

TREATMENT UPDATE:

Cutaneous T-Cell Lymphoma

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Cutaneous T-cell lymphoma (CTCL) is a group of rare forms of non-Hodgkin lymphoma that primarily affect the skin.

In CTCL, T-cells (a type of white blood cell) grow inappropriately and accumulate in the body.

The majority of people with CTCL have a specific subtype called mycosis fungoides, in which the malignant (abnormal) T-cells are typically slow-growing and primarily (or often only) affect the skin. Sézary syndrome is a much less common type of CTCL in which the malignant T-cells affect the blood as well as the skin. There are other subtypes of CTCL that are even less common.

Diagnosis

The accumulation of T-cells in the skin cause most people with CTCL to develop skin lesions described as patches (a flat, red rash) or plaques (a raised patch), both of which may be dry, itchy and/or scaly. Larger, significantly raised nodules (tumors) may develop, but are less common.

CTCL is diagnosed through a skin biopsy, in which a small sample of skin is removed and then examined by a pathologist to determine if it contains abnormal T-cells that reflect the presence of CTCL. Other conditions, such as psoriasis and eczema, can result in abnormal T-cells in the skin, and multiple biopsies are sometimes needed for an accurate diagnosis.

In addition to a skin biopsy, diagnostic tests performed when CTCL is suspected include:

- **Blood tests.** Blood is drawn and analyzed for the presence of abnormal T-cells.
- **Imaging tests.** Tests such as PET scans, CT scans and/or MRIs are used to determine if the CTCL has spread to other parts of the body, such as lymph nodes or organs.



Treatment Options

There are a number of options for the treatment of CTCL. The recommended treatment approach is individualized and is based on a number of factors, including:

- The CTCL subtype
- How much of the skin is affected
- The types of lesions that have developed
- Whether or not the malignant cells have spread beyond the skin

Other factors are also considered, such as the person's age, other medical conditions they may have and their overall health.

In many cases, a combination of approaches is used. You and your doctor will discuss the approach that is right for you based on your individual circumstances.

Skin-Directed Therapy

People with mycosis fungoides can often be treated solely with therapies that are directed to the skin, rather than delivered to the whole body. Skin-directed therapies are also an important component of treatment for Sézary syndrome and other types of CTCL.

The type of skin-directed therapy used in the treatment of CTCL is based on the extent of the affected skin and the type of patch that is present.

Topical Treatments

CTCL that affects only a small area of the skin can often be treated with medicated gels, ointments or creams that are applied directly to the skin.

Topical treatments include:

- **Retinoids.** Retinoids are chemically related to vitamin A and appear to interfere with genes that control cell growth. In the treatment of CTCL, the retinoids bexarotene (Targretin) gel and acitretin (Soriatane) can slow cancer growth by damaging T-cell lymphoma cells on the skin.
- **Corticosteroids.** Steroids can help reduce the itching and redness caused by CTCL itself and by other topical treatments, such as retinoids.
- **Chemotherapy.** Mechlorethamine gel (Valchlor) is approved for the treatment of early stage CTCL in people who received prior skin-directed therapy.
- **Immune-stimulating creams.** Imiquimod cream (Aldara and Zyclara) works by stimulating the release of specific proteins that boost the immune system. Imiquimod cream is more commonly used for other skin conditions, and is usually used to treat CTCL only after other treatments have stopped working.



Phototherapy

In phototherapy, the skin is exposed to ultraviolet light (UVA or UVB) which destroys the malignant T-cells near the surface of the skin. Drugs called psoralens are sometimes administered to make the cancer cells more likely to be destroyed by the ultraviolet light.

Radiation

Electron beam radiation therapy, which targets the skin and does not deliver radiation deep into the body, is used in the treatment of CTCL. Electron beam radiation therapy can be directed to a specific area or to the entire surface of the skin, depending on the extent of the affected skin and the goals of treatment.

Systemic Therapy (Medications)

Systemic therapy is treatment in which one or more drugs are delivered to the entire body. Systemic therapy is used in the treatment of CTCL when skin-directed therapy does not adequately work or the CTCL has spread beyond the skin. It can also be used as an initial treatment approach for Sézary syndrome. Different types of systemic therapies are sometimes given in combination.

Chemotherapy

Chemotherapy is broadly defined as drugs used to kill cancer cells, usually by stopping the ability of the cells to grow and divide. Chemotherapy drugs used to treat CTCL include:

- **Bexarotene (Targretin)** binds to specific receptors within cancer cells called “Retinoid X-Receptors,” slowing the growth of the abnormal T-cells.

- **Romidepsin (Istodax) and vorinostat (Zolinza)** are enzyme inhibitors that can slow or stop the growth of cancer cells. These therapies are used to treat CTCL that has progressed or recurred (come back).
- **Methotrexate (Rheumatrex, Trexall, others)** prevents abnormal T-cells from using folic acid to create more cancer cells.
- **Pralatrexate (Folotyn)** is approved for the treatment of an advanced type of CTCL known as transformed mycosis fungoides.

Extracorporeal photopheresis (ECP)

In ECP, blood is drawn from the body and separated into white blood cells, red blood cells and platelets. The white blood cells are exposed to ultraviolet (UV) light after being treated with methoxsalen, which sensitizes the lymphoma cells to being destroyed by the UV light. All of the drawn blood is returned to the body where the immune system is stimulated to help fight CTCL.

Immunotherapy

Our immune system is constantly working to keep us healthy. It recognizes and fights against danger, such as infections, viruses and growing cancer cells. In general terms, immunotherapy uses our own immune system as a treatment against cancer.

Interferons (IFNs) are naturally occurring substances that can boost the immune system. IFN alfa and IFN gamma are given intravenously (into a vein) to treat CTCL.

In August 2018, the U.S. Food and Drug Administration (FDA) approved the immunotherapy mogamulizumab-kpkc (Poteligeo) for the treatment of patients with CTCL who have received at least one prior systemic therapy. Mogamulizumab-kpkc is a monoclonal antibody that targets CCR4, a protein found on some cancer cells (including CTCL).

Targeted Therapy

Targeted therapy focuses on specific molecules and cell mechanisms thought to be important for cancer cell survival and growth, taking advantage of what researchers have learned in recent years about how cancer cells grow.

In November 2017, the FDA approved brentuximab vedotin (Adcetris) for the treatment of mycosis fungoides in which CD30 (a protein that can lead to the growth of cancer cells) is present. Brentuximab vedotin is an antibody-drug conjugate that directs a medication called auristatin to cells that have CD30. The approval is specific to people who have received prior systemic therapy.

Stem Cell Transplant

Stem cell transplantation is a very aggressive therapy. It is not commonly used in the treatment of CTCL, but may be recommended if the abnormal T-cells are growing rapidly or extensively and are not adequately controlled by skin-directed or systemic therapies.

In an allogenic stem cell transplant, stem cells are removed from a close relative or an unrelated person who is a “match” (has a similar tissue type). Concurrently, the person with CTCL is given treatment to destroy as many cancer cells as possible. The stem cells are then “infused” (transplanted) into the body, forming new healthy blood cells and boosting the body’s defense against the cancer cells.

Treatment Side Effects

All treatments for CTCL can cause side effects. It's important that you report any side effects you experience to your health care team so they can help you manage them. Report them right away—don't wait for your next appointment. Doing so may improve your quality of life and allow you to adhere to your treatment plan. It's important to remember that not all patients experience all side effects, and patients may experience side effects not listed here.

CTCL itself carries a risk of infection, and treatment can increase that risk. Your doctor may recommend a medication to help prevent infection. Notify your health care team immediately if you experience a fever, as this is a sign of a possible infection.

Side Effects of Skin-Directed Therapy

Topical Treatments

With long-term use, corticosteroids can cause skin to become thinner, so doctors carefully manage the dose and length of use. Retinoids, chemotherapy and protein-stimulating creams can cause inflammation and redness of the skin, which is usually relieved by the use of topical steroids.

Phototherapy and Radiation

The ultraviolet light used in phototherapy can cause skin to become itchy and red. Nausea, headache and fatigue may occur if phototherapy is combined with the use of psoralen drugs.

Changes to the skin are the most common side effects of radiation therapy; those changes can include dryness, swelling, peeling, redness and blistering.

If a reaction occurs when being treated with phototherapy or radiation, contact your health care team so the appropriate treatment can be prescribed. It's especially important to contact your health care team if there is any open skin or painful areas, as this could indicate an infection. Infections can be treated with an oral antibiotic or topical antibiotic cream.

Side Effects of Systemic Therapy

Chemotherapy

The side effects specific to chemotherapy depend on the drug or drugs given, their dose and the length of time they are used. Side effects can include:

- Hair loss
- Easy bruising or bleeding
- Changes in memory or thinking
- Peripheral neuropathy (numbness or tingling in hands and feet)
- Mouth sores

Extracorporeal photopheresis (ECP)

The side effects of ECP are generally minimal, but may include a low-grade fever, slight nausea, dizziness and increased redness of the skin.

Immunotherapy

Immunotherapy travels through the bloodstream, helping to prompt what is called an "immune response." Because immunotherapy can attack healthy cells as well as cancer cells, certain side effects may be experienced. The most common side effects of treatment with mogamulizumab-kpkc include rash, fatigue, diarrhea, musculoskeletal pain and upper respiratory tract infection.

Targeted Therapy

Targeted therapy drugs don't have the same effect on the body as do chemotherapy drugs, but they can still cause side effects. It's important to have a discussion with your health care team to understand the potential side effects of your treatment, how to reduce the risk of their occurring, and how to minimize their impact should you experience them.



General Side Effects

There are certain side effects that may occur across different treatment approaches. The following tips and guidance may help you manage these side effects.

Managing Digestive Tract Symptoms

Nausea and vomiting

- Avoid food with strong odors, as well as overly sweet, greasy, fried or highly seasoned food.
- Eat meals cold or at room temperature, which often makes food more easily tolerated.
- Nibble on dry crackers or toast. These bland foods are easy on the stomach.
- Having something in your stomach when you take medication may help ease nausea.

Diarrhea

- Drink plenty of water. Ask your doctor about using drinks such as Gatorade that provide electrolytes as well as liquid. Electrolytes are body salts that must stay in balance for cells to work properly.
- Over-the-counter medicines such as loperamide (Imodium A-D and others) and prescription drugs are available for diarrhea but should be used only if necessary. If the diarrhea is bad enough that you need medicine, discuss it with your doctor or nurse.
- Choose foods that contain soluble fiber, like beans, oat cereals and flaxseeds. High-pectin foods such as peaches, apples, oranges, grapefruit, bananas and apricots can also help to avoid diarrhea.

- Low fat food choices are less likely to cause diarrhea than fatty, greasy or fried foods. The fats you eat should come from healthy sources, such as olive oil, canola oil, avocado, olives, nuts and seeds.

Loss of appetite

- Eating small meals throughout the day is an easy way to take in more protein and calories, which will help maintain your weight. Try to include protein in every meal.
- To keep from feeling full early, avoid liquids with meals or take only small sips (unless you need liquids to help swallow). Drink most of your liquids between meals.
- Keep high-calorie, high-protein snacks on hand such as hard-boiled eggs, peanut butter, cheese, ice cream, granola bars, liquid nutritional supplements, puddings, nuts, canned tuna or trail mix.
- If you are struggling to maintain your appetite, talk to your health care team about whether appetite-building medication could be right for you.



Managing Fatigue

Fatigue (extreme tiredness not helped by sleep) is one of the most common side effects of many cancer treatments. If you are taking a medication, your doctor may lower the dose of the drug, as long as it does not make the treatment less effective. If you are experiencing fatigue, talk to your doctor about whether taking a smaller dose is right for you.

There are a number of other tips for reducing fatigue:

- Take several short naps or breaks.
- Take short walks or do some light exercise, if possible.
- Try easier or shorter versions of the activities you enjoy.
- Ask your family or friends to help you with tasks you find difficult or tiring.

There are also prescription medications that may help. Your health care team can provide guidance on whether medication is the right approach for your individual circumstances.

Fatigue can be a symptom of other illnesses, such as anemia, diabetes, thyroid problems, heart disease, rheumatoid arthritis and depression. Be sure to ask your doctor if he or she thinks any of these conditions may be contributing to your fatigue.

Managing Pain

There are a number of options for pain relief, including prescription and over-the-counter medications. It's important to talk to a member of your health care team before taking any over-the-counter medication to determine if they are safe and whether they will interfere with your treatments. Many pain medications can lead to constipation, which may make your pain worse. Your doctor can prescribe medications that help to avoid constipation.

Physical therapy, acupuncture and massage may also be of help in managing your pain. Other techniques, such as mindfulness meditation, deep breathing exercises and yoga may also be helpful. Consult with a member of your health care team before beginning any of these activities.



Communicating With Your Health Care Team

As you manage your CTCL, it's important to remember that you are a consumer of health care. The best way to make decisions about health care is to educate yourself about your diagnosis and the members of your health care team, including nurses, social workers, nutritionists and patient navigators.

Here are some tips for improving communication with your health care team:

Start a health care journal. Having a health care journal or notebook will allow you to keep all of your health information in one place. You may want to write down the names and contact information of the members of your health care team, as well as any questions for your doctor. Keep a diary of your daily experiences with cancer and treatment. You can separate your journal or notebook into different sections to help keep it organized.

Prepare a list of questions. Before your next medical appointment, write down your questions and concerns. Because your doctor may have limited time, you should ask your most important questions first, and be as specific and brief as possible.

Bring someone with you to your appointments. Even if you have a journal and a prepared list of questions or concerns, it's always helpful to have support when you go to your appointments. The person who accompanies you can serve as a second set of ears. He or she may also think of questions to ask your doctor or remember details about your symptoms or treatment that you may have forgotten.

Write down your doctor's answers. Taking notes will help you remember your doctor's responses, advice and instructions. If you cannot write down the answers, ask the person who accompanies you to do that for you. If you have a mobile device, like a tablet or smartphone, ask if you can use it to take notes. Writing notes will help you review the information later.

Record your visit if your doctor allows it. Recording the conversation with your doctor gives you a chance to hear specific information again or share it with family members or friends.

Incorporate other health care professionals into your team.

Your medical oncologist is an essential member of your health care team, but there are other health care professionals who can help you manage your diagnosis and treatment:

- Your primary care physician should be kept updated about your cancer treatment and any test results.
- Dermatologists and radiation oncologists are an important part of the multi-disciplinary team approach in the treatment of patients with CTCL.
- Your local pharmacist is a great source of knowledge about the medications you are taking. Have all of your prescriptions filled at the same pharmacy to avoid the possibility of harmful drug interactions.
- Make sure your oncologist knows of any other medical conditions you have or any pain you are experiencing so that they can consult with your primary care physician or specialist as needed.

Remember, there is no such thing as over-communication.

CancerCare's Free Support Services and Programs

It can be very difficult to receive a diagnosis of CTCL, and adjusting to the necessary changes in your life can be challenging.

CancerCare can help. We are a national nonprofit organization providing free, professional services to anyone affected by cancer. Our licensed oncology social workers can provide support and education, help in navigating the complicated health care system, and provide information on support groups and other resources.

To learn more about how CancerCare helps, call us at 800-813-HOPE (4673) or visit www.cancercare.org.

You will likely also build your own personal support network comprised of family and friends. In doing so, it's best to take some time to think about the people in your life and how they are best suited to help. Match the task to their strengths—ask a family member who loves to shop to pick up something for you at the store; ask a friend who's a good listener to come over for a chat.



Frequently Asked Questions

Q: Is CTCL curable? Can it recur?

A: Although there is currently no known cure for CTCL outside of stem cell transplantation, it is highly treatable. As a result of clinical trials, additional treatment options have become available for people living with CTCL. Most people with mycosis fungoides type CTCL are diagnosed at an early stage and have a normal life expectancy. Some people may live symptom-free for years, although the patches and plaques associated with CTCL tend to recur and require additional treatment.

Q: How can I manage the itch associated with CTCL?

A: Severe itching of the skin, also called pruritus, is experienced by most people with CTCL. The itching associated with CTCL can be treated with prescription-strength medications that are applied directly to the skin or taken as tablets. These medications are typically either antihistamines (drugs used to treat allergic reactions) or corticosteroids (drugs that act against inflammation). The itching often increases in severity at bedtime, in which case a sedative antihistamine may help.

Additionally, it's important to be gentle with your skin: shower or bathe in lukewarm water, choose skin cleansers without fragrance or alcohol, and avoid after-bath powders that may aggravate itching.

Q: What is a treatment summary and why is important?

A: A treatment summary, sometimes called a “shadow chart,” is a document that you create and keep in your possession. Maintaining your own records allows you and your family members to have instant access to the specifics of your CTCL diagnosis and treatment. A treatment summary should include:

- Your name and date of birth
- Date of diagnosis
- Prescribed therapy/therapies, including start dates, end dates and dosages when appropriate
- Dates and types of post-diagnosis testing and the results of these tests
- Other medications and supplements you are taking
- Names, affiliations and contact information of all members of your health care team

Talk to your doctor or a member of your health care team about your intention to create a treatment summary, and ask what else they suggest be included. Take your treatment summary with you when you visit any doctor, not just your oncologist or dermatologist.

Resources

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American Cancer Society

800-227-2345
www.cancer.org

Cancer.Net

Patient information from the
American Society of Clinical
Oncology
888-651-3038
www.cancer.net

National Cancer Institute

800-422-6237
www.cancer.gov

Cancer Support Community

888-793-9355
www.cancersupportcommunity.org

**National Coalition for
Cancer Survivorship**

877-622-7937
www.canceradvocacy.org

Cutaneous Lymphoma Foundation

248-644-9014
www.clfoundation.org

Lymphoma Research Foundation

800-500-9976
www.lymphoma.org

The Leukemia & Lymphoma Society

800-955-4572
www.lls.org

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