

Anaplastic large cell lymphoma (ALCL)

This page is about anaplastic large-cell lymphoma (ALCL), a type of T-cell lymphoma.

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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.

What is ALCL?

Anaplastic large cell lymphoma (ALCL) is a type of **T-cell lymphoma** – a **non-Hodgkin lymphoma** that develops from white blood cells called **T cells**. Under a microscope, the cancerous cells in ALCL look large, undeveloped and very abnormal ('anaplastic').

There are four main types of ALCL. They have complicated names based on their features and the types of proteins they make:

- ALK-positive ALCL (also known as ALK+ ALCL) is the most common type. In ALK-positive ALCL, the abnormal T cells have a genetic change (mutation) that means they make a protein called 'anaplastic lymphoma kinase' (ALK). In other words, they test positive for ALK. ALK-positive ALCL is a **fast-growing (high-grade) lymphoma**.

- ALK-negative ALCL (also known as ALK- ALCL) is a high-grade lymphoma that accounts for around 3 in every 10 cases of ALCL. The abnormal T cells do not make the ALK protein – they test negative for ALK.
 - Breast implant-associated ALCL is a T-cell lymphoma that develops **extremely rarely** following silicone breast implantation, particularly with textured rather than smooth implants. Although it develops in the breast, it is not a type of breast cancer. It is usually **slow-growing (low-grade)**. Scientists think it might be caused by an inflammatory reaction to the implant.
 - Primary cutaneous ALCL is a low-grade T-cell lymphoma that develops in the skin. We have more information on this type of ALCL on our page about **T-cell skin lymphoma**.
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Who gets ALCL?

ALCL is rare. Each year, fewer than 200 people are diagnosed with ALCL in the UK.

Different types of ALCL usually affect different groups of people:

- ALK-positive ALCL usually affects children and young adults, commonly in their 30s. It affects three times more males than females.
- ALK-negative ALCL tends to affect older adults, typically around 40 to 65 years old. It is slightly more common in men than women.
- Breast implant-associated ALCL is extremely rare. It affects around 80 in every million people who have had breast implants. It is more common in people with implants that have a rough, textured surface than people with smooth implants. It typically develops 8 to 10 years after having the implant but it can develop sooner.

If you are under 25 and have been diagnosed with ALCL, or if you have a child who has been diagnosed with ALCL, you might find our information on **lymphoma in children and young people** useful.

Symptoms of ALCL

People with ALK-positive and ALK-negative ALCL typically have **swollen lymph nodes** and **B symptoms** (fevers, night sweats and weight loss). ALCL is also commonly found outside the lymph nodes (extranodal areas), where it can cause many different **symptoms**.

- Lymphoma affecting the gut (gastrointestinal system) might cause bloating, pain, sickness, diarrhoea or vomiting.
- If you have lymphoma in your chest, you might feel short of breath, develop a cough or have pain or a feeling of pressure in your chest.
- Your **bone marrow** (the spongy tissue in the centre of bones where blood cells are made) might be affected, which can lead to **anaemia** (low red blood cells) and **thrombocytopenia** (low platelet levels).
- You might develop a rash.

Most people with ALK-positive and ALK-negative ALCL are diagnosed at an **advanced stage** (stage 3 or 4), which means the lymphoma affects several parts of the body.

People with breast implant-associated ALCL typically develop a build-up of fluid or a lump around the implant. It can be uncomfortable. This type of ALCL does not usually spread outside the affected breast. It is usually diagnosed at an **early stage** (stage 1 or 2).

My illness had come on quickly at the beginning of the year. I was so exhausted, I was having to drag myself to school to pick my daughter up. I also had a cough that wouldn't go away, a persistently high temperature, swollen lymph nodes and night sweats. I was also out of breath.

Katherine, diagnosed with ALCL in 2016

Treatment of ALCL

The treatment you need depends on what type of ALCL you have.

ALK-positive and ALK-negative ALCL

ALCL is rare. This makes it difficult to determine which treatment gives the best outcome. 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 and to find out what the best treatment is for ALCL.

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 ALCL is CHOP:

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

You might have a chemotherapy drug called **etoposide** in addition to CHOP (CHEOP), or you might have a different chemotherapy regimen altogether. The number of cycles and the exact type of chemotherapy you have depends on several factors like your age, the **stage** of your lymphoma and whether it is ALK-positive or ALK-negative. Some people with early-stage ALK-positive ALCL might also have **radiotherapy** to the affected area.

A targeted drug called **brentuximab vedotin** has recently been licensed, in combination with cyclophosphamide, hydroxydaunorubicin and prednisolone (CHP), to treat adults with ALCL who have not been treated before. However, it is not yet available for this use on the NHS.

Brentuximab vedotin is a strong anti-cancer drug joined to an antibody that binds to a protein called CD30 on the surface of T cells. This carries the drug directly to the T cells and kills them.

If you have ALK-negative ALCL and you respond to chemotherapy, your doctor might 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 have ALK-positive ALCL, you might not need a stem cell transplant.

Breast implant-associated ALCL

If you have breast implant-associated ALCL, you have the implant and any lumps or fluid removed by surgery. This might be all the treatment you need. You are then followed-up carefully to make sure all the lymphoma has been removed. If the lymphoma is more widespread, you are likely to have **chemotherapy**, usually with CHOP.

Relapsed and refractory ALCL

If your lymphoma doesn't respond (refractory) or comes back (**relapses**) after treatment, you might be treated with **brentuximab vedotin**. If brentuximab vedotin is not suitable for you, or if it is not available in your area, you might have a different **chemotherapy regimen** or a **targeted drug**, usually through a clinical trial.

If you respond to more treatment and you are fit enough, you might have a **donor (allogeneic) stem cell transplant** to increase your chance of staying in remission.

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 ALCL include:

- **cell signal blockers**, including a drug called crizotinib, which blocks the activity of ALK, and alisertib, which blocks a protein called Aurora A kinase (AAK)
- **antibody–drug conjugates** that bind to a protein called CD30 on T cells, such as brentuximab vedotin
- **proteasome inhibitors** such as bortezomib
- **HDAC inhibitors** such as vorinostat
- 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**.

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.

Acknowledgements

- With thanks to Prof Tim Illidge, Professor of Targeted Therapy and Oncology and Honorary Consultant Oncologist, University of Manchester, the Christie NHS Foundation Trust, for reviewing this information.
- We would like to thank the members of our Reader Panel who gave their time to review this information.

Content last reviewed: August 2019

Updated April 2020

Next planned review: August 2022

LYMweb0209ALCL2019v3

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