

T-cell Lymphomas

Expert review by:

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What is lymphoma?

Lymphoma is a cancer of the white blood cells, namely lymphocytes, that happen to constitute the lymphatic system. The two main types of lymphoma are Hodgkin lymphoma (HL) and non-Hodgkin lymphoma (NHL). Lymphoma is the most common blood cancer and the third most common cancer of childhood. Lymphoma occurs when lymphocytes grow abnormally. The body has two types of lymphocytes: B lymphocytes, or B-cells, and T lymphocytes, or T-cells. Although both cell types can develop into lymphomas, B-cell lymphomas are more common. Like normal lymphocytes, those that turn malignant can grow in many parts of the body, including the lymph nodes, spleen, bone marrow, blood or other organs.

What is non-Hodgkin lymphoma?

Of the more than 35 types of lymphoma, 30 are classified as non-Hodgkin lymphoma. The average age of diagnosis is in the 60's. While scientists do not know the exact causes of NHL, they do know that it is not caused by injury or by coming into contact with someone else with the disease. Most people diagnosed with NHL have no known risk factors, though increasingly many scientists believe infections may play an important role in causing select types of NHL.

What is T-cell lymphoma?

T-cell lymphoma accounts for only about 15 percent of all cases of NHL in the United States, although some forms of T-cell lymphoma are more common in Asia and other parts of the world. There are many different types of T-cell lymphoma and some are extremely rare, occurring in only a few patients per year throughout the world. Most T-cell lymphomas are diagnosed by taking a small sample (called a biopsy) of the tumor and looking at the cells under a microscope. However, many forms of T-cell lymphoma are unusual in that the cells may look the same, making the diagnosis difficult. Thus, other blood tests can be helpful in diagnosing T-cell lymphoma. One of the most common forms of T-cell lymphoma is cutaneous, or skin lymphoma, because it starts in the lymphocytes in the skin. Cutaneous lymphoma actually describes many different disorders with various signs and symptoms, outcomes, and treatment considerations.

Brief descriptions of the main types of T-cell lymphoma appear below.

Peripheral T-cell lymphoma is actually any of a large number of different T-cell lymphomas. Together, these lymphomas account for about 10 percent of all cases of non-Hodgkin lymphoma and can occur anytime during adulthood. Some varieties of peripheral T-cell lymphoma are quite rare in the United States, but more common in countries such as Japan and China, where an infection called HTLV-1 is very

common. HTLV-1 infection can make people more likely to develop some types of peripheral T-cell lymphoma. Chemotherapy is often used as treatment.

Anaplastic large cell lymphoma can appear in skin, like cutaneous T-cell lymphoma (CTCL), but it can also appear in other organs throughout the body (then it is called **systemic anaplastic large cell lymphoma**). The skin form grows quite slowly, and prognosis is usually excellent after radiation, surgery or occasionally chemotherapy as treatment. Systemic anaplastic large cell lymphoma responds fairly well to chemotherapy medications used for treating other large cell lymphomas and is potentially curable. A condition called **lymphomatoid papulosis (LyP)**, while not classified as a lymphoma, is often a precursor of cutaneous anaplastic large cell lymphoma. LyP is poorly understood and fairly rare.

Angioimmunoblastic lymphoma is a fast-growing T-cell lymphoma. It accounts for 1 to 2 percent of all cases of non-Hodgkin lymphoma in the United States. Symptoms include high fever, night sweats, skin rash and certain molecular abnormalities that can be detected with blood tests. Angioimmunoblastic lymphoma is often first treated with steroids, although this lymphoma often progresses to other types of lymphoma, which then require other medications. For more advanced cases, doctors may use bone marrow transplantation.

T-cell leukemias also arise from T-cells and can behave like T-cell lymphoma. These include **T-cell promyelocytic leukemia**, **T-cell granular lymphocytic leukemia**, **aggressive NK-cell leukemia** and **adult T-cell lymphoma/leukemia**. The medication Arranon (nelarabine) has recently been approved to treat T-cell leukemia. In addition, several chemotherapy combinations are currently being tested against T-cell leukemias.

Blastic NK-cell lymphoma is a very rare form of T-cell lymphoma, affecting only a few people (usually adults) internationally each year. This cancer is very fast-growing and difficult to treat. It can arise anywhere in the body, although cases appearing in the skin have a better outlook. Since this cancer is so rare, patients should consult with their medical team, who can search the medical literature for promising therapies or clinical trials.

Nasal T-cell lymphomas are also rare in the United States, but relatively common in Asia and parts of Latin America. For this reason, researchers suspect that some ethnic groups may be more prone to this cancer that affects both children and adults. It is a fast-growing lymphoma that is treated with various combinations of chemotherapy (i.e., CHOP) and radiation. As with other rare cancers, patients should discuss treatment options with their medical team and consider participating in clinical trials.

Treatment-related T-cell lymphomas sometimes arise after transplantation of solid organs or bone marrow. The considerable immune system suppression that is required for transplant patients can put these patients at risk for developing post-transplant lymphoproliferative disorders, certain unusual forms of peripheral T-cell lymphoma and other types of non-Hodgkin lymphoma. These post-treatment cancers may require therapy that differs from the standard treatments normally used to treat these conditions.

Other T-cell lymphomas include **enteropathy-type T-cell lymphoma**, which appears in the intestines and **hepatosplenic gamma-delta T-cell lymphoma**, which starts in the liver or spleen.

Rare immune system cancers include a group of rare cancers that are neither B-cell or T-cell lymphomas, but are cancerous forms of other types of white blood cells. These include histiocytic and dendritic cell neoplasms. Patients diagnosed with these cancers should seek advice from their medical team about clinical trials that may be recruiting patients with these cancers.

Lymphoblastic lymphoma can appear in both B-cells and T-cells, but it is much more common in T-cells, comprising 80 percent of all lymphoblastic lymphoma. This T-cell lymphoma is most often seen in children, and with intensive chemotherapy the complete remission rate can be as high as 96 percent. The medication Arranon has recently been approved to treat T-cell lymphoblastic lymphoma.

Cutaneous T-cell lymphoma, often abbreviated CTCL, accounts for about 2 to 3 percent of all cases of NHL lymphoma and mostly affects adults. CTCL is a slow-growing cancer that usually starts in the skin. Mycosis fungoides is the most common type of CTCL. It appears as skin patches, or plaques, and usually progresses over many years. A much rarer form of CTCL is Sézary syndrome,

characterized by widespread reddening of the skin. Sézary syndrome is generally thought of as an advanced, or variant, form of mycosis fungoides. Because some common skin conditions have similar symptoms, like eczema or psoriasis, CTCL may be missed for years. Doctors treat CTCL based on its stage, or how far it has progressed in the body. Earlier stages respond well to electron beam radiation, ultraviolet light therapy or chemotherapy skin creams, whereas later stages require standard chemotherapy given through the blood.

What treatments are available?

Because there are so many different types of T-cell lymphoma, treatment varies widely. Standard lymphoma therapies may be effective; these include chemotherapy, radiation, bone marrow transplantation and surgery. Treatments that use ultraviolet light therapy or electron beam radiation (a type of radiation that does not penetrate to internal organs) are effective for the many forms of T-cell lymphoma that appear in skin. Other drugs that have been recently approved for use in skin forms of T-cell lymphoma include Targretin (bexarotene), Ontak (Denileukin Diftitox) and Zolinza (vorinostat). Because T-cell lymphoma is uncommon, treatment is often not well-defined. For this reason, patients should consider talking to their medical team about participating in a clinical trial as initial therapy. Many of the improvements in survival have been made using investigational therapies aiming to improve on the best available conventional treatments. For a listing of treatments currently being tested in clinical trials, visit www.clinicaltrials.gov and type “T-cell lymphoma” in the search box. Assistance in locating clinical trials is also available by contacting the *Lymphoma Helpline and Clinical Trials Information Service*.

Are complementary and alternative therapies safe and effective?

Complementary and alternative therapies are non-standard therapies that may help patients cope with their cancer and its treatment, but that should not be used as the only treatment. No alternative therapy has been proven effective against lymphoma. However, complementary therapies such as meditation, exercise, prayer, diet and relaxation techniques may provide comfort and emotional strength. It is very important for patients considering

alternative or complementary treatments to discuss the matter openly with their healthcare team. Certain unproven treatments, including some herbal supplements, can interfere with standard lymphoma treatments or may cause serious side effects.

What about follow-up appointments?

During and after treatment, patients should play an active role in their healthcare, including keeping a master file of medical records, asking questions, reporting new symptoms, exercising and eating a balanced diet. In addition, patients who smoke should strongly consider stopping. Follow-up visits, usually scheduled every few months, typically include CT scans and a variety of blood tests. After treatment, it is very important to keep these appointments. Since lymphoma symptoms may resemble those of less serious illnesses, like colds or viral infection, regular medical care is very important. In addition to looking for signs of the cancer coming back, follow-up care can help identify and resolve unusual side effects of treatment.

How can I find support?

A lymphoma diagnosis may provoke a range of feelings and bring many concerns. In addition, cancer treatment can cause physical and emotional discomfort. Connecting with other people who have lymphoma, or have been cured of it, can help a great deal. Support groups and on-line message boards are often useful. One-to-one peer support programs, such as the Lymphoma Research Foundation’s *Lymphoma Support Network*, matches lymphoma survivors (or caregivers) with volunteers who have gone through similar experiences. The Cutaneous Lymphoma Foundation offers an informative website www.clfoundation.org, newsletter and e-mail listserv for patients with T-cell lymphoma of the skin.

How can I stay informed?

The Lymphoma Research Foundation offers a wide range of resources that address treatment issues, the latest research advances, and coping with all aspects of lymphoma. For more information about any of these resources:

Call 800-500-9976

Email: helpline@lymphoma.org

Web: www.lymphoma.org

Contact Us

For more information about *Getting the Facts* or information about the



Please contact:

Los Angeles

Patient Education, Services and Support
8800 Venice Boulevard, Suite 207
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The Lymphoma Research Foundation (LRF) offers a comprehensive series of patient education and support programs including:

- *Lymphoma Helpline & Clinical Trials Information Service*
- *Lymphoma Support Network*
- Patient Aid Grant Program
- Publications and newsletters
- Informational teleconferences and webcasts
- In-person conferences
- National Chapter Network

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Glossary of Terms

Biopsy: Removal of a small piece of tissue (for example, a lymph node) for evaluation under a microscope

Chemotherapy: Treatment with drugs ("chemo") to stop the growth of rapidly dividing cancer cells, including lymphoma cells

Complete remission: Term used when all signs of the disease have disappeared after treatment

Electron beam radiation: Radiation of the skin that does not penetrate to internal organs

Lymph nodes: Small bean-shaped glands located in the small vessels of the lymphatic system; thousands are located throughout the body; they are most easily felt in the neck, armpits, and groin

Lymphatic system: The vessels, tissues, and organs that store and carry lymphocytes that fight infection and other diseases

Lymphocyte: A type of white blood cell

Ultraviolet light therapy: Use of UVB ultraviolet light exposure to slow the rapid growth of skin cells; light boxes with full-body exposure are used to deliver ultraviolet rays that can treat CTCL

About LRF

The mission of the Lymphoma Research Foundation (LRF) is to eradicate lymphoma and serve those touched by this disease. The Foundation is the nation's largest lymphoma-focused voluntary health organization devoted exclusively to funding lymphoma research and providing patients and healthcare professionals with critical information on the disease. More than 85 cents of every dollar spent support research and programming. People affected by lymphoma can receive free personalized information tailored to their diagnosis, help with finding a clinical trial, and easy-to-understand information on lymphoma, current treatments, and promising research. Please call 800-500-9976, email helpline@lymphoma.org, or visit the website www.lymphoma.org