

B-cell  
Prolymphocytic  
Leukemia  
**B-PLL**

## WHAT YOU NEED TO KNOW

You or your loved one has been diagnosed with B-cell prolymphocytic leukemia (B-PLL). What does it mean and how will it affect you?

This fact sheet will help you:

Learn about  
B-PLL and how  
it is diagnosed

Get an overview  
of treatment  
options

Understand  
what happens  
next



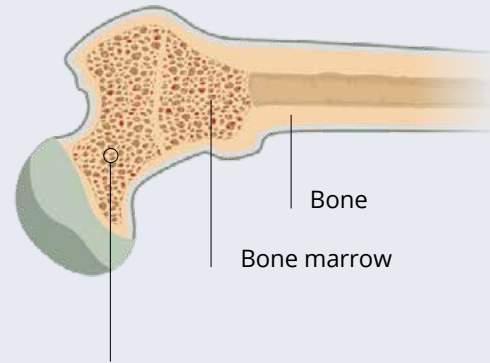
## What is leukemia?

Leukemia is a cancer of the blood and bone marrow. Bone marrow is the soft, spongy material inside bones. Blood cells are formed in the bone marrow. Three kinds of blood cells develop from stem cells:

- **Red blood** cells carry oxygen
- **Platelets** help your blood to clot (stop bleeding)
- **White blood cells** help your body fight infection

When you have leukemia, cancerous blood cells form and push out healthy blood cells.

Blood is created in the **bone marrow** (the spongy part inside the bone).



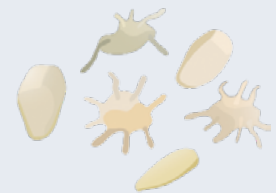
**Stem cell**



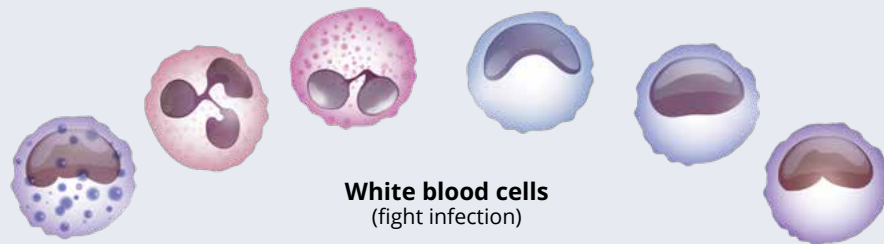
**Three kinds of blood cells** develop from stem cells:



**Red blood cells**  
(carry oxygen)



**Platelets**  
(allow blood to clot)



**White blood cells**  
(fight infection)

**Leukemia is a cancer of the blood and bone marrow.**



**B-PLL often evolves from a more slow-growing B-cell cancer such as chronic lymphocytic leukemia (CLL).**

**It's a type of *prolymphocytic leukemia (PLL)* with cells that are large and abnormal. These cells are not normally found in the blood.**

## About B-cell prolymphocytic leukemia

- B-PLL is a rare and often aggressive cancer
- It affects a specific type of white blood cell known as a B-cell (also known as a B-lymphocyte) that is part of your immune system and helps your body fight infections
- The B-cells grow out of control
- It is slightly more common in men than women
- It usually affects older adults: the average age of diagnosis is 69 years

## Signs and symptoms

It can be difficult to diagnose B-PLL as it presents in a similar way to other B-cell cancers. There are a number of common signs and symptoms.

You may experience:

- Fatigue and weakness
  - When your red blood cell count is low (anemia)
- Bruising and bleeding easily
  - When your platelet count is low (thrombocytopenia)
- Feeling bloated, full, or unable to eat a full meal
  - When your spleen is enlarged (splenomegaly)
- Fatigue, fever, and poor appetite
  - When your lymphocyte count is high (lymphocytosis)
- Fevers, night sweats, and weight loss
  - When you have what is known as B-symptoms
- Skin rash



## Your diagnosis

With a diagnosis, your doctor can determine the right treatment for you. Your test results help your doctor predict how B-PLL will likely progress and how you may respond to treatment. Here are some possible tests you may do:

Name of test	Description
<b>Medical history and physical exam</b>	The doctor reviews past illnesses, injuries, and symptoms. They examine your lungs, heart, and other organs.
<b>Complete blood count</b>	<p>This test measures the number of red blood cells, white blood cells, and platelets in a sample of your blood to find out if the counts are high or low.</p> <p>With B-PLL, you will often have a high number of lymphocytes (a type of white blood cell) and low red blood cell and platelet counts.</p>
<b>Bone marrow aspiration and biopsy</b>	These two tests look at bone marrow cells for anything unusual in your chromosomes (for example, the presence of cancer cells). They are usually done at the same time.
<b>Cytogenetics</b>	Karyotyping examines the number and structure of your chromosomes.
<ul style="list-style-type: none"><li>• Karyotyping</li><li>• Fluorescence <i>in situ</i> hybridization (FISH)</li></ul>	FISH looks at genes and chromosomes in cells to detect changes to the chromosomes of B-PLL cells.
<b>Immunophenotyping by flow cytometry</b>	During this test, cells are taken from your blood or bone marrow to detect which proteins or markers (antigens) you have. This helps to determine if you have B-PLL.
<b>Molecular testing</b>	This genetic test looks for a specific mutation in your genes.
<b>Peripheral blood smear</b>	This test looks at blood cells under a microscope to see the number, size, shape, type, and pattern of cells. It also looks for blast cells, which are not usually present in healthy people.

B-PLL is diagnosed when either:

- More than 55% of the lymphocytes in the peripheral blood are prolymphocytes
- A lymph node or bone marrow sample shows that most of the lymphocytes are prolymphocytes

## B-PLL treatment

B-PLL is an extremely rare disease. There is no standard treatment: it is considered not curable with standard treatments. The possible exception is a hematopoietic stem cell transplant, which is available only to a small number of young and healthy people. It's a good idea to enroll in a clinical trial if you are eligible.

### Types of treatment

#### Watch and wait or active surveillance

delays treatment until the disease progresses. This applies to about 10 to 15% of people who don't have symptoms when they are diagnosed.

#### Chemotherapy

uses medicine (chemicals) to kill cancer cells. A combination chemotherapy procedure uses two or more chemotherapy drugs.

#### Targeted therapies

include a type of drug therapy to target specific substances on the cancer cell. The drug is often given in pill form and is more commonly used for high-risk people.

#### Hematopoietic stem cell transplantation

transfers a healthy person's (donor) stem cells to your body to slow the growth of the disease. This is an option for some people with B-PLL who have had a remission following initial drug therapy. It is also an option for people who are younger and fitter and have responded to the initial therapy.

### Factors that affect treatment

Discuss your treatment options with your doctor to make sure you understand the benefits and risks of each approach. Your treatment plan is based on:

- Your age and overall health
- Your medical history
- The type and extent of symptoms
- Physical exam and lab test results
- Your preference

**Clinical trials are research studies that aim to improve the care and treatment of people living with cancer.**

**For some people with a blood cancer, a clinical trial may be the best treatment choice. Talk to your healthcare team for more information.**

### Treatment side effects

When you begin treatment for B-PLL, you may have mild to severe side effects, depending on your age, your overall health, and your treatment plan. Side effects can vary from person to person. Most side effects disappear once your treatment ends. New drugs and therapies can help control side effects, such as nausea and vomiting. Speak to your doctor if you are having side effects.

### Common side effects

You may experience side effects such as:

- Aches, diarrhea, and constipation from chemotherapy and radiation
- Fatigue, infections, and low blood pressure from chemotherapy
- Low counts of platelets, red blood cells, and white blood cells from chemotherapy
- Mouth sores, upset stomach, and vomiting from chemotherapy

### Long-term or late effects of treatment

Medical follow-up is important after treatment for B-PLL. You will need blood tests with or without repeat bone marrow tests to determine if you need further treatment. Your medical team should provide you with a care plan listing how often you will need follow-up visits and the tests you will have at those visits.

- **Long-term side effects** are common and can last for months or years after treatment ends. Examples include chronic fatigue, problems concentrating (known as chemo brain), and increased risk of infection.
- **Late effects** are medical problems that do not show up until years after treatment ends. See your doctor to get follow-up care for possible early detection of secondary cancers and fertility problems.



Living with B-PLL can be overwhelming. Seek medical help if you feel “down” or “blue” or do not want to do anything and your mood does not improve over time. These could be signs of depression, an illness that should be treated even when you are undergoing treatment for B-PLL. Treatment for depression has important benefits for people living with cancer. Remember, you are not alone.

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