

Cutaneous  
Lymphoma  
Basics

A Patient's Guide To Understanding  
**Cutaneous Lymphoma**

Third Edition

A comprehensive guide  
for patients, care partners and others  
affected by cutaneous lymphoma





# CUTANEOUS LYMPHOMA FOUNDATION

## WELCOME!

If you or someone close to you has been given a diagnosis of cutaneous lymphoma, you probably have many questions and concerns. Living with cutaneous lymphoma and the many changes that this diagnosis brings to your life can leave you feeling overwhelmed, confused and lonely. You may not even know what kinds of questions to ask.

This guide was created so you can find valuable information to help you understand the disease and know what questions to ask. Inside these pages you will learn the basics about treatments, how to find specialists, access support, and more to empower you to feel confident to make informed decisions about your treatment and care.

The Cutaneous Lymphoma Foundation is dedicated to providing anyone impacted by cutaneous lymphoma, up-to-date information that you can turn into knowledge to help you live life fully even with this diagnosis. Every program or supportive service is created with a focus on being your guide throughout your personal journey.

We are here for you.

You are not alone.

You are part of a knowledgeable, caring, resourceful, and compassionate community.

Get in touch anytime. Give us a call or drop us an email. Participate in an educational event live or via the web. Ask all your questions. Give us your feedback and let us know how we can serve you.

We wish you all the best in your journey.

*The Staff and Board of Directors of the Cutaneous Lymphoma Foundation*

# A Patient's Guide to Understanding CUTANEOUS LYMPHOMA

This guide is an educational resource published by the Cutaneous Lymphoma Foundation providing general information on cutaneous lymphoma. Publication of this information is not intended to take the place of medical care or the advice of your physician(s). Individuals are strongly encouraged to talk to their physician(s) for complete information on how their disease should be diagnosed, treated and followed. Before starting treatment, individuals should discuss the potential benefits and side effects of any therapy.



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To see a list of all those who have contributed to this publication, please visit [clfoundation.org/guide\\_workgroup](http://clfoundation.org/guide_workgroup)

This version of the guide was made possible through the generous support of our donors and the following corporate partners:



***Disclaimer: The Cutaneous Lymphoma Foundation does not endorse any drugs, treatments or products in this guide. Information is provided for informational purposes only. Because the symptoms and severity of cutaneous lymphomas vary among individuals, the Cutaneous Lymphoma Foundation recommends that all drugs and treatments be discussed with reader's physician(s) for proper evaluation, treatment and medical care.***

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# **PART 1: LEARNING THE BASICS**

## Chapter 1

# OVERVIEW OF LYMPHOMAS

You've been diagnosed with cutaneous lymphoma—the first thing you may ask is: What does that mean and how might it affect me?

The easiest way to face any diagnosis is to start to understand it—by breaking down the information into manageable pieces. First, let's look at what lymphoma is in general.

Lymphoma is a cancer of a family of white blood cells called lymphocytes. When an infection invades the body, lymphocytes are the cells responsible for recognizing the specific germs responsible for the infection, clearing them, and providing long-term immunity against them. Because lymphocytes travel, reside, and work within the lymphatic system (page 2, Figure 1), the lymph nodes swell and grow tender, one sign that an infection has settled in.

There are two types of lymphocytes: B-cells and T-cells. Both work toward similar goals: they identify and fight infections or abnormal cells. B-cells work mostly indirectly: they produce antibody proteins that attach to abnormal cells or infectious agents and alert the immune system to get working. T-cells find the germs, help B-cells make the antibodies, do some direct destroying, and turn on and off the immune response.

A diagnosis of lymphoma means that one or more lymphocytes (B-cell or T-cell) have mutated and are multiplying uncontrollably. This is the hallmark of cancer. Naming a specific cancer is based on the type of cells that are involved and from where it starts, not from where it may travel to. For example, if a patient has breast cancer and it migrates to a lymph node or to the bone, it's still breast cancer, not lymphoma or bone cancer. Like all cancers, lymphomas are now named for the specific type of cells which make them. Thus we have B-cell lymphomas and T-cell lymphomas. This was not always true, however.

Historically, the only diagnostic tool the doctors had was the microscope. Not much was known about B-cells and T-cells, and only two kinds of lymphoma were recognized: Hodgkin's lymphoma (HL) and non-Hodgkin's lymphoma (NHL). HL was the first lymphoma to be identified, and all others were



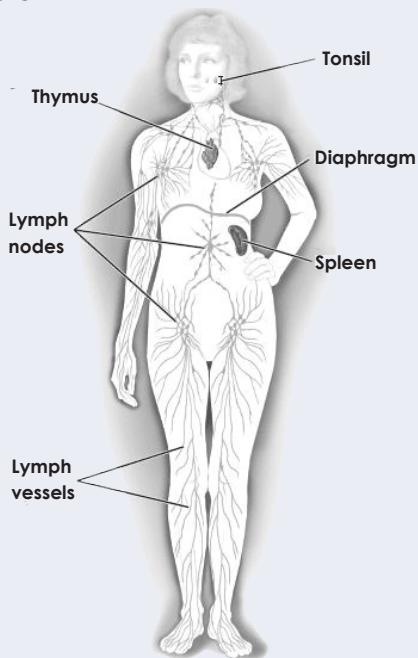
called NHL by exclusion. With the progress in genetics, molecular biology, and immunology, we have learned that the two old classifications actually comprise more than 67 subtypes of lymphoma—six primary types of HL and as many as 61 types of NHL. Now that each type of NHL can be specifically recognized, the distinction between HL and NHL is much less important. All types of cutaneous lymphoma, B-cell and T-cell, belong to the NHL family, of which they represent a very distinct subset.

If one considers all NHLs, B-cell NHLs are far more common than T-cell NHLs: 85% versus 15%. The reason for this is not known. However, if we look only at cutaneous lymphomas, the opposite is true: B-cell lymphomas are far less common than T-cell lymphomas (20-25% versus 75-80%). This suggests that the development of cutaneous lymphomas is distinct from that of other lymphomas.

Because the lymphatic system is connected and works together with the blood and the bone marrow systems, lymphomas are considered blood cancers, like leukemia and myeloma. Lymphoma is the most common blood cancer in the United States and the third most common childhood cancer. The disease is more common in men than in women, with no scientifically identifiable risk factors or causes. Most cases of NHL are found in adults. The average age of diagnosis is in the early-to-mid 60s.

Many lymphomas are potentially curable—meaning you can rid yourself of the disease forever, not just put it into remission. This may require very strong treatments, though, especially in the case of an aggressive form. Physicians think about lymphoma in terms of its behavior—aggressive or indo-

**Figure 1**



Lymphatic system diagram from the  
National Cancer Institute (NCI) booklet  
*What You Need To Know About Non-Hodgkin's Lymphoma*

lent. Simply put, aggressive lymphomas grow more quickly. If not treated, the disease can make a person very sick or become life-threatening within months. Most cutaneous lymphomas are indolent (not aggressive and slow growing).

An indolent lymphoma proceeds over years, often does not make people sick and can be treated multiple times. In most cases, these conditions are chronic rather than curable, and physicians look to manage the situation over the course of a person's life. Most skin lymphomas are very good examples of indolent lymphomas.

**Please refer to our website for additional and updated information:**

**[clfoundation.org](http://clfoundation.org)**

## Chapter 2

# OVERVIEW OF CUTANEOUS LYMPHOMAS

In Chapter 1 we learned that cutaneous lymphomas are a distinct subset of non-Hodgkin’s lymphoma (NHL), and that they can be divided into cutaneous B-cell lymphomas and cutaneous T-cell lymphomas. Unlike most other types of lymphoma, which develop in lymph nodes, people with cutaneous lymphoma have a cancer of lymphocytes that develops primarily in the skin. While the skin is not typically considered a lymphatic organ, it is the largest surface of interaction between the human body and the environment, and as such is a very important battleground for normal immune responses.

Billions of lymphocytes normally travel to the skin and make it their home for variable lengths of times. When one of these lymphocytes mutates and starts growing uncontrollably, people develop cutaneous lymphoma. The course of the disease will vary depending upon the specific type (B-cell or T-cell), the patient conditions and the stage of diagnosis.

This chapter provides an overview of terms and diagnoses that you might encounter. It is important to know and use the most specific name possible for your disease, so that you can understand the proper course of treatment, what to expect, and potential outcomes. Talk with your healthcare provider about your specific case or condition so you are armed with the most up-to-date and thorough knowledge possible.

## CUTANEOUS T-CELL LYMPHOMA (CTCL)

CTCL is the acronym for cutaneous T-cell lymphoma, a general term for several types of lymphomas of the skin that derive from T-cells, including mycosis fungoides, Sézary syndrome, primary cutaneous anaplastic large cell lymphoma, lymphomatoid papulosis, granulomatous slack skin disease, pagetoid reticulosis, and subcutaneous panniculitis-like T-cell lymphoma, to name a few. Most CTCLs typically fall into the category of indolent (i.e., chronic) lymphomas—treatable and usually not life-threatening.

In CTCL, malignant T-cells travel to the upper layers of the skin, causing a rash, which leads to diagnosis. CTCL is sometimes wrongly referred to as a skin cancer because it affects the skin, but this is not a precise use of the term “skin cancer.” Skin cancer is the designation for cancers that develop from

other, non-lymphoid cells of the skin, including epidermal cells (which lead to squamous cell carcinoma) and melanocytes or pigment cells (which lead to melanoma).

As a group, CTCL is a relatively rare family of diseases with approximately 3,000 new cases reported in the United States every year. More common among men than women, CTCL occurs more in patients older than 50 years of age than in younger people. It is important to know, too, that CTCL is not contagious. It is not an infection and cannot be passed from person to person.

Some patients enter long-term remission with treatment and live symptom-free for many, many years. The most recent research indicates that patients diagnosed with the early stages of the most common type of CTCL—mycosis fungoides (which makes up about 70% of CTCL)—have a normal life expectancy. With advances in research and new treatment options resulting from physician collaboration and clinical trials, CTCL patients are experiencing better care and an array of effective treatment options that work for them.

Treatments vary from patient to patient, depending on symptoms, stage of disease and personal health profile. Treatments are either directed at the skin (skin-directed therapies) or the entire body (systemic therapies). It is important that patients discuss fully with their physician all the treatment options before deciding on a course of action.

Skin-directed therapies include ultraviolet light (PUVA, UVB, narrow-band UVB), topical steroids, topical chemotherapy (nitrogen mustard, carmustine), topical retinoids, local radiation to single lesions or total skin electron beam (TSEB) radiation. Full-body, or systemic, therapies include oral retinoids, photopheresis, interferon, biologic agents/targeted therapies and systemic chemotherapy. All treatments may be prescribed alone or in combination, with the goal of achieving the best, long-term benefits for the patient.

The two most common types of CTCL are mycosis fungoides (MF) and Sézary syndrome (SS). Together, they make up about three quarters of all CTCL.

## **MYCOSIS FUNGOIDES (MF)**

Mycosis fungoides is the most common form of CTCL. Because of that, the terms MF and CTCL are often used interchangeably, and sometimes imprecisely. For example, much of the research on MF reported in the medical journals is listed under the name CTCL, even though MF is just one type of CTCL.

This can be a source of confusion. All cases of MF are CTCL, but not all CTCL cases are MF. Mycosis fungoides can appear anywhere on the body, but tends to affect areas of the skin protected from sun by clothing.

MF is an indolent type of CTCL, follows a slow, chronic course and very often does not spread beyond the skin. Over time, in about 10% of cases, it can progress to lymph nodes and internal organs. Symptoms of MF can include flat, red, scaly patches, thicker raised lesions called plaques, and sometimes large nodules called tumors. The disease can progress over many years, often decades.

Although there is continuing research, at this time no single factor has been proven to cause this disease. There is no supportive research indicating that it is genetic or hereditary. Studies have failed to show connections between chemical exposure, environment, pesticides, radiation, allergies and occupations. Exposure to Agent Orange may be a risk factor for developing CTCL for veterans of the Vietnam War, but no direct cause-effect relationship has been established.

While the number of new cases diagnosed each year is relatively low (about 3,000), it is estimated that, since patients have a very long survival, there may be as many as 30,000 patients living with cutaneous lymphoma in the United States and Canada. Due to the difficulty of diagnosing the disease in its early stages and the lack of an accurate reporting system, these numbers are estimates.

One of the challenges in describing this disease is that it doesn't look the same for all patients. Patches, plaques and tumors are the clinical names for different skin manifestations and are generally defined as "lesions." Patches are usually flat, often scaly and look like a "rash." Plaques are thicker, raised lesions. MF patches and plaques are often mistaken for eczema, psoriasis or "non-specific" dermatitis until an exact diagnosis is made.

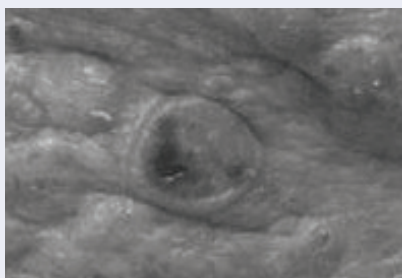
Tumors are raised "bumps" or "nodules" which may or may not ulcerate (open sores). A common characteristic is itching, although not all patients experience this symptom. Most of the time patients present with patches or plaques. Only rarely are the tumors the presenting lesion. While it is possible to have all three of these types of lesions at the same time, most people who have had the disease for many years experienced only one or two types of lesions, generally patches and plaques.



**Patches**



**Plaques**



**Tumors**

Photos courtesy of Dr. Henry K. Wong

While, on average, MF is an indolent, chronic disease, the course in individual patients is unpredictable. It can be slow, rapid or static. Most patients will only experience skin symptoms without serious complications. About 10% will see the disease progress to lymph nodes or other internal organs with serious complications. Unlike other types of skin cancer, chiefly melanoma, MF almost never progresses to lymph nodes and internal organs without showing very obvious signs of progression in the skin. Many patients live normal lives while treating their disease, some remaining in remission for long periods of time.

MF is very difficult to diagnose in early stages, as symptoms and skin biopsy findings are similar to other skin conditions, leading to frequent misdiagnosis. Patients may go on for years before a definitive diagnosis is established. Both the clinical findings (based on both history and examination) and the skin biopsy findings are essential for diagnosis. Physicians will examine lymph nodes and often order blood tests

including a test for Sézary cells in the blood. Other screening tests such as computed tomography (CT) or positron emission tomography (PET) scans may be indicated, depending on the patient's history.

## **PEDIATRIC MYCOSIS FUNGOIDES**

A diagnosis of cutaneous lymphoma is rare in the pediatric population and information about children diagnosed is limited. Recent studies of children diagnosed with this rare lymphoma show that hypopigmented mycosis fungoides

(loss of skin color) is the most prevalent, and most children present with early stage disease. Based on the limited data in the pediatric population undergoing treatment, most patients achieve good responses with phototherapy. The disease generally follows a slow growing course. Additional research into this patient population is needed to gain a better understanding of prognosis and treatment impact over the long-term disease history. If possible, children should be evaluated by a specialist in the field of cutaneous lymphoma and periodically followed up by a specialist to manage and monitor the treatment plan.

## **SÉZARY SYNDROME (SS)**

Sézary syndrome is a less common but more aggressive type of CTCL that is related to MF but presents with very severe itching, total body redness (erythroderma), intense scaling of the skin, and frequent hair loss. Lymph nodes are usually enlarged, and the malignant T-cells found in the skin are also seen circulating in the bloodstream.

SS is the only type of CTCL that always affects the skin and the blood. The skin may be red from head to toe. Tumor cells are found in the blood, and lymph nodes are larger than usual. The skin may be hot, sore, extremely itchy, occasionally flaking and burning. Oozing of clear fluid from the skin is common. Because much heat is lost through the skin, people often feel cold. Symptoms may be accompanied by changes in nails, hair or eyelids. Approximately 15% of patients with CTCL have SS. This disease usually occurs in adults older than 50 and is found more in men than women.

Both MF and SS are rare cancers requiring a definitive diagnosis in order to proceed with treatments. Like all cancer diagnoses, both are classified according to stage, which defines how advanced the disease is at the time of diagnosis.

## **FOLLICULOTROPIC MYCOSIS FUNGOIDES (FMF)**

Folliculotropic mycosis fungoides (FMF) affects less than 10% of the cutaneous lymphoma patient population. This subtype can be categorized into two distinct variations: indolent (slow growing) and advanced, each with its own distinctive clinical and pathological pathway.

Clinical presentation of FMF is somewhat different from MF but a biopsy is required to diagnose this entity. Scalp and facial involvement are common presentations, with hair loss being one of the more common attributes. Roughly 10% of patients have large cell transformation. Large-scale studies on the clinical course and treatment response with newer agents are needed.

Slow growing or early stage FMF patients benefit from non-aggressive standard skin-directed therapies similar to classic mycosis fungoides. For FMF, however, a higher number of treatments often are needed to achieve a response compared to treatment of classical MF. More advanced stage FMF is less responsive to standard skin-directed therapies. More directed treatments like PUVA, various forms of radiotherapy or combination therapies may be more effective than a single type of treatment.

FMF prognosis depends upon the variant (slow growing or advanced). And prognosis appears to be consistent with classic mycosis fungoides (MF).

## **CD30+ LYMPHOPROLIFERATIVE DISORDERS**

Lymphomatoid papulosis (LyP) is a disease of the immune system that manifests with self-healing small bumps and spots on the skin that come and go. It can be persistent with frequent, recurring eruptions or it can disappear for an extended period of time before showing up again. Stress is often reported to trigger breakouts.

LyP belongs to a family of conditions called primary cutaneous CD30-positive lymphoproliferative disorders (pcCD30+LPD). The name lymphoproliferative disorder is used to define a broad range of diseases of the immune system that share a common biology (in this case the presence of CD30-positive T-cells) and may span from non-malignant, inflammatory or infectious, all the way to full-blown lymphoma. In the family of pcCD30+LPD, LyP is usually classified as non-malignant or as a CTCL precursor, though some experts say it is a very low-grade form of CTCL.

This is a one-in-a-million disease. LyP's overall prevalence is only 1.2-1.9 cases per 1 million people. That said, onset can happen at any time in life – from early childhood to middle age, affecting both genders equally. Black-skinned individuals seem less affected than other racial groups.

LyP is characterized by red-brown bumps and spots, which heal on their own by turning scaly or forming a crust. Lesions can be unnoticed or itchy and painful, taking 2-3 months to run their course.

This disease is not contagious, and there has been no supporting evidence to indicate that it is genetic-based or hereditary. In fact, scientists have yet to find a single risk factor for LyP.



The good news is that, in more than 90% of cases, LyP is a persistent but limited disease that does not affect a patient's overall health. There is no known cure, but there are many treatments for living with the disease.

At the other end of the spectrum of pcCD30+LPD, anaplastic large cell lymphoma (ALCL) is a true T-cell lymphoma, which comprises about 20% of all CTCL, but only 3% of all non-Hodgkin's lymphomas in adults. The disease is characterized by the large size and misshape of the cells under the microscope and by the uniform expression of a special marker on the lymphoma cells called CD30.

Like MF, primary cutaneous ALCL (pcALCL) is an indolent, slow-growing type of CTCL, with a good prognosis. Because they are part of the same spectrum of diseases, pcALCL and LyP often are found together in the same patient. LyP, unlike pcALCL, almost always shows spontaneous self-healing, and is occasionally a precursor to the development of pcALCL or other lymphomas, most commonly MF.

Characteristic features of pcALCL include single or multiple raised red skin lesions and nodules, which are larger than those seen in LyP, may itch, do not typically crust and they have a tendency to ulcerate. These lesions may appear anywhere on the body and grow very slowly, so they may be present for a long time before being diagnosed.

## **CUTANEOUS B-CELL LYMPHOMA (CBCL)**

As discussed in Chapter 1, in general, B-cell non-Hodgkin's lymphomas are much more common than T-cell non-Hodgkin's lymphomas (85% versus 15%). However, in the skin, the opposite is true: CTCL makes up about 75-80% of all cutaneous lymphomas, whereas CBCL makes up about 20-25%. CBCLs are B-cell non-Hodgkin's lymphomas which originate in skin-based B-cells. The fact that most skin-resident lymphocytes are T-cells, rather than B-cells, may explain the difference. (Please note the percentage of B-cell lymphomas vs. T-cell lymphomas are derived only from US statistics.)

Since systemic or nodal B-cell non-Hodgkin's lymphomas can secondarily involve the skin, when a skin biopsy shows B-cell lymphoma it is very important to make sure that the skin is the only organ involved and that there is not a systemic non-Hodgkin's lymphoma. There is no way to tell the difference between a CBCL and a systemic B-cell lymphoma just based on the biopsy. The three primary types of CBCL are primary cutaneous marginal

zone B-cell lymphoma (PCMZL); primary cutaneous follicle center lymphoma (PCFCL); and primary cutaneous diffuse large B-cell lymphoma, leg type (PCDLBCL-LT).

PCMZL and PCFCL are slow-growing lymphomas and are classified as indolent. They respond well to mild treatments, though they often recur. Relapses of slow-advancing CBCL are mostly confined to the skin and rarely life-threatening. Given the slow, non-threatening nature of this type of the disease, some cases may not even warrant treatment.

PCFCL is the most common type of CBCL. It most commonly shows on the head, neck or upper torso. Lesions are pink or red nodules, or slowly-developing tumors. They rarely become open sores or ulcers. Some patients find nodules in many locations on the body, but more often it is a single tumor or small group of nodules.

PCMZL is also a slow-growing B-cell lymphoma which is related to a type of non-Hodgkin's lymphoma known as extranodal lymphoma of mucosa-associated lymphoid tissue (MALT) type. This is the second most common form of CBCL. Patients find pink or red papules, nodules or, less frequently, tumors. It can occur anywhere on the skin, but tends to show up on arms, legs or torso.

Diffuse large B-cell lymphoma, leg type, also known as PCLBCL-LT, is a rare and more dangerous type of CBCL that looks much different under the microscope, and most of the time is found in the lower legs, where it can reach a very significant size. This version of the disease was first recognized as a more aggressive type of CBCL that showed up on the legs of elderly women. Today, experts recognize this type of lymphoma by spotting the unique molecular features that differentiate it from slow-growing versions of CBCL.

While PCLBCL-LT most often shows up on the legs, it can occur anywhere on the skin; other, slower-growing types of CBCL can show up on the legs so it is imperative not to make a rash diagnosis or an assumption just based on where the disease is first noticed. A biopsy is always needed. PCLBCL-LT often grows into large tumors that extend deep into the fat of the body, growing quickly and becoming open sores. Unlike slow-growing types of lymphoma, this one has a high likelihood of spreading outside the skin.

To determine a diagnosis and classification of CBCL, it is important and appropriate to biopsy the affected skin. Once a diagnosis has been made, clinical stag-

## Staging

Following are stages for mycosis fungoides and Sézary syndrome:

- **Stage IA:** Less than 10% of the skin is covered in red patches or plaques.
- **Stage IB:** 10% or more of the skin is covered in patches or plaques.
- **Stage IIA:** Any amount of the skin surface is covered with patches or plaques and lymph nodes are enlarged and inflamed, but the cancer has not spread to the nodes.
- **Stage IIB:** One or more tumors are found on the skin, lymph nodes may be enlarged, but cancer has not spread to the nodes.
- **Stage III:** Nearly all of the skin is reddened and may have patches, plaques or tumors; lymph nodes may be enlarged, but cancer has not spread to them.
- **Stage IVA:** Most of the skin is reddened and malignant cells are found in the blood; cancer has spread to the lymph nodes.
- **Stage IVB:** Most of the skin is red, any amount of skin is covered in patches, plaques or tumors, cancer has spread to other organs.

ing establishes the course of the disease and informs treatment decisions. The prognosis for most cases of CBCL is usually excellent. Studies show a 96% survival rate at 5 years after diagnosis and a 90% survival rate after 10 years.

Prognosis for the more aggressive PCLBCL-LT is much less optimistic with no more than 50% of patients surviving at 5 years. However, while the mortality rate of patients with PCLBCL-LT is much higher than other forms of CBCL, it is important to remember that most diagnoses occur when patients are in their 80s or 90s and survival rates at 5 years include patients who have succumbed to other natural causes.

## Chapter 3

# SIGNS & SYMPTOMS

*A **sign** is something a healthcare provider (or the patient) detects on physical examination (objective). A **symptom** is something a patient notices and states to their healthcare provider (subjective).*

One of the challenges in definitively diagnosing cutaneous lymphoma is that its signs and symptoms are not the same for all patients. Patches, plaques, and tumors are clinical names for a variety of skin presentations (also known as lesions) that can be clues that lead to diagnosis.

Patches are usually flat, possibly scaly, and look like a rash. Plaques are thicker, raised lesions. Patches and plaques are often mistaken for eczema, psoriasis, or non-specific dermatitis until a definitive diagnosis is made. Tumors are raised “bumps” or “nodules” which may or may not ulcerate. To be called a tumor, generally a nodule has to be at least 1 cm in size, or greater. A common symptom is itching, although some patients do not experience this. It is possible to have one or all three of these types of lesions. Some people have the disease for years and only experience one.

The most common form of cutaneous lymphoma, mycosis fungoides, often presents with an area of red, slightly scaly skin, usually in sun-protected parts of the body, with variable size and shape. Common locations for these symptoms are the buttocks, trunk, upper thighs—all areas that are typically shielded from sun exposure. Patients with cutaneous lymphoma find their outbreaks in sun-protected areas of the skin because the natural UV component of sunlight may have a protective effect against mycosis fungoides. The exact reason, however, is not known.

Approximately 25% of people diagnosed with mycosis fungoides present with plaques, which are raised, elevated skin lesions. In the most advanced stages, symptoms may include round, dome-shaped lumps or bumps that can break down and ulcerate.

Other forms of cutaneous lymphoma present in varying ways. Sézary syndrome (SS) is one type that can present in generalized redness affecting 80% or

## Common Signs & Symptoms of Mycosis Fungoides

- Patches
- Plaques
- Itching
- Skin ulcers
- Tumors

## Common Signs & Symptoms of Sézary Syndrome

- Diffuse scaling skin (erythroderma)
- Thickening of palms and soles (hyperkeratosis)
- Hair thinning
- Eyelid margin thickening (ectropion)
- Itching
- Enlarged lymph nodes

more of the skin's surface. Patients with SS tend to experience very intense itching, perhaps the most intense and relentless itching that has ever been described. These patients will also experience a fair amount of scaling, redness and dryness. They often lose large amounts of skin during the night and may find their bed sheets covered with skin flakes in the morning. This variation presents more dramatically than other types of the disease, making it easier to diagnose because the presentation is more unusual. Sézary syndrome patients will likely also feel tired, have enlarged lymph nodes, may run a fever and just generally feel sick.

Many patients find it takes quite a while to get a definitive diagnosis. A patient may see many physicians who may say "You have eczema" or "It's just dry skin." The reality is that the signs and symptoms of this disease are quite similar to other conditions, and it is very difficult to pinpoint a firm diagnosis of cutaneous lymphoma without a biopsy.

Taking the step to biopsy the affected skin can often provide answers. It is almost impossible to have cutaneous lymphoma without at least some signs or symptoms. This type of disease rarely is unnoticed. However, the milder symptoms are commonly confused with other conditions. Some patients, when misdiagnosed, try treatments prescribed for other conditions like eczema or allergies. When those do not work, it can be a clue in the puzzle and can help move a patient toward a proper diagnosis or a referral to a specialist.

### **Cutaneous Lymphoma Symptoms Are Variable**

Due to the many different types of cutaneous lymphoma, no two people have exactly the same signs and symptoms, and each person's physical findings and symptoms can change over time.

One person may experience only one symptom while another may experience many more. Most of the symptoms of cutaneous lymphoma can be managed very effectively with medication and other health management strategies.

## Chapter 4

# GETTING A DIAGNOSIS

It can be very difficult to make a diagnosis of cutaneous lymphoma, and this is largely because the signs and symptoms very often look like other conditions such as eczema, allergies, or drug reactions.

Diagnosis of the many subtypes of cutaneous lymphomas can vary and sometimes it takes a long time before it is confirmed. Regardless, the process for diagnosis is similar for all types and may include a physical exam and history; blood tests to identify antigens, or markers, on the surface of cells in the blood; and a skin biopsy (removal of a small piece of tissue) for examination under the microscope by a pathologist (a doctor who studies tissue and cells to identify disease). In the presence of more advanced disease, more testing may be done to determine if the cancer has spread.

Both cutaneous T-cell lymphoma (CTCL) and cutaneous B-cell lymphoma (CBCL) require equal consideration and physician attention in order to reach a proper diagnosis. A definitive diagnosis will help inform treatment decisions and potentially yield better outcomes over time. One of the key concepts for patients and care partners to understand is that a clear diagnosis may take time. While a prompt diagnosis should always be the goal, and a late diagnosis always means deferred relief or resolution of symptoms, it is also important to consider that, in many cases of cutaneous lymphoma, how early a diagnosis is made does not have a major impact on response to treatment and survival. The exceptions are Sézary syndrome and primary cutaneous diffuse large B-cell lymphoma, leg type (PCLBCL-LT) where a prompt diagnosis is very important. The best approach is to collaborate and work with the healthcare team to confirm the disease type.

In milder cases of cutaneous lymphoma, ruling out non-cancerous reactive rashes and lesions resulting from medication, external environmental or drug exposure, or inflammatory conditions of the skin is crucial. From there, physicians and patients can determine the appropriate next steps in care and treatment. With a number of different types and subtypes, building a case for diagnosis based on multiple elements is vital. Assessment of a patient's symptoms, coupled with a history of skin lesions and consistent biopsies, are criti-



## Common Procedures For Obtaining A Proper Diagnosis

- Physical exam and history
- Blood tests
- Skin biopsies

cal components that help build a patient’s individual case and accelerate the diagnostic process.

In some cases, test results, including skin biopsies, are not conclusive. When this happens, patients should discuss options with their healthcare provider. Some may receive the recommendation to seek relief from symptoms by starting a non-systemic treatment such as a topical lotion, steroid cream or ointment, or even ultraviolet light therapy. Others may prefer to wait for a recommendation to take additional biopsies of the skin lesion to aim for more conclusive results.

### IMPORTANCE OF THE SKIN BIOPSY

A biopsy is a simple outpatient procedure that involves removing a small sample of tissue from the skin for examination. When a biopsy is performed, local anesthetic is given prior to the procedure. Following the procedure, the skin is sewn together with a couple of stitches, leaving a very small, almost invisible scar.

Given the complexity of cutaneous lymphomas, it is important to have the diagnosis confirmed by a dermatopathologist or a hematopathologist – these pathologists are experts in diagnosing lymphomas. The pathologist analyzes biological tissue and reviews all test results. Typically, the presence of an excess of certain kinds of T-lymphocyte cells, arranged in certain patterns within the tissue, indicates a skin lymphoma. A definitive diagnosis cannot be obtained without a biopsy, and multiple biopsies are necessary to confirm the presence of excess T-lymphocyte cells or B-lymphocyte cells, and consistent patterns. The physician and pathologist will need to work together to determine a proper diagnosis.

To conduct an effective, informative biopsy, patients need to be off topical steroids and ultraviolet light treatment regimens for at least four to six weeks. While these treatments may provide temporary symptom relief, they can also mask potential symptoms of skin lymphomas and thus delay a patient's definitive diagnosis. Consistent biopsies over time are an integral step in obtaining a proper diagnosis.

## **WHY IT TAKES TIME TO GET A DIAGNOSIS**

Cutaneous T-cell lymphoma is a complex disorder which often takes a significant amount of time to diagnose. Various studies indicate that the average time from first appearance of symptoms to confirmed diagnosis of the disease ranges from two to seven years.

This delay can lead to frustration for both the patient and healthcare providers. There are two main reasons why diagnosis can take time to confirm. The first is that the disease presents differently on the skin. Only after patients do not respond to traditional therapies for less severe conditions do some physicians consider an alternate diagnosis. Also, there is no single test that will be positive for cutaneous lymphoma every time. The most useful test is a skin biopsy because lesions that appear very similar on the skin may look quite different under the microscope. Many patients require multiple biopsies before a satisfying and complete diagnosis is made.

The need for multiple, sequential biopsies can be exasperating and difficult for patients to understand. Some pathologists compare making the diagnosis from a single biopsy to experiencing a party through a keyhole. One glance may tell you what kind of party it is, but you will not get the full picture until you walk through the door.

Mycosis fungoides is difficult to diagnose in early stages as the symptoms and skin biopsy findings are similar to those of other conditions. It is often misdiagnosed for this exact reason and patients need to be persistent. It also may require seeking out a cutaneous lymphoma specialist at a larger health center. These specialists see many patients with cutaneous lymphoma and work with a multidisciplinary team when diagnosing patients.

## Chapter 5

# WORKING WITH YOUR HEALTHCARE TEAM

The best way to manage a disease like cutaneous lymphoma is by assembling the right team of physicians and support individuals to guide your treatment course and help you through each stage of the journey.

Members of your healthcare team may include a combination of the following individuals:

- **You** – You will feel better and more in control when you play an active role in your own health care. It is important to become educated about your disease, your diagnosis, and the options available to you.
- **Your Care Partner(s)** – Family members and friends can offer great support and accompany you to treatment appointments or just be there to talk through the various emotions you may experience as you follow the course of your disease.
- **Dermatologist** – A doctor who specializes in diagnosis and treatment of skin diseases. Some dermatologists specialize in treating cutaneous lymphoma.
- **Oncologist** – A doctor who specializes in diagnosing and treating cancer. Some oncologists specialize in cutaneous lymphoma.
- **Radiation Oncologist** – A doctor who specializes in radiation to treat cancer.
- **Nurse Practitioner** – A registered nurse with advanced education and training who can help your doctors manage the disease, symptoms, and side effects.
- **Social Worker/Clinic Coordinator/Patient Navigator** – This person is tasked with looking at logistics and helping patients plan their course of treatments. This is a person who can help guide patients when they face treatment challenges such as having to travel 50-100 miles for light therapy, communicate with insurance companies, etc.
- **Pharmacist** – An expert in medications who understands and can advise on the use and interactions of medications, and help doctors review allergies and drug to drug interactions (which are common).
- **Financial Counselor** – This is an individual, often within the doctor's office, who helps empower patients to navigate their insurance policy,

plan for additional uncovered treatments and otherwise meet the financial demands of their course of treatment.

- **Psychosocial Therapist** – This person can counsel you through the emotions you have and outcomes of your disease.
- **Spiritual Guide** – Some patients find it comforting to include a spiritual advisor from their faith community in their support team.
- **Nutritionist** – This person is an expert in providing strategies for finding the right foods and nutrients you'll need to sustain you through treatments.
- **Community of Patients** – Individuals diagnosed with any form of a serious disease find comfort in connecting with others who have walked a similar path. Your healthcare provider or your treatment center can connect you with others who have experience with your particular diagnosis. The Cutaneous Lymphoma Foundation is an excellent resource for helping you connect with others. The Foundation website ([clfoundation.org](http://clfoundation.org)) is a good place to start.

Before selecting individuals to guide you through treatment, educate yourself on the available options and interview potential providers before including them in your team. Keep in mind the following steps as you proceed:

- Gather information to make treatment decisions with your healthcare team.
- Ask questions.
- Tell your healthcare team how you are feeling – physically AND emotionally.
- Learn about treatment choices/options and potential side effects.
- Seek relief from itching or pain.
- Ask others for support.
- Assign important tasks or chores to friends and family when needed.
- Ask care partners to help with insurance matters as they arise.
- Find a reliable partner to drive you to and from treatment.

The importance of creating a multidisciplinary team to support your treatment course is vital. At some treatment centers, multimodality clinics exist where you will find all of these support individuals under one roof, but in many cases you will have to compile your own team from the available resources in your town or treatment network. Try to find physicians and support individuals who can work well together, especially with regard to reading test results and communicating with ease over the variables of your diagnosis.

<b>Cancer Specialists</b>	<b>What They Do</b>	<b>Types of Sub-Specialists</b>
Dermatologists	Trained to diagnose and treat skin diseases, including cancers of the skin.	Some dermatologists may specialize in treating patients with cutaneous lymphoma.
Medical Oncologists	Trained in internal medicine and in diagnosing and treating cancer with chemotherapy, biological therapy and other treatments. They often manage the patient's course of treatment, especially when the cancer reaches advanced stages. Medical oncologists may offer consultation services to other physicians treating cancer patients or refer patients to other specialists.	Additional subspecialties can be hematology-oncology.
Radiation Oncologists	Radiologists specialize in treating cancer patients using complex radiologic imaging systems to administer various types of radiation treatment. They work closely with surgical and medical oncologists and other physicians such as dermatologists, who treat cancer patients to help coordinate their overall care.	Radiation oncologists may focus on treating specific types of cancer such as lymphoma and may provide specific types of radiation therapy such as electron beam or laser therapy.
Oncology Nurses and Other Healthcare Professionals	These professionals may specialize in working with specific types of cancer and specific treatment modalities such as photopheresis or dermatology.	Registered nurses, clinical nurse specialists, advanced practice nurses, chemotherapy nurses, radiation therapists and oncology social workers.

## WHEN SHOULD YOU WORK WITH AN ONCOLOGIST?

Oncologists are cancer specialists who treat cancer with chemotherapy or other systemic therapies, such as immunotherapy, vaccines, and biological drugs. That is why for cancers that require additional treatment modalities, such as surgery or radiation (e.g., breast cancer, colon cancer, or lung cancer), you always need to see a surgeon and a radiation specialist, in addition to the oncologist.

Cutaneous lymphomas are approached in a slightly different way. Since the treatment for cutaneous lymphoma may consist of skin-directed therapies (usually administered by dermatologists) or systemic therapies (usually administered by oncologists), depending on the stage, a step approach has often been adopted.

Early stages have typically been treated with skin-directed therapy by dermatologists, with little oncology input, and advanced stages have typically been treated with systemic therapy by oncologists, with little dermatology input. While this system may superficially appear to be simple and economical in terms of time management, visits, tests, and payments, and has produced acceptable outcomes in many cases, it has the big disadvantage that patients are often lost in the critical transition from one type of therapy to the other.

Furthermore, adequate pre-planning for systemic therapy in a patient beginning to display resistance to skin-directed therapy cannot be accomplished even by most dermatologists. Likewise, optimal skin care, which remains an essential component of each patient's treatment plan, even in advanced stages, is rarely provided by the oncologist alone. This fact results in one of two frequently observed outcomes: 1) the patient has to keep shuttling back and forth between the dermatologist's and the oncologist's office, often with poor or no communication, which defies the purpose of efficiency and economy; or 2) the patient simply gets inadequate care. Neither outcome is acceptable. Therefore, rather than focusing on the title and specialty of the healthcare provider, you should try to find a physician who has shown expertise, personal interest, dedication, and leadership in the care of patients with cutaneous lymphoma, regardless of his or her specialty. In some cases it will be a dermatologist, in others an oncologist. Once found, that person will be your "captain," "steward," and "advocate" and will help coordinate your care with any other relevant specialist. Basic rule: one leader.

The only better alternative to this scenario is the ideal situation of a multidisciplinary clinic, where the entire team of doctors is wholly focused and dedicated to the care of patients with cutaneous lymphoma. Multidisciplinary clinics, by definition, have an oncologist and a dermatologist on site, both in a leading role, in addition to a large number of additional supporting staff. Such clinics, unfortunately, are available only in a small number of selected cancer centers.

**Please refer to our website for additional and updated information:**

**[clfoundation.org](http://clfoundation.org)**





**PART 2:  
TREATING CUTANEOUS  
LYMPHOMA**

## Chapter 6

# WHAT YOU SHOULD KNOW BEFORE STARTING TREATMENT

Before you start treatment, it's important to get informed.

The more knowledge you have about what to expect, the more comfortable you will be as you receive treatment. Armed with information, you will be able to better determine, with your healthcare provider, treatment team and family members, that all the options have been weighed and assessed accordingly. Know the full extent of what is available to you before making any decisions. While there is no one perfect pill to make things disappear, a combination of treatments, medications and other approaches—along with a heavy dose of patience—will be your personal remedy. Take into consideration your own capabilities, lifestyle and work demands and other daily details before deciding on a treatment course. It is important to share any potential limiting factors you have (financial, insurance, transportation, distance to treatment centers) with your healthcare providers so that your treatment plan is realistic for you and your care partner(s).

Following is information that can be helpful in preparing yourself for specific treatment options:

### **PREPARING THE SKIN**

For most topical treatments, patients may need to clean and prepare their skin. Patients find better penetration through the skin if their skin is well-moisturized before applying prescribed treatments to the skin. Thus, bathing or taking a shower using warm, not hot, water so the skin is well-hydrated and still slightly damp before applying medication is a good idea. Many patients experience dry skin with their disease; moisturizers work better if the skin is soaked first and then sealed with emollient.

### **PREPARING FOR PUVA**

For patients getting PUVA (psoralen medication +UVA light) treatment, eye care is essential. There exists a theoretical, though minimal, risk of patients developing cataracts due to exposure to UVA lights. With that in mind, it's a good idea to have an eye exam prior to the start of treatment to determine that your eyes are healthy. Also, make sure you have protective eyewear,

UVA-blocking (labeled as 100% UVA-blocking) wrap-around sunglasses, to wear for 24 hours on the days of your PUVA treatments, starting from the time psoralen is taken. This eyewear may be removed at sundown but continued at sunrise the following day until the 24 hour period is up. These are available at many superstores and also treatment centers that administer phototherapy.

Because you will be sensitive to light after undergoing PUVA, you'll want to schedule your treatment sessions accordingly. Getting PUVA first thing in the morning may not be a wise choice as you'll likely spend the rest of the day going in and out of sunlight. Discuss your specific lifestyle demands and routines with your healthcare provider to determine the best time of day to schedule light treatments.

Also discuss with your healthcare team the potential side effects of this or any treatment (which can include nausea) because certain natural remedies, like ginger products, may aid in alleviating such symptoms.

## **PREPARING FOR PHOTOPHERESIS**

The better informed a patient is about photopheresis, the better the procedure will go. Certain blood tests are recommended before beginning photopheresis, where the collected blood is treated with a drug that is later activated by exposure to ultraviolet light. Physicians often check a patient's T-cells, red blood cell count, and blood-clotting to have a baseline before beginning this course of treatment.

Eye care is an issue with photopheresis, so make sure you have your protective eyewear before beginning treatment. Hydration is also important in this course, so patients should make sure to be well-hydrated for several days before each treatment session. The better hydrated a person is, the easier it will be to access veins for treatment. Many doctors recommend that patients tour the photopheresis unit before undergoing treatment and have their veins checked to determine ease of intravenous access for the procedure.

Because the treatment takes time, bring reading material or movies to watch to make the time go faster. You will likely develop relationships with others in the treatment facility. Because many people remain on the same schedule, they often see familiar faces each time.

## RELYING ON FRIENDS AND FAMILY

For skin-directed treatments, you won't need a friend or relative to drive you home. However, you may find comfort in the support of someone close to you when you go for photo therapies, topical therapies or radiation therapies.

When it comes to systemic treatments, especially for the first cycle before you know how you will react, it is a good idea to bring someone with you. A caring companion can allay nerves or fears and, in the event that you have an allergic reaction to a treatment and need relief from an antihistamine agent like Benadryl®, which can be sedating, you will take comfort in the presence of another person who can help you throughout the treatment experience and bring you home.

Even with chemotherapies, most people are not affected immediately upon receiving treatment. Related nausea, vomiting, and other effects may take several days to occur, but it is always comforting to have someone at your side so that anxiety doesn't overwhelm you.

## POST-TREATMENT EXPECTATIONS

Certain types of medicines are bigger offenders for side effects like fatigue, nausea and other issues. Pace yourself and plan your time wisely. Do not expect to be operating at the top of your game. Make sure to communicate any and all side effects with your healthcare team, as many symptoms are treatable, even nausea and vomiting. Be aware of your resources and take advantage of your team of experts, who can guide you toward maintaining optimal health and energy levels during treatments.

## MANAGING ANXIETY

Because some treatment methods can be confining or claustrophobic, you may experience feelings of anxiety before or during treatment. Integrative processes like hypnotherapy can be helpful in maintaining calm and balance as you face such challenges. (See Chapter 16 for more information on integrative medicine). Complementary or supportive care is often helpful in achieving balance for your emotions and your physical symptoms. Consider consulting with a nutritionist, Reiki therapist, massage therapist, hypnotherapy expert, acupuncturist, acupressure therapist, yoga teacher, qigong instructor or others to help you handle stress and maintain a sense of control. Speak to your treatment team about referrals for any of the above complementary/supportive care strategies.

## **PHYSICAL LIMITATIONS DURING TREATMENT**

Many patients experience fatigue or depression during their treatment course. Knowing this is a real possibility helps you prepare for such an outcome by seeking out the help of a therapist to talk with during treatment and altering your schedule to account for reduced energy levels.

Healthcare providers are likely to ask if you have trouble falling asleep, if you have bad dreams and nightmares, and if you're feeling blue or tired. Don't be afraid to seek out antidepressants to ease such side effects. Some treatment medications can actually cause depression. Supportive and complementary care and various medications are available to help you through.

Pace yourself. Do not overload your schedule with commitments. Try to postpone any non-essential appointments until your treatment course finishes. Most importantly, don't hesitate to talk with your healthcare team about how you are feeling and how your treatments are affecting you.

## **DON'T BELIEVE EVERYTHING YOU READ**

In this information age, many patients read research related to their disease on the Internet before ever meeting with their healthcare team. There is so much information out there, and the Internet does not discern between your particular situation and other conditions. Information can produce fear and anxiety – especially if it is not specifically related to your case. Try to resist the temptation to dig out any and all information online. Educate yourself with the resources your healthcare provider recommends and ignore the rest. Hospital-approved patient education materials give you a window into the world of your specific situation, the therapies available to you and approved by your providers, and the outcomes you can anticipate. The Cutaneous Lymphoma Foundation is a comprehensive platform containing educational resources and extensive information about treatment options and the latest therapies.

Many effective treatment options exist for cutaneous lymphoma. Your health-care team will determine the best course of treatment for you based on a set of variables:

- Goals of therapy
- Stage of disease
- Prior treatments and the response to those treatments
- Age and activity level
- Costs, coverage, and accessibility
- Other health conditions such as diabetes, heart disease, etc.
- General health concerns and lifestyle considerations

In treating cutaneous lymphomas, unlike most other cancers, healthcare providers often use the same treatment more than once, such as phototherapy and radiation over the course of your treatment. What worked once often will work again. Patients with early-stage cutaneous T-cell lymphoma (CTCL) can often achieve long-lasting remissions with skin-directed therapies. In CTCL, since malignant T-cells are thought to spend the majority of their time in the skin and are dependent on the skin for survival, therapies aimed at the skin are likely to be effective for long periods of time.

Throughout the United States, there are several cancer centers that have dedicated multidisciplinary cutaneous lymphoma treatment centers. However, you may receive care in a variety of settings, which may include your dermatologist's or medical oncologist's office or a local hospital. It is important during and after treatment to keep a master file of medical records for future reference as needed.

The goal of treatment for cutaneous lymphoma is to clear up all patches, plaques, or tumors; to reduce the number of T-lymphocytes in the blood (for Sézary syndrome); and to relieve symptoms such as pain, itching, burning, and redness. Additionally, patients tend to handle treatments better when they maintain a healthy diet and exercise regimen and report any new symptoms or changes to their physicians during treatment.

Treatment choices for cutaneous lymphoma are directed at either the skin (topical) or the entire body (systemic). It's important to discuss with your healthcare team the benefits and disadvantages of both before making a decision on your treatment course. There have been very few studies done to compare the effectiveness of one therapy for cutaneous lymphoma with another, so it is an individual matter of trial and error until your healthcare team finds the right combination of treatments for you.

Here are some common treatment options:

<b>Skin-Directed Therapies</b>
Topical Corticosteroids
Topical Chemotherapy
Retinoids
Phototherapy
Psoralen with UVA (320-400NM) (PUVA)
Narrowband UVB (nbUVB) and Broadband Ultraviolet B (bbUVB)
Fluorescent Light Therapy
Radiation Therapy
<b>Systemic Therapies</b>
Extracorporeal Photopheresis (ECP)
Interferon
Retinoids
Antibodies
Histone Deacetylase (HDAC)
Chemotherapy

Additional information about treatment options can be found on the Foundation's website, [www.clfoundation.org](http://www.clfoundation.org), and in the *Understanding Treatment Options in Cutaneous Lymphoma* publication.

Before you begin a treatment protocol, you will want to learn as much as possible about the course of action that your healthcare team has designed. Write down questions about your illness, treatments, or side effects to discuss with your doctor. Take someone with you when you go for doctor visits – that way, you won't miss important information with an extra set of ears

present. You may even want to record your healthcare visits for later review, providing you obtain permission from your doctor beforehand.

To help you decide which treatment path is right for you, there are some good questions to ask your healthcare team besides “What are the side effects?” and “How well does this treatment work?” (See page 34 for suggested questions.)

One way to help you track all the details and information is to create a chart of your options and the factors that are important to you. You’ll find a sample worksheet on our website to help you keep track of details ([clfoundation.org/treatment-basics](http://clfoundation.org/treatment-basics)).

There are many effective therapies available for all types of cutaneous lymphoma. It is not always easy to decide which treatment course is optimal for you. Follow these tips to help in your decision-making process:

- Look for a healthcare provider who is comfortable with treating your cancer. Because this is a rare disease, your healthcare provider may need to work with a cutaneous lymphoma specialist to help guide your treatment.
- Ask questions. It is hard to remember what questions you want to ask when you are in the exam room. Write down questions in advance and after each visit so you don’t forget. Take someone with you so you can talk over what the doctor told you.
- Get a second opinion. Many people like to get information from several sources so they feel confident before making treatment decisions and moving forward.
- Take your time. There is a lot to be said for taking time to consider your options and not rush into any decisions. While it is important to start treatment as soon as possible, do not rush to make an immediate decision.
- Make sure your healthcare provider understands you. Share any and all personal details of your lifestyle, schedule, routine, and concerns so that he or she understands where you’re coming from and what factors will affect your treatment choices. Talk to your healthcare team about what is important to you. Depending on your situation, one treatment may be better than another. Ask your healthcare provider about all the possible treatment options so you can have thorough information before making a decision.



## Questions To Ask About Treatment

- 1. How confident are you in my diagnosis of cutaneous lymphoma?** The less confident your doctor is in the diagnosis, the less risk you should take with therapies.
- 2. What type of cutaneous lymphoma do I have?** Patients with mycosis fungoides should expect a different menu of treatment options than patients with Sézary syndrome and cutaneous B-cell lymphoma. For example, topical creams are not effective for Sézary syndrome but may be perfect for mycosis fungoides.
- 3. What stage of cutaneous lymphoma do I have and what is my prognosis?** This question is KEY. In general, early-stage patients (IA, IB, IIA) should consider topically-applied medications or ultraviolet light therapy over pills and IV medications because they are usually very effective, have fewer side effects, and the prognosis is usually very good. Because more advanced-stage patients (IIB, III, IV) have a worse prognosis, these patients may consider taking more risks with therapy choices. However, it's important to recognize that there is a wide variation of how the disease looks among patients within the same stage. Your physician will tailor your treatment regimen(s) based upon the complete picture.
- 4. What are the short and long-term side effects?** How likely are they? Are they reversible, and how long may they last? These are important questions that are a major factor in many patients' decisions.
- 5. How inconvenient are the treatment options? (How often? Where? For how long?).** Unfortunately, all treatments create at least some inconvenience for patients, but each patient will view the details of their protocol differently as to how it fits into their life.
- 6. How expensive are the treatment options and how likely is it that my insurance will pay for them?** Don't hesitate to ask this question! Your clinician may not have an immediate answer, but he or she should help you find one. There's no harm in trying the least expensive option if it is as effective as a more expensive choice.

# COPING WITH TREATMENT SIDE EFFECTS

Treatments are intended to relieve the symptoms of your disease, but it is often the side effects from such treatments that may create discomfort and make daily life challenging. Let your doctor know if you experience any of these side effects from treatments:

- Skin irritation and inflammation
- Redness, rash, and itching
- Heat, tenderness, or burning sensation (like a sunburn)
- Fatigue or depression
- Flu-like symptoms

Many oral medications and IV chemotherapy can also cause a change in appetite or loss of appetite entirely. If you experience chronic nausea, vomiting or weight loss, be sure to seek out a nutritionist to discuss options for increasing nutrient intake in a way that your body can handle and maintain. Other problematic side effects can occur in the GI tract—diarrhea, constipation and the like. Try altering the time of day when you take the medication, which may alleviate appetite or GI side effects.

Many cutaneous lymphoma patients experience extreme fatigue as a side effect of treatment. Anticipate it and pace yourself, building rest periods into your day as needed, and adjusting your work schedule wherever possible. Reorganizing your life to accommodate the changes that come from this disease and its treatments is important. It is also essential to focus on eating right, getting enough sleep and being kind to yourself, rather than checking things off a regular to-do list and pleasing others. Self-care is essential now and will affect the outcomes of your treatment and healing process.

If you experience treatment-induced depression, talk with your physician and don't hesitate to request antidepressants to help boost your mood and energy. Always drink enough liquids such as water and non-caffeinated beverages to maintain hydration. Dietary supplements and protein drinks can help maintain nutrient intake and prevent weight loss.

Generally, side effects may be more or less intense at times, some with cumulative side effects, and some worsening as treatments increase. Patients should understand that each specific treatment protocol leads to its own menu of side effects, so communicate with your doctor.

Skin-related issues can be addressed with soothing emollients, extra moisturization, and increasing fluid intake to keep the skin moist. Apply moisturizers immediately after bathing. Wear non-irritating clothing, and protect your skin from the sun. Watch for signs of infection and report any as they appear. Protective clothing and/or protective dressings may help if a particular area is troublesome or painful.

For excessive itching, oral antihistamines and other medications that help with nerve related itching are available and can offer relief. Good skin care goes hand in hand with healing. Apply topical agents that your healthcare provider prescribes, avoid scratching lesions, and take recommended antihistamines.

If you experience heat or a burning sensation in the skin, there are remedies that can help. Cool soaks, moisturization, and cooling agents with menthol can be helpful. In addition, consider cold compresses or ice applications for severely affected areas.

Always check with your doctor or a pharmacist or both before using any over-the-counter products or supplements to make sure they do not contain harmful ingredients.

## Chapter 9

# ACCESS TO MEDICATIONS AND TREATMENTS

Treatment paths for cutaneous lymphoma vary from creams and ointments to prescriptions, procedures and oral/intravenous or light therapies. In the United States, there are many qualified centers where patients with cutaneous lymphoma can be treated, especially during the early stage of the disease. For patients whose disease has advanced, treatment may fall under the realm of cutaneous lymphoma specialists in dermatology and oncology, cancer center or infusion/chemotherapy clinics.

If you live in a rural area or small town, you may find that it is difficult, if not impossible, to obtain certain treatments. You may need to travel to have your treatments, and there may be costs involved with that. If the costs become too prohibitive, some clinics have individuals appointed to guide patients through the process of finding, affording and implementing the proper treatments. Most major medical centers have provisions for helping people stay for lengths of time to fulfill their treatments.

The Cutaneous Lymphoma Foundation, physicians, nurses, pharmacists and other healthcare providers can be powerful resources full of information and ideas to help you get what you need. There are organizations that specialize in assisting patients with cancer through their challenges with obtaining medications along with financial, employment, insurance and other issues.

You can find many helpful organizations on the Cutaneous Lymphoma Foundation website for up-to-date resources.

## Chapter 10

# CHILDREN AND YOUNG ADULTS WITH CUTANEOUS LYMPHOMA

Cutaneous lymphoma is a rare disease in general. It is even rarer to find it in children. In Europe and the United States, up to 5% of all cases are diagnosed before the age of 20. However, in some parts of the world, such as the Arab populations in the Middle East, children constitute up to 60% of the patients with mycosis fungoides. Prevalence rates differ greatly among geographic regions and populations.

Many researchers believe that mycosis fungoides, in young people in particular, may have a genetic component and may be triggered by environmental factors. In fact, there may be a link between skin type and early incidence of this disease. For example, African Americans are diagnosed with mycosis fungoides before the age of 40 significantly more often than Caucasians.

Despite the infrequent occurrence of this disease in children, the principles of diagnosis remain the same regardless of age. However, there are some special considerations regarding this age group. For example, physicians devote extra attention to determining a course of treatment for younger patients, weighing the benefit of therapy against any potentially long-term complications more prevalent among the pediatric age group than adults. Phototherapy is the preferred treatment for mycosis fungoides, but if a child is very young, they are technically unsuitable for it. However, each treatment plan is individual and all factors, including age, are taken into consideration.

Among children, the distinguishing features of this disease can be misleading, making it ever more difficult to diagnose cutaneous lymphoma in children. Another complicating factor is that the medical community is usually hesitant to perform biopsies on children unless there is absolute indication of the presence of this disease—and biopsy is the best way to firmly diagnose cutaneous lymphoma. For these reasons, diagnosis in children is often delayed much longer than in adults. The prognosis for children is similar to adults.

When children have chronic inflammatory dermatitis, which are ongoing skin conditions that are not classified with a certain diagnosis (an unusual

presentation of psoriasis, eczema or atopic dermatitis), physicians must consider the possibility of mycosis fungoides. The challenges of diagnosing and treating such complex and often ambiguous diseases are magnified by parental concerns, expectations, and a child's own level of understanding. It has only been in the last several years that investigators have truly begun to understand the characteristics of pediatric presentations of cutaneous lymphoma as more data have become available.

Treatment decisions for pediatric patients are difficult to make because younger patients are more sensitive to the adverse effects of certain therapies. Of particular concern, given their young age, pediatric patients have more time to develop and experience the long-term adverse effects of anti-cancer therapies. When treated with oral/topical psoralen plus ultraviolet A (PUVA) or narrowband UVB phototherapy, local radiation therapy, or total skin electron beam therapy, younger patients are inherently subject to greater risk for developing skin cancers because they have a longer expected lifetime ahead of them.

Similarly, pediatric patients in need of systemic chemotherapy may develop leukemia later on in life, and patients taking retinoids such as isotretinoin (Accutane®) are at risk of premature stunting of bone growth. Topical corticosteroids and nitrogen mustard may be among the safer treatments for early-stage disease with less cancer risk. In the end, proper treatment for pediatric cutaneous lymphoma is individualized to the unique occurrence and needs of each patient.

Although rare, some younger adults are diagnosed with cutaneous lymphoma. For patients newly-married or in the dating scene, this can be a devastating diagnosis. For some patients, the hardest part is the physical presentation of the disease—especially if you're in the workplace or actively dating. A facial rash or other obvious outbreak can make you self-conscious. "You feel like you're a broken individual," one patient said, "like you're not whole anymore."

Treat your diagnosis like any other serious medical diagnosis—this is cancer, after all, and if you put it in those terms to colleagues and friends, they may be more understanding. The good news is that with appropriate, effective treatments in the beginning, there is the hope that you'll experience a long remission. Many patients do.

It helps to connect with others in your situation who are experiencing similar symptoms and situations. Connecting with others who have experienced what you're going through can make the journey a lot easier.

**Please refer to our website for additional and updated information related to children and young adults:**

**[clfoundation.org](http://clfoundation.org)**





# **PART 3:**

# **SKIN CARE**

## Chapter 11

# SKIN CARE

Cutaneous lymphoma, as well as some treatments for this disease, can make skin dry, itchy, and scaly. Because cutaneous lymphoma occurs in the skin, with effects that can make the skin irritated, the skin care recommendations for patients are similar to those for patients with other chronic skin conditions such as eczema. Following, you'll find tips to help with your skin care routine.

### MOISTURIZING

Adequate moisturizing is a very important part of keeping skin healthy as it helps thicken the barrier function and keeps our skin feeling comfortable. Keeping skin moisturized and decreasing dryness can alleviate itch too. A simple and effective way to combat dry skin is to frequently apply moisturizers or emollients.

With so many different moisturizers on the market today, it may be difficult to decide which ones to use. Here are some tips to use when comparing products:

- Ointments and creams provide the greatest moisturizing power. They are the best moisturizing products to use because they contain a high content of oil that leads to greater penetration and more staying power.
- Avoid lotions, as they are made with mostly water and little oil. Avoid gels, as they contain alcohol or acetone that can be drying to skin. Avoid moisturizers that contain perfume and dyes.
- Apply moisturizers frequently, at least 2 to 3 times daily, to keep skin from drying.
- You may need to try different products before finding those that work best for you.
- Avoid products with fragrances.

Moisturize the skin while damp because moisturizing lotions and lubricating ointments can trap water against the skin and provide longer relief from dryness. When skin gets dry and flaky, wet the skin with water then apply a lubricating ointment with lanolin such as petroleum jelly or Aquaphor®. Patients often need to lubricate and moisturize skin many times daily. Visit [nationaleczema.org](http://nationaleczema.org) for a list of eczema products approved by the National Eczema Association, which are also recommended for patients with cutaneous lymphoma.

## **DRYNESS**

Dryness accompanies almost all presentations of cutaneous lymphoma. Because of this, dryness (also known as xerosis) is the most common cause of itching, which contributes to a high degree of patient discomfort.

There is, of course, wide variability as to how dryness shows up with cutaneous lymphoma. Some patients have dry patches primarily during colder months when humidity is low. Other patients experience shedding of dry flakes of skin throughout the year as their disease involves more and more of the skin's surface. In patients with extensive skin involvement, especially those with redness (also known as erythroderma), diffuse shedding of skin can occur (also known as desquamation).

This can be concerning for patients. Some feel embarrassed when skin noticeably sheds or flakes in public and at home. These feelings are understandable and completely normal. Patients can cope with these symptoms in a variety of ways, including lubricating the skin with thick layers of ointment-based products such as Vaseline® or petrolatum, which can reduce shedding and scaling for at least 2-5 hours before needing to reapply.

Adding fragrance-free bath oils to bath water and soaking for 10 minutes several times a week can ease flaking and shedding for some patients. Be aware that this can cause skin to be very slippery so patients should be careful when emerging from the tub.

Over-the-counter products that contain lactic acid can help remove dry flakes from scaly skin. Most of these products contain 12% lactic acid in a fragrance-free lotion (an example is AmLactin® lotion and there are generic equivalents) or 10% urea.

## **ITCH**

Most people with cutaneous lymphoma experience itching (also known as pruritus) and often are unsuccessful in finding relief from this chronic, aggravating symptom. Why are there no good treatments available to relieve itch? The reason is that remarkably little was known or investigated about the basic mechanisms that underlie itch until the past several years. The medical community may not have recognized the widespread nature and importance of the problem and the fascination that comes along with understanding itch. That is changing rapidly.

Patients who experience itch will begin scratching the area that's itching, which sends a signal to the brain and back to the skin to initiate an itching frenzy. It can be helpful to apply an ice pack, bag of frozen vegetables, or crushed ice in a Ziploc® bag covered in a paper towel and apply it to the itchy area for as long as 10 minutes. This will “put out the fire” so to speak. Think of itching in this disease like a wildfire—if we can eliminate the instance of itch, we can prevent it from spreading all over the skin.

For most patients with cutaneous lymphoma, itching can range from a minor irritation to a tormenting sensation that can significantly decrease a patient's quality of life. Cutaneous lymphoma-related itching is particularly troubling since it can start small and take over the body. In a study conducted by the late Dr. Marie-France Demierre of the Boston University School of Medicine, 88% of cutaneous lymphoma patients ranked itch among the top causes of distress associated with this disease.

Pruritus is a physiological condition at its root, and appropriate treatment is more likely to be prescribed if the degree of suffering is explained in detail to your physician. When meeting with your doctor, try to describe the severity of itchiness on a scale of 1 to 10, with 1 being little to no itch and 10 being unbearable and preventing smooth daily functioning.

Although a definitive cause for itch has yet to be determined, some treatments are available. A common first-line treatment is antihistamines. Some options include Allegra® and Claritin® for daytime use or Benadryl® and Atarax®, which are more commonly used at night because they have sedative properties. Some of these medications can be obtained over-the-counter while others require a prescription. These medications block redness, swelling, and itch. For most sufferers, itching becomes noticeably greater just prior to falling asleep, so sedative antihistamines can be particularly effective.

There are easy home remedies to alleviate itch too. Soak for 15 minutes in an oatmeal bath. Apply a cold compress to a particularly itchy area to calm irritation and reduce the urge to itch. Apply open-wet dressings which are a simple, safe, and effective way to alleviate itch, as well as decrease redness, burning, and weeping of skin lesions. Ask for detailed instructions about how to apply these dressings when you visit your healthcare provider.

There are other medications that help alleviate itch. Some options are:

- Gabapentin (Neurontin<sup>®</sup>) is an anticonvulsant prescribed for individuals prone to seizures, which has been effective in treating itch.
- Mirtazapine (Remeron<sup>®</sup>) is an antidepressant prescribed for individuals at bedtime who have difficulty sleeping due to itching.
- Aprepitant (Emend<sup>®</sup>) is a prescription medication used for preventing chemotherapy-induced nausea and vomiting, which has been shown to be effective in reducing itch.
- Phototherapy is a viable treatment option for patients with mycosis fungoides, especially those suffering with itch.
- Topical steroids may be effective when used in conjunction with other treatment methods, but tend to be impractical in treating severe cases due to the large surface area.
- The most effective over-the-counter lotions for less severe cases are those that contain pramoxine, an anesthetic that reduces the transmission of the itch sensation from skin nerves, or menthol as its cooling properties can overpower the itching sensation. Be cautious of using analgesics, as they have been shown to aggravate itching episodes.

In addition, some relief of itch has been reported through alternative methods such as acupuncture and biofeedback.

## **FISSURES**

A fissure is a straight or linear crack in the skin, which often extends into the second layer of skin, where it causes considerable pain and discomfort. These primarily occur on the palm side of the fingers and hands. These fissures can make it difficult to use fingers for fine motor skills like dressing, writing, cooking, or eating. Fissures can happen at any time, to anybody, particularly during winter, and especially in individuals who have thick, scaly skin on their palms because of an illness like cutaneous lymphoma.

Most physicians believe this occurs in patients with scaly skin because a small crack in the skin gets infected with bacteria, particularly staph, and that causes a crack to widen and become more painful. It happens more often in winter than summer because the humid conditions of the summer minimize fissuring by virtue of increased moisture.

Patients can take preventative measures to avoid fissures by keeping thick, scaly hands and feet moisturized frequently throughout the day with fragrance-free creams or a thick layer of petroleum jelly products. This may include

applying a thick layer at bedtime to palms and soles and sleeping with white cotton gloves or cotton socks.

Once fissures form, it's important to clean them at least twice a day with soap and water and apply an antibiotic ointment to cracks or fissures to speed healing. Most dermatologists encourage patients to avoid triple antibiotic ointment (such as Neosporin®) because the neomycin ingredient in Neosporin® can be an allergen that causes a rash. If fissuring does not improve, patients should consult with their healthcare provider for prescription-strength topical antibiotics.

Occasionally, a fissure is so deep and wide that physicians may instruct patients to use superglue to glue the fissure together. If instructed to do so, patients should clean the fissure with soap and water and dispense a tiny droplet of superglue in the fissure, pinching surrounding skin together for 60-90 seconds to help close it. It dries very quickly. However, patients need to be careful not to glue their fingers together.

In addition, consider using New-Skin®, an antibiotic solution containing a light adhesive that also provides a barrier over fissures and a mixture of chemicals that creates a polymer layer which binds to the skin, keeping away dirt and germs and retaining moisture. Look for products named liquid band-aid or the like.

## INFECTION

Skin infections are not uncommon in cutaneous lymphoma patients. Some patients with more extensive skin involvement may find that their skin is colonized with a bacteria called staphylococcus aureus. While some forms of staph bacteria exist normally on our skin, this version is the most common bacteria to infect the skin of patients with cutaneous lymphoma. When they occur, infections usually arise in skin lesions of cutaneous lymphoma. Infection is the greatest concern for cutaneous lymphoma patients because, if left untreated, its consequences can be life-threatening.

With that in mind, it's important to recognize the signs of skin infection and know when to contact your physician. Signs of skin infection may include the following:

- A red area on the skin that is painful, swollen, and may be scabbed over or weeping fluid
- Tender redness surrounding a skin lesion

- A skin lesion that does not itch but hurts
- Lesions that develop a thin, yellowish crust

You'll want to urgently contact your healthcare provider if any of the following occur (which could indicate the beginning of cellulitis or blood infection):

- You develop fever and chills associated with abrupt onset of fatigue and weakness.
- All of your skin lesions suddenly become tender and red, especially with streaks of redness, extending from the lesions toward your trunk (armpits or groin).

## **BATHING AND SHOWERING**

When bathing or showering, be sure to use lukewarm water – not hot – as hot water tends to melt the natural oils from our skin that keep it hydrated and can leave our skin drier than before. Hot water also causes a surge of blood flow to the skin, which can increase itching once patients emerge from the shower or bath. Excessively hot water may temporarily relieve itch, but it is not recommended because it may aggravate skin and worsen itch long-term.

Keep baths and showers brief, no longer than 15 minutes. Also, the best time to apply emollients is after bathing, when skin is still moist. Applying moisturizer (or topical medication in an emollient) on damp skin helps seal hydration into the skin's outer layers.

You may reduce the amount of itching and skin infections with the use of “bleach baths,” which is like turning your bathtub into a swimming pool. Bleach baths entail soaking for 15 minutes in a  $\frac{3}{4}$  full bath tub with warm water and  $\frac{1}{4}$  cup of plain household bleach (fragrance free, sodium hypochlorite 6-8%) three times a week. Bleach baths not only may reduce the risk of infection but they have been shown to be anti-inflammatory.

## **SOAPS**

With regard to soaps, less is better. When patients feel the need to use soap, try a moisturizing soap that contains extra oils. Avoid heavily-fragranced soaps because, when a product is infused with fragrance, it must first be dissolved in alcohol, which is drying to the skin. Fragrances can be irritating as well, and potential allergens. Choose soaps and moisturizers that are labeled fragrance-

free (Dove<sup>®</sup>, Eucerin<sup>®</sup>, Lubriderm<sup>®</sup>, Curel<sup>®</sup>, CeraVe<sup>®</sup> and Cetaphil<sup>®</sup> are some examples). Avoid antibacterial or deodorant soaps as they may be too drying.

## LAUNDRY DETERGENTS

Laundry detergents can affect skin too. Be sure to use a fragrance-free detergent. Also, fabric softeners often have fragrances, so look for one that is fragrance-free. Fabric softener sheets added to the dryer are often the worst product for patients with sensitive skin because the fragrances directly coat clothing, which comes into direct contact with the skin, and can be a constant source of irritation.

## SUN PROTECTION

Too much sun is damaging to skin. Use sunscreen, wear a hat with a brim, and consider wearing long-sleeves and pants year-round. If you receive light therapy as part of your treatment, your healthcare provider may advise you to wear UV-blocking sunglasses. However, for some people with cutaneous lymphoma, abbreviated periods (15-20 minutes) of sunlight may be beneficial for your skin. Discuss this with your healthcare team to determine which protocol is best for you.

## OTHER TIPS

You can keep your skin comfortable by also following these helpful tips:

- Wear loose-fitting, comfortable fabrics that ‘breathe.’ Cotton is best. Avoid rough, scratchy fabrics like wool. Avoid tight-fitting clothing.
- Do not scrub skin or rub harshly.
- Keep fingernails short to prevent infection and skin damage.
- Avoid getting overheated; sweating worsens itch.
- Find ways to manage stress as stress can trigger flare-ups in cutaneous lymphoma and increase itching.



**PART 4:  
LIVING WITH  
CUTANEOUS LYMPHOMA**

## Chapter 12

# OVERVIEW

You have cancer.

Those three words can unleash a riptide of emotions, from disbelief to anger to sadness to fear. All of those emotions and more are normal and can be calmed by asking questions, gaining information, and talking with individuals who have either experienced the disease or understand its normal course of action.

Many patients say they experience denial when they first hear the diagnosis. “What do you mean I have cancer?” Cutaneous lymphoma does not seem like cancer, feel like it, or look like cancer the way we have come to understand it. Other patients feel an intense desire to fix it—find the treatment or medication that can make it go away. The reality is that cutaneous lymphoma is a chronic disease with multiple options for treatment, but no cure – at least not yet.

The purpose of these treatments is to help patients feel better, as the symptoms due to the disease can be uncomfortable, unpleasant and sometimes debilitating. Patients often complain of discomfort, pain, fatigue, depression, itching and insomnia when their disease is not well controlled. During periods of worse disease control, you may find lesions, tumors, scaling or plaques, or other presentations that appear in places that others can see. Patients often become self-conscious when they find skin lesions on noticeable areas like the face, neck or leg.

Over the course of your disease, there may be periods where these symptoms can be intense, but there may also be times where the disease is under good control. Therefore, in patients who have cutaneous lymphomas, the goal of the therapy is for quality of life on therapy to be better than quality of life off of therapy. Balancing your own priorities, and being honest with your doctors about how you are feeling is an important part of shared decision making.

While most patients who have this disease will remain in the stage that they started in, the course, appearance, and symptoms of disease can change over

time. Similarly, the treatments may also change over time. Most patients will change from one treatment to another, and at times even retry treatments they have previously had. Be prepared to try a variety of treatments before finding the one, or a combination of treatments, that works for you at any given point in time. Given assessing quality of life is a central component of shared medical decision making, it is important to work closely with your healthcare team to determine the best course of treatment.

Any chronic disease changes your life significantly, and cutaneous lymphoma is no different. You may face physical, logistical, and emotional hurdles because of your disease. Talking with trusted friends and family, your healthcare team, and others close to you can ease the challenges and make it easier for you to ride the waves of your disease.

In every situation, talk with your healthcare providers about how to manage or ease symptoms. Keeping a diary of your symptoms and feelings can be helpful, and feel free to share this with your healthcare team. Some people find that the cancer itself does not make them sick, but medications or treatments might. Communicate this to your physician so that you can try other treatment methods to alleviate negative side effects. There are ways to control symptoms and side effects and treat infections. Ask your doctor about medications that can help calm these discomforts.

In addition to treating symptoms and discomfort, it is important to continue doing things that bring you happiness within your physical capability. This may include physical activity, exercise, meditation, music or enjoying time with your family.

Cutaneous lymphoma is a disease that can change the way you look. Dry patches, skin redness, plaques, skin ulcers, and tumors can show up anywhere on the body. Chronic itching and discomfort, plus a change in appearance, can alter how you feel about yourself. You may lose interest in socializing, which can lead to feelings of loneliness or depression. That's when it is extra important to talk with someone—whether that be family, friends, a healthcare professional, a licensed counselor or social worker. Talking to others who have also lived with, and experienced, what you're going through can also be helpful. If you find caring for yourself physically difficult, your healthcare provider or insurance company can recommend skilled-care individuals to help with skin care, dressing changes, or to check on your condition.

Ultimately, know that it's ok and normal to have fears, concerns, and feelings of frustration. You do not have to face this disease alone. Gathering supportive friends and family around, and asking others for help, are two ways to ease fears and challenges.

There are many ways to help yourself, including the following:

- Learn as much as you can about your diagnosis and treatment options. Knowledge is empowering and can help you feel more in control of your situation. Ask questions!
- Find a support group. Like-minded individuals who have experienced, or are experiencing, the same things can be great to share feelings with, offer coping tips, and learn more about disease and treatment options. The Cutaneous Lymphoma Foundation is a great resource for finding support.
- Talk with someone one-on-one—a doctor or nurse, counselor, spiritual advisor, close friend or relative.
- Have hope. Some research suggests that hope may actually help the body face challenges and heal. Remember: more people are surviving cancer today than in the past, and many people with cutaneous lymphoma lead active, normal lives, even during treatment.

## Chapter 13

# HOW TO TELL FAMILY AND FRIENDS

*“Telling family and friends that you have cancer is worse than having cancer,” says one patient. “You see their face and know you just ruined their week. They look at you differently, and it also makes them think ‘Hey, if he got it, and he’s always been healthy and active, it could happen to me.’ It makes people consider their own mortality.”*

To make matters more complicated, cutaneous lymphoma is not like other cancers. It is a chronic, live-a-long-time-with-it disease, and most people understand cancer to be dire and often immediately deadly. Additionally, because the disease is on the skin, patients often feel that others cannot see them beyond the disease.

Because this is a rare disease, most of your family and friends may not be aware of it. It can be difficult for patients to explain about their disease repeatedly. Everyone responds differently and some people may become distant or go into denial. Expect to do a fair amount of educating the people around you so they can become sensitive to what you are going through. Encourage them to contact an organization like the Cutaneous Lymphoma Foundation to obtain their own information or speak with someone who can help them gain an understanding of what you are going through.

It can also be difficult for others to understand your physical symptoms, like itchiness. Many patients claim that dealing with a symptom like relentless itching can be frustrating and even a mental struggle—something that you cannot explain to others. You may find that the people who are most supportive are not the ones you would have expected. Feel free to talk to others about your disease, if you’re comfortable. Surround yourself with only those people who help you, not hinder the healing process.

Developing lymphomas in areas where others can see it readily can be particularly difficult. Such symptoms can affect your social life. Some people prefer to remain at home during such outbreaks. This feeling of isolation can be very difficult for some patients.

It can be hard to make plans. Some patients find that, on a good day, they schedule an outing or a get-together with friends but, when the date arrives, their symptoms or treatment render them unable to fulfill the promised activity. The people in your life should be understanding. There will be times that you can keep plans, take vacations or otherwise make good on a scheduled outing, and there will be times when it's just not possible.

Ultimately, it is imperative to surround yourself with people who care about you and try their hardest to understand what you're going through and provide the kind of support you need. Whether it is a local community organization, family group or faith organization, find a place that provides non-judgmental support and rely on those individuals when the going gets tough. Find additional, professional support and assistance if you need to supplement the support from family and friends. A professional psychologist or other type of healthcare provider can assist in working through disturbing feelings and depression.

**Please refer to our website for additional and updated information:**

**[clfoundation.org](http://clfoundation.org)**

## Chapter 14

# DEALING WITH FATIGUE

People in the modern world are tired, and that tiredness is often more intense for people living with cancer and other chronic illnesses. Fatigue can range from daytime drowsiness to feeling too exhausted to get out of bed. It has been identified as one of the main symptoms that negatively impact a patient's quality of life in multiple surveys, including the Lymphoma Coalition's Global Patient Surveys. In the most basic of descriptions, fatigue makes it hard to enjoy life. Given that cutaneous lymphomas are primarily chronic conditions, it follows that fatigue may be a long-term problem that many people diagnosed with a cutaneous lymphoma face.

Multiple factors contribute to fatigue; some common causes include:

- poor sleep
- the severity of illness - sometimes called “disease burden”
- pain
- chemotherapy
- radiation therapy
- certain medications
- diet
- thyroid dysfunction
- low blood counts causing anemia
- mental health issues such as depression

The Cutaneous Lymphoma Foundation defines fatigue as a decreased capacity for activity, often accompanied by feelings of weariness, sleepiness, or irritability. This is not the usual fatigue you might feel after physically working very hard all day long. This kind of fatigue doesn't improve after getting a full night's sleep and it interferes with how you function in your daily life. Naturally, this level of fatigue is upsetting and definitely can impact a person's quality of life. Historically, common advice given to cancer patients, in dealing with fatigue, includes anticipating fatigue and pacing yourself, building rest periods into your day as needed, and adjusting your work schedule wherever possible. These suggestions remain valuable in coping with fatigue. However,

other strategies are now recommended for dealing with fatigue in addition to the above tried and true methods.

## **ACTIVITY TO FIGHT FATIGUE**

While exercising when feeling fatigued might seem like bad advice, there is increasing evidence that it is reasonable and even good advice. For example, according to the American Association for Cancer Research, the evidence continues to mount that those who suffer from fatigue and engage in exercise, experience numerous physical and mental health benefits, including increased activity tolerance, improved quality of life, and decreased depression and anxiety.

Adding to this idea, the American College of Sports Medicine (ACSM) Roundtables found that cancer survivors could safely engage in enough exercise training to improve physical fitness and reduce fatigue. For cancer survivors, their other recommendations included avoiding inactivity and engaging in moderate intensity aerobic training three times per week. Doing this can decrease cancer-related fatigue, both during and after treatment. The ACSM encourages cancer healthcare providers, as a standard of care, to “Assess, Advise, and Refer.” This means connecting cancer survivors to the most appropriate exercise programming. You can find the programming registry, along with other helpful resources, at [exerciseismedicine.org/movethruca](http://exerciseismedicine.org/movethruca).

## **OTHER CAUSES OF FATIGUE FOR CUTANEOUS LYMPHOMA PATIENTS**

Fatigue can come from multiple sources, some of which have been listed at the beginning of this chapter. Poor sleep, high and/or prolonged stress levels, medications and treatments, and other mental health/medical conditions all can contribute to fatigue. That is why it’s so important to discuss your fatigue with your healthcare providers so that all possible sources of fatigue are sorted out. Hopefully a plan of action is developed to deal with your fatigue that takes care of cancer-related fatigue or fatigue coming from other causes.

## **WHAT ELSE CAN BE DONE FOR CANCER-RELATED FATIGUE?**

The National Comprehensive Cancer Network (NCCN) guide for patients with cancer fatigue includes some other helpful suggestions. In addition to searching for and treating the cause of the fatigue when possible, the NCCN guidelines also recommend a team-based approach that brings in other specialists to help you:



- Physical and/or rehabilitative therapy can help with an exercise plan or think of ways to help you overcome other challenges you might face in daily life.
- Mental health care by professionals can assess and provide help with depression or anxiety or other symptoms associated with your cancer treatments. Talking with a therapist or attending support groups with other patients can also help.
- Palliative care addresses ways to help improve your quality of life during cancer treatment. Examples of this would be helping you with cancer pain and fatigue.
- Integrative medicine specialists can help by providing additional therapies along with your prescribed cancer treatment. These therapies might include things such as yoga, meditation, behavioral therapy, acupuncture, massage, guided relaxation and mindfulness techniques.

## WHAT CAN YOU DO?

The most important thing you can do is talk about your fatigue to the healthcare team responsible for your care. Don't assume that it isn't important or that it is to be expected because you have a cancer diagnosis and are undergoing treatment. Most importantly, don't assume that nothing can be done about it. When talking with your providers, try to rate your fatigue, just as pain is rated, on a 0-10 scale where 0 would be no fatigue and 10 is the worst fatigue you can possibly imagine. Ask about any other physical conditions that might be adding to, or causing, your fatigue and if there are treatments that could specifically address those problems. Finding emotional support from other patients with a similar diagnosis can often be helpful. To this end, you might consider joining a Cutaneous Lymphoma Foundation online networking group, attending an in-person education/support event or participating in any other programs offered on a local, regional, or national level that provide exposure to experts, other patients, and helpful information. Your goal is to decrease your fatigue and improve your quality of life in the process.

*Sources referenced in addition to those mentioned:*

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## Chapter 15

# SEXUALITY

Being intimate can be challenging when you have cutaneous lymphoma for the simple reason that plaques, lesions and rashes can be so uncomfortable or sensitive that you don't want to be touched. Also, sometimes people with cutaneous lymphoma feel self-conscious or uninterested in being intimate because of changes to their physical appearance. Some symptoms make the skin feel raw and a physical experience would be aggravating. From itch to burning to lathering up with ointment after a shower, the details of this disease can surely create an obstacle to intimacy.

Couples work through this. There are times when you just won't be sexual, and hopefully your partner understands this. Other times, when there aren't flare-ups, you'll likely feel as sexual as you normally would.

It is important to feel supported when you're not feeling your best. One patient noted that, "When you have a disease, any kind of terrible disease, you need a partner who's very, very understanding. It's frustrating, from the care side, to not know what you can do to make the person you love feel better."

Communication is key throughout the course of this disease. Letting your partner know when it's ok to touch and when it's not, explaining gently that it's not a rejection of the other person, but a symptom of the condition of your skin, is important. As one patient says, "You have to tell your partner, 'I love you with all my heart, but I can't explain what I'm going through' and I'm sure the partner will understand, knowing that you love each other."

## FERTILITY

Cutaneous lymphomas are most often chronic illnesses. Because they are chronic illnesses, treatments are often given continuously over many months to years. Both doctors and patients look for mild, safe, well tolerated, and effective therapies that can treat and control the lymphoma for many months, and often years. In fact, when studying new treatments, doctors consider both duration of response (how long the treatment is effective for) and how many people get better to decide if a new treatment is effective.

When it comes to family planning, using even mild therapies for a long period of time can pose challenges. Some therapies are not considered to be safe during pregnancy or for conceiving a child, as they can lead to complications in the fetus or the pregnant woman. Additionally, most therapies are not studied in pregnant women. Much of our knowledge comes from studies in animals, understanding how drugs work, or a few cases where patients conceived or became pregnant while on one of these therapies. As there is limited information and each patient's treatment needs are unique, please see the "Cutaneous Lymphoma: Effect of Treatment on Reproductive Health and Implications for Patient Education" chart available for download online on the CL Foundation's website. The chart is for general information to start a discussion with your treating physician and not as a specific recommendation for your care.

Therefore, talking about family planning with your healthcare team is important. This includes fertility (the ability or potential to become pregnant or get someone pregnant), conception (the actual process of getting pregnant and getting someone pregnant), pregnancy, and breastfeeding after pregnancy.

**Please refer to our website for the "Cutaneous Lymphoma: Effect of Treatment on Reproductive Health and Implications for Patient Education" chart:**

**[clfoundation.org/fertility](http://clfoundation.org/fertility)**

Integrative medicine is an approach to healthcare that utilizes all available evidence-based treatments and therapies, including traditional Western medicine to complementary and alternative methods including acupuncture, herbal supplements, nutrition, lifestyle, meditation, yoga, and more. The combination of these modalities, and a focus on the therapeutic relationship that is patient-centered and inclusive of the physical, mental, emotional, and environmental influences, provides for a more comprehensive approach to addressing patients with chronic diseases including cancer. The integrative approach to cancer is intended to support the immune system, improve overall health status, and provide options that assist patients cope with cancer, treatments, and treatment side effects. These treatment options are not recommended to replace traditional medical care; however, when used in conjunction with medical care, they can provide valuable support and enhance quality of life and general recovery.

### **TAKING CARE OF THE WHOLE PERSON: BODY, MIND AND SPIRIT**

Today, the definition of a cancer survivor has dramatically changed. The Institute of Medicine defines a survivor as: *“Any individual from the point of their cancer diagnosis moving forward is a SURVIVOR.”* The goal is to help individuals not only SURVIVE with their cancer but to live their life as full as possible. The goal of providers is to help patients achieve their goals throughout their cancer journey.

Therefore, it is essential to recognize and help reduce the emotional stress associated with a cancer diagnosis in order to improve quality of life for patients and their loved ones. Patients and families are often so focused on “making it” through a course of treatment that they often overlook the need for self-care and emotional support—important aspects of life that we now know are equally important in helping patients get well.

It is imperative to assess how a cancer diagnosis affects the “whole” person. What happens to a patient on every level—physical, behavioral, emotional, cognitive? Integrative medicine modalities can help achieve that ever-important balance of mind-body wellness.

In 2007, the Institute of Medicine published a report entitled “Cancer Care for the Whole Person: Addressing the Psychosocial Health Needs of Cancer Patients.” This report reveals how important it is to ensure a person’s psychosocial health, and the impact psychological well-being plays in the course of cancer. Cancer doesn’t occur just to the patient; cancer affects every member of a family.

The following mind-body techniques have proven helpful for patients and family members when facing a major life stress such as cancer:

## **STRESS MANAGEMENT**

There are many effective ways to manage stress. These include physical exercise, medication, mindfulness exercises, doing activities that find meaning or joy, and reading or listening to materials that also decrease your stress.

Keep in mind some general thoughts as you develop coping mechanisms to manage stress that can work for you. Your cancer treatments do not need to overtake you and your life. Feel free to stick to your routines, and organize your schedule in a way that best meets your needs. Observe and respect the natural limits of your body. If you are tired—rest!

Review your normal routine and learn what makes you tick. What stresses you out? What soothes you? Understanding yourself and having realistic expectations of your nature will guide you in determining effective coping methods. Look at your past efforts to manage stress and consider what has or has not worked. Know your strengths and exploit them. Ask for help and take suggestions. You may opt to see a professional counselor or speak to your health care provider about managing stress. In this disease, the goal of treatment is meant to allow patients to live a normal, quality life.

## **MANAGING ANXIETY**

Anxiety can be one of the toughest emotions for patients and loved ones. Anxiety often peaks around the time of doctor visits, with the start of new treatments or when new symptoms arise. This is completely normal. Sometimes anxiety fades; sometimes it does not. A recent study of anxiety found that more than one-third of patients in remission reported levels of clinical anxiety about the possibility of relapse. It’s normal to be concerned about your disease.

The best way to manage anxiety is to identify when it is at its highest for you. Then, there are numerous techniques to manage it. You do not need to manage

your anxiety alone. Feel free to seek support from a mental health professional. Remember that there are various healthy and effective ways to manage support using behavioral modification. Some find it helpful to talk with others who have cutaneous lymphoma or have battled it in the past and who share other characteristics with you—age, geography, family situation, etc.

People with indolent (slow-growing) lymphomas learn to adapt to the cycle of starting active treatment, finishing treatment, wondering if the treatment worked, re-establishing a life routine and then wondering when the disease will return. You will discover your own unique adaptation. If you are newly-diagnosed, take solace in the fact that others have been through your experience.

Oftentimes the unknown and uncertainty about the future fuels a significant amount of anxiety. A study of young adults with relapsed/refractory lymphoma in their twenties revealed that those with indolent disease adapt to “living in the gray” by finding a balance between knowing lymphoma will return at some point, and not letting worrying thoughts about “when” affect daily life. Many patients note their anxiety improves as they can see themselves living with this disease and especially when they feel they are less limited by it.

## RELAXATION TECHNIQUES

Relaxation techniques offer strategies to help manage how we respond to stressors. One technique includes basic diaphragmatic or belly breathing—breathing fully into your lungs and expanding your belly. Belly breathing essentially shuts off the fight or flight response and triggers a relaxation response. The goal is to learn to recognize stress-induced rapid breathing and instead take deep breaths to create calm.

## GUIDED IMAGERY

Imagine lying on a beach under the warm sun, a cool breeze caressing your face. You are listening to the ocean lap at the shore. Imagine that you are sitting in front of a fire on a cool autumn night. The fire crackles and sparks. You stare at no particular point but rather to the flickering orange flames. The night sky is filled with sparkly stars. All is quiet.

Using guided imagery, or visualizing images like these, may cause a person to feel less distressed, taking their minds away from a stressful situation. When dealing with a cancer diagnosis, our minds may race with conflicting, scary, unanswerable thoughts. Some patients find that using guided imagery helps to ground them.

## **MEDICAL HYPNOSIS**

Medical hypnosis is defined as a state of focused awareness. This is simply a relaxed state where individuals “put to sleep” thoughts from their conscious mind and tap into a deeper level of consciousness to increase focus. Similar to relaxation and guided imagery, in medical hypnosis a patient hears a set of suggestions that have meaning for that individual, transporting them into a calmer state of being. This should be performed by a licensed professional.

## **MINDFULNESS**

Modern Western psychology has many definitions of mindfulness including “a psychological quality that involves bringing one’s complete attention to the present experience on a moment-to-moment basis.” In a fast-paced world, with scary news like a cancer diagnosis, it becomes easy to become preoccupied with what the future may hold or what the next test, scan, or treatment will reveal. Mindfulness allows us to be present and disseminate random, scary, unknowable thoughts coursing through the mind. There are many books, podcasts, and programs that focus on mindfulness. Studies of mindfulness techniques in other cancers have been proven to improve stress, and even cognitive changes that can occur with chemotherapy.

## **YOGA**

Yoga can provide both a restoring physical activity as well as mindfulness. Practicing yoga during cancer treatments can resolve insomnia, improve mood, and enhance quality of life. Additional benefits include enhanced breathing, improved appetite and bowel habits, increased sense of peace and tranquility, and fewer side effects. A recent study found that lymphoma patients who practiced yoga had improved sleep quality and decreased use of sleep medications.

## **FOCUS ON WELLNESS**

Remember that health is much more than the absence of disease! You can have lymphoma and still focus on being healthy and active. Many patients don’t realize that they can alter the course of their disease, the intensity of their symptoms, their response to treatment, their experience of side-effects, and their emotional state by focusing on wellness. Through nutrition, lifestyle, and complementary and alternative therapies, many aspects of one’s health can be supported. An integrative practitioner can help to identify the best ways to support you!

Today there are many options for adding to your care and support system while living with cutaneous lymphoma. There are some free programs available to

patients with cancer to improve their physical wellbeing as well. Check out your local gym, yoga studio or holistic health center and try a new class. See what works for you and incorporate that into your overall long-term healthy living program.

**Please refer to our website for additional and updated information:**

**[cflfoundation.org /integrative\\_medicine](https://cflfoundation.org/integrative_medicine)**



**LIVING WITH  
CUTANEOUS LYMPHOMA:  
PATIENT AND  
CARE PARTNER STORIES**

## Continuing My Journey

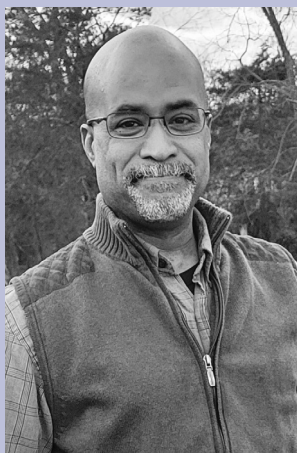
My story with cutaneous lymphoma is one of hope. Like so many, I didn't receive a definitive diagnosis immediately but underwent continued surveillance and blood tests to rule out other conditions. I didn't have symptoms like itching, but instead, non-raised patches along my arms, so I was prescribed topical creams and UVB treatment. At 43 years of age, I was informed that I had suspicion for mycosis fungoides and it was indolent in terms of its trajectory.

I am grateful to have a UVB lamp for treating my MF, but it can be hard to stay on the regimen. I set an intention every day to make things work for me so that I can create my life from within it. To help make the treatments seem less mundane, I will meditate or do box breathing to relax. I may stretch or move around rather than standing still. I also listen to podcasts that are uplifting to relax my mind.



Joining the Cutaneous Lymphoma community has allowed me to share what is going on with my life and hearing other people's stories uplifts my spirit. I can also be there for others when they are not feeling so well in a certain phase of their life. I am now 54 years old and continuing this journey, not knowing where it will lead me. Following up with my appointments with my doctor is key and although there is no cure for mycosis fungoides at this point in time, I find ease in knowing I am not alone.

**Karla R.  
Patient**



## Two Are Better Than One!

In 2018, after my wife was diagnosed, our physician gave us a copy of the Cutaneous Lymphoma Foundation's (CL Foundation) guidebook, *A Patient's Guide to Understanding Cutaneous Lymphoma*. This book has been a tremendous help to us, as it provides information essential to making informed decisions, communicating effectively, and taking appropriate actions.

As husband and caregiver, the guide facilitates my intense need to help (i.e., to do something!). It tempered my initial fears, and the section on treatment options has been an invaluable resource, serving as both an educational starting point and an ongoing reference for understanding and participating in treatment decision-making processes. I've assisted in asking relevant questions, weighing treatment-option pros and cons, and influencing treatment-related decisions.

However, the CL Foundation's role in helping us navigate this disease far exceeds the Patient's Guide. Its website is a cornucopia of beneficial content, and we thank God for the dedicated staff and volunteers who make it possible.

I highly recommend attending the Foundation's patient/caregiver online meetings, as they provide a warm, safe environment where we mutually share our challenges and insights while receiving comfort and encouragement. The facilitators, participants, and caregivers are all kind and welcoming, and especially comforting to attendees who are newly diagnosed and feeling frightened.

To my fellow caregivers, it is critically important to view your loved one's cutaneous lymphoma (CL) as if you both have the disease. From day one, I have viewed my wife's CL as "our" disease, and we've navigated the journey together. This includes attending all appointments and treatments, asking questions, and monitoring and reporting noticeable skin issues that she may not see. This mindset—this pragmatic expression of love—encourages her, lightens her burden, and mitigates my sense of helplessness.

**Tom G.**  
**Husband of Patient**

## The "C" Word That Wasn't The End!

I was about 16 when I first noticed the round dark circle on the top of my left arm that didn't seem to go away. Over the years, I tried different things to remove the marks but nothing worked. I went through the whole rigmarole of playing “guess what I’ve got” with my doctor. I kept asking, “Are you sure it’s not a skin cancer?”

In 2005, I finally received a referral for a second opinion from my local doctor in Staffordshire, England. The Dermatology Consultant was baffled. She asked for my permission to have a few other doctors look at my skin - who, excited by what they saw, took biopsies from a few areas of light and dark skin.

About a week later, I was informed they finally had a diagnosis for me, mycosis fungoides, but they didn’t know how to treat it. I was gobsmacked, shocked and scared. The Consultant gave me a piece of paper with the CL Foundation’s website so I could get in touch with other sufferers for support and advice!

Joining the Foundation’s online networking group was one of the best things I ever did. Another member suggested I ask my doctor to refer me to Guy’s and St Thomas’ Hospital in London. I was referred to a Consultant who examined me and arranged for me to start PUVA treatment in my local hospital in Staffordshire. I insisted that I stay with this Consultant, as I felt safe and knew he was fully aware of my disease.

When I was first diagnosed I thought I was going to die straight away or in a few years, so I started to put my life in order. I asked my girlfriend, now my wife, to marry me and made sure everything was in order in case anything happened to me. Now 19 years after my diagnosis, I’m still being treated at the hospital in London for annual checkups and treatment plans.

As I’m a night owl, I jump on the laptop at midnight to attend the networking group meetings. It has been a great source of comfort and support for my mental health and well-being. I can’t tell you how great it is to speak to people who know what I’m going through, even though we’re miles apart. I’ve found out so much information and am able to share my experiences and knowledge with any newbies too.



**Ronni B.**  
**Patient**

# **PART 5:**

# **CLINICAL TRIALS**

## Chapter 17

# CLINICAL TRIALS

A clinical trial is a research study designed to answer basic and clinically relevant questions about a new treatment or a new way of using an old treatment. A clinical trial provides information or data by which treatments can be measured—how effective they are (efficacy), how long they are effective before the disease returns or gets worse (time to progression), and how safe they are (side effects).

Clinical trials are incredibly important for determining new and more effective treatments. They require voluntary participation and each participant must be fully informed about the details of the clinical trial before enrollment. To be fully informed about the clinical trial, an informed consent process must take place between the patient and the study doctor. This requires a thorough review of the consent form with the patient, study staff, and study doctor prior to signing the consent form or undergoing any study procedures. This helps to ensure that the patient and their family understand what is involved and required for their participation, including any travel or financial obligations the trial may require. The consent process should take as long as is needed for you to feel confident, including the option to take the consent document home to read thoroughly and formulate any questions you may have. Make sure all of your questions are answered before signing the consent document.

Safeguards are required to ensure that each clinical trial is scientifically valid, has fair subject selection, a favorable risk-benefit ratio, and ensures the respect and safety of all those enrolled. The clinical trial is reviewed and approved by an independent board, called an Institutional Review Board, to ensure all of these safeguards are in place before the trial can begin at any specific hospital, clinic, or center.

While participating in a clinical trial can be time-consuming, the results can be potentially groundbreaking. Many of the cancer treatments we take for granted today were, at some point, studied in a clinical trial. Patients and physicians must discuss and evaluate all associated factors before deciding a course of action, as there are always risks associated with any form of treatment. Patients typically choose to participate in clinical trials because they are looking for

availability and access to new treatments that may be effective for them when other treatment methods have proved unsuccessful or limiting.

The medications used to treat cutaneous lymphoma are evaluated in three ways: formal clinical trials, years of post-marketing experience, and case studies. In medical literature, over the last decade, evidence-based criteria have been used for evaluating the quality and impact of published data that demonstrates a particular treatment's effectiveness. This information is published and made available for use by doctors, researchers, and patients.

The best evidence for evaluating whether a treatment is effective for cutaneous lymphoma is a controlled clinical trial in which a new drug is directly compared to another, previously FDA-approved drug (the control). The best situation is when it is a double-blind trial, where the patient as well as the physician does not know what drug is being administered, and, at the end of the trial, data are evaluated to determine each drug's effectiveness. More common in cancer medicine are so-called open-label trials, where everyone is aware of which medication is being administered.

In addition to clinical trials, there are case studies. A case study is a publication where doctors review their records retrospectively over a course of many years to determine the numbers of patients with a certain disease and evaluate what treatments have been effective. The results are published with a general estimate of the treatment's success rate and safety. The problem with retrospective case studies is that, since the information was not collected in real time going forward, data may be missing or inaccurate. Therefore, the quality of the evidence from case studies is much inferior to that obtained in prospective clinical trials, and conclusions about treatment efficacy and safety should always be confirmed prospectively.

Scientific advances and education are important cornerstones to continuing the progress in finding effective new treatments for cutaneous lymphoma, as well as for continuing to monitor existing therapies or new combinations of therapies that can move us closer to finding a cure. Patients can contribute to these efforts by being open to exploring participation in a clinical trial with their physicians. By taking an active role and being a voice in support of continued governmental or private funding for new treatments and research for cutaneous lymphoma, patients can have a big impact on the future of funding new developments leading to a potential cure one day.

## WHAT EXACTLY IS A CLINICAL TRIAL?

A cancer clinical trial is a carefully controlled research study conducted by doctors to improve the care and treatment of people who have cancer. A treatment that is proven safe and effective in a cancer clinical trial is often approved by the U.S. Food and Drug Administration (FDA) for use as a standard treatment if it meets one or both of the following criteria:

- It's more effective than the current standard treatment.
- It has fewer side effects than the current standard treatment.

The purpose of cancer clinical trials is to:

- Improve treatment options
- Increase survival
- Improve quality of life

Advances in the treatment for cutaneous lymphoma depend on clinical trials of new therapies or new therapy combinations. Different types of cancer clinical trials are designed to develop and test new and better ways to:

- Diagnose and treat cancer in people
- Prevent or relieve treatment side effects
- Help prevent a return of cancer
- Improve comfort and quality of life for people with cancer

## WHO'S ON THE CLINICAL TRIAL TEAM?

A clinical trial team is made up of doctors, nurses, social workers and other healthcare professionals. The team members:

- Check each participant's health at the beginning of the trial
- Give specific instructions for taking part in the trial
- Monitor each participant's health throughout the trial
- In some cases, follow up with patients after the trial is over

Your regular doctor or healthcare provider coordinates with the research team to ensure that other ongoing drugs or treatments you're receiving won't interfere with the study treatment.

*Source: The Leukemia & Lymphoma Society; lls.org*



## Phases of a Clinical Trial

### Phase 1

Phase I involves administering a new drug to a small group of patients to determine the side effects and best dosage of the new drug.



### Phase 2

Phase II works with a slightly larger group of patients to test the effectiveness of the drug.



### Phase 3

Phase III is an even larger trial that usually involves comparing the new treatment to a traditional or current standard treatment.



If the results are favorable for the new drug, they may be presented to the FDA for approval of the treatment; and...

### Phase 4

Phase IV, or post marketing studies, assess information such as the drug's risks, benefits and optimal use.

## QUESTIONS TO ASK YOUR HEALTHCARE PROVIDER

- Is a clinical trial right for me? What are my options?
- Where does the clinical trial “fit” into the standard skin-directed and systemic therapies?
- Will I have to stop any of my treatments to participate in this trial? Are there risks to stopping some or all of my current treatments?
- What will change with my skin care in order to participate in this trial?
- Will my healthcare provider team be directly involved in the care during the clinical trial period?
- Will there be an opportunity to continue clinical trial agents after the study ends?
- What if I need to withdraw from the clinical trial?
- What are the requirements related to logistics, location and the visits I will need to make?
- What are the risks and benefits of this trial?
- Are there any anticipated side effects?
- What are the costs associated with the clinical trial?
- How do I make sure my insurance covers these costs?
- Will there be any anticipated costs that my insurance will not cover?
- What kind of care partner support will be required?

**Please refer to our website to find available clinical trials:**

**[clfoundation.org/directory](http://clfoundation.org/directory)**

# **GLOSSARY OF MEDICAL TERMS**

**Aggressive lymphomas**

Lymphomas that are fast-growing and generally need to be treated immediately: typically considered intermediate-grade or high-grade lymphomas.

**Alemtuzumab**

A monoclonal antibody directed against CD52, an antigen (or marker) found on both B and T lymphocytes. The drug is used most often to treat chronic lymphocytic leukemia and has been used in the treatment of advanced CTCL. (Campath<sup>®</sup> is the brand name for alemtuzumab.)

**Allogeneic transplant**

A procedure in which a patient receives bone marrow or stem cells donated by another person.

**Alopecia**

Hair loss. Alopecia from systemic chemotherapy is almost always temporary; hair grows back when therapy is finished.

**Anemia**

A shortage of red blood cells, causing weakness and fatigue.

**Angiogenesis**

The process of developing new blood vessels.

**Antiangiogenesis therapies**

Drugs that prevent tumors from developing new blood vessels, thereby stopping or limiting tumor growth.

**Antibody**

A complex protein made by B-lymphocytes that reacts with antigens on toxins, bacteria and some cancer cells and either kills or marks them for removal.

**Antiemetic**

A drug that reduces or prevents nausea and vomiting.

## **Antigen**

Identifying proteins located on the surface of all cells. The immune system uses antigens to determine whether cells are a necessary part of the body or need to be destroyed.

## **Apheresis**

Process of separation of components of whole blood (white blood cells, red blood cells, platelets, stem cells, plasma) performed in a specialized apparatus.

## **Autologous transplant**

A type of bone marrow or stem cell transplantation in which a patient receives his or her own cells.

## **BCNU**

A chemotherapy agent that is used topically in CTCL. (Also known as carmustine.)

## **Bexarotene**

Medications in both capsule and gel forms that have been shown to be effective in treating CTCL. (Targretin® [tar-GRET-in] is a name brand of bexarotene [beks-AIR-oh-teen].)

## **Biologic therapy**

Treatment that uses or stimulates the immune system in directing a response against an infection or disease.

## **Biomarker**

A compound (usually a protein) used to measure the presence of a disease.

## **Biopsy**

Removal of tissue for evaluation under a microscope for diagnostic purposes.

## **Bone marrow**

Spongy material found inside the bones containing stem cells that develop into three types of cells: red blood cells that deliver oxygen to the body and take away carbon dioxide; white blood cells that protect the body from infection; and platelets that help the blood to clot.

**Campath®**

A monoclonal antibody directed against CD52, a antigen (or marker) found on both B- and T- lymphocytes. The drug is used most often to treat chronic lymphocytic leukemia and has been used in the treatment of advanced CTCL. (Campath® is the brand name for alemtuzumab.)

**Cancer**

Abnormal cell growth that cannot be controlled by the body's natural defenses. Cancerous cells can grow and eventually form tumors.

**Carmustine**

A chemotherapy agent that is used topically in CTCL. (Also known as BCNU.)

**Catheter (Intravenous access)**

A device that is temporarily or permanently placed into a vein that makes it easier to give medications.

**Chemotherapy**

Treatment with drugs to stop the growth of rapidly dividing cancer cells, including lymphoma cells.

**Chemotherapy cycle**

Term used to describe the process in which chemotherapy is given, followed by a period of rest in which the body is allowed to recover.

**Chemotherapy regimen**

Combinations of anticancer drugs given at a certain dose in a specific sequence according to a strict schedule.

**Clinical trial**

A research study in which a new treatment is given to patients to determine whether it is safe, more effective or less toxic than current therapies. Clinical trials are an important part of the process of understanding diseases and have been instrumental in providing information to the Food and Drug Administration for approval of new therapies.

### **Combination chemotherapy**

Several drugs given together to increase response rate of certain tumors.

### **Complete remission (CR)**

Term used when all signs of disease have disappeared after treatment.

### **CT or CAT (computerized axial tomography) scan**

This imaging test provides a series of detailed pictures of the inside of the body using an X-ray machine linked to a computer.

### **Cutaneous T-cell lymphoma (CTCL)**

A general term for many lymphomas of the skin including mycosis fungoides, Sézary syndrome, lymphomatoid papulosis, cutaneous anaplastic large cell lymphoma, adult T-cell leukemia/lymphoma, peripheral T-cell lymphoma, lymphomatoid granulomatosis, granulomatous slack skin disease, and pagetoid reticulosis, to name a few. All cases of mycosis fungoides are CTCL, but not all CTCLs are mycosis fungoides.

### **Dermatologist**

Physician who specializes in the diagnosis and treatment of skin diseases.

### **Disease progression**

The terms used if the disease worsens despite treatment (also called treatment failure).

### **DNA**

Abbreviation for deoxyribonucleic acid, an essential component of genes.

### **Dose intensity**

A term used to describe giving the highest possible doses of drugs over a specific period of time with acceptable side effects.

### **Durable remission**

When a complete response lasts for years.

### **Electron beam therapy**

A form of radiation therapy that only treats the superficial portions of the skin. It is highly effective in clearing all forms of lesions of CTCL from the skin. It

can be used to treat portions of the skin or the entire skin surface. When used to treat all of the skin it is referred to as total skin electron beam (TSEB) therapy.

### **Fatigue**

A decreased capacity for activity that is often accompanied by feelings of weariness, sleepiness or irritability.

### **Generalized disease**

A cancer that has spread throughout the body.

### **Genes**

The basic building blocks of heredity that, are present in all cells. Genes are comprised of DNA and other materials.

### **Grade**

A method of classifying a tumor on the basis of how aggressively it is growing.

### **Graft versus host disease (GVHD)**

Occurs when a donor's bone marrow (graft) recognizes the recipient of the marrow (host) as foreign. In response, the immune cells in the donor marrow attack the cells in the host. This is a side effect of bone marrow transplantation.

### **Harvesting**

A procedure in which stem cells are obtained from the blood or bone marrow for use in repopulating the body's cells after high-dose chemotherapy.

### **Hematologist**

A physician who specializes in treating diseases of the blood and blood-forming tissues.

### **Histology**

The study of tissue characteristics that may lead to identifying a specific type of tumor.

### **Idiotype**

A unique "fingerprint" portion of an antibody present on the surface of B-cells.



### **Idiotype vaccine**

A lymphoma vaccine that is custom made to attack an individual patient's lymphoma and contains an idiotype (unique) tumor materials and an immune stimulant.

### **Immune system**

One of the body's defense mechanisms involved in fighting infections and recognizing foreign tissues. All CTCLs and lymphomas are diseases of the immune system.

### **Immunological tests**

Blood tests that detect the presence of diagnostic proteins or antigens on a tumor.

### **Immunotherapy**

See biologic therapy.

### **Indolent lymphoma**

Lymphoma that is slow-growing and has few symptoms. Also called low-grade lymphoma.

### **Interferon**

A systemic therapy that has been shown to be very effective in treating CTCL. (Intron<sup>®</sup> and Roferon are name brands of interferon.)

### **Interferons**

Naturally occurring compounds that stimulate the immune system in infections and inflammation. Synthetic forms are used to treat viral infections, autoimmune diseases and cancers.

### **Intron<sup>®</sup>A**

A systemic therapy that has been shown to be very effective in treating CTCL. (Intron<sup>®</sup> is a name brand of interferon.)

### **Lactate dehydrogenase (LDH)**

An enzyme measured in the blood and used as a biomarker to measure the extent or spread of cancers.

**Leukopenia**

An abnormally low level of circulating white blood cells resulting in the inability to fight infections.

**Local therapy**

A therapy that is directed to specific and limited areas.

**Localized disease**

A cancer that is only present in a limited part of the body — for example, the neck or armpits.

**Low-grade lymphoma**

Lymphoma that grows slowly and has few symptoms. Also called indolent lymphoma.

**Lymph**

The watery fluid in the lymph system that contains white blood cells (lymphocytes).

**Lymph node**

Small bean-shaped glands located in the small vessels of the lymphatic system. Thousands are located throughout the body with clusters of them in the neck, under the arms, the chest, abdomen and groin. Lymph nodes filter lymph fluid, trapping and destroying potentially harmful bacteria and viruses.

**Lymphatic system**

The channels, tissues and organs that store and carry lymphocytes that fight infection and other diseases.

**Lymphocyte**

A type of white blood cell. Lymphocytes, carried along by the lymph fluid, are part of the immune system and fight infection.

## **Lymphoma**

A cancer of lymphocytes involving lymph nodes, organs and tissues of the lymphatic system (immune system). Hodgkin lymphoma is one type of lymphoma; the other major type is non-Hodgkin lymphoma. Cutaneous T-cell lymphoma is a non-Hodgkin lymphoma that starts in the skin. There are approximately 61 types of non-Hodgkin lymphoma.

## **Lymphomatoid Papulosis (LyP)**

A lymphatic system disorder that manifests itself in self-healing nodules and papules ("bumps" and "spots") that come and go spontaneously. It looks like cancer under the microscope, but frequently and for no reason, it suddenly disappears of its own accord for weeks or months at a time before reactivating.

## **Malignant**

Cancerous—a malignant tumor is a cancerous tumor.

## **Matrex<sup>®</sup>**

A chemotherapy that is given as a treatment for some types of cancer. (Matrex<sup>®</sup> is a name brand of methotrexate.)

## **Mechlorethamine**

A medication used topically to treat CTCL. Also known as topical nitrogen mustard.

## **Medical oncologist**

A physician who specializes in the use of chemotherapy, hormone therapy and many other types of biologic therapies to treat cancer.

## **Memory cells**

Types of B-lymphocytes and T-lymphocytes. After a foreign invader or unwanted cell has been destroyed, surviving B- and T-lymphocytes develop into specialized memory cells that remain on watch and can provide protection if the invader is encountered in the future.

**Metastasize**

To spread to other organs of the body. Cancer may spread from its primary site of origin to other sites or organs.

**Methotrexate**

A chemotherapy that is given as a treatment for some types of cancer. (Matrex® is a name brand of methotrexate.)

**Monoclonal antibodies**

Antibodies that act specifically against a particular antigen. Scientists can produce large amounts of an antibody that can be directed to a single target (or antigen) on the cell's surface. Monoclonal antibodies are used to classify lymphomas by identifying surface proteins on lymphocytes. Monoclonal antibodies are also used as therapeutic agents and have been developed to combat specific cancers including cutaneous T-cell lymphomas.

**MRI (magnetic resonance imaging)**

MRI uses magnets and radio frequency waves to produce images of inside the body. MRIs can provide information about tissues and organs that is not available from other imaging techniques.

**Mucositis**

Inflammation of the lining of the tissues and organs. In the mouth, it is characterized by sores or inflammation.

**Mustargen**

A medication used topically to treat CTCL. (Also known as mechlorethamine.)

**Mycosis fungoides (MF)**

Term for the most common type of CTCL. It is typically a low-grade lymphoma which primarily affects the skin. Generally it has a slow course and often remains confined to the skin. Over time, in about 10% of the cases, it can progress to the lymph nodes and internal organs.

**Myelosuppression**

A reduction in the bone marrow's ability to make red blood cells, white blood cells and platelets.

## **Neutropenia**

An abnormally low level of neutrophils (the white blood cells responsible for fighting bacterial infections).

## **Neutrophils**

The primary type of white blood cells found in the blood that fight bacteria.

## **Nitrogen mustard (NM)**

A medication used topically to treat CTCL. (Also known as mechlorethamine.)

## **Non-bulky tumor**

A small tumor, usually less than five centimeters (approximately two inches).

## **Non-Hodgkin lymphoma (NHL)**

A group of several closely related cancers that arise from the lymphatic system. Although the different types of NHL have some things in common, they differ in what the cancer cell looks like under a microscope, how the cells grow and how the tumor affects the body. CTCLs are a type of NHL.

## **Oncologist**

A physician who specializes in treating cancer. Some specialize in chemotherapy (medical oncologist), radiotherapy (radiation oncologist) or surgery (surgical oncologist).

## **Palliation**

Treatment that is given to remove or relieve symptoms.

## **Partial remission (PR)**

The term used when a cancer has shrunk in size by at least half but has not totally disappeared. The cancer can still be detected and other treatments may be recommended.

## **Pathologist**

A physician who specializes in studying disease through microscopic evaluation of body tissues and organs (biopsy). Any tissue suspected of being cancerous must first be examined by a pathologist to confirm the diagnosis.

**PCR (Polymerase chain reaction)**

A molecular test that can identify small amounts of genetic material.

**Pentostatin**

A medication used in the treatment of CTCL.

**PUVA**

Combining treatment with UVA with psoralen, a drug that promotes sensitivity to light.

**Sézary syndrome (SS)**

The leukemic variant of CTCL. Patients usually present with SS, but rarely patients with early stage mycosis fungoides develop SS. The presenting features of SS include widespread redness and scaling of the skin (erythroderma), often with severe itching. Lymph nodes are enlarged and the malignant T-cells found in the skin are also found circulating in the bloodstream.

**Stable disease**

The disease does not get better or worse following therapy.

**Stage**

The extent of cancer at the time of diagnosis. It discriminates if the cancer is localized to its site of origin, spread to neighboring regions or distant sites of the body.

**Standard therapy**

The most widely used primary therapy.

**Stem cell therapy**

A therapy used in experimental stages in the treatment of CTCL.

**Synergism**

The term used when two or more drugs given together provide a better anti-cancer effect than expected from the additive effects from the medications alone.

### **Systemic chemotherapy**

A chemotherapy with single agents along with combination chemotherapy is usually reserved for advanced stages (Stage III and IV) that are recalcitrant to other forms of therapy and administered orally or intravenously.

### **Targeted therapy**

A treatment that is directed to specific genes or proteins (targets) unique or abnormally expressed in a cancer cell.

### **Targretin®**

Targretin® (tar-GRET-in) is a name brand of the medication bexarotene (beks-AIR-oh-teen). It is available in capsule and gel form.

### **Thrombocytopenia**

A shortage of platelets in the blood, which reduces the ability of the blood to clot.

### **Thymus gland**

A gland located behind the sternum (breastbone) that enhances the reproduction and development of lymphocytes. T-lymphocytes are processed in the thymus.

### **Topical nitrogen mustard**

A medication used topically to treat CTCL. (Also known as mechlorethamine.)

### **Topical steroids**

High potency topical steroids have been shown to have activity in CTCL, and induce clearing in early stage disease (Stage I-A and I-B). Topical steroids are easy to apply and are not associated with many complications like those seen with other skin based treatments for CTCL.

### **Toxicities**

The unwanted side effects of cancer therapies, such as a decrease in blood cells, nausea and vomiting, and hair loss.

**TSEB (Total skin electron beam)**

A form of radiation therapy that only treats the superficial portions of the skin. It is highly effective in clearing all forms of lesions of CTCL from the skin. Also known as electron beam therapy.

**Tumor**

An abnormal mass or swelling of tissue. Tumors may occur anywhere in the body. A tumor may be benign (non-cancerous) or malignant (cancerous).

**UVB — broadband**

A form of phototherapy that uses ultraviolet light involving the entire range of UVB wavelengths.

**UVB — narrowband**

A form of phototherapy that concentrates ultraviolet output in a narrow range of UVB wavelengths. It can be an effective treatment for patch-stage CTCL as well as other skin diseases.

**Vaccine**

A substance or group of substances meant to stimulate the immune system to respond. A vaccine can help the body recognize and destroy cancer cells. Lymphoma vaccines often combine cancer antigens with a substance to stimulate the patient's own natural defenses to fight the disease. These vaccines are custom-made for each patient using a sample of tumor obtained from the patient's lymph nodes.

**X-ray**

Radiation that is used in low doses to provide images of the inside of the body and in high doses to treat cancer.















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