

Marginal Zone Lymphoma (MZL)

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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

Marginal Zone Lymphoma (MZL) is a subtype of non-Hodgkin Lymphoma (NHL). If you've been diagnosed with MZL you have a type of slow growing blood cancer. This blood cancer affects a type of white blood cells called B-cell lymphocytes (B-cells). Some people with MZL may be cured with treatment, however not everyone can be cured. Some types of MZL do not respond to any of the treatments we have yet. But, because MZL are slow growing cancers, many people still live a long and healthy life with MZL – even without treatment.

Understanding B-cell lymphocytes

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 B-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 can travel to any part of your body. This means your lymphoma cells can also travel to any part of your body.

MZL subtypes

There are more than 80 different types of lymphoma, and MZL is a common subtype. One in every 10 people diagnosed with non-Hodgkin lymphoma has MZL. There are 3 subtypes of MZL, named because of where the lymphoma cells gather. They are called:

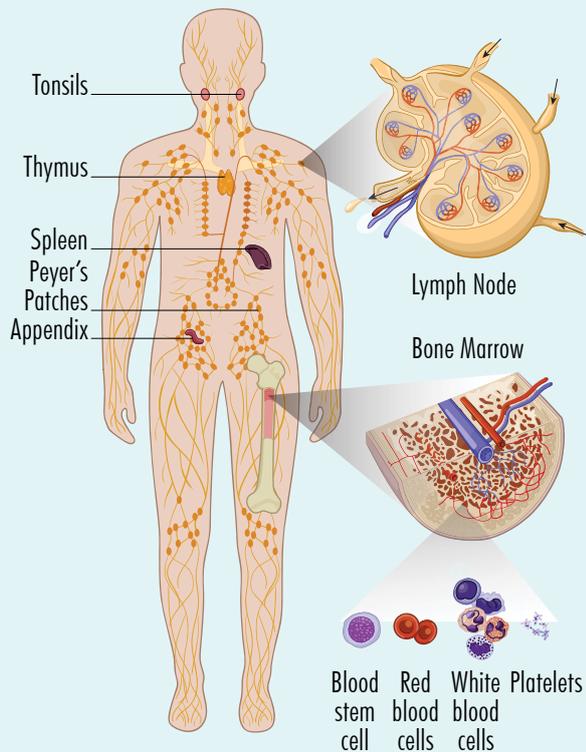
- **Nodal MZL** – this can also be called **monocytoid B-cell lymphoma**. This subtype develops in the outer edges of your lymph nodes.
- **Extra-nodal MZL** – this can also be called **mucosa-associated lymphoid tissue (MALT) lymphoma**. This is a type of MZL that develops outside of your lymph nodes. It can develop in your stomach, small intestine, salivary gland, thyroid gland, eyes, or lungs. It may be called “gastric” if it affects your stomach or small intestines, and “non-

gastric" if it affects the other areas.

- **Splenic MZL** - This subtype develops in your spleen. Your spleen is an organ that sits just under your stomach on the left side of your abdomen, and plays an important role in keeping your blood healthy. It also stores a lot of your disease fighting B-cells.

Most people that get MZL are over the age of 60 years, but younger people can get it too.

LYMPHATIC SYSTEM



Signs and Symptoms

Because MZL grows slowly, you may not have any symptoms at first. Many people are diagnosed when they have a blood test, scans, or a physical exam for something else. If you do have symptoms, they may be general symptoms of lymphoma, or specific symptoms based on where the lymphoma is located.

General symptoms of lymphoma may include:

- feeling unusually tired (fatigued)
- feeling out of breath

SWOLLEN LYMPH NODE



- 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 (this is caused by lymphoma cells gathering in the follicles of your lymph nodes and making it grow larger). These lumps may or may not hurt, depending on where they are located.

You may have more specific symptoms depending on your subtype:

Nodal MZL –The main symptom of nodal MZL is a lump in your neck, armpit or groin. It might feel rubbery to touch. More information on Nodal MZL can be found on our webpage [here](#).

Extra-nodal or MALT MZL –If you have gastric MALT MZL you may have symptoms related to your gut including pain in your tummy, feeling sick, and heartburn (indigestion). We are not sure what causes this to happen but, people who have infections with a bacteria that causes stomach ulcers, may be at increased risk of developing a gastric MALT MZL. This bacterium is called helicobacter pylori (H. Pylori).

For non-gastric MALT MZL, the symptoms will depend on the area affected. If the MZL is in your thyroid, you may have a lump in your throat or changes to your hormones. If your eyes are affected you may have changes to your sight, and if it is in your lungs you may have changes to your breathing, or chest pain. More information on MALT MZL can be found on our webpage [here](#).

Splenic MZL – The most common symptom you might get with this subtype is feeling full sooner than you normally would, even if you have only a little bit to eat. You may also have pain in your tummy. Like with MALT MZL, we're not sure what causes splenic MZL to occur, but people who have had a viral infection of their liver, called hepatitis C have an increased risk of developing splenic MZL. More information on splenic MZL can be found on our webpage [here](#).

Diagnosis and Staging

Your doctor may suspect you have lymphoma when they get your blood test results, X-ray or other scan results, or do a physical exam. But to diagnose MZL, you will need a biopsy.

A biopsy is a procedure to remove part, or all of an affected lymph node, tumour or bone marrow. Your bone marrow is the spongy part inside your bones where your blood cells are made. The biopsy is then checked by scientists to see if there are changes that help the doctor diagnose MZL.

When 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.

If you have MZL, your doctor will organise more tests to check if the cancer has spread to other parts of your body.

Staging Tests

- [Positron emission tomography \(PET\) scan](#)
- [Computed tomography \(CT\) scan](#)
- [Bone marrow biopsy](#)

Additional tests

- Bronchoscopy – A long thin tube with a camera is passed through your mouth or nose into your lungs so your doctor can see your lungs better, and take biopsies if needed
- Gastroscopy – (endoscopy or colonoscopy) – A long thing tube with a camera is passed down your throat into your stomach, or through your rectum (bottom) into your bowels to look and take biopsies if needed
- Other tests to see how well your kidneys, heart and other organs are working.

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 (or your child) 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.

Treatment options

Because MZL is generally slow-growing, you may not need treatment. But you will see your specialist doctor regularly. This time can be called "Watch and Wait", but active monitoring is a better description because your doctor will continue to watch how your **MZL** grows.

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 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. Your health will not be improved, and you will not live longer by starting treatment earlier.

Your doctor will consider several things when making choices about your treatment. These include:

- What subtype of MZL you have and if you have or have had infections in the past
- how fast your lymphoma is growing
- how bad your symptoms are
- your age
- your overall health.

Treatments can include:

Antibiotics – Depending on your subtype and stage of MZL you may only need antibiotics. As mentioned above, some infections increase your risk of MZL. By treating the infection, the MZL may also be controlled. Once the infection is gone, it may still take months or several years for the lymphoma to go away completely, but without the infection, your lymphoma may not progress.

Radiation / Radiotherapy - Radiation treatment uses high doses of radiation to kill cancer cells and shrink tumours. Before having radiation, you will have a planning session. This session is important for the radiation therapists to plan how to target the radiation to the lymphoma, and avoid nearby healthy tissue. Radiation therapy usually lasts between 2-4 weeks. During this time, you will need to go to the

radiation centre everyday (Monday-Friday) for treatment.

Surgery – An operation to remove the tumour, spleen (splenectomy) or affected lymph node/s.

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. 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) – MABs are 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. This helps your own immune system fight the MZL more effectively. Some people may have a MAB and chemo.

Targeted therapy – Targeted therapy is taken as a tablet at home or in hospital. Targeted therapies attach to lymphoma cells and block signals needed for them to grow and produce more lymphoma cells. This stops the cancer from growing, and causes the lymphoma cells to die off.

Stem-cell transplant – if your first treatment did not work, or you've had treatment in the past and your MZL has

come back, you may be offered a stem cell transplant. Stem cell transplants are only suitable for some patients. Your doctor will be able to talk to you about this. To learn more about stem cell transplants please see out factsheets

All factsheets are available on our website [here](#).) If you do not have access to a computer and would like a paper copy, please call us on 1800 359 081 or email us at enquiries@lymphoma.org.au.

Starting 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 some people, you may need treatment again, but not for weeks, months or even years.

First-line treatment

If you have stage one or two lymphoma and need treatment, you may be offered radiation treatment. Some people will only ever need to have one course of radiotherapy, and will not need any other treatment. Some people may have radiation treatment and chemotherapy. This is called chemo-radiation.

If you have stage 3 or 4 MZL you may need more treatment. This can be a combination of treatments such as chemotherapy and a monoclonal antibody. You may also be eligible for a clinical trial – Ask your doctor if there are any clinical trials you could join.

Second-line treatment – first relapse, or refractory disease

Some people will not need treatment again at all. But, if your MZL “flares up again” it is called a relapse. Treatment after a relapse is called second-line treatment (or third-line etc). But some people may not respond to their first line treatment. If your MZL 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.

Second-line treatments may be a combination of the above - radiation, chemo and monoclonal antibodies, or for some people, a stem cell transplant. You may also be eligible for a clinical trial – Ask your doctor if there are any clinical trials you could join.

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 MZL in the future. Some targeted therapies are the focus of current clinical trials for MZL. Clinical trials can also offer you a chance 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 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

- MZL is usually a slow growing (indolent) B-cell lymphoma.
- There are different subtypes of MZL and your treatment and diagnostic tests will depend on your subtype (or

suspected subtype).

- When MZL is caused by bacteria, treatment may include antibiotics.
- Ask your doctor what subtype of MZL you have and how this will affect your treatment options.
- Report and new 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 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

Understanding clinical trials



Definitions



eviQ Lymphoma Treatments



Factsheets



Health translations



MZL webpage



Questions to ask your doctor



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.



