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About Castleman Disease

Overview and Types

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

- [What Is Castleman Disease?](#)

Key Statistics

Castleman disease is rare. Learn which form tends to be more common.

- [Key Statistics for Castleman Disease](#)
- [What's New in Castleman Disease Research?](#)

What Is Castleman Disease?

Castleman disease (CD) is a rare disease of lymph nodes and related tissues. It was first described by Dr. Benjamin Castleman in the 1950s. It is also known as Castleman's disease, giant lymph node hyperplasia, and angiofollicular lymph node hyperplasia (AFH). CD is not cancer. Instead, it is called a **lymphoproliferative disorder**. This means there is an abnormal overgrowth of cells of the lymph system that is similar in many ways to [lymphomas](#)¹ (cancers of lymph nodes).

Even though CD is not officially a cancer, one form of this disease (known as

multicentric Castleman disease) acts very much like lymphoma. In fact, many people with this disease eventually develop lymphomas. And like lymphoma, CD is often treated with [chemotherapy](#)² or [radiation therapy](#)³. This is why it is included in the American Cancer Society's cancer information. (For information about lymphoma, see [Hodgkin Disease](#)⁴ and [Non-Hodgkin Lymphoma](#)⁵).

About lymph nodes and lymphoid tissue

To understand Castleman disease, it helps to know about the body's lymph system.

Lymphoid tissue, also known as lymphatic tissue, is the main part of the immune system. It is formed by different types of cells that work together to help the body fight infections. The main cells in lymphoid tissue are lymphocytes, a type of white blood cell. There are 2 main types of lymphocytes: B cells and T cells.

Lymphoid tissue is found in many places throughout the body, including:

- **Lymph nodes:** bean-sized collections of lymphocytes found in small groups throughout the body, including inside the chest, abdomen, and pelvis. They can sometimes be felt under the skin in the neck, under the arms, and in the groin.
- **Thymus:** a small organ behind the upper part of the breastbone and in front of the heart. The thymus plays a vital role in development of T cells.
- **Spleen:** an organ under the lower part of the rib cage on the left side of the body. The spleen makes lymphocytes and other immune system cells to help fight infection. It also stores healthy blood cells and helps filter the blood.
- **Tonsils and adenoids:** collections of lymphoid tissue at the back of the throat. They help protect the body against germs that are breathed in or swallowed.
- **Bone marrow:** the soft inner part of certain bones that makes red blood cells, blood platelets, and white blood cells (including lymphocytes).
- **Digestive tract:** the stomach, intestines, and other organs, which also have lymphoid tissue.

Types of Castleman disease

Doctors can group CD in a number of ways, and they are still trying to determine which classification (or combination of them) provides the most helpful information.

CD is classified by on how much of the body it affects. The main forms of CD are called

localized and **multicentric**. They affect people very differently.

Localized (unicentric) Castleman disease

This is the more common type of CD. Localized CD only affects a single group of lymph nodes. It is not widespread. Lymph nodes in the chest or abdomen are affected most often. CD causes these lymph nodes to grow.

Enlarged lymph nodes in the chest can press on the windpipe (trachea) or smaller breathing tubes going into the lungs (bronchi), causing breathing problems. If the enlarged nodes are in the abdomen, the person might have pain, a feeling of fullness, or trouble eating. Sometimes the enlarged nodes are in places such as the neck, groin, or underarm area and are first noticed as a lump under the skin.

People with localized CD are usually cured when the affected lymph nodes are removed with [surgery](#)⁶.

Multicentric Castleman disease

Multicentric Castleman disease (MCD) affects more than one group of lymph nodes. It can also affect other organs containing lymphoid tissue. This form sometimes occurs in people infected with human immunodeficiency virus (HIV), the virus that causes AIDS. Multicentric CD is more serious than the localized type, particularly in people with HIV infection. You can read more about HIV infection in [HIV, AIDS, and Cancer](#)⁷.

People with MCD often have problems such as serious infections, fevers, weight loss, fatigue, night sweats, and nerve damage that can cause weakness and numbness. Blood tests often show too few red blood cells (anemia) and high levels of antibodies in the blood (hypergammaglobulinemia).

MCD can weaken the body's immune system, making it hard to fight infection. Infections in people with MCD can be very serious, even life threatening. MCD also increases the risk of developing [lymphoma](#)⁸, a cancer of lymphoid tissue, which can often be hard to treat.

Microscopic subtypes of CD

Castleman disease can also be classified based on how the lymph node tissue looks under a microscope. These are called **microscopic subtypes**.

- The **hyaline vascular** type is most common. It tends to be localized, in which case

people often have few symptoms and usually have a good outlook, but in rare cases it can be multicentric.

- The **plasma cell** type is more likely to cause symptoms and to be multicentric, but it is sometimes localized.
- The **mixed** subtype shows areas of both hyaline vascular and plasma cell types. It occurs less often.
- The **plasmablastic** type was recognized more recently. Like the plasma cell type, it is usually multicentric, usually causes symptoms, and has a less favorable outlook.

In choosing [treatments](#)⁹, doctors believe that the microscopic type is less important than whether the disease is localized or multicentric.

Subtypes of CD based on viral infections

Infection with certain viruses plays a role in at least some cases of CD.

Multicentric CD is more common in people infected with [HIV](#)¹⁰, the virus that causes AIDS. Doctors sometimes group patients with multicentric CD into those who are infected with HIV (HIV positive) and those who are not infected (HIV negative).

In recent years, it's become clear that another virus, known as human herpesvirus-8 (HHV-8) or Kaposi sarcoma herpesvirus (KSHV), is often found in the lymph node cells of people with multicentric CD. In fact, HHV-8 is found in the lymph nodes of nearly all CD patients who are HIV positive. Some doctors have suggested classifying CD based on whether the cells contain HHV-8.

Hyperlinks

1. www.cancer.org/cancer/lymphoma.html
2. www.cancer.org/cancer/castleman-disease/treating/chemotherapy.html
3. www.cancer.org/cancer/castleman-disease/treating/radiation.html
4. www.cancer.org/cancer/hodgkin-lymphoma.html
5. www.cancer.org/cancer/non-hodgkin-lymphoma.html
6. www.cancer.org/cancer/castleman-disease/treating/surgery.html
7. www.cancer.org/cancer/cancer-causes/infectious-agents/hiv-infection-aids.html
8. www.cancer.org/cancer/lymphoma.html
9. www.cancer.org/cancer/castleman-disease/treating.html
10. www.cancer.org/cancer/cancer-causes/infectious-agents/hiv-infection-aids.html

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

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Key Statistics for Castleman Disease

We aren't sure how many people are diagnosed with Castleman disease (CD) each year. The National Cancer Institute keeps track of how many people have each type of cancer, but because CD is not a cancer it is not included. We do know that CD is rare,

especially in people who are otherwise healthy. Recent studies that looked at medical records of patients with CD suggest there may be about 4,300 to 5,100 new cases of CD per year in the US.

- The localized (unicentric) form of CD is more common than the multicentric form (MCD).
- MCD is much more likely to occur in people infected with HIV. Over the past few decades, as the number of people with HIV infection has increased, the number of people diagnosed with MCD has also gone up.
- Modern anti-viral treatments have helped people with HIV live much longer, but these drugs don't seem to lower the chance of getting MCD.
- CD can affect children as well as adults. Younger people are more likely to have the localized form.
- Older adults and those with HIV infection are more likely to have the multicentric form.

For statistics related to survival, see [Survival Rates for Castleman Disease¹](#).

Hyperlinks

1. www.cancer.org/cancer/castleman-disease/detection-diagnosis-staging/outlook.html

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What's New in Castleman Disease Research?

Important research on Castleman disease (CD) is going on in many university hospitals, medical centers, and other institutions around the world. Scientists are learning more about what causes the disease and how best to treat it. Unfortunately, research into this disease is slowed by the fact that CD is rare.

Causes

Doctors have learned a great deal about some of the possible causes of CD in recent years. For example, many people with multicentric CD (MCD) have been found to have evidence of infection with a virus known as human herpesvirus-8 (HHV-8) in their lymph nodes. Genetic mutations that happen throughout life, and inflammation, are also possible causes of certain kinds of CD. Knowing this type of information might help researchers come up with new ways of treating this disease. But there's a lot we still don't know about the exact causes of CD.

Treatment

Researchers have begun to study several promising new drugs for use against CD in recent years.

Many patients with MCD have too much of a protein called interleukin-6 (IL-6) in their blood. Drugs that target IL-6 have shown promise against MCD. One of these drugs, siltuximab, is a monoclonal antibody that binds to IL-6, which stops it from reaching lymphocytes. This drug is now approved for use in people with MCD who are not infected with HIV or HHV-8 (see [Immunotherapy for Castleman Disease¹](#)).

Another monoclonal antibody, tocilizumab (Actemra[®]), blocks the action of IL-6 by binding to its receptor on lymphocytes. This drug is being studied for use against MCD. It is already approved to treat rheumatoid arthritis in the United States, and some doctors use it to treat MCD as well.

It is unclear how helpful these drugs will be in MCD patients who are HIV-positive, since few HIV-positive patients have been in the studies so far. Still, these drugs offer hope for the future treatment of MCD.

Other drugs being studied for treatment of CD include:

- Sirolimus, cyclosporine, and mycophenolate mofetil: These drugs suppress the immune system, so they are often used in autoimmune diseases or to help prevent the rejection of organ transplants, but they may also be helpful in CD.
- Suramin: This drug is thought to work by stopping IL-6 from attaching to and affecting lymphocytes.
- Bortezomib (Velcade): This drug is used mainly to treat multiple myeloma, but some reports also suggest it might help some people with CD.
- Rituximab (Rituxan): This drug is used to treat certain kinds of lymphoma and may be helpful in CD.
- Silmitasertib or CX-4945: This oral drug blocks CK2, a protein that helps some cells grow and divide. It is still in the earliest phases of testing for CD.

Hyperlinks

1. www.cancer.org/cancer/castleman-disease/treating/immunotherapy.html

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