

Splenic Marginal
Zone Lymphoma
SMZL



WHAT YOU NEED TO KNOW

You or your loved one has been diagnosed with splenic marginal zone lymphoma (SMZL). What does it mean and how will it affect you?

This fact sheet will help you:

Learn about
SMZL and how
it is diagnosed

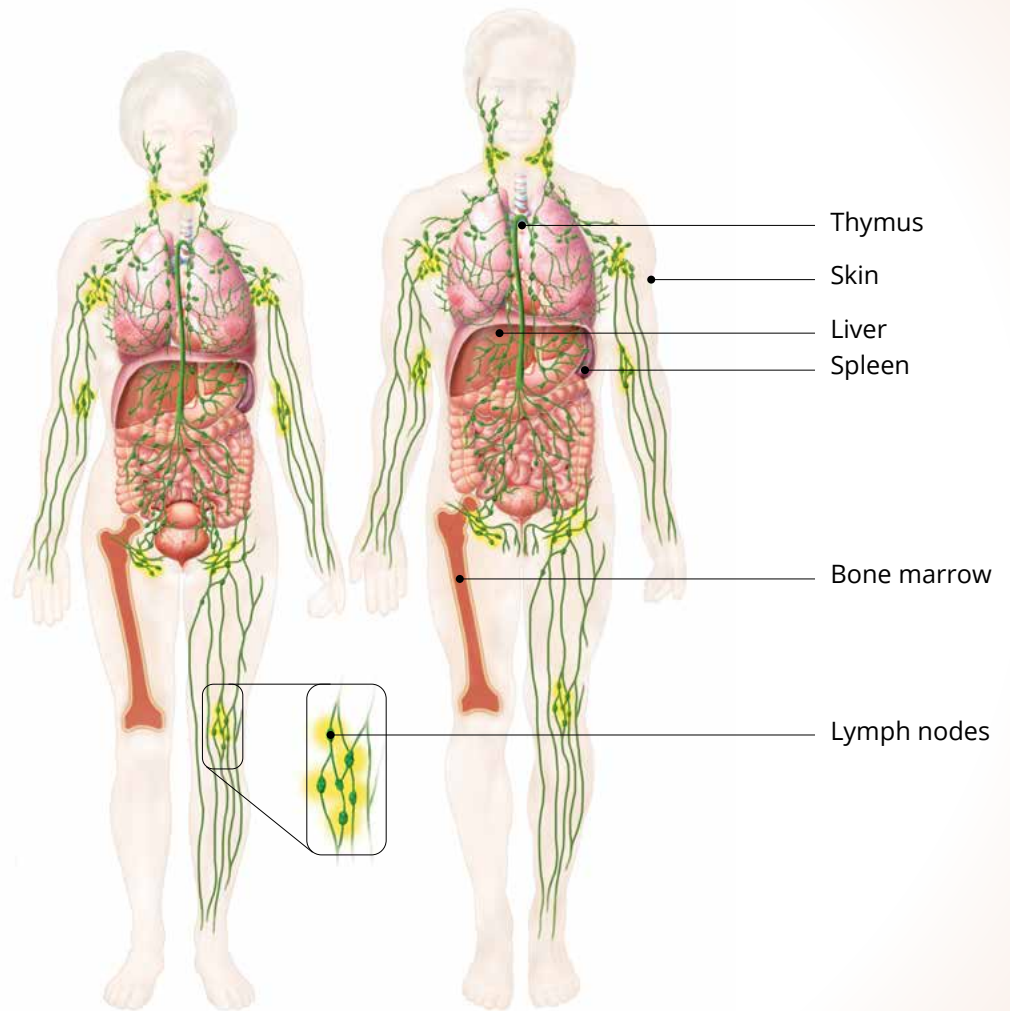
Get an overview
of treatment
options

Understand what
happens next

About lymphoma

Lymphoma is cancer of the lymphatic system, which includes your bone marrow, lymph nodes, thymus, liver, skin, and spleen.

Your lymphatic system defends your body against infection by creating white blood cells called **lymphocytes**. If these cells become abnormal, you may develop lymphoma.



What is lymphoma?

Lymphoma is the name for a group of blood cancers that develop in your lymphatic system. The two main types are Hodgkin lymphoma (HL) and non-Hodgkin lymphoma (NHL).

About SMZL

- SMZL is a rare, slow-growing (indolent) B-cell subtype of NHL
- It is one of three subtypes of marginal zone lymphoma
- It often involves the spleen, bone marrow, blood, and abdomen
- It typically affects adults over age 60
- It can be linked to autoimmune conditions or viral infections, including hepatitis C
- It can change into a fast-growing type of NHL (not common)

Signs and symptoms

In some people with SMZL, there are no obvious signs or symptoms (they are asymptomatic). The disease may be detected during a doctor's visit with a routine blood test.

You may experience:

- Feeling bloated, full, or unable to finish a meal
 - When you have an enlarged spleen
- Abdominal pain when your spleen is enlarged
- Fatigue and weakness
 - When your red blood cell count is low (anemia)
- Infection
 - When your white blood cell count is low (neutropenia)
- Bruising easily and ongoing bleeding from minor cuts
 - When your platelet count is low (thrombocytopenia)
- Fevers, night sweats, or unexplained weight loss
 - Due to your lymphoma

Your diagnosis

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

Name of test	Description
Medical history and physical exam	The doctor will review past illnesses, injuries, and symptoms, examining your lungs, heart, and other organs.
Blood tests	Blood tests help determine your need for treatment and the extent of your disease. They also help identify several non-Hodgkin lymphoma subtypes.
Bone marrow biopsy	This test will confirm whether the lymphoma has spread to your bone marrow. This will help your doctor determine the benefits of specific therapies.
Imaging tests	In these tests, a computed tomography (CT) scan uses a computer linked to an X-ray machine to make a series of detailed pictures of areas inside your body.
Lymph node biopsy	In this test, a sample of the tumour or lymph node is used to look at the size, shape, and arrangement of the lymphoma cells. This may be necessary in some cases for diagnosis and treatment.
Splenectomy	This is a surgical procedure to remove the spleen, an organ located in the upper part of the abdomen. It is performed to help with discomfort and to improve blood cell counts. In some cases, it may be necessary for diagnosis and treatment.

Stages of SMZL

Identifying the stage of your disease is an important step in planning your treatment. The stage of lymphoma refers to where your disease is located and how much of it is in your body.

Your doctor will determine the stage of your disease using imaging, lab tests, and a physical exam to:

- Figure out your lymphoma subtype, such as SMZL
- Monitor how it progresses

Stages I and II are early stages. Stages III and IV are advanced. Many people with SMZL are diagnosed at an advanced stage.

Stages

Stage I	The lymphoma is in one group of lymph nodes or in one extranodal site (outside the lymph nodes).
Stage II	The lymphoma is in two or more groups of lymph nodes on the same side of your diaphragm
Stage III	The lymphoma is in lymph nodes above and below your diaphragm
Stage IV	The lymphoma is found in many areas of your body (in the lymph nodes and beyond)

SMZL treatment

Treatment will help manage the symptoms and complications of SMZL, including infections and fatigue. The types of treatment for SMZL can vary widely.

Types of treatment

- **Active surveillance** (watch and wait) delays treatment until the disease progresses. This approach is for people with slow-growing SMZL who have no symptoms.
- **Monoclonal antibody immunotherapies** are lab-made antibodies that target lymphoma and boost the immune system to help the body fight cancer. These antibodies may be used alone or in combination with chemotherapy. They are given to help symptoms resulting from an enlarged spleen or low blood counts.
- **Splenectomy** is the removal of your spleen, an organ found in your upper abdomen. This surgery may be used to help with discomfort and to improve blood cell counts.
- **Radiation therapy** uses X-rays or other high-energy rays to kill lymphoma cells.
- **Chemotherapy** uses medicine (chemicals) to kill cancer cells. It is often given in combination with immunotherapy.
- **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 may be useful when the disease has relapsed after other treatments.

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.



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 symptoms and blood counts
- The stage of SMZL

Treatment side effects

When you begin your treatment for SMZL, you may experience mild to severe side effects. This will depend on your age, your overall health, and your treatment plan. Side effects can affect people in different ways. Most side effects disappear once your treatment ends. New drugs and therapies can help control most side effects. Speak to your doctor if you are having side effects.

Common side effects

You may experience side effects such as:

- Infusion reactions (fever, wheezing, hives, racing heart) or infections due to immunotherapy
- Nausea, diarrhea, vomiting, loss of appetite, brain fog from chemotherapy or radiation treatments
- Infection from a decrease in white blood cells that can cause side effects such as fever or chills, coughing, sore throat, frequent/loose bowel movements, mouth sores, and rashes

Long-term or late effects of treatment

Medical follow-up is important after treatment for SMZL. You may need blood tests, bone marrow tests, or imaging 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 which tests you will have at those visits.

- **Long-term side effects** can last for months or years after treatment ends. Examples include fatigue or infections.
- **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 heart disease, secondary cancers, fertility problems, thyroid problems, problems concentrating, or chronic fatigue.





Living with SMZL can be overwhelming. Seek medical help if you feel “down” or “blue” or don’t 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’re undergoing treatment for SMZL. Treatment for depression has important benefits for people living with cancer. Remember, you are not alone.

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