There is an online support group for people affected by sarcoma, hosted by GroupSpaces.com. It 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.

groupspaces.com/sarcoma is owned and run by a group of dedicated volunteers, who greet new members and monitor activity on the site. Sarcoma UK offers support to members by providing useful sarcoma information.

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 specialist or a presentation from a patient.
invited speaker on a related topic, discussion and questions, informal chat with other group members, and refreshments.

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 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.

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.

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.

<table>
<thead>
<tr>
<th>Key Worker/Clinical Nurse Specialist (CNS)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Offers support and advice, and acts as an important point of contact for patients when they have a concern.</td>
</tr>
<tr>
<td>Name</td>
</tr>
<tr>
<td>Direct telephone</td>
</tr>
<tr>
<td>Notes</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Oncologist</th>
</tr>
</thead>
<tbody>
<tr>
<td>A doctor who specialises in cancer treatments other than surgery, for example chemotherapy or radiotherapy.</td>
</tr>
<tr>
<td>Name</td>
</tr>
<tr>
<td>Direct telephone</td>
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<tr>
<td>Notes</td>
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<table>
<thead>
<tr>
<th>Surgeon</th>
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</thead>
<tbody>
<tr>
<td>Treats cancer through the removal of tumours.</td>
</tr>
<tr>
<td>Name</td>
</tr>
<tr>
<td>Direct telephone</td>
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<tr>
<td>Notes</td>
</tr>
</tbody>
</table>
Physiotherapist
Advises on exercises to help with rehabilitation before, during and after treatment with surgery, radiotherapy and chemotherapy.

Name

Direct telephone Email

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

Pathologist
A doctor who identifies diseases by studying tissue samples.

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.

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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.

<table>
<thead>
<tr>
<th>Name</th>
<th>Contact details and notes</th>
</tr>
</thead>
</table>

**Appointment diary**

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

<table>
<thead>
<tr>
<th>Date</th>
<th>Hospital</th>
<th>Appointment/Treatment</th>
</tr>
</thead>
</table>
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.

References to the source of information used to write this booklet and an acknowledgement of the members of the Information Review Panel who reviewed the booklet are available from Sarcoma UK – info@sarcoma.org.uk

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.
Awareness • Research
Support & Information

sarcoma.org.uk
020 7250 8271
info@sarcoma.org.uk
@Sarcoma_UK
uk.sarcoma
49-51 East Road, London N1 6AH

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