Anaplastic Large Cell Lymphoma (ALCL)

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Learning about your lymphoma can be like learning a new language. It takes time and practice. Please keep this document handy so you can refer back to it as often as you need to. **It will become easier to understand the more you read it.**

Overview

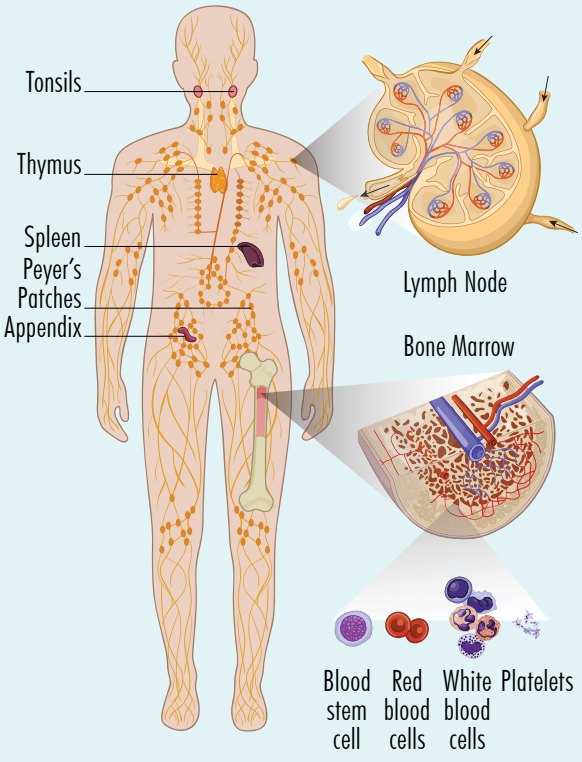
Anaplastic Large Cell Lymphoma (ALCL) is a rare subtype of T-cell Non-Hodgkin Lymphoma. It is cancer that begins in a type of white blood cell, called T-cell lymphocytes (T-cells).

T-cells are specialized immune cells that fight infection and disease, help other immune cells work more effectively, and have an immunological memory. This means they remember infections you had in the past so that if you get the same infection again, you can fight it more effectively and quickly.

When T-cell lymphocytes become cancerous, they do not grow properly. They look and behave very differently to normal T-cells, and are unable to protect you from infection and disease. They can also grow uncontrollably which means they sometimes affect the way other good cells grow, because they can take up so much room that the good cells have no room to grow.

More about T-cells

Unlike other blood cells, most T-cell lymphocytes do not live in our blood. They are made in our bone marrow, the



spongey middle part of our bones, and then they move out of our bone marrow and travel through our lymphatic system to our thymus, where they continue to mature. However, because T-cells are specialized immune cells, they do not stay in the thymus. They can travel to any part of our body including our skin, to fight infection and disease. This means that ALCL can also be found in any part of your body.

What does Anaplastic Large Cell Lymphoma mean?

Anaplastic means the cells look very different from normal healthy T-cells. They are larger than normal cells, but also have not developed in the way a normal cell develops, so they are unable to function like a normal cell would.

Symptoms of ALCL

Some symptoms of ALCL will depend on the subtype of ALCL, and the part of your body that is affected by the lymphoma. Symptoms specific to subtypes will be listed below under the different subtype headings. However, there are some symptoms you could get, with any of the subtypes. They include:

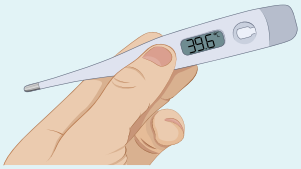
- A lump or lumps (swollen lymph node/s) in your neck, armpit or groin that does not go away after a couple of weeks
- Loss of appetite

B-SYMPTOMS

B-symptoms are a group of three distinct symptoms that some people with lymphoma can get. They often occur together and may indicate that your lymphoma is more advanced. **Contact your doctor as soon as possible if you get B-symptoms.**

Drenching night sweats – where your clothes and bedding become saturated.

Losing weight without trying, and without other reason.



A high fever of 37.5˚ or more that keeps coming back or does not go away even when you don't have an infection. You may even get chills.

- Shortness of breath, cough or pain in your chest
- Stomach or abdominal pain
- Pain
- Fatigue – extreme tiredness that does not improve after a rest or sleep
- Itchy skin
- Changes to your blood counts on a blood test
- B-symptoms (see picture on page 2).

Types of ALCL

There are different subtypes of ALCL including:

- Primary Cutaneous ALCL
- Breast implant associated ALCL
- Systemic ALCL
  - ALK positive systemic ALCL
  - ALK negative systemic ALCL.

Primary cutaneous ALCL is considered an indolent lymphoma. This means it grows relatively slowly, and can go through stages where it is sleeping. It is less likely for indolent ALCL to spread to other parts of your body.

Breast implant associated ALCL can be indolent, or behave more aggressively, and Systemic ALCL is an aggressive subtype of lymphoma and grows quickly and can spread to other parts of your body. Treatment is needed more urgently with aggressive lymphomas.

Primary Cutaneous ALCL (pcALCL) – mainly affects your skin. It is more common in men than women, and usually starts when you're over 50 years of age. It usually does not spread, but if it does spread to areas inside your body including your lymph nodes, you will be given the same type of treatment as someone with systemic ALCL.

Symptoms of pcALCL include one or more lumps on your skin, or an area where your skin breaks down, meaning that you may develop sores that do not heal like you would expect. It may also look more like a rash. If you have any of these symptoms that do not go away after two or three weeks, see your doctor.

Breast Implant-associated ALCL (BIA-ALCL) – This type of lymphoma is only found in people who have had breast implants. It is thought to happen as a result of long-term inflammation. It is more common in people with silicone textured implants, rather than those with a smooth surface.

You may notice a lump or a build-up of fluid around your implant, or it may show up on mammogram or other scan. Although this cancer develops in the breast, it is not a type of breast cancer.

Most people with breast implants will not develop BIA-ALCL, but it's important to talk to your doctor about your risks if you have implants, or are considering getting them.

Your doctor will likely recommend you have your implants removed if you have this subtype of lymphoma. They will be able to talk to you about options and risks of other implants if needed.

Systemic ALK positive ALCL – This is the most common subtype of ALCL and usually affects children and young adults less than 40 years of age. It is fast-growing, and called ALK positive, because the lymphoma cells have a genetic mutation that causes them to make a protein called **Anaplastic Lymphoma Kinase** (ALK). Systemic ALK positive ALCL can start in any part of your body, and you may get any of the symptoms listed above. You may also have a high level of lactate dehydrogenase (LDH) in your blood tests, which indicates the lymphoma cells are growing and expanding quickly.

Systemic ALK negative ALCL – This is similar to ALK positive lymphoma, but your lymphoma cells do not make the Anaplastic Lymphoma Kinase (ALK) protein. It is more common in people who are 40-65 years old. It can be found in any part of your body and is a little more common in men than in women.

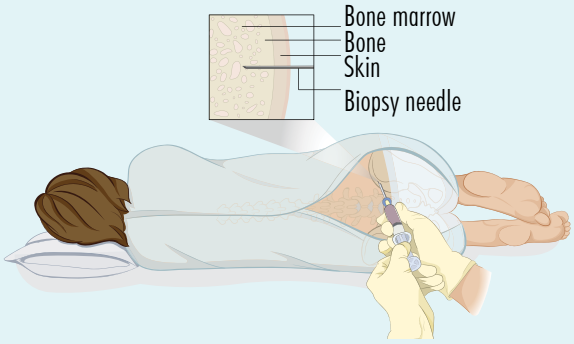
Diagnosis of ALCL

You will need a biopsy to diagnose ALCL. The type of biopsy will depend on where in your body the suspected lymphoma is. If it is affecting your skin, you will need a biopsy of the area of skin, or lump. If your lymph nodes are swollen you will need a lymph node biopsy or, if one of your organs are involved you will need a biopsy of that organ.

Types of biopsy you may have include:

- Core needle biopsy – a needle is used to remove a sample of tissue that may be from your skin, lymph node or breast.
- Excisional biopsy – a whole lymph node is removed.
- Incisional biopsy – a section of your lymph node or tissue is removed.
- Bone marrow biopsy – a needle is used to remove bone marrow from the middle of your bone.

BONE MARROW ASPIRATE AND TREPHINE



Staging ALCL

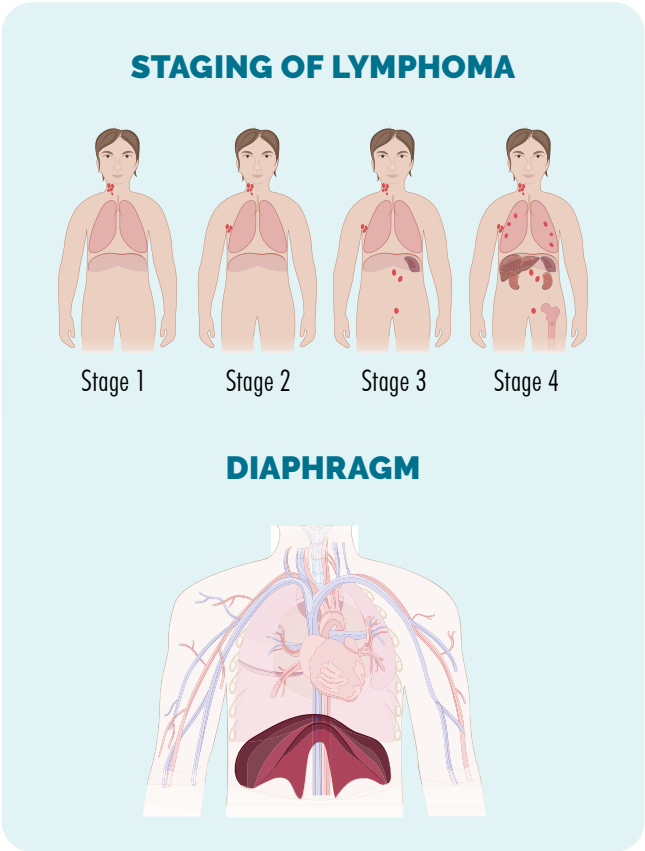
Once your diagnosis is confirmed, you will need more tests to see how much of your body is affected by the lymphoma. These tests are called “staging” and can include:

- PET scan - PET scans provide an image of the inside of your whole body, and lights up areas that are affected by lymphoma. You will be given an injection of a radioactive medicine that any cancerous cells absorb, making them stand out on the PET scan. It takes around 30-60 minutes to do, but you should allow at least 2 hours for the appointment over all.
- CT scan - A CT scan is a scan that can look at the inside of your body and give a 3D image. It is usually used when only a certain part of your body needs to be seen, such as your chest or abdomen. They can provide an image of your body from front to back and top to bottom. Scans are often used to check for tumours, swollen lymph nodes and other conditions.
- MRI - MRI scans use magnets and radio waves to create a picture of the inside of your body. It is similar to a CT scan in that you will lay on a bed and be moved in and out of the MRI machine. However, MRI scans can take longer, and depending on what part of your body is being scanned, can take 15 – 90 minutes (1 and half hours). It is also a very noisy scan as the magnets move around inside the machine.
- Ultrasound - An ultrasound is a scan that uses sound waves to make a picture. The ultrasonographer (person doing the ultrasound) will put some gel over the area being checked, and use a wand-like device to run over your skin, which sends sound waves into your body. As the waves bounce back it creates a picture of the inside of your body.
- X-ray

Staging is classed as stage 1, stage 2, stage 3 or stage 4.

Stage 1 and 2 are considered early-stage lymphoma, and stage 3 and 4 are considered advanced stage.

Stage 1 and 2 are when the lymphoma is only on one side of your diaphragm, the muscle that runs along bottom of your lungs and separates your chest from your abdomen.



Stage 3 and 4 are when the lymphoma is on both sides of your diaphragm, with stage 4 also having lymphoma in at least one of your organs such as your liver, bone marrow, lungs or brain.

You can learn more about the different types of biopsies and scans at our website https://www.lymphoma.org.au/lymphoma/tests-diagnosis-and-staging/

If you do not have access to the internet, you can contact our lymphoma care nurses on 1800 953 081 for more information.

Before you start treatment

Before you start treatment, you will also need to have some baseline tests done. These can include blood tests to check how well your liver and kidneys are working, scans of your heart (echocardiogram), or lung function tests.

Baseline tests are done to make sure that you are well enough to have treatment without it causing you to become too unwell. Throughout your treatment you will have regular blood tests which will be compared to these baseline tests. You may also have further lung tests and heart scans to make sure that the treatment has not caused any damage to your organs.

Questions to ask your doctor

It is important that you ask your doctor any questions you may have before you start treatment. But it can be hard to know what questions to ask. Click here, or scan the QR code at the end of this document to download questions to consider asking your doctor.

Fertility

Some cancer treatments can make it harder to fall pregnant, or to get somebody pregnant. If you (or your child) are planning to have children later in life, talk to your doctor about how to preserve your fertility **before you start treatment.**

Treatment for ALCL

Treatment for ALCL will depend on:

- The subtype and stage of ALCL you have.
- Whether your lymphoma is indolent or aggressive.
- Your overall health and well being.
- Whether you have ever had treatment for the ALCL in the past.
- Your personal preferences once you have all the information about risks and benefits of the different treatments available.

If you have breast implant-associated ALCL you may need surgery to remove the implants, and any signs of infection or lymphoma from the breast. If your lymphoma is early stage, this may be the only treatment you need.

Other types of treatment you may be offered include:

Radiation/radiotherapy

Radiotherapy is a treatment that uses high doses of radiation to kill cancer cells and shrink tumours. If you are having radiotherapy you will have a planning session before you start which helps the radiation therapists work out how to target the radiation to your lymphoma and avoid damaging nearby healthy tissue. It is usually given every day Monday to Friday and can last between three and five weeks. You may or may not have radiation treatment with chemotherapy.

If you live far away from the radiation centre and you need help with accommodation, **talk to your doctor or nurse about what help is available to you.** You can also **contact the Cancer Council or Leukaemia Foundation in your state** to see if they can help with somewhere to stay.

Monoclonal antibody (MAB) and antibody-drug treatments

MABs are a type of treatment that help your own immune system to fight the lymphoma more effectively. They are given by a drip and move through your bloodstream to the lymphoma cells and attach to these cells. By attaching to these lymphoma cells, MABs attract your own immune cells to the lymphoma so they can begin to attack. In ALCL the MAB attaches to a protein called **CD30** that is found on your lymphoma cells. It is then able to move inside the cancerous cell and kill it from the inside.

Brentuximab vedotin is a MAB that targets CD30 and may be given alone, or with chemotherapy. It is a special type of MAB called a conjugated MAB because the monoclonal antibody (brentuximab) is joined to a toxic drug (vedotin) and delivers the vedotin into the cancerous cells causing them to be destroyed from the inside.

Chemotherapy (chemo)

Chemotherapy are types of medications that kill fast-growing cells. Because they kill fast-growing cells, they can be very effective at treating lymphoma. Unfortunately, chemotherapy cannot tell the difference between healthy cells and lymphoma cells, so you can get unwanted side-effects from chemo. These can include hair loss, a sore mouth, nausea and vomiting, diarrhea or constipation. Chemotherapy you may be offered include:

- **CHOP** – a protocol including chemotherapy medications called cyclophosphamide, doxorubicin and vincristine, plus a steroid called prednisolone.
- **CHEOP** – a protocol with the same medications as CHOP, plus an extra chemotherapy called etoposide.
- **BV-CHP** – a protocol including the conjugated monoclonal antibody

brentuximab vedotin (BV), plus the chemotherapy medicines cyclophosphamide, doxorubicin and the steroid prednisolone.

- **Pralatrexate** – a chemotherapy on its own.

Watch and Wait

If you have an indolent ALCL and are not having any troubling symptoms, you may not need to have any active treatment. Instead, you will be on active monitoring – called Watch and Wait. This is where you still see your doctor regularly and have tests to make sure the lymphoma is not growing or spreading, and that you are not getting any symptoms.

By having these regular check-ups, your doctor will be able to pick up early if the lymphoma starts to become more aggressive, and will be able to start you on treatment as soon as it would be beneficial for you.

Watch and Wait is more common if you have pcALCL or BIA-ALCL and have previously had surgery to remove the lymphoma.

Topical treatments – include creams that are rubbed into the skin lesion. These may be used if you have pcALCL.

Intralesional steroids – Steroids are poisonous to lymphocytes, including those that have become cancerous. If your skin lesion (or rash) has not responded to topical treatments, you may have an injection directly into the lesion. These may be used if you have pcALCL.

Stem cell transplant

You may be offered a stem cell transplant if you have **Systemic ALK negative ALCL**. Stem Cell transplants are complex treatments that involve you having high doses of chemotherapy followed by an infusion of stem cells. Stem cells are the most immature type of blood cell that can develop into any type of blood cell you need. The stem cells you receive may be your own stem cells, or they may come from a donor. You can learn more about stem cell transplants at https://www.lymphoma.org.au/lymphoma/treatments/stem-cell-transplants/.

Clinical Trials

Clinical trials are important because they help find new treatments. They can offer you the opportunity to try something new for free, that you would otherwise not have access to. Some clinical trials may look at:

- how to treat ALCL
- safe doses of new medications or new combinations of existing medications to treat ALCL with less side-effects
- managing side-effects
- improving quality of life.

It is always worth asking your doctor what clinical trials you are eligible for to see if any are of interest to you. You can also look online for clinical trials yourself at the ClinTrial Refer website https://clintrialrefer.

org.au/. If you would like more information on clinical trials, please see our fact sheet Understanding Clinical Trials here.

Refractory or Relapsed ALCL

Sometimes, your lymphoma may not improve with treatment. When this happens, your lymphoma is called refractory. In other cases, your lymphoma may improve and you will go into remission – meaning there is no more signs of lymphoma in your body. But after some time – and this varies between people - the lymphoma may come back. This is called a relapse.

If your lymphoma is refractory or relapses, you will need more treatment. You may be offered the same treatment again if you have been in remission for a long time, or you may be offered a different type of treatment, or a clinical trial. Your doctor will be able to talk to you about the best options for you.

Follow up

Once you finish treatment, you will still see your specialist doctor on a regular basis. How often will depend on your personal situation. Your doctor will let you know how often they want to see you. The purpose of these appointments is to:

- See how you are coping after finishing treatment – both mentally and physically.
- Check that your body is recovering from the effects of the treatment.
- Monitor you for any signs of lasting or delayed side-effects.
- Monitor you for signs of relapse (lymphoma coming back), so that any further treatment can start early.

Summary

- There are different subtypes of ALCL and your symptoms may depend on your subtype, and where the lymphoma is in your body.
- There are different types of treatment for ALCL and the best option for you will depend on your subtype, overall health, stage and other factors.
- If you get B-symptoms you need to contact your doctor.
- Primary cutaneous ALCL is an indolent lymphoma.
- Breast implant associated ALCL may be indolent or behave more aggressively.
- Systemic ALCL may be ALK positive – which is more common in children and people less than 40 years, or ALK negative, which is more common in people 40-65 years of age – Both are aggressive lymphomas.

Resources and support

Lymphoma Australia offers a wide range of resources and support for people living with lymphoma or CLL, and their carers. How to access our resources:

- **Visit** our website www.lymphoma.org.au

for more information.

- **Phone** our Lymphoma Care Nurse Hotline on 1800 953 081.
- **Email** our Lymphoma Care Nurses nurse@lymphoma.org.au
- **Booklet:** Understanding Non-Hodgkin Lymphoma (NHL)
- **Downloadable information:** Visit our website, or give us a call if you would like some more information on a variety topics related to lymphoma
- **Join** our Facebook page Lymphoma Down Under (make sure you complete all the membership questions when you join).

Cancer Council offers a range of services, including free counselling, to support people affected by cancer, including patients, families and friends. Services may be different depending on where you live. You can contact them at www.cancer.org.au or by phone on 13 11 20.

Medicare Australia: Check with your GP if you are eligible for a Mental Health Treatment Plan (MHTP). This plan is funded by Medicare and can provide you with up to 10 sessions with a registered psychologist. More information can be found here.

WeCan is an Australian supportive care website to help find the information, resources and support services you may need following a diagnosis of cancer. You can visit their website at www.wecan.org.au.

Canteen provides support for young people aged 12-25 years who have cancer, or, who have a parent with cancer. Find out more at their website here www.canteen.org.au.

Health Translations: A collection of health related information collected by the Victorian Government with resources in different languages. You can visit their website at www.healthtranslations.vic.gov.au.

Useful links

ALCL website

Bone marrow biopsy

Definitions

Health translations

Lymphoma Australia

Tests diagnosis staging

SCAN ME

Treatments

SCAN ME

Understanding clinical trials

SCAN ME

Disclaimer: Lymphoma Australia has taken every precaution to make sure the information in this document is accurate and up-to-date. However, this information is intended for educational purposes only and does not substitute for medical advice. If you have any concerns about your health or wellbeing, please contact your treating team.

Lymphoma Australia

Notes