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Early Detection, Diagnosis, and Staging of Castleman Disease

Detection and Diagnosis

Some people with Castleman disease may have signs and symptoms that can be noticed, but that is not always the case.

- [Signs and Symptoms of Castleman Disease](#)
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Staging and Outlook (Prognosis)

Learn how staging provides information about the extent of disease and anticipated response to treatment.

- [Castleman Disease Stages](#)
- [Survival Rates for Castleman Disease](#)

Questions To Ask About Castleman Disease

Here are some questions you can ask to help you better understand your diagnosis and treatment options.

- [Questions to Ask About Castleman Disease](#)
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Signs and Symptoms of Castleman Disease

Castleman disease (CD) can cause a lot of different types of symptoms, and in some people it might not cause any symptoms at all. If symptoms do occur, they are often like those seen with other diseases, such as infections, autoimmune diseases, or even some types of cancer. Because of this, doctors might not suspect CD at first.

Common symptoms of localized CD

In the localized form of CD, symptoms are found in a particular part of the body. Localized CD often starts as an enlarged lymph node. If the node is just under the skin, such as in the neck or underarm area, it might be seen or felt as a lump. But if it's in the chest or abdomen (belly), it might not be noticed until it grows large enough to cause other symptoms:

- An enlarged node in the chest might press on the windpipe, which could cause trouble breathing, wheezing, a cough, or a feeling of fullness in the chest.
- An enlarged node in the abdomen can cause trouble eating, pain, or just a feeling of fullness.

In general, most people with localized CD feel well otherwise. In fact, some people have no symptoms at all, and CD is found only when the doctor does a test for another reason. On the other hand, some people with localized CD can also have some of the other symptoms listed below.

Common symptoms of multicentric CD

People with multicentric CD have more than one area of enlarged lymph nodes. The enlarged nodes can be in the chest or abdomen, but multicentric CD often affects lymph nodes in the groin, the underarm area, and on the sides of the neck, which can often be seen or felt as lumps under the skin.

Multicentric CD can also affect lymphoid tissue of internal organs, causing the liver, spleen, or other organs to enlarge. Enlarged organs might be seen or felt as masses under either side of the rib cage. They can also cause problems eating or a sense of fullness (or even pain) in the abdomen.

Other symptoms of CD

In addition, people with either type of CD can have other symptoms (although these symptoms are much more common in people with multicentric CD):

- Fever
- Night sweats (that soak the sheets)
- Weight loss
- Loss of appetite
- Weakness
- Fatigue (tiredness)
- Shortness of breath
- Nausea and vomiting
- Nerve damage that leads to numbness and weakness (neuropathy)
- Leg swelling (edema)
- Skin rashes

Some of these symptoms might come and go over time.

Amyloidosis, a condition where abnormal proteins build up in body tissues, can occur in CD. This can lead to kidney damage, heart damage, nerve damage, and intestinal problems, mainly diarrhea. If CD is treated successfully, the amyloidosis may improve or even go away.

Anemia(having too few red blood cells) is very common in multicentric CD, and can lead to problems such as fatigue and shortness of breath.

CD is rare, and the symptoms above often have other causes. Still, if you have any of these symptoms and they don't go away within a few weeks (or they get worse), see a doctor so that the cause can be found and treated, if needed.

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

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Tests for Castleman Disease

People with Castleman disease (CD) may see their doctor because of [symptoms](#) they are having, or because they just don't feel well and go in for a checkup. Sometimes CD is found in people without symptoms when it's found on an [imaging or blood test](#)¹being

done for another reason.

CD is rare, and its symptoms are often like those caused by other diseases (including infections, autoimmune diseases, and [lymphomas](#)²), so doctors often suspect it is something else at first. The diagnosis of CD does involve looking at any symptoms the patient may have and blood test results. However, an actual diagnosis is made when doctors remove an affected lymph node and look at it with a microscope. This procedure, known as a **biopsy**, is described below.

Medical history and physical exam

If your symptoms or the results of a test suggest you might have a lymph node problem such as CD, your doctor will want to get a thorough medical history, including the details of any symptoms, possible [risk factors](#)³, family history, and other medical conditions.

Next, the doctor will examine you, paying special attention to lymph nodes and other areas of your body that could be involved, including the spleen and liver. Because infections are the most common cause of enlarged lymph nodes, the doctor will look for an infection in the part of the body near the swollen lymph nodes.

If the doctor suspects that CD or another serious problem (such as lymphoma) might be causing the symptoms, he or she will probably order blood tests, imaging tests, and/or do a biopsy of an affected lymph node (see below).

Blood tests

Blood tests are likely to be done if the doctor suspects CD, some other type of immune system problem, a serious infection, or other conditions. Some of the types of blood tests that can be abnormal in people with CD include:

- Complete blood count (CBC)
- Interleukin-6 (IL-6)
- Erythrocyte sedimentation rate (ESR)
- C-reactive protein (CRP)
- Kidney function tests

Having abnormal results for these lab tests doesn't prove someone has CD, because other conditions can also cause these problems. But these and other blood tests can help doctors make the right diagnosis.

Imaging tests

Imaging tests use x-rays, magnetic fields, sound waves, or radioactive particles to create pictures of the inside of the body. These tests may be done for a number of reasons, including

- To look for enlarged lymph nodes or organs that might be causing symptoms
- To look for enlarged nodes in other parts of the body
- To help determine if [treatment](#)⁴ is working

People who might have CD (or another lymph node problem) may have one or more of the following tests.

Computed tomography (CT) scan

The [CT scan](#)⁵ uses x-rays to make detailed cross-sectional images of your body. Unlike a regular x-ray, CT scans can show the detail in soft tissues (such as internal organs). This scan can help tell if any lymph nodes or organs in your body are larger than normal.

CT-guided needle biopsy: In some cases, CT scans can also be used to guide a biopsy needle precisely into a lymph node. A small sample of the node is then removed and looked at with a microscope. A needle biopsy can't diagnose CD by itself, but it can sometimes help diagnose or exclude other diseases that can cause large lymph nodes.

Magnetic resonance imaging (MRI)

[MRI](#)⁶ is not used as often as CT scans for lymph node problems, but if your doctor is concerned about areas near your spinal cord or brain, MRI is very useful for looking at these areas.

Like CT scans, MRI scans show detailed images of soft tissues in the body. But MRI scans use radio waves and strong magnets instead of x-rays.

Chest x-ray

This test might be done if you're having breathing problems, to find out if there are enlarged lymph nodes in your chest (usually in the center part of the chest between the lungs, the **mediastinum**)..

Ultrasound

[Ultrasound](#)⁷ uses sound waves and their echoes to create pictures of internal organs or masses.

Ultrasound can be used to look at lymph nodes near the surface of your body or to look inside your abdomen for enlarged lymph nodes or organs such as the liver and spleen. It can also show kidneys that have become swollen because the outflow of urine has been blocked by enlarged lymph nodes. (It can't be used to look at organs or lymph nodes in the chest because the ribs block the sound waves.)

Positron emission tomography (PET) scan

[PET scans](#)⁸ are helpful in finding small collections of fast-growing cells that might not be visible on a CT scan. PET is not often used to diagnose CD, but sometimes it can help the doctor determine the cause of enlarged lymph nodes.

For a PET scan, you are injected with a slightly radioactive form of sugar, which collects mainly in cancer cells. A special camera is then used to create a picture of areas of radioactivity in the body. The picture is not detailed like a CT or MRI scan, but a PET scan can look for possible areas of cancer spread in all areas of the body at once.

Some newer machines can do both a PET and CT scan at the same time (PET/CT scan). This lets the doctor see areas that "light up" on the PET scan in more detail.

Gallium scan

For this test, you are injected with a solution containing slightly radioactive gallium. It is attracted to lymph tissue in the body. A few days later a special camera is used to detect the radioactivity, showing the location of the gallium. A gallium scan can sometimes find unsuspected sites of CD disease, but it is not always reliable since the gallium might not be taken up by all of the lymph nodes affected by CD.

This test is not used as much now as in the past, as many doctors do a PET scan instead.

Lymph node biopsy

A doctor might suspect you have Castleman disease based on your symptoms or the results of exams or tests, but it can only be diagnosed by removing an enlarged lymph node and examining it under the microscope. This procedure is called a **biopsy**.

Different types of biopsies can be used, based on where the lymph node is.

Excisional or incisional biopsy: If the lymph node is near the skin surface, a surgeon can often remove the node using local anesthesia (numbing medicine). The surgeon cuts the skin over the enlarged lymph node, removes the node, and then stitches the cut closed.

- If the procedure removes the entire lymph node, it is called an excisional biopsy.
- If only part of the node is removed, it is called an incisional biopsy.

If the lymph node is in the chest or the abdomen, the surgeon might need to make a large incision to get into either of these places. This type of surgery might require general anesthesia (where you are in a deep sleep), but it might be needed to learn why the lymph node is enlarged.

Sometimes, lymph nodes in the chest can be removed by **mediastinoscopy**. In this procedure, a small cut is made in the front of the neck and a thin, hollow, lighted tube (called a mediastinoscope) is inserted behind the sternum (breast bone) and in front of the windpipe to look at the area. Special instruments can be passed through this tube to remove all or part of a lymph node.

The same type of procedure can be used to sample lymph nodes in the abdomen. In this case, the test is known as **laparoscopy**. The doctor makes a small cut in the abdomen and inserts a thin, hollow, lighted tube (called a laparoscope) and other instruments to look at the area and remove all or part of a lymph node.

Fine needle aspiration (FNA) or core needle biopsy: Sometimes lymph nodes are biopsied by putting a hollow needle into the node to remove a small amount of tissue. In a fine needle aspiration (FNA) biopsy, the doctor uses a very thin needle to withdraw (aspirate) a small amount of tissue from the enlarged node. For a core needle biopsy, the doctor uses a larger needle to remove a slightly larger piece of tissue.

Doctors have found that diagnosing CD by needle biopsy is sometimes possible, but biopsy methods that remove larger samples of tissue are usually recommended because they are thought to be more accurate.

Lab tests of biopsy samples

No matter what procedure is used to take a biopsy, the cells from the biopsy are then sent to a lab. Using a microscope, a doctor called a pathologist looks at them and might do other [tests](#)⁹. Since CD is so rare, the pathologist might ask another pathologist with

special training in diagnosing blood and lymph node diseases (called a hematopathologist) to look at the biopsy.

Sometimes it's hard to tell if the lymph node is affected by CD or by lymphoma. In these cases, other tests might be done on the lymph node tissue to help figure this out. Some tests look at the proteins on the surface of the cells, while others look for gene or chromosome changes within the cells. Examples of these lab tests include:

- Immunohistochemistry
- Flow cytometry
- Cytogenetics
- Fluorescent in situ hybridization (FISH)
- Polymerase chain reaction (PCR)

These tests, are described in more detail in [Non-Hodgkin Lymphoma](#)¹⁰.

Hyperlinks

1. www.cancer.org/treatment/understanding-your-diagnosis/tests.html
2. www.cancer.org/cancer/lymphoma.html
3. www.cancer.org/cancer/castleman-disease/causes-risks-prevention/risk-factors.html
4. www.cancer.org/cancer/castleman-disease/treating.html
5. www.cancer.org/treatment/understanding-your-diagnosis/tests/ct-scan-for-cancer.html
6. www.cancer.org/treatment/understanding-your-diagnosis/tests/mri-for-cancer.html
7. www.cancer.org/treatment/understanding-your-diagnosis/tests/ultrasound-for-cancer.html
8. www.cancer.org/treatment/understanding-your-diagnosis/tests/nuclear-medicine-scans-for-cancer.html
9. www.cancer.org/treatment/understanding-your-diagnosis/tests/testing-biopsy-and-cytology-specimens-for-cancer.html
10. www.cancer.org/cancer/non-hodgkin-lymphoma.html

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

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Castleman Disease Stages

After someone is diagnosed with cancer, doctors will try to figure out if it has spread and if so, how far. This process is called **staging**. The stage of a cancer describes how much cancer is in the body. It helps determine how serious the cancer is and how best to [treat](#)¹ it. Doctors also use a cancer's stage when talking about survival statistics.

Since Castleman disease (CD) is not a cancer, it doesn't have a formal staging system. Instead, doctors use other important pieces of information to help decide on the best treatment and to give them an idea of how well a patient might do.

The most important factor when deciding on treatment is whether the CD is [localized/unicentric or multicentric](#).² Localized/unicentric CD affects only a single lymph node (or lymph node group). The multicentric type affects 2 or more groups of lymph nodes in different parts of the body. It may also affect organs like the spleen or liver. Tests are done to see what lymph nodes and organs are affected to learn which type of CD a patient has. Doctors use the results of physical exams and imaging, such as a chest x-ray and either a CT scan or MRI of the chest and abdomen. These are described in [Tests for Castleman Disease](#).

Another factor is the microscopic subtype of the CD, which is a description of the patterns of cells seen under the microscope in the biopsy sample. These subtypes are described in [What Is Castleman Disease?](#)³

A third important factor is whether or not the patient is infected with the human immunodeficiency virus (HIV), the virus that causes AIDS. Just about all people infected with HIV who develop CD will have the multicentric form of the disease.

Hyperlinks

1. www.cancer.org/cancer/castleman-disease/treating.html
2. www.cancer.org/cancer/castleman-disease/about/what-is-castleman-disease.html
3. www.cancer.org/cancer/castleman-disease/about/what-is-castleman-disease.html

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Survival Rates for Castleman Disease

Doctors often use survival rates as a standard way of discussing a person's outlook (prognosis). Survival rates tell you what portion of people with the same type and stage of disease are still alive a certain amount of time after they were diagnosed. They can't tell you how long you will live, but they may help give you a better understanding about how likely it is that your treatment will be successful.

It's hard to get accurate numbers on the outlook for people with CD because it is rare. Most statistics on the disease come from small numbers of patients who were treated at a single center or hospital. These numbers might not accurately reflect the outcomes for all people with CD.

The numbers below come from a study of nearly 200 people treated for CD in many different centers. The study divided people into [groups](#)¹ based on whether the CD was unicentric or multicentric, its microscopic subtype, and whether the patient was infected with HIV (HIV+) or not infected (HIV-).

The 3-year **disease-free survival (DFS)** rate refers to the percentage of patients who were still alive and had no signs of CD at least 3 years after it was diagnosed. Of course, many people went much longer than 3 years without any signs of CD (and many were likely cured).

Type of Castleman Disease	3-Year Disease-Free Survival Rate
Unicentric, hyaline vascular, HIV-	93%
Unicentric, plasma cell or mixed, HIV-, OR Multicentric, hyaline vascular, HIV-	79%
Multicentric, plasma cell, HIV-	46%
HIV+ (multicentric)	28%

Even when taking the factors above into account, disease-free survival rates are at best rough estimates. If you have CD, your doctor can tell you how well these numbers might apply to you, as he or she knows your situation best.

Hyperlinks

1. www.cancer.org/cancer/castleman-disease/about/what-is-castleman-disease.html

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

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Questions to Ask About Castleman Disease

It's important to have honest, open discussions with your cancer care team. Ask any question, no matter how small it might seem. Some questions to consider:

When you're told you have Castleman disease (CD)

- Is my CD [localized or multicentric](#)¹?
- Has my biopsy been reviewed by a pathologist who is an expert on CD?

- Do I also have [HIV infection and AIDS](#)²? If so, how does it influence my prognosis (outlook) and treatment of CD?
- Do I need other [tests](#) before we can decide on treatment?
- Are there other doctors I need to see?

When deciding on a treatment plan

- How much experience do you have treating CD?
- Should I get a second opinion before starting treatment? Can you suggest a doctor or treatment center?
- What [treatment choices](#)³ do I have? Do we need to start treatment right away?
- Am I eligible for [clinical trials](#)⁴ of any new treatments?
- Which treatment do you recommend, and why?
- What are the side effects of the treatments that you recommend?
- What can I do to help reduce the side effects I may have from the treatment?
- What should I do to be ready for treatment?
- How long will treatment last? What will it be like? Where will it be done?
- How will treatment affect my daily activities?
- What is my outlook for survival?
- What are the chances of the CD coming back with these treatment plans?

During treatment

- What would we do if the treatment doesn't work or if the CD comes back?
- What type of [follow-up](#)⁵ will I need after treatment?
- What symptoms or side effects should I tell you about right away?
- How can I reach you on nights, holidays, or weekends?
- Do I need to change what I eat during treatment?
- Are there any limits on what I can do?
- Should I exercise? What should I do, and how often?
- Can you suggest a mental health professional I can see if I start to feel overwhelmed, depressed, or distressed?

After treatment

- Are there any limits on what I can do?
- What symptoms should I watch for?
- What kind of exercise should I do now?
- What type of follow-up will I need after treatment?
- How often will I need to have follow-up exams and tests?
- How will we know if the cancer has come back? What should I watch for?
- What will my options be if the cancer comes back?

Along with these sample questions, be sure to write down some of your own. For instance, you might want more information about recovery times so that you can plan your work or activity schedule.

Keep in mind that doctors aren't the only ones who can give you information. Other health care professionals, such as nurses and social workers, can answer some of your questions. For more about speaking with your health care team, see [Talking With Your Doctor](#)⁶.

Hyperlinks

1. www.cancer.org/cancer/castleman-disease/about/what-is-castleman-disease.html
2. www.cancer.org/cancer/cancer-causes/infectious-agents/hiv-infection-aids.html
3. www.cancer.org/cancer/castleman-disease/treating.html
4. www.cancer.org/treatment/treatments-and-side-effects/clinical-trials.html
5. www.cancer.org/cancer/castleman-disease/after-treatment.html
6. [/ssLINK/talking-with-your-doctor-toc](#)

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

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