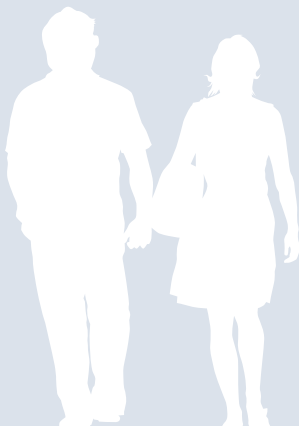




Cutaneous T-cell
Lymphoma
CTCL



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

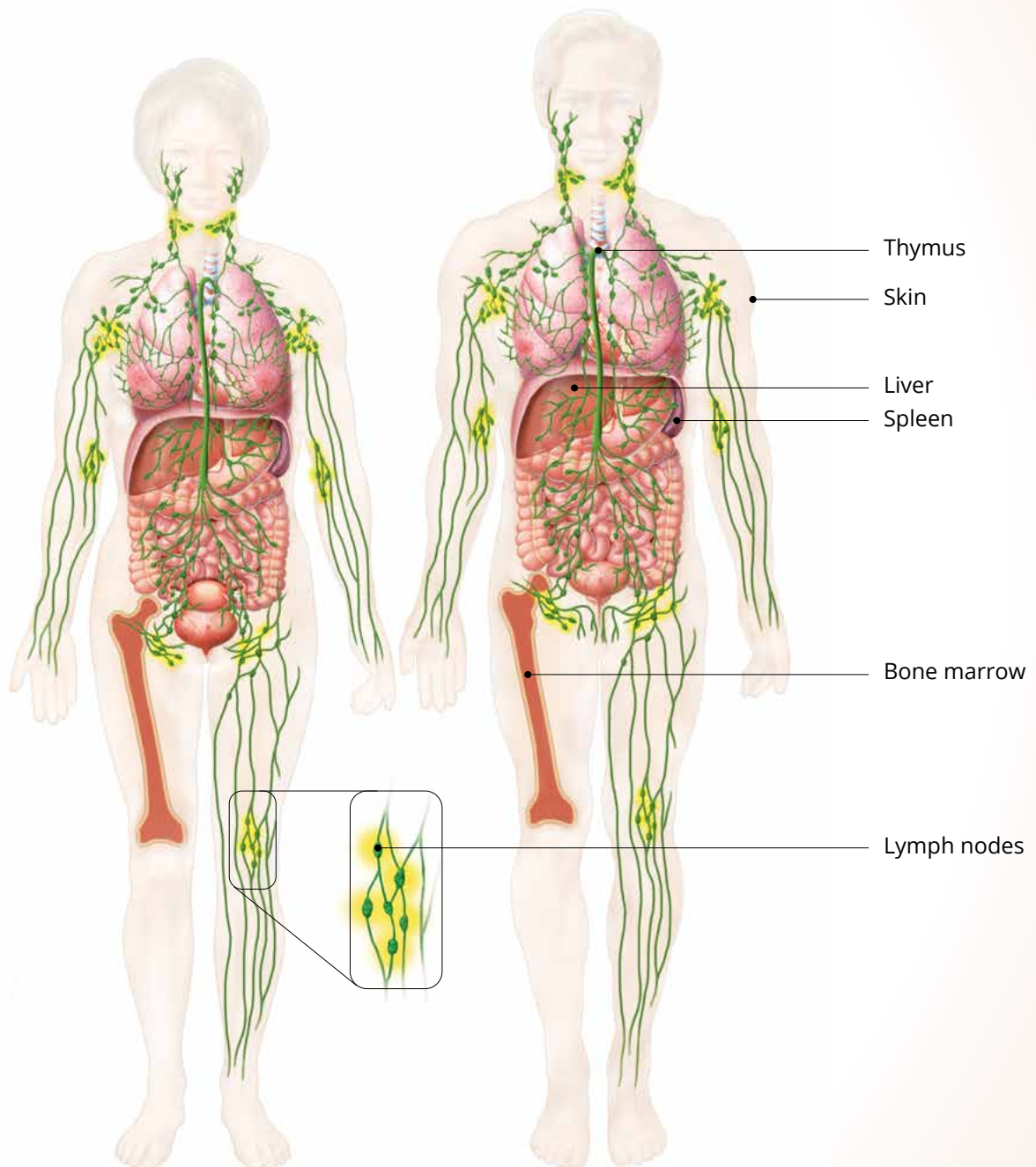
Get an overview
of treatment
options

Understand
what happens
next

About lymphoma

Lymphoma is a 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).

About CTCL

- CTCL is a slow-growing (indolent) subtype of NHL
- It is rare, representing 4% of NHL cases
- It starts in white blood cells (lymphocytes) that reside in the skin, called T-cells
- CTCL develops when T-cells start growing uncontrollably and build up in the skin, causing skin lesions
- The two most common types (60–80% of cases) are mycosis fungoides (MF) and Sézary syndrome (SS)
- MF is generally a chronic condition that requires ongoing care and is usually not life-threatening
- SS is a leukemic form of CTCL that is not chronic
- CTCL may progress to a more advanced stage
- It can happen at any age, but usually appears in adults aged 50 to 60
- CTCL is twice as likely to affect men than women

Symptoms of CTCL

The symptoms vary depending on the type of CTCL you have. This fact sheet focuses on MF and SS. Both types can feature skin lesions.

With **MF**, you may experience:

- Flat, scaly pink, or red skin patches
- Small, solid raised bumps at the hair follicles (papules)
- Abnormal thick skin patches that are raised or hard (plaques)
- Solid, dome-shaped masses (tumours) that are at least 1 cm in diameter
- Redness covering more than 80% of your skin (erythroderma)

With **SS**, you may experience:

- Large masses in your neck, armpit, or groin
- Painless swelling in one or more lymph nodes
- Skin swelling, thick palms, and thick soles of the feet
- Abnormal fingernails and toenails
- Hair loss
- High white blood cell count
- Difficulty controlling body temperature



Your diagnosis

With a 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.

Here are some possible tests you may undergo:

Name of test	Description
Medical history and physical exam	The doctor reviews past illnesses, injuries, and symptoms, and will examine your lungs, heart, and other organs. The doctor will examine your skin lesions.
Lymph node biopsy	A sample of the tumour or lymph node is examined 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 (cancerous T cells) in your blood.
Skin biopsy	This test studies the size and shape of the cells in your skin. It looks at how the cells are arranged in your skin's layers and around things like your hair follicles.
Lactate dehydrogenase (LDH)	LDH 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 within a blood sample to confirm a diagnosis. It identifies the lymphoma cells as B cells, T-cells, or natural killer cells.
Imaging tests	<p>Computed tomography (CT) scan uses a computer linked to an X-ray machine to make a series of detailed pictures of areas inside the body.</p> <p>Magnetic resonance imaging (MRI) uses magnetic fields and radio waves to create images of the body's organs, including the brain and tissues.</p> <p>Positron emission tomography (PET) uses radioactive material to create a 3D image of the body. It can identify whether lymphoma cells are in the bone marrow and other parts of the body.</p>
Molecular testing	These DNA tests identify specific changes (genetic mutations) in CTCL cells. In people with MF and SS, there are often changes in specific genes.



Stages of CTCL

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 your body.

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

- Figure out the lymphoma subtype, such as CTCL
- Determine if the CTCL has spread and, if it has, how far

The stage of the disease depends on:

- The extent of tumours present
- If lymph nodes are involved and if they contain cancer cells
- If it has spread to other internal organs
- The amount of lymphoma cells in your blood

CTCL treatment

Most cases of CTCL are chronic and can often be managed with local treatment. Sézary syndrome (SS) can be more challenging to treat and may need more care, including chemotherapies.

You may experience mild to severe side effects during treatment, depending on your age, overall health, and treatment plan. If so, let your doctor 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.

New treatment approaches might help manage CTCL symptoms and complications, including infections and fatigue. The types of treatment can vary widely and could include clinical trials.

Your treatment is based on the stage of the disease:

- **Early-stage disease** (stages 1 and 2) may respond well to therapies applied to the skin
- **Advanced disease** (stages 3 and 4) may need a combination of therapies applied to the skin and entire body (systemic) therapies

Types of treatment

Common CTCL treatments and possible side effects include:

Watch and wait, or active surveillance

- Delays treatment until the disease progresses.
- This approach is for people with early-stage MF 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.

- **Potential side effects:** thinning of the skin and stretch marks from long-term use

Topical retinoids can slow the growth of cancer cells. They are applied directly on skin lesions for people with early-stage CTCL.

- **Potential side effects:** skin irritation

Topical chemotherapy is applied directly on skin lesions for people with early-stage CTCL who have had skin treatment (topical retinoids).

- **Potential side effects:** redness, itching, swelling, skin ulcers or blisters, infection, and darkening of the skin

Topical immunotherapy modifies immune system activity to help the body destroy cancer cells.

- **Potential side effects:** redness, itching, flaking, scaling, or skin thickening

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. Electron beam therapy, a form of radiation therapy that does not penetrate as deeply, is often used to treat skin lesions.

- **Potential side effects:** redness, dryness, itching, blistering, nausea, diarrhea, vomiting, loss of appetite, headaches, swelling, fatigue, shortness of breath, diseases or disorders affecting the heart (cardiopathy), temporary hair loss, and secondary cancers

Phototherapy (light therapy) uses ultraviolet light to direct the radiation beam to an area on the skin.

- **Potential side effects:** red, painful skin (similar to a sunburn)

Systemic therapies (affecting the entire body)

Extracorporeal photopheresis is a combination of UV light (phototherapy) and white blood cell removal (leukapheresis) that treats T cells outside your body.

- **Potential side effects:** no major side effects (treatment is well tolerated)

Oral retinoids are used to treat skin inflammation.

- **Potential side effects:** high number of fat particles in the bloodstream, low levels of thyroid hormones, dry skin, muscle and joint pain, headaches, and sensitivity to light

Systemic therapies
(affecting the entire body)

Interferons (immunotherapy) boost your immune system to fight cancer.

- **Potential side effects:** low white blood cell count, flu-like symptoms, fatigue, nausea, vomiting, depression, hair thinning, and heart and liver damage

Monoclonal antibodies (immunotherapy drugs) use your immune system to target the cancer cells. One example is mogamulizumab, which is given by infusion (injection into the vein).

- **Potential side effects:** low blood cell counts (white, red, and platelets), infection, infusion-related reactions (chills, fever, heart issues), rashes, issues with the liver and lungs, fatigue, diarrhea, nausea, vomiting, and increased blood sugar levels

Antibody-drug conjugates are a type of targeted therapy that kills cancer cells.

- **Potential side effects:** low white and red blood cell counts, nerve damage to the hands and feet (peripheral neuropathy), nausea, fatigue, constipation, diarrhea, and vomiting

Systemic chemotherapy is often given to people with advanced-stage MF or SS whose disease has returned (relapsed) many times and who do not have many other treatment options.

- **Potential side effects:** fever or chills, fatigue, nausea, loss of appetite, mouth sores, peripheral neuropathy, changes in blood cell counts, infection, rash, vomiting, diarrhea, shortness of breath, swelling, and temporary hair loss

Stem cell transplant

Your body relies on stem cells to produce blood cells. A stem cell transplant (SCT) gives you healthy stem cells to replace those damaged by cancer or intense chemotherapy and radiation treatments.

There are two main types of SCT used to treat blood cancers:

Autologous: The stem cells come from your own body. This allows you to receive high doses of chemotherapy (sometimes with radiation). By using your own stem cells, it helps your bone marrow produce new blood cells and reset your immune system.

Allogeneic: This treatment may be used for post-remission therapy. Doctors transfer a healthy person's stem cells into your body to slow the growth of CTCL and restore your body's ability to make normal cells following chemotherapy.

- **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 the digestive system, skin and hair problems, issues with organs or the central nervous system, and possible graft-versus-host disease (GvHD) or veno-occlusive disease (affecting the small vessels leading to the liver)

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 CTCL
- How much of your skin is affected and the type of lesions you have
- If the 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 an enzyme called LDH (lactate dehydrogenase) in your blood
- The presence of an MF subtype that involves your hair follicles (called large-cell transformation or folliculotropic MF)

Long-term or late effects of treatment

Medical follow-up is important after treatment for CTCL. You may need blood, bone marrow, or imaging tests to determine if you need further treatment. Your medical team will provide a care plan listing follow-up visits and the 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 nerve damage (neuropathy).
- **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 issues, thyroid problems, trouble concentrating, or chronic fatigue.



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

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