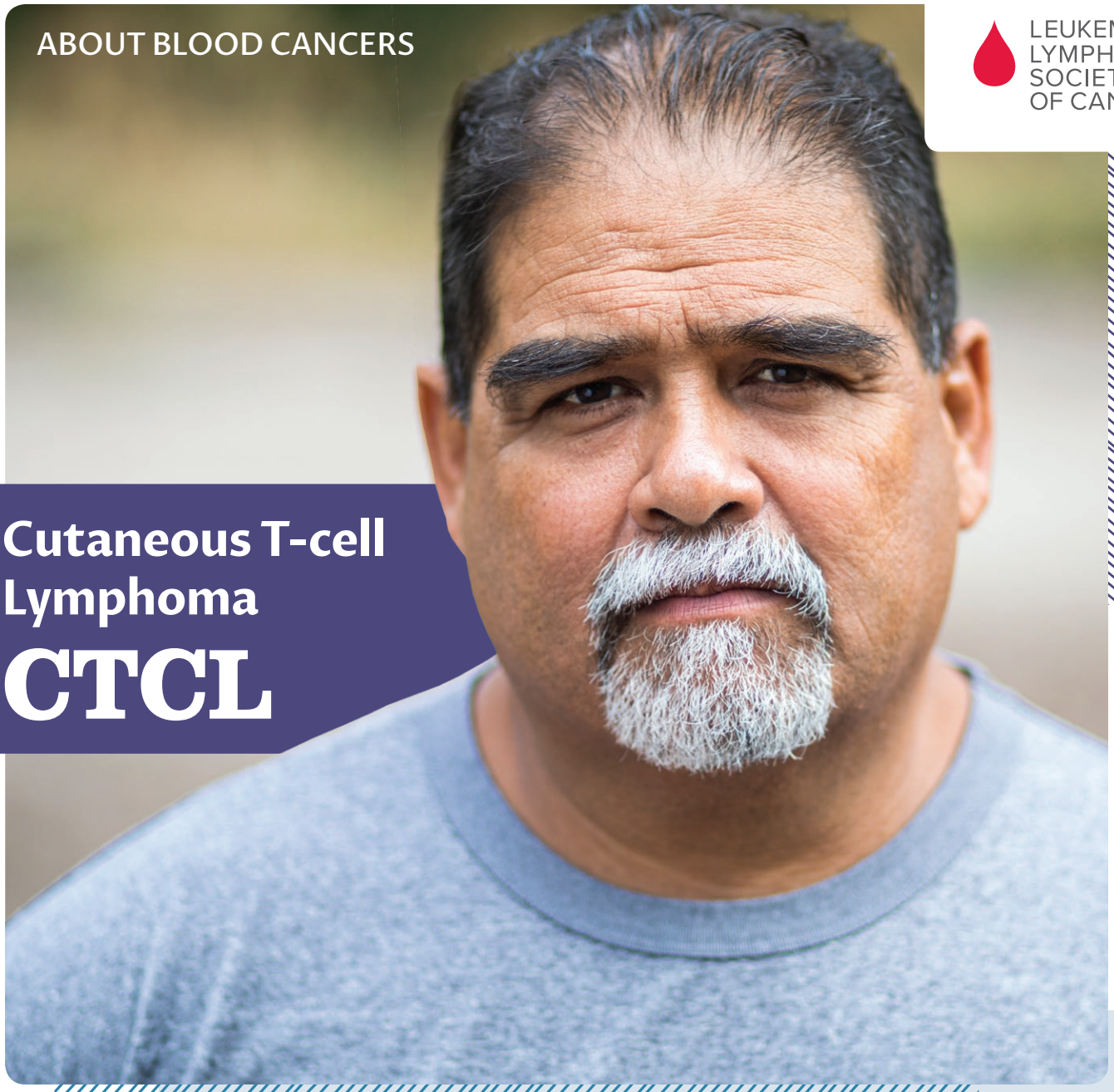
LEUKEMIA & LYMPHOMA SOCIETY OF CANADA®



Cutaneous T-cell Lymphoma

CTCL

WHAT YOU NEED TO KNOW

You or your loved one has been diagnosed with cutaneous T-cell lymphoma (CTCL). What does it mean and how will it affect you?

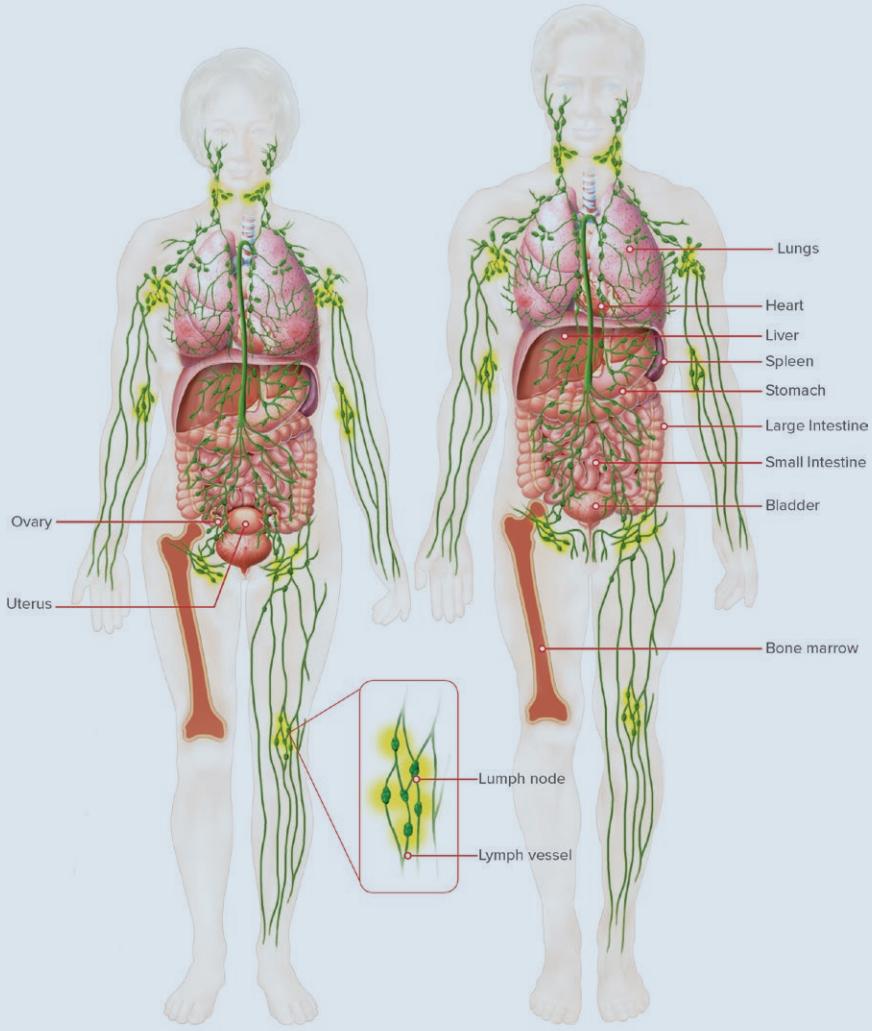
This fact sheet will help you:

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

About lymphoma

Lymphoma is cancer of the lymphatic system. This system 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 and non-Hodgkin lymphoma. CTCL is a type of non-Hodgkin lymphoma.

About CTCL

- CTCL is a slow-growing (indolent) subtype of non-Hodgkin lymphoma.
- 4% of all non-Hodgkin lymphoma cases are CTCL.
- CTCL involves lymphocytes called T-cells.
- CTCL develops from a malignant (cancerous) change in a T cell in the skin.
- T-cells start growing uncontrollably and build up in the skin, causing skin lesions.
- The two most common types of CTCL are mycosis fungoides and Sézary syndrome.
- CTCL can happen at any age, but usually appears in adults aged 50 to 60.
- CTCL is as twice as common in men.

Signs and symptoms

The signs and symptoms vary depending on the type of CTCL you have. This fact sheet focuses on mycosis fungoides (MF) and Sézary syndrome (SS).

Both can feature skin lesions, including:

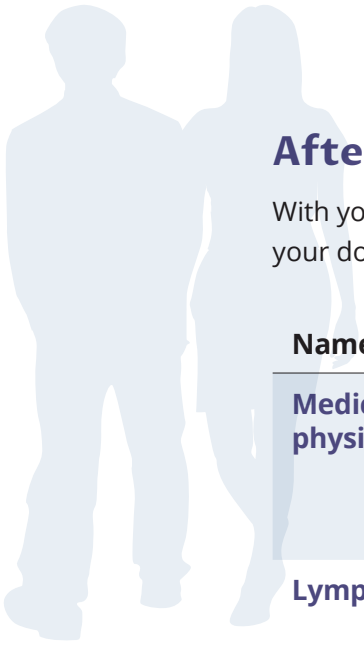
- Patches: flat, scaly pink or red areas.
- Plaques: abnormal thick patches that are raised or hard.
- Tumours: solid, dome-shaped masses that are at least 1 cm in diameter.
- Erythroderma: redness covering more than 80% of your skin.

With **mycosis fungoides (MF):**

- Papules: small, solid raised bumps by hair follicles.
- Skin lesions can appear when your T-cells start growing and building up in your skin.
- There can be one lesion, or lesions can appear on a larger area of skin.

With **Sézary syndrome (SS),** you may also experience:

- Large masses in your neck, armpit, or groin and/or painless swelling in one or more lymph nodes.
- Skin swelling, thick palms (hands) and soles of your feet, abnormal fingernails and toenails, hair loss.
- High white blood cell count.



After your diagnosis

With your diagnosis, your doctor can determine the right treatment for you. Your test results help your doctor predict how CTCL will likely progress and how you may respond to treatment.

Name of test	Description
Medical history and physical exam	The doctor reviews past illnesses, injuries, and symptoms. They examine your lungs, heart, and other organs. They will examine your skin to determine the type of skin lesions and how much of your skin is affected.
Lymph node biopsy	A biopsy draws out a sample of the tumour or lymph node. This is used to look at the size, shape, and arrangement of the lymphoma cells.
Complete blood cell count (CBC)	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.
Sézary screen	This test identifies the number of abnormally shaped Sézary cells in your blood.
Skin biopsy	This test studies the size and shape of the cells in your skin. It looks at how they are arranged in your skin's layers and around things like your hair follicles.

Lactate dehydrogenase (LDH)	TLDH is an enzyme in your blood. A high level may be a sign of tissue damage and a possible aggressive lymphoma.
Immunophenotyping	This test helps find specific types of cells in a blood sample to confirm a non-Hodgkin lymphoma diagnosis. It will also identify lymphoma cells as B-cells, T-cells, or natural killer cells.
Imaging 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. **Magnetic resonance imaging (MRI)** uses magnetic fields and radio waves to create images of the body's organs and tissues. The **positron emission tomography (PET) scan** uses radioactive material to create a 3D image of your cells to look for changes in the bone marrow and pockets of lymphoma cells.
Molecular testing	These DNA tests identify specific genetic mutations (changes) in CTCL cells. In people with MF and SS, there are often changes in specific genes.

Stages of CTCL

Identifying the stage of your disease is an important step in planning your treatment. The stage of CTCL refers to how your disease has progressed. **It does not determine how well you will respond to treatment.**

Your doctor will determine the stage of your disease using imaging, lab tests, and a physical exam of your body. This helps them find out:

- If your CTCL has spread
- If it has spread, how far

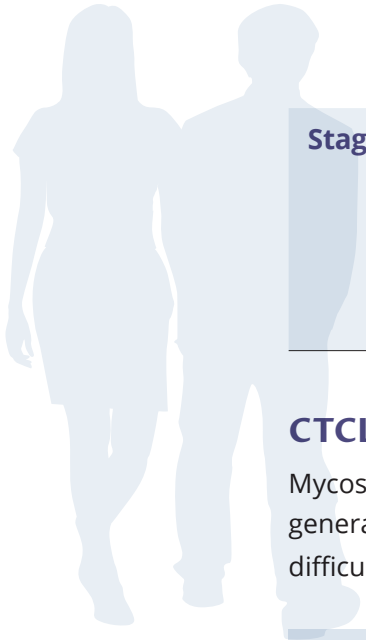
Staging for MF and SS is based on four factors:

- **T (tumour):** percentage of your skin that is affected by the lymphoma and the type of lesions
- **N (node):** if lymph nodes are involved and if they contain cancer cells
- **M (internal organs):** if your organs are affected by the cancer
- **B (blood):** number of lymphoma cells in your blood

These four factors are then grouped together to find out what your stage is:

Name of stage	Description
Stage IA	• Skin lesions (patches, papules, and/or plaques) cover less than 10% of the skin surface. No skin tumours. • Lymph nodes are not enlarged. Lymphoma cells have not spread to other organs. • Blood contains few or no Sézary cells.
Stage IB	• Skin lesions cover 10% or more of the skin surface. No skin tumours. • Lymph nodes are not enlarged. Lymphoma cells have not spread to other organs. • The number of Sézary cells in the blood is low.
Stage IIA	• Skin lesions cover up to 80% of the skin surface. No skin tumours. • Lymph nodes are enlarged. They do not contain cancerous cells. • Lymphoma cells have not spread to other organs. • The number of Sézary cells in the blood is low.
Stage IIB	• At least one of the skin lesions is a tumour. • Lymph nodes may be enlarged but do not contain cancerous cells. • Lymphoma cells have not spread to other organs. • The number of Sézary cells in the blood is low.
Stage IIIA	• Skin lesions cover at least 80% of the skin. • Lymph nodes are either normal or enlarged. They do not contain cancerous cells. • Lymphoma cells have not spread to other organs. • The number of Sézary cells in the blood is low.
Stage IIIB	• Skin lesions cover at least 80% of the skin, and the skin is reddened. • Lymph nodes may be enlarged. They do not contain cancerous cells. • Lymphoma cells have not spread to other organs. • The number of Sézary cells in the blood is low.

Stage IVA$_1$	• Skin lesions cover any amount of skin. • Lymph nodes are either normal or enlarged. Cells do not look very abnormal under the microscope. • Lymphoma cells have not spread to other organs. • The number of Sézary cells in the blood is high.
Stage IVA$_2$	• Skin lesions cover any amount of skin. • Some lymph nodes are enlarged. Cells appear abnormal under the microscope. • Lymphoma cells have not spread to other organs. • The number of Sézary cells in the blood is either high or low.
Stage IVB	• Skin lesions cover any amount of skin. • Lymph nodes are either normal or abnormal. • Lymphoma cells have spread to other organs. • Sézary cells may or may not be in the blood.



CTCL treatment

Mycosis fungoides (MF) and Sézary syndrome (SS) are chronic conditions. Although they are generally considered incurable, they are treatable and are usually not life-threatening. SS is more difficult to treat.

Your treatment is based on the stage of your disease:

- **Early-stage disease** may respond well to therapies applied to the skin.
- **Advanced disease** may need a combination of therapies applied to the skin and systemic (entire body) therapies.

Types of treatment

Depending on the stage of your disease, there are three treatment approaches:

Watch and wait or active surveillance	• Delays treatment until the disease progresses. • This approach is for people with MF stage IA with a low risk of the disease progressing.

Therapies applied to the skin	• **Topical corticosteroids** are applied to skin lesions to help relieve red, swollen, and inflamed skin. • **Topical chemotherapy** is applied directly on skin lesions. This is for people with MF stage IA and IB who have already had skin treatment. • **Topical retinoids** can slow the growth of certain types of cancer cells. These are applied directly on skin lesions for people with stage IA and IB CTCL. • **Electron beam therapy** uses x-rays and other high-energy rays to treat skin lesions. It can be given locally (in one area) or over the entire skin surface, depending on your stage of CTCL. • **Phototherapy** (light therapy) uses ultraviolet light, which is made up of ultraviolet A and B rays. A special lamp or laser directs the radiation beam to a specific area on your skin.
Systemic therapies (affecting the entire body)	• **Oral retinoids** are used to treat skin inflammation. • **Interferons** boost your immune system to fight cancer. • **Histone deacetylase inhibitors** cause cancer cells to stop dividing and die. • **Monoclonal antibodies** are drugs that use your immune system to target the cancer cells. • **Targeted therapies** target a specific molecule on the cancer cell, killing the cancer cells and not affecting the healthy cells. • **Systemic chemotherapy** is often given to people with advanced stage MF or SS whose disease has relapsed (returned) many times and who have few other treatment options.

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 prognosis (the likely outcome of the disease)
- The stage of your disease
- How much of your skin is affected and the type of lesions you have
- If your disease has spread from your skin to the lymph nodes or other organs (extracutaneous)
- The level of Sézary cells in your blood
- The level of lactate dehydrogenase in your blood
- The presence of large-cell transformation or folliculotropic MF (a subtype of MF that involves your hair follicles)

Treatment side effects

When you begin your treatment for CTCL, you may experience reactions to it. 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:

- Nausea and fatigue
- Fever, chills, and dizziness
- Blood clots
- Infertility
- Severe birth defects from retinoid treatment. Women of childbearing age must use birth control when taking these medications.

Living with CTCL

Medical follow-up is important with CTCL. Your medical team should provide you with a care plan with information on how often you will need follow-up visits and the tests you will have at those visits.

Be sure to attend follow-up visits with your doctor. Schedule an appointment if you have concerns. This can help with early detection of heart disease and secondary cancers.

Living with CTCL can be overwhelming. Seek medical help if you are feeling "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 CTCL. Treatment for depression has important benefits for people living with cancer. Remember, you are not alone.

This fact sheet was reviewed by:

Alyson Parker, RN, MSc(A)
Clinical Nurse Educator
Leukemia/Bone Marrow Transplant Program
of British Columbia
Vancouver General Hospital