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Treating Castleman Disease

How is Castleman disease treated?

Several types of treatment can be used to treat Castleman disease, including:

- [Surgery for Castleman Disease](#)
- [Radiation Therapy for Castleman Disease](#)
- [Corticosteroids for Castleman Disease](#)
- [Chemotherapy for Castleman Disease](#)
- [Immunotherapy for Castleman Disease](#)
- [Anti-viral Drugs for Castleman Disease](#)

Common treatment approaches

Your treatment options will be based on whether the CD is localized (unicentric) or multicentric, as well as other factors when these are important. Because CD is rare, it has been hard to do studies to learn the best ways to treat it. Of course, no two patients are exactly alike, so treatment is tailored to each person's situation.

- [Treatment of Localized \(Unicentric\) Castleman Disease](#)
- [Treatment of Multicentric Castleman Disease](#)

Who treats Castleman disease?

Based on your treatment options, you might have different types of doctors on your treatment team, including:

- A **surgeon**
- A **hematologist**: a doctor who treats disorders of the blood and lymph system,

including CD

- A **medical oncologist**: a doctor who treats cancer and similar diseases with medicines
- A **radiation oncologist**: a doctor who treats cancer and similar diseases with radiation therapy

You might have other specialists on your treatment team as well, including physician assistants (PAs), nurse practitioners (NPs), nurses, psychologists, nutrition specialists, social workers, rehabilitation specialists, and other health professionals.

- [Health Professionals Associated With Cancer Care¹](#)

Making treatment decisions

It's important to discuss all of your treatment options, including the goals of treatment and possible side effects, with your doctors to help make the decision that best fits your needs. In choosing a treatment plan, consider your health and the type of CD. Be sure that you understand all the risks and side effects of the various treatments before making a decision. Ask your health care team questions.

CD is a rare disease, so not many doctors have much experience treating it. If time allows, it's often a good idea to seek a second opinion. Getting a second opinion can give you more information and help you feel confident about the treatment plan that you choose. Your doctor should be willing to help you find another doctor who can give you a second opinion.

- [Questions to Ask About Castleman Disease²](#)
- [Seeking a Second Opinion³](#)

Thinking about taking part in a clinical trial

Clinical trials are carefully controlled research studies that are done to get a closer look at promising new treatments or procedures. Clinical trials are one way to get state-of-the-art cancer treatment. In some cases they may be the only way to get access to newer treatments. They are also the best way for doctors to learn better methods to treat illnesses such as Castleman disease. Still, they're not right for everyone.

If you would like to learn more about clinical trials that might be right for you, start by asking your doctor if your clinic or hospital conducts clinical trials.

- [Clinical Trials](#)⁴

Considering complementary and alternative methods

You may hear about alternative or complementary methods that your doctor hasn't mentioned to treat your cancer or relieve symptoms. These methods can include vitamins, herbs, and special diets, or other methods such as acupuncture or massage, to name a few.

Complementary methods refer to treatments that are used along with your regular medical care. Alternative treatments are used instead of a doctor's medical treatment. Although some of these methods might be helpful in relieving symptoms or helping you feel better, many have not been proven to work. Some might even be harmful.

Be sure to talk to your care team about any method you are thinking about using. They can help you learn what is known (or not known) about the method, which can help you make an informed decision.

- [Complementary and Alternative Medicine](#)⁵

Help getting through treatment

Your care team will be your first source of information and support, but there are other resources for help when you need it. Hospital- or clinic-based support services are an important part of your care. These might include nursing or social work services, financial aid, nutritional advice, rehab, or spiritual help.

The American Cancer Society also has programs and services – including rides to treatment, lodging, and more – to help you get through treatment. Call our National Cancer Information Center at 1-800-227-2345 and speak with one of our trained specialists.

- [Find Support Programs and Services in Your Area](#)⁶

Choosing to stop treatment or choosing no treatment at all

Because CD is not a cancer but may lead to cancer, it's important to talk through your decision with your doctors. For some people, when treatments may not be helpful or are no longer working, it may be time to weigh the benefits and risks of continuing to try new treatments. Whether or not you continue treatment, there are still things you can do to

help maintain or improve your quality of life.

- [Palliative or Supportive Care](#)⁷

The treatment information given here is not official policy of the American Cancer Society and is not intended as medical advice to replace the expertise and judgment of your cancer care team. It is intended to help you and your family make informed decisions, together with your doctor. Your doctor may have reasons for suggesting a treatment plan different from these general treatment options. Don't hesitate to ask him or her questions about your treatment options.

Surgery for Castleman Disease

Surgery is often used to get a tissue sample to diagnose Castleman disease (CD). A lymph node biopsy (described in [Tests for Castleman Disease](#)¹) is usually a minor procedure, and patients can often go home afterward.

Surgery also works well to treat localized (unicentric) CD. The type of surgery depends on where the disease is located.

If the affected lymph node or nodes are in a place that is easy to get to, such as in the armpit, then surgery is usually straightforward. In many cases the person may even be able to go home the same day after the surgery.

When the enlarged lymph nodes are in a place that is hard to get to, like deep in the chest or abdomen, surgery is more complex and might require a stay in the hospital for a few days after the operation.

Another option is to have [radiation](#) or treatment with [chemotherapy](#) or another drug first. This can shrink the lymph nodes or tumors, which can make them easier to remove with surgery.

Surgery might also be used to help treat symptoms rather than to try to cure the disease. For example, the spleen can be removed if it has grown large and is causing symptoms.

Potential side effects of surgery depend on several factors, including the extent of the

operation and a person's health before surgery. Most people will have at least some [pain](#)² after the operation, but it usually can be controlled with medicines if needed. Other problems can include reactions to anesthesia, damage to nearby organs during the operation, bleeding, blood clots in the legs, and skin infections at the incision sites.

Even though Castleman disease is not a cancer, surgery is often used in much the same way as it is for cancer. You can read more in [Cancer Surgery](#)³.

Hyperlinks

1. www.cancer.org/cancer/castleman-disease/detection-diagnosis-staging/diagnosis.html
2. www.cancer.org/treatment/treatments-and-side-effects/physical-side-effects/pain.html
3. www.cancer.org/treatment/treatments-and-side-effects/treatment-types/surgery.html

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disease with following adjuvant radiotherapy. *SAGE Open Med Case Rep.*2017;5:2050313X17744481.

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

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Radiation Therapy for Castleman Disease

Radiation therapy uses high-energy radiation to kill cells. It is sometimes used to treat localized Castleman disease (CD), especially if the affected lymph nodes can't be removed completely with [surgery](#). Radiation can also be used as part of the treatment for multicentric CD.

Before your treatment starts, the radiation team will take careful measurements to find the correct angles for aiming the radiation beams and the proper dose of radiation. This planning session, called **simulation**, usually includes getting [imaging tests](#)¹ such as CT or MRI scans.

Radiation therapy is much like getting an x-ray, but the radiation is much stronger. The treatment itself is painless. It lasts only a few minutes, although the setup time – getting you into place for treatment – usually takes longer. You might get radiation treatment for several days in a row.

Possible side effects

Depending on where the radiation is aimed, side effects may include:

- Skin changes in areas getting radiation, ranging from redness to blistering and peeling

- Nausea and vomiting
- Diarrhea
- Fatigue
- Low blood counts

Most [side effects](#)² go away a short while after treatment ends, although fatigue and skin changes may last longer. Talk with your doctor about the possible side effects and the ways to reduce or relieve them.

Even though Castleman disease is not a cancer, radiation is often used in the same way as it is when people have cancer. To learn more, see [Radiation Therapy](#)³⁴.

Hyperlinks

1. [/ssLINK/imaging-radiology-tests-for-cancer](#)
2. www.cancer.org/treatment/treatments-and-side-effects/physical-side-effects.html
3. www.cancer.org/treatment/treatments-and-side-effects/treatment-types/radiation.html
4. [/content/cancer/en/treatment/treatments-and-side-effects/treatment-types/radiation/radiation-therapy-guide.html](#)

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Corticosteroids for Castleman Disease

Corticosteroids are a group of drugs related to hormones made in the body by the adrenal glands. These drugs weaken the immune system, so they are useful in treating people with certain immune system diseases and cancers that develop from immune system cells, such as [lymphomas](#)¹.

In some patients with multicentric Castleman disease (CD), these drugs can be helpful, either alone or along with [chemotherapy](#).

Corticosteroids are often taken as pills, but they can also be injected into a vein. Prednisone is the corticosteroid pill most often used to treat CD.

Side effects of corticosteroids (especially long-term use) can include increased blood sugar (which can lead to diabetes), mood changes, increased risk of [infections](#)², weakened bones, [fatigue](#)³, muscle weakness, weight gain, fluid retention, and high blood pressure. Most of these side effects improve after the drug is stopped.

Hyperlinks

1. www.cancer.org/cancer/lymphoma.html
2. www.cancer.org/treatment/treatments-and-side-effects/physical-side-

[effects/infections.html](#)

3. www.cancer.org/treatment/treatments-and-side-effects/physical-side-effects/fatigue.html

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Chemotherapy for Castleman Disease

Chemotherapy (chemo) is the use of anti-cancer drugs that are injected into a vein or a muscle or are taken by mouth. These drugs enter the bloodstream and reach all areas of the body, making this treatment very useful for multicentric Castleman disease (CD). Chemo may be used alone, in combination with [corticosteroids](#) or other drugs, or combined with [radiation therapy](#) (called **chemoradiation**).

Many chemo drugs can be used to treat patients with multicentric CD. The drugs used most often include:

- Carmustine
- Cladribine
- Chlorambucil
- Cyclophosphamide
- Doxorubicin
- Etoposide
- Melphalan
- Vinblastine
- Vincristine

Often several drugs are combined. Because CD is similar to [lymphomas](#)¹ in many ways, doctors often use chemo combinations like those used for lymphoma. But because CD is so rare, there is not a lot of information on which chemo treatment is best or even how well it works.

Doctors give chemo in cycles, in which a period of treatment is followed by a rest period to give the body time to recover. Each chemo cycle generally lasts for several weeks. Most chemo treatments are given on an outpatient basis (in the doctor's office or clinic or hospital outpatient department) but some might require a hospital stay. Sometimes a patient takes one drug combination for several cycles and then later is switched to a different one.

Possible side effects

Chemo drugs attack cells that are dividing quickly, which is why they work against cancer and diseases like CD. But other cells in the body, such as those in the bone marrow (where new blood cells are made), the lining of the mouth and intestines, and the hair follicles, also divide quickly. These cells can also be affected by chemo, which can lead to certain side effects.

The [side effects](#)² of chemo depend on the type and dose of drugs given and the length

of time they are taken. These side effects can include:

- Hair loss
- Mouth sores
- Loss of appetite
- [Nausea and vomiting](#)³
- Diarrhea
- Increased risk of [infections](#)⁴ (due to a shortage of white blood cells)
- Easy bruising and bleeding (due to a shortage of blood platelets)
- [Fatigue](#)⁵ and weakness (due to a shortage of red blood cells)

Along with the risks above, some chemo drugs can cause other side effects. Ask your health care team about what side effects you can expect based on the specific drugs you will get. Be sure to tell your doctor or nurse if you do have side effects, as there are often ways to help with them. For example, drugs can be given to help prevent or reduce nausea and vomiting.

Even though Castleman disease is not a cancer, chemo is often used in much the same way as it is for cancer. To learn more, see [Chemotherapy](#)⁶.

Hyperlinks

1. www.cancer.org/cancer/lymphoma.html
2. www.cancer.org/treatment/treatments-and-side-effects/physical-side-effects.html
3. www.cancer.org/treatment/treatments-and-side-effects/physical-side-effects/nausea-and-vomiting.html
4. www.cancer.org/treatment/treatments-and-side-effects/physical-side-effects/infections.html
5. www.cancer.org/treatment/treatments-and-side-effects/physical-side-effects/fatigue.html
6. www.cancer.org/treatment/treatments-and-side-effects/treatment-types/chemotherapy.html

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Immunotherapy for Castleman Disease

Immunotherapy is treatment that either boosts the patient's own immune system or uses man-made versions of the normal parts of the immune system.

Monoclonal antibodies

Antibodies are proteins made by the body's immune system to help fight infections. Man-made versions, called **monoclonal antibodies**, can be designed to attack a specific target, such as a substance on the surface of lymphocytes, the cells in which Castleman disease (CD) starts. Sometimes they are given along with [chemotherapy](#).

Siltuximab (Sylvant): This monoclonal antibody is used to treat some patients with multicentric CD. These patients often have high levels of a protein called IL-6. Siltuximab binds to IL-6, which keeps the protein from acting on lymphocytes. This drug doesn't seem to bind to IL-6 that is made from viruses, so it isn't meant for patients who are infected with either HIV or HHV-8. (IL-6, HIV, and HHV-8 were discussed in [What Causes Castleman Disease?](#)¹)

This drug is given as an infusion into a vein (IV), usually every 3 weeks.

Side effects tend to be mild and can include rash, itching, swelling, and weight gain. Some patients can also have side effects during the infusion, such as flushing (skin redness with a feeling of warmth), chest pain, back pain, nausea, and rapid heartbeat.

Rituximab (Rituxan): This monoclonal antibody is used widely for [lymphoma](#)². It can also be helpful in treating CD. Rituximab attaches to a protein called CD20 that is found on the surface of some lymphocytes. This attachment causes the cell to die.

Side effects of rituximab are most common during the infusion, and can include chills, fever, nausea, rashes, fatigue, and headaches. Rarely, more severe side effects occur during the infusion, such as trouble breathing and low blood pressure. Unlike regular chemotherapy, rituximab does not cause low blood counts or hair loss.

This drug can also increase a person's risk of certain infections. In people who have ever been infected with the hepatitis B virus, this drug can sometimes cause the infection to become active again. Your doctor may check your blood for signs of a prior hepatitis infection before starting this drug to see if it is safe.

Tocilizumab (Actemra): This monoclonal antibody is used to treat some kinds of arthritis that can affect the immune system. Tocilizumab works by blocking certain messenger proteins, known as cytokines, that send signals between some cells of the immune system. Tocilizumab is sometimes used in unicentric and multicentric CD after other treatments have stopped working.

Side effects of tocilizumab include runny or stuffy nose, sinus pain, sore throat, headache, dizziness, itching, mild stomach cramps, or urinary tract infection (UTI). This drug also increases the risk for developing serious infections. Your doctor will monitor you closely during treatment.

Rituximab and tocilizumab are drugs that are given through infusion into a vein (IV) at the doctor's office or clinic.

Other antibodies: Newer antibodies that attack other targets are also being studied for

use against CD. These are discussed in [What's New in Research of Castleman Disease?](#)³

Immunomodulating drugs (IMiDs)

Drugs such as thalidomide (Thalomid) and lenalidomide (Revlimid) are used to treat certain cancers of immune cells such as [multiple myeloma](#)⁴ and some types of lymphoma, but they have also helped some patients with CD.

These drugs are thought to work by affecting parts of a person's immune system. It's not exactly clear how they do this, but it seems to be at least in part by working against interleukin-6 (IL-6).

The drugs can cause side effects such as drowsiness, fatigue, constipation, low blood cell counts, and neuropathy (painful nerve damage). There is also an increased risk of serious blood clots (that start in the leg and can travel to the lungs). These tend to be more likely with thalidomide.

Because of concerns these drugs can cause severe birth defects if taken during pregnancy, they can only be obtained through special programs run by the drug company that makes them.

Interferon-alfa

Interferon-alfa is a hormone-like protein made by white blood cells in the body to help the immune system fight infections. Some patients with CD have improved with man-made interferon treatment.

Interferon is given by an injection, either daily or several times a week. This may be into a vein (IV), under the skin (SubQ), or into a muscle (IM). It may be given in a doctor's office, or you or a family member can be taught how to give the medicine under the skin.

Side effects of this treatment can include fatigue, fever, chills, headaches, muscle and joint aches, and mood changes. Because of its side effects, interferon is not used very often. It may be given to some patients in addition to chemotherapy.

Hyperlinks

1. www.cancer.org/cancer/castleman-disease/causes-risks-prevention/what-causes.html

2. www.cancer.org/cancer/lymphoma.html
3. www.cancer.org/cancer/castleman-disease/about/new-research.html
4. www.cancer.org/cancer/multiple-myeloma.html

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[disease/references.html](#))

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Anti-viral Drugs for Castleman Disease

Multicentric Castleman disease (CD) is sometimes associated with the virus HHV-8. Doctors have had success in treating some patients with multicentric CD with drugs that kill this virus, such as ganciclovir, valganciclovir, and foscarnet.

Many patients with [HIV infection](#)¹ are treated with anti-retroviral therapy to keep the HIV in check. But the effect of therapy for HIV on CD is not clear.

Hyperlinks

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

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Treatment of Localized (Unicentric) Castleman Disease

Surgery is the recommended treatment for people with localized Castleman disease (CD) whenever possible. Removing the abnormal lymph node(s) usually cures the disease. Symptoms such as fever and fatigue that are caused by the CD go away when the lymph node is removed. Relapses are rare.

Sometimes if the diseased area is too large to be removed by surgery, medicines such as corticosteroids or rituximab might be given first. This can shrink the tumor and make it easier to remove.

Radiation can also be used to treat localized CD, either in people who can't have surgery for some reason or if surgery cannot remove all of the disease. But radiation is not used as often as surgery as the main treatment.

Some patients with localized CD develop secondary amyloidosis, a condition in which abnormal proteins build up in the kidneys, skin, and some other organs. This protein build-up stops once the lymph node(s) affected by CD is removed.

The outlook for localized CD is very good if the affected lymph node(s) can be removed with surgery (or treated with radiation). But sometimes not all of the disease can be removed or treated safely. This doesn't necessarily mean it will continue to grow and get worse. Even partial removal may help, and the disease may not grow back.

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

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Treatment of Multicentric Castleman Disease

Multicentric Castleman disease (MCD) is usually much harder to treat than localized CD. There is no standard therapy for MCD, and no single treatment works for all patients. Several types of treatment have been shown to help some patients. But because MCD is rare, it has been hard for doctors to compare different treatments against each other in clinical trials.

Surgery is used to biopsy an affected lymph node to make the diagnosis, but MCD is too widespread to remove it all with [surgery](#) or treat it effectively with [radiation](#). Still, some people are helped by these treatments. For example, surgery is sometimes used to remove an enlarged spleen that is causing symptoms, or radiation can be used to shrink tumors in a specific area that are causing problems.

More often, doctors use treatments that can reach all parts of the body, such as [corticosteroids](#), [chemotherapy](#), or [immunotherapy](#), when treating MCD. Doctors usually try one or a combination of these treatments to try to put the disease into remission. [Anti-viral drugs](#) including anti-HIV treatment may also help.

In some patients the disease shrinks or even goes away completely with treatment, at least for a time. In others, the benefit does not last long and symptoms come back once treatment is stopped. Some patients may not be helped by these drugs at all.

In people whose MCD is no longer responding to other treatments, some doctors might recommend high-dose chemotherapy followed by a stem cell transplant. There are a

few reported cases of this being successful. This is a complex, serious, and often expensive treatment, so it's important to understand what it might entail if you are considering this option. For more on this treatment, see [Stem Cell Transplant for Cancer](#).¹

The long-term outlook (prognosis) for people with MCD is often not as good as for people with localized CD. Treatment can often help for a time, but the disease tends to come back within a couple of years. A major concern is that people with MCD are at risk of dying from other causes, like serious infections or progression of the MCD to a fast-growing form of lymphoma that is hard to treat. (For more information about lymphoma, see [Non-Hodgkin Lymphoma](#).²)

The outlook for MCD tends to be worse if the person also has [HIV/AIDS](#).³ Even if the HIV infection is under control with drug treatment, the MCD is not likely to go away. The treatment and outlook of MCD can also be complicated by the presence of [Kaposi sarcoma](#)⁴ and other AIDS-related conditions. These conditions may be less of a problem if the patient is on anti-HIV treatment.

Because MCD can be hard to treat, taking part in a [clinical trial](#)⁵ of newer treatments can be a good option for some people. (See [What's New in Research of Castleman Disease?](#)⁶ for a description of some newer treatments.)

Hyperlinks

1. www.cancer.org/treatment/treatments-and-side-effects/treatment-types/stem-cell-transplant.html
2. www.cancer.org/cancer/non-hodgkin-lymphoma.html
3. www.cancer.org/cancer/cancer-causes/infectious-agents/hiv-infection-aids.html
4. www.cancer.org/cancer/kaposi-sarcoma.html
5. www.cancer.org/treatment/treatments-and-side-effects/clinical-trials.html
6. www.cancer.org/cancer/castleman-disease/about/new-research.html

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Written by

The American Cancer Society medical and editorial content team
(www.cancer.org/cancer/acs-medical-content-and-news-staff.html)

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