



# Causes, Risk Factors, and Prevention

## Risk Factors

A risk factor is anything that affects your chance of getting a disease such as cancer. Learn more about the risk factors for soft tissue sarcoma.

- [What Are the Risk Factors for Soft Tissue Sarcomas?](#)
- [Do We Know What Causes Soft Tissue Sarcomas?](#)

## Prevention

The only way to prevent some soft tissue sarcomas is to avoid exposure to [risk factors](#) whenever possible. Most sarcomas, however, develop in people with no known risk factors, so there is no way known at this time to prevent most cases. And for people receiving radiation therapy, there is usually little choice.

## What Are the Risk Factors for Soft Tissue Sarcomas?

A *risk factor* is anything that changes your chance of getting a disease like cancer. Different cancers have different risk factors. For example, unprotected exposure to strong sunlight is a risk factor for skin cancer. Smoking is a risk factor for cancers of the lung, and many other cancers. But risk factors don't tell us everything. Having a risk factor, or even several, doesn't mean that you will get the cancer. Also, many people get cancer without having a risk factor.

Scientists have found a few risk factors that make a person more likely to develop soft tissue sarcomas. These include:

- Radiation
- Certain family cancer syndromes
- A damaged lymph system
- Exposure to certain chemicals

Injury and lifestyle factors such as smoking, diet, and exercise are NOT linked to the risk of soft tissue sarcoma.

## Radiation given to treat other cancers

Patients might develop sarcomas from radiation given to treat other cancers, like breast cancer or lymphoma. The sarcoma often starts in the area of the body that had been treated with radiation. The average time between the radiation treatments and the diagnosis of a sarcoma is about 10 years. Radiation exposure accounts for less than 5% of sarcomas.

Radiation therapy techniques have improved steadily over several decades. Treatments now target cancers more precisely, and more is known about selecting radiation doses. These advances are expected to reduce the number of cancers caused by radiation therapy. But because these cancers take so long to develop, the results of these changes may not be seen for a long time. Still, radiation therapy is used only when its benefits (improved survival rate and relief of symptoms) outweigh the risk of cancer and other complications. For more information, see [Second Cancers in Adults](#).

## Family cancer syndromes

Family cancer syndromes are disorders caused by gene defects (mutations) that people are born with (often inherited from a parent) that are linked to a high risk of getting certain cancers. Some family cancer syndromes increase a person's risk of developing soft tissue sarcomas.

### Neurofibromatosis

Neurofibromatosis usually runs in families and is characterized by many benign tumors that form in nerves under the skin and in other parts of the body (neurofibromas). It is also known as *von Recklinghausen disease*. It is caused by a defect (mutation) in a gene called *NF1*. About 5% of people with neurofibromatosis will develop a malignant peripheral nerve sheath tumor in a neurofibroma.

### Gardner syndrome

Gardner syndrome is a disease caused by defects in the *APC* gene. This syndrome is a type of familial adenomatous polyposis, and people with it get many polyps in the colon (and intestines) and have a high risk of getting colon cancer. It also causes problems outside the colon including desmoid tumors (these were discussed in What Is a Soft Tissue Sarcoma?).

## Li-Fraumeni syndrome

Li-Fraumeni syndrome is caused by inherited defects in the *TP53* gene. People affected by this syndrome have a high risk of cancer, such as [breast cancer](#), [brain tumors](#), and sarcomas. People with this syndrome are sensitive to the cancer-causing effects of radiation. If their cancer is treated with radiation, they have a very high chance of developing a new cancer in the part of the body that received the radiation.

## Retinoblastoma

[Retinoblastoma](#) is an eye cancer in children that can be caused by defects in the *RB1* gene. Children with one of these gene defects also have a higher risk of developing bone or soft tissue sarcomas, especially if treated for cancer with radiation.

## Werner syndrome

Werner syndrome is caused by defects in the *RECQL2* gene. Children with this syndrome have problems like those seen in the elderly. These include cataracts, skin changes, and clogged heart arteries (arteriosclerosis) which can lead to heart attacks. They also have an increased risk of cancer, including soft tissue sarcomas.

## Gorlin syndrome

Gorlin syndrome, also called *nevroid basal cell carcinoma syndrome*, is caused by defects in the *PTCH1* gene. People with this syndrome have a high risk of developing many [basal cell skin cancers](#). They also have an increased risk of getting fibrosarcoma and [rhabdomyosarcoma](#).

## Tuberous sclerosis

Tuberous sclerosis can be caused by a defect in the *TSC1* gene. It can also be caused by a defect in another gene: *TSC2*. People with this syndrome often have seizures and learning problems. They get benign tumors in many different organs. They also get kidney problems, often along with a kidney tumor called *angiomyolipoma*. People with

tuberous sclerosis have an increased risk of getting [rhabdomyosarcoma](#).

## Damaged lymph system

Lymph is a clear fluid containing immune system cells that is carried throughout the body by a series of lymph vessels. These vessels connect lymph nodes (small bean-shaped collections of immune system cells). When lymph nodes have been removed by surgery or damaged by radiation therapy, lymph fluid can build up and cause swelling. This is called [lymphedema](#).

Lymphangiosarcoma (a malignant tumor that develops in lymph vessels) is a very rare complication of chronic lymphedema.

## Chemicals

Exposure to vinyl chloride (a chemical used in making plastics) is a risk factor for developing sarcomas of the liver, but it has not been proven to cause soft tissue sarcomas. [Arsenic](#) has also been linked to a type of liver sarcoma but not soft tissue sarcoma. Exposure to dioxin and to herbicides that contain phenoxyacetic acid at high doses (such as might occur in people who work on farms) may also be risk factors, but this is not known for certain. There is no evidence that herbicides (weed killers) or insecticides, at levels encountered by the general public, cause sarcomas.

## Injury

An injury is **not** a risk factor for developing sarcomas. But this issue has caused some confusion in the past. One reason is that an injured area might swell. That swelling could resemble a tumor but is not a true tumor. Also, when you are injured, the pain may draw your attention to the injured area. The area may be examined closely, and x-rays or other imaging studies may be obtained. This can make it more likely that any sarcoma that is present will be discovered, even though it may have been present for some time.

- [References](#)

[See all references for Soft Tissue Sarcoma](#)

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# Do We Know What Causes Soft Tissue Sarcomas?

Scientists still don't know exactly what causes most cases of soft tissue sarcoma, but they have found several risk factors that can make a person more likely to develop these cancers. And research has shown that some of these risk factors affect the DNA of cells in the soft tissues.

Researchers have made great progress in understanding how certain changes in DNA can cause normal cells to become cancerous. Our DNA carries the instructions for nearly everything our cells do. We usually look like our parents because they are the source of our DNA. However, DNA affects more than the way we look.

The DNA is divided into units called *genes*. Genes carry the recipes for making proteins, the molecules that determine all cell functions. Some genes contain instructions for proteins that control when our cells grow and divide.

Certain genes that promote cell division are called *oncogenes*. Others that slow down cell division or cause cells to die at the right time are called *tumor suppressor genes*. Cancers can be caused by DNA *mutations* (defects) that turn on oncogenes or turn off tumor suppressor genes.

Several family cancer syndromes have been found in which inherited DNA mutations cause a very high risk of developing breast, colon, kidney, eye, or other cancers. In some of these, there is also an increased risk of developing soft tissue sarcomas (these were noted in [What Are the Risk Factors for Soft Tissue Sarcomas?](#)) They are caused by defects (mutations) in genes that can be inherited from a parent. These gene defects can be found through testing. For more on this topic, see [Family Cancer Syndromes](#) and [Genetics and Cancer](#).

DNA mutations in soft tissue sarcoma are common. They are usually acquired during life rather than having been inherited before birth. Acquired mutations may result from exposure to radiation or cancer-causing chemicals. In most sarcomas, they occur for no apparent reason.

Researchers still do not know why most soft tissue sarcomas develop in people who

have no apparent risk factors.

- [References](#)

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## Can Soft Tissue Sarcomas Be Prevented?

The only way to prevent some soft tissue sarcomas is to avoid exposure to [risk factors](#) whenever possible. Most sarcomas, however, develop in people with no known risk factors, so there is no way known at this time to prevent most cases. And for people receiving radiation therapy, there is usually little choice.

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