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](#)
- [How Is Castleman Disease Diagnosed?](#)

Staging and Outlook (Prognosis)

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

- [How Is Castleman Disease Staged?](#)
- [Outlook (Prognosis) for People With 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.

- [What Should You Ask Your Doctor About Castleman Disease?](#)

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**

The localized form of 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 the CD is only found 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.

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 tissues around the body, can occur in CD. This can lead to kidney damage, heart damage, nerve damage, and intestinal problems, mainly diarrhea. If the 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.

- References
  See all references for Castleman Disease

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**How Is Castleman Disease Diagnosed?**

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 seen on an imaging test being done for another reason.

CD is rare, and its symptoms are often like those caused by other diseases (including infections, autoimmune diseases, and lymphomas), so doctors often suspect it is
something else at first. The actual diagnosis of CD is made when doctors remove an affected lymph node and look at it under 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, family history, and other medical conditions.

Next, the doctor will examine you, paying special attention to the lymph nodes and other areas of the body that might 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 might 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)

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 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 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.

Instead of taking one picture like a regular x-ray, a CT scanner takes many pictures as it rotates around you. A computer then combines these pictures into detailed images of the part of your body being studied.

Before the scan you might be asked to drink 1 or 2 pints of a contrast agent. This helps outline the intestine so that certain areas aren’t mistaken for tumors. You might also need an intravenous (IV) line through which a different kind of contrast dye is injected. This helps better outline structures in your body. The injection can cause some flushing (redness and warm feeling, especially in the face). Some people are allergic and get hives. Rarely, more serious reactions like trouble breathing and low blood pressure can occur. Be sure to tell your doctor if you have any allergies or have ever had a reaction to any contrast material used for x-rays. You can be given medicine to help prevent or treat allergic reactions.

A CT scanner has been described as a large donut, with a narrow table that slides in and out of the middle opening. You will need to lie still on the table while the scan is being done. CT scans take longer than regular x-rays, and you might feel a bit confined by the ring while the pictures are being taken.

**CT-guided needle biopsy:** CT scans can also be used to guide a hollow biopsy needle precisely into an enlarged lymph node. For this procedure, you remain on the CT scanning table while a doctor moves a biopsy needle through the skin and toward the lymph node. CT scans are repeated until the needle is within the lymph node. A small sample of the node is then removed to be looked at under 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)**
This test is not used as often as CT scans for lymph node problems, but if your doctor is concerned about areas near the spinal cord or brain, MRI is very useful for looking at these areas.

Like CT scans, MRI scans provide detailed images of soft tissues in the body. But MRI scans use radio waves and strong magnets instead of x-rays. The energy from the radio waves is absorbed by the body and then released in a pattern formed by the type of body tissue and by certain diseases. A computer translates the pattern of radio waves given off by the tissues into a very detailed image of parts of the body. A contrast material might be injected just as with CT scans but is used less often.

MRI scans take longer than CT scans – often up to an hour – and are a little more uncomfortable because you have to lie inside a narrow tube, which is confining and can upset people with a fear of enclosed spaces. Newer, more open MRI machines may be another option. The MRI machine makes loud buzzing and clicking noises that you may find disturbing. Some places provide headphones or earplugs to help block this noise out.

**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 called the *mediastinum*.

**Ultrasound**

Ultrasound uses sound waves and their echoes to create pictures of internal organs or masses. For this test, a small, microphone-like instrument called a *transducer* is placed on the skin (which is first lubricated with a gel). It gives off sound waves and picks up the echoes as they bounce off the organs. The echoes are converted by a computer into a black and white image that is displayed on a computer screen.

Ultrasound can be used to look at lymph nodes near the surface of the 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.)

This is an easy test to have done, and it uses no radiation. For most ultrasounds, you simply lie on a table, and a technician moves the transducer over the part of your body being looked at.
**Positron emission tomography (PET) scan**

PET scans 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, a form of radioactive sugar (known as fluorodeoxyglucose or FDG) is injected into the blood. (The amount of radioactivity used is very low and will pass out of the body over the next day or so.) After about an hour, you are moved onto a table in the PET scanner. You lie on the table for about 30 minutes while a special camera creates a picture showing areas of radioactivity in the body.

Any cancer cells in the body will be growing quickly, so they absorb large amounts of the radioactive sugar. Although CD cells don’t take up the sugar as much as cancer cells, they do seem to take it up more than normal cells. The picture from a PET scan is not detailed like a CT or MRI scan, but it can provide helpful information about your whole body.

Often the PET scan is combined with a CT scan, which is more detailed. This helps the doctor determine if abnormal areas seen on the CT scan are CD, cancer, or something else.

**Gallium scan**

For this test, a solution containing slightly radioactive gallium is injected into a vein. It is attracted to lymph tissue in the body. A few days later a special camera is used 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**

All biopsy specimens are looked at under a microscope by a pathologist (a doctor specially trained to diagnose disease), who studies the size and shape of the cells and how they are arranged. 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 our document Non-Hodgkin Lymphoma.

- References
See all references for Castleman Disease

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How Is Castleman Disease Staged?

When talking about cancer, the stage is a description of how far it has spread. The stage helps doctors determine the best treatment and the likely outlook (prognosis) for the patient. Most cancers have a formal staging system that lets doctors sum up the extent of the cancer.

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. 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 internal 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. These tests often include some of the imaging tests mentioned in the section “How is Castleman disease diagnosed?” such as a chest x-
ray and either a CT scan or MRI of the chest and abdomen.

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 the section “What is Castleman disease?”

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.

- References
  See all references for Castleman Disease

Outlook (Prognosis) for People With Castleman Disease

Survival rates are often used by doctors as a standard way of discussing a person’s outlook (prognosis). Some people may want to know statistics on the outlook for those in similar situations, while others may not find the numbers helpful, or may even not want to know them. If you don’t want to know about the outlook for people with different types of Castleman disease (CD), stop reading here and skip to the next section.

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 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).

<table>
<thead>
<tr>
<th>Type of Castleman Disease</th>
<th>3-Year Disease-Free Survival</th>
</tr>
</thead>
<tbody>
<tr>
<td>Unicentric, hyaline vascular, HIV-</td>
<td>93%</td>
</tr>
<tr>
<td>Unicentric, plasma cell or mixed, HIV-, OR</td>
<td>79%</td>
</tr>
<tr>
<td>Multicentric, hyaline vascular, HIV-</td>
<td>46%</td>
</tr>
<tr>
<td>Multicentric, plasma cell, HIV-</td>
<td>28%</td>
</tr>
</tbody>
</table>

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.

- References
  See all references for Castleman Disease

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What Should You Ask Your Doctor About Castleman Disease?

As you cope with Castleman disease (CD) and its treatment, we encourage you to have honest, open discussions with your doctors. Feel free to ask any question that’s on your mind, no matter how small it might seem. Here are some questions you might want to ask. Be sure to add your own questions as you think of them. Nurses, social workers, and other members of the treatment team might also be able to answer many of your questions.

- Is my CD localized or multicentric?
- Has my biopsy been reviewed by a pathologist who is an expert on CD?
- Do I also have HIV infection and AIDS? 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?
- 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 do I have? Do we need to start treatment right away?
- 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?
- What would we do if the treatment doesn’t work or if the CD comes back?
- What type of follow-up will I need after treatment?
- Am I eligible for clinical trials of any new treatments?

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.

- References
  See all references for Castleman Disease

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