



NCCN  
GUIDELINES  
FOR PATIENTS®

2024

# Cutaneous B-Cell Lymphomas



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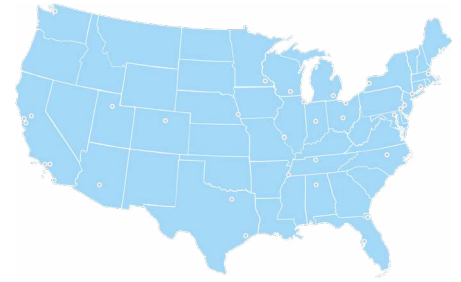
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# About the NCCN Guidelines for Patients®



Did you know that top cancer centers across the United States work together to improve cancer care? This alliance of leading cancer centers is called the National Comprehensive Cancer Network® (NCCN®).



Cancer care is always changing. NCCN develops evidence-based cancer care recommendations used by health care providers worldwide. These frequently updated recommendations are the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®). The NCCN Guidelines for Patients plainly explain these expert recommendations for people with cancer and caregivers.

**These NCCN Guidelines for Patients are based on the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®) for Primary Cutaneous Lymphomas, Version 2.2024 – May 6, 2024.**

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## Contents

- 4 CBCL basics
- 8 Testing for CBCL
- 23 Treating CBCL
- 34 PCFCL and PCMZL
- 40 Making treatment decisions
- 53 Words to know
- 55 NCCN Contributors
- 56 NCCN Cancer Centers
- 58 Index

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National Comprehensive Cancer Network (NCCN) and NCCN Foundation  
3025 Chemical Road, Suite 100, Plymouth Meeting, PA 19462 USA

# 1

## CBCL basics

- 5 Lymphatic system
- 6 Lymphocytes
- 6 Primary cutaneous lymphomas
- 7 Key points

**Cutaneous B-cell lymphoma (CBCL) starts in B cells, a type of white blood cell called a lymphocyte. These cells make antibodies to fight infections and are an important part of the lymphatic system. CBCL appears as a nodule under the skin that might look like small pimples called papules.**

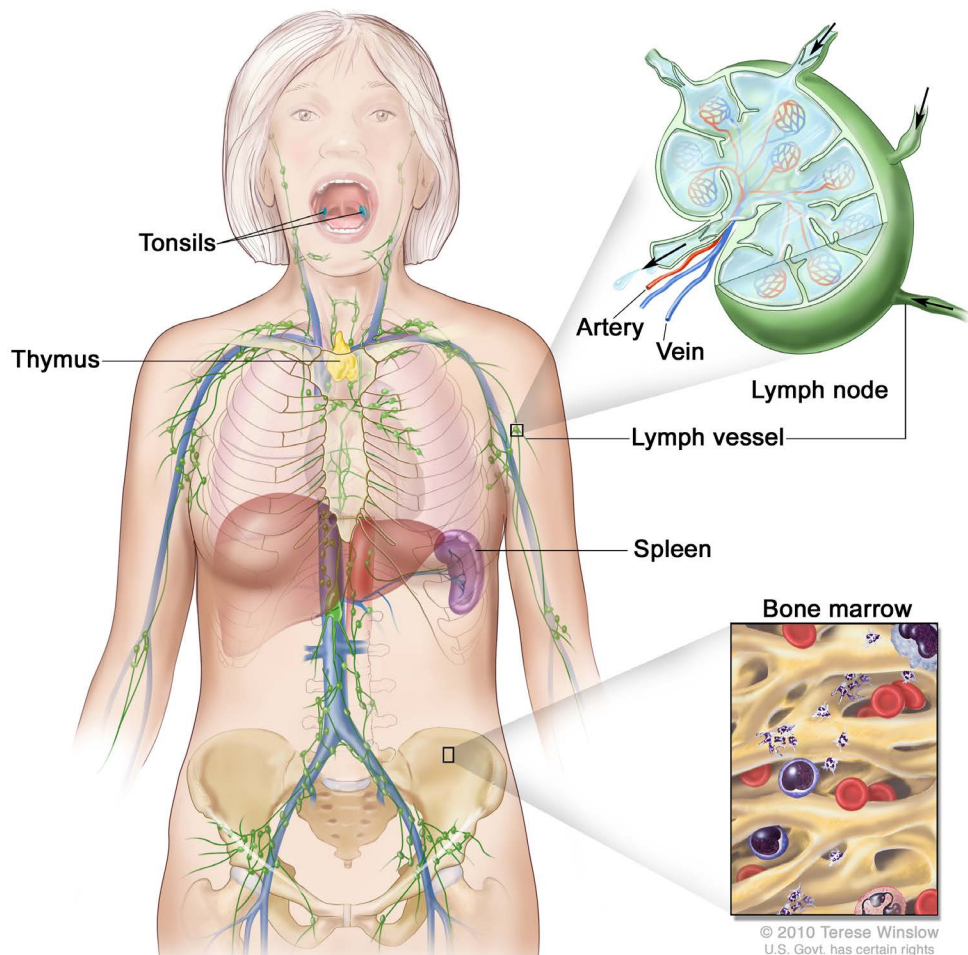
## Lymphatic system

Lymphoma is the most common type of blood cancer. It affects the lymphatic system. The lymphatic or lymph system is a major part of the body's immune system. It is a germ-fighting network of tissues and organs that includes the bone marrow, spleen, thymus, lymph nodes, and lymphatic vessels.

Lymphatic vessels are a network of thin tubes that carry lymphatic fluid (lymph) and white blood cells into all the tissues of the body. Lymph gives cells water and food. White blood cells, such as lymphocytes, help fight infection and disease.

### Lymphatic system

The lymphatic or lymph system is part of the immune system. It includes lymph vessels, lymph nodes, tonsils, thymus, spleen, and bone marrow.



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As lymph travels throughout your body, it passes through hundreds of small bean-shaped structures called lymph nodes. Lymph nodes make immune cells that help the body fight infection. They also filter the lymph fluid and remove foreign material such as bacteria and cancer cells.

## Lymphocytes

Non-Hodgkin lymphoma (NHL) is a cancer of lymphocytes. A lymphocyte is a type of white blood cell that helps fight and prevent infection. Lymphocytes are found in blood and lymph tissue, and every organ in the body. Lymph tissue includes lymph vessels and lymph nodes. Lymphocytes normally grow in response to infection or inflammation. When they grow on their own without proper regulation, they can develop into lymphoma.

There are 3 main types of lymphocytes:

- **B lymphocytes or B cells** make antibodies. An antibody is a protein that fights infection.
- **T lymphocytes or T cells** help kill tumor cells and help control immune responses.
- **Natural killer (NK) cells** have granules (small particles) with enzymes that can kill tumor cells or cells infected with a virus.

## Primary cutaneous lymphomas

Primary cutaneous lymphomas (PCL) or lymphomas of the skin are a rare group of NHLs. PCL is not a type of skin cancer. Skin cancer develops from skin cells. PCL develops from abnormal B or T lymphocytes. A lymphocyte is a type of white blood cell.

There are 2 types of PCL:

- Cutaneous T-cell lymphoma (CTCL)
- Cutaneous B-cell lymphoma (CBCL)

### Cutaneous B-cell lymphomas

Cutaneous B-cell lymphoma (CBCL) develops from abnormal B cells. B cells produce antibodies that are used to attack invading bacteria, viruses, and toxins. The antibody molecules latch onto and destroy invading viruses or bacteria.

CBCL often appears as nodules under the skin that might look like small pimples called papules.

Main types include:

- Primary cutaneous marginal zone lymphoma (PCMZL), also called PCMZ lymphoproliferative disorder
- Primary cutaneous follicle center lymphoma (PCFCL)
- Primary cutaneous diffuse large B-cell lymphoma, leg type (PC-DLBCL, leg type)

Most primary cutaneous B-cell lymphomas (PCBCLs) are indolent or slow-growing.



This book will discuss treatment options for the following CBCLs:

- PCMZL (PCMZ lymphoproliferative disorder)
- PCFCL
- Treatment for PC-DLBCL, leg type can be found in *NCCN Guidelines for Patients: Diffuse Large B-Cell Lymphomas* at [NCCN.org/patientguidelines](https://www.nccn.org/patientguidelines) and on the [NCCN Patient Guides for Cancer](#) app.

### Cutaneous T-cell lymphomas

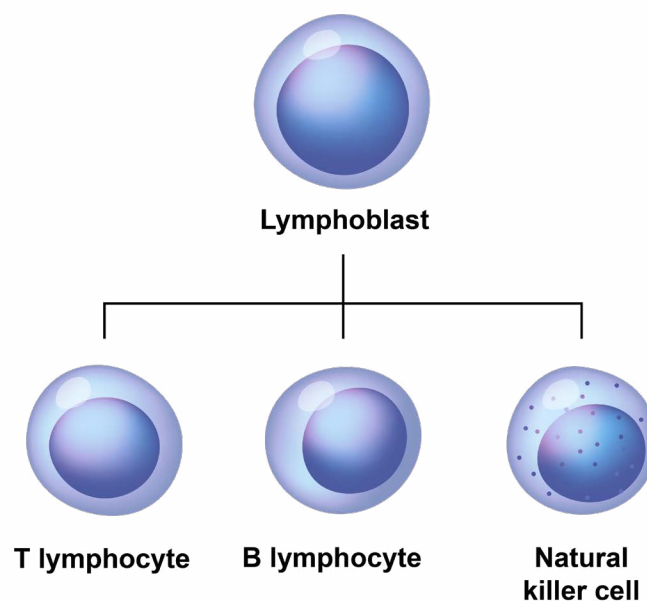
Cutaneous T-cell lymphoma (CTCL) develops from abnormal T cells. Information on types of CTCL such as mycosis fungoides, Sézary syndrome, and primary cutaneous CD30+ T-cell lymphoproliferative disorders (PCLPDs) can be found at [NCCN.org/patientguidelines](https://www.nccn.org/patientguidelines) and on the [NCCN Patient Guides for Cancer](#) app.

## Key points

- Non-Hodgkin lymphoma (NHL) is a cancer that develops from lymphocytes, a type of white blood cell. Primary cutaneous lymphoma (PCL) is an NHL found in the skin. It is not skin cancer.
- Lymphocytes normally grow in response to infection or inflammation. When they grow on their own without proper regulation, they can develop into lymphoma.
- Cutaneous B-cell lymphomas (CBCLs) are lymphomas of the skin that appear as lumps that look like small pimples called papules.

### Lymphocytes

A lymphocyte is a type of white blood cell. In primary cutaneous lymphoma (PCL), abnormal lymphocytes cause skin lesions.



# 2

## Testing for CBCL

- 9 Test results
- 10 General health tests
- 11 Skin exam
- 12 Fertility (all genders)
- 13 Blood tests
- 14 Imaging tests
- 15 Biopsy
- 16 Immunophenotyping
- 17 Testing for CBCL biomarker and genetic changes
- 20 TNM scores
- 22 Key points

**Treatment planning starts with testing. Accurate testing is needed to diagnose and treat lymphomas of the skin. This chapter presents an overview of possible tests you might receive and what to expect.**

### Test results

Accurate testing is needed to diagnose and treat cutaneous B-cell lymphoma (CBCL). Results from blood and tissue tests, imaging studies, and biopsy will determine your treatment plan. It is important you understand what these tests mean. Ask questions about your test results. Online patient portals are one way to access your test results. Please wait to discuss the results with your doctor or health care team.

Keep these things in mind:

- It's beneficial to have a support system in place during diagnosis and treatment. Enlist the help of friends, family members, or peers who can provide transportation, meals, and emotional support. These can be different people for different tasks or change over time.
- Consider bringing someone with you to doctor visits if possible, or have someone on the phone or join you for telehealth visits.
- Don't hesitate to ask questions and take notes during appointments. Write down questions and ask a friend or family

member to take notes. Caregivers should ask questions, too.

- Organize your medical documents, including insurance forms, medical records, and test results. Keep a list of contact information for your care team and update your primary care physician (PCP) regarding any changes. Include details about the specific type of cancer, treatment, and dates in your contact list.
- Set up a patient portal or health record account if it's available, which can help you track your appointments and communicate with your care team. In many places the MyChart or portal messages are not immediately seen by a nurse or physician, so ask your care team how best to communicate with them, especially in an emergency.

For possible tests and procedures, **see Guide 1.**

## 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 have had in your life. Be prepared to list any illness or injury and when it happened. Bring a list of old and new medicines and any over-the-counter (OTC) medicines, herbals, or supplements you take. Some supplements interact with and affect medicines that your care team may prescribe. Tell your care team about any symptoms you have. A medical history, sometimes called a health history, will help determine which treatment is best for you.

### Physical exam

During a physical exam, your health care provider may:

- Check your temperature, blood pressure, pulse, and breathing rate
- Check your height and weight
- Listen to your lungs and heart
- Look in your eyes, ears, nose, and throat
- Feel and apply pressure to parts of your body to see if organs are of normal size, are soft or hard, or cause pain when touched
- Feel for enlarged lymph nodes in your neck, underarm, and groin
- Conduct a complete skin exam

### Guide 1

#### Tests to plan treatment

Skin biopsy with pathology review and tumor/lesion testing

Medical history and physical exam, including complete skin exam

Complete blood count (CBC) with differential, comprehensive metabolic panel (CMP), and lactate dehydrogenase (LDH)

CT with contrast of chest, abdomen, and pelvis (C/A/P) and/or PET/CT scan

Pregnancy test for those of childbearing age

Possible:

- Bone marrow biopsy
- Serum protein electrophoresis (SPEP) and other blood tests such as flow cytometry
- HIV testing
- Hepatitis B and hepatitis C testing
- Discussion of fertility preservation

## Skin exam

A dermatologist is an expert in the skin and diseases of the skin. It is important to find a dermatologist experienced in lymphomas of the skin to conduct a skin exam. Cutaneous B-cell lymphoma (CBCL) might appear as a rash, lumps, bumps, or tumor. A rash is an area of irritated or swollen skin. Many rashes are itchy, red, and painful. As a rash, CBCL might come and go. This doesn't mean the cancer is cured.

The amount of cancer 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 the differences in where the skin looks normal and 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

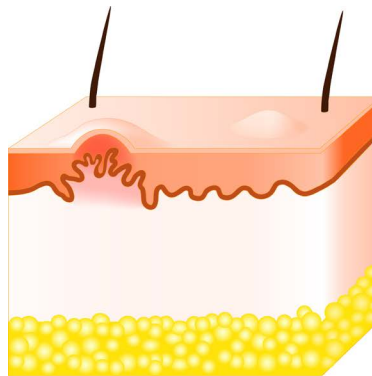
Skin is the largest organ in your body. A skin lesion is a change in color or texture in an area of the skin. Skin lesions can appear anywhere on the body, but are most common on the lower abdomen, upper thighs, buttocks, and breasts. Some words to describe skin lesions include patch, papule, plaque,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

### Patch

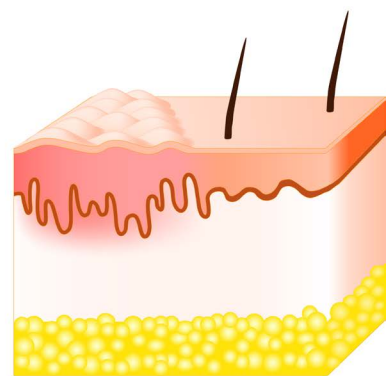
A patch is a flat, thin, pink or red lesion of any size that forms on the skin. Patches may be dry, scaly, itchy, and may look like eczema or psoriasis. They can be lighter or darker than surrounding skin. The patches may sometimes become plaques (raised or hardened lesions) on the skin.

### 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.**



Papule



Plaque

### 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, and itchy, and may look like eczema or psoriasis. Plaques sometimes become tumors on the skin.

### Papulonodular

Papulonodular is a combination of papules and nodules found on the skin. Nodules are raised higher on the skin than papules.

### Tumor

A tumor is a firm, dome-shaped mass at least 1 centimeter in size.

### Ulcer

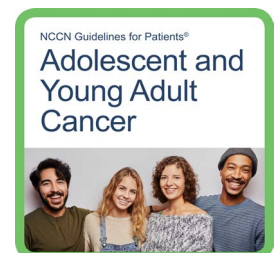
A skin ulcer is an open sore or wound on the skin.

## Fertility (all genders)

Some types of treatment can affect your fertility, the ability to have children. If you think you want children in the future, ask your care team how cancer and cancer treatment might change your fertility. To preserve your fertility, you may need to take action before starting cancer treatment. Those who want to have children in the future should be referred to a fertility specialist to discuss the options before starting treatment.

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

More information on fertility preservation in adolescents and young adults is available at [NCCN.org/patientguidelines](https://www.nccn.org/patientguidelines) and on the [NCCN Patient Guides for Cancer](#) app.



### Changes in fertility

Treatment might cause your fertility to be temporarily or permanently impaired or interrupted. This loss of fertility is related to your age at time of diagnosis, treatment type(s), treatment dose, and treatment length. Talk to your care team about your concerns and if you are planning a pregnancy.

# 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 a vein in your arm.

## Complete blood count and differential

A complete blood count (CBC) measures the levels of red blood cells (RBCs), white blood cells (WBCs), and platelets (PLTs) in your blood. A CBC is a key test that gives a picture of your overall health. A differential counts the number of each type of WBC (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 (CMP) measures substances in your blood. A CMP provides important information about how well your kidneys and liver are working, among other things.

## Hepatitis B and hepatitis C

Hepatitis B and hepatitis C are types of liver disease caused by a virus. A hepatitis blood test will show if you had hepatitis in the past or if you have it today. Some cancer treatments can wake up (or reactivate) the virus. If this happens, it can cause harm to the liver.

## HIV

Human immunodeficiency virus (HIV) causes acquired immunodeficiency syndrome (AIDS). An HIV antibody test checks for HIV antibodies in a sample of blood, urine, or saliva. It's important to let your doctor know if you have ever been infected with HIV.

## HTLV

Human T-lymphotropic virus (HTLV) testing is used to detect an HTLV infection that could be the cause of a T-cell lymphoma. In the United States, all donated blood is screened for HTLV.

## Lactate dehydrogenase

Lactate dehydrogenase (LDH) or lactic acid dehydrogenase is a protein found in most cells. Dying cells release LDH into the blood. Fast-growing cells also release LDH and cause levels of this protein to be elevated in the blood.

## Pregnancy test

If planned treatment might affect pregnancy, then those who can become pregnant will be given a pregnancy test before treatment begins.

## SPEP

Serum protein electrophoresis (SPEP) examines specific proteins in the blood called globulins, which may be increased in certain conditions such as multiple myeloma.



# 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 doctor. While these reports might be available to you through your patient portal or patient access system, please wait to discuss these results with your care team.

## Contrast material

Contrast material is used to improve the pictures of the inside of the body. Contrast materials are substances that help enhance and improve the images of several organs and structures in the body. It is used to make the pictures clearer. The contrast is not permanent and will leave your body in your urine immediately after the test. The types of contrast vary and are different for CT and MRI.

Tell your care team if you have had allergic reactions to contrast in the past. This is important. You might be given medicines to avoid the effects of those allergies. Contrast might not be used if you have a serious allergy or if your kidneys aren't working well.

## CT scan

A CT or CAT (computed tomography) 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 one detailed picture. A CT scan of your head, neck, chest, abdomen, and pelvis may be one of the tests to look for cancer. In most cases, contrast will be used.

## MRI scan

An MRI (magnetic resonance imaging) scan uses radio waves and powerful magnets to take pictures of the inside of the body. It does not use x-rays. Because of the very strong magnets used in the MRI machine, tell the technologist if you have any metal in your body. During the test, you will likely be asked to hold your breath for 10 to 20 seconds as the technician collects the images. Contrast is often used.

A closed MRI has a capsule-like design where the magnet surrounds you. An open MRI has a magnetic top and bottom, which allows for an opening on each end. Closed MRIs are more common than open MRIs, so if you have claustrophobia (a dread or fear of enclosed spaces), be sure to talk to your care team about it.

## PET scan

A PET (positron emission tomography) scan uses a radioactive drug called a tracer. A tracer is a substance injected into a vein to see where cancer cells are in the body and if they are using sugar produced by your body to grow. Cancer cells show up as bright spots on PET scans. However, not all tumors will appear on a PET scan. Also, not all bright spots are cancer. It is 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 is called a PET/CT scan.

## X-ray

An x-ray is a type of radiation. In small doses, it is used to make pictures of the inside of the body. It might be referred to as a radiograph.



# Biopsy

A biopsy is the removal of a sample of tissue such as skin, lymph node, and/or bone marrow. It is an important part of an accurate diagnosis of lymphoma. Your sample should be reviewed by a pathologist who is an expert in the diagnosis of CBCL. The pathologist will note the overall appearance and the size, shape, and type of your cells. This review is often referred to as histology, histopathology, or hematopathology review. Tests will be done on the biopsied cells. Ask questions about your biopsy results and what they mean for your treatment.

### Skin lesion biopsy

A sample of your lesion will be removed and tested to confirm the type of CBCL. A skin lesion biopsy can be incisional or excisional. An incisional biopsy removes an area of skin using a scalpel blade. Stitches are usually required after an incisional biopsy. An excisional biopsy removes a larger area of skin, tumor, or lesion.

### Skin punch biopsy

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

### Skin shave biopsy

A skin shave biopsy removes the top layer of skin using a tool like a razor. This type of biopsy may not be recommended because it doesn't take a deep enough sample. Abnormal lymphocytes are often found under the surface of the skin.

### Lymph node biopsy

Lymph nodes are usually too small to be seen or felt. Sometimes, lymph nodes can feel swollen, enlarged, hard to the touch, or don't move when pushed (fixed or immobile). A lymph node biopsy can be 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:

- Bone marrow aspirate
- Bone marrow biopsy

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

For many, this is an uncomfortable procedure. Your care team will try to make you as comfortable as possible. The samples are usually taken from the back of the hip bone (pelvis). You will likely lie on your belly or side. For an aspirate, a hollow needle will be pushed through your skin and into the bone. Liquid bone marrow will then be drawn into a syringe. For the biopsy, a wider needle will be used to remove a small piece of the bone. You may feel bone pain in your hip for a few days. Your skin may bruise.

### Immunophenotyping

Immunophenotyping is a process that uses antibodies to detect the presence or absence of certain antigens. Antigens are proteins or markers that can be found on the surface of or inside all cells, including white blood cells. Specific groupings of antigens are normal. However, some specific patterns of antigens called the immunophenotype are found on abnormal cells including non-Hodgkin lymphoma (NHL) and CBCL.

Immunophenotyping can be done using specialized techniques called flow cytometry or immunohistochemistry (IHC). These techniques are used to distinguish CBCL from other types of lymphoma. Immunophenotype can change as cancer progresses.

#### Flow cytometry

Flow cytometry (FCM) is a laboratory method used to detect, identify, and count specific cells. Flow cytometry 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, things like the size and shape of the cells, and other unique features of the cells. Flow cytometry may be used on cells from circulating (peripheral) blood, bone marrow, or a biopsy. The most common use of flow cytometry is in the identification of markers on cells, particularly in the immune system (called immunophenotyping).



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

#### Immunohistochemistry

Immunohistochemistry (IHC) is a special staining process that involves adding a chemical marker to immune cells. The cells are then studied using a microscope. IHC looks for the immunophenotype of cells from a skin biopsy. IHC panel of skin biopsy can include testing for CD3, CD4, CD8, CD10, CD20, CD30, CD56, ALK, BCL2, BCL6, IRF4/MUM1, and others.

## Testing for CBCL biomarker and genetic changes

Biomarker and genetic tests are used to learn more about your type of CBCL, to guide treatment, and to determine the likely path your cancer will take (prognosis). This genetic testing is different from family history genetic testing or genetic cancer risk testing. This testing looks for changes only in the CBCL cells that have developed over time, and not 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 chromosomes. Each chromosome contains thousands of genes. Genes are coded instructions for the proteins your cells make. A mutation is when something goes wrong in the genetic code.

Proteins are written like this: BCL6. Genes are written with italics like this: *BCL6*. When a gene or protein is found (expressed), it is shown with a plus sign (+) like this: CD30+. When a gene or protein has not been found, it is written with a negative sign (-) like this CD30-.

### CBCL mutation testing

A sample of your skin, lymph node, blood, or bone marrow will be used to see if the CBCL cancer cells have any specific mutations. Some mutations can be targeted with specific therapies. This is separate from the genetic testing for mutations that you may have inherited from your biological parents.



**Testing of your lymphoma cells can gather specific information about your lymphoma to help guide treatment.**

CBCL cells can have changes in genes and chromosomes. Mutation testing using methods such as high-throughput sequencing (HTS), comparative genomic hybridization (CGH), next-generation sequencing (NGS), karyotype, fluorescence in situ hybridization (FISH), and polymerase chain reaction (PCR) are used to look for these changes or abnormalities. Some mutations may determine the type of treatment given. Subtle new drug-resistant mutations may occur over time. Mutations can also happen during treatment. Mutation testing is used to look for these new mutations. Some mutations lead to resistance to certain targeted therapies. There are many possible mutations.

### Comparative genomic hybridization

Comparative genomic hybridization (CGH) is a technique that compares DNA samples from normal tissue and tumor tissue. It is used to detect abnormal chromosomes.

### 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, lymph node, or blood sample.

FISH can find translocations that are too small to be seen with other methods. A translocation occurs when parts of two chromosomes switch with one another. However, FISH can only be used for known changes. It cannot detect all the possible changes found within the chromosomes or genes.

### Gene rearrangements

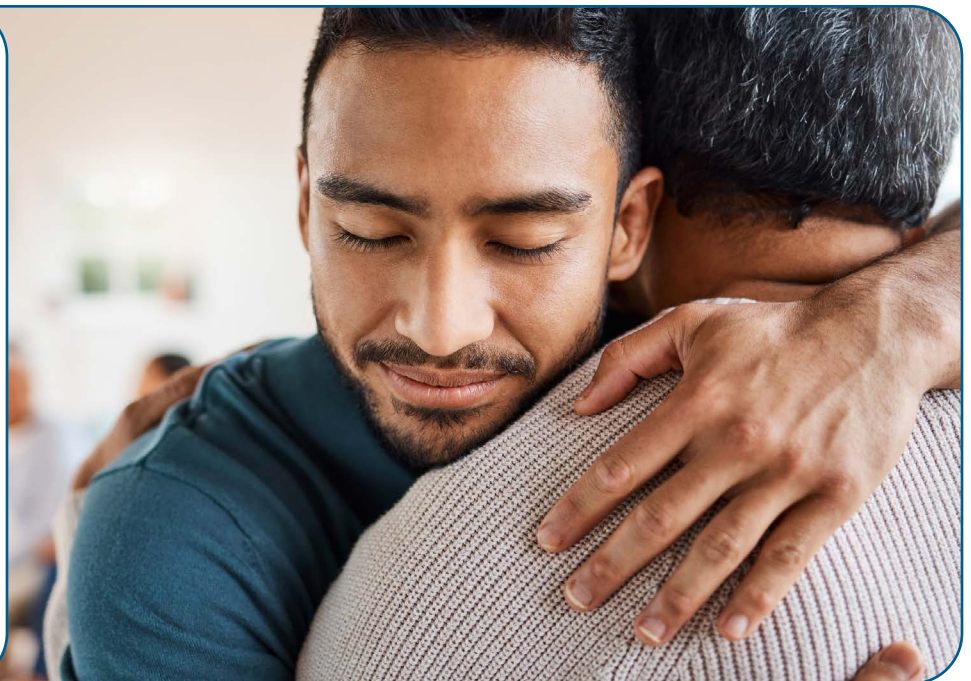
Normal B cells and T cells break their DNA in certain ways to create diversity within your immune system. In a tumor, all cancer cells derive from the same original cell. In CBCL that cell is a B cell. When that one B cell divides many times, the entire group of B cells is called clonal or the tumor is described as having clonality. In clonal cells, the same gene rearrangements are found in a group of cancer cells. Pathologists have tests they can use to determine if a group of cells is clonal or not.

Biomarker and genetic testing is used to detect B-cell rearrangements commonly found in CBCL. This information can be helpful when diagnosing and treating CBCL.

### High-throughput sequencing

High-throughput sequencing (HTS) is capable of sequencing hundreds of millions of DNA molecules at a time.

**Seek out support groups at your local hospital, through social media, or through resources listed in the back of this book. Look to friends, relatives, neighbors, and coworkers for social support.**



## Karyotype

A karyotype is a picture of chromosomes. Normal human cells contain 23 pairs of chromosomes for a total of 46 chromosomes. A karyotype will show extra, missing (deletion), rearranged, or abnormal pieces of chromosomes within the lymphoma cells.

## Next-generation sequencing

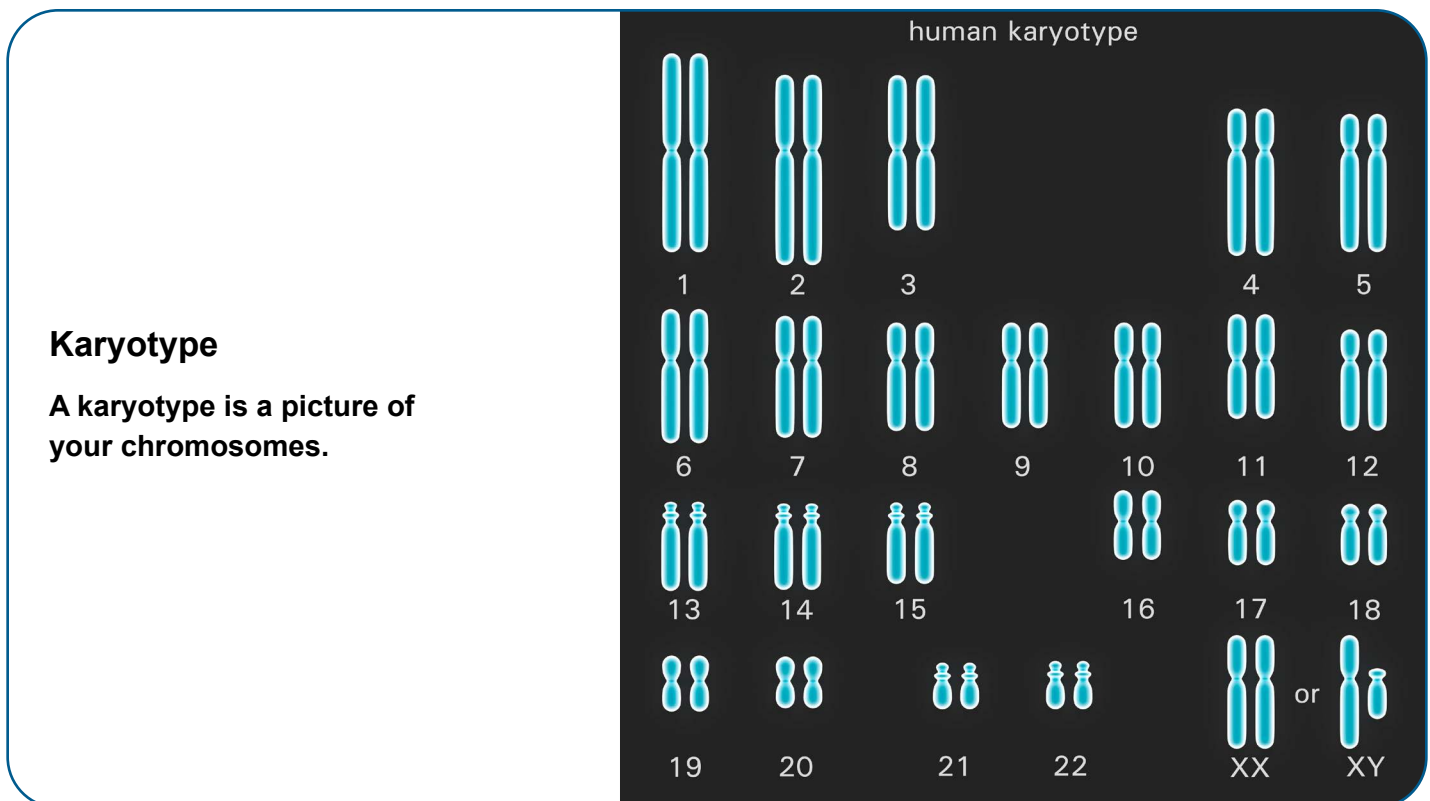
Next-generation sequencing (NGS) is a method used to determine a portion of a person's DNA sequence. It shows if a gene has any mutations that might affect how the gene works. NGS looks at the gene in a more detailed way than other methods and can find mutations that other methods might miss.

## PCR

A polymerase chain reaction (PCR) is a lab process that can make millions or billions of copies of your DNA or RNA (genetic information). PCR is very sensitive. It can find 1 abnormal cell among more than 100,000 normal cells. These copies, called PCR product, might be used for next-generation sequencing (NGS). This is important when testing for treatment response or remission.

## Translocations

Translocation is a switching of parts between two chromosomes. A translocation between chromosome 14 and 18 is written as t(14;18). This translocation is rare in CBCL. Specific translocations such as t(14;18) can help distinguish between types of blood cancers and disorders.





## TNM scores

The tumor, node, metastasis (TNM) system is used to stage many cancers such as CBCL. In this system, the letters T, N, and M describe different areas of cancer growth. Based on biopsy and other test results, your doctor will assign a score or number to each letter. The higher the number, the larger the tumor or the more the cancer has spread to lymph nodes or other organs. A TNM example might look like this: T1N0M0 or T1, N0, M0.

- **T is for skin** – Tumor refers to size, type, and number of tumors or lesions covering the skin.
- **N is for lymph node** – Cancer can spread to lymph nodes.
- **M is for metastasis** – Cancer can spread to distant parts of the body.

Cancer staging is often done twice.

- **Clinical stage (c)** is the rating given before any treatment. It is based on a physical exam, biopsy, and other tests.
- **Pathologic stage (p)** or surgical stage is determined by examining tissue removed during surgery such as in the removal of a lymph node. Pathologic features include size, shape, and type of cell.

### T = Skin

Lesions or tumors will be measured by their depth, height, size, and region of the body. Lesions are often measured in centimeters (cm). Body regions are based on regional lymph node drainage patterns. Body regions include head/neck, chest, upper arm, lower arm and hand, abdomen and genitals, upper leg, lower leg and feet, upper back, lower back, and buttocks.

- **T1** – Solitary (one) skin lesion
  - **T1a** is one lesion smaller than 5 centimeters (cm). For comparison, a golf ball is about 4 cm.
  - **T1b** is one lesion 5 cm or larger.
- **T2** – Regional skin involvement is multiple lesions limited to 1 body region or 2 body regions next to one another (contiguous).
  - **T2a** is disease limited to an area smaller than a 15-cm wide circle.
  - **T2b** is disease limited to an area the size of a 15-cm to 30-cm wide circle.
  - **T2c** is disease found in a 30-cm or larger area.
- **T3** – Generalized skin involvement
  - **T3a** is multiple lesions involving 2 body regions not next to one another.
  - **T3b** is multiple lesions involving 3 or more body regions.

## N = Node

There are hundreds of lymph nodes throughout your body. Lymph nodes work as filters to help fight infection and remove harmful things. Lymph node regions are based on the Ann Arbor Staging System. Peripheral sites include antecubital, cervical, supraclavicular, axillary, inguinal-femoral, and popliteal. Central sites include mediastinal, pulmonary hilar, para-aortic, and iliac.

- **N0** means no cancer found in lymph nodes.
- **N1** means involvement of 1 peripheral lymph node region that drains an area of current or prior skin involvement. Lymph node biopsy tests positive for lymphoma.
- **N2** means involvement of 2 or more peripheral lymph node regions or involvement of any lymph node region that does not drain an area of current or prior skin involvement. Lymph node biopsy tests positive for lymphoma.
- **N3** means cancer is found in central lymph nodes. Lymph node biopsy tests positive for lymphoma.

## M = Metastasis

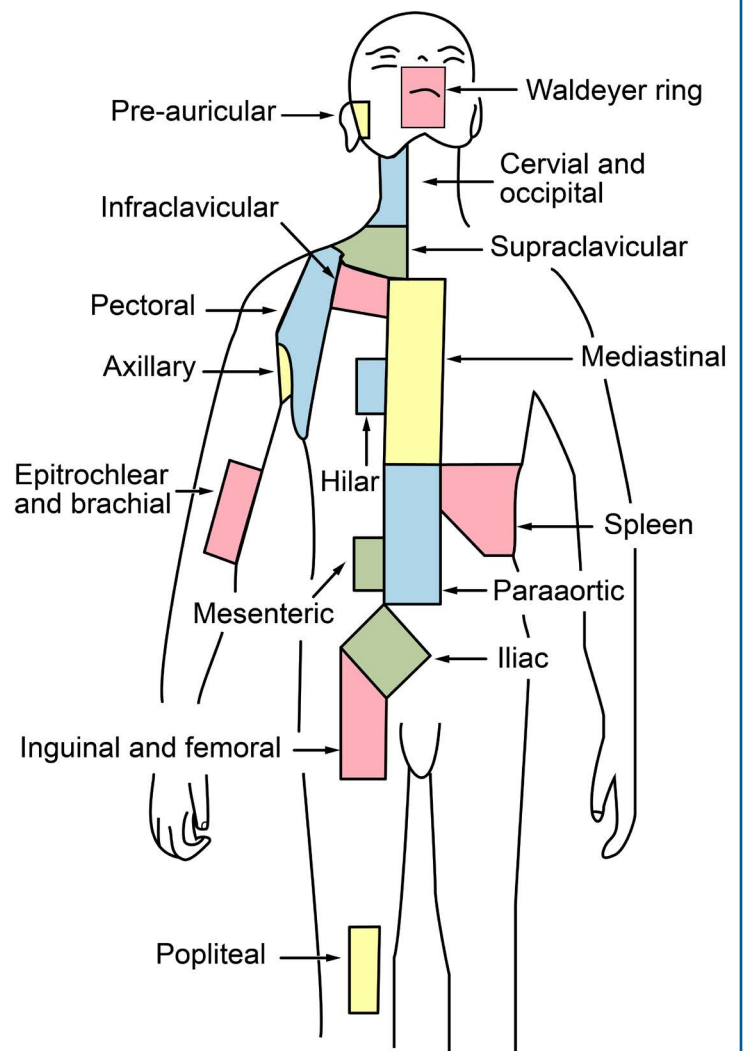
Cancer that has spread to distant parts of the body is called metastatic. Cancer can metastasize to internal (visceral) organs like the liver or spleen.

- **M0** means no cancer is found in visceral organs.
- **M1** means cancer is found in visceral organs.

## Lymph node regions

### Lymph node regions based on the Ann Arbor Staging System.

[https://commons.wikimedia.org/wiki/File:Lymph\\_node\\_regions.svg](https://commons.wikimedia.org/wiki/File:Lymph_node_regions.svg)



### Key points

- Tests check for signs of disease, determine how well organs are working, and assess treatment results.
- Skin lesions can appear anywhere on the body. Lesions may look like papules, patches, plaques, or nodules.
- A skin biopsy is needed to diagnose cutaneous B-cell lymphoma (CBCL). Your sample should be reviewed by a pathologist who is an expert in the diagnosis of lymphomas of the skin.
- Immunophenotyping is used to distinguish CBCL from other types of lymphoma.
- A sample from your biopsy may undergo lab tests to learn more about your CBCL and choose the best treatment for you.
- Biomarker testing includes tests of genes or their products (proteins). It identifies the presence or absence of mutations and certain proteins that might suggest treatment.
- The tumor, node, metastasis (TNM) system might be used to describe your cancer.



### **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](https://www.nccn.org/patients/response)



# 3

## Treating CBCL

- 24 Care team
- 26 Preventing pregnancy during treatment
- 27 Surgery
- 27 Skin-directed therapy
- 28 Radiation therapy
- 29 Systemic therapy
- 30 Clinical trials
- 32 General supportive care
- 33 Key points

**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 is best for you.**

## Care team

Cutaneous B-cell lymphomas (CBCL) are treatable and may be curable in certain cases.

Treating cancer takes a team approach. Treatment decisions should involve a multidisciplinary team (MDT). An MDT is a team of health care and psychosocial care professionals from different professional backgrounds who have knowledge (expertise) and experience in your type of cancer. This team is united in the planning and implementing of your treatment. Ask who will coordinate your care.

Some members of your care team will be with you throughout cancer treatment, while others will only be there for parts of it. Get to know your care team and help them get to know you.

Depending on your diagnosis, your team might include the following specialists:

- **A dermatologist** specializes in the diagnosis and treatment of skin diseases.
- **A hematologist or hematologic oncologist** is a medical expert in blood diseases and blood cancers. Other types of oncologists include medical, radiation, and surgical oncologists.
- **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 (RT).
- **Oncology nurses** provide your hands-on care, like giving systemic therapy, managing your care, answering questions, and helping you cope with side effects.
- **An advanced practice nurse (APN) or a physician assistant (PA)** help provide an extra layer of support with your cancer-related symptoms.
- **Oncology pharmacists** are experts in knowing how to use medicines to treat cancer and to manage symptoms and side effects.
- **Palliative care specialists** concentrate on preventing and alleviating suffering and improving quality of life.
- **An occupational therapist** helps people with the tasks of daily living.
- **A physical therapist** helps people move with greater comfort and ease.

- **Psychologists and psychiatrists** are mental health experts who can help manage issues such as depression, anxiety, or other mental health conditions that can affect how you think and feel.
- **Social workers** help people solve and cope with problems in their everyday lives. Clinical social workers also diagnose and treat mental, behavioral, and emotional issues. The anxiety a person feels when diagnosed with cancer might be managed by a social worker in some cancer centers. They, or other designated professionals, can help navigate the complexities of financial and insurance stresses.
- **Spiritual care specialists** identify and support those with spiritual distress or unmet spiritual needs.
- **A research team** helps to collect research data and coordinate care if you are in a clinical trial. Clinical trials help bring new therapies to patients and advance the treatment for everyone. Consider asking your care team about access to clinical trials.



## You know your body better than anyone

Help your care team understand:

- ✓ How you feel
- ✓ What you need
- ✓ What is working and what is not

Keep a list of names and contact information for each member of your team. This will make it easier for you and anyone involved in your care to know whom to contact with questions or concerns.

**Get to know your care team and help them get to know you.**

## Preventing pregnancy during treatment

Cancer and cancer treatment can affect the ovaries and damage sperm. If you become pregnant during chemotherapy, radiation therapy, or other types of systemic therapy, serious birth defects can occur. Speak with your care team about preventing pregnancy while being treated for cancer. Hormonal birth control may or may not be recommended, so ask your doctor about options such as intrauterine devices (IUDs) and barrier methods. Types of barrier methods include condoms, diaphragms, cervical caps, and the contraceptive sponge.

### Those with ovaries

Those who can become pregnant will have a pregnancy test before starting treatment. Cancer treatment can hurt the developing baby if you are or become pregnant during treatment. Therefore, birth control to prevent pregnancy

during and after treatment is recommended. If you are pregnant or breastfeeding at the time of your cancer diagnosis, certain treatments will need to be avoided.

Menstruation, menses, menstrual flow, or your period may stop during treatment, but often returns within 2 years after treatment in those 35 years of age and under. It is still possible to become pregnant even though you might not have a period. Therefore, birth control is recommended during and after treatment. Consult your doctor for the best time to plan a pregnancy.

### Those with testicles

Cancer and cancer treatment can damage sperm. Therefore, use contraception (birth control) such as condoms to prevent pregnancy during and immediately after cancer 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 regimen that is considered standard of care. Ask your care team what treatment options are available and if a clinical trial might be right for you.**



## Surgery

Surgery is an operation or procedure to remove a lesion or tumor from the body. The type of surgery depends on size, location, and number of tumors. It is also called excision or excisional surgery. The usual role for surgery in CBCL is to biopsy suspicious lesions, not to completely remove (excise) the lymphoma.

## Skin-directed therapy

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

- **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 used to treat certain types of flat, scaly growths on the skin. Brand names include Aldara and Zyclara.
- **Nitrogen mustard (mechlorethamine hydrochloride)** stops or slows the growth of cancer. Brand names include Valchlor and Ledaga.
- **Retinoids** are products related to vitamin A. Examples include bexarotene

(Targretin gel) and tazarotene (Tazorac Gel, Tazorac Cream).

### Local radiation therapy

Local radiation therapy (RT) treats the skin lesion. Involved-site radiation therapy (ISRT) is a type of local radiation that might be used to treat skin lesions.

### Phototherapy

Phototherapy uses different ultraviolet (UV) light wavelengths to treat skin lesions or tumors. Phototherapy prescribed by your treatment team uses carefully selected wavelengths to target the skin lymphoma. It is very different from the radiation given in tanning beds. Using tanning beds increases your risk of other skin cancers and is not recommended.

Types include:

- **Ultraviolet B (UVB)** – exposes the skin to an artificial UVB light source for a set length of time on a regular schedule.
- **Narrowband ultraviolet B (NB-UVB)** – uses a very specific UV wavelength.
- **Photochemotherapy or psoralen plus ultraviolet A (PUVA)** – combines psoralen (P) 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.
- **Ultraviolet A1 (UVA1)** - penetrates deep into the skin causing T cells to die.

UV can increase your risk of some skin cancers. Phototherapy may not be favored

in those with a history of skin cancer, or who have had melanoma.

### Radiation therapy

Radiation therapy (RT) uses high-energy radiation from x-rays, photons, electrons, and other sources to kill cancer cells and shrink tumors. RT can be given alone or with other treatments. Treatment may focus on individual tumors, a small area/region of the body, the entire surface of the skin, or specific lymph nodes. RT may be used as supportive care or palliative care to help ease pain or discomfort caused by cancer.

#### EBRT

External beam radiation therapy (EBRT) uses a machine outside of the body to aim radiation at the tumor(s) or areas of the body.

The most common type of EBRT that may be used to treat CBCL is:

- ▶ **Involved-site radiation therapy (ISRT)** targets a specific area of skin. It can also be used to treat specific lymph nodes with cancer.

Less common types of EBRT that may be used to treat CBCL include:

- ▶ **Three-dimensional conformal radiation therapy (3D-CRT)** uses computer software and CT images to aim beams that match the shape of the tumor.
- ▶ **Intensity-modulated radiation therapy (IMRT)** uses small beams of different strengths to match the shape of the tumor.
- ▶ **Stereotactic body radiation therapy (SBRT)** uses high-energy radiation beams to treat cancers in five or fewer treatments.
- ▶ **Stereotactic radiosurgery (SRS)** uses special equipment to position the body and give one precise, large dose of radiation.
- ▶ **Particle beam RT** uses protons, carbon ions, or other heavy ions to treat cancer.



## Systemic therapy

Systemic therapy works throughout the body. It includes retinoids, chemotherapy, targeted therapy, and immunotherapy. Systemic therapy might be used alone or with other therapies.

### Chemotherapy

Chemotherapy kills fast-growing cells throughout the body, including cancer cells and normal cells. Methotrexate is an example of a chemotherapy. When more than one chemotherapy is used, it is called multiagent.

### Targeted therapy

Targeted therapy focuses on specific or unique features of cancer cells. Targeted therapies 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.

### Immunotherapy

Immunotherapy is a drug therapy that increases the activity of your immune system. By doing so, it improves your body's ability to find and destroy cancer cells. Immunotherapy can be given alone or with other types of treatment. Rituximab (Rituxan) is an example of an immunotherapy.

### 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.



### Warnings about supplements and drug interactions

You might be asked to stop taking or avoid certain herbal supplements when on a systemic therapy. Some supplements can affect the ability of a drug to do its job. This is called a drug interaction.

**It is critical to speak with your care team about any supplements you may be taking. Some examples include:**

- ▶ Turmeric
- ▶ Ginkgo biloba
- ▶ Green tea extract
- ▶ St. John's Wort
- ▶ Antioxidants

Certain medicines can also affect the ability of a drug to do its job. Antacids, heart or blood pressure medicine, and antidepressants are just some of the medicines that might interact with a systemic therapy or supportive care medicines given during systemic therapy. Therefore, it is very important to tell your care team about any medicines, vitamins, over-the-counter (OTC) drugs, herbals, or supplements you are taking.

**Bring a list with you to every visit.**

## Clinical trials

A clinical trial is a type of medical research study. After being developed and tested in a laboratory, 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. Food and Drug Administration (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. Treatment trials are done in phases.

- **Phase 1** trials study the dose, safety, and side effects of an investigational drug or treatment approach. They also look for early signs that the drug or approach is helpful.
- **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 long-term safety and benefit of an FDA-approved treatment.

### Who can enroll?

Every clinical trial has rules for joining, called eligibility criteria. The rules may be about age, cancer type and stage, treatment history, lab tests, or general health. These requirements ensure that participants are alike in specific ways and that the trial is as safe as possible for the participants.

### Informed consent

Clinical trials are managed by a group of experts called a research team. The research team 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 with family, friends, or others whom you trust. Keep in mind that you can leave and seek treatment outside of the clinical trial at any time.

### Start the conversation

Don't wait for your doctor to bring up clinical trials. Start the conversation and learn about all of your treatment options. If you find a study that you may be eligible for, ask your treatment team if you meet the requirements. If you have already started standard treatment you may not be eligible for certain clinical trials. Try not to be discouraged if you cannot join. New clinical trials are always becoming available.



### Frequently asked questions

There are many myths and misconceptions surrounding clinical trials. The possible benefits and risks are not well understood by many with cancer.

#### Will I get a placebo?

Placebos (inactive versions of real medicines) are almost never used alone in cancer clinical trials. It is common to receive either a placebo with a standard treatment, or a new drug with a standard treatment. You will be informed, verbally and in writing, if a placebo is part of a clinical trial before you enroll.

#### Do I have to pay to be in a clinical trial?

It depends on the study, your health insurance, and the state in which you live. In general, procedures, drugs, or tests that are considered standard of care will be billed to you or your insurance, whereas those considered research are covered by the trial sponsor. Your treatment team and the research team can help determine if you are responsible for any costs.



### Finding a clinical trial

#### In the United States

NCCN Cancer Centers  
[NCCN.org/cancercenters](https://www.nccn.org/cancercenters)

The National Cancer Institute (NCI)  
[cancer.gov/about-cancer/treatment/clinical-trials/search](https://www.cancer.gov/about-cancer/treatment/clinical-trials/search)

#### Worldwide

The U.S. National Library of Medicine  
(NLM)  
[clinicaltrials.gov](https://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](https://www.cancer.gov/contact)

### General supportive care

Supportive care will be specific to your needs. Supportive care is health care given to prevent, reduce, and relieve suffering, and to improve quality of life. Supportive care might include pain relief, palliative care, emotional or spiritual support, financial aid, or family counseling. Tell your care team how you are feeling and about any side effects so they can be managed. Supportive care, best supportive care, and palliative care often mean the same thing.

It is very important to take care of yourself by eating well, drinking plenty of fluids, exercising, and doing things that make you feel energized.

#### Side effects

All cancer treatments can cause unwanted health issues called side effects. Side effects depend on many factors. These factors include the drug type and dose, length of treatment, and the person. Some side effects may be harmful to your health. Others may just be unpleasant. Treatment can cause several side effects. Some are very serious.

Ask for a complete list of side effects of your treatments. Also, tell your treatment team about any new or worsening symptoms. There may be ways to help you feel better. There are also ways to prevent some side effects. You will be monitored closely for side effects.

#### Late effects

Late effects are side effects that occur months or years after a disease is diagnosed or after treatment has ended. Late effects may be caused by cancer or cancer treatment. They may include physical, mental, and social health

**All cancer treatments can cause unwanted health issues called side effects. It is important to tell your care team about all of your side effects so they can be managed.**

issues, and second cancers. The sooner late effects are treated the better. Ask your care team about what late effects could occur. This will help you know what to look for.

#### Survivorship

A person is a cancer survivor from the time of diagnosis until the end of life. After treatment, your health will be monitored for side effects of treatment and the return of cancer. This is part of your survivorship care plan. It is important to keep any follow-up doctor visits and imaging test appointments. Seek good routine medical care, including regular doctor visits for preventive care and cancer screening.

A personalized survivorship care plan will contain a summary of possible long-term effects of treatment called late effects and list follow-up tests. Find out how your primary care provider will coordinate with specialists for your follow-up care.

## Key points

- ▶ Cutaneous B-cell lymphomas (CBCLs) are treatable and may be curable in certain cases.
- ▶ Skin-directed therapy focuses on the skin and includes topical therapy, local radiation, and phototherapy.
- ▶ Systemic therapy works throughout the body. It includes chemotherapy, targeted therapy, immunotherapy, and retinoids.
- ▶ Radiation therapy (RT) uses high-energy radiation from x-rays, electrons, photons, and other sources to kill cancer cells and shrink tumors.
- ▶ Clinical trials study how safe and helpful tests and cancer treatments are for people.
- ▶ Supportive care is health care that relieves symptoms caused by cancer or its treatment and improves quality of life.
- ▶ All cancer treatments can cause unwanted health issues called side effects. You will be monitored for side effects, infection, and other treatment-related issues.

**Those with lymphomas of the skin should be treated at centers experienced in this type of cancer.**

# 4

## PCFCL and PCMZL

- 35 Overview
- 36 Diagnosis
- 36 Treatment
- 39 Key points

**There is more than one type of cutaneous B-cell lymphoma (CBCL). This chapter discusses treatment options for those with primary cutaneous follicle center lymphoma (PCFCL) and primary cutaneous marginal zone lymphoma (PCMZL).**

## Overview

In primary cutaneous B-cell lymphoma (PCBCL or CBCL) abnormal B cells (B lymphocytes) attack the skin causing nodules or lesions. Most PCBCLs are slow-growing (indolent).

Types include:

- Primary cutaneous follicle center lymphoma (PCFCL)
- Primary cutaneous marginal zone lymphoma (PCMZL), also called PCMZ lymphoproliferative disorder
- Primary cutaneous diffuse large B-cell lymphoma, leg type (PC-DLBCL, leg type)

### PCFCL

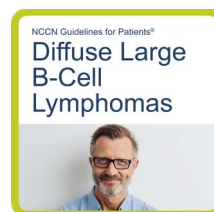
PCFCL (or CFCL) is generally slow-growing and most often found in the scalp and forehead. PCFCL develops slowly over months or years and may look like a single tumor or several nodules grouped together. Tumors are pink or reddish and slightly raised and smooth. Usually, PCFCL is BCL6-positive, CD20-positive, IRF4/MUM1-negative, CD3-negative, and FOXP1-negative. Treatment for PCFCL will be discussed in this chapter.

### PCMZL

PCMZL (or CMZL) is also called PCMZ lymphoproliferative disorder. It is generally slow-growing and most often found in the torso and limbs. It appears as pink or red papules, nodules, and/or tumors. PCMZL is always negative for BCL6 and CD10, and often BCL2-positive. Treatment for PCMZL will be discussed in this chapter.

### PC-DLBCL, leg type

Treatment for PC-DLBCL, leg type can be found in *NCCN Guidelines for Patients: Diffuse Large B-Cell Lymphomas* at [NCCN.org/patientguidelines](https://www.nccn.org/patientguidelines) and on the [NCCN Patient Guides for Cancer](#) app.

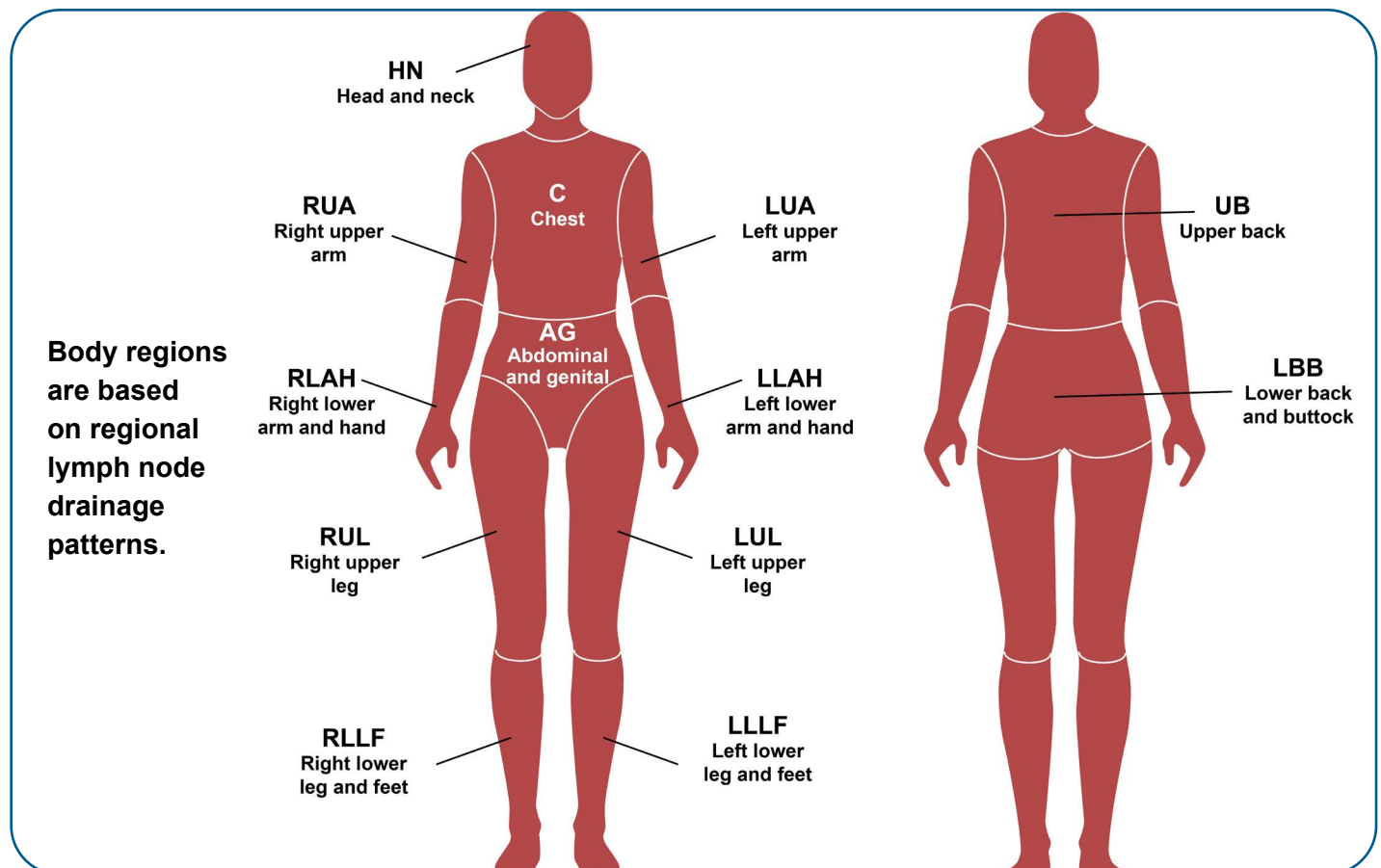


## Diagnosis

A skin biopsy is an important part of diagnosis. Since it is very difficult to distinguish between PCFCL and PC-DLBCL, leg type, biopsy results should be reviewed by an expert pathologist. Immunophenotyping will help determine the specific type of primary cutaneous B-cell lymphoma (PCBCL).

## Treatment

After diagnosis, further testing will be done to plan treatment. This section discusses treatment for PCFCL and PCMZL. Treatment is based on the number of skin lesions and their location. Disease may be solitary, regional, generalized skin only, or generalized extracutaneous.



### Solitary or regional disease

A solitary lesion is one lesion (T1). Regional lesions can be multiple lesions limited to one body region or two adjoining regions (T2). Disease area will be measured.

Options include:

- Local involved-site radiation therapy (ISRT) (preferred)
- In some cases, observation, removal of lesion (excision), skin-directed therapy, or intralesional steroids

Local ISRT is the preferred initial treatment, but not necessarily the preferred treatment for relapse. Initial treatment is the first treatment used. It might be called primary treatment.

After initial treatment, response will be assessed. You might enter a period of observation if there are no signs of disease. If cancer returns or progresses, treatment will depend on the number of lesions and regions, and whether or not there are any symptoms.

### Generalized skin-only disease

Generalized skin-only disease covers a larger area of the body than regional disease. There are multiple lesions that involve 2 or more body regions (T3) not next to one another. Disease is not found in lymph nodes, blood, or other organs.

Treatment options include:

- Observation
- Skin-directed therapy
- Local ISRT
- Intralesional steroids
- Rituximab
- Other systemic therapy

Treatment targets the skin lesions. Some treatments are placed directly on or in the lesion, while others work inside the body. ISRT targets a specific area of skin. Observation or watch-and-wait might be an option for someone without symptoms (asymptomatic). After initial treatment, response will be assessed. You might enter a period of observation if there are no signs of disease.

- **Relapse** is cancer that returns after a disease-free period. If cancer relapses or progresses, treatment will be based on if disease is generalized (skin only) or extracutaneous (outside the skin).
- **Refractory disease** is disease that is not responding to treatment. It may be resistant at the beginning of treatment or it may become resistant during treatment. For refractory disease, a different therapy will be given.



### **Extracutaneous disease**

Extracutaneous disease is found in areas other than the skin. This is cancer that might be found in the lymph nodes, blood, or organs. If you have skin-only disease and later develop extracutaneous disease, you will keep your original diagnosis, but the stage will change.

If you have a skin lesion and testing finds extracutaneous disease at the time of initial diagnosis, you often (but not always) would be classified as having systemic lymphoma. Treatments are based on the type of B-cell lymphoma.

**An intralesional steroid is injected directly into a lesion on or immediately below the skin.**

**Your preferences about treatment are always important. If you have any religious or personal beliefs about certain kinds of treatment, share them with your care team and make your wishes known.**



## Key points

- Primary cutaneous B-cell lymphoma (PCBCL or CBCL) starts in B cells, a type of white blood cell.
- A skin biopsy is an important part of diagnosis. Results should be reviewed by an expert pathologist.
- Immunophenotyping will help determine the specific type of PCBCL.
- Lesions or tumors will be measured by their depth, height, size, and region of the body. Lesions are often measured in centimeters (cm).
- Body regions are based on regional lymph node drainage patterns. Body regions include head/neck, chest, upper arm, lower arm and hand, abdomen and genitals, upper leg, lower leg and feet, upper back, lower back, and buttocks.
- Treatment is based on the number of skin lesions and their location.
- Generalized extracutaneous disease is found in areas other than the skin. Cancer may be in lymph nodes, blood, or internal organs such as the liver or spleen.



### **We want your feedback!**

**Our goal is to provide helpful and easy-to-understand information on cancer.**

**Take our survey to let us know what we got right and what we could do better.**

[NCCN.org/patients/feedback](https://www.nccn.org/patients/feedback)

# 5

## Making treatment decisions

- 41 It's your choice
- 41 Questions to ask
- 51 Resources

**It's important to be comfortable with the cancer treatment you choose. This choice starts with having an open and honest conversation with your care team.**

### It's your choice

In shared decision-making, you and your care team share information, discuss the options, and agree on a treatment plan. It starts with an open and honest conversation between you and your care team.

Treatment decisions are personal. What is important to you may not be important to someone else. Some things that may play a role in your decision-making:

- What you want and how that might differ from what others want
- Your religious and spiritual beliefs
- Your feelings about certain treatments
- Your feelings about pain or side effects
- Cost of treatment, travel to treatment centers, and time away from school, work, or family
- Quality of life and length of life
- How active you are and the activities that are important to you

Think about what you want from treatment. Discuss openly the risks and benefits of specific treatments and procedures. Weigh options and share concerns with your

care team. If you take the time to build a relationship with your care team, it will help you feel supported when considering options and making treatment decisions.

### Second opinion

It is normal to want to start treatment as soon as possible. While cancer can't be ignored, there is time to have another doctor review your test results and suggest a treatment plan. This is called getting a second opinion, and it's a normal part of cancer care. Even doctors get second opinions!

Things you can do to prepare:

- Check with your insurance company about its rules on second opinions. There may be out-of-pocket costs to see doctors who are not part of your insurance plan.
- Make plans to have copies of all your records sent to the doctor you will see for your second opinion.

### Support groups

Many people diagnosed with cancer find support groups to be helpful. Support groups often include people at different stages of treatment. Some people may be newly diagnosed, while others may be finished with treatment. If your hospital or community doesn't have support groups for people with cancer, check out the websites listed in this book.

### Questions to ask

Possible questions to ask your care team are listed on the following pages. Feel free to use these questions or come up with your own. Be clear about your goals for treatment and find out what to expect from treatment.

## Questions about testing and diagnosis

1. What type of cancer do I have? What does this mean in terms of my prognosis and treatment options?
2. Is cancer in my blood, lymph nodes, or other organs?
3. Is there a cancer center or hospital nearby that specializes in this type of cancer?
4. What tests will I have? How often will they be repeated?
5. Will my insurance pay for this test?
6. When will I know the test results and who will explain them to me?
7. Would you give me a copy of the pathology report and other test results?
8. Who will talk with me about the next steps? When?
9. Will treatment start before the test results are in?

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## Questions about skin

1. Can I use lotions or oils on my skin or hair other than what you give me? What are the best types of soap, shampoo, sunscreen, hair dye, or makeup for me to use?
2. Is it better to wear long sleeves and/or pants, or cover the rash/lesions in some way? Or should I let my skin be exposed to the air as much as possible?
3. Should I take time to inspect my skin? If so, how often?
4. If I notice any changes in my skin whom should I call and when?
5. Will keeping a diary and photo journal help? What should I include in the diary? How often should I take photos?
6. Can I go out in the sun? Should I wear sunscreen, long sleeves, or a hat?
7. Are there any changes that I can make to my diet, or activity and stress level that will help my condition?

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## Questions about your care team's experience

1. What is your experience treating this type of cancer?
2. What is the experience of those on your team?
3. What types of cancer do you treat?
4. I would like to get a second opinion. Is there someone you recommend?
5. How many people like me (of the same age, gender, race) have you treated?
6. Will you be consulting with experts to discuss my care? Whom will you consult?
7. How many procedures like the one you're suggesting have you done?
8. Is this treatment a major part of your practice?
9. What types of complications are possible?
10. Who will manage my day-to-day care?

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## Questions about options

1. What will happen if I do nothing?
2. Am I a candidate for a clinical trial?
3. Can I join a clinical trial at any time?
4. Which option is proven to work best for my cancer, age, overall health, and other factors?
5. What if I am pregnant or am planning to get pregnant soon?
6. Should I see a fertility specialist or genetic counselor?
7. Can I stop treatment at any time? What will happen if I stop treatment?
8. What are my options if treatment doesn't work as expected?
9. What decisions must be made today? Is there a social worker or someone who can help me decide about treatment?
10. Is there a hospital or treatment center you can recommend for treatment?

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## Questions about treatment

1. Which treatment(s) do you recommend and why?
2. Does this treatment offer a cure? If not, how well can treatment stop the cancer from growing?
3. Does the order of treatment matter?
4. When will I start treatment and how long will treatment likely take?
5. What should I expect from treatment?
6. What will you do to make me comfortable during treatment?
7. How much will my insurance pay for treatment?
8. Are there programs to help me pay for treatment?
9. What are the chances my cancer will return?
10. What is my risk for developing another kind of cancer, such as skin cancer?

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## Questions about radiation therapy

1. What type of radiation therapy (RT) will I have?
2. What will you target?
3. What is the goal of this radiation treatment?
4. Will RT be used with other therapies?
5. How many treatment sessions will I require?
6. Do you offer this type of radiation here? If not, can you refer me to someone who does?
7. What side effects can I expect from RT?
8. Will I be given medicine to help me relax during RT?
9. What should I wear?

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## Questions about clinical trials

1. What clinical trials are available for my type and stage of cancer?
2. What are the treatments used in the clinical trial?
3. What does the treatment do?
4. Has the treatment been used before? Has it been used for other types of cancer?
5. What are the risks and benefits of this treatment?
6. What side effects should I expect? How will the side effects be controlled?
7. How long will I be in the clinical trial?
8. Will I be able to get other treatments if this doesn't work?
9. How will you know the treatment is working?
10. Will the clinical trial cost me anything? If so, how much?

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## Questions about side effects

1. What are the side effects of this treatment?
2. How long will these side effects last?
3. Do any side effects lessen or worsen in severity over time?
4. What side effects are expected and which are life threatening?
5. When should I call the doctor? Can I text?
6. What should I do for an issue on weekends and other non-office hours?
7. What medicines can I take to prevent or relieve side effects?
8. Will you stop treatment or change treatment if there are side effects? What do you look for?
9. What can I do to lessen or prevent side effects? What will you do?
10. What side effects are life-long and irreversible even after completing treatment?

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## Questions about resources and support

1. Who can I talk to about help with housing, food, and other basic needs?
2. What help is available for transportation, childcare, and home care?
3. How much will I have to pay for treatment?
4. What help is available to pay for medicines and other treatment?
5. What other services are available to me and my caregivers?
6. How can I connect with others and build a support system?
7. How can I find in-person or online support?
8. Who can help me with my concerns about missing work or school?
9. Who can I talk to if I don't feel safe at home, at work, or in my neighborhood?
10. How can I get help to stop smoking or vaping?

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## Resources

**AnCan Foundation**

[AnCan.org](https://AnCan.org)

**NMDP**

[nmdp.org](https://nmdp.org)

**BMT InfoNet (Blood & Marrow Transplant Information)**

[bmtinfonet.org](https://bmtinfonet.org)

**The Leukemia & Lymphoma Society (LLS)**

[LLS.org/PatientSupport](https://LLS.org/PatientSupport)

**CancerCare**

[Cancercare.org](https://Cancercare.org)

**Triage Cancer**

[Triagecancer.org](https://Triagecancer.org)

**Cutaneous Lymphoma Foundation**

[Clfoundation.org](https://Clfoundation.org)

**Imerman Angels**

[Imermanangels.org](https://Imermanangels.org)

**Lymphoma Research Foundation**

[lymphoma.org](https://lymphoma.org)

**MedlinePlus**

[medlineplus.gov](https://medlineplus.gov)

**National Bone Marrow Transplant Link (nbmtLINK)**

[nbmtlink.org](https://nbmtlink.org)

**National Cancer Institute (NCI)**

[cancer.gov/types/lymphoma](https://cancer.gov/types/lymphoma)

**National Coalition for Cancer Survivorship**

[canceradvocacy.org](https://canceradvocacy.org)





# Words to know

**biopsy**

The removal of a sample of tissue for testing.

**chemotherapy**

Drugs that kill fast-growing cells, including cancer cells and normal cells.

**clinical trial**

A type of research that assesses health tests or treatments.

**complete blood count (CBC)**

A lab test that includes the number of blood cells.

**dermatologist**

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

**external beam radiation therapy (EBRT)**

A cancer treatment with radiation received from a machine outside the body.

**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 cell traits involved in abnormal cell growth.

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

Targets a specific area of skin. It can also be used to treat specific lymph nodes with cancer.

**lymph**

A clear fluid containing white blood cells.

**lymphatic system**

Germ-fighting network of tissues and organs that includes the bone marrow, spleen, thymus, lymph nodes, and lymphatic vessels. Part of the immune system.

**lymph node**

A small, bean-shaped, disease-fighting structure.

**papule**

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

**papulonodular**

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.

**phototherapy**

Uses different ultraviolet (UV) light wavelengths to treat skin lesions or tumors.

**plaque**

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

**progression**

The growth or spread of cancer after being tested or treated.

**radiation therapy (RT)**

A treatment that uses high-energy rays or related approaches to kill cancer cells.

**refractory**

Cancer that does not respond to multiple treatments.

**relapse**

The return or worsening of cancer after a period of improvement.

**remission**

There are minor or no signs of disease.

**retinoids**

Products related to vitamin A.

**side effect**

An unhealthy or unpleasant physical or emotional response to treatment.

**skin-directed therapy**

Treatment focused on the skin. Includes topical therapy, local radiation, and phototherapy.

**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.

**targeted therapy**

A drug treatment that targets and attacks specific cancer cells.

# NCCN Contributors

This patient guide is based on the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®) for Primary Cutaneous Lymphomas, Version 2.2024. It was adapted, reviewed, and published with help from the following people:

**Dorothy A. Shead, MS**  
Senior Director  
Patient Information Operations

**Tanya Fischer, MEd, MSLIS**  
Senior Medical Writer

**Susan Kidney**  
Senior Graphic Design Specialist

The NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®) for Primary Cutaneous Lymphomas, Version 2.2024 were developed by the following NCCN Panel Members:

**Steven M. Horwitz, MD/Chair**  
Memorial Sloan Kettering Cancer Center

**Stephen Ansell, MD, PhD/Vice-Chair**  
Mayo Clinic Comprehensive Cancer Center

**Weiyun Z. Ai, MD, PhD**  
UCSF Helen Diller Family  
Comprehensive Cancer Center

**Jeffrey Barnes, MD, PhD**  
Mass General Cancer Center

**Stefan K. Barta, MD, MRCP, MS**  
Abramson Cancer Center  
at the University of Pennsylvania

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

**Mark W. Clemens, MD**  
The University of Texas  
MD Anderson Cancer Center

**Utpal P. Davé, MD**  
Indiana University Melvin and Bren Simon  
Comprehensive Cancer Center

**Ahmet Dogan, MD, PhD**  
Memorial Sloan Kettering Cancer Center

**Francine Foss, MD**  
Yale Cancer Center/Smilow Cancer Hospital

**Zachary Frosch, MD, MSHP**  
Fox Chase Cancer Center

**Aaron M. Goodman, MD**  
UC San Diego Moores Cancer Center

**Joan Guitart, MD**  
Robert H. Lurie Comprehensive Cancer  
Center of Northwestern University

**Ahmad Halwani, MD**  
Huntsman Cancer Institute  
at the University of Utah

**Bradley M. Haverkos, MD, MPH, MS**  
University of Colorado Cancer Center

**Francisco Hernandez-Ilizaliturri, MD**  
Roswell Park Comprehensive Cancer Center

**Richard T. Hoppe, MD**  
Stanford Cancer Institute

**Eric Jacobsen, MD**  
Dana-Farber/Brigham and  
Women's Cancer Center

**Deepa Jagadeesh, MD, MPH**  
Case Comprehensive Cancer Center/  
University Hospitals Seidman Cancer  
Center and Cleveland Clinic Taussig  
Cancer Institute

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

**Youn H. Kim, MD**  
Stanford Cancer Institute

**Kiran Kumar, MD, MBA**  
UT Southwestern Simmons  
Comprehensive Cancer Center

**Neha Mehta-Shah, MD, MSCI**  
Siteman Cancer Center at Barnes-  
Jewish Hospital and Washington  
University School of Medicine

**Elise A. Olsen, MD**  
Duke Cancer Institute

**Saurabh A. Rajguru, MD**  
University of Wisconsin  
Carbone Cancer Center

**Peter Riedell, MD**  
The UChicago Medicine  
Comprehensive Cancer Center

**\*Sima Rozati, MD, PhD**  
The Sidney Kimmel Comprehensive  
Cancer Center at Johns Hopkins

**\*Jonathan Said, MD**  
UCLA Jonsson  
Comprehensive Cancer Center

**Aaron Shaver, MD, PhD**  
Vanderbilt-Ingram Cancer Center

**\*Lauren Shea, MD**  
O'Neal Comprehensive  
Cancer Center at UAB

**Michi M. Shinohara, MD**  
Fred Hutchinson Cancer Center

**Lubomir Sokol, MD, PhD**  
Moffitt Cancer Center

**Matthew Stephany, MD**  
Fred & Pamela Buffett Cancer Center

**\*Susan Thornton**  
Patient Advocate

**\*Carlos Torres-Cabala, MD**  
The University of Texas  
MD Anderson Cancer Center

**Ryan Wilcox, MD, PhD**  
University of Michigan Rogel Cancer Center

**Peggy Wu, MD, MPH**  
UC Davis Comprehensive Cancer Center

**Jasmine Zain, MD**  
City of Hope National Medical Center

## NCCN

**Mary Dwyer, MS**  
Senior Director, Guidelines Operations

**Hema Sundar, PhD**  
Senior Manager, Global Clinical Content

\* Reviewed this patient guide.

For disclosures, visit [NCCN.org/disclosures](https://www.nccn.org/disclosures).

# NCCN Cancer Centers

**Abramson Cancer Center**  
at the University of Pennsylvania  
Philadelphia, Pennsylvania  
800.789.7366 • [penmedicine.org/cancer](https://penmedicine.org/cancer)

**Case Comprehensive Cancer Center/  
University Hospitals Seidman Cancer Center and  
Cleveland Clinic Taussig Cancer Institute**  
Cleveland, Ohio  
UH Seidman Cancer Center  
800.641.2422 • [uhhospitals.org/services/cancer-services](https://uhhospitals.org/services/cancer-services)  
CC Taussig Cancer Institute  
866.223.8100 • [my.clevelandclinic.org/departments/cancer](https://my.clevelandclinic.org/departments/cancer)  
Case CCC  
216.844.8797 • [case.edu/cancer](https://case.edu/cancer)

**City of Hope National Medical Center**  
Duarte, California  
800.826.4673 • [cityofhope.org](https://cityofhope.org)

**Dana-Farber/Brigham and Women's Cancer Center |  
Mass General Cancer Center**  
Boston, Massachusetts  
877.442.3324 • [youhaveus.org](https://youhaveus.org)  
617.726.5130 • [massgeneral.org/cancer-center](https://massgeneral.org/cancer-center)

**Duke Cancer Institute**  
Durham, North Carolina  
888.275.3853 • [dukecancerinstitute.org](https://dukecancerinstitute.org)

**Fox Chase Cancer Center**  
Philadelphia, Pennsylvania  
888.369.2427 • [foxchase.org](https://foxchase.org)

**Fred & Pamela Buffett Cancer Center**  
Omaha, Nebraska  
402.559.5600 • [unmc.edu/cancercenter](https://unmc.edu/cancercenter)

**Fred Hutchinson Cancer Center**  
Seattle, Washington  
206.667.5000 • [fredhutch.org](https://fredhutch.org)

**Huntsman Cancer Institute at the University of Utah**  
Salt Lake City, Utah  
800.824.2073 • [healthcare.utah.edu/huntsmancancerinstitute](https://healthcare.utah.edu/huntsmancancerinstitute)

**Indiana University Melvin and Bren Simon  
Comprehensive Cancer Center**  
Indianapolis, Indiana  
888.600.4822 • [www.cancer.iu.edu](https://www.cancer.iu.edu)

**Johns Hopkins Kimmel Cancer Center**  
Baltimore, Maryland  
410.955.8964  
[www.hopkinskimmelcancercenter.org](https://www.hopkinskimmelcancercenter.org)

**Mayo Clinic Comprehensive Cancer Center**  
Phoenix/Scottsdale, Arizona  
Jacksonville, Florida  
Rochester, Minnesota  
480.301.8000 • Arizona  
904.953.0853 • Florida  
507.538.3270 • Minnesota  
[mayoclinic.org/cancercenter](https://mayoclinic.org/cancercenter)

**Memorial Sloan Kettering Cancer Center**  
New York, New York  
800.525.2225 • [mskcc.org](https://mskcc.org)

**Moffitt Cancer Center**  
Tampa, Florida  
888.663.3488 • [moffitt.org](https://moffitt.org)

**O'Neal Comprehensive Cancer Center at UAB**  
Birmingham, Alabama  
800.822.0933 • [uab.edu/onealcancercenter](https://uab.edu/onealcancercenter)

**Robert H. Lurie Comprehensive Cancer Center of  
Northwestern University**  
Chicago, Illinois  
866.587.4322 • [cancer.northwestern.edu](https://cancer.northwestern.edu)

**Roswell Park Comprehensive Cancer Center**  
Buffalo, New York  
877.275.7724 • [roswellpark.org](https://roswellpark.org)

**Siteman Cancer Center at Barnes-Jewish Hospital  
and Washington University School of Medicine**  
St. Louis, Missouri  
800.600.3606 • [siteman.wustl.edu](https://siteman.wustl.edu)

**St. Jude Children's Research Hospital/  
The University of Tennessee Health Science Center**  
Memphis, Tennessee  
866.278.5833 • [stjude.org](https://stjude.org)  
901.448.5500 • [uthsc.edu](https://uthsc.edu)

**Stanford Cancer Institute**  
Stanford, California  
877.668.7535 • [cancer.stanford.edu](https://cancer.stanford.edu)

**The Ohio State University Comprehensive Cancer Center -  
James Cancer Hospital and Solove Research Institute**  
Columbus, Ohio  
800.293.5066 • [cancer.osu.edu](https://cancer.osu.edu)

**The UChicago Medicine Comprehensive Cancer Center**  
Chicago, Illinois  
773.702.1000 • [uchicagomedicine.org/cancer](https://uchicagomedicine.org/cancer)

**The University of Texas MD Anderson Cancer Center**  
Houston, Texas  
844.269.5922 • [mdanderson.org](https://mdanderson.org)

UC Davis Comprehensive Cancer Center  
Sacramento, California  
916.734.5959 • 800.770.9261  
[health.ucdavis.edu/cancer](http://health.ucdavis.edu/cancer)

UC San Diego Moores Cancer Center  
La Jolla, California  
858.822.6100 • [cancer.ucsd.edu](http://cancer.ucsd.edu)

UCLA Jonsson Comprehensive Cancer Center  
Los Angeles, California  
310.825.5268 • [uclahealth.org/cancer](http://uclahealth.org/cancer)

UCSF Helen Diller Family  
Comprehensive Cancer Center  
San Francisco, California  
800.689.8273 • [cancer.ucsf.edu](http://cancer.ucsf.edu)

University of Colorado Cancer Center  
Aurora, Colorado  
720.848.0300 • [coloradocancercenter.org](http://coloradocancercenter.org)

University of Michigan Rogel Cancer Center  
Ann Arbor, Michigan  
800.865.1125 • [rogelcancercenter.org](http://rogelcancercenter.org)

University of Wisconsin Carbone Cancer Center  
Madison, Wisconsin  
608.265.1700 • [uwhealth.org/cancer](http://uwhealth.org/cancer)

UT Southwestern Simmons  
Comprehensive Cancer Center  
Dallas, Texas  
214.648.3111 • [utsouthwestern.edu/simmons](http://utsouthwestern.edu/simmons)

Vanderbilt-Ingram Cancer Center  
Nashville, Tennessee  
877.936.8422 • [vicc.org](http://vicc.org)

Yale Cancer Center/Smilow Cancer Hospital  
New Haven, Connecticut  
855.4.SMILOW • [yalecancercenter.org](http://yalecancercenter.org)



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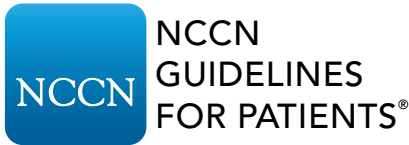
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# Index

- B cell (or B lymphocyte)** 6
- biomarkers** 17–19
- biopsy** 15
- cancer stages** 20–21
- chemotherapy** 29
- clinical trials** 30–31
- cutaneous B-cell lymphoma (CBCL)** 6–7
- cutaneous T-cell lymphoma (CTCL)** 7
- external beam radiation therapy (EBRT)** 28
- fertility** 12, 26
- gene rearrangements** 18
- immunotherapy** 29
- involved-site radiation therapy (ISRT)** 27, 28
- local therapy** 27
- lymph node** 5, 15, 21, 36
- lymphocytes** 6
- mutations** 17–19
- phototherapy** 27
- pregnancy** 12, 26
- primary cutaneous lymphoma (PCL)** 6
- primary cutaneous diffuse large B-cell lymphoma (PC-DLBCL), leg type** 6–7, 35
- primary cutaneous follicle center lymphoma (PCFCL)** 6–7, 35
- primary cutaneous marginal zone lymphoma (PCMZL) or PCMZ lymphoproliferative disorder** 6–7, 35
- radiation therapy (RT)** 27, 28
- retinoids** 27, 29
- side effects** 32
- skin-directed therapy** 27
- skin** 11–12, 20
- surgery** 27
- survivorship** 32
- targeted therapy** 29
- topical therapy** 27







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