

Mantle cell lymphoma

This page is about mantle cell lymphoma, an uncommon type of lymphoma that usually behaves like a high-grade (fast-growing) lymphoma.

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Quick overview

This section is an overview of the information on this page. There is more detail in the sections below.

What is it?

Mantle cell lymphoma is usually a fast-growing type of lymphoma, but some cases grow slowly. It can be successfully treated, but usually comes back and needs more treatment.

What are the symptoms and how is it diagnosed?

Most people with mantle cell lymphoma have swollen lymph nodes (glands). The lymphoma can also affect other parts of the body, such as the bone marrow, bowel and spleen. You need a biopsy of the abnormal area to confirm you have mantle cell lymphoma. Other tests are done to work out where the lymphoma is and how your body is being affected by it.

How is it treated?

Treatment for mantle cell lymphoma depends on how fit you are. Most people have chemotherapy with an antibody treatment called rituximab. If possible, you have strong chemotherapy and a stem cell transplant, which could keep mantle cell lymphoma under control for longer. Some people need gentler treatments. Newer drugs are increasingly used for mantle cell lymphoma when it comes back.

What is mantle cell lymphoma?

Mantle cell lymphoma develops from abnormal B lymphocytes (B cells; a type of white blood cell). It is called 'mantle cell' because the abnormal B cells come from an area called the 'mantle zone' in lymph nodes (glands).

There are **types of lymphoma** that grow quickly (high-grade lymphoma) and types that grow slowly (low-grade or 'indolent' lymphoma). Mantle cell lymphoma is unusual as it often has features of both.

- The abnormal B cells in mantle cell lymphoma are small – they look like low-grade lymphoma cells under a microscope.
 - Like low-grade lymphomas, mantle cell lymphoma is likely to relapse (come back) after it has been treated.
 - Mantle cell lymphoma usually grows quickly, like a high-grade lymphoma. A few people have an 'indolent' form of mantle cell lymphoma, which means it grows slowly, like a low-grade lymphoma.
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Who gets mantle cell lymphoma and why?

About 500 people are diagnosed with this type of lymphoma each year in the UK. Mantle cell lymphoma is much more common in men than in women. It is usually diagnosed in people who are middle-aged or older. It is very rare in young people.

My diagnosis was mantle cell lymphoma stage 3 with my lymph nodes and spleen affected. I now realise that at the age of 63 and a man, I am the classic profile of someone with this type of lymphoma.

– Stephen

In most cases, the **cause** of mantle cell lymphoma is unknown.

A certain genetic change is found in the abnormal cells in almost all cases of mantle cell lymphoma, but it is not known why it occurs. This genetic change is called a 'translocation'. It happens when two of the chromosomes (chromosome 11 and chromosome 14) in a B cell break and then join up with each other. This translocation means the cells make too much of a protein called 'cyclin D1' that helps to control cell growth. Too much cyclin D1 causes uncontrolled growth; too many new B cells are made and mantle cell lymphoma develops.

Symptoms

The most common **symptom** of mantle cell lymphoma is one or more swollen lymph nodes.

Most people have mantle cell lymphoma at an advanced stage when it is diagnosed, meaning that other parts of the body are likely to be affected as

well as lymph nodes. Your symptoms depend on which areas of your body the lymphoma is affecting.

Areas that are commonly affected by mantle cell lymphoma include:

- bone marrow – this can stop enough normal blood cells being made, causing low blood counts
- bowel – this can cause problems such as diarrhoea
- spleen – your spleen may be swollen, causing abdominal (tummy) pain or discomfort.

Rarely, mantle cell lymphoma spreads to the brain and spinal cord (the central nervous system or CNS) too. This is called '**secondary CNS lymphoma**'. Lymphoma in the CNS causes symptoms such as headaches, dizziness and confusion. Secondary CNS lymphoma is more likely to develop if mantle cell lymphoma comes back, rather than when it is first diagnosed, but it is still uncommon.

Diagnosis and staging

Mantle cell lymphoma is usually diagnosed by examining a **biopsy** of a swollen lymph node or another abnormal area. Sometimes, the abnormal cells can be detected in blood or bone marrow samples. The abnormal B cells can look like other types of lymphoma under the microscope, for example, **chronic lymphocytic leukaemia (CLL)** or **splenic marginal zone lymphoma**. However, most cases of mantle cell lymphoma have characteristic genetic changes. Tests are done on the sample to look for changes in the genes and proteins of the abnormal cells so you can be given the correct diagnosis.

Other tests find out more about your general health. Tests are also needed to find out which parts of your body are affected by lymphoma – this is called '**staging**'. These tests usually include:

- a physical examination
- **blood tests** to look at your general health, including your blood cell counts

- a **scan** – usually a CT scan
- a **bone marrow biopsy** to see if the lymphoma is affecting your bone marrow.

If your lymphoma is in your bowel, you might have other tests such as a colonoscopy (a tiny camera passed through your anus to look at your bowel).

If your doctor suspects your lymphoma might be in your CNS, you might need a **lumbar puncture** and possibly an MRI scan.

Your doctor might recommend other tests depending on your symptoms and your general health.

Although **waiting for the results** of your tests can be difficult, your doctor is collecting important information during this time. It is important that your doctor knows exactly what type of lymphoma you have so they can give you the most appropriate treatment.

Outlook

First-line treatment is usually successful at putting mantle cell lymphoma into remission, but this type of lymphoma is difficult to cure. It almost always relapses and needs more treatment.

People who are fit enough to have intensive (strong) treatments usually have a better outlook but there are increasing treatment options, including newer, targeted drugs, for people who cannot have such treatments. Targeted drugs also offer new treatment options if the lymphoma relapses.

Your doctor is best placed to advise you on your outlook based on your individual circumstances and any risk factors you have.

Treatment

Most people have advanced-stage (stage 3 or 4) mantle cell lymphoma when they are diagnosed and need systemic treatment (treatment that affects the

whole body). A few people have localised disease (stage 1 or 2) and may be able to have **radiotherapy** to the affected areas.

Treatment for mantle cell lymphoma that needs systemic treatments depends on several factors including:

- how your lymphoma is behaving, for example whether it is growing quickly or slowly
- what symptoms you have
- your age and general health
- your feelings about the treatment options.

Treatment for indolent (slow-growing) mantle cell lymphoma

‘Indolent’ means the lymphoma is growing slowly – it might be called ‘low-grade mantle cell lymphoma’. If you have indolent mantle cell lymphoma, you might have lymphoma cells in your bloodstream but your swollen lymph nodes might be small and not growing much.

Your doctor might suggest **active monitoring** (also called ‘watch and wait’) if you have an indolent form of mantle cell lymphoma and do not have troublesome symptoms. Active monitoring involves regular check-ups but you do not receive treatment until the lymphoma is causing you problems. Treating you before the lymphoma is causing problems does not make you live longer.

When the lymphoma is causing problems, your doctor usually considers the same treatment options that are used for fast-growing mantle cell lymphoma.

Treatment for fast-growing mantle cell lymphoma

Mantle cell lymphoma usually grows quickly and is treated like a high-grade lymphoma. Most people with mantle cell lymphoma have intravenous **chemotherapy**, usually combined with an **antibody treatment** such as **rituximab**. You might hear this called ‘chemo-immunotherapy’.

If the lymphoma is in your CNS, it is treated as **secondary CNS lymphoma**.

Standard chemo-immunotherapy regimens

Chemo-immunotherapy regimens used for mantle cell lymphoma include:

- R-CHOP, which is the antibody rituximab with CHOP chemotherapy
- bendamustine and rituximab.

Your doctor might recommend a different regimen. A targeted drug called **bortezomib** might be used in combination with chemo-immunotherapy. Your doctor might suggest modifying the regimen, for example, by reducing the dose, if you are less fit. Some people need gentler treatments.

Note: after having treatment with bendamustine, you must be given specially prepared irradiated blood if you need to have **blood transfusions** in future. This is so that you don't develop a rare but serious complication of blood transfusion called 'transfusion-associated graft-versus-host disease'.

Chemotherapy including cytarabine

People who are fit enough usually have the drug cytarabine (also known as Ara-C) given at a high dose as part of their treatment. It might be given:

- on its own between cycles of chemo-immunotherapy
- as part of another regimen, such as R-DHAP.

This is then followed by an **autologous stem cell transplant** (using your own cells) in people fit enough to have one.

This treatment can give a longer remission than standard chemo-immunotherapy alone, but it is a more intensive form of treatment and can cause more side effects. Most older people (over 65) and those who have other illnesses are not able to have such intensive treatment because of the risk of serious side effects.

Gentler treatment

Gentler kinds of chemotherapy might be used for people not fit enough for standard chemo-immunotherapy. This might include chlorambucil or cyclophosphamide tablets. These treatments can help control the lymphoma

and ease symptoms with fewer side effects than standard chemo-immunotherapy. They are less likely to give a long-lasting remission.

Newer, targeted drugs might also be an option for people not able to have standard chemo-immunotherapy. You may be able to enter a clinical trial testing a gentler treatment for mantle cell lymphoma.

Maintenance treatment

People who respond well to treatment may be offered **maintenance treatment** with rituximab. Maintenance treatment aims to make your remission last longer.

Rituximab is usually given on its own once every 2 months for maintenance treatment. Most people have maintenance for a couple of years, but it could be longer or shorter than this. Maintenance is not suitable for everyone. It can cause side effects, although these are mild for most people. Your doctor can advise you if they think maintenance treatment might benefit you and how long they recommend you continue maintenance.



Part of my reason for taking part in the trial was that I understand how important it is to get people involved in trials in order to have data to analyse. But I also realised that I would still get good treatment and a lot of care and follow-up.

– Stephen, who was treated for mantle cell lymphoma as part of a clinical trial.

Follow-up

You have regular **follow-up appointments** after treatment, usually every 2–3 months.

If you are worried about your health at any time, contact your GP or medical team. They can reassure you or arrange an appointment for you to have a check-up.

Relapsed and refractory mantle cell lymphoma

The length of remission (time where the lymphoma is under control) after treatment varies greatly. Mantle cell lymphoma **relapses** at some time after treatment in most people. The treatment at this stage depends on:

- what treatment or treatments you've had before
- how well your previous treatment worked
- how long your remission lasted
- how your lymphoma is behaving now
- your age, general health and thoughts about further treatment.

Sometimes, mantle cell lymphoma is refractory (doesn't respond) to the first-line treatment. In this case, a different treatment is usually needed.

Mantle cell lymphoma can relapse several times and you might have different treatments each time.

Possible treatment options for relapsed or refractory mantle cell lymphoma include:

- the same chemo-immunotherapy again if it worked well previously, although it may not work as well or for as long when used again
- a different or stronger chemotherapy for those who are fit enough
- an autologous or allogeneic stem cell transplant for those who are fit enough and haven't already had one
- the targeted drug **ibrutinib** (Imbruvica®)
- another **targeted drug** or experimental treatment as part of a clinical trial
- **palliative care** to control symptoms
- rarely, a period of active monitoring if the lymphoma is growing slowly and is not causing any problems.

Newer, targeted drugs are often available first for people with relapsed and refractory lymphoma. There are several targeted drugs already approved for mantle cell lymphoma and many clinical trials testing new treatments for this type of lymphoma. Our **targeted drugs page** has the latest information on drugs available for mantle cell lymphoma and other types of lymphoma. You can also search our clinical trials information service, Lymphoma TrialsLink, to find clinical trials suitable for people with mantle cell lymphoma.

References

These are some of the sources we used to prepare this information. The full list of sources is available on request. Please contact us by email at **publications@lymphoma-action.org.uk** or phone on **01296 619409** if you would like a copy.

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Further reading

- [What is lymphoma?](#)
- [Chemotherapy](#)
- [Ibrutinib](#)
- [Glossary](#)

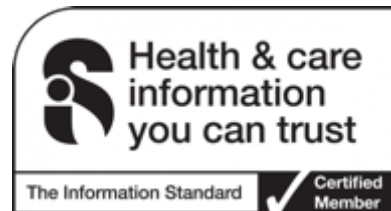
Acknowledgements

We would like to thank the Expert Reviewers and members of our Reader Panel who gave their time to review this information.

Content last reviewed: February 2018

Updated: April 2018

Next planned review: February 2021



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