

Chronic Lymphocytic Leukemia (CLL) and Small Cell Lymphoma (SLL)

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

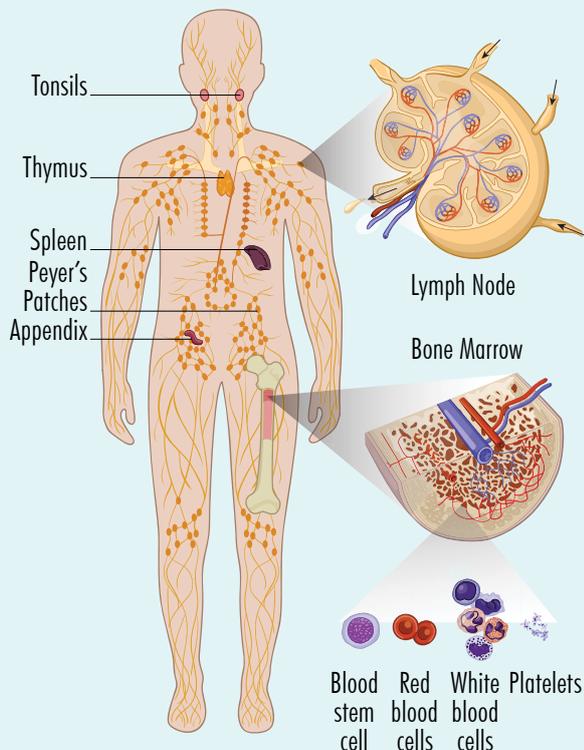
Learning about your lymphoma can be like learning a new language. It takes time and practice. Please keep this document handy so you can refer back to it as often as you need to. **It will become easier to understand the more you read it.**

Introduction

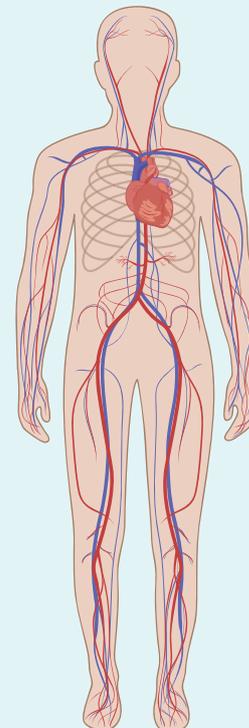
If you've been diagnosed with Chronic Lymphocytic Leukemia (CLL) or Small Lymphocytic Lymphoma (SLL), you have

a type of slow growing blood cancer. This blood cancer affects a type of white blood cell called a B-cell lymphocytes (B-cells).

LYMPHATIC SYSTEM - WHERE SLL STARTS



CIRCULATORY SYSTEM - WHERE CLL STARTS



B-cells fight infection and diseases to keep you healthy. They remember infections you had in the past, so if you get the same infection again, your immune system can fight it more effectively. When these cells don't grow or work properly, you can be diagnosed with CLL or SLL. This means you will have cancerous B-cells called lymphoma cells. Lymphoma cells can travel to any part of your body.

CLL and SLL are basically the same disease. The difference between them is where the lymphoma cells are located. When lymphoma cells are mostly in the bloodstream and bone marrow, it is called CLL. When the lymphoma cells are mostly in the lymphatic system, it is called SLL.

Symptoms

You may not have symptoms at first. Many people are diagnosed when they have a blood test, or a physical exam for something else. But if you do have symptoms, they might include:

- feeling unusually tired (fatigued)
- feeling out of breath
- bruising or bleeding more easily than usual
- infections that don't go away, or keep coming back (recurrent)
- sweating at night more than usual
- losing weight without trying
- a new lump in your neck, under your arms, your groin, or other areas of your body.

Diagnosis and Staging

You may only need a blood test to diagnose CLL/SLL. But you may also need a biopsy to check for changes in your genes. A biopsy is a procedure to remove part, or all of an affected lymph node or bone marrow. The bone marrow is the spongy part inside your bones where your blood cells are made. If you have a biopsy, you may have a local or general anaesthetic. This will depend on what part of your body the biopsy is taken from.

Your doctor will organise extra tests, to check if the lymphoma has spread to other parts of your body.

Staging tests

Staging tests may include:

- [Positron Emission Tomography \(PET\) scan](#)
- [Computed Tomography \(CT\) scan](#)
- [Bone Marrow Biopsy](#)
- [Lumbar Puncture](#)

You can find more information on staging tests at our website. Just look down the left-hand side to find the test you want more information on: <https://www.lymphoma.org.au/lymphoma/tests-diagnosis-and-staging/>.

Cytogenetic tests

This is where your blood and tumour sample is checked for genetic variances

that may be involved in your disease. All patients with CLL/SLL should have genetic testing before starting treatment. These include:

- IgHV mutation status: **before the first treatment** (does not change over time)
- FISH: (13q, 17p, Trisomy 12) **before first and every treatment** (can change over time)
- TP53 mutation status: **before first and every treatment** (can change over time)

Cytogenetics are tests done on your blood and biopsies to look for changes in your chromosomes or genes.

We usually have 23 pairs of chromosomes, but if you have CLL/SLL your chromosomes may look a little different. On your chromosomes are many genes that tell the cells of your body how to look or act. If there is a change in these chromosomes or genes, you can develop different diseases.

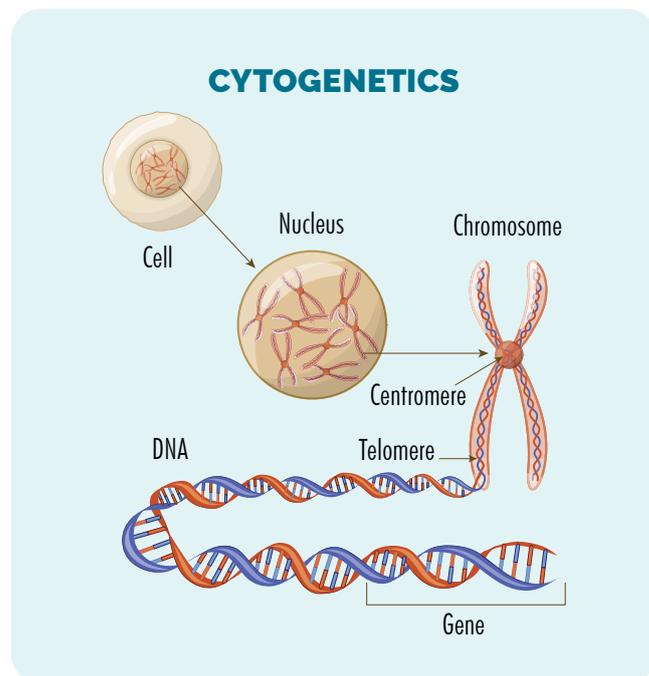
Types of cytogenetic changes in CLL/SLL

The three main changes that can happen with CLL/SLL are called a deletion, a translocation and a mutation.

A deletion is when part of your chromosome is missing. If your deletion is a part of the 13th or 17th chromosome it is called either "del(13q)" or "del(17p)". The "q" and the "p" tell the doctor which part of the chromosome is missing. It is the same for other deletions.

If you have a translocation, it means that a small part of two chromosomes - chromosome 11 and chromosome 14 for example, swap places with each other. When this happens, it's called "t(11:14)".

If you have a mutation, it may mean you have an extra chromosome. This is called Trisomy 12 (an extra 12th chromosome). Or you may have other mutations called IgHV mutation or Tp53 mutation. All these changes can help your doctor work out the best treatment for you, so please make sure you ask your doctor to explain your individual changes.



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. These are 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.

Fertility – some cancer treatments can make it harder to fall pregnant, or to get somebody pregnant. If you are planning to have children, talk to your doctor about how to preserve your fertility

Questions for your doctor before you start treatment

Starting treatment can feel overwhelming, and even knowing what questions to ask can be difficult. To help get you started, we have put together some questions you may like to consider asking. Click [here](#) to download our Questions to ask your Doctor" factsheet or scan the QR code at the end of this document.

Treatment Options

Because CLL/SLL are generally slow growing cancers, you may have it for years without any signs or symptoms. When you are diagnosed, you still may not need any treatment at all. Many people will only need to see their doctor and have a check-up (including some blood tests

or scans) to watch if the CLL/SLL starts growing more quickly. This is called 'watch and wait'.

Some patients call this "watch and worry", because it can be uncomfortable not doing anything to fight the cancer. But, watch and wait is a great way to start. It means your own immune system is fighting the cancer and doing a good job keeping it under control. If your immune system is keeping it under control, you will not need extra help to fight the cancer. Extra medicine that can make you feel quite sick or cause long term side effects, are unlikely to help at this point.

Research shows there is no benefit starting treatment early, when your type of cancer will not respond. Your health will not be improved, and you will not live longer by starting treatment earlier. If your CLL/SLL starts to grow more, or you start to get symptoms from your disease, you might start treatment.

Starting active treatment

If you need treatment, it's important to have genetic testing done before you start. These tests tell your doctor what treatment might work best for you. There are many choices available to treat CLL/SLL and more medicines are being developed and tested in clinical trials. Your doctor will consider several things when

making choices about your treatment. These include:

- your genetic changes
- how fast your lymphoma is growing
- how bad your symptoms are
- your age
- your overall health

Treatments can include:

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 actively growing lymphoma and CLL. 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.

Monoclonal Antibody (MAB) – Given as an infusion in a cancer clinic or hospital. MABs attach to the lymphoma cell and attract other disease fighting white blood cells and proteins to the cancer so your own immune system can fight the CLL/SLL.

Targeted therapy – taken as a tablet either at home or in hospital. Targeted therapies attach to the lymphoma cell and block signals it needs to grow and produce more cells. This stops the cancer from growing and causes the lymphoma cells to die off.

How your cytogenetics can affect your treatment options

If you have a normal IgHV - also called an **unmutated IgHV**, or if you have **17p deletion**, or a **mutation in your TP53 gene**, your CLL/SLL **will not respond to chemotherapy**, but it may respond to one of these treatments instead:

- Ibrutinib - a targeted therapy called a BTK inhibitor
- Acalabrutinib - a targeted therapy (BTK inhibitor)
- Venetoclax & Obinutuzumab - venetoclax is a type of targeted therapy called a BCL-2 inhibitor, obinutuzumab is a monoclonal antibody
- Idelalisib & rituximab - idelalisib is a targeted therapy called a PI3K inhibitor, and rituximab is a monoclonal antibody
- You may also be eligible to participate in a clinical trial – Ask your doctor about this.

If you have a **mutated IgHV**, or variation other than the ones above, you may be offered standard treatments for CLL/SLL including chemo. Depending on what markers your lymphoma has, you may also get a monoclonal antibody. There are a few different choices your doctor can choose from if you have a **mutated IgHV** and these include:

- Fludarabine, cyclophosphamide & rituximab (FC-R). Fludarabine and cyclophosphamide are chemotherapy and rituximab is a monoclonal antibody

- Chlorambucil & Obinutuzumab - chlorambucil is a chemotherapy tablet and obinutuzumab is a monoclonal antibody
- Chlorambucil - a chemotherapy tablet
- Bendamustine & rituximab (BR) – bendamustine is a chemotherapy and rituximab is a monoclonal antibody. They are both given as an infusion.
- You may also be eligible to participate in a clinical trial.

The first time you start treatment it is called first-line treatment. Once you finish your first-line treatment, you may not need treatment again for weeks, months or even years. If your CLL/SLL gets to a point where you need treatment again, it is called a “relapse”. But some people may not respond to their first line treatment. If your CLL/SLL does not respond to the treatment, it is called refractory disease. In these cases, you may need to start a different type of treatment.

First relapse, or refractory disease – Second line treatment

CLL/SLL usually respond well to treatment, but it is very common for these types of lymphoma to come back (relapse). If it does, you may need a different type of treatment. Recommended standard treatments for relapsed/refractory CLL/SLL include:

- Venetoclax & rituximab - venetoclax is a targeted therapy and rituximab is a monoclonal antibody

- Ibrutinib – a targeted therapy (BTK inhibitor)
- Acalabrutinib - a targeted therapy (BTK inhibitor)
- Bendamustine (not PBS listed) - a chemotherapy
- Idelalisib & rituximab - idelalisib is a targeted therapy (PI3K inhibitor) and rituximab is a monoclonal antibody
- Allogeneic stem cell transplant (Bone marrow transplant using a healthy donors cells – only suitable for some patients)
- You may also be eligible to participate in a clinical trial.

Further information on different treatment protocols can be viewed [here](#).

Clinical Trials

Clinical trials are important to find new medicines, or combinations of medicines to better treat people with CLL/SLL in the future. They may also offer you the opportunity to try a new medicine, before it would otherwise be available. If you are interested in participating in a clinical trial, ask your doctor what clinical trials are available to you. You can also read our ['Understanding Clinical Trials' Fact Sheet](#) to find a clinical trial.

Some medications being tested for CLL/SLL in clinical trials include:

- Zanubrutinib
- Umbralisib

- Ublituximab
- Pembrolizumab - an Immune Checkpoint Inhibitor currently used effectively with some other cancers
- Chimeric Antigen Receptor (CAR) T-cell therapy

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 CLL/SLL relapsing, and your doctor will also want to make sure you're not having any 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.

If you have any concerns or worries please contact your healthcare team or contact our lymphoma care nurses on 1800 953 081. You can also email us on nurse@lymphoma.org.au.

Summary

- CLL and SLL are both indolent blood cancers affecting your B-cell lymphocytes - immune cells that fight infection and disease.

- CLL starts in your blood or bone marrow, while SLL starts in your lymphatic system. In every other way, CLL and SLL are the same disease with the same diagnostic tests and treatment.
- You may not need treatment for your CLL/SLL - but you will be actively monitored while on watch and wait.
- Ask your doctor about the cytogenetics of your CLL or SLL.
- If you have an unmutated IgHV, 17p deletion or mutation in your TP53 gene, you will not respond to chemotherapy, so targeted therapies or monoclonal antibodies will work best for you.
- If you have none of the above mutations, you may be offered standard chemotherapy with a monoclonal antibody.
- Many people live a long and healthy life with CLL/SLL.

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:** Living with CLL/SLL

- **Downloadable information:** Visit our [website](#), or give us a call if you would like some more information on a variety of 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

Questions to ask your Doctor



CLL website



Definitions



Facebook – Lymphoma Down Under Patient Group



Health translations



Treatments Website



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.



