

Understanding desmoid-type fibromatosis



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The bone & soft tissue cancer charity

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About this booklet

This booklet is for anyone affected with desmoid-type fibromatosis (DF). It explains what DF is, how it is diagnosed and the treatment options available to you. It is your personal guide with space available to keep all your diagnosis information and details of your next hospital appointments. You can also include contact details for your doctor, clinical nurse specialist and other healthcare professionals caring for you.

You may find it useful to share the information in this booklet with your partner or family members to help them understand about DF. If you have any questions about anything you read in this booklet please contact Sarcoma UK's Support Line. 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 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 cancer.

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



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Symbols used in this booklet



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

What is desmoid-type fibromatosis (DF)?

Desmoid-type fibromatosis (DF) is sometimes called Desmoid Tumour or aggressive fibromatosis. It is a rare type of benign (non-cancerous) tumour. DF develops from fibroblasts. These are a type of cell that provide cell support for the body's tissues. DF can occur anywhere in the body but it is mostly found in the arms, legs and abdomen (tummy). DF can affect anyone but it mostly affects young adults, especially women.

“DF can affect anyone but it mostly affects young adults, especially women.”

It is difficult to predict how DF will develop. Sometimes they can be slow growing and they have also been known to get smaller without any treatment.

There is a small number of people with DF who have a condition called familial adenomatous polyposis (FAP) and Gardner Syndrome. This is a rare condition that can run in families. People affected by FAP may be diagnosed with DF following routine tests and scans to investigate the progression of their condition, such as a colonoscopy. Usually their DF will be in the abdomen and treatment will be different to those who do not have FAP.

How is DF diagnosed?

People with DF that is not related to having FAP may discover a lump and visit their GP who will refer them to a specialist doctor.

A specialist will do:

- A clinical examination – looking at and feeling the lump
- A scan – taking pictures of the inside of the body using ultrasound, CT or MRI
- A biopsy – taking and testing a tissue sample

Types of diagnostic scans and tests

X-ray

Uses x-radiation to take images of dense tissues inside the body such as bones or tumours.

Ultrasound

A scan that uses soundwaves to create images from within the body.

CT

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

Colonoscopy

This test uses a thin, flexible tube called a colonoscope to look inside the colon.

MRI

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

A biopsy of DF should be done by a specialist sarcoma team who has expertise in diagnosing and treating DF. A diagnosis of DF should be confirmed by an expert soft tissue pathologist.

 Has my DF been confirmed by a soft tissue pathologist?

“A colonoscopy uses a thin, flexible tube called a colonoscope to look inside the colon.”

“The MDTs with the experience to treat DF are usually only found in sarcoma centres.”

Histopathology

Examination of a tissue sample by a pathologist under a microscope to identify disease. The tissue sample can be taken during a biopsy or from a tumour removed during surgery.

Who should treat me?

DF patients are treated under an oncology team. Oncology is the branch of medicine that treats cancer. DF is not a cancer but oncologists (doctors who specialise in the treatment of cancer) have the experience and skills to look after DF patients. Your treatment plan will need to be managed by a team of experts from a wide range of health care professions called a multidisciplinary team (MDT). The MDTs with the experience to treat DF are usually only found in sarcoma centres. Your MDT will include your clinical nurse specialist, surgeon and oncologists with experience of treating DF. See page 12 for more information on your MDT.

What treatment is available?

Treatment for DF depends on the area of the body that is affected, the size of the tumour and how fast it is growing. It also depends on how close your tumour is to important structures in your body like nerves, a major blood vessel or an organ. Your doctor should explain your treatment options to you and why your MDT has made their decisions. DF tumours can be unpredictable, they can stabilise (stop growing) and even regress (shrink). No one knows why this happens. If a DF tumour stabilises or shrinks then you may not need treatment.

In the past, treatment for DF has relied on surgery. Now you are more likely to be under a Watchful Waiting programme. See the box (right) for more details on how experts came to this decision.

Watchful Waiting

Watchful Waiting is usually the first approach to treat people who are newly diagnosed with DF. This is sometimes known as a ‘watch and wait’ policy, ‘active monitoring’ or ‘active observation’. Watchful Waiting ensures effective treatment can be held in reserve for when you need it. You will be constantly monitored to ensure treatment is given at the earliest appropriate time.

People diagnosed with DF are usually on a watch and wait programme for 1 -2 years. You may be on a watch and wait programme for a shorter or longer time depending on your individual circumstances. If your condition stabilises or regresses watch and wait will continue. If your condition progresses then other treatment options will be considered for you.

If you are on a watch and wait programme you may be worried that your DF is not being treated immediately. However, there are advantages to watch and wait programme:

- Treatment options for DF come with side effects. Although these treatments can be very helpful, it is important to use them only if you are having symptoms that affect your quality of life or your tumour has grown greatly.

In 2015 a consensus paper was written with the involvement of doctors and nurses from across Europe who treat DF patients. Previously different countries had treated DF patients in different ways and there is currently little research into what are the best treatment options. The group found that it is best to have understanding of the growth of the tumour before deciding if any treatment is needed. This can be achieved by reviewing a patient with DF under a Watchful Waiting programme.

“People with DF are usually on a watch and wait programme for 1-2 years.”

If you are having symptoms that are affecting your quality of life you should be offered symptom management. Ask your GP for help. They may ask your local pain management team or symptom control team for further advice. You can also speak to your clinical nurse specialist if you are worried about any symptoms you are experiencing.

During pregnancy the rate of growth of the DF may change, sometimes the tumour can grow and then may settle down after pregnancy. If you are considering getting pregnant please speak to your specialist doctor or clinical nurse specialist first.

- Some treatments cannot be given more than once, for example, radiotherapy. Radiotherapy can be helpful to improve your symptoms but it may not shrink the tumour.
- It is possible that having repeated surgery may cause the tumour to be more aggressive in its recurrence and growth.

Monitoring

If you are on a Watchful Waiting Programme you will be monitored regularly by your specialist team. Usually every three months. At each appointment you will be checked for signs that you may need further treatment. This will be done by:

- Giving you a chance to discuss your symptoms
- A clinical examination - looking at and feeling the tumour
- You may also have a scan, such as an ultrasound or MRI

The reasons you may need further treatment could include:

- You are experiencing symptoms that are affecting your quality of life. For example, increasing pain.
- Your tumour is growing quickly
- Your tumour has grown and is getting close to important structures in the body like nerves, a major blood vessel or an organ.

Surgery

It is sometimes possible for your surgeon to remove the tumour. Many patients are keen to have their tumour removed. In the past, surgery used to include taking the tumour out along with a wide area of normal tissue too. This is known as taking a wide margin. Now it is thought that removing a large amount of tissue around the tumour does not always make a difference to whether it will regrow, so surgeons now aim to just remove the tumour.

Sometimes after surgery for DF, the part of the body where the tumour was removed does not work properly or you are left with cosmetic changes to your physical appearance. Surgery also does not guarantee the tumour will not return. This is why, for most DF patients, surgery will only be offered after a period of Watchful Waiting and after their case has been discussed by an MDT.

Chemotherapy

Chemotherapy is a treatment that uses drugs to destroy tumour cells. Chemotherapy can help switch off the tumour and make it dormant. This may help with symptoms such as pain. The tumour may get smaller with chemotherapy treatment or stop growing. Chemotherapy can cause side effects so the decision to use it needs to be balanced with how your symptoms are affecting you and how the tumour is growing.

Tyrosine Kinase Inhibitors

Tyrosine Kinase Inhibitors (TKIs) are a newer type of treatment called a targeted therapy. They work by blocking growth signals inside the tumour cells. The main drug used to

The treatment you receive will be based on your individual circumstances. Your MDT will discuss your case and decide on what treatment is best for you. You may receive any of the following treatments depending on your specific circumstances.

“Chemotherapy can help switch off the tumour and make it dormant.”



Ask your clinical nurse specialist for more information on TKIs

treat DF this way is called Imatinib. Special funding is needed to make imatinib available for DF patients which is not always approved. You can ask your clinical nurse specialist for more information on this type of treatment.

Anti-inflammatory drugs

Nonsteroidal anti-inflammatory drugs (NSAIDs) can be used to treat DF. They have an impact on the nature of the tumour and can reduce any pain or swelling you may be experiencing.

Hormone treatment

Hormones are substances produced naturally in the body that control the activity of cells and organs. Hormonal therapies use drugs to interfere with the way hormones are made or how they work in the body. Hormone therapy can be used to treat some DF patients as it may make the tumour smaller or stop the tumour growing. The main drug used is Tamoxifen, a hormonal therapy drug that is used to treat breast cancer. This is often used with high dose anti-inflammatory drugs.

Radiotherapy

Radiotherapy uses high-energy radiation beams to destroy tumour cells. It can be used after surgery or as a treatment on its own. It is used to improve symptoms and to try and get the tumour to switch itself off and get smaller. DF patients are usually fit and healthy and radiotherapy can have significant long term side effects, so the decision to use radiotherapy needs to be balanced with how significant the symptoms are. It is rarely used to treat children and younger adults.

Isolated Limb Perfusion

Isolated Limb Perfusion (ILP) is a treatment where chemotherapy drugs are given directly into an arm or leg to

treat a tumour. It is used as an alternative to surgery to treat DF in the arm or leg where surgery will cause damage to the arm or leg or make it not work properly. Not all specialist centres offer this treatment. If ILP is considered appropriate for your case, you might need to be referred to a specialist centre with expertise in this procedure.

What happens after my treatment?

After any treatment you receive, you will have regular follow up appointments. These appointments are to check your health and to check the site of your DF. They are also a good opportunity to talk to your doctor or nurse about any concerns you may have.

- A chance to discuss symptoms
- A physical examination to look for any signs of your DF returning
- You may have a scan such an ultrasound or MRI

What if my DF comes back?

Sometimes DF can come back after it is first treated. This is called a recurrence. Sometimes your DF can be stable for a long time and then start growing again. If you do have a recurrence or your DF starts growing again your doctors will discuss your care in an MDT meeting and consider the best treatment options for you.

What support is available?

Sarcoma UK Support Line

Our Support Line is here for everyone affected by sarcoma – we



You may experience side effects from hormone therapy. Ask your clinical nurse specialist about side effects if you are offered this treatment.

support DF patients too as you will be treated under a sarcoma MDT.



supportline@sarcoma.org.uk

- 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

Symptom management

This is care that aims to help you live as well as possible with the effects of DF. It could include social, psychological or spiritual support. It may also include dietary services, physiotherapy and occupational therapy. Your clinical nurse specialist may be able to suggest useful services to you and your GP can help you access local services in your area.

Peer support

There is a UK support page on Facebook for anyone affected by DF called Desmoid United UK. This is a



closed group where DF patients can share their stories and offer and receive support. To join, search for "Desmoid United UK" in the search bar.

Other sources of support

- British Pain Society
Resources for people living with pain
britishpainsociety.org
- NHS Pain Support
Tips and advice on how to manage pain
nhs.uk/Livewell/Pain

? Ask your clinical nurse specialist about what supportive services are available to you

My diagnosis details

Date of diagnosis

Details of diagnosis

Treatment plan

My 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 health 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. An oncologist will usually take the lead in the treatment of people with DF.

Name

Direct telephone

Email

Notes

Surgeon

Treats DF by removing the tumour.

Name

Direct telephone

Email

Notes

Radiologist

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

Notes

Radiographer

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

Notes

Pathologist

A doctor who identifies diseases by studying tissue samples.

Notes

Other professionals

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

Name

Contact details and notes

<i>Name</i>	<i>Contact details and notes</i>

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