

Angioimmunoblastic T-cell lymphoma (AITL) and follicular T-cell lymphoma (FTCL)

This page is about angioimmunoblastic T-cell lymphoma (AITL) and follicular T-cell lymphoma (FTCL). These are types of T-cell lymphoma that are treated in the same way.

On this page

[What are AITL and FTCL?](#)

[Who gets them?](#)

[Symptoms](#)

[Treatment](#)

[Relapsed and refractory AITL and FTCL](#)

[Research and targeted treatments](#)

What are AITL and FTCL?

Angioimmunoblastic T-cell lymphoma (AITL) and follicular T-cell lymphoma (FTCL) are types of **T-cell lymphoma – non-Hodgkin lymphomas** that develop from white blood cells called **T cells**. Many T-cell lymphomas have complicated names based on the type of cell they develop from or particular proteins they make.

AITL and FTCL are officially classed as **slow-growing (low-grade)** lymphomas but they often behave like **fast-growing (high-grade)** types. They develop from a particular type of white blood cell called a 'follicular helper T cell'. They are sometimes grouped together under the name 'nodal T-cell lymphomas with T-follicular helper (TFH) phenotype'.

Although they develop from the same type of cell, **lymph node biopsies** from people with AITL and FTCL look different under a microscope. In AITL, affected **lymph nodes** contain abnormal immune cells and lots of new, small blood vessels.

In FTCL, abnormal immune cells are mainly found in parts of the lymph nodes called the 'follicles'.

About 1 in 3 people with AITL or FTCL have abnormal **B cells** as well as abnormal T cells. This can make it difficult to diagnose AITL and FTCL accurately. Very occasionally, the abnormal B cells can develop into a type of **B-cell non-Hodgkin lymphoma**.

Who gets AITL or FTCL?

AITL and FTCL account for about 1 in every 5 cases of T-cell lymphoma. Only around 140 people are diagnosed with AITL or FTCL in the UK each year. These types of lymphoma usually affect older people, typically around the age of 70. They affect men and women equally.

Scientists don't know what causes AITL or FTCL. They seem to be linked to infection with viruses called Epstein-Barr virus (EBV) and human herpesvirus 6 (HHV-6). However, the relationship between having these infections and AITL or FTCL is not clear.

Symptoms of AITL and FTCL

AITL often causes symptoms that affect your whole body (systemic symptoms). FTCL is usually less widespread when it is diagnosed and is more likely to cause symptoms related to where the lymphoma is growing (local symptoms), rather than whole body symptoms. The most common symptoms of AITL and FTCL are:

- fever, night sweats and unexplained weight loss ('B symptoms')
- **swollen lymph nodes**, usually in several places
- **a swollen liver and spleen**, which might make you feel bloated or cause tummy (abdominal) pain
- **itching**
- **skin rash**.

AITL and FTCL often affect your bone marrow (the spongy tissue in the centre of your bones where blood cells are made), which can lead to low levels of red blood cells (**anaemia**) and platelets (**thrombocytopenia**). Some people have a build-up of fluid in their tummy or around their lungs (pleural effusion), which can cause bloating or breathing difficulties.

The lymphoma can also affect how well your **immune system** works so you might find it **harder than usual to fight off infections**.

In some people with AITL and FTCL, abnormal immune cells might produce too many antibodies (also known as 'immunoglobulins'). Antibodies are proteins that B cells usually make to help you fight off infection. If you have AITL or FTCL, you might make abnormal antibodies that react against healthy cells. This is called an 'autoimmune reaction'. It can cause a variety of symptoms, including:

- a low red blood cell count (**autoimmune haemolytic anaemia**), which might make you feel tired or short of breath
- low platelet levels (**immune thrombocytopenia**), which might make you bleed or bruise more easily than normal
- painful, swollen joints
- inflammation of your blood vessels
- thyroid problems.

Most people with AITL have **advanced stage** lymphoma (stage 3 or 4) when they are diagnosed. People with FTCL are more likely to be diagnosed at an earlier stage.

Treatment of AITL and FTCL

Treatment of AITL or FTCL varies from simple treatment with **steroids** to combination chemotherapy.

Several **targeted drugs** are being tested and different chemotherapy regimens (combinations of drugs) are being compared in **clinical trials**. Your doctor might ask you if you would like to take part in a clinical trial to help test new treatments and to find out what the best treatment is for AITL and FTCL.

If you don't want to take part in a clinical trial, or if there isn't one that is suitable for you, you are likely to be treated with **chemotherapy**. The most common chemotherapy **regimen** used for AITL and FTCL is CHOP: cyclophosphamide, doxorubicin (or hydroxydaunorubicin), vincristine (also known as **Oncovin**[®]) and prednisolone.

You might have a chemotherapy drug called **etoposide** in addition to CHOP (CHEOP), or you might have a different chemotherapy regimen altogether.

Some people with AITL or FTCL have a high number of abnormal B cells in their body. If this is the case, you might have **antibody treatment** such as **rituximab** as well as chemotherapy. This destroys B cells.

If you respond to chemotherapy and you are well enough, your doctor is likely to recommend that you have a **self (autologous) stem cell transplant**. This gives you a better chance of staying in remission (no evidence of lymphoma).

If you are not fit enough to have standard chemotherapy, you might have less intensive treatment, such as gemcitabine, bendamustine or cyclosporine.

Relapsed or refractory AITL and FTCL

It is common for AITL and FTCL to come back (**relapse**) after being in remission. Sometimes, AITL and FTCL don't respond to treatment (**refractory lymphoma**). In these cases, your doctor might consider:

- a different **chemotherapy regimen**, such as DHAP (**dexamethasone, high-dose cytarabine** [also known as **Ara-C**] and cisplatin [also known as **Platinol®**]) or ICE (**ifosfamide, carboplatin and etoposide**)
 - a **donor (allogeneic) stem cell transplant** if your lymphoma responds to more chemotherapy and you are well enough
 - a **targeted drug**, usually through a clinical trial.
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Research and targeted treatments

Many new treatments are being tested to see if they can help people with T-cell lymphoma. Drugs that are being tested in AITL and FTCL include:

- **antibody treatments** such as alemtuzumab, which binds to a protein called CD52 on T cells
- **antibody–drug conjugates** such as **brentuximab vedotin**
- **immune checkpoint inhibitors** such as avelumab
- **proteasome inhibitors** such as bortezomib or carfilzomib
- **HDAC inhibitors** such as vorinostat, romidepsin or belinostat
- a chemotherapy drug called **pralatrexate**.

Some of these might be available to you through a clinical trial. If you are interested in taking part in a clinical trial, ask your doctor if there is a trial that might be suitable for you. To find out more about clinical trials or search for a trial that might be suitable for you, visit [Lymphoma TrialsLink](#).

We have separate information about the topics in **bold font**. Please get in touch if you'd like to request copies or if you would like further information about any aspect of lymphoma. Phone 0808 808 5555 or email information@lymphoma-action.org.uk.

References

The full list of references for this page is available on our website. Alternatively, email publications@lymphoma-action.org.uk or call 01296 619409 if you would like a copy.

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