



# Understanding retroperitoneal sarcoma

[sarcoma.org.uk](http://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 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 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 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.**

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# What is retroperitoneal sarcoma?

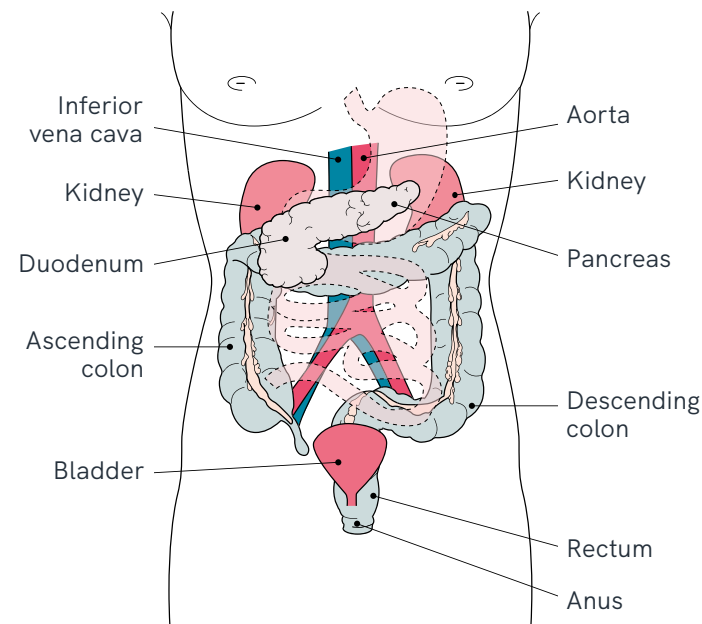
What are the different types of retroperitoneal sarcoma?

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. The retroperitoneum is deep in the abdomen (tummy) 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.



## Signs and symptoms

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.

*"The retroperitoneum is deep in the abdomen and pelvis, behind the abdominal lining."*

# How is retroperitoneal sarcoma diagnosed?

*“Some retroperitoneal sarcomas are discovered through investigations for another medical condition.”*

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
- A core needle biopsy - taking or testing a tissue sample. Core needle biopsies use a core (meaning hollow) needle to remove the tissue.

A clear diagnosis will be made after a pathologist with experience of sarcoma has examined a tissue sample. The biopsy can also help to tell the difference between a sarcoma and other conditions occurring in the abdomen. It can also help to decide the correct treatment for other retroperitoneal conditions.

# What tests or scans might I have?

## 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. In the majority of cases your ultrasound will be carried out by a specialist musculoskeletal radiologist.

## X-ray

Uses x-radiation to take images of the dense tissues inside the body such as bones and tumours. An x-ray is sometimes used to check for tumours in other parts of the body particularly in the lungs.

## 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.

## Blood test

Laboratory analysis of a blood sample.

*“CT images can be used to plan surgery.”*

**“A biopsy may not always 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.”**

*“An ultrasound CT scan is used to help your clinician find the best place to take the sample.”*

#### Biopsy

Taking and testing a sample of tissue from the tumour. For RPS a core needle biopsy is used. This is usually carried out by a specialist sarcoma team under local anaesthetic. An ultrasound CT scan is used to help your clinician find the best place to take the sample.

#### Histopathology

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

# Understanding your diagnosis

The diagnosis of a retroperitoneal sarcoma is mostly made from a needle biopsy and analysis of tissue by a pathologist in combination with the CT scan appearance. 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.

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



What grade is my tumour?

*“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.”*

## 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

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

# 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.

People with a retroperitoneal sarcoma should be referred to a sarcoma centre with special expertise in managing this type of tumour. Treating retroperitoneal sarcomas has particular challenges, and experience of treating this tumour type will vary between sarcoma treatment centres. Outcomes for people with retroperitoneal sarcoma will be improved by concentrating their treatment at centres with staff who are experienced in managing this type of tumour .

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 21 for more details.

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. **Contact Sarcoma UK for more information on high-volume centres.**



Who is my sarcoma clinical nurse specialist?

*Find out more about your sarcoma MDT and specialist centres at [sarcoma.org.uk](http://sarcoma.org.uk)*

# 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 option for this type of sarcoma. The surgeon will remove the tumour and will aim to take out an area of normal tissue too when possible. This is known as taking a margin. Retroperitoneal sarcomas can sometimes touch or press on surrounding organs. In these cases the surgeon will aim to remove the tumour along with any organs next to it 'en bloc' meaning as a whole.

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 next to your tumour. A small number of people will need a colostomy or urostomy following surgery. In cases where it is not possible to remove the tumour completely, surgery is unlikely to be recommended.

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.

Surgery for retroperitoneal sarcoma can be complex and only a small number of surgeons in the UK are specialists in this type of surgery. Your surgeon should also be a core or extended member of your sarcoma MDT. The quality of surgery received is critical to a patient's outcome so it is important that the surgeon operating on you has sufficient expertise in operating on sarcomas that occur in the retroperitoneum. Surgeons with this expertise include gastrointestinal surgeons.

Rarely 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).

If you have had your bladder removed you will need a urostomy. A urostomy is formed during surgery to allow drainage of urine 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 bladder and urethra. The potential need for a colostomy or urostomy should be discussed with your surgeon.

## Radiotherapy

This treatment uses high-energy radiation beams to destroy cancer cells. It is not routinely used to treat RPS. In some cases, radiotherapy can be used before surgery to treat the tumour when it is close to vital structures, such as the organs in the retroperitoneum. It can also be used after surgery to kill any local cancer cells. If the tumour is located near to an organ and there is a risk that



What are the likely side effects of my surgery?

*"Surgery may not always be possible due to where the tumour is situated."*

*"Radiotherapy can also be used after surgery to kill off any local cancer cells."*



the organ will be damaged by radiation, then this treatment will not be used.

### Chemotherapy

This treatment uses anti-cancer drugs to destroy cancer cells. Different sarcoma sub-types respond in different ways to chemotherapy. The use of chemotherapy as a treatment for RPS is dependent on the types of cells that make up the tumour.

In some sub-types 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 after surgery to prevent the cancer coming back. Chemotherapy is also used to treat sarcomas that have spread to other parts of the body.

Discuss chemotherapy treatment with your specialist team who will be able to give you specific information about your sub-type.

### 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.

Find out more about clinical trials on our website.

# 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 clinical nurse specialist. The usual practice will include:

- A chance to discuss symptoms
- A physical examination and CT, ultrasound or MRI to look for any signs of the sarcoma returning.

# Will my cancer come back?

Retroperitoneal sarcoma can reappear in the same area after treatment. 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 guidance on further management from the specialist sarcoma centre. Your further treatment will be assessed on an individual basis.

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

*"If the sarcoma does reappear, it is important to get guidance on further management from the specialist sarcoma centre."*



There is information on accessing dietary services at [sarcoma.org.uk/rehabilitation](http://sarcoma.org.uk/rehabilitation)

returning, contact your doctor or nurse, they may decide to bring forward the date of your follow-up appointment to investigate your concerns.

You may experience side effects from your treatment. If you have had major abdominal surgery it may affect how you eat. Specialist dietitians can give you advice and support on making changes to your diet.

## What if my cancer spreads?

Sometimes sarcoma can 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.

*"If the sarcoma does reappear, it is important to get treated as quickly as possible."*

## What support is available?

### Emotional support

#### 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

0808 801 0401

[supportline@sarcoma.org.uk](mailto:supportline@sarcoma.org.uk)

#### 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.

#### Talking to others affected by sarcoma

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 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



Is there a support group in my area?

*"There are a number of sarcoma support groups supported by Sarcoma UK around the country."*

**“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.”**

*“There is also online support available for people affected by all types of sarcoma.”*

specialists or doctors. A typical meeting may include a talk from an invited speaker on a related topic, discussion and questions, informal chats 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.

#### 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.

Find out more about local and online support groups at [sarcoma.org.uk](http://sarcoma.org.uk)

## 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**

### 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**

### 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

### 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.**

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](mailto: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.



**Sarcoma**UK

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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[sarcoma.org.uk](https://www.sarcoma.org.uk)

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