



## **DERMATOFIBROSARCOMA PROTUBERANS (DFSP)**

### **What are the aims of this leaflet?**

This leaflet has been written to help you understand more about dermatofibrosarcoma protuberans (DFSP). It tells you about what it is, what causes it, what can be done about it and where you can find out more about it.

### **What is DFSP?**

DFSP is a very rare type of skin cancer. It usually occurs on the trunk, often the chest and shoulders; however, it can also affect the limbs, head and neck and rarely the genitalia. It starts in the deep layer of the skin (the dermis) and can spread to surrounding structures such as fat and muscle. Although it grows very slowly, it can become quite large. It can often be completely removed with a wide margin of normal tissue, or with a specialised form of surgery called Mohs surgery. DFSP can come back even if completely removed by surgery. It is very rare for DFSP to spread to other parts of the body (occurring in approximately 5% of cases).

### **What causes DFSP?**

Trauma or injury to the skin may be partly responsible in some cases. It is not known why or how the tumour occurs.

### **Is DFSP hereditary?**

No. It occurs in people of all races and ages, usually in early adult life.

### **What does DFSP look like?**

DFSP is usually a slow growing, painless thickened lumpy area of skin. It can range from skin coloured to pink/brown and occasionally can have a blue appearance. It can also feel like a soft indented area on the skin which can make diagnosis especially difficult. If left for several years DFSP can grow through the top layer of the skin, producing an ulcer.

### **What are the symptoms of DFSP?**

Most people with DFSP do not have any symptoms. They may notice a thick or discoloured patch of skin, an indented area or a lump increasing in size.

### **How is DFSP diagnosed?**

As DFSP is so rare, the diagnosis is often not suspected and delayed. It may be mistaken for common harmless skin conditions such as cysts, [dermatofibromas](#) or [keloids](#). If DFSP is suspected, a piece of the abnormal skin will be removed under local anaesthetic (a biopsy) and examined under the microscope to make the diagnosis.

### **Can DFSP be cured?**

Yes, DFSP can be cured if completely removed. However, it can sometimes come back. Long term follow-up with regular visits to the dermatology clinic for at least five to ten years is recommended. If DFSP has spread to other parts of the body removal may not be possible and cure is much less likely.

### **How can DFSP be treated?**

DFSP is treated by surgical excision (removal). Treatment plans for people with DFSP are agreed by members of specialist skin cancer multidisciplinary team. There are two main types of surgery:

**Wide excision** – this involves removal of the DFSP with a margin of normal skin around the edges to ensure no abnormal cells are left behind. The wound will then be reconstructed (closed) using the most suitable type of surgery. This may involve a skin flap or graft. This may happen under local or general anaesthetic depending on the size and location of the DFSP, and the preference of the patient and surgeon.

**Mohs micrographic surgery** – this is a specialised surgical technique, performed by a dermatologist, in which the DFSP is surgically removed in stages, usually all on the

same day, under local anaesthetic. The abnormal tissue is removed and examined under the microscope immediately. A dressing is placed over the wound until the results are ready, and if there are any remaining abnormal cells this process is repeated until the DFSP has been completely removed. The wound will then be repaired by either the dermatologist or a plastic surgeon, depending on its size and location. This type of surgery offers a higher cure rate than wide excision.

Very rarely, DFSP may spread to other parts of the body. Further treatment may then be required, such as radiotherapy or chemotherapy. Radiotherapy or medication (see below) can also be used if the DFSP can't be removed completely by surgery.

### **Drug treatment for DFSP**

Imatinib is a chemotherapy medicine used for DFSP that cannot be removed with an operation or has spread within the body (a metastatic DFSP). It is taken as a tablet and most people have very few side effects.

### **What tests do I need?**

Most people only need surgery for DFSP. If there are concerns that it may have spread, you may need X-rays, ultrasound, MRI or CT scans. If any lumps develop in the scar, the lump will need to be removed surgically for examination under the microscope.

### **Self care (what can I do?)**

DFSP can be difficult even for doctors to detect. After having had a DFSP, you should do regular checks of your scar at home; if you have any changes in the scar such as new lumps which don't settle within a week or two, you should contact your primary care physician or dermatologist.

### **Where can I get more information about dermatofibrosarcoma protuberans?**

*Links to patient support groups:*

*Macmillan Cancer Support*

Helpline (for information): **0808 808 00 00**

Website: <https://www.macmillan.org.uk/>

*Web links to detailed leaflets:*

<http://www.pcds.org.uk/clinical-guidance/dermatofibrosarcoma-protuberans>

<http://www.dermnetz.org/lesions/dfsp.html>

<https://www.aad.org/dermatology-a-to-z/diseases-and-treatments/a---d/dermatofibrosarcoma-protuberans/signs-and-symptoms>

<http://emedicine.medscape.com/article/1100203-overview>

For details of source materials used please contact the Clinical Standards Unit ([clinicalstandards@bad.org.uk](mailto:clinicalstandards@bad.org.uk)).

**This leaflet aims to provide accurate information about the subject and is a consensus of the views held by representatives of the British Association of Dermatologists: individual patient circumstances may differ, which might alter both the advice and course of therapy given to you by your doctor.**

*This leaflet has been assessed for readability by the British Association of Dermatologists' Patient Information Lay Review Panel*

**BRITISH ASSOCIATION OF DERMATOLOGISTS  
PATIENT INFORMATION LEAFLET  
PRODUCED MARCH 2012  
UPDATED JUNE 2015, AUGUST 2018  
REVIEW DATE AUGUST 2021**

