Waldenstrom's Macroglobulinemia (WM)

Lymphoma Australia
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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.**

Introduction

Waldenstrom's Macroglobulinemia (WM) is a rare sub-type of non-Hodgkin lymphoma. If you have WM, you have a type of slow-growing blood cancer. WM affects a type of white blood cell called B-cell lymphocytes (B-cells).

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 lymphoma. This means you will have cancerous B-cells called lymphoma cells.

B-cells are made in your bone marrow (the spongy part in the middle of your bones) but usually live in your lymph nodes and spleen. They can travel to any part of your body to fight infection.

B-cells fight infection by producing proteins called immunoglobulins. Immunoglobulins are sometimes called antibodies. The antibodies recognise infection or disease, and either fight it themselves or, draws other cells to the area to fight the infection or disease. WM happens when the B-cells in your bone marrow become cancerous, and produce too much of an Immunoglobulin called "Macroglobulin" or IgM.

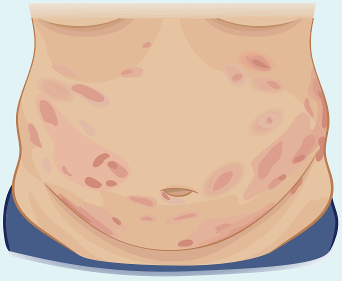
Macroglobulinemia means you have too much IgM in your blood and bone marrow. The high level of IgM can crowd your bone marrow so much, that your healthy cells do not have room to grow. As a result, you may have low levels of your healthy cells. The high number of cancerous B-cells and antibodies can make your blood thicker than normal.

Signs and Symptoms

Some of the healthy cells affected can include your red blood cells. Red blood cells carry oxygen around your body, so when these are low you can get tired, short of breath and feel dizzy.

Your platelets may also be affected. Platelets stop you from bleeding or bruising too much. When these are low you may notice you bruise and bleed more easily. You may even develop a spotty purple/ red rash though this is quite rare.

ABDOMEN RASH



Your B-cells are not your only infection fighting white cells. You also have other white cells made in your bone marrow that can be affected. Because your white cells fight infection, you might have trouble getting rid of an infection, or you might get lots of different infections.

Other symptoms you may experience include:

- tingling, numbness or pain in your fingers and toes (this is called peripheral neuropathy)
- headaches
- bleeding from your nose or gums
- weight loss
- fevers
- muscle cramps
- difficulty concentrating or remembering things.

However, because WM is a slow growing blood cancer, you may not have any symptoms at all.

Diagnosis and Staging

You may be diagnosed with WM after you have a blood test. But your doctor will want to do some more tests to confirm this, and look at what parts of your body are affected. Some of these tests will include:

- Blood tests and urine (wee) tests
- Computed tomography (CT) scan
- Bone marrow biopsy
- Tests on your heart, liver and kidneys.

Treatment

If you have a slow growing WM, you may not need treatment, but you will see your specialist doctor regularly. This time can be called 'Watch and Wait", because your doctor will continue to monitor your levels of WM cells and IgM levels.

Some patients call this "watch and worry", because it can be uncomfortable not doing anything to fight the cancer. However, watch and wait is a great way to start. It means your own immune system is fighting the cancer and doing a better job keeping it under control than any current treatment could do. 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 WM becomes more fast-growing, or starts causing you symptoms, you may need treatment. The treatment choices your doctor offers you will depend on:

- how quickly your WM cells are growing and producing IgM
- how bad your symptoms are
- how thick your blood is
- if your healthy blood cells or organs are being affected
- your age
- your overall health.

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 later in life, talk to your doctor about how to preserve your fertility.

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.

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 WM. 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 WM.

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. For more information on these treatments, please see our oral therapies factsheet here.

Plasma Exchange – A procedure where you will have a drip put in your arm, and be connected to a machine called an apheresis machine. This machine will remove thickened blood from your body. It will then filter out the bad cells and antibodies, remove them and return your good cells along with some replacement plasma into you. Plasma is the watery part of your blood that carries some of your antibody producing B-cells called Plasma Cells. This is done if your blood becomes too thick with all the extra IgM antibodies.

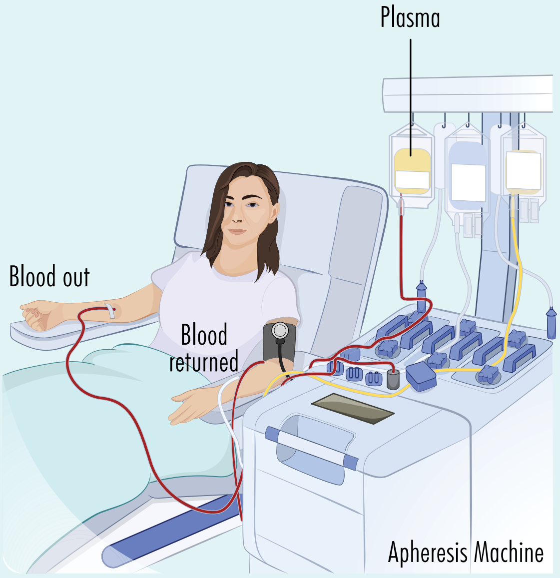
First-line Treatment

The first time you start treatment it's called first-line treatment. Once you finish your first-line treatment, you may not need treatment again for weeks, months or even years. Some people will not need treatment again at all. Some first-line treatments you may be offered include:

- **BR** – Bendamustine (chemo infusion) and rituximab (monoclonal antibody).
- **DRC** – Rituximab - monoclonal antibody (or a medication similar to rituximab called a biosimilar), cyclophosphamide (chemo tablet) and a corticosteroid called dexamethasone.
- **Zanubrutinib** (target therapy – BTK inhibitor) – If you can't have chemotherapy or a monoclonal antibody.

*BTK is an enzyme that helps WM to grow. By blocking this enzyme with BTK inhibitors, the WM cannot grow.

APHERESIS



Step 1: Blood is removed from your IV line in your arm, or from your central line.

Step 2: Blood is processed in the apheresis machine and the plasma is separated and removed from your blood.

Step 3: Blood is returned to you (Minus the plasma). You will receive new plasma from a donor.

- **Apheresis - Plasma exchange (plasmapheresis)** – This may be offered if your blood has become too thick – this helps to improve symptoms, rather than treat your WM.
- You may also be offered to join a clinical trial – More information on clinical trials is below.

Second-line Treatment

If your WM gets to a point where you do need treatment again, it is called a "relapse". Treatment after a relapse is called second-line treatment (or third-line etc). However, some people may not respond to their first line treatment. If your WM does not respond to the treatment, it is called "refractory" disease. In these cases, you may need to start a different type of treatment. If you have refractory disease, and start a new treatment, this is also called second-line treatment.

The type of second-line treatment you are offered will depend on a few things. Your doctor will consider how long you have been in remission for, how well you feel and what symptoms you have.

You may be offered the same treatment you had for first-line treatment if you have been in remission, or not needed treatment for more than 5 years. If it has been less than 2 years since you last had treatment, then your doctor might want to try something different. Some options include:

- **Rituximab** – monoclonal antibody (or a medication similar to rituximab called a biosimilar) with one or more different types of chemotherapy or zanubrutinib (targeted therapy – BTK inhibitor).
- **Plasma exchange** – This may be offered if your blood has become too thick – this helps to improve symptoms, rather than treat your WM.
- You may also be offered to join 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 improve treatment of WM in the future. They can also offer you a chance to try a new medicine, or new combinations, before it would otherwise be available. If you are interested in participating in a clinical trial, ask your doctor what clinical trials you are eligible for. You can also read our 'Understanding Clinical Trials' Fact Sheet to find a clinical trial.

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, 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

- WM is usually a slow growing (indolent) lymphoma that may not need treatment.
- Some faster growing (more aggressive) WM may need treatment, and while you may not be cured from WM, many people go into remission after treatment.
- WM occurs when your cancerous B-cell make too many immunoglobulins called IgM, which can make your blood thicker and cause problems with your blood flow.
- Report any concerns or worsening symptoms to your medical team.

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

Car T-cell therapy

SCAN ME

Definitions

SCAN ME

eviQ Lymphoma Treatments

SCAN ME

Factsheets

SCAN ME

Health translations

SCAN ME

Questions to ask your doctor

SCAN ME

Understanding clinical trials

SCAN ME

WM webpage

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.

Notes