Adult T-cell Leukemia Lymphoma (ATLL)

Lymphoma Australia
Nurse hotline: 1800 953 081
nurse@lymphoma.org.au

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.

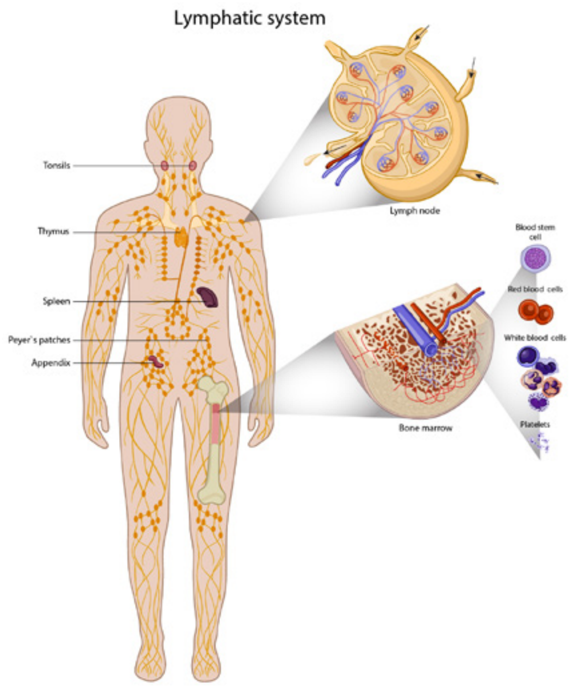
Adult T-cell Leukemia Lymphoma (ATLL) is a rare subtype of T-cell Non-Hodgkin Lymphoma. It can be fast growing (aggressive) or slow-growing (indolent). It starts when your white blood cells, called T-cell Lymphocytes, become cancerous.

These cancerous T-cells can be found in your blood and bone marrow, which is why it has the word leukemia in it, but also in your lymphatic system, which is why it is a subtype of lymphoma.

This is still one cancer – but the cancerous T-cells are found in both your blood and lymphatic system.

We don't know what causes ATLL, but you have a higher chance of developing ~~ATLL~~ if you have had an infection with a virus called Human T-Cell Lymphotropic Virus 1 (HTLV-1). However, most people with HTLV-1 will never develop ATLL (less than 5 people in every 100 with HTLV-1)

ATLL is more common in men than women, though some women develop it too.



About T-cell Lymphocytes

Although T-cell lymphocytes are a type of blood cell, they live mostly in our lymphatic system rather than in our blood. They are made in our bone marrow – the spongey, middle part of bones, but then move out, into our lymphatic system and to an organ in the middle of our chest behind our breastbone, called our thymus (see picture). Once in our thymus, T-cells continue to grow and mature.

Some T-cells are also found in our lymph nodes and spleen. However, because of their specialised function of fighting infection and disease, they can travel to any part of our body, including our skin, blood, and different organs.

There are also patches of **lymphoid tissue** – including lymphocytes in different areas of our body, such as in our stomach or bowels – These areas are called 'Peyer's Patches' (see the Lymphatic system picture).

Subtypes, Signs & Symptoms

There are different subtypes of ATLL, most are fast-growing (aggressive) but rarely they can be slow-growing (indolent). It is important that you understand what subtype you have, so you can better understand what to expect, what questions to ask, and what treatments might work best for you.

Ask your doctor what subtype of ATLL you have, and what it means for your treatment.

	Acute ATLL	**Lymphomatous ATLL**	**Chronic ATLL**	**Smoldering ATLL**
Behaviour	Aggressive	Aggressive	Indolent	Indolent
Symptoms	Fatigue – tiredness that doesn't go away with rest or sleep. Rash on your skin. Swollen lymph nodes in your neck, armpits or groin. Constipation. Changes in your blood tests including – High white blood cell count and high calcium levels. B-symptoms	Fatigue – tiredness that doesn't go away with rest or sleep. Rash on your skin. Swollen lymph nodes in your neck, armpits or groin. May or may not have changes to blood tests such as high white blood cells. B-symptoms.	Fatigue – tiredness that doesn't go away with rest or sleep. Rash on your skin. Swollen lymph nodes in your neck, armpits or groin. High lymphocytes in blood tests. Swollen (enlarged) spleen or liver.	Generally very mild symptoms. May include some skin lesions or rash.

High calcium levels is called hypercalcemia and can cause changes to your heart rhythm, muscle~~s~~ twitches, confusion and constipation

Diagnosis

You will need a biopsy to diagnose ATLL. A biopsy is a procedure to remove part or all of an affected lymph node, a sample of your bone marrow or other area affected by the lymphoma. Once the sample is taken it is sent to pathology where scientists check for cancerous changes to your cells.

You will have either a local or general anaesthetic when you have the biopsy. A general anaesthetic is given to you in the operating room if your biopsy is being taken from a lymph node or organ deep inside your body. These anaesthetics are given so you sleep through the procedure.

You will feel no pain during the biopsy, but may wake up with a small wound and stitches (sutures).

A local anaesthetic is usually used to numb the area the biopsy is being taken from if it is being taken from your bone marrow or lymph nodes close to your skin.

Young children will have a general anaesthetic regardless of where the biopsy is being taken from so that they can remain still and safe during the procedure.

Your nurse will talk to you about when you will have the stitches removed and how to keep your wound clean when you are at home. They will also let you know how to manage any pain ~~if you get any~~.

Staging

If your biopsy comes back positive for ATLL, which means you do have this type of lymphoma, your doctor will organise more tests to see if it has spread to other parts of your body. These may include:

- Positron Emission Tomography (PET) scan
- Computed Tomography (CT) scan
- Bone Marrow Biopsy

Once all your test results are back your doctor will be able to tell you:

- what subtype you have
- if it is indolent or aggressive
- if it is high, standard or low risk, and how this will affect your treatment
- if you need treatment - what treatment will likely work best for you.

If you haven't had this conversation with your doctor, make sure you ask them to explain these things to you.

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 on your heart, 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 these 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. We have put together some questions to get you started. Click here, or scan the QR code at the end of this document to download Questions to ask 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.

Treatment

Watch and Wait

If you have an indolent ATLL 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.

Indolent ATLL is very rare.

First-line Treatment

You will need to start treatment soon after being diagnosed if your subtype of ATLL is aggressive. These can include:

- Radiotherapy
- Topical steroids (creams) if your skin is affected

Or a combination of chemotherapy medications such as:

- CHOP – cyclophosphamide, doxorubicin, vincristine and a steroid called prednisolone or,
- CHOEP – same medications as CHOP but with an extra one called etoposide or,
- Dose-adjusted EPOCH – same medications as CHOEP but may use different timing and doses
- BV-CHP (if your lymphoma cells have a protein called CD30 on them) - brentuximab vedotin, cyclophosphamide, doxorubicin, prednisolone
- Hyper-CVAD – given as part A alternating with part B
    - Part A -cyclophosphamide, vincristine, doxorubicin and a steroid called dexamethasone alternating
    - Part B – high dose methotrexate and cytarabine
- High dose chemotherapy followed by a Stem cell transplant – click here or scan QR code at end of document for more information.
- Clinical trials

Second-line Treatment

In some rare cases, your ATLL may not get better with the first-line treatment. When this happens it is called refractory.

In other cases your ATLL may get better and you go into "remission", which is where you no longer have ATLL in your body. But sometimes, after a time of remission the ATLL can come back – which is called a relapse.

If your ATLL is refractory or relapses, you will be offered different chemotherapy treatments which can include:

- **DHAP** – cytarabine, cisplatin and a steroid called dexamethasone
- **ESHAP** – etoposide, cytarabine, cisplatin and dexamethasone
- **GDP** - gemcitabine, cisplatin and dexamethasone
- **ICE** – ifosphamide, carboplatin and etoposide
- **Gemcitabine and Vinorelbine** with pegfilgratim
- **Pralatrexate**
- **Romidepsin**

For more information on these treatment protocols please see the eviQ Treatments QR code at the end of this document.

Note: The chemotherapy protocols listed in this document are an example of some of the more common treatments. The protocol your doctor suggests for you may differ from these and will be based on your individual circumstances such as your

- **Age**
- **Overall well-being**
- **Other medications you may be taking**
- **Own preferences once you have all the information you need to make a decision.**

Clinical Trials

Clinical trials are an important way to find new medicines, or combinations of medicines to improve treatment or lessen side-effects. They may also offer you the opportunity to try treatment options that you would not otherwise be able to access.

New clinical trials are starting all the time to try and find better ways to treat lymphoma, while making sure you continue to have the best quality of life. If you are interested in joining a clinical trial, ask your doctor what clinical trials are available to you.

You can also read our "Understanding clinical trials" fact sheet for more information, and links to websites that you can visit to find a clinical trial yourself.

Follow up

Finishing treatment can be a time of mixed emotions. You may feel relieved and excited, or you may feel worried and scared. You may even alternate between all of these emotions. This is very normal. However, you will not be alone.

You will continue to see your specialist team on a regular basis, and be checked for any signs and symptoms of your lymphoma relapsing. Your doctor will also want to make sure you're not having any late or long lasting side-effects from your treatment.

Your doctor will let you know how often they want to see you, however the longer time you are in remission the less often they will need to see you.

Contact your health care team if you have any concerns or worries. You can also contact our lymphoma care nurses on **1800 953 081** or email them on nurse@lymphoma.org.au.

Summary

- ATLL is a subtype of T-cell lymphoma.
- There are different types of ATLL – most are aggressive. It is very rare to have an indolent ATLL.
- If you don't already know, ask your doctor what subtype of ATLL you have.
- Not everyone with ATLL will need treatment, though most people will. If you have an aggressive ATLL you will need treatment soon after diagnosis.
- If you need treatment it may include radiotherapy, topical treatments, chemotherapy or stem-cell transplant.
- You are not alone, if you need support, call our Lymphoma Care Nurses.

Additional Links

<QR codes will go here>

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 at www.canteen.org.au.

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.

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