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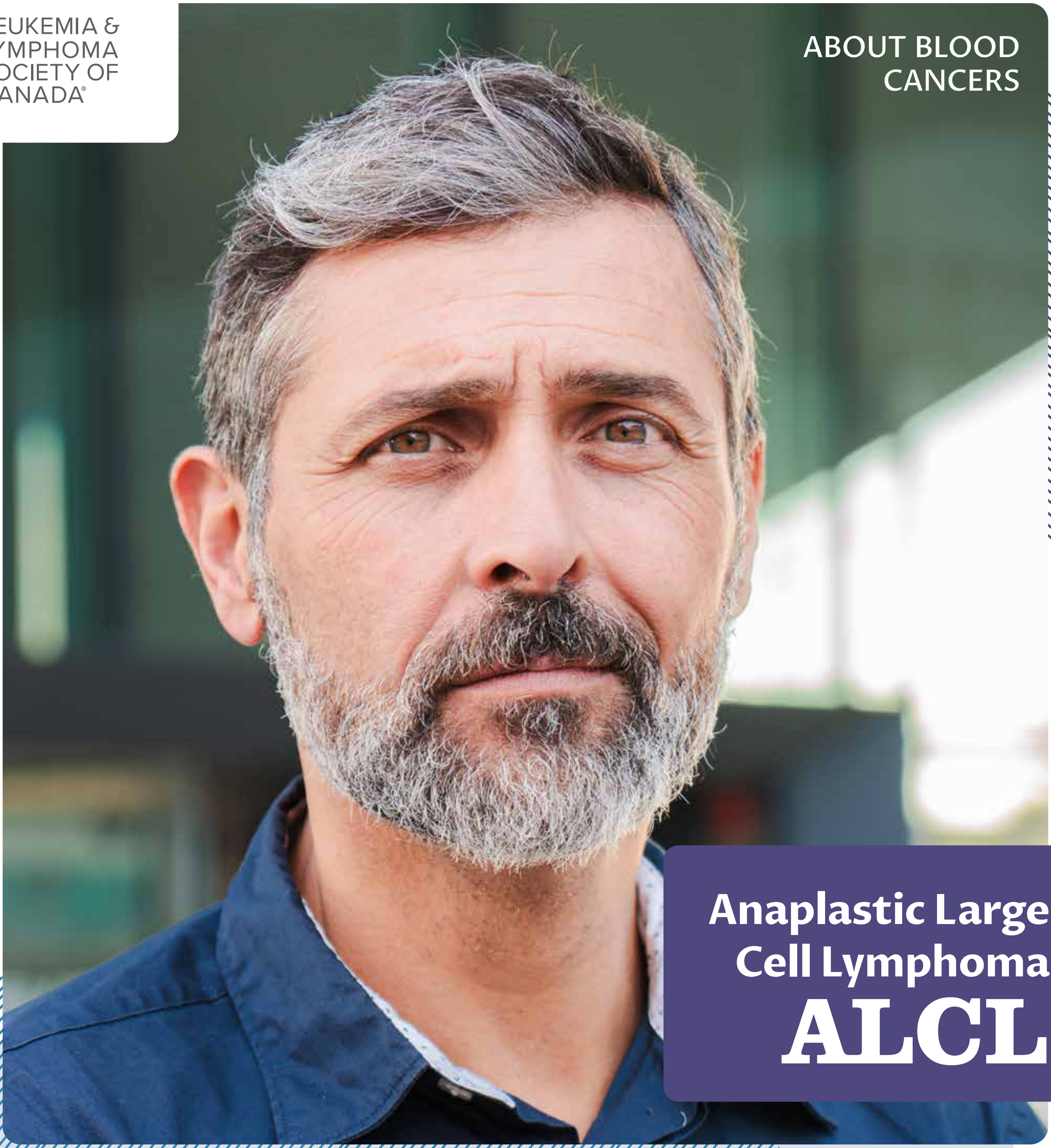
ABOUT BLOOD
CANCERS



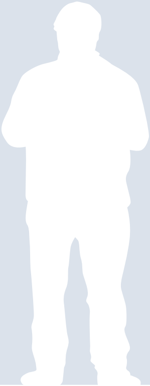
Anaplastic Large Cell Lymphoma ALCL

WHAT YOU NEED TO KNOW

You or your loved one has been diagnosed with anaplastic large cell lymphoma (ALCL). What does it mean and how will it affect you?

This fact sheet will help you:

- Learn about ALCL and how it is diagnosed
- Get an overview of treatment options
- Understand what happens next

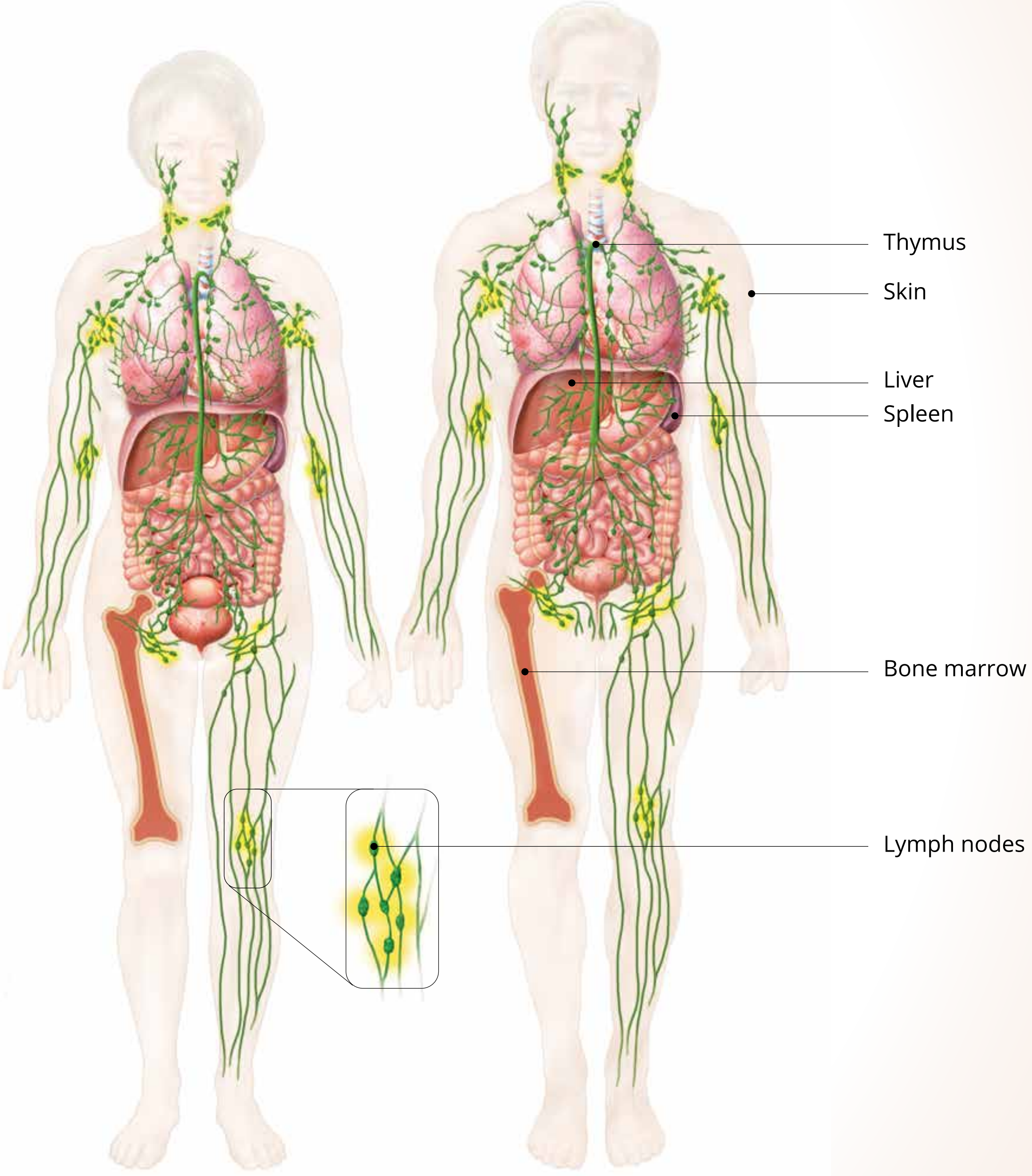


About lymphoma

Lymphoma is the most common form of blood cancer.

Lymphoma is cancer of the lymphatic system, which includes the 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 of a group of blood cancers that develop in your lymphatic system. The two main types are Hodgkin lymphoma (HL) and non-Hodgkin lymphoma (NHL). Anaplastic large cell lymphoma is a subtype of NHL.

About ALCL	• It is a rare form of lymphoma • It accounts for about 15% of NHLs in children and young adults (about 2% of adult NHL cases) • It involves white blood cells (lymphocytes) called T cells that grow abnormally • It can be fast-growing (aggressive), depending on the type • It is more common in men

Subtypes of ALCL

Systemic ALCL

Systemic ALCL is fast growing and affects more than one part of the body, including lymph nodes and organs. There are two subtypes. The difference depends on a genetic change that causes cells to make too much of a protein known as anaplastic lymphoma kinase (ALK).

The two subtypes of systemic ALCL include:

ALK-positive ALCL	• Abnormal type of protein drives uncontrolled cell growth • It is more common in children and young adults • Usually responds well to chemotherapy treatments
ALK-negative ALCL	• Abnormal protein is not present • More common in adults over 50 years of age (more common in men) • Often returns after a short remission and may need more aggressive therapy

Primary cutaneous ALCL

Develops in the skin, causing bumps or rashes. It is slow growing and is more common in adults over 40.

Breast implant-associated ALCL

Forms around breast implants and is very rare. It is slow growing and is more common in adults over 40.

Symptoms of ALCL

Systemic ALCL may affect the lymph nodes and other organs such as the liver, lungs, and bones. You may experience:	• Painless swelling in your neck, armpits, or groin - When your lymph nodes are enlarged or swollen • Nausea, stomach (abdomen) pain, bloating, and diarrhea - When the lymphoma is found in the abdomen • Fatigue and weakness - When your red blood cell count is low (anemia) • Shortness of breath, coughing, and chest pain - When the lymphoma is in the chest • Backache - When expanding lymph nodes are placing pressure on nerves • Skin rash - A response from your immune system • Fever and night sweats - Possibly a response from your immune system • Weight loss and loss of appetite - When you are eating less or using more energy
Primary cutaneous ALCL affects the skin and usually never spreads beyond it. You may experience:	• Bumps or a rash on the skin that may become itchy and get bigger over time - When abnormal cells multiply in the skin • Bumps that form a sore and scab over - When the skin becomes irritated from the abnormal growth of cells
Breast implant-associated ALCL (BIA-ALCL) forms around textured breast implants. It may happen 10 years or more after reconstruction or implants. You may experience:	• Swelling - When fluid builds up around the implant • Pain - When irritation, fluid buildup, or scarring occur around the implant

Your diagnosis

With a diagnosis, your healthcare team can determine the right treatment for you. Your healthcare team includes an oncologist (the primary cancer doctor), who works with a range of professionals such as your family doctor, nurse practitioners, nurses, pharmacist, and social worker to manage your treatment and provide support. Depending on the type of cancer, other specialists like a pathologist, radiologist, or surgeon may also be involved in your care.

Your test results help your healthcare team predict how ALCL will likely progress and how you may respond to treatment.

Here are some possible tests you may undergo:

Name of test	Description
Medical history and physical exam	Your healthcare provider will review past illnesses, injuries, and symptoms and will examine your lungs, heart, and other organs.
Blood tests	Blood tests help determine the need for treatment and the extent of the disease. They also help identify the ALCL subtype.
Tissue biopsy	A biopsy is done to confirm an ALCL diagnosis. A small tissue sample is taken from the swollen lymph node or affected body area and examined under a microscope to determine the type of ALCL.
Bone marrow biopsy	This test confirms whether the lymphoma has spread to the bone marrow. It will help your healthcare team determine the benefits of specific therapies.
Imaging tests	**Computed tomography (CT)** uses a computer linked to an X-ray machine to make detailed pictures of areas inside your body. **Positron emission tomography (PET)** uses radioactive material to create a 3D image of your body. It can identify whether lymphoma cells are in the bone marrow and other parts of your body. **Magnetic resonance imaging (MRI)** uses magnetic fields and radio waves to create images of the body's organs, including the brain and tissues.
Immunophenotyping	Immunophenotyping helps find specific types of cells within a blood sample to confirm a diagnosis. It identifies the lymphoma cells as B cells, T cells, or natural killer cells.

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.

Stages of ALCL

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

Your healthcare team will use imaging, lab tests, and a physical exam to determine the stage of the disease, figure out the lymphoma subtype, and monitor how it progresses.

Stages I and II (1 and 2) are early stages. Stages III and IV (3 and 4) are advanced.

Stage I	The lymphoma is in one group of lymph nodes or 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 the diaphragm
Stage III	The lymphoma is in lymph nodes above and below the diaphragm
Stage IV	The lymphoma is found in many areas of the body (in the lymph nodes and beyond)

ALCL treatment

New treatment approaches will help manage your symptoms and complications, including infections and fatigue. The type of treatment for ALCL depends on the subtype and how aggressive it is. Treatment types can vary widely and could include clinical trials.

You may have mild to severe side effects during treatment, depending on your age, overall health, and treatment plan. If you do, let your healthcare team know.

Side effects can affect people in different ways. Most side effects improve or go away after treatment ends. New drugs and therapies can help control most side effects.

Types of treatment

Common SLL treatments and possible side effects include:

- **Chemotherapy** uses medicine (chemicals) to kill cancer cells. It also affects your body's healthy, fast-growing cells (such as hair, nails, and blood cells). Chemotherapy is often given in combination with immunotherapy. It is the standard treatment for systemic ALCL and often eliminates cancer in the short term.
  - **Potential side effects:** low blood cell counts (white, red, and platelets), infection, bleeding, anemia, nausea, diarrhea, vomiting, loss of appetite, brain fog (chemo brain), fatigue, shortness of breath, diseases or disorders affecting the heart (cardiopathy), temporary hair loss, mouth sores, rashes, secondary cancers, and nerve damage (neuropathy)
- **Radiation therapy** uses high-energy X-rays or other types of radiation to kill cancer cells. It may be combined with other treatments, such as chemotherapy. Radiation therapy is a key treatment for primary cutaneous ALCL, depending on how many areas of the skin are affected. If the bumps or sores are in multiple areas of the body, your healthcare team may recommend chemotherapy.
  - **Potential side effects:** redness, dryness, itching, blistering, nausea, diarrhea, vomiting, loss of appetite, headaches, swelling, fatigue, shortness of breath, cardiopathy, temporary hair loss, and secondary cancers
- **Surgery** is another key treatment for primary cutaneous ALCL, depending on how many areas of the skin are affected. Surgery is the main treatment used to remove the breast implant and surrounding cancer cells for BIA-ALCL. If the cancer cells have spread beyond the breast or return after surgery, your healthcare team may recommend chemotherapy or radiation.
  - **Potential side effects:** infection, scarring, pain, bleeding, and nerve damage
- **Targeted therapies** are a type of drug therapy that targets specific substances on cancer cells. These drugs are often given in pill form. This type of therapy is used to treat ALK-positive systemic ALCL. Targeted therapy may also be given when ALCL relapses or is resistant to other treatments. Some targeted therapies are available in Canada; others are only available through a clinical trial.
  - **Potential side effects:** low blood cell counts (white, red, and platelets), infection, bleeding, anemia, skin problems, high blood pressure, fatigue, diarrhea, neuropathy, and slower healing time for wounds
- **Immunotherapy** uses an intravenous drug that can either boost or pause your immune system to help your body fight cancer. Immunotherapy is done in addition to chemotherapy. In Canada, it may be available in your province or territory or through a clinical trial.
  - **Potential side effects:** rashes, fatigue, diarrhea, nausea, vomiting, and decreased thyroid hormone levels
- A **stem cell transplant** (SCT) gives you healthy stem cells to replace those damaged by cancer or chemotherapy and radiation treatments. Your body relies on stem cells to produce blood cells. If your healthcare team thinks the ALCL may return, they may recommend a stem cell transplant while you are in remission.

Types of treatment (cont'd)

Two main types of stem cell transplant are used to treat blood cancers:

- **Autologous:** The stem cells are removed from your body before treatment and placed back into your body after treatment. This allows you to receive high doses of chemotherapy (sometimes with radiation). Using your own stem cells helps your bone marrow produce new blood cells and reset your immune system.
  - **Potential side effects:** low white blood cell count (increased risk of infection), low platelet count (increased risk of bleeding or bruising), low red blood cell count (causes fatigue, dizziness, shortness of breath, and feeling unwell), pain and issues with your digestive system, skin and hair problems, and issues with your organs or central nervous system
- **Allogeneic:** The stem cells come from a donor. These donor cells replace the damaged ones in your bone marrow, potentially offering a long-term cure.
  - **Potential side effects:** low white blood cell count (increased risk of infection), low platelet count (increased risk of bleeding or bruising), low red blood cell count (fatigue, dizziness, shortness of breath, feeling unwell), pain and issues with your digestive system, skin and hair problems, issues with your organs or central nervous system, and possible graft-versus-host disease (GvHD) or veno-occlusive disease (affecting the small vessels leading to your liver)

Factors that affect treatment

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

- Your age and overall health
- ALCL subtype and stage
- The status of the disease (first diagnosis or relapse, which is when the cancer returns after initial treatment)
- Any other health issues you may have
- Your lifestyle and preferences

Long-term or late effects of treatment

Medical follow-up is important after treatment for ALCL. You may need blood, bone marrow, or imaging tests to determine if you need further treatment. Your healthcare team should provide a care plan listing how often you will need follow-up visits and which tests you will have at those visits.

You may experience long-term or late effects of your treatment:

- **Long-term side effects** can last for months or years after treatment ends. Examples include fatigue, fertility issues (the ability to conceive and have children), secondary cancers, and neuropathy.
- **Late effects** are medical problems that do not show up until years after treatment ends. See your healthcare team to get follow-up care for possible early detection of heart disease, secondary cancers, fertility issues, thyroid problems, trouble concentrating, or chronic fatigue.

Living with ALCL 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 ALCL. Treatment for depression has important benefits for people living with cancer. Remember, you are not alone.

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