

# Mantle Cell Lymphoma (MCL)

## Introduction

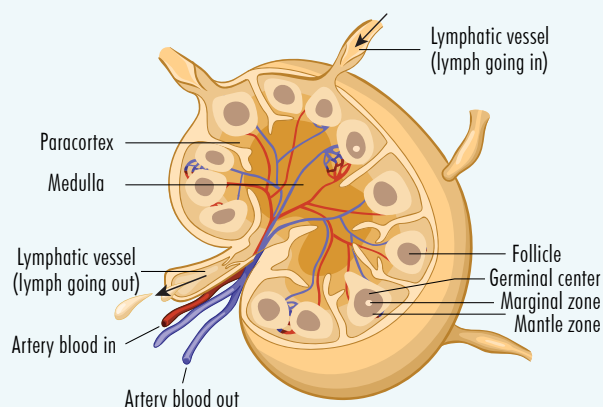
Mantle cell lymphoma (MCL) is a subtype of non-Hodgkin Lymphoma (NHL). If you've been diagnosed with MCL, you have a type of blood cancer. Most people with MCL have a fast growing (aggressive) lymphoma, but for some people it may grow more slowly. There are more than 80 different types of lymphoma, and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

### 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 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. These tests are called "staging" and may include:

- Blood tests
- Positron emission tomography (PET) scan
- Computed tomography (CT) scan
- Bone marrow biopsy (Your doctor

will use a needle to take a sample of marrow from the middle of your bone - usually hip, but sometimes the sample may be taken from a different bone. This will be done with local anaesthetic.

- Lumbar puncture - (Your doctor will use a needle to take a sample of fluid from near your spine. This is done if the doctor needs to check if your lymphoma is in your brain or spinal cord)
- Echocardiogram (this is an ultrasound to take a picture of your heart and see how well it is working. You may need this if you are going to have certain chemotherapy medications)
- Cytogenetic tests - This is where your blood and tumour sample are checked for genetic variances that may be involved in your disease.

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.

### Questions for your doctor before you start treatment

1. What tests have been done?  
What tests still need to be done before treatment?
2. Do I have any genetic abnormalities in my blood or biopsies?
3. Can you explain these results to me?
4. What treatment will work best for me based on these results?
5. What is the intent of this treatment? (e.g., Will you be cured, or go into remission)
6. How long will I be at the cancer center/hospital on treatment days?  
How often?
7. Do I have any other choices?
8. What are the best treatments that are available for my type of MCL, even if they're not available in Australia?
9. What are the main, and the worst side effects of this treatment?
10. What clinical trials am I eligible for?
11. Will I need to have time off work? If yes, can I see a social worker to help me arrange finances?
12. What support services are available to help me cope with my diagnosis and learn strategies to avoid stress and talk to my family and friends about my diagnosis and treatment.

### Additional Questions if you live in rural Australia

13. Can I have my appointments and treatment close to home?
14. Are there better treatments for me in larger centres that are not available here?
15. Is telehealth an option for me?
16. How long will I need to be away from home for treatment?
17. How often will I need to come to the city (or be away from home)?
18. What financial, accommodation and travel assistance is available to me?
19. Is there a social worker who can help me arrange these things?
20. Are there any clinical trials I can join, and if I do, can I stay where I am or do I need to travel to another centre?

### 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)** – These medications might be taken as a tablet or be given as a drip (infusion) into your blood at a cancer clinic or hospital. Chemo kills fast growing cells so can also affect some of your good cells that grow fast

**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](mailto: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)
- 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

You will still see your doctor for blood tests and scans after your treatment ends. Your doctor will check you for signs and symptoms of the FL coming back, and for side-effects you may have from your treatment.

### Resources and support

**Lymphoma Australia** offers a wide variety of resources and support for people with lymphoma and their carers. Please visit our website [www.lymphoma.org.au](http://www.lymphoma.org.au) for further information. Lymphoma Australia Fact sheets and booklets include:

- Booklet: What you need to know about lymphoma
- Lymphoma subtypes
- Understanding Clinical Trials
- Emotional impact of a lymphoma diagnosis and treatment
- Fear of cancer recurrence and anxiety

Visit the [Lymphoma Australia YouTube Channel](#) for presentations and interviews on a variety of topics about

## Mantle Cell Lymphoma (MCL)

lymphoma subtypes, management and supportive care.

Contact the Lymphoma Nurse Support Line on 1800 953 081, email: [nurse@lymphoma.org.au](mailto:nurse@lymphoma.org.au) or join the private Facebook group: [Lymphoma Down Under](#).

2022\_MCL\_FactSheet