

Understanding DFSP

If you have received a diagnosis of dermatofibrosarcoma protuberans (DFSP), you probably have many questions. This sheet can help you learn more about DFSP so you can take an active role in your treatment. Remember that you have reason to feel hopeful. Doctors learn more about DFSP every day.

What is DFSP?

DFSP is a rare type of cancer that starts in the second layer of the skin. It is most often found on the trunk of the body (the chest, abdomen, or back), but it can occur elsewhere. DFSP may be first noticed as a very small bump on the skin. It grows slowly into a mass that bulges outward, or protrudes. Because the top layer of skin stretches so tightly over the mass, the skin may become tender and bleed.

DFSP rarely spreads to other areas of the body. But it may invade deeper tissues under the skin, including the fat, muscle, and bone. DFSP may come back after it is removed by surgery. In rare cases DFSP does spread to other parts of the body. It can be very serious.

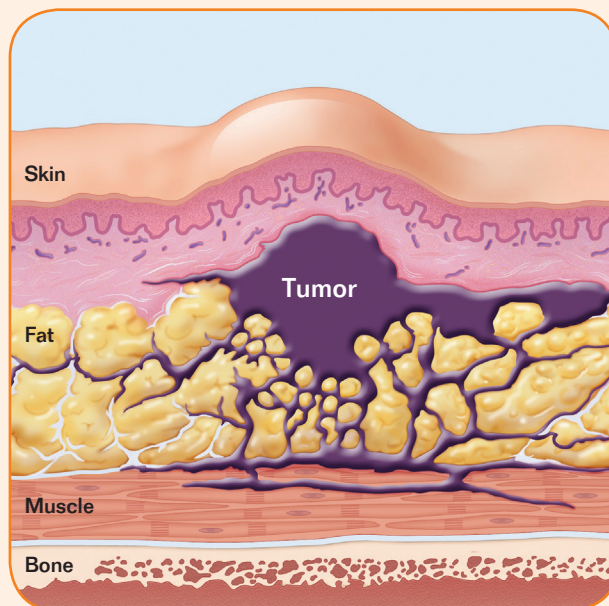
DFSP can occur at any age and in people of all races. It most often affects adults between the ages of 20 and 50 years.

How is DFSP diagnosed?

To diagnose DFSP, the doctor removes a small sample of the mass and sends it to a laboratory to be examined under a microscope. This is called a *biopsy*.

How does DFSP start?

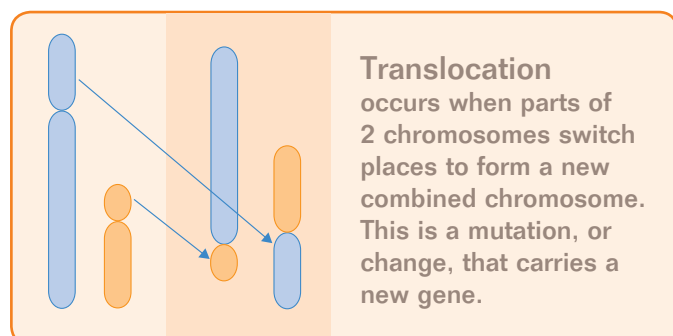
DFSP is believed to start with an immature cell, such as a stem cell. (Stem cells are early cells that mature into different types of blood cells and other cells.) With time, DFSP may spread into surrounding tissue by sending out long, fingerlike projections.



What causes DFSP?

The cause of DFSP is currently not known. It does not appear to run in families. However, researchers have found that abnormal genes and chromosomes may contribute to the development of DFSP. Chromosomes are found inside each cell in the body. They contain genes that tell each cell what to do in the body.

In DFSP, 2 chromosomes switch parts of themselves. This switch, called *translocation*, results in the joining together of 2 genes that were once apart. This combined gene produces an abnormal form of *platelet-derived growth factor (PDGF)*. Normal PDGF activates a specific type of *tyrosine kinase*. Tyrosine kinases are proteins that normally regulate cell operations but, in DFSP, the abnormal PDGF triggers overactivity of DFSP cell growth. DFSP then becomes larger and spreads.



Translocation occurs when parts of 2 chromosomes switch places to form a new combined chromosome. This is a mutation, or change, that carries a new gene.

How is DFSP treated?

A few treatments are available for DFSP. You and your doctor can talk about which treatments are best for you. Because DFSP may come back, you should visit your doctor at least every 6 to 12 months for an examination.

Treatment	How it works
Surgery	<p>The main treatment for DFSP is surgery to remove as many cancerous cells as possible</p> <p>One of 2 procedures may be used:</p> <p>Wide excision — Removal of the cancerous cells and a large amount of the tissue surrounding them</p> <p>Mohs micrographic surgery — Removal of the cancerous cells, followed by bit-by-bit removal of the edges of tissue around the area. Each bit of tissue is examined under a microscope to look for signs of cancerous cells, and the removal continues until there is no longer any sign of them</p>
Radiation therapy	<p>The use of high-energy x-rays or other types of radiation to kill cancer cells — sometimes used in addition to surgery to kill any cancerous cells that might be left behind</p>
Molecularly targeted therapy	<p>Targets the source of DFSP by blocking the overactivity of tyrosine kinase resulting from abnormal PDGF, which causes the growth and spread of DFSP cells</p>

Resources for learning more about DFSP

Please contact your national cancer organization for more information about DFSP. Other sources of information are listed below.

■ **American Academy of Dermatology**
USA
1-866-503-7546 (Toll-free)
1-847-240-1280 (International)
www.aad.org

■ **Genetic and Rare Diseases Information Center**
USA
1-888-205-2311 (Toll-free)
1-301-251-4925 (International)
rarediseases.info.nih.gov/GARD

■ **Sarcoma Alliance**
USA
1-415-381-7236
www.sarcomaalliance.org

■ **European Organisation for Rare Diseases (EURORDIS)**
France
33-1-56-53-52-10
www.eurordis.org

■ **Sarcoma Foundation of America**
USA
1-301-253-8687
www.curesarcoma.org