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# About Kaposi Sarcoma

## Overview and Types

If you have been diagnosed with Kaposi sarcoma or are worried about it, you likely have a lot of questions. Learning some basics is a good place to start.

- [What Is Kaposi Sarcoma?](#)

## Research and Statistics

See the estimates for cases of Kaposi sarcoma in the US and what research is currently being done.

- [Key Statistics About Kaposi Sarcoma](#)
- [What's New in Kaposi Sarcoma Research?](#)

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# What Is Kaposi Sarcoma?

Cancer starts when cells in the body begin to grow out of control. Cells in nearly any part of the body can become cancer and can spread. To learn more about how cancers start and spread, see [What Is Cancer?](#)<sup>1</sup>

Kaposi sarcoma (KS) is a cancer that develops from the cells that line lymph or blood vessels. It usually appears as tumors on the skin or on mucosal surfaces such as inside the mouth, but these tumors can also develop in other parts of the body, such as in the

lymph nodes (bean-sized collections of immune cells throughout the body), the lungs, or digestive tract.

The abnormal cells of KS form purple, red, or brown blotches or tumors on the skin. These affected areas are called **lesions**. The skin lesions of KS most often show on the legs or face. They may look bad, but they usually cause no symptoms. Some lesions on the legs or in the groin area may cause the legs and feet to swell painfully.

KS can cause serious problems or even become life threatening when the lesions are in the lungs, liver, or digestive tract. KS in the digestive tract, for example, can cause bleeding, while tumors in the lungs may cause trouble breathing.

## Types of Kaposi sarcoma

There are four different types of KS defined by the different populations it develops in, but the changes within the KS cells are very similar.

### Epidemic (AIDS-associated) Kaposi sarcoma

The most common type of KS in the United States is epidemic or AIDS-associated KS. This type of KS develops in people who are infected with HIV, the virus that causes AIDS.

HIV stands for human immunodeficiency virus. A person infected with HIV (someone who is HIV-positive) does not necessarily have AIDS, but the virus can be present in the body for a long time, often many years, before causing major illness. The disease known as AIDS begins when the virus has seriously damaged a person's immune system, which means they can get certain types of infections (such as Kaposi sarcoma-associated herpesvirus, KSHV) or other medical complications, including KS.

KS is considered an **AIDS defining illness**. This means that when KS occurs in someone infected with HIV, that person officially has AIDS (and is not just HIV-positive).

In the United States, treating HIV infection with highly active antiretroviral therapy (HAART) has resulted in fewer cases of AIDS-associated KS. Still, some patients can develop KS in the first few months of HAART treatment.

For most patients with HIV, HAART can often keep advanced KS from developing. Still, KS can occur in people whose HIV is well controlled with HAART. Even if KS develops, it is still important to continue HAART.

In areas of the world where it is not easy to get HAART, KS in AIDS patients can advance quickly.

### **Classic (Mediterranean) Kaposi sarcoma**

Classic KS occurs mainly in older people of Mediterranean, Eastern European, and Middle Eastern heritage. Classic KS is more common in men than in women. People typically have one or more lesions on the legs, ankles, or the soles of their feet. Compared to other types of KS, the lesions in this type do not grow as quickly, and new lesions do not develop as often. The immune system of people with classic KS is not as weak as it is in those who have epidemic KS, but it may be weaker than normal. Getting older can naturally weaken the immune system a little. When this occurs, people who already have a KSHV (Kaposi sarcoma--associated herpesvirus) infection are more likely to develop KS.

### **Endemic (African) Kaposi sarcoma**

Endemic KS occurs in people living in Equatorial Africa and is sometimes called African KS. Kaposi sarcoma--associated herpesvirus (KSHV) infection is much more common in Africa than in other parts of the world, so the risk of KS is higher. Other factors in Africa that weaken the immune system (such as malaria, other chronic infections, and malnutrition) also probably contribute to the development of KS, since the disease affects a broader group of people that includes children and women. Endemic KS tends to occur in younger people (usually under age 40). Rarely a more aggressive form of endemic KS is seen in children before puberty. This type usually affects lymph nodes and other organs and can progress quickly.

Endemic KS used to be the most common type of KS in Africa. Then, as AIDS became more common in Africa, the epidemic type became more common.

### **Iatrogenic (transplant-related) Kaposi sarcoma**

When KS develops in people whose immune systems have been suppressed after an organ transplant, it is called **iatrogenic**, or **transplant-related KS**. Most transplant patients need to take drugs to keep their immune system from rejecting (attacking) the new organ. But by weakening the body's immune system, these drugs increase the chance that someone infected with KSHV (Kaposi sarcoma--associated herpesvirus) will develop KS. Stopping the immune-suppressing drugs or lowering their dose often makes KS lesions go away or get smaller.

## **Hyperlinks**

1. [www.cancer.org/treatment/understanding-your-diagnosis/what-is-cancer.html](http://www.cancer.org/treatment/understanding-your-diagnosis/what-is-cancer.html)
2. [www.cancer.gov/types/soft-tissue-sarcoma/hp/kaposi-treatment-pdq](http://www.cancer.gov/types/soft-tissue-sarcoma/hp/kaposi-treatment-pdq)
3. [www.nccn.org/professionals/physician\\_gls/pdf/kaposi.pdf](http://www.nccn.org/professionals/physician_gls/pdf/kaposi.pdf)
4. [www.cancer.gov/types/soft-tissue-sarcoma/hp/kaposi-treatment-pdq](http://www.cancer.gov/types/soft-tissue-sarcoma/hp/kaposi-treatment-pdq)
5. [www.nccn.org/professionals/physician\\_gls/pdf/kaposi.pdf](http://www.nccn.org/professionals/physician_gls/pdf/kaposi.pdf)

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## Key Statistics About Kaposi Sarcoma

Before the AIDS epidemic, Kaposi sarcoma (KS) was rare in the United States. At that time, only about 2 new cases of KS were found for every million people in the United

States each year. Most often, the types of KS that occurred were classic and transplant-related.

With the AIDS epidemic, the rate of KS in this country increased more than 20 times — peaking at about 47 cases per million people (per year) in the early 1990s.

With new treatments for HIV and AIDS, KS has become less common in the United States, and it now occurs at a rate of about 6 cases per million people each year. It is still seen most often in people infected with HIV. In the United States, KS is much more common in men than in women, and it is rarely seen in children. It is also more common in African Americans than in White individuals in the United States. Transplant recipients are another group that gets KS. About 1 in 200 transplant patients in the United States gets KS. Most of these people were already infected with Kaposi sarcoma associated herpesvirus (KSHV) before the transplant, but the virus didn't cause problems because their immune system kept it in check. The drugs the transplant patients take to suppress their immune system allow KS to develop.

In areas of the world (such as Africa) where KSHV and HIV infection rates are high, both endemic and epidemic (AIDS-associated) KS are seen, and can occur in men, women, and children.

Visit the [American Cancer Society's Cancer Statistics Center](#)<sup>1</sup> for more key statistics.

## Hyperlinks

1. [cancerstatisticscenter.cancer.org/](https://cancerstatisticscenter.cancer.org/)

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## What's New in Kaposi Sarcoma Research?

A great deal of research is being done to find more effective ways to prevent and treat Kaposi sarcoma (KS).

### Prevention

Probably the most important advance in the prevention of AIDS-related KS has been the development of drugs that help control HIV infection and AIDS. This has reduced the chance of getting KS.

Testing for Kaposi sarcoma--associated herpesvirus (KSHV), the virus that causes KS, could help manage patients at risk for KS, including those who are HIV-positive or those who will be having an organ transplant and will be taking drugs to suppress their immune system.

Newer drugs, such as valganciclovir, used to treat related herpesviruses such as cytomegalovirus (CMV) can also help treat KSHV infections. These drugs stop the KSHV-infected cells from making more of the virus, although they haven't been found to help treat KS once it has developed.

### Treatment

Researchers are studying new and different ways to treat KS.

KS lesions depend on the formation of new blood vessels for their growth. Drugs called **angiogenesis inhibitors**, which block the growth of blood vessels within tumors, may

help treat these lesions. For example, the drug bevacizumab (Avastin<sup>®</sup>) has been shown to cause some KS lesions to shrink or stop growing in a small study. This drug and other angiogenesis inhibitors, such as sirolimus and everolimus, are being studied further.

Drugs called **immunomodulating agents** both boost the immune system and affect blood vessel growth, so these drugs may be helpful against KS. The oldest of these drugs, thalidomide (Thalomid<sup>®</sup>), has been shown to help shrink some KS lesions in early studies, but this drug can have side effects that make it hard to take. Related drugs, such as lenalidomide (Revlimid<sup>®</sup>) and pomalidomide (Pomalyst<sup>®</sup>), which tend to have fewer side effects, are now being studied.

Some other drugs that are already used to treat other cancers are also being studied for use against KS, including bortezomib (Velcade<sup>®</sup>), imatinib (Gleevec<sup>®</sup>), and sorafenib (Nexavar<sup>®</sup>).

Research into HIV vaccines and antiretroviral drugs also may have a great impact on AIDS-related KS.

KSHV also offers a new target for KS drugs and biologic therapy. [Clinical trials](#)<sup>1</sup> are testing whether antiviral drugs that target KSHV may be used for KS.

## Hyperlinks

1. [www.cancer.org/treatment/treatments-and-side-effects/clinical-trials.html](http://www.cancer.org/treatment/treatments-and-side-effects/clinical-trials.html)

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