

T-cell lymphomas

This page gives an overview of T-cell lymphomas – non-Hodgkin lymphomas that develop from a type of white blood cell called a T lymphocyte or T cell.

On this page

[What are T-cell lymphomas?](#)

[Who gets T-cell lymphomas?](#)

[Symptoms](#)

[Diagnosis and staging](#)

[Types of T-cell lymphoma](#)

[Outlook](#)

[Treatment](#)

[Follow-up](#)

What are T-cell lymphomas?

Lymphoma is a type of blood cancer that develops when white blood cells called **lymphocytes** grow out of control. Lymphocytes are part of your **immune system**. They travel around your body in your **lymphatic system**, helping you fight infections. There are two types of lymphocyte: **T lymphocytes (T cells)** and **B lymphocytes (B cells)**.

Lymphomas can be grouped as **Hodgkin lymphomas** or **non-Hodgkin lymphomas**, depending on the types of cell they contain. T-cell lymphomas are non-Hodgkin lymphomas that develop from T lymphocytes.

Some T-cell lymphomas develop in the skin. We have separate information on **T-cell skin lymphomas**.

Who gets T-cell lymphomas?

T-cell lymphomas are rare. They account for around 1 in 10 cases of non-Hodgkin lymphoma. Around 700 people are diagnosed with T-cell lymphomas in the UK each year.

Most T-cell lymphomas develop from mature T cells. They usually affect older adults, typically people in their mid-60s. They are more common in men than in women. Occasionally, T-cell lymphoma can develop from immature T cells. This is known as **T-cell lymphoblastic lymphoma**. It tends to affect **children and young adults**.

In most cases, it is not known what **causes** T-cell lymphomas. In a few types of T-cell lymphoma, research has shown that certain viral infections or medical conditions can increase the risk of developing lymphoma.

- **Human T-lymphotropic virus type 1** (HTLV-1) is linked with development of **adult T-cell leukaemia/lymphoma (ATL)**.
- Past infection with Epstein-Barr virus (EBV) is linked to the development of a range of lymphomas, including **angiimmunoblastic T-cell lymphoma (AITL)**.
- **Enteropathy-associated T-cell lymphoma (EATL)** is linked with coeliac disease.

Remember that T-cell lymphomas are very rare and most people with these conditions do not develop lymphoma.

Symptoms of T-cell lymphomas

Symptoms of T-cell lymphomas can vary depending on the particular type of lymphoma you have and where in your body it develops.

One of the most common **symptoms of lymphoma** is a swollen lymph node or nodes. However, many T-cell lymphomas develop outside the lymph nodes or in the organs – for example, in the liver, **bone marrow**, gut or **skin**. Lymphoma that starts outside of the lymph nodes is called ‘extranodal’ lymphoma.

Extranodal lymphoma can cause many different symptoms, such as a swollen liver or spleen, a skin rash, or abnormal **blood counts**. These varied symptoms can make it difficult to diagnose T-cell lymphomas.

People with T-cell lymphomas often experience fevers, night sweats and unexplained weight loss. These three symptoms are known as ‘B symptoms’.

Diagnosis and staging of T-cell lymphomas

T-cell lymphomas are rare and can be difficult to diagnose. Your doctor might consult other centres that have expertise in T-cell lymphoma. In some cases, your doctor might refer you to one of those centres. It is important to find out exactly what type of T-cell lymphoma you have and what parts of your body are affected, so that your doctor can choose the best treatment for you. A **number of different healthcare professionals** are likely to be involved in your care.

T-cell lymphoma is usually diagnosed based on a **biopsy**. A sample of tissue that is affected by lymphoma, such as a swollen lymph node, is removed and examined by an expert lymphoma **pathologist**. The pathologist does tests on the tissue to find out what type of lymphoma you have.

You also have **blood tests** to look at your general health, check your **blood cell counts**, make sure your kidneys and liver are working well and rule out infections that could flare up when you have treatment.

You have other tests to find out which areas of your body are affected by lymphoma. This is called **staging**. Staging usually involves having a **PET scan** and a **CT scan**. Some people, particularly children, may have an **MRI scan**. You might have a sample of your bone marrow cells taken (a **bone marrow biopsy**), to check if you have lymphoma cells in your bone marrow. Rarely, you might have a **lumbar puncture** or MRI scan to check if you have lymphoma in your brain or spinal cord (central nervous system).

As the lymphatic system is all over the body, it isn't unusual for T-cell lymphoma to be at an advanced stage when it is diagnosed. Although this sounds alarming, there are treatment options for advanced stage lymphoma.

You usually have your tests done as an outpatient. It can take a few weeks to get all the results together to be able to confirm the correct diagnosis. **Waiting for test results** can be a worrying time, but it is important for your doctor to gather all of this information in order to plan the best treatment for you.

Types of T-cell lymphoma

There are many different types of T-cell lymphoma. They are sometimes grouped depending on whether they mainly affect lymph nodes (nodal T-cell lymphomas) or other parts of the body (extranodal T-cell lymphomas).

We have dedicated pages outlining the symptoms and treatment for the most common types. They have complicated names based on the type of cell they develop from or the proteins they make.

Nodal T-cell lymphomas include:

- **peripheral T-cell lymphoma not otherwise specified (PTCL-NOS)**
- **angioimmunoblastic T-cell lymphoma (AITL), follicular T-cell lymphoma (FTCL)** and other T-cell lymphomas that develop from follicular helper T cells
- **anaplastic large cell lymphoma (ALCL)** including breast implant-associated T-cell lymphoma.

Extranodal T-cell lymphomas include:

- **intestinal T-cell lymphomas** including enteropathy-associated T-cell lymphoma (EATL) and monomorphic epitheliotropic intestinal T-cell lymphoma (MEITL)
- **extranodal NK/T-cell lymphoma, nasal type**
- **hepatosplenic T-cell lymphoma.**

T-cell lymphomas that can be either nodal or extranodal include:

- **adult T-cell leukaemia/lymphoma (ATL).**

T-cell lymphoblastic lymphoma, which develops from immature T cells, tends to affect younger people. We cover **lymphoblastic lymphoma** briefly on our page about **lymphoma in children**.

T-cell lymphomas that start in the skin behave differently, and are treated differently, from other types of T-cell lymphoma. We have separate information on **T-cell skin lymphomas**.

Outlook of T-cell lymphomas

Your specific outlook depends on the stage of your lymphoma, the exact type of T-cell lymphoma you have, your general health and many other individual factors. Your medical team might use the results of your tests and other individual factors (for example, your age and how fit you are) to calculate a score that helps predict how well you are likely to respond to treatment.

Your doctor is best placed to advise you on your outlook based on your individual circumstances.

Most people do not find it helpful to look at survival statistics. Survival statistics can be confusing as they don't tell you what your **individual** outlook is – they only tell you how a group of people with the same diagnosis did over a period of time. Remember that treatments are improving all the time and survival statistics are usually measured over 5 or 10 years after treatment. This means that statistics only tell you how people did in the past. Those people may not have received the same treatment that you will receive. It also includes the whole range of different outlooks from those who have a poor prognosis and to those who may be long-term survivors.

Treatment of T-cell lymphomas

We have information on the treatment of particular types of T-cell lymphoma on our separate pages.

Lymphoma specialists and research scientists are investigating novel approaches to improve the treatments and outcomes for T-cell lymphomas. Several **targeted drugs** are being tested in **clinical trials**. Your doctor might ask you if you would like to take part in a clinical trial to help test new treatments.

Outside clinical trials, most T-cell lymphomas have in the past been treated with **chemotherapy**. The most common combination chemotherapy regimen that has been used is CHOP:

- cyclophosphamide
- doxorubicin (or hydroxydaunorubicin)
- vincristine (also known as **Oncovin**®)
- prednisolone (a steroid).

Some specialists might add a chemotherapy drug called **etoposide** to CHOP (CHEOP), or you might have a different chemotherapy regimen altogether. If you are not fit enough to have CHOP or CHEOP, you might be treated with less intensive chemotherapy, such as gemcitabine or bendamustine.

Your doctor might recommend a **self (autologous) stem cell transplant** for younger people with T-cell lymphoma who have responded well to initial chemotherapy and are well enough. For some types of T-cell lymphoma, this could give you a better chance of staying in remission (no evidence of lymphoma) after treatment.

Follow-up of T-cell lymphomas

After treatment for T-cell lymphoma, you have regular **follow-up appointments** at the hospital. In general, you are seen every 2 to 3 months when your treatment first ends, although this can vary depending on your particular circumstances and your hospital's policy. Your appointments usually become less frequent over time.

Your follow-up appointments are to check that:

- you are recovering well from treatment
- you have no signs of the lymphoma coming back (relapsing)
- you are not developing any **late effects** (side effects that develop months or years after treatment).

People who stay in **remission** (no evidence of lymphoma) after treatment for T-cell lymphoma are usually followed up for at least 2 years after the end of their treatment. Some hospitals offer follow-up for 5 years or longer. If you have been treated as part of a **clinical trial**, you might be followed up for longer.

Contact your medical team if you develop any **symptoms of lymphoma** or if you have other concerns between your appointments. Your specialist might bring your appointment forward if they think they need to see you sooner.

After your follow-up period ends, your GP usually becomes your main point of contact if you have any concerns or notice anything unusual. Your GP should have a record of your diagnosis and all the treatment you've had. When you visit your GP, it is a good idea to remind them you have been treated for lymphoma in the past so they are aware of any health problems you may be at risk of.

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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