



# Chronic Lymphocytic Leukemia (CLL) - Staging and Treatment

## What is staging for cancer?

Staging is a way to find out how much cancer is in your body, how far it has spread, and what treatment can be used. Staging helps providers learn about your cancer and your health so that they can plan the best treatment for you. Most cancers form tumors, but CLL does not. It is found in the blood, bone marrow, and organs of the body. Tests like blood counts, flow cytometry, bone marrow biopsy, and blood tests are done to help stage your cancer.

## How is CLL staged?

There are two different staging systems for CLL:

- **Rai Staging System:** Based on results of blood tests (especially the number of white blood cells) and physical exam.
- **Binet Staging System:** Based on the number of affected lymphoid tissue groups and if you have anemia (low red blood cell count) or thrombocytopenia (low platelet count).

### Rai Staging System

In each stage, there is lymphocytosis. Lymphocytosis is when your white blood cell (WBC) count is higher than it should be. The extra WBCs are found in the blood and bone marrow.

- **Stage 0:** Lymphocytosis but your lymph nodes, spleen, and liver are not enlarged (bigger than they should be), and red blood cell and platelet counts are almost normal.
- **Stage I:** Lymphocytosis with enlarged lymph nodes. The spleen and liver are not affected, and red blood cell and platelet counts are almost normal.
- **Stage II:** Lymphocytosis with an enlarged spleen and in some cases an enlarged liver. Lymph nodes may or may not be enlarged. Red blood cell and platelet counts are almost normal.
- **Stage III:** Lymphocytosis plus the lymph nodes, spleen or liver may or may not be enlarged. Red blood cell count is low, and platelets are near normal.
- **Stage IV:** Lymphocytosis plus enlarged lymph nodes, spleen, or liver. Red blood cell counts may be low or near normal and the platelet count is low.

Each stage is also put into a risk group that helps decide which treatments are best:

- **Stage 0:** Low risk.
- **Stage I and II:** Intermediate risk.
- **Stage III and IV:** High Risk.

### Binet Staging System

The Binet system looks at lymphoid tissue, anemia, and thrombocytopenia. Lymphoid tissue groups include the neck lymph nodes, groin lymph nodes, underarm lymph nodes, spleen, and liver.

- **Stage A:** Fewer than 2 areas of lymphoid tissue are enlarged, no anemia, and no thrombocytopenia.
- **Stage B:** 3 or more areas of enlarged lymphoid tissue, no anemia, and no thrombocytopenia.
- **Stage C:** Any number of lymphoid areas are enlarged, and you have anemia and thrombocytopenia.

Although not part of the formal staging systems, prognostic factors can help guide treatment for CLL. Prognostic factors can be called adverse or favorable. Prognostic factors can be things like: age, gender, levels of protein in the body, and chromosome changes. Talk with your provider about if you have any prognostic factors and how they will affect your treatment.

## How is CLL treated?

Treatment for CLL is based on the stage of your disease, your current health, and your goals. You and your care team will decide on the best course of treatment. Treatments can include:

- Supportive Care.
- [Chemotherapy](#).
- [Targeted Therapy](#).
- [Immunotherapy](#).
- [Surgery](#).
- [Bone Marrow Transplant](#).
- [Clinical Trials](#).

## Supportive Care

People with early-stage or less aggressive disease can have better outcomes if treatment is not started until symptoms start or there is a shorter doubling time (doubling time is the amount of time it takes for your lymphocyte count to double). The treatment for stage 0 CLL is often watching to see how you are doing.

Even if CLL is not being treated, supportive care can be given. Supportive care is used to manage side effects of the disease. CLL affects a person's ability to fight infection. The best way to prevent infection is to make sure you and the people you are with wash their hands. Avoid large crowds, like at shopping malls or festivals. The common cold, flu, or pneumonia can be life-threatening for a person with CLL. Vaccines, like the flu vaccine, are less effective in people with CLL. You should talk to your provider about if and when you should get vaccines.

People with anemia can have fatigue, shortness of breath, or look pale. Medications and blood transfusions can be used to treat anemia. Thrombocytopenia can lead to bleeding that is hard to stop. This can be small like gums bleeding when brushing the teeth or a nosebleed, or can be very serious like bleeding that can lead to a stroke. You should not play contact sports, shave with a razor (using an electric razor is okay), or do any activities that increase your risk of bleeding or bruising. Tell your provider if you have symptoms of thrombocytopenia. You are also at risk for [tumor lysis syndrome](#) (TLS) which is the rapid breakdown of cells that causes an imbalance of chemicals and electrolytes in the body. TLS impacts levels of uric acid, potassium, phosphate, and calcium, and can cause kidney failure, seizures, and heart rhythm changes.

Radiation is the use of high-energy x-rays to kill cancer cells. It is not used to treat CLL but can be used as a supportive care measure. Radiation may be used to help shrink an organ that has become enlarged and is causing pain. It can also be used to treat bone pain. Leukemia cells grow in the bone marrow and can cause this pain.

## Chemotherapy

[Chemotherapy](#) is the use of anti-cancer medicine that goes through your whole body to kill cancer cells. You will likely get a few chemotherapies, called a regimen. A regimen can be given over the course of days to

weeks. Some chemotherapy medications used to treat CLL are [bendamustine](#), [chlorambucil](#), [pentostatin](#), [cyclophosphamide](#), [fludarabine](#), and [cladribine](#). Steroids may be given with chemotherapy or alone to treat CLL. Examples of steroids used to treat CLL are [prednisone](#) and [methylprednisolone](#).

## Targeted Therapy

Targeted therapies are medications that target something specific to the cancer cells, stopping them from growing and dividing. Targeted therapies that may be used in the treatment of CLL are:

- **Monoclonal antibodies:** Man-made antibodies are made in a lab and target a specific protein on cancer cells. Some monoclonal antibodies send a message to the immune system to kill the cell and some stop cell growth. Examples of monoclonal antibodies are [obinutuzumab](#), [ofatumumab](#) and [rituximab](#). These are all given intravenously (IV, into a vein).
- **Kinase inhibitors:** These block growth signals within cancer cells, reducing the production of new cancer cells. Examples of kinase inhibitors are [acalabrutinib](#), [ibrutinib](#), [duvelisib](#), and [idelalisib](#).
- **Venetoclax:** This is another targeted therapy that may be used in the treatment of CLL.

## Immunotherapy

Immunotherapy is the use of your own immune system to kill cancer cells. CAR T-cell Therapy takes T-cells from your blood, changes them in a lab to attack cancer cells, and then puts them back in your body. The only CAR T-cell therapy currently approved for CLL is [Lisocabtagene Maraleucel](#).

## Surgery

If you have splenomegaly (enlarged spleen) you may have a [splenectomy](#) (getting your spleen removed). The spleen helps to filter old, unneeded cells from the blood. People without a spleen are at a [higher risk](#) of infection.

## Bone Marrow Transplant

Transplants can be done using a donor's bone marrow or stem cells ([allogeneic](#)) or your own bone marrow or stem cells ([autologous](#)). Allogeneic transplants are more common for people with CLL. One thing that is a very important part of all allogeneic transplants is called the "[graft versus tumor effect](#)." This is the effect that the donor's immune system (which is part of the marrow that they donate) has on your cancer cells. The hope is that the healthy donor immune system can attack any cancer cells that survived the treatment before the transplant.

## Clinical Trials

You may be offered a clinical trial as part of your treatment plan. To find out more about current clinical trials, visit the [OncoLink Clinical Trials Matching Service](#).

## Making Treatment Decisions

Your care team will make sure you are part of choosing your treatment plan. This can be overwhelming as you may be given a few options to choose from. It feels like an emergency, but you can take a few weeks to meet with different providers and think about your options and what is best for you. This is a personal decision. Friends and family can help you talk through the options and the pros and cons of each, but they cannot make the decision for you. You need to be comfortable with your decision - this will help you move on to the next steps. If you ever have any questions or concerns, be sure to call your team.

You can learn more about CLL at [OncoLink.org](#).

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