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.

- What Are the Key Statistics About Kaposi Sarcoma?
- What's New in Kaposi Sarcoma Research and Treatment?

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 other areas of the body. To learn more about how cancers start and spread, see What Is Cancer?

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 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 appear 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**

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

**Epidemic (AIDS-related) Kaposi sarcoma**

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

A person infected with HIV (that is, who is HIV-positive) does not necessarily have AIDS. 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 the immune system, which results in certain types of infections or other medical complications, including KS. When HIV damages the immune system, people who also are infected with a certain virus (the Kaposi sarcoma associated herpesvirus or KSHV) are more likely to develop 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 epidemic KS. Still, some patients develop symptoms of 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 still occur in people whose HIV is well controlled with HAART. Once KS develops it is still important to continue HAART.

In areas of the world where HAART is not easy to obtain, 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.

Patients typically have one or more lesions on the legs, ankles, or the soles of the 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. KSHV (Kaposi sarcoma associated herpesvirus) 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 the 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.

Kaposi sarcoma in HIV negative men who have sex with men
There have been reports of KS developing in men who have sex with men who are not infected with HIV. In this group, the cases of KS are often mild, similar to cases of classic KS.

- References
See all references for Kaposi Sarcoma

What Are the 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. Early in the AIDS epidemic, patients infected with HIV in the United States were estimated to have a 1 in 2 chance of developing KS.

With new treatments for 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 whites 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 these people 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 HIV-associated KS are seen, and KS occurs in men, women, and children.

Visit the American Cancer Society’s Cancer Statistics Center for more key statistics.

- References
  See all references for Kaposi Sarcoma

Last Medical Review: August 8, 2014 Last Revised: February 9, 2016

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What’s New in Kaposi Sarcoma Research and Treatment?

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

Several drugs 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. The drugs seem to lower the risk of developing KS in patients who are at high risk, 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.

Imiquimod (Aldara) is a topical drug that modulates the immune system. It can be applied to the skin to treat certain kinds of warts. There have been reports of this drug helping shrink KS skin lesions, as well.

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®) has been shown to cause some KS lesions to shrink or stop growing in a small study. Further studies of this drug and other angiogenesis inhibitors are now being done.

Boosting the immune system is another promising approach to treating KS. Interferon alfa was used for many years to help treat KS, although its use is limited today because of its side effects. Studies looking at similar drugs, such as interleukin-12 (IL-12), have shown good results in early studies.

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®), 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®) and pomalidomide (Pomalyst®), 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®), imatinib (Gleevec®), and sorafenib (Nexavar®).

Of course, research into HIV vaccines and antiretroviral drugs also may have a great impact on AIDS-related KS. Maraviroc (Selzentry®) is an antiretroviral drug currently being studied for its effect on KS.

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