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Causes, Risk Factors, and Prevention of Castleman Disease

Risk Factors for Castleman Disease

A risk factor is anything that affects your chance of getting a disease.

- [Castleman Disease Risk Factors](#)

Causes of Castleman Disease

Researchers continue to investigate the causes of Castleman disease. Learn more about what is known.

- [What Causes Castleman Disease?](#)

Castleman Disease Risk Factors

A risk factor is anything that might change a person's chance of getting a disease. Some risk factors, like smoking, can be changed. Others, like a person's age or family history, can't be changed. But having a risk factor, or even several, doesn't mean that a person will get the disease. And, many people who get the disease may have few or no known risk factors.

Most patients with Castleman disease (CD) don't have any known risk factors.

The only clear risk factor for CD is infection with [HIV, the virus that causes AIDS](#)¹. The multicentric form of Castleman disease is much more common in people with HIV infection, particularly in those who have developed AIDS. This might be because these people tend to have weakened immune systems, which allows the growth of another virus known as HHV-8 (see [What Causes Castleman Disease?](#)). It's not clear if people who have weakened immune systems for other reasons are also at higher risk.

Hyperlinks

1. www.cancer.org/cancer/cancer-causes/infectious-agents/hiv-infection-aids.html

References

Castleman Disease Collaborative Network. *About Castleman disease*. <http://www.cdcn.org/about-castleman-disease/>. Accessed January 5, 2018.

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Wu D, Lim MS, Jaffe ES. Pathology of Castleman disease. *Hematol Oncol Clin North Am*.2018;32(1):37-52.

See all references for Castleman Disease (www.cancer.org/cancer/castleman-disease/references.html)

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What Causes Castleman Disease?

We do not know exactly what causes Castleman disease (CD). The main feature of CD is an overgrowth of lymphocytes (immune cells) called B cells. The cause of this overgrowth is not known for sure, but it seems to be related to problems with the way a

person's immune system is working. Many people with CD have abnormally high blood levels of certain substances made by immune system cells.

For example, in the multicentric form of CD (MCD), the body often makes too much of a protein called interleukin-6 (IL-6). IL-6 normally helps regulate immune function. Too much IL-6 can cause lymphocytes to grow and divide too quickly. But it's not clear what causes the high levels of IL-6.

One cause seems to be infection with human herpesvirus-8 (HHV-8), also known as Kaposi sarcoma herpesvirus (KSHV) (because it can cause [Kaposi sarcoma](#)¹). HHV-8 is often found in the lymph node cells in people who have MCD, especially those who are HIV positive. HHV-8 can cause infected cells to make a form of IL-6, which could explain how it leads to CD.

Many people are infected with HHV-8, but in people with normal immune systems the virus doesn't seem to cause problems. People infected with HIV, however, often have weakened immune systems, which might allow HHV-8 to grow and cause problems. This could explain why people infected with HIV are more likely get MCD. Still, some people with HIV who develop MCD do not have weakened immune systems, so it's not clear if this is the only reason.

HHV-8 hasn't been found in all cases of MCD. And it's not clear what causes the localized (unicentric) form of CD. Researchers are still looking for the causes of CD in these other cases.

Hyperlinks

1. www.cancer.org/cancer/kaposi-sarcoma.html

References

National Organization for Rare Disorders. *Castleman disease*. Accessed at <https://rarediseases.org/rare-diseases/castlemans-disease/> on January 5, 2018.

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