

## Angioimmunoblastic T-Cell Lymphoma

### Overview

Lymphoma is the most common blood cancer. The two main forms of lymphoma are Hodgkin lymphoma (HL) and non-Hodgkin lymphoma (NHL). Lymphoma occurs when lymphocytes, a type of white blood cell, grow abnormally. The body has two main types of lymphocytes that can develop into lymphomas: B-lymphocytes (B-cells) and T-lymphocytes (T-cells). Cancerous lymphocytes can travel to many parts of the body, including the lymph nodes, spleen, bone marrow, blood or other organs, and can accumulate to form tumors.

Angioimmunoblastic T-cell lymphoma is a rare, aggressive (fast-growing) T-cell lymphoma that accounts for between 1 percent and 2 percent of all NHL cases in the United States. Symptoms include high fever, night sweats, skin rash and some types of autoimmune disorders, such as autoimmune hemolytic anemia (AIHA) and immune thrombocytopenic purpura (ITP), in which the body does not recognize its own cells. As a result of these autoimmune disorders, the body makes antibodies against and destroys its own cells and tissues, such as platelets (in the case of ITP) and red blood cells (in the case of AIHA).

As with other types of NHL, diagnosing angioimmunoblastic T-cell lymphoma requires taking a small sample of the tumor tissue, called a biopsy, and looking at the cells under a microscope. Once a diagnosis has been made, a series of other diagnostic tests, such as blood, CT (computerized axial tomography), MRI (magnetic resonance imaging) and PET (positron emission tomography) scans and a bone marrow biopsy, may be done to determine the extent, or stage, of the disease.

The majority of patients with angioimmunoblastic T-cell lymphoma are diagnosed with stage III (disease is found in lymph nodes both above and below the diaphragm) or stage IV disease (disease has spread beyond the lymph nodes and spleen to one or more other organs, such as the bone, bone marrow, skin or liver). Although the actual cause of angioimmunoblastic T-cell lymphoma is unknown, this lymphoma subtype has been linked to the Epstein Barr Virus (EBV).

### Treatment Options

Angioimmunoblastic T-cell lymphoma is usually first treated with chemotherapy regimens such as CHOP (cyclophosphamide, doxorubicin, vincristine, prednisone), dose intense regimens such as Hyper-CVAD (cyclophosphamide, vincristine, doxorubicin, dexamethasone), or non-adriamycin based chemotherapy, radiation or high-dose chemotherapy followed by an autologous stem cell transplant (stem cell transplant in which a patient receives their own stem cells). Once a patient has disease that has relapsed, alternative therapies are indicated such as pralatrexate (Folotyn), other salvage chemotherapies such as a gemcitabine (Gemzar) containing regimen, or an allogeneic stem cell transplant (stem cell transplant in which a patient receives stem cells from a donor).

### Treatments Under Investigation

While standard therapy has not proven to be very effective against angioimmunoblastic T-cell lymphoma (only 10 percent to 15 percent of patients have long-term remission), several drugs currently being tested in clinical trials are showing promise. For example, a new class of drugs, known as histone deacetylase inhibitors, which include the drugs suberoylanilide hydroxamic acid (SAHA) and vorinostat (Zolinza), have been effective in the treatment of a variety of T-cell lymphomas.

Some treatments under investigation in clinical trials for the treatment of angioimmunoblastic T-cell lymphoma include:

- CHOP (cyclophosphamide, doxorubicin, vincristine, prednisone) plus bevacizumab (Avastin)
- Depsipeptide (Istodax)
- Lenalidomide (Revlimid)
- PEGS (cisplatin, etoposide, solimmedrol, gemcitabine) chemotherapy
- Vorinostat (Zolinza)

These clinical trials are in various phases of development. It is

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The Lymphoma Research Foundation offers the following patient education and support programs:

- *Lymphoma Helpline*
- Clinical Trials Information Service
- Lymphoma Support Network
- Publications
- Teleconferences
- Webcasts & podcasts
- In-person conferences

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critical to remember that today's scientific research is continuously evolving. Treatment options may change as new treatments are discovered and current treatments are improved. Therefore, it is important that patients check with the Lymphoma Research Foundation or their physician for any treatment updates that may have recently emerged.

## Participating in Clinical Trials

Clinical trials are crucial in identifying effective drugs and determining optimal doses for lymphoma patients. Because angioimmunoblastic T-cell lymphoma is such a rare disease, clinical trial enrollment is critical for establishing more effective, less toxic treatments. Patients interested in participating in a clinical trial should talk to their physician. Contact the Lymphoma Research Foundation's *Helpline* for an individualized clinical trial search by calling (800) 500-9976 or emailing [helpline@lymphoma.org](mailto:helpline@lymphoma.org).

## Follow Up

Once treatment is completed, a follow-up care plan should be created by the patient's physician. The plan should provide a treatment summary, which includes information such as age and date of diagnosis, staging, the types of treatment administered and response to treatment. The plan should also indicate the type and frequency of medical tests that a survivor should routinely undergo over their lifespan.

Lymphoma survivors should receive regular medical exams from a physician who is familiar with their medical history as well as the treatments they have received. Survivors and their caregivers are encouraged to keep copies of all medical records and test results as well as information on the types, amounts and duration of all treatments received. This documentation will be important for keeping track of any effects resulting from treatment or potential disease recurrences.

## Finding Support

A lymphoma diagnosis may provide a range of feelings and raise 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 provide enormous relief. Support groups and online message boards are often useful. One-to-one peer support programs, such as the Lymphoma Research Foundation's Lymphoma Support Network, match lymphoma survivors (or caregivers) with volunteers who have gone through similar experiences.

## Resources

The Lymphoma Research Foundation offers a wide range of resources that address treatment options, the latest research advances and how to cope with all aspects of lymphoma. For a more comprehensive source of NHL information, including currently approved treatments and treatments under investigation, please visit the Foundation's website to view or order the publication entitled *Understanding Non-Hodgkin Lymphoma: A Guide for Patients, Survivors and Loved Ones*.

The Foundation also provides many educational activities, from in-person meetings to teleconferences and webcasts. For more information about any of these resources, visit the website at [www.lymphoma.org](http://www.lymphoma.org), e-mail the *Helpline* at [helpline@lymphoma.org](mailto:helpline@lymphoma.org) or call at (800) 500-9976.