National Comprehensive Cancer Network®

2026

NCCN Guidelines for Patients®

Cancer care recommendations from leading experts at the National Comprehensive Cancer Network® (NCCN®)

# Cutaneous T-Cell Lymphomas



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# NCCN Guidelines for Patients®

The essential guide for people facing cancer.

Based on care recommendations from leading cancer experts.

Explains high-quality cancer care provided at state-of-the-art cancer centers.

Reviewed and revised every year.

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Because cancer care is always evolving, NCCN develops and frequently updates evidence-based cancer care recommendations used by health care providers worldwide. These recommendations are known as the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®).

The NCCN Guidelines for Patients plainly explain these expert recommendations, so you can talk with your care team about the best care for you.

**These NCCN Guidelines for Patients are based on the NCCN Guidelines® for Cutaneous Lymphomas, Version 2.2026 — February 13, 2026**

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# Contents

4 Chapter 1
About CTCL

8 Chapter 2
Testing for CTCL

19 Chapter 3
Cancer staging

25 Chapter 4
Types of treatment

34 Chapter 5
Mycosis fungoides

45 Chapter 6
Sézary syndrome

50 Chapter 7
CD30+ PCTLD

56 Chapter 8
Subcutaneous panniculitis-like T-cell lymphoma

62 Chapter 9
Primary cutaneous indolent LPD

65 Chapter 10
Other resources

69 Words to know

72 NCCN Contributors

73 NCCN Cancer Centers

74 Index

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# 1
# About CTCL

5 What is cutaneous T-cell lymphoma?

6 How is CTCL treated?

7 How can I get the best care?

Cutaneous T-cell lymphoma (CTCL) develops when abnormal lymphocytes, a type of white blood cell, grow in the skin. This can cause redness of the skin, slightly raised or scaly rough patches, plaques, and sometimes skin tumors. CTCL is treatable, but generally not curable. Read this guide to learn more about the types of CTCL, and how they're diagnosed and treated.

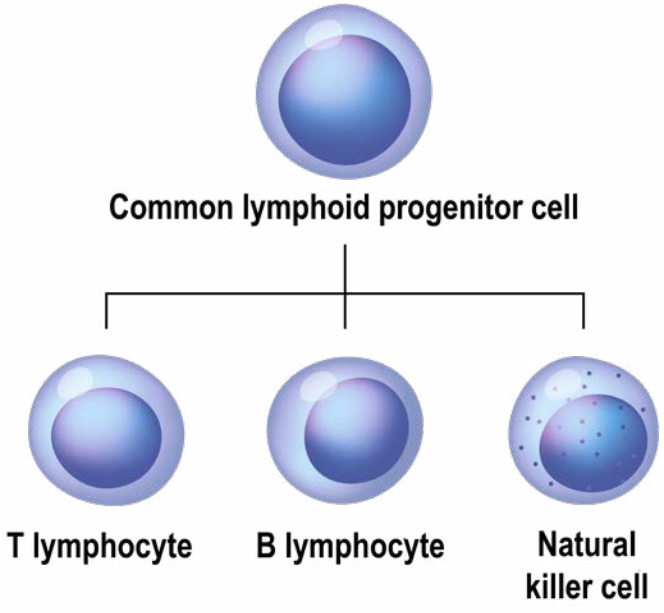
## What is cutaneous T-cell lymphoma?

Cutaneous T-cell lymphoma (CTCL) is a cancer of lymphocytes. Lymphocytes are part of the body's immune system. Their job is to protect the body against infections and to prevent or fight cancer. Lymphocytes normally grow in response to infection or inflammation. When they grow on their own without proper regulation, they can develop into lymphomas. In CTCL, abnormal lymphoma cells are attracted to and settle in the skin. These abnormal cells can also grow in the lymph nodes, blood, and other areas of the body. It's not skin cancer. Skin cancer develops from skin cells. Lymphomas of the skin develop from abnormal B or T lymphocytes.

CTCL is one of many types of T-cell lymphomas.

### Lymphocytes

**A lymphocyte is a type of white blood cell. In cutaneous T-cell lymphoma, abnormal lymphocytes cause skin lesions.**



### Types of lymphocytes

There are 3 main types of lymphocytes:

- **B lymphocytes**, or B cells, make antibodies and eventually mature into plasma cells. An antibody is a protein that specifically targets infections or cancer cells and recruits other parts of the immune system.
- **T lymphocytes**, or T cells, help fight infections and cancer, and control immune responses. You have normal T cells throughout your body, including in your skin.
- **Natural killer cells** can kill tumor cells and virus-infected cells, among other functions.

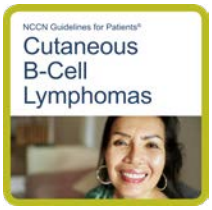
Lymphocytes are found in lymph nodes, in blood, and all over the body, including your skin. When lymphoma grows mainly in the skin, it's called primary cutaneous lymphoma.

There are 2 types of cutaneous lymphomas:

- Cutaneous B-cell lymphomas
- Cutaneous T-cell lymphomas

### Cutaneous B-cell lymphoma

Cutaneous B-cell lymphoma (CBCL) develops from abnormal B cells. It's the second-most common type of cutaneous lymphoma, making up about 1 out of every 5 cases. Information on types of CBCL, such as primary cutaneous follicle center lymphoma and primary cutaneous marginal zone lymphoma, can be found in *NCCN Guidelines for Patients: Cutaneous B-Cell Lymphomas,* available at NCCN.org/patientguidelines and on the NCCN Patient Guides for Cancer app.




### Cutaneous T-cell lymphoma

Cutaneous T-cell lymphoma (CTCL) develops from abnormal T cells. It's the most common type of cutaneous lymphoma, making up about 4 out of every 5 cases. Information on types of CTCL, such as mycosis fungoides, Sézary syndrome, and primary cutaneous T-cell lymphoproliferative disorders, will be discussed in this guide.

## How is CTCL treated?

CTCL is treatable, but generally not curable. While CTCL is typically not life-threatening, advanced disease can be very challenging to treat and should involve a team of specialists with experience in CTCL.

Treatment for the following types of CTCL are explained in this guide:

- Mycosis fungoides and Sézary syndrome (or simply referred to as MF/SS)
- Primary cutaneous CD30+ T-cell lymphoproliferative disorders
- Subcutaneous panniculitis-like T-cell lymphoma
- Primary cutaneous indolent lymphoproliferative disorders

In general, treatment focuses on the skin and might be a combination of therapies. Some treatments are placed directly on the lesion, while others work inside the body. The goal is to improve your condition and to maintain this improvement for as long as possible. Treatment will be personalized for you and your type of CTCL.

## How can I get the best care?

Advocate for yourself. You have an important role to play in your care. Many people feel more satisfied when they actively take part in planning their cancer care.

The NCCN Guidelines for Patients will help you play a larger role in your care. Discuss the recommendations in this guide with your care team. Ask questions about your options and share your goals and concerns.

Don't know what to ask? You're not alone. That's why we include suggested questions to ask at the end of chapters.

Keep reading to find the best care for you.

### How this guide can help you

Making decisions about cancer care is stressful. There's a lot to learn, and you don't know what the future holds.

Use this guide to get the information and support you need.

Patients, doctors, and other health care professionals trust the NCCN Guidelines for Patients. This guide uses clear, everyday language to explain current cancer care recommendations made by respected experts in the field. Their recommendations are based on the latest research and practices at leading cancer centers.

Your health is unique to you, so your cancer care should be, too. As you read this guide, you'll learn which treatments are likely to provide the best results for you. And you'll be better prepared to talk with your care team.

# 2
# Testing for CTCL


9 General health tests

10 Skin exam

11 Fertility (all genders)

12 Blood tests

12 Imaging tests

13 Biopsy

15 Immunophenotyping

16 Testing for CTCL biomarkers and genetic changes

18 Key points

18 Questions to ask

Treatment planning starts with testing. Accurate testing is essential to diagnose and treat lymphomas of the skin. This chapter presents an overview of tests you might receive and what to expect. It's okay to skip over information that doesn't interest you.

**Cutaneous T-cell lymphoma (CTCL) might appear on the skin as:**

- Papules (pimple-like lesions)
- Nodules (larger papules)
- Patches (flat, scaly lesions)
- Plaques (thicker, raised lesions)
- Tumors (raised lumps)
- Erythroderma (a red rash)

## General health tests

Some general health tests are described next.

### Medical history

A medical history is a record of all health issues and treatments you've had in your life. You'll be asked about past surgeries and current health conditions. Bring a list of old and new medicines, topical ointments, and any over-the-counter medicines, herbals, or supplements you take. Some supplements interact with and affect medicines your care team may prescribe. Since there's often a lack of published information about how certain supplements interact with standard medicines, your care team may recommend that many of them be discontinued during treatment.

### Physical exam

During a physical exam, your health care provider may:

- Feel and apply pressure to parts of your body to see if organs are a normal size, are soft or hard, or cause pain when touched.
- Feel for enlarged lymph nodes in your neck, armpit, and groin.
- Feel for an enlarged spleen or liver.
- Conduct a complete skin exam.

### Distress screening

Dealing with a cancer diagnosis can be stressful and may cause further distress. Distress is an unpleasant experience of a mental, physical, social, or spiritual nature. It can affect how you feel, think, and act. Distress might include feelings of sadness, fear, helplessness, worry, anger, and guilt. You may also have depression, anxiety, and sleep issues. Your treatment team will screen your level of distress. This is part of your cancer care.

## Skin exam

A dermatologist is an expert in skin. It's important to find an experienced dermatologist to conduct a skin exam. Cutaneous T-cell lymphoma (CTCL) might appear as a rash, lumps, bumps, or tumor. A rash is an area of irritated or swollen skin. Many rashes are itchy, red, or painful.

The amount of lymphoma is measured using the size of your hand. One hand is equal to 1 percent (1%) of your total body surface area (BSA). In addition, any tumors will be measured by their depth, height, size, and region of the body. Keeping a photo journal might help track your skin changes over time.

You know your skin better than anyone. Tell your doctor about your normal skin color. Show them where your skin looks normal and where it looks different to you. Describe any changes. Does the area itch or burn? Is it dry? Is it red or warm to the touch? Are there bumps or a raised, smooth area? Is there an odor? Share any photos.

### Skin lesions

The skin is the largest organ in your body. A skin lesion is a change in color or texture in an area of the skin. Skin lesions can appear anywhere on the body, but are most common on the lower abdomen, upper thighs, buttocks, and breasts. Some words to describe skin lesions include patch, papule, plaque, erythroderma, tumor or nodule, and ulcer.

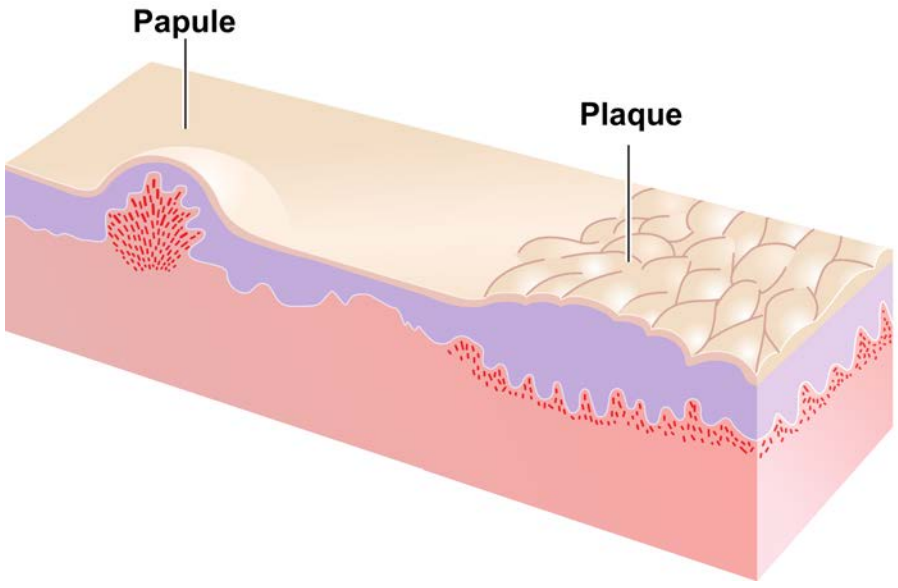
### Patch

A patch is a flat, thin, pink or red lesion of any size that forms on the skin. Patches may be dry, scaly, and itchy, and may look like eczema or psoriasis. They can be lighter or darker than surrounding skin. The patches may sometimes become plaques (raised or hardened lesions) on the skin.

**Skin lesions**

**A papule is a very small, solid bump. A plaque is a raised or hardened lesion that forms on the skin, larger than a papule. Plaques sometimes become tumors on the skin.**

Credit: https://commons.wikimedia.org/wiki/File:Papule_and_Plaque.svg



### Papule

A papule is a very small, solid lump that might look like a very small pimple. Usually, papules are found in groups. Papules may be red, purple, brown, or pink.

### Plaque

A plaque is a raised (elevated) or hardened (indurated) lesion of any size that forms on the skin. Plaques may be red, scaly, crusty, and itchy, and may look like eczema or psoriasis. Plaques sometimes become tumors on the skin.

### Erythroderma

Erythroderma is redness of over 80 percent (80%) of the body's skin surface. It's an important part of recognizing and treating mycosis fungoides and Sézary syndrome. Erythroderma can look like sunburn or large blotches on the skin.

### Tumor or nodule

A tumor or nodule is a firm, dome-shaped mass at least 1 centimeter in size (higher than a papule).

### Ulcer

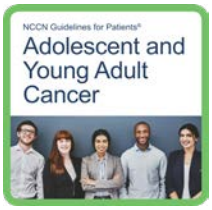
A skin ulcer is an open sore or wound on the skin.

## Fertility (all genders)

Treatment with systemic therapy can affect your fertility, or the ability to have children. If you think you want children in the future, ask your care team how lymphoma treatment might affect your fertility.

Fertility preservation is all about keeping your options open, whether you know you want to have children later in life or aren't sure. Fertility and reproductive specialists can help you sort through what may be best for your situation.

More information on fertility preservation can be found in *NCCN Guidelines for Patients: Adolescent and Young Adult Cancer,* available at NCCN.org/patientguidelines and on the NCCN Patient Guides for Cancer app.




### Preventing pregnancy during treatment

Preventing pregnancy during treatment is important. Lymphoma treatment, especially some of the systemic treatments, can affect the ovaries, damage sperm, and hurt a developing baby. Therefore, becoming pregnant or having one's partner become pregnant during treatment should be avoided. If you're pregnant or breastfeeding at the time of your lymphoma diagnosis, treatments may need to be adjusted.

## Blood tests

Blood tests check for signs of disease and how well organs are working. They require a sample of your blood, which is removed through a needle placed into your vein.

Some possible blood tests are described next.

### Complete blood count and differential

A complete blood count measures the levels of red blood cells, white blood cells, and platelets in your blood. A differential counts the number of each type of white blood cell (neutrophils, lymphocytes, monocytes, eosinophils, and basophils). It also checks if the counts are in balance with each other.

### Comprehensive metabolic panel

A comprehensive metabolic panel measures different substances in your blood. It provides important information about how well your liver and kidneys are working, among other things.

### Hepatitis B and hepatitis C

Hepatitis B and hepatitis C virus infections can affect the liver. A blood test shows if you had hepatitis in the past or if you have it now. Some lymphoma treatments can wake up (or reactivate) the virus. If this happens, it can harm the liver.

### HTLV

Human T-lymphotropic virus (HTLV) testing may be useful to evaluate a different type of T-cell lymphoma associated with certain at-risk geographic locations.

### Lactate dehydrogenase

Lactate dehydrogenase (LDH), or lactic acid dehydrogenase, is a protein found in most cells, including red blood cells and white blood cells. Dying cells release LDH into the blood. Fast-growing cells, including lymphoma cells, also release LDH.

### Pregnancy test

People who can become pregnant will have a pregnancy test before treatment begins.

## Imaging tests

Imaging tests take pictures of the inside of your body to look for cancer deposits. A radiologist, an expert in interpreting imaging tests, will write a report and send this report to your care team.

### CT scan

A CT, or CAT, scan uses x-rays and computer technology to take pictures of the inside of the body. It takes many x-rays of the same body part from different angles. All the images are combined to make 1 detailed picture. A CT scan of your neck, chest, abdomen, and pelvis may be used to look for lymphoma. In most cases, contrast is used. Contrast is a substance used to improve the quality of CT imaging scans. It might be taken by mouth (oral) or given through a vein (intravenous, or IV).

### PET scan

A PET scan uses a radioactive substance called a tracer. The radiotracer fluorodeoxyglucose (FDG) mimics the behavior of sugar once injected into a vein in your body. The FDG uptake gives a clue to where

lymphoma cells are in the body and how much sugar they take up. This provides information on how fast the lymphoma cells are growing. Lymphomas often show up as bright spots on PET scans. However, not all tumors appear on a PET scan, and not all bright spots are lymphoma. It's normal for the brain, heart, kidneys, and bladder to be bright on PET. Inflammation or infection can also show up as a bright spot. When a PET scan is combined with CT, it's called a PET/CT scan.

## Biopsy

A biopsy removes a tissue sample, such as skin, lymph node, and/or bone marrow, for testing. It's an important part of an accurate diagnosis of lymphoma. Your sample should be reviewed by a pathologist who's an expert in lymphoma. The pathologist (or dermatopathologist) will note the overall appearance and size, shape, and type of cells and write a report. This review is often referred to as a histology, histopathology, or hematopathology review. Tests are done on the biopsied cells to accurately diagnose cutaneous T-cell lymphoma (CTCL).

### Skin lesion biopsy

A sample of your skin lesion will be removed and tested to confirm the type of CTCL. The biopsy may be a shave biopsy or punch biopsy.

### Skin shave biopsy

A skin shave biopsy removes the top layer of skin, using a tool like a razor. Stitches aren't needed.

### Skin punch biopsy

In a skin punch biopsy, a small sample of skin and connective tissue is removed using a handheld tool. Stitches are often used to close the opening in the skin.

### PET scan

**A PET scan uses a safe, radioactive substance, called a tracer, to look for areas of lymphoma in the body.**

Credit: https://commons.wikimedia.org/wiki/File:Positron_emission_tomography.png



## Lymph node biopsy

When CTCL involves lymph nodes, they can be swollen, enlarged, or hard, or may not move when pushed. A lymph node biopsy is done using a needle biopsy procedure or as a small surgery to remove (excise) a lymph node.

## Bone marrow tests

Bone marrow tests might be done in certain cases.

There are 2 types of bone marrow tests that are often done at the same time:

- Aspiration
- Biopsy

Your bone marrow is like a sponge holding liquid and cells. Aspiration takes some of the liquid and cells out of the marrow, and a biopsy removes a piece of the marrow.

Bone marrow aspiration and biopsy are often bedside procedures done under local anesthesia. They aren't surgeries and don't usually require an operating room. Your care team will try to make you as comfortable as possible during the procedures. The biopsy can also be done by an interventional radiologist when needed.

The samples are usually taken from the back of the pelvis. You'll likely lie on your belly or side. For the aspiration, a hollow needle is pushed through your skin and into the bone marrow. Liquid bone marrow is then drawn into a syringe. For the biopsy, a wider needle is used to remove a small piece of bone marrow. You may feel pain at your hip for a few days. Your skin may bruise.

**While cutaneous T-cell lymphoma (CTCL) is typically not life-threatening, advanced disease can be very challenging to treat and should involve a team of specialists with experience in CTCL.**



# Immunophenotyping

Immunophenotyping is a process that uses antibodies to detect the presence or absence of white blood cell antigens, called biomarkers. These antigens are proteins that can be found on the surface of or inside white blood cells. They help the pathologist tell what type of cells make up a tumor.

Specific groupings of antigens are normal. However, some specific patterns of antigens, called the immunophenotype, are found on abnormal cells, including CTCL.

Immunophenotyping can be done using specialized techniques, such as flow cytometry or immunohistochemistry. These techniques are used to distinguish CTCL from other types of lymphoma. The immunophenotype can change as lymphoma progresses.

- **Mycosis fungoides and Sézary syndrome** cells are typically characterized by the following immunophenotype: CD2+, CD3+, CD5+, CD4+, CD8-, and CD7- and CD26-. Sézary syndrome cells can have a clonal rearrangement of the T-cell receptor gene (*TCR*) in the blood.
- **Primary cutaneous CD30+ T-cell lymphoproliferative disorders** have more than 75% T cells that are CD30+.
- **Subcutaneous panniculitis-like T-cell lymphoma** cells are typically characterized by the following immunophenotype: CD3+, CD8+, betaF1+, CD2+, CD5+, and CD7+.
- **Primary cutaneous indolent lymphoproliferative disorders** can have a *TCR* gene rearrangement and are CD4+ or CD8+.

## Immunohistochemistry

Immunohistochemistry (IHC) is a special staining process that involves adding a chemical marker to immune cells. The cells are then studied under a microscope. IHC looks for the immunophenotype of cells collected during a skin biopsy.

CD4 and CD8 are proteins that are on two families of T lymphocytes. CD4 T cells (helper cells) help regulate functions of the immune system. CD8 T cells (killer cells) break down or rid the body of foreign substances. Most cases of CTCL come from CD4 T cells. An IHC will look for these cells and others. An IHC panel of skin biopsy may include testing for CD2, CD3, CD4, CD5, CD7, CD8, CD20, and CD30. Others might be included.

## Flow cytometry

Flow cytometry is a laboratory method used to detect, identify, and count specific cells. It involves adding a light-sensitive dye to cells. The dyed cells are passed through a beam of light in a machine. The machine measures the number of cells, as well as things like their size and shape.

Flow cytometry may be used on cells from circulating (peripheral) blood, bone marrow, or biopsy. The most common use of flow cytometry is to identify markers on cells, particularly in the immune system (called immunophenotyping). In CTCL, flow cytometry is often used to count the number of Sézary cells or CTCL cells in the blood.

# Testing for CTCL biomarkers and genetic changes

Biomarker and genetic tests might be used to learn more about your type of CTCL to guide treatment and to determine the likely path your lymphoma will take (prognosis). This genetic testing is different from family history genetic testing or genetic cancer risk testing. It looks for changes only in the CTCL cells that have developed over time and not for changes in the rest of your body's cells.

Inside our cells are DNA molecules. These molecules are tightly packaged into what is called a chromosome. Chromosomes contain most of the genetic information in a cell. Normal human cells contain 23 pairs of chromosomes, for a total of 46. Each chromosome contains thousands of genes. Most genes contain information for making a specific protein. A mutation is when something goes wrong in the genetic code.

Proteins are written without italics like this: CD4. Genes are written with italics like this: *TCR.* When a gene or protein is found (expressed), it's shown with a plus sign (+) like this: CD4+. When a gene or protein isn't found, it's written with a negative sign (-) like this: CD4-.

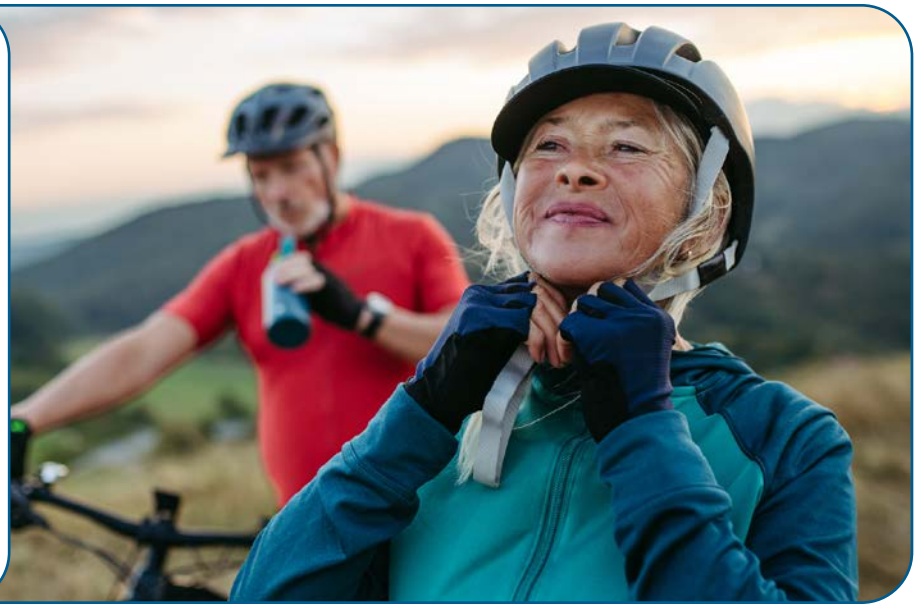
## CTCL molecular testing

Molecular testing, using methods such as fluorescence in situ hybridization (FISH), polymerase chain reaction (PCR), high-throughput sequencing (HTS), next-generation sequencing (NGS), and multigene panel testing, looks for changes or abnormalities unique to CTCL cells. A sample of your skin, lymph node, blood, or bone marrow might be used to see if the CTCL cells have any specific abnormalities or mutations.

## Epstein-Barr virus testing

Epstein-Barr virus-encoded RNA in situ hybridization (EBER-ISH) is used to detect Epstein-Barr virus (EBV).

**"Don't let yourself stop doing the things you enjoy doing. Whatever your hobbies are, whatever things you like to do, keep doing them. It'll help you feel more like yourself and stay positive during a stressful time."**



## FISH

Fluorescence in situ hybridization (FISH) is a method that involves special dyes, called probes, that attach to pieces of DNA. Since this test doesn't need growing cells, it can be performed on bone marrow, a lymph node, or a blood sample.

FISH can find translocations that are too small to be seen with other methods. A translocation occurs when parts of 2 chromosomes switch with one another. However, FISH can only be used for known specific changes. It can't detect all the possible changes found within the chromosomes or genes. For example, FISH is used to detect *TP63* gene rearrangements by attaching probes to *TP63* (3q28) and *TBL1XR1/TP63.*

## Gene rearrangement

Gene rearrangement is a normal process that happens as B cells and T cells mature. The genes that code for antibodies, or T-cell receptors, are cut and reshuffled in a specific pattern to create the large diversity of receptors the immune system needs. A normal, healthy lymphocyte is made up of many different cells with many different rearrangement patterns, called a polyclonal mix. Lymphoma, on the other hand, arises when a single, abnormal lymphocyte multiplies out of control. This means the lymphoma has 1 dominant, identical rearrangement pattern repeated across all the cancerous cells, called a monoclonal population. Testing for this helps distinguish a true cancer (monoclonal) from a reactive, benign immune response (polyclonal), which can look very similar under a microscope.



**Testing takes time. It might take days or weeks for all test results to come in.**

In cutaneous T-cell lymphoma, a tumor might be described as having clonality. Clonal T cells should have the same T-cell receptor and the same T-cell receptor gene sequence. A T-cell receptor (TCR) test determines if a group of tumor cells is clonal or not. However, positive or negative TCR clonality should be interpreted in the clinical context by experienced physicians. Sézary syndrome can have a TCR clonal rearrangement found in the blood. Gene rearrangements within *TCR*, such as *TCRB* and *TCRG,* are also used for diagnosis of other types of CTCL.

## Genetic testing for *HAVCR2* mutation

Genetic testing is done to look for gene mutations inherited from your biological (birth) parents, called germline mutations. Testing for a *HAVCR2* gene mutation may aid in diagnosis and treatment decisions if there's concern for subcutaneous panniculitis-like T-cell lymphoma.

## Key points

- Blood and imaging tests check for signs of disease beyond the skin, how well organs are working, and treatment results.
- Skin lesions can appear anywhere on the body. Lesions can be patches, papules, plaques, erythroderma, tumors or nodules, or ulcers.
- Usually more than 1 skin biopsy is needed to diagnose cutaneous T-cell lymphoma (CTCL). Your skin samples should be reviewed by a pathologist who's an expert in cutaneous lymphomas.
- Immunophenotyping is used to distinguish CTCL from other types of lymphoma.
- A sample from your biopsy may undergo lab tests to learn more about your CTCL and to choose the best treatment for you.
- Biomarker and genetic tests are used to learn more about your type of CTCL, to guide treatment, and to determine the likely path your lymphoma will take (prognosis).

## Questions to ask

- What's the name of my lymphoma and can you write it down for me?
- What can you tell me about my type of CTCL?
- What websites about my condition do you recommend? Is there any information I can take home to read?
- What type of biopsy will I have?
- What types of biomarker and genetic tests will be done on my biopsy sample?

## What's next?

The next chapter provides information on cancer staging and how it's used for the 2 main types of CTCL.

# 3
# Cancer staging


20 TNMB scores

21 MF/SS stages

24 Key points

24 Questions to ask

Cancer staging is used to guide treatment decisions in mycosis fungoides (MF) and Sézary syndrome (SS). In MF/SS, staging describes areas of lymphoma found in the skin, lymph nodes, blood, or other organs.

## TNMB scores

Cancer staging is used to guide treatment decisions in some lymphomas. In mycosis fungoides (MF) and Sézary syndrome (SS), the tumor, node, metastasis, and blood (TNMB) system is used to describe areas of lymphoma found in the skin, lymph nodes, blood, or other organs. Based on biopsy and other test results, each TNMB letter is assigned a number, or score. A TNMB example might look like this: T4N1M0B1 or T4, N1, M0, B1.

- **T is for skin** – Refers to the type and number of lesions covering the skin. Staging looks for the presence of lesions, patches, papules, plaques, tumors, or reddening of the skin (erythema) and how much body surface area (BSA) is affected.
- **N is for node** – Refers to abnormal T lymphocytes found in lymph nodes.
- **M is for metastasis** – Refers to lymphoma found in internal (visceral) organs or bone marrow.
- **B is for blood** – Refers to abnormal T lymphocytes found in circulating (peripheral) blood.

### T = Tumors in the skin

In MF/SS, the amount of lymphoma is measured by evaluating what percent of your skin is affected by lymphoma. One hand is equal to 1 percent (1%) of your total body surface area (BSA). In addition, tumors are measured by their depth, height, size, and region of the body. Tumors/tumor areas are often measured in centimeters (cm).

- **T1** – Patches, papules, and/or plaques cover less than 10% BSA
- **T2** – Patches, papules, and/or plaques cover 10% or more BSA
  - **T2a** is patches only
  - **T2b** is plaques with or without patches
- **T3** – One or more tumors 1 cm or more in size
- **T4** – Reddening, thickening, or involvement of the skin (erythema) covering 80% or more BSA

### N = Node

There are hundreds of lymph nodes throughout your body. Lymph nodes work as filters to help fight infection and remove harmful substances. They also produce lymphocytes. As abnormal T lymphocytes multiply, they can distort, or overtake, the lymph node.

Abnormal lymph nodes are any that can be felt on physical exam as firm, irregular, clustered, fixed, or 1.5 cm or more in diameter. Imaging tests are used to confirm abnormal lymph nodes.

- **N0** – No abnormal T lymphocytes found in lymph nodes.
- **N1** – Some abnormal T lymphocytes found in lymph nodes.
- **N2** – Many abnormal T lymphocytes or clusters found in lymph nodes.
- **N3** – Abnormal T lymphocytes have altered the structure of lymph nodes. This is called lymph node (nodal) disease.

### M = Metastasis

Lymphoma that has spread to distant parts of the body is usually called metastatic. In MF/SS, lymphoma can spread to bone marrow or visceral (internal, solid) organs, such as the spleen, liver, or lungs.

- **M0** – No lymphoma found in bone marrow or internal organs.
- **M1A** – Lymphoma found in bone marrow only.
- **M1B** – Lymphoma found in visceral organs only.

### B = Blood

Peripheral blood circulates throughout your body (bloodstream). The amount of abnormal T cells found in the blood will be measured.

- **B0 (no or very low blood involvement)** – No blood involvement or very small amounts (less than 250 cells per microliter, or 250/µL) of CD4+/CD26- or CD4+/CD7- cells found in blood.
- **B1 (low blood tumor burden)** – More than 250/µL but less than 1,000/µL of CD4+/CD26- or CD4+/CD7- cells found in blood.
- **B2 (high blood tumor burden)** – More than 1,000/µL CD4+/CD26-, CD4+/CD7-, or other abnormal T cells found in blood. A diagnosis of SS requires B2 involvement.

## MF/SS stages

Stages range from 1 to 4, with 4 being the most advanced. You might see these stages written as stage I, stage II, stage III, and stage IV. Not all cancers are described this way. Stages are defined by TNMB scores.

For mycosis fungoides and Sézary syndrome (MF/SS) cancer stages, see **Guide 1**.

### Stage 1A – Limited skin involvement

Stage 1A is mycosis fungoides.

Limited patches, papules, and/or plaques cover less than 10% of the skin (T1). Lymphoma isn't found in lymph nodes (N0) or visceral organs (M0).

### Stage 1B – Skin-only disease

Stage 1B is mycosis fungoides.

Stage 1B has more skin involvement than stage 1A. Patches, papules, and/or plaques cover 10% or more of the skin (T2). Lymphoma isn't found in lymph nodes (N0) or visceral organs (M0).

### Stage 2A

Stage 2A is mycosis fungoides.

Any amount of the skin surface is covered with patches or plaques (T1 or T2). Lymphoma is found in the lymph nodes (N1 or N2).

### Stage 2B – Tumor stage disease

Stage 2B is mycosis fungoides.

One or more tumors 1 cm or more in size are found on the skin (T3). Lymphoma may (N1 or N2) or may not (N0) be in the lymph nodes. It may (B1) or may not (B0) be found in blood.

### Stage 3A – Erythrodermic disease

Stage 3A is mycosis fungoides.

In erythrodermic disease, nearly all the skin (more than 80%) is reddened (erythema) (T4), but little disease is found inside the body. Patches, plaques, or tumors may also be found on the skin. Lymphoma may (N1 or N2) or may not (N0) be found in lymph nodes. There's no visceral or significant blood involvement.

**Guide 1**
**Mycosis fungoides and Sézary syndrome cancer stages**

| | | |
|---|---|---|
| **Stage 1**<br>**Mycosis fungoides** | **Stage 1A**<br>**Limited skin involvement** | T1, N0, M0, B0 or B1 |
| | **Stage 1B**<br>**Skin-only disease** | T2, N0, M0, B0 or B1 |
| **Stage 2**<br>**Mycosis fungoides** | **Stage 2A** | T1 or T2, N1 or N2, M0, B0 or B1 |
| | **Stage 2B**<br>**Tumor stage disease** | T3, N0 or N1 or N2, M0, B0 or B1 |
| **Stage 3**<br>**Mycosis fungoides** | **Stage 3A**<br>**Erythrodermic disease** | T4, N0 or N1 or N2, M0, B0 |
| | **Stage 3B**<br>**Erythrodermic disease** | T4, N0 or N1 or N2, M0, B1 |
| **Stage 4**<br>**Sézary syndrome (SS) or mycosis fungoides (MF)** | **Stage $4A_1$**<br>**Sézary syndrome** | Any T, N0 or N1 or N2, M0, B2 |
| | **Stage $4A_2$**<br>**Can be SS or MF** | Any T, N3, M0, any B |
| | **Stage 4B**<br>**Visceral disease (SS or MF)** | Any T, any N, M1A or M1B, any B |

### Stage 3B – Erythrodermic disease

Stage 3B is mycosis fungoides.

This is the same as 3A, but a small amount of lymphoma is found in the blood (B1).

### Stage $4A_1$ – Sézary syndrome

Stage $4A_1$ is Sézary syndrome.

There's significant disease in the blood, including a high number of Sézary cells (B2). Skin can be any stage (any T). Lymphoma may (N1 or N2) or may not (N0) be in the lymph nodes.

### Stage $4A_2$ – Sézary syndrome or mycosis fungoides

Stage $4A_2$ can be Sézary syndrome or mycosis fungoides.

There's significant disease in the lymph nodes. Skin can be any stage (any T). Abnormal T lymphocytes have altered the structure of the lymph node (N3). Lymphoma may be in the blood. A diagnosis of Sézary syndrome requires B2 blood involvement (more than 1,000/μL Sézary cells found in blood).

### Stage 4B – Visceral disease

Stage 4B can be Sézary syndrome or mycosis fungoides.

There's significant disease in the organs. Skin can be any stage (any T). Lymph nodes can be any stage (any N). Lymphoma has spread to bone marrow (M1A) or internal (visceral) organs (M1B). There may be blood involvement (any B).

**Erythroderma**

**Mycosis fungoides and Sézary syndrome often cause erythroderma. Erythroderma is severe inflammation of most of the body's skin surfaces. It can look like sunburn or large splotches.**

Credit: https://commons.wikimedia.org/wiki/File:Sezery2.jpg



## Key points

- Cancer staging is used to guide treatment decisions in mycosis fungoides (MF) and Sézary syndrome (SS).
- Staging for MF/SS is slightly different from that for other cancers. It uses a TNMB system that stands for tumor, node, metastasis, and blood.
- T refers to the type and number of lesions covering the skin. N describes if there are any abnormal lymphocytes found in lymph nodes. M describes lymphoma found in internal organs or bone marrow. B refers to abnormal lymphocytes in the blood.
- Staging may change as cancer progresses.
- MF can be stage 1, 2, 3, $4A_2$, or 4B.
- In SS, cancerous T cells, called Sézary cells, are found in the skin, lymph nodes, and blood. SS can be stage $4A_1$, $4A_2$, or 4B. To be diagnosed with SS, a certain number of Sézary cells need to be found in the blood.

## Questions to ask

- Do I have mycosis fungoides or Sézary syndrome?
- What stage is the lymphoma and what does this mean?
- How does the cancer stage influence treatment options?
- Is there lymphoma beyond the skin (in the lymph nodes or blood)?
- How well will treatment control or reduce the lymphoma?

## What's next?

The next chapter provides information on the types of treatment and what to expect from treatment.

# 4
# Types of treatment


26 Your care team

27 Skin-directed therapy

28 Radiation therapy

29 Systemic therapy

30 Clinical trials

31 Hematopoietic cell transplant

32 Supportive care

33 Key points

33 Questions to ask

This chapter presents an overview of the possible types of treatment and what to expect. Not everyone will receive the same treatment. Treatment options are based on many factors. Together, you and your care team will choose a treatment plan that's best for you.

## Your care team

Treating cutaneous T-cell lymphoma (CTCL) takes a team approach. Treatment decisions should involve a team of health care and psychosocial care professionals from different backgrounds who have knowledge and experience in your type of CTCL. This team is united in planning and implementing your treatment.

It's recommended your care team involve the following experts:

- **A dermatologist** specializes in the diagnosis and treatment of skin diseases.
- **A hematologist or an oncologist** specializes in cancers, like lymphoma.
- **A pathologist, dermatopathologist, or hematopathologist** analyzes the cells and tissues removed during a biopsy and provides cancer diagnosis, staging, and information about biomarker testing.
- **A radiation oncologist** treats cancer using radiation therapy.

**"I'm glad I found a care team that I could talk to. We discussed treatment options together and they listened when I spoke about how I was feeling. This shared decision-making made me feel like I had a say about my care."**



# Skin-directed therapy

Types of therapy focused on the skin include topical therapy, local radiation therapy, and phototherapy.

## Topical therapy

A topical treatment is put on the surface of the skin. It might be a lotion (cream), gel, or ointment. Types of topical therapy are described next.

- **Calcineurin inhibitors** block an enzyme that causes inflammation and itching. These are only used in some cases. Examples include pimecrolimus (Elidel) and tacrolimus (Protopic).
- **Carmustine** is a chemotherapy that stops or slows the growth of cancer. Topical carmustine (BiCNU) is applied to lesions.
- **Corticosteroids (steroids)** are used to reduce inflammation. Steroids can be topical or intralesional. An intralesional steroid is injected directly into a lesion on or just below the skin.
- **Imiquimod** is a cream used to treat certain types of flat, scaly growths on the skin.
- **Mechlorethamine (nitrogen mustard)** gel (Valchlor) stops or slows the growth of cancer.
- **Retinoids** are products related to vitamin A. Bexarotene (Targretin gel) and tazarotene (Tazorac gel, Tazorac cream) are retinoids.

## Local radiation therapy

Local radiation therapy treats the skin lesion directly. Involved-site radiation therapy is a type of local radiation. It can be used on lymph nodes and/or skin lesions. The type of radiation is usually electron beam therapy (EBT). Some people with cutaneous T-cell lymphoma (CTCL) may see a benefit with low doses of radiation.

## Phototherapy

Phototherapy uses different wavelengths of ultraviolet (UV) light to treat skin lesions or tumors.

Types include:

- **Ultraviolet B (UVB)** exposes the skin to an artificial UVB light source for a set length of time on a regular schedule. This is less commonly used.
- **Narrowband ultraviolet B (NB-UVB)** uses a very specific UV wavelength. This is more commonly used.
- **Photochemotherapy or psoralen plus ultraviolet A (PUVA)** combines psoralen with UVA. Psoralen is a type of medicine taken by mouth (orally) that causes your skin to be sensitive to light. After taking psoralen, the skin is exposed to long-wave UV light. It has a deeper penetration than UVB.
- **Ultraviolet A1 (UVA1)** penetrates even deeper into the skin, causing T cells to die.

Often, NB-UVB is used for patches or thin plaques, and PUVA or UVA1 is used for thicker plaques.

UV may increase your risk of some skin cancers in certain cases. Phototherapy may not be favored in people with a history of melanoma or other types of skin cancer.

## Radiation therapy

Radiation therapy (RT) uses high-energy beams to kill lymphoma cells and shrink tumors. Most lymphomas of the skin are very sensitive to radiation. Because of this, RT is often the preferred initial treatment for lymphoma limited to the skin. The dose of radiation typically prescribed for CTCL is relatively low. Treatment usually uses electron beams or superficial x-rays that only target the skin and tissues a couple of millimeters deep. This limits the amount of radiation that reaches deeper tissues and organs.

Before treatment begins, you'll have a planning visit called simulation (or mapping). For most skin lesions, this involves a clinical simulation. Your radiation oncologist will examine your skin and outline the treatment area, including a small border around the lymphoma. Your treatment team will also find the best position for you during treatment. You'll typically return on a separate day to begin RT, which is delivered by a machine outside your body.

In some cases, depending on the size, depth, or location of the lymphoma, a planning CT scan may be needed. Your radiation oncologist will use the scan and computer software to create a 3D treatment plan.

The most common types of radiation therapy used to treat CTCL include:

- **Involved-site radiation therapy (ISRT)** targets a specific area of skin. It can also be used to treat specific lymph nodes with lymphoma.
- **Total skin electron beam therapy (TSEBT)** treats the entire skin surface. You might stand on a platform that slowly rotates horizontally to receive this treatment.

### Radiation therapy

**Most cutaneous T-cell lymphomas (CTCLs) are extremely sensitive to radiation. Because of this, radiation therapy is widely used to treat CTCL.**



## Systemic therapy

Systemic therapy works throughout the body and is more commonly used to treat lymphoma in areas outside the skin, such as lymph nodes, blood, or bone marrow. Types that may be used to treat CTCL are described next.

### Extracorporeal photopheresis

Photopheresis, also known as extracorporeal photopheresis, is a medical treatment that removes blood from the body using a machine. The machine separates out the white blood cells. These white cells are exposed to a light-sensitizing medicine, called 8-methoxypsoralen (8-MOP), followed by ultraviolet A radiation. Then the blood with the treated white blood cells is returned to your body.

### Chemotherapy

There are a few chemotherapies used to treat CTCL. Examples include liposomal doxorubicin (Doxil), methotrexate (Jylamvo), gemcitabine (Avgemsi), pralatrexate (Folotyn), and romidepsin (Istodax).

### Immunotherapy

Immunotherapy helps your immune system recognize and fight lymphoma. Examples include interferon alfa-2a (Pegasys), lenalidomide (Revlimid), mogamulizumab (Poteligeo), and pembrolizumab (Keytruda). Denileukin diftitox-cxdl (Lymphir) and brentuximab vedotin (Adcetris) are antibody-drug conjugates that combine the features of immune-targeting and chemotherapy.

### Immunosuppressive agents

Immunosuppressive agents decrease the body's immune response.

### About NCCN recomendations for therapy

Cutaneous T-cell lymphomas (CTCLs) are treated with skin-directed therapy, radiation therapy, and systemic therapy.

All treatments listed in this guide are deemed recommended and appropriate, by our NCCN panel members for cutaneous lymphomas. This panel often assign levels of preferrence to the treatment recommendations as described below.

- **Preferred therapies** have the most evidence they may work better and be safer than other therapies.
- **Other recommended therapies** can provide effective results but may have less evidence, more side effects, or may not work quite as well as preferred therapies.
- **Therapies used in certain cases** work best for individuals with specific cancer features or health circumstances.

### Retinoids

Retinoids are products related to vitamin A that can stop the growth of cancer cells. When taken by mouth (orally), they work throughout the body. Bexarotene (Targretin), acitretin, and isotretinoin are retinoids.

### Targeted therapy

Targeted therapy focuses on specific or unique features of cancer cells. They seek out how cancer cells grow, divide, and move in the body. These drugs stop the action of molecules that help cancer cells grow and/or survive. Duvelisib (Copiktra) is a targeted therapy.

## Clinical trials

You may also be able to receive treatment through a clinical trial. A clinical trial is a type of medical research study. After being developed and tested in a lab, potential new ways of treating cancer need to be studied in people.

If found to be safe and effective in a clinical trial, a drug, device, or treatment approach may be approved by the U.S. FDA.

Everyone with cancer should carefully consider all of the treatment options available for their cancer type, including standard treatments and clinical trials. Talk to your doctor about whether a clinical trial may make sense for you.

### Phases

Most cancer clinical trials focus on treatment and are done in phases.

- **Phase 1** trials study the safety and side effects of an investigational drug or treatment approach.
- **Phase 2** trials study how well the drug or approach works against a specific type of cancer.
- **Phase 3** trials test the drug or approach against a standard treatment. If the results are good, it may be approved by the FDA.
- **Phase 4** trials study the safety and benefit of an FDA-approved treatment.

### Who can enroll?

It depends on the clinical trial's rules, called eligibility criteria. The rules may be about age, cancer type and stage, treatment history, or general health. They ensure that participants are alike in specific ways and that the trial is as safe as possible for the participants.



### Finding a clinical trial

#### In the United States

NCCN Cancer Centers
NCCN.org/cancercenters

The National Cancer Institute (NCI)
cancer.gov/about-cancer/treatment/clinical-trials/search

#### Worldwide

The U.S. National Library of Medicine (NLM)
clinicaltrials.gov

#### Need help finding a clinical trial?

NCI's Cancer Information Service (CIS)
1.800.4.CANCER (1.800.422.6237)
cancer.gov/contact

### Informed consent

Clinical trials are managed by a research team. This group of experts will review the study with you in detail, including its purpose and the risks and benefits of joining. All of this information is also provided in an informed consent form. Read the form carefully and ask questions before signing it. Take time to discuss it with people you trust. Keep in mind that you can leave and seek treatment outside of the clinical trial at any time.

### Will I get a placebo?

Placebos (inactive versions of real medicines) are almost never used alone in lymphoma clinical trials. It's common to receive either a placebo added to a standard treatment, or a new drug with a standard treatment.

### Are clinical trials free?

There's no fee to enroll in a clinical trial. The study sponsor pays for research-related costs, including the study drug. But you may need to pay for other services, like transportation or childcare, due to extra appointments. During the trial, you will continue to receive standard cancer care. This care is often covered by insurance.

## Hematopoietic cell transplant

A hematopoietic cell transplant (HCT) is a cellular therapy that replaces a person's bone marrow and immune system with their own healthy cells or donor cells. More specifically, an HCT replaces hematopoietic stem cells that have been destroyed by high doses of chemotherapy and/or radiation therapy as part of the transplant process. A hematopoietic stem cell is an immature cell that can develop into any type of blood cell. HCTs are performed in specialized centers.

There are 2 types of HCTs:

- **Autologous** – Stem cells come from you. It's also called high-dose therapy with autologous stem cell rescue (HDT/ASCR). Before high-dose chemotherapy, your healthy stem cells are removed (rescued) from your blood or bone marrow.
- **Allogeneic** – Stem cells come from a matched donor. These stem cells may be able to detect and eliminate lymphoma cells better than your own immune system.

In some cases, an allogeneic HCT is a treatment option. It's used to cure mycosis fungoides and Sézary syndrome. The steps of an allogeneic HCT are described next.

**Before an allogeneic HCT**, a treatment called conditioning is needed to destroy your bone marrow cells. Conditioning creates room for transplanted donor stem cells. It also weakens your immune system, so your body won't kill the transplanted cells. After conditioning, you'll receive a transfusion of healthy stem cells from a donor who's matched to you.

### After conditioning

A transfusion is a slow injection of blood products into a vein. This can take several hours. The transplanted stem cells from a donor travel to your bone marrow and grow. New, healthy blood cells form. This is called engraftment, and it usually takes about 2 to 4 weeks. Until then, you'll have little or no immune defense. You may need to stay in a very clean room at the hospital or be given antibiotics to prevent or treat infection. Transfusions are also possible. A red blood cell transfusion is used to prevent bleeding and to treat anemia (a below-normal red blood cell count). A platelet transfusion is used to treat a low platelet count or bleeding. While waiting for the cells to engraft, you'll likely feel tired and weak. This treatment has very serious and life-threatening side effects.

The goal of the transplant is for the new immune system to recognize the lymphoma as foreign and destroy it and to provide you with a new, healthy immune system.

## Supportive care

Supportive care is an important part of cancer care. The goal is to improve your quality of life during and after cancer treatment. Supportive care is for everyone with cancer and their families, not just for those at the end of life.

Supportive care includes a wide range of services. Supportive care prevents or manages the symptoms of lymphoma and the side effects of treatment, like pain and cancer-related fatigue. It also addresses the mental, social, emotional, and spiritual concerns faced by people with cancer.

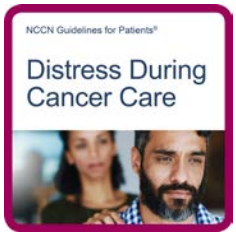
Supportive care provides help with additional needs, such as:

- Making treatment decisions
- Coordinating your care
- Paying for care
- Planning for advance care and end of life

Some health care professionals refer to palliative care as supportive care. They think these types of care are alike or overlap. Others think there are important differences, but both help people with cancer to live as well as they can. Your cancer center may have palliative care specialists, who can give you additional support and help coordinate your supportive care needs.

Read more about the types of support you may receive in *NCCN Guidelines for Patients: Palliative Care* and in *NCCN Guidelines for Patients: Distress During Cancer Care,* available at NCCN.org/patientguidelines and on the NCCN Patient Guides for Cancer app.



## Key points

- Treating cutaneous T-cell lymphoma (CTCL) takes a team of professionals experienced in your type of CTCL.
- Skin-directed therapy focuses on the skin and includes topical therapy, local radiation, and phototherapy.
- Radiation therapy uses high-energy beams to kill lymphoma cells and shrink tumors. Radiation therapy in CTCL often uses electrons, which only target the skin and tissues just beneath it.
- Systemic therapy works throughout the body. It includes extracorporeal photopheresis, chemotherapy, immunotherapy, and targeted therapy.
- Clinical trials study how safe and helpful tests and cancer treatments are for people.
- A hematopoietic cell transplant (HCT) replaces a person's bone marrow and immune system with their own healthy cells or donor cells.
- Supportive care is health care that relieves symptoms caused by cancer or its treatment and improves quality of life.

## Questions to ask

- What is your experience in treating CTCL?
- Is there a cancer center or hospital nearby that specializes in treating CTCL?
- How long will treatment take, and in what order will it be given?
- Can I work during treatment?
- Will I spend time in the hospital during treatment?

## What's next?

The next chapter explains diagnosis and treatment of mycosis fungoides.

# 5
# Mycosis fungoides


35 Overview

36 Testing

36 Goal of treatment

36 Stage 1A

38 Stage 1B and 2A

39 Stage 2B

41 Stage 3

42 Stage 4

43 Large cell transformation

44 Key points

44 Questions to ask

Mycosis fungoides is a chronic disease that starts in the skin, but in advanced stages can spread to the lymph nodes, blood, or other organs. Treatment might be a skin-directed therapy or a combination of systemic therapy with skin-directed therapy. The goal of treatment is to improve your condition and to maintain this improvement for as long as possible.

## Overview

Mycosis fungoides (MF) is the most common type of cutaneous T-cell lymphoma (CTCL). MF starts in the skin, but in advanced stages, it can spread to the lymph nodes, blood, or other organs, such as the spleen, liver, or lungs. MF is usually slow growing (indolent) and appears as patches, plaques, and tumors. A combination of patches, plaques, and tumors with open sores (ulceration) is possible. Symptoms include rash and itchy skin.

There are several different types (variants) of MF:

- **Folliculotropic MF** affects hair follicles. Lesions are found in the head, eyebrow, or neck area and often with follicular papules and plaques. Loss of hair in affected sites is common.
- **Pagetoid reticulosis, or Woringer-Kolopp disease**, is characterized mostly by a single, persistent, scaly plaque, commonly involving the limbs.
- **Granulomatous slack skin** is a rare type of CTCL where bulky, hanging skin folds are found in the armpit or groin.
- **Hypopigmented MF** is characterized by patches lighter than the surrounding skin.

**Mycosis fungoides**

**An example of mycosis fungoides skin lesions found on the arm.**
Credit: https://commons.wikimedia.org/wiki/File:Mycosis_fungoide.JPG



## Testing

A skin biopsy is needed to get the right diagnosis. In fact, multiple skin biopsies are needed in most cases. Tests are done on the biopsied cells to confirm the diagnosis and learn more about your type of mycosis fungoides. For tests to diagnose mycosis fungoides, see **Guide 2**.

In addition to skin biopsies, other tests are needed to plan treatment. For tests to plan treatment, see **Guide 3**.

## Goal of treatment

The goal of treatment is to improve your condition and to maintain this improvement for as long as possible. Treatment is based on the cancer stage, if any, what treatments were given before, and other factors. If lymphoma in the blood is suspected, then treatment may follow stage 3 (erythrodermic disease).

Treatment for mycosis fungoides based on cancer stage is explained next.

## Stage 1A

In stage 1A, cancer is limited to a small area on the skin. The amount of cancer is measured by evaluating what percent of your skin is affected by lymphoma. One hand is equal to 1 percent (1%) of your total body surface area (BSA). In stage 1A, less than 10% of the skin (BSA) is covered in patches, papules, and/or plaques.

### Treatment

Treatment for stage 1A mycosis fungoides is skin-directed therapy. A skin-directed therapy could be radiation therapy, phototherapy, or topical creams, gels, or ointments, such as topical steroids, imiquimod, mechlorethamine, retinoids, or carmustine.

**Guide 2**
**Tests to diagnose mycosis fungoides**

| | |
|---|---|
| **Needed** | • Skin biopsy of lymphoma sites and pathology review<br>• Immunophenotyping using immunohistochemistry (IHC) panel of skin biopsy that may include: CD2, CD3, CD4, CD5, CD7, CD8, CD20, CD30<br>• Biomarker testing to detect clonal *TRB* and *TRG* gene rearrangements |
| **In some cases** | • Flow cytometry and other tests to look for Sézary cells<br>• IHC panel of skin biopsy that may include: CD25, CD56, TIA1, granzyme B, TCR beta, TCR delta, CCR4, CXCL13, inducible T-cell co-stimulator (ICOS), and programmed cell death protein 1 (PD-1)<br>• Fluorescence in situ hybridization (FISH) and multigene panel testing<br>• Human T-lymphotropic virus (HTLV) testing |

Treatment response might be described in the following ways:

- **A complete response** is remission or a disease-free period.
- **In a partial response**, treatment is working, but lymphoma remains. You'll likely continue the same treatment until you have a complete response.

## Relapsed disease

When cancer returns after a disease-free period, it's called a relapse. Treatment is based on cancer stage.

## Disease progression

In disease progression, mycosis fungoides is growing or spreading. If it progresses higher than stage 1A, then treatment is based on the new stage.

## Persistent disease

Disease that persists, but hasn't progressed, may be treated with a different skin-directed therapy.

## Refractory disease

When mycosis fungoides appears resistant to multiple therapies, it's called refractory. Treatment might be skin-directed therapy with or without systemic therapy (see the next section, "Stage 1B and 2A"), radiation therapy if not used before, or a clinical trial.

### Guide 3
### Tests to plan treatment

| |
|---|
| Skin biopsy with pathology review and tumor/lesion testing |
| Medical history and physical exam, including complete skin exam (noting amount of body surface area affected and type of skin lesion, such as patch/plaque, tumor, or erythroderma) |
| Complete blood count with differential, comprehensive metabolic panel, and lactate dehydrogenase (LDH) |
| Whole body PET/CT scan (preferred) with CT with contrast of chest, abdomen, and pelvis |
| Distress screening |
| Possible:<br>• Bone marrow biopsy<br>• Lymph node biopsy or biopsy of other sites<br>• Neck CT with contrast if whole body PET/CT scan not done<br>• Pregnancy test for people of childbearing age<br>• Discussion of fertility preservation |

## Stage 1B and 2A

In stage 1B, patches, papules, and/or plaques cover 10% or more of the skin. There's no lymphoma in lymph nodes or visceral organs.

In stage 2A, any amount of the skin surface is covered with patches or plaques. There's some involvement of lymph nodes, but not enough to alter the structure of the lymph nodes. If lymphoma is found in the blood, it might be treated as stage 3 erythrodermic disease.

Treatment for stage 1B and 2A is based on the amount of skin disease called burden. The lower the amount of skin disease, the lower the burden.

### Lower skin disease burden

Lower skin disease burden is mostly patch disease—a limited area of the skin is involved. It's treated with skin-directed therapies, which may be used alone or in combination with other skin-directed therapies.

### Higher skin disease burden

Higher skin disease burden is mostly plaque disease. The goal of treatment is to improve your condition and to reduce the amount of cancer.

Treatment options include:

- Skin-directed therapies for general skin involvement.
- Systemic therapy alone or with skin-directed therapies. For systemic therapy options, see **Guide 4**.

### Relapsed disease

Treatment for relapse is based on the cancer stage, what treatments were given before, and other factors.

### Disease progression

If mycosis fungoides progresses higher than stage 1B or 2A, then treatment is based on the new stage.

### Persistent disease

Persistent disease may be treated with a different skin-directed therapy.

**Guide 4**
**Systemic therapies: Stage 1B and 2A**

| | |
|---|---|
| **Preferred therapies** | • Bexarotene<br>• Brentuximab vedotin<br>• Interferon alfa<br>• Methotrexate<br>• Mogamulizumab<br>• Romidepsin<br>• Vorinostat |
| **Therapies used in certain cases** | • Acitretin<br>• Denileukin diftitox-cxdl<br>• Extracorporeal photopheresis (ECP)<br>• Interferon gamma-1b<br>• Isotretinoin<br>• Alemtuzumab<br>• Gemcitabine<br>• Liposomal doxorubicin<br>• Pembrolizumab<br>• Pralatrexate |

### Refractory disease

Treatment options for refractory disease include:

- Clinical trial
- Total skin electron beam therapy (TSEBT), if not used before
- Treatments found under “Stage 2B,” explained next

## Stage 2B

Stage 2B is also called tumor stage disease. In this stage, 1 or more tumors 1 cm or larger in size are found on the skin. Treatment is based on if the tumors are limited or widespread. Abnormal T cells may be found in the lymph nodes and/or blood.

### Limited tumors

Treatment options include:

- Local radiation therapy (RT) and/or skin-directed therapy.
- Systemic therapy with or without local RT. Skin-directed therapy might be added. For systemic therapy options, see **Guide 5**.

### Widespread tumors

The goal is to reduce the amount of cancer in the body and to prevent further spread.

Treatment options include:

- TSEBT.
- Systemic therapy alone or with skin-directed therapy, or combination therapies.



**Let us know what you think!**

**Please take a moment to complete an online survey about the NCCN Guidelines for Patients.**
**NCCN.org/patients/response**

**Guide 5**
**Systemic therapies: Stage 2B limited**

| | |
|---|---|
| **Preferred therapies** | • Bexarotene<br>• Brentuximab vedotin<br>• Interferon alfa<br>• Methotrexate<br>• Mogamulizumab<br>• Romidepsin |
| **Other recommended therapies** | • Vorinostat<br>• Pembrolizumab |
| **Therapies used in certain cases** | • Acitretin<br>• Denileukin diftitox-cxdl<br>• Extracorporeal photopheresis (ECP)<br>• Interferon gamma-1b<br>• Isotretinoin |

- For preferred systemic therapy options, see **Guide 6**.

### Relapsed disease

Depending on the cancer stage, the same or another treatment might be given.

### Disease progression

If cancer spreads or advances to a stage higher than 2B, then the cancer will be restaged. Treatment will be based on the new stage.

### Persistent disease

Persistent disease is treated with a systemic therapy not used before.

### Refractory disease

When mycosis fungoides appears resistant to multiple therapies, it's called refractory.

Treatment options include:

- TSEBT
- Systemic therapy options under "Large cell transformation" on page 43
- Other systemic therapies not used before
- Clinical trial
- Allogeneic hematopoietic cell transplant (HCT)

**Guide 6**
**Systemic therapies: Stage 2B widespread lesions**

| | | |
|---|---|---|
| **Preferred therapies** | • Bexarotene<br>• Brentuximab vedotin<br>• Denileukin diftitox-cxdl<br>• Gemcitabine<br>• Interferon alfa | • Liposomal Doxorubicin<br>• Methotrexate<br>• Mogamulizumab<br>• Pralatrexate<br>• Romidepsin |
| | Combination therapy:<br>• Retinoid plus Interferon alfa | |
| **Other recommended therapies** | • Pembrolizumab<br>• Vorinostat | |
| **Therapies used in certain cases** | • Acitretin<br>• Extracorporeal photopheresis (ECP)<br>• Interferon gamma-1b<br>• Isotretinoin | |
| | Combination therapy:<br>• ECP plus Interferon alfa or retinoid<br>• ECP plus Interferon alfa plus retinoid | |

HCT is not an option for everyone. It works best when your lymphoma is responding to therapy.

## Stage 3

Stage 3 is also called erythrodermic disease. In erythrodermic disease, nearly all the skin is reddened (erythema). Cancer may be in lymph nodes (any N) or blood (B1). Treatment is systemic therapy with skin-directed therapy. Skin-directed therapy might include phototherapy, topical corticosteroids, topical mechlorethamine, and total skin electron beam therapy (TSEBT). For systemic therapy options, see **Guide 7**.

**Guide 7**
**Systemic therapies for stage 4 mycosis fungoides**

| | | |
|---|---|---|
| **Preferred therapies** | • Bexarotene<br>• Brentuximab vedotin<br>• Extracorporeal photopheresis (ECP) | • Interferon alfa<br>• Methotrexate<br>• Mogamulizumab<br>• Romidepsin |
| | Combination therapy:<br>• ECP plus Interferon alfa or retinoid<br>• ECP plus Interferon alfa plus retinoid<br>• Retinoid plus Interferon alfa | |
| **Other recommended therapies** | • Vorinostat | |
| **Therapies used in certain cases** | • Acitretin<br>• Alemtuzumab<br>• Denileukin diftitox-cxdl<br>• Gemcitabine<br>• Interferon gamma-1b | • Isotretinoin<br>• Liposomal Doxorubicin<br>• Pembrolizumab<br>• Pralatrexate |
| | Skin-directed therapy:<br>• Phototherapy<br>• TSEBT | |

### Relapsed disease

Relapsed disease is treated as stage 4.

### Disease progression

If cancer spreads or advances to a stage higher than 3, then it's treated as stage 4.

### Persistent disease

For persistent disease, treatment is a systemic therapy not used before.

### Refractory disease

Mycosis fungoides resistant to multiple therapies is treated with a clinical trial, a systemic therapy not used before, or an HCT.

**Systemic therapies for stage 4 mycosis fungoides**

**Preferred therapies:**

- Bexarotene
- Gemcitabine
- Liposomal Doxorubicin
- Pralatrexate
- Romidepsin

**Other recommended therapies:**

- Mogamulizumab
- Systemic therapies used to treat T-cell lymphomas

## Stage 4

Treatment for stage 4 mycosis fungoides is explained below. Stage $4A_1$ is Sézary syndrome and is discussed in *Chapter 6*: *Sézary syndrome.*

### Non-Sézary disease (stage $4A_2$)

In non-Sézary stage $4A_2$ disease, abnormal T lymphocytes have altered the structure of lymph nodes. Lymphoma may be in blood. Treatment is systemic therapy. Radiation therapy might be added to treat skin lesions. A clinical trial or allogeneic hematopoietic cell transplant (HCT) is possible. An HCT is not for everyone. Ask your care team why one treatment might be recommended over another.

### Visceral disease (stage 4B)

Visceral disease is lymphoma that's spread, or metastasized, to a solid organ, such as the spleen or liver. Imaging tests are used to confirm visceral disease and might be used to see how the lymphoma is responding to treatment.

In stage 4B, cancer has metastasized to internal (visceral) organs or bone marrow. Treatment is systemic therapy alone or with radiation therapy. Systemic therapy works throughout the body to reduce the amount of lymphoma in the organs and blood. It includes retinoids, chemotherapy, targeted therapy, and immunotherapy. Radiation therapy might be added to treat skin lesions. A clinical trial or allogeneic HCT is possible.

## Large cell transformation

Large cell transformation (LCT) occurs when a specific group of mycosis fungoides tumor cells undergo molecular and/or genetic changes that cause them to become larger. LCT can happen at any stage and is identified when large cells make up more than 25 percent (25%) of tumor cells in a skin or lymph node biopsy. This means that a large cell is found in more than 1 out of every 4 tumor cells. Typically, mycosis fungoides grows and progresses slowly, but sometimes it transforms into LCT and may become more aggressive. The goal of treatment is to slow the growth of transformed cells.

### Limited lesions with LCT

If there is a limited number of lesions with LCT, they might be treated with radiation therapy.

### Widespread lesions with LCT

Treatment options for widespread skin lesions with LCT include total skin electron beam therapy or systemic therapy with skin-directed therapy. Systemic therapy helps control LCT found in any organs and skin lesions. Multi-agent combination chemotherapy plus a hematopoietic cell transplant (autologous or allogeneic) is often needed to achieve lasting disease control. Imaging and other tests should be performed to evaluate responses.

### Relapse

You might have the same treatment as before, a clinical trial, or an allogeneic hematopoietic cell transplant (HCT).

### Systemic therapies for large cell transformation

**Preferred therapies:**

- Bexarotene
- Gemcitabine
- Liposomal Doxorubicin
- Pralatrexate
- Romidepsin
- Systemic therapies used to treat T-cell lymphomas

**Other recommended therapies:**

- Pembrolizumab

### Persistent disease

For LCT that persists, but hasn't progressed, treatment focuses on reducing the amount of lymphoma. It will likely be a different systemic therapy.

### Refractory disease

When cancer appears resistant to multiple therapies, it's called refractory. Treatment might be a clinical trial, systemic therapy, or an allogeneic HCT.

## Key points

- In stage 1A, cancer is limited to a small area on the skin. Less than 10 percent (10%) of the skin (body surface area, or BSA) is covered in patches, papules, and/or plaques. Treatment is skin-directed therapy.
- Treatment for stage 1B and 2A is based on the amount of skin disease.
- Stage 2B is also called tumor stage disease. Treatment is based on if the tumors are limited or widespread.
- Stage 3 is also called erythrodermic disease. In erythrodermic disease, nearly all of the skin is reddened (erythema). Treatment is systemic therapy with skin-directed therapy.
- In stage $4A_2$ disease, abnormal T lymphocytes have altered the structure of lymph nodes. Cancer may be found in blood.
- In stage 4B visceral disease, cancer is found in a solid, internal organ, such as the spleen or liver, or in the bone marrow.
- Large cell transformation can happen at any stage and is identified by the presence of large cells on a skin or lymph node biopsy.

## Questions to ask

- What is the mycosis fungoides stage? What does this mean?
- Do I have skin-only disease? Is it limited or widespread?
- Is there lymphoma in the lymph nodes, internal organs, or blood?
- What treatment do you recommend and why? Can I manage the treatment at home, or do I need to come to the clinic?
- What are the side effects of the recommended treatment?

## What's next?

The next chapter explains diagnosis and treatment of Sézary syndrome.

# 6
# Sézary syndrome


46 Overview

46 Treatment

49 Key points

49 Questions to ask

In Sézary syndrome, cancerous T cells called Sézary cells may be found in the skin, blood, and lymph nodes. Cancer may be found in a solid, internal organ, such as the liver or spleen, or in the bone marrow. Treatment is a combination of systemic therapy with skin-directed therapy.

## Overview

Sézary syndrome (SS) is a rare type of cutaneous T-cell lymphoma (CTCL). It's closely related to mycosis fungoides (MF) but has its own unique features. SS often causes skin redness (erythema) over most of the body, called erythroderma. In SS, abnormal T cells called Sézary cells are found in the skin and blood and may cause swollen and enlarged lymph nodes (lymphadenopathy). A characteristic of Sézary cells is an abnormal, brain-shaped nucleus, described as cerebriform. In the skin, SS and MF can look alike. Blood tests are used to tell the difference. For a person to have SS, Sézary cells must be present in the blood.

SS uses the same staging system as mycosis fungoides but is treated differently. SS can be stage $4A_1$, $4A_2$, or 4B. A diagnosis of SS requires B2 blood involvement, which means that a high percentage of Sézary cells are found in blood. Cancer may be found in bone marrow or internal organs.

## Treatment

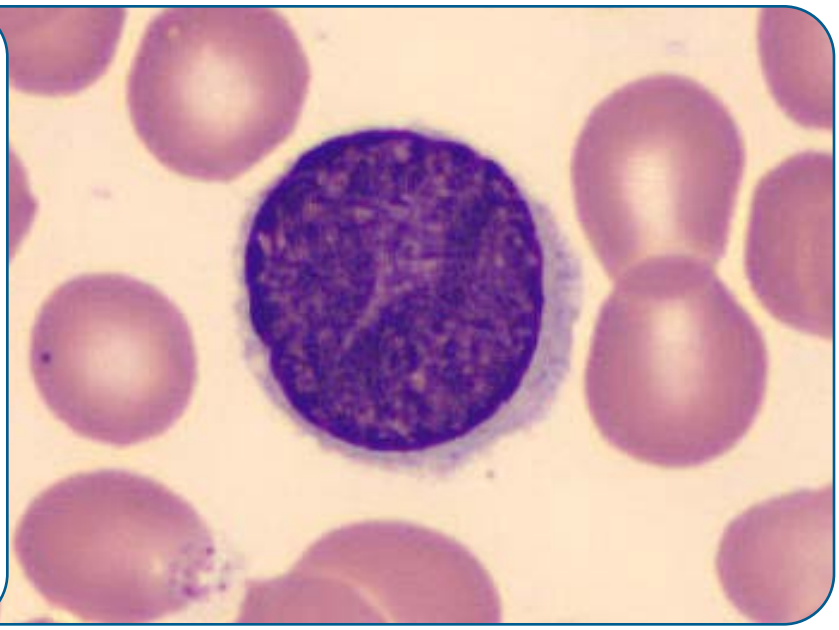
Treatment is based on whether disease burden is low or high. Burden refers to the amount of disease in the body.

- **In low blood tumor burden,** more than 250/μL but less than 1,000/μL of peripheral (circulating) blood lymphocytes are Sézary cells.

**Sézary syndrome**

**A characteristic of Sézary cells is an abnormal, brain-shaped nucleus, described as cerebriform. For a person to have Sézary syndrome, Sézary cells must be present in the blood.**

Credit: https://commons.wikimedia.org/wiki/File:Hem1SezaryCell.jpg



- **In high blood tumor burden**, more than 1,000/μL Sézary cells are found. A diagnosis of SS requires B2 involvement.

The goal of treatment is to reduce disease burden by combining skin-directed therapy with treatment that works from inside the body (systemic therapy). Treatment options are listed in **Guide 8**.

**Guide 8**
**Sézary syndrome: Treatment options based on disease burden**

| | Low to intermediate burden | High burden |
|---|---|---|
| **Preferred therapy** | • Bexarotene<br>• Extracorporeal photopheresis (ECP)<br>• Interferon alfa<br>• Methotrexate<br>• Mogamulizumab<br>• Romidepsin<br>• Vorinostat<br>• ECP plus interferon alfa or retinoid<br>• ECP plus interferon alfa plus retinoids<br>• Retinoid plus interferon alfa | • Mogamulizumab<br>• Romidepsin<br>• ECP plus interferon alfa or retinoid<br>• ECP plus interferon alfa plus retinoids<br>• Retinoid plus interferon alfa |
| **Other recommended therapies** | • Alemtuzumab<br>• Brentuximab vedotin<br>• Gemcitabine<br>• Liposomal Doxorubicin<br>• Pembrolizumab<br>• Pralatrexate | • Alemtuzumab<br>• Bexarotene<br>• Brentuximab vedotin<br>• ECP<br>• Gemcitabine<br>• Interferon alfa<br>• Liposomal Doxorubicin<br>• Methotrexate<br>• Pembrolizumab<br>• Pralatrexate<br>• Vorinostat |
| **Therapies used in certain cases** | • Acitretin<br>• Interferon gamma-1b<br>• Isotretinoin | • Acitretin<br>• Interferon gamma-1b<br>• Isotretinoin |

## Relapse

When cancer returns after a disease-free period, it's called a relapse. Relapse can happen after a complete or partial response. Treatment is based on disease burden. It might be the same treatment as before or a different one from the list.

## Persistent

Persistent disease should be treated with a different systemic therapy from before.

## Refractory

When lymphoma appears resistant to multiple therapies, it's called refractory.

Treatment options include:

- Clinical trial
- Systemic therapy
- Allogeneic HCT

An HCT is not for everyone. Ask your care team which option might be best for your type of SS. Your wishes are always important.

## Visceral disease

Visceral disease is lymphoma that has spread, or metastasized, to a solid, internal organ, such as the spleen or liver. Lymphoma might also have spread to the bone marrow, lymph nodes, or blood. Imaging tests are used to confirm visceral disease and might be used to see how your body is responding to treatment.

Treatment is systemic therapy alone or with radiation therapy. Systemic therapy works throughout the body to reduce the amount of lymphoma in the organs and blood. Radiation therapy might be used to treat skin lesions.

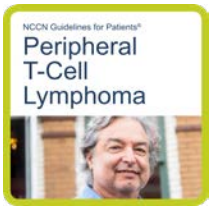
### Systemic therapy options after multiple therapies

- Alemtuzumab
- Chlorambucil
- Cyclophosphamide
- Duvelisib
- Etoposide
- Lenalidomide
- Pembrolizumab
- Pentostatin
- Temozolomide
- Other combined chemotherapies

Preferred systemic therapy options for visceral disease include:

- Brentuximab vedotin
- Gemcitabine
- Liposomal doxorubicin
- Pralatrexate
- Romidepsin

Other recommended systemic therapy options are mogamulizumab and those found in *NCCN Guidelines for Patients: Peripheral T-Cell Lymphoma,* available at NCCN.org/patientguidelines and on the NCCN Patient Guides for Cancer app.



Visceral disease that relapses may be treated with the same treatment as before, a clinical trial, or an allogenic HCT.

For visceral disease that persists or is resistant to multiple therapies, options include a clinical trial, an allogenic HCT, or therapies used for peripheral T-cell lymphoma.

## Key points

- In Sézary syndrome (SS), abnormal T cells, called Sézary cells, are found in the skin and blood.
- SS often causes skin redness (erythema) over most of the body called erythroderma.
- Enlarged lymph nodes (lymphadenopathy) are common.
- Treatment is a combination of systemic (drug) therapy with skin-directed therapy.
- The goal of treatment is to reduce the amount of lymphoma in the body and to improve your condition.
- A clinical trial or allogeneic hematopoietic cell transplant are possible treatment options.

## Questions to ask

- What is your experience treating cutaneous lymphomas?
- Is there a cancer center or hospital nearby that specializes in treating cutaneous lymphomas?
- How long will treatment take, and in what order will it be given?
- Will I spend time in the hospital?
- Are there resources to help pay for treatment or other care I may need?

## What's next?

The next chapter explains diagnosis and treatment of a group of skin diseases that test positive for a protein called CD30.

# 7
# CD30+ PCTLD


51 Overview

52 Primary cutaneous ALCL

53 Lymphomatoid papulosis

54 Key points

55 Questions to ask

In primary cutaneous CD30+ T-cell lymphoproliferative disorders (PCTLDs), T-cell lymphocytes that are CD30-positive (CD30+) grow and cause skin lesions or nodules. Types of PCTLD include cutaneous anaplastic large cell lymphoma, lymphomatoid papulosis, and borderline cases.

**Keeping a photo journal might help track skin changes over time.**

## Overview

Primary cutaneous CD30+ T-cell lymphoproliferative disorders (PCTLDs) are a group of diseases whose cells test positive for the CD30 protein (CD30+). These CD30+ cells grow out of control (proliferate) and cause skin lesions or nodules. Typically, more than 75 percent (75%) of T cells in PCTLD are CD30+. This means that at least 3 out of every 4 abnormal T cells have the CD30 protein.

Types of PCTLDs include cutaneous anaplastic large cell lymphoma (ALCL), lymphomatoid papulosis (LyP), and borderline cases with overlapping features. Other types of skin lymphomas, such as mycosis fungoides, can be CD30+. Non-skin lymphomas can also be CD30+. Therefore, an accurate diagnosis is needed to determine the appropriate treatment. Diagnosis is based on the appearance of the lesions and how the disease behaves over time. A skin biopsy might be done.

Types of PCTLDs discussed in this chapter include:

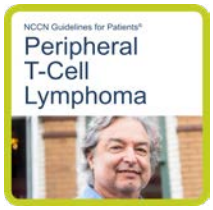
- **Primary cutaneous anaplastic large cell lymphoma (PC-ALCL)** is defined by the presence of anaplastic large cells. Lesions appear on the skin as 1 or more raised, red lesions or nodules. Anaplastic cells are fast-growing, abnormal cells. PC-ALCL starts in the skin and may spread to the lymph nodes.
- **Lymphomatoid papulosis (LyP)** is a benign, chronic, recurring, self-healing condition. Papulonodular skin lesions appear on the skin as small bumps and nodules.

Mycosis fungoides or Sézary syndrome that's CD30+ isn't the same as PCTLD. In addition, a person can have both PCTLD and mycosis fungoides. This is why getting an accurate diagnosis is so important.

# Primary cutaneous ALCL

Anaplastic large cell lymphoma (ALCL) can be found in the skin, lymph nodes, or organs throughout the body. When ALCL appears in the skin, it's called primary cutaneous ALCL (PC-ALCL) or cutaneous ALCL. PC-ALCL may also be found in a nearby (regional) lymph node. Sometimes, cutaneous ALCL can extend beyond the skin and lymph nodes to organs. If this occurs, it's usually treated as systemic ALCL.

For treatment of systemic ALCL, see *NCCN Guidelines for Patients: Peripheral T-Cell Lymphoma* at NCCN.org/patientguidelines and on the NCCN Patient Guides for Cancer app.




## Treatment

PC-ALCL is often a slow-growing (indolent), persistent disease with lesions that may go away (regress) in 2 out of 5 cases. Lesions often return but respond well to treatment. Not everyone's disease responds to treatment the same way. Treatment for PC-ALCL is based on the number of lesions and their location and will be personalized for you.

### 1 lesion or lesions grouped together

Treatment aims to remove the lesion. This can be achieved using involved-site radiation therapy (ISRT), surgery (excision), or both. ISRT is more common.

After a response to treatment, you'll be monitored for recurrence. Recurrence or relapse is the return of 1 or more lesions. If a lesion returns and is confined to the skin, then you can be re-treated with ISRT, surgery, or both. Sometimes, observation (no treatment) is an option if the lesions aren't causing symptoms (asymptomatic). If there's no response or lesions seem resistant (refractory) to treatment, then you'll be treated as described below in "Multiple lesions."

### Multiple lesions

When there are lesions in multiple locations, the preferred treatment is a systemic therapy called brentuximab vedotin (Adcetris). Other systemic therapies might be given alone or with skin-directed therapy.

### PC-ALCL with lymph node involvement

PC-ALCL may involve the skin and nearby (regional) lymph nodes. A lymph node biopsy is needed to confirm disease. Treatment aims to reduce the amount of skin lesions and disease in lymph nodes. The preferred treatment is brentuximab vedotin (Adcetris) with or without involved-site radiation therapy (ISRT) to lymph nodes and primary skin lesions. ISRT may be used alone in some cases. Other systemic therapies might be given.

### Relapsed or refractory disease

After a response to treatment, you'll be observed for recurrence. Recurrence or relapse is the return of disease. When cancer appears resistant to multiple therapies, it's called refractory.

For relapse, no response, or lesions that seem resistant (refractory) to treatment, then options include:

- Clinical trial
- Same treatment as before
- Different treatment from before
- Other systemic therapies, such as those discussed in “Large cell transformation” in *Chapter 5: Mycosis fungoides.*

## Lymphomatoid papulosis

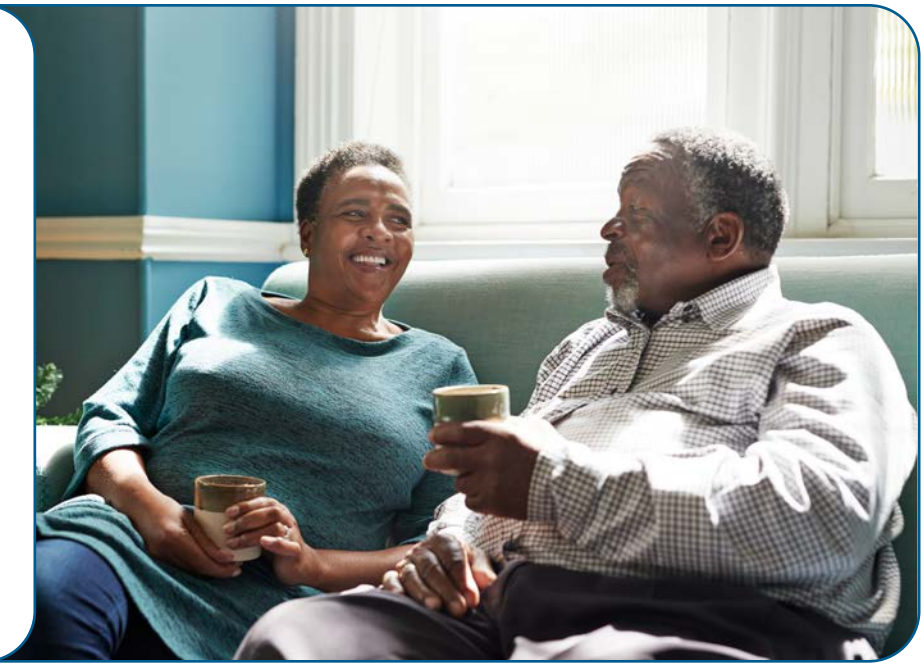
Lymphomatoid papulosis (LyP) is a benign, chronic, recurring disease of the immune system. Lesions appear on the skin as small bumps and nodules, called papulonodular skin lesions. LyP is a primary cutaneous CD30+ T-cell lymphoproliferative disorder. Lymphoproliferative disorders are often treated like cancer, but they are not cancer.

### Treatment for limited lesions

Treatment is based on if the lesions are causing symptoms. If you don’t have symptoms, you are asymptomatic. Observation is the preferred option for asymptomatic people with limited lesions. Topical steroids are also an option. If you have symptoms, treatment aims to reduce these symptoms using topical steroids or phototherapy. Observation is also an option.

- **If disease is responding to treatment**, then you’ll continue the same treatment.
- **If there’s no response or disease is refractory**, then the options are a different treatment or clinical trial. Refractory means disease is not responding to treatment.

**Standard of care is the best-known way to treat a particular disease based on past clinical trials. There may be more than one treatment that’s considered standard of care. Ask your care team what treatment options are available and if a clinical trial might be right for you.**



### Treatment for widespread lesions

Treatment for widespread lesions aims to reduce the number of lesions and any discomfort they may cause. This may be done using methotrexate, phototherapy, systemic retinoids, topical steroids, or nitrogen mustard. Observation is preferred in people without symptoms (asymptomatic).

- **If disease is responding to treatment**, then you'll be observed for recurrence. Recurrence or relapse is the return of disease.
- **For relapse**, options are a clinical trial, observation, the same treatment as before, or a different treatment from before.
- **If there's no response or disease is refractory**, then the options are a treatment not used before or a clinical trial.

### Monitoring

People with LyP are at risk of developing another type of lymphoma. Lifelong follow-up is needed. Your doctor should conduct a thorough skin exam during each follow-up visit. Ask how you'll be monitored and how often you should have a checkup.

## Key points

- In primary cutaneous CD30+ T-cell lymphoproliferative disorders (PCTLDs), T-cell lymphocytes that are CD30-positive (CD30+) grow and cause skin lesions or nodules.
- Typically, in PCTLD, more than 75 percent (75%) of T cells are CD30+. This means that at least 3 out of every 4 abnormal T cells have the CD30 protein.
- Types of PCTLD include primary cutaneous anaplastic large cell lymphoma, lymphomatoid papulosis, and borderline cases.
- Other types of skin lymphomas, such as mycosis fungoides, can be CD30+. Non-skin lymphomas can also be CD30+. Therefore, an accurate diagnosis is needed to determine the appropriate treatment.
- Primary cutaneous anaplastic large cell lymphoma (PC-ALCL) is a slow-growing, persistent disease that starts in the skin and may spread to lymph nodes.
- Lymphomatoid papulosis (LyP) is a benign, chronic, recurring disease of the immune system. Lesions appear on the skin as small bumps and nodules, called papulonodular skin lesions.
- People with LyP are at risk of developing another type of lymphoma. Lifelong follow-up is needed.

## Questions to ask

- What type of CD30+ lymphoproliferative disorder do I have? Is it cancer or benign?
- Can I wait to start treatment? What treatment do you recommend?
- What side effects can I expect from this treatment and how will they be managed?
- If this is a chronic disease, how will it be treated over time?
- How often should I see a dermatologist? Is there a dermatologist you recommend?

## What's next?

The next chapter explains diagnosis and treatment of a type of T-cell lymphoma that grows in the fat under the skin causing lumps that can be confused with inflammation and other health conditions.



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**NCCN.org/patients/feedback**

# 8

# Subcutaneous panniculitis-like T-cell lymphoma


57 Overview

57 Testing

58 Treatment

61 Key points

61 Questions to ask

In subcutaneous panniculitis-like T-cell lymphoma (SPTCL), abnormal lymphocytes grow in the fat under the skin, causing deep nodules. SPTCL is usually found in the legs, but lesions may develop anywhere on the body. SPTCL is difficult to diagnose because it looks and acts like other conditions.

## Overview

In subcutaneous panniculitis-like T-cell lymphoma (SPTCL), abnormal white blood cells grow in the fat under the skin, causing lumps (nodules) to form. It's a slow-growing lymphoma that looks and acts like fat inflammation. Nodules and plaques are often found in the legs, but lesions may develop anywhere on the body. Common symptoms (called B symptoms) include fever, fatigue, and excessive sweating during sleep.

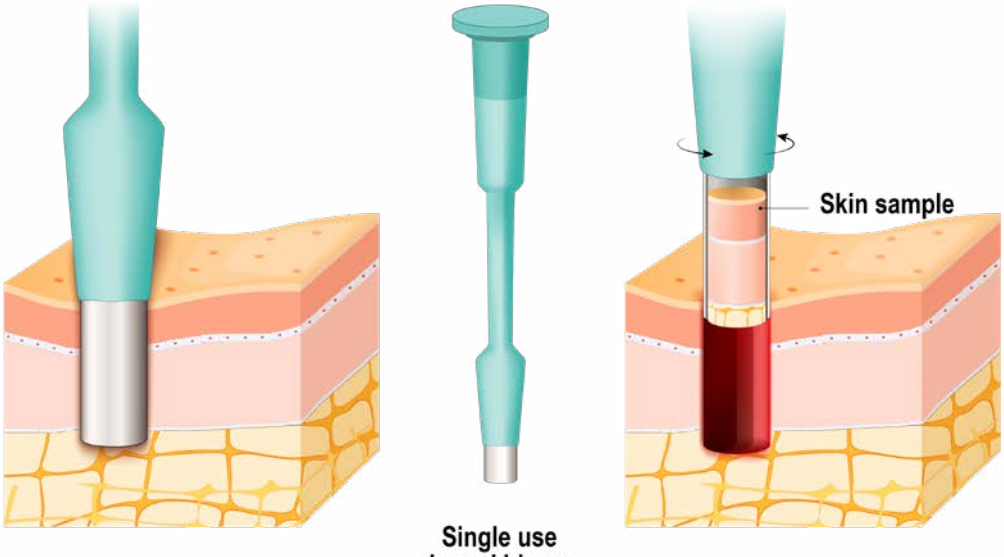
People with SPTCL tend to be in their 30s and may have a personal or family history of autoimmune disorders, such as systemic lupus erythematosus.

## Testing

A precise diagnosis of SPTCL is challenging and requires several methods for an accurate diagnosis. These methods include a deep skin biopsy with immunophenotyping, blood tests to rule out immune and rheumatoid conditions, and imaging.

### Skin punch biopsy

**To diagnose subcutaneous panniculitis-like T-cell lymphoma (SPTCL), a skin punch biopsy must be deep enough to collect fat cells underneath the skin.**



People with SPTCL generally have multiple, painless, red or dark nodules or plaques on the legs, arms, or torso. One or more of these areas may be biopsied. A skin biopsy needs to go deep enough to collect fat cells. Tests, such as immunophenotyping, are done on the biopsied cells. SPTCL is commonly CD3+, CD8+, βF1+, CD2+, CD5+, and CD7+.

Tests to diagnose SPTCL can be found in **Guide 9**.

## Treatment

Developing a treatment plan for SPTCL can be challenging. SPTCL is a rare lymphoma that can be complicated by the presence of an immune condition called hemophagocytic lymphohistiocytosis (HLH). About 1 in 4 people with SPTCL also have this rare, severe, and life-threatening immune system disorder.

Treatment options are based on the presence of HLH and if the tumors are limited or widespread.

In HLH, the body overreacts, causing white blood cells to damage organs like the liver and spleen. Fever and enlargement of the spleen are the most common symptoms of HLH. There are many other possible symptoms.

If you have HLH, certain white blood cells—histiocytes and lymphocytes—attack your other blood cells. These abnormal blood cells collect in your spleen and liver, causing these organs to enlarge. If you have SPTCL with HLH, then you'll be treated for both.

**Guide 9**
**Tests to diagnose SPTCL**

| | |
|---|---|
| **Needed** | • Deep skin biopsy (including fat cells) of 1 or more lesions and pathology review<br>• Immunophenotyping using immunohistochemistry (IHC) panel that may include: TCR beta, TCR delta, CD2, CD3, CD20, CD4, CD5, CD7, CD8, CD30, and CD56<br>• Epstein-Barr virus-encoded RNA in situ hybridization (EBER-ISH)<br>• Biomaker testing to detect clonal *TRB* and *TRG* gene rearrangements or clonality |
| **In some cases** | • Additional IHC panel to establish subtype may include Ki-67, CD123, TIA1, perforin, granzyme-B, CD1a, TdT, TCL1<br>• Flow cytometry to rule out other T-cell lymphoma subtypes<br>• Germline testing for *HAVCR2* mutation<br>• Multigene panel testing |

Tests to plan treatment can be found in **Guide 10**.

## SPTCL with HLH or widespread disease

SPTCL with HLH or SPTCL that's widespread is treated with cyclosporine, pralatrexate, or romidepsin. Prednisone might be added. Another option is etoposide-based chemotherapy.

- **If disease responds to treatment**, then you'll be observed for recurrence. You might also be given cyclosporine, pralatrexate, romidepsin, methotrexate, or bexarotene at a lower dose.
- **If disease doesn't respond**, then you'll have a treatment not used before.

## SPTCL without HLH and limited disease

Treatment options for new or returning SPTCL include cyclosporine, methotrexate, or bexarotene. Prednisone might be added. Involved-site radiation therapy or intralesional steroids are also an option.

- For lesions that aren't responding to treatment, a different therapy not used before, and pralatrexate or romidepsin with or without prednisone, are possible.
- For lesions that grow during treatment or persist, a clinical trial or an etoposide-based combination chemotherapy will be considered.

After treatment, you might be observed. It's also possible you'll continue taking

**Guide 10**
**Tests to plan treatment**

| |
|---|
| Skin biopsy with pathology review and tumor/lesion testing |
| Medical history, family history, and physical exam, including complete skin exam |
| Complete blood count with differential, comprehensive metabolic panel, lactate dehydrogenase (LDH), iron, and other blood tests |
| Whole body FDG-PET/CT scan |
| Distress screening |
| Possible:<br>• Pregnancy test for people of childbearing age<br>• Epstein-Barr virus testing<br>• Blood tests to rule out autoimmune conditions<br>• Bone marrow biopsy<br>• Biopsy of enlarged lymph nodes or suspected lymphoma sites |

cyclosporine, methotrexate, or bexarotene, but at a lower dose.

### Relapse

SPTCL can relapse more than once. For a relapse, you might have the same treatment as before or a different treatment, depending on the length of time that has passed since your last treatment.

### After multiple previous therapies

After multiple relapses or if SPTCL is resistant to multiple therapies, options include a clinical trial, an allogeneic hematopoietic cell transplant, or other systemic therapies.

## Systemic therapy options after multiple therapies

**Preferred therapies:**

- Belinostat
- Brentuximab vedotin if CD30+
- Duvelisib
- Pralatrexate
- Romidepsin

**Other recommended therapies:**

- Alemtuzumab
- Bendamustine
- Cyclophosphamide and/or Etoposide
- Gemcitabine
- Lenalidomide
- Ruxolitinib

## Key points

- Subcutaneous panniculitis-like T-cell lymphoma (SPTCL) grows in the fat under the skin, causing painless red or dark nodules or plaques on the legs, arms, or torso to form. SPTCL looks and acts like fat inflammation but is made of abnormal white blood cells.
- A deep skin biopsy that includes the layer of fat under the skin is an important part of diagnosis.
- Immunophenotyping helps determine SPTCL from other types of lymphoma and non-lymphoma conditions.
- A person can have both SPTCL and an immune system disorder called hemophagocytic lymphohistiocytosis (HLH). In HLH, white blood cells overreact, causing damage to organs like the liver and spleen.
- If you have SPTCL with HLH, then you'll be treated for both at the same time.
- Systemic (drug) therapy is used to treat SPTCL. Radiation therapy or intralesional steroids might be used to treat small lesions.

## Questions to ask

- Do I have SPTCL with HLH? How does this affect my treatment options?
- What treatment order do you recommend and why?
- Can I work during treatment?
- Will I spend time in the hospital during treatment?
- After completing treatment, what follow-up testing will I have?

## What's next?

The next chapter provides information on slow-growing skin disorders that don't cause symptoms and tend to regress (go away) after biopsy.

# 9

# Primary cutaneous indolent LPD


63 Overview

63 Testing

63 Treatment

64 Key points

64 Questions to ask

Primary cutaneous indolent lymphoproliferative disorders (LPDs) appear as a single papule, nodule, or plaque. They usually don't cause symptoms, tend to regress (go away) after biopsy, and rarely return. Lymphoproliferative disorders are often treated like cancer, but they are not cancer.

## Overview

In primary cutaneous indolent lymphoproliferative disorders (LPDs), abnormal but not cancerous cells form a single, isolated nodule or plaque.

There are 2 main types of primary cutaneous indolent LPDs:

- Primary cutaneous CD4+ small/medium T-cell LPD (PCSM-TLPD)
- Primary cutaneous CD8+ acral T-cell LPD (PCA-TLPD)

## Testing

Testing is needed to diagnose primary cutaneous indolent LPDs and to distinguish them from other types of lymphoid disorders. A skin biopsy will be taken and tested.

## Treatment

Primary cutaneous indolent LPDs tend to go away on their own after a biopsy. Doctors call this spontaneous remission. These types of LPDs usually don't cause symptoms. If you

**"Do whatever you can to connect with a support group. Having a support network of people with similar experiences will help you navigate your diagnosis with confidence. Knowing that you aren't the only one to be going through this is priceless."**



have symptoms, treatment options include topical or intralesional steroids, low-dose radiation therapy, or surgery to remove the lesion. Systemic (drug) therapy isn't needed and can be avoided because this type of skin lesion doesn't spread and isn't cancer. However, it's important to have regular skin exams and report any skin changes to your health care provider.

## Key points

- Primary cutaneous indolent lymphoproliferative disorders (LPDs) appear as a single papule, nodule, or plaque.
- There are 2 main types of primary cutaneous indolent LPDs: primary cutaneous CD4+ small/medium T-cell LPD and primary cutaneous CD8+ acral T-cell LPD.
- A skin biopsy is an important part of diagnosis.
- Immunophenotyping helps distinguish primary cutaneous indolent LPDs from other diseases or conditions.
- Primary cutaneous indolent LPDs tend to go away on their own after a biopsy. Doctors call this spontaneous remission.
- Treatment can wait until you have symptoms. If you have symptoms, treatment options include topical or intralesional steroids, low-dose radiation therapy, or surgery to remove the lesion.
- These types of LPDs don't spread to other sites in the body and rarely return.

## Questions to ask

- Can I wait to start treatment?
- How will I know it's time to start treatment?
- What treatment do you recommend and why?
- How often should I see a dermatologist?
- Is my LPD related to another condition or disorder?

## What's next?

The next chapter provides information about resources and where you can get support.

# 10
# Other resources

66 What else to know

66 What else to do

66 Where to get help

67 Questions to ask

Want to learn more? Here's how you can get additional help.

## What else to know

This guide helps you know your options so you can make informed decisions and improve your cancer care. But it's not the only resource that you have.

Ask for as much information and help as you need. Many people are interested in learning more about:

- Finding a health care team that's experienced in treating lymphomas of the skin
- Understanding treatment options, including participating in a clinical trial
- Coping with the stress of having lymphoma
- Getting financial help and other types of support

## What else to do

Your health care center can help you with next steps. It often has on-site resources to help meet your needs and find answers to your questions. Health care centers can also inform you of resources in your community.

In addition to help from your providers, the resources listed in the next section provide support for many people like yourself.

## Where to get help

Look through the list below and visit the provided websites to learn more about these organizations.

**Blood Cancer United**
bloodcancerunited.org/PatientSupport

**Cancer*Care***
cancercare.org

**Cutaneous Lymphoma Foundation**
clfoundation.org

**GRACE**
cancergrace.org

**Imerman Angels**
imermanangels.org

**Stupid Cancer**
stupidcancer.org

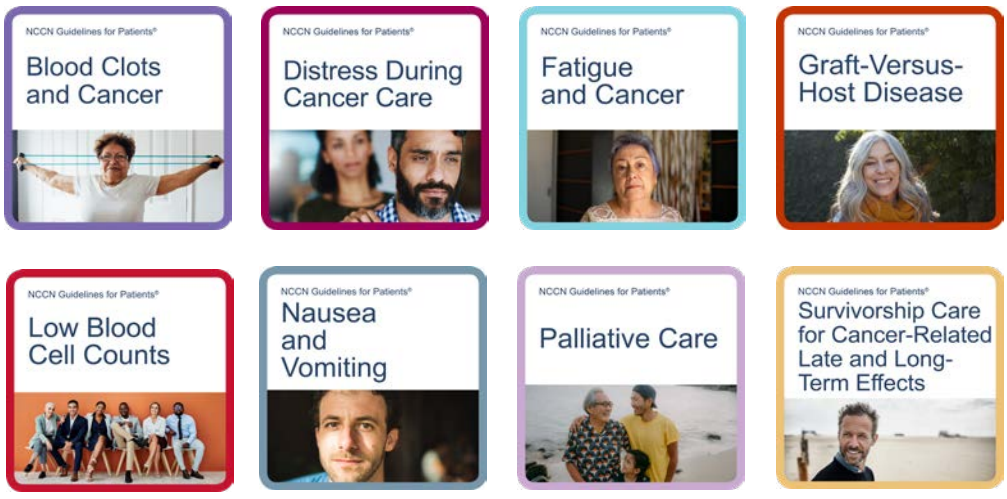
**Triage Cancer**
triagecancer.org

## Questions to ask

- Who can I talk to about help with housing, food, and other basic needs?
- What help is available for transportation, childcare, and home care?
- What other services are available to me and my caregivers?
- How can I connect with others and build a support system?
- Who can I talk to if I don't feel safe at home, at work, or in my neighborhood?

### Supportive care resources

More information on supportive care is available at NCCN.org/patientguidelines and on the NCCN Patient Guides for Cancer app.





# Words to know

**biomarker testing**
A lab test of any molecule in your body. Also called molecular testing.

**biopsy**
A procedure that removes fluid or tissue samples to be tested for a disease.

**body surface area (BSA)**
The measured, or calculated, surface area of the human body. One hand is equal to 1 percent (1%) of your total body surface area.

**chromosome**
Structures within cells that contain coded instructions for cell behavior.

**clinical trial**
A type of research that assesses health tests or treatments.

**computed tomography (CT)**
A test that uses x-rays from many angles to make a picture of inside the body.

**contrast**
A substance put into your body to make clearer pictures during imaging tests.

**cutaneous**
Having to do with the skin.

**deoxyribonucleic acid (DNA)**
A chain of chemicals in cells that contains coded instructions for making and controlling cells.

**dermatologist**
A doctor who specializes in the diagnosis and treatment of skin diseases.

**erythema**
Reddening of the skin.

**erythroderma**
Redness of over 80 percent (80%) of the body's skin surface.

**extracorporeal photopheresis (ECP)**
A procedure where blood is removed from the body and treated with ultraviolet light and drugs that become active when exposed to light. The blood is then returned to the body.

**flow cytometry**
A lab test of substances on the surface of cells to identify the type of cells present.

**fluorescence in situ hybridization (FISH)**
A lab test that uses special dyes to look for abnormal chromosomes and genes.

**gene**
Coded instructions in cells for making new cells and controlling how cells behave.

**histology**
The structure of cells, tissue, and organs as viewed under a microscope.

**imaging test**
A test that makes pictures (images) of the insides of the body.

**immune system**
The body's natural defense against infection and disease.

**immunohistochemistry (IHC)**
A lab test of cancer cells to find specific traits involved in abnormal cell growth.

**immunophenotyping**
A lab test that detects the type of cells present based on the cells' surface proteins.

**involved-site radiation therapy (ISRT)**
Radiation therapy that treats cancer found in a small region or 1 area of the body. It can also be used to treat specific lymph nodes with cancer.

**lactate dehydrogenase (LDH)**
A protein in blood that helps to make energy in cells.

**large cell transformation (LCT)**
When a specific group of mycosis fungoides tumor cells undergo molecular and/or genetic changes that cause them to become larger.

**lymph node**
A small, bean-shaped, disease-fighting structure.

**lymphocyte**
A type of white blood cell that is part of the immune system.

**mutation**
An abnormal change in the instructions within cells for making and controlling cells.

**nodule**
A firm, dome-shaped mass at least 1 centimeter in size (higher than a papule). Also called a tumor.

**papule**
A small, solid, raised bump on the skin that might look like a small pimple. Papules may be red, purple, brown, or pink.

**papulonodular**
A combination of papules and nodules found on the skin.

**patch**
A flat, thin, pink or red skin lesion of any size.

**pathologist**
A doctor who is an expert in testing cells and tissue to find disease. Also called a hematopathologist.

**plaque**
A raised (elevated) or hardened (indurated) skin lesion of any size.

**positron emission tomography (PET)**
A test that uses radioactive material to see the shape and function of body parts.

**prognosis**
The likely course a disease will take.

**progression**
Growth or spread of lymphoma.

**radiation therapy (RT)**
Treatment that uses high-energy rays or related approaches to kill lymphoma cells.

**refractory**
Lymphoma that doesn't respond to multiple treatments.

**relapse**
The return of lymphoma.

**remission**
No signs of lymphoma.

**retinoids**
Treatment that uses vitamin A. Can be a topical or systemic treatment.

**side effect**
An unhealthy or unpleasant physical or emotional response to treatment.

**skin-directed therapy**
Treatment that focuses on the skin, such as topical therapy and local radiation therapy.

**supportive care**
Health care that includes symptom relief but not cancer treatment. Also called palliative care or best supportive care.

**systemic therapy**
Treatment that works throughout the body.

**total skin electron beam therapy (TSEBT)**
A type of radiation therapy using electrons that's directed at the entire surface of the body. This type of radiation goes into the outer layers of the skin.

**translocation**
Switching of parts between 2 chromosomes.

**tumor**
A firm, dome-shaped mass at least 1 centimeter in size (higher than a papule). Also called a nodule.




**Take our survey and help make the NCCN Guidelines for Patients better for everyone!**

NCCN.org/patients/comments

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For contact information, visit NCCN.org/cancercenters.

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*Philadelphia, Pennsylvania*

Case Comprehensive Cancer Center/
University Hospitals Seidman Cancer Center and
Cleveland Clinic Taussig Cancer Institute
*Cleveland, Ohio*

City of Hope National Medical Center
*Duarte, California*

Dana-Farber Cancer Institute
*Boston, Massachusetts*

Duke Cancer Institute
*Durham, North Carolina*

Fox Chase Cancer Center
*Philadelphia, Pennsylvania*

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*Omaha, Nebraska*

Fred Hutchinson Cancer Center
*Seattle, Washington*

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Johns Hopkins Kimmel Cancer Center
*Baltimore, Maryland*

Mass General Brigham Cancer Institute
*Boston, Massachusetts*

Mayo Clinic Comprehensive Cancer Center
*Phoenix/Scottsdale, Arizona*
*Jacksonville, Florida*
*Rochester, Minnesota*

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Moffitt Cancer Center
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*Chicago, Illinois*

Roswell Park Comprehensive Cancer Center
*Buffalo, New York*

Siteman Cancer Center at Barnes-Jewish Hospital
and WashU Medicine
*St. Louis, Missouri*

St. Jude Children's Research Hospital/
The University of Tennessee Health Science Center
*Memphis, Tennessee*

Stanford Cancer Institute
*Stanford, California*

The Ohio State University Comprehensive Cancer Center -
James Cancer Hospital and Solove Research Institute
*Columbus, Ohio*

The UChicago Medicine Comprehensive Cancer Center
*Chicago, Illinois*

The University of Texas MD Anderson Cancer Center
*Houston, Texas*

UC Davis Comprehensive Cancer Center
*Sacramento, California*

UC San Diego Moores Cancer Center
*La Jolla, California*

UCLA Jonsson Comprehensive Cancer Center
*Los Angeles, California*

UCSF Helen Diller Family Comprehensive Cancer Center
*San Francisco, California*

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*Aurora, Colorado*

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Vanderbilt-Ingram Cancer Center
*Nashville, Tennessee*

Yale Cancer Center/Smilow Cancer Hospital
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# Index

**anaplastic large cell lymphoma (ALCL)** 51–52

**biomarkers** 15–17

**biopsy** 13–14

**cancer stages** 21–23, 36–42

**CD4+** 15, 21, 63

**CD8+** 15, 63

**CD30+** 15, 51, 53

**clinical trials** 30–31

**cutaneous T-cell lymphoma (CTCL)** 5–7, 15

**distress** 9

**Epstein-Barr virus** 16

**extracorporeal photopheresis (ECP)** 29

**fertility** 11

**folliculotropic mycosis fungoides** 35

**gene rearrangement** 11

**granulomatous slack skin** 35

**hemophagocytic lymphohistiocytosis (HLH)** 58–60

**hematopoietic cell transplant (HCT)** 31–32

**hyperpigmented mycosis fungoides** 35

**immunophenotype** 15

**immunohistochemistry (IHC)** 15

**involved-site radiation therapy (ISRT)** 28

**large cell transformation (LCT)** 43

**lymph node biopsy** 14

**lymphocytes** 5–6

**lymphomatoid papulosis (LyP)** 51, 53–54

**lymphoproliferative disorders (LPDs)** 15, 63–64

**mutations** 16–17

**mycosis fungoides (MF)** 15, 21–23, 35–42

**pagetoid reticulosis (Woringer-Kolopp disease)** 35

**phototherapy** 27

**radiation therapy (RT)** 27–28

**retinoids** 29

**Sézary syndrome (SS)** 15, 23, 46–49

**skin biopsy** 13, 57

**skin-directed therapy** 27

**skin exam** 10–11

**subcutaneous panniculitis-like T-cell lymphoma (SPTCL)** 15, 57–60

**systemic therapy** 29–30

**T cell (or T lymphocyte)** 5–6

**T-cell receptor (TCR)** 15, 17

**topical therapy** 27

**total skin electron beam therapy (TSEBT)** 28

**tumor, node, metastasis, and blood (TNMB) scores** 20–21

**visceral disease** 23, 42, 48



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PAT-N-2014-1126