

Understanding Angioimmunoblastic T-Cell Lymphoma

Angioimmunoblastic T-cell lymphoma (AITL) is a rare and often fast-growing form of peripheral T-cell lymphoma (PTCL). AITL and the follicular helper subtype of PTCL (PTCL-FH) share a common biology and are often grouped together in treatment approaches. AITL accounts for about 13 percent of PTCLs.

AITL is more common in older people but can sometimes affect young adults as well. Symptoms of AITL include high fever, night sweats, skin rash, and autoimmune disorders such as autoimmune hemolytic anemia (AIHA) and immune thrombocytopenia (ITP). As a result of these autoimmune disorders, the body's immune system attacks its own cells and tissues, such as red blood cells (in the case of AIHA) or platelets (in the case of ITP).

Diagnosing AITL requires taking a *biopsy* (sample of the tumor tissue) and looking at the cells under a microscope. Other tests may be done to determine the extent, or stage, of the disease. These can include blood tests, a computed tomography (CT) scan, a positron emission tomography (PET) scan, a magnetic resonance imaging (MRI) scan, and a bone marrow biopsy. Rarely, a lumbar puncture might also be recommended.

Most patients with AITL are diagnosed with advanced-stage disease, either Stage III or Stage IV. In Stage III, affected lymph nodes are found both above and below the diaphragm. In Stage IV, one or more organs beyond the lymph nodes are affected, such as the bone, bone marrow, skin, or liver. Less-extensive disease, Stage I or II, is rare in AITL. Patients with Stage I have localized disease that has not spread beyond one lymph node or other location; Stage II disease has spread only to nearby lymph nodes.



TREATMENT OPTIONS

Patients with AITL are most commonly treated with combinations of chemotherapy drugs. Recommended *frontline* (initial) therapy for the treatment of AITL is either a clinical trial or a course of chemotherapy using a combination of drugs, such as CHOP (cyclophosphamide, doxorubicin, vincristine, prednisone) or CHOEP (doxorubicin, vincristine, cyclophosphamide, etoposide, prednisone). Sometimes radiation therapy or higher doses of chemotherapy, followed by stem cell transplantation, may be added at the end of treatment with the multitargeted chemotherapy. Brentuximab vedotin (Adcetris), a therapy that targets a specific marker on the surface of cancer cells called "CD30," may be used in combination with cyclophosphamide, doxorubicin, and prednisone for cancers expressing this marker. The goal of such therapy is often to get rid of the lymphoma permanently or at least for a long time. In cases where the goal is to treat the disease more mildly, mostly to relieve symptoms, steroids may be used.

Disease *relapse* (returns after treatment) is common with this cancer. If the cancer returns or is *refractory* (does not respond to treatment), there are several other treatment options available. Romidepsin (Istodax) and belinostat (Beleodaq) are approved for treatment of relapsed or refractory PTCL; patients with AITL were included in the clinical studies that led to these approvals. Romidepsin and belinostat are histone deacetylase

(HDAC) inhibitors that block tumor cells from growing and dividing, causing cell death. Pralatrexate (Folotyn) was the first drug to be approved by the U.S. Food and Drug Administration (FDA) for the treatment of relapsed or refractory PTCL; patients with AITL were included in the clinical study that supported this approved use. Brentuximab vedotin also may be used in patients with T-cell lymphomas with CD30 expression including AITL. Treatments following relapse may include high-dose chemotherapy followed by an *autologous stem cell transplant* (patients receive their own stem cells) or an *allogeneic stem cell transplant* (patients receive stem cells from a donor). For additional information about stem cell transplantation, view the *Understanding Stem Cell Transplantation* publication on the Lymphoma Research Foundation's (LRF's) website at lymphoma.org/publications.



OTHER TREATMENT POSSIBILITIES

Lenalidomide (Revlimid) has been tested alone or in combination with current chemotherapy regimens and shows promise for the treatment of AITL. Some other drugs used in other types of lymphoma that may occasionally be considered for the treatment of patients with AITL include gemcitabine (Gemzar), bortezomib (Velcade), and bendamustine (Treanda), or other chemotherapies. Alemtuzumab (Campath) is also occasionally considered, although it is no longer commercially available and is provided only through the Campath Distribution Program.

TREATMENTS UNDER INVESTIGATION

New treatments for AITL are being researched all the time. There are several drugs currently in clinical trials that are showing promising results, including:

- Duvelisib (Copiktra)
- Cerdulatinib
- Tipifarnib (Zarnestra)
- Ruxolitinib (Jakafi)
- Azacitidine (Vidaza)
- MEDI-570
- Nivolumab (Opdivo)
- Pembrolizumab (Keytruda)
- Plitidepsin (Aplidin)
- Valemetostat

New drugs such as lenalidomide (Revlimid), romidepsin (Istodax), and others are also being studied in combination with chemotherapy such as CHOP (doxorubicin, vincristine, cyclophosphamide, prednisone) or CHOEP (doxorubicin, vincristine, cyclophosphamide, etoposide, prednisone) as frontline therapy. In relapsed patients, combinations of some of the new drugs listed above are also being studied. It is important to remember that today's scientific research is continuously evolving. Treatment options may change as new treatments are discovered and current treatments are improved. Because the science is always changing, it is important for patients to check in with their doctor or with LRF to find out about any new treatments that become available.

CLINICAL TRIALS

Clinical trials are crucial in identifying effective drugs and determining optimal doses for patients with lymphoma. Because AITL is a rare disease, clinical trial enrollment is critical for establishing more effective, less toxic treatments. The rarity of the disease also means that the most novel treatments are often available only through clinical trials. Patients interested in participating in a clinical trial should view the *Understanding Clinical Trials* fact sheet on LRF's website at lymphoma.org/publications, and the *Clinical Trials Search Request Form* at lymphoma.org, talk to their physician, or contact the LRF Helpline for an individualized clinical trial search by calling (800) 500-9976 or emailing helpline@lymphoma.org.

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FOLLOW-UP

Disease relapse and infections are common with this cancer. It is important to seek medical attention for fever or other symptoms related to improper functioning of the immune system.

Patients with lymphoma should have regular visits with a physician who is familiar with their medical history and the treatments they have received. Medical tests (such as blood tests and CT scans or at times PET scans) may be required at various times during remission to evaluate the need for additional treatment.

Some treatments can cause long-term side effects or late side effects, which can vary based on duration and frequency of treatments, age, gender, and the overall health of each patient at the time of treatment. A physician will check for these side effects during follow-up care. Visits may become less frequent the longer the disease remains in remission.

Patients 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 side effects resulting from treatment or potential disease recurrences. LRF's award-winning *Focus On Lymphoma* mobile app (lymphoma.org/mobileapp) or the *Lymphoma Care Plan* (lymphoma.org/publications) can help patients manage this documentation.

PATIENT AND CAREGIVER SUPPORT SERVICES

A lymphoma diagnosis often triggers a range of feelings and concerns. In addition, cancer treatment can cause physical discomfort. One-to-one peer support programs, such as LRF's *Lymphoma Support Network*, connect patients and caregivers with volunteers who have experience with AITL, similar treatments, or challenges, for mutual emotional support and encouragement. Patients and loved ones may find this information useful whether the patient is newly diagnosed, in treatment, or in remission.

Resources

LRF offers a wide range of resources that address treatment options, the latest research advances, and ways to cope with all aspects of lymphoma, including our award-winning mobile app. LRF also provides many educational activities, from in-person meetings to teleconferences and webinars for people with lymphoma and AITL, as well as patient guides and e-Updates that provide the latest disease-specific news and treatment options. To learn more about any of these resources, visit our websites at lymphoma.org/AITL or lymphoma.org, or contact the LRF Helpline at (800) 500-9976 or helpline@lymphoma.org.

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