Mantle Cell Lymphoma (MCL)

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

Mantle cell lymphoma (MCL) is a subtype of non-Hodgkin Lymphoma (NHL). If you've been diagnosed with MCL, you have a type of blood cancer. Most people with MCL have a fast growing (aggressive) lymphoma, but for some people it may grow more slowly. There are more than 80 different types of lymphoma, and for every 20 people diagnosed with lymphoma, 1 or 2 people (5-10%) will have MCL.

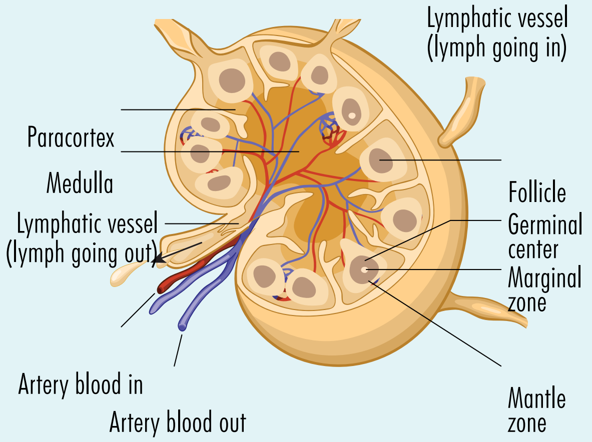
MCL affects a type of white blood cell called B-cell lymphocytes (B-cells). B-cells fight infection and diseases to keep you healthy. They remember infections you had in the past, so if you get the same infection again, your immune system can fight it more effectively. When these cells don't grow or 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 live in your spleen and your lymph nodes. MCL happens when the B-cells in the outer edge of your lymph node (called the mantle zone) become cancerous.

Symptoms of MCL

If you have a slow-growing MCL, your spleen may be bigger than normal and you may have some lymphoma cells in your blood. If you have a fast-growing MCL, your first symptom might be a swollen lymph node in your neck, armpit, groin or abdomen.

STRUCTURE OF A LYMPH NODE



Other symptoms, you might get include:

- feeling unusually tired (fatigued)
- feeling out of breath
- bruising or bleeding more easily than usual
- blood in your poo (this can happen if you have MCL in your stomach or bowels)
- 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 mantle zone of your lymph nodes and making it grow larger). These lumps may or may not hurt, depending on where they are.
- B-symptoms.

Diagnosis and Staging

Your doctor may suspect you have lymphoma when they get your blood test results, X-ray, other scan results, or do a physical exam. But to diagnose MCL, you will need a biopsy. A biopsy is a procedure to remove part, or all of an affected lymph node or bone marrow. The biopsy is then checked by scientists in a laboratory to see if there are changes that help the doctor diagnose MCL.

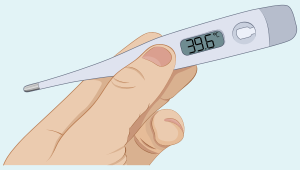
When you have a biopsy, you may have a local or general anaesthetic. This will

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.

depend on what part of your body the biopsy is taken from.

Lymphoma cells can travel to any part of your body so, if you have MCL your doctor will organise more tests to check if it is anywhere else.

Staging tests

Staging tests may include:

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

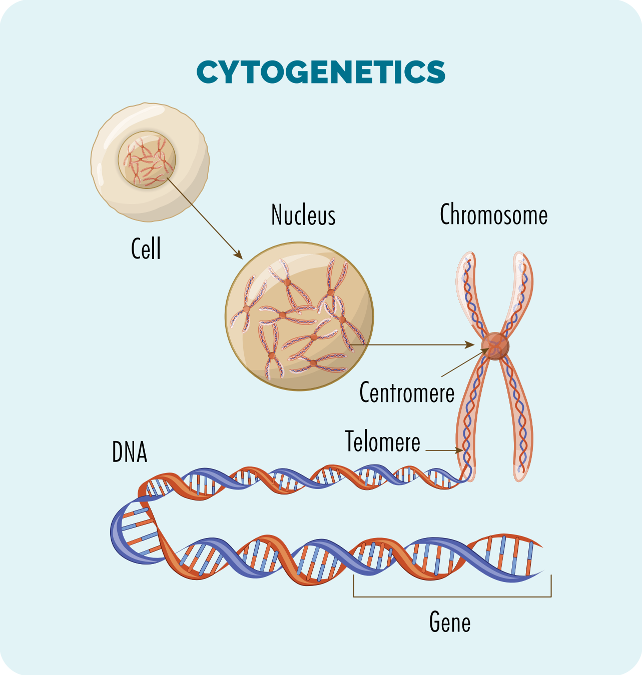
You can find more information on staging tests at our website. Just look down the left-hand side to find the test you want more information on: https://www.lymphoma.org.au/lymphoma/tests-diagnosis-and-staging/.

Stage I (1) and II (2) lymphoma are considered early-stage lymphomas, meaning that only one or two areas either above OR below your diaphragm have lymphoma. Stage III (3) lymphoma is considered an advanced stage and means that you have lymphoma on both sides of your diaphragm.

Stage IV (4) lymphoma is the most advanced stage, and means you have lymphoma on both sides of your diaphragm, and in one of your organs such as your liver, lungs, brain, bones or bone marrow.

Cytogenetic tests

Cytogenetics are tests on your blood and biopsies that look for changes in your chromosomes or genes. We usually have 23 pairs of chromosomes, and they are numbered according to their size. If you have MCL, your chromosomes may look a little different.



On your chromosomes are many genes that tell the proteins and cells in your body how to look or act. If there is a change (variation) in these chromosomes or genes, your proteins and cells will not work properly. As a result, you can develop different diseases, including lymphoma.

The most common cytogenetic change in MCL is when a small part of two of your chromosomes swap places. This usually involves chromosomes 11 and 14, or the bottom part of chromosomes 13 and 32. When this happens it is called a translocation. It is often written as **t(11:14) or (13q:32q)**. The "q" tells the doctor it is the bottom part of the chromosomes. If it is the top part of the chromosome it will have a "p".

Sometimes you might have a part of the chromosome missing. When this happens it is called a deletion. If the top part of your 17th chromosome is missing it will be written as **del(17p).**

It is important to find out what type of cytogenetic changes you have before you start treatment, because different changes need different types of treatment.

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.

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

If you have a slow growing MCL, 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 watch how your MCL 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. 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 you have a fast-growing MCL you may need treatment. The treatment choices your doctor will offer you will depend on:

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

Treatments can include:

Radiation/Radiotherapy – Radiation therapy is a cancer treatment that uses high doses of radiation to kill lymphoma 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 damaging healthy cells. 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.

If you live a long way from the radiation centre and need help with a place to stay during treatment, please talk to your doctor or nurse about what help is available to you. You can also contact the Cancer Council or Leukemia Foundation in your state and see if they can help with somewhere to stay.

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.

Monoclonal Antibody (MAB) – Given as an infusion at 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 MCL.

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.

Stem-cell transplant – to learn more about stem cell transplants please see our factsheets

- Transplants in Lymphoma
- Allogeneic stem cell transplants
- Autologous stem cell transplants

Chimeric antigen receptor therapy (CAR T-cell therapy) – Please see our factsheet for more information on CAR T-cell therapy

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 weeks, months or even years.

First-line treatment

For early stage 1 or 2 MCL, you may have radiation therapy. This may be with or without chemotherapy. If you need to start treatment, you may have more than one medicine, including several different types of chemotherapy and a monoclonal antibody. When you have these treatments, you will have them in cycles. That means you will have the treatment, then a break. then another round of treatment. First-line treatments may include:

R-CHOP - monoclonal antibody called rituximab (or a medication similar to rituximab called a biosimilar), and chemotherapy called cyclophosphamide, doxorubicin, vincristine and a steroid called prednisolone

R-DHAP – monoclonal antibody called rituximab (or a medication similar to rituximab called a biosimilar), and chemotherapy called cisplatin and cytarabine and a steroid called dexamethasone.

R-maxi CHOP – The same as R-CHOP above, but without the rituximab (or biosimilar) on the first cycle

R-HiDAC – monoclonal antibody called rituximab (or a medication similar to rituximab called a biosimilar), and chemotherapy called cytarabine

Bendamustine & rituximab – (or a medication similar to rituximab called a biosimilar) a chemotherapy and a monoclonal antibody

In some cases your doctor may also decide to alternate your treatment each cycle. For example, R-CHOP one cycle and R-DHAP the next.

Second-line treatment

If you previously had treatment, finished treatment and had time not needing treatment, then your MCL 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). But some people may not respond to their first line treatment. If your MCL 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. Some of the treatment options you may be offered include:

- Ibrutinib – (Targeted therapy called a BTK inhibitor – Blocks signals the cancer needs to survive)

- Acalabrutinib - (Targeted therapy called a BTK inhibitor - Blocks signals the cancer needs to survive)
- Lenalidomide – (Immunomodulator – Increased your immune cells and boosts their ability)
- Chemotherapy – may include one of the treatments above
- If you are less than 70 years old and fit, you may be offered a stem cell transplant.
- 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 MCL in the future. They can also offer you a chance to try a new medicine, combination of medicines or other treatments such as CAR T-cell therapy that you would not be able to get outside of the trial. 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

- Mantle Cell Lymphoma is type of blood cancer affecting B-cells lymphocytes. B-cells are immune cells that fight infection infection and disease, and live mostly in your lymphatic system.
- MCL is usually an aggressive type of lymphoma, but in some cases it may be indolent.
- If you have aggressive MCL, you will start treatment soon after you are diagnosed.
- If you have indolent MCL, you may not need treatment. However, you will be actively monitored by your specialist doctor while on watch and wait.
- Your cytogenetic tests provides your

doctor more information to help plan your care. If you don't know if you have any genetic mutations - Ask your doctor.

- Talk to your doctor abut preserving your fertility before starting treatment if you are planning to have children.
- Report any B-symptoms or other concerns to your doctor as soon as possible.

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

Questions to ask your doctor

SCAN ME

Definitions

SCAN ME

Facebook: Lymphoma Down Under

SCAN ME

Health Translations

SCAN ME

MCL website

SCAN ME

Treatments website

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.

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