

Mantle cell lymphoma

Mantle cell lymphoma is a rare type of non-Hodgkin lymphoma.

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What is mantle cell lymphoma?

Lymphoma is a type of blood cancer that develops when white blood cells called **lymphocytes** grow out of control. Lymphocytes are part of your **immune system**. They travel around your body in your **lymphatic system** and blood, helping you fight infections. There are two types of lymphocyte: **T lymphocytes (T cells)** and **B lymphocytes (B cells)**.

There are lots of different **types of lymphoma**. Mantle cell lymphoma is a type of non-Hodgkin lymphoma that develops from B cells. It is called 'mantle cell' lymphoma because the abnormal B cells usually develop in a part of your **lymph nodes** called the 'mantle zone'.

Usually, **non-Hodgkin lymphomas** are divided into **fast-growing (high-grade) lymphomas** and **slow-growing (low-grade) lymphomas**. Mantle cell lymphoma is unusual because it often has features of both.

- Mantle cell lymphoma usually grows quickly, like a high-grade lymphoma. Some people have a form of mantle cell lymphoma that grows more slowly.
- Mantle cell lymphoma is likely to come back (relapse) after treatment and need more treatment. This is like low-grade lymphoma.

Types of mantle cell lymphoma

There are two main types of mantle cell lymphoma:

- **Classical mantle cell lymphoma** accounts for most cases. It affects lymph nodes but often spreads to other parts of the body, such as the **bone marrow**, spleen, bowel and liver. It is usually fast-growing, although it can sometimes have a slower course.
- **Leukaemic non-nodal mantle cell lymphoma** is less common. People with this type typically have a swollen spleen, and lymphoma cells in the bloodstream and bone marrow. This type of mantle cell lymphoma tends to grow slowly.

Who gets mantle cell lymphoma?

Around 600 people are diagnosed with mantle cell lymphoma each year in the UK. It is 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, diagnosed with mantle cell lymphoma

Read [Stephen's story](#).

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

Most cases of mantle cell lymphoma have a particular genetic change (mutation) in the abnormal cells. Scientists don't know why this genetic change develops. The mutation means the B cells make too much of a protein called cyclin D1. Too much cyclin D1 makes the B cells grow out of control, and lymphoma develops.

Symptoms

The most common symptom of mantle cell lymphoma is a lump, or lumps, which often develop in several parts of your body. These are **swollen lymph nodes**.

Many people with mantle cell lymphoma have a swollen spleen. Your doctor might notice it when they are examining your tummy (abdomen) during a routine examination. If your spleen becomes very big, you might feel full very quickly when you eat, or get pain or discomfort behind your ribs.

Many people with mantle cell lymphoma have lymphoma cells in their **bone marrow** (the spongy centre of larger bones where blood cells are made) when they are diagnosed. This might lead to:

- **anaemia** (low red blood cell count), which can cause tiredness and shortness of breath
- **thrombocytopenia** (low platelet count), which makes you more likely to bruise and bleed
- **neutropenia** (low neutrophil count – a type of white blood cell), which makes you more prone to infection.

Some people experience **unexplained weight loss**, **night sweats** or **fever**. These are known as 'B symptoms' and can occur together.

Mantle cell lymphoma commonly grows outside the lymphatic system (extranodal lymphoma). Your symptoms depend on which areas of your body the lymphoma is affecting. It often grows in the tummy (abdomen) and bowel, which might cause tummy or back pain, diarrhoea or swelling of your tummy. It might also develop in other parts of your body.

Rarely, mantle cell lymphoma spreads to the brain and spinal cord (the central nervous system or CNS). This is called **secondary CNS lymphoma**. Lymphoma in the CNS can cause symptoms such as headaches, dizziness and confusion.

Diagnosis and staging

Mantle cell lymphoma is typically diagnosed by a **biopsy**. This is a small procedure to remove a sample of tissue, such as a swollen lymph node. If you have leukaemic mantle cell lymphoma, the lymphoma might be diagnosed through a **blood test**.

An expert lymphoma **pathologist** looks at the blood and tissue samples in the laboratory. In most cases of mantle cell lymphoma, there are characteristic proteins and genetic features that help the pathologist confirm the diagnosis. The pathologist also tests the samples to find out how quickly the cells are dividing, and, in some cases, to look for particular genetic changes (mutations). This can help your medical team decide on the most appropriate **treatment** for you.

I noticed a couple of lumps in my neck and booked an appointment with my GP. She suspected a viral infection but she also referred me straightaway to specialists in both ENT and haematology. After various examinations and tests, I had an ultrasound scan and fine needle biopsy, which suggested mantle cell lymphoma. I had to wait for an operation to remove a lymph node from my neck for biopsy and a CT scan to ascertain the extent of the disease.

Peter, diagnosed with mantle cell lymphoma

You have other tests to find out which areas of your body are affected by lymphoma. This is called **staging**. These tests might include:

- a CT or **PET/CT** scan
- a **blood test** to look for lymphoma cells in your bloodstream
- a **bone marrow biopsy** to check for lymphoma cells in your bone marrow
- a **lumbar puncture** (a procedure to take a sample of fluid from your spine using a thin needle) and an **MRI scan** if your doctor thinks you might have lymphoma affecting your brain or spinal cord
- an **endoscopy** (a camera examination of your stomach or bowel using a thin, flexible tube passed through your mouth or bottom) if your doctor thinks you might have lymphoma in your digestive tract.

You also have **blood tests** to look at your general health, check your **blood cell counts** and make sure your kidneys and liver are working well. Other blood tests check for viral **infections that might have led to your lymphoma** or that could flare up (reactivate) during treatment.

Depending on what treatment your doctor recommends for you, you might have a heart scan to test your heart function before you start treatment.

Not everyone needs all of these tests. Your **medical team** will decide what tests you need based on your individual circumstances.

Waiting for the results of your tests can be difficult. However, it's important for your medical team to know exactly what type of lymphoma you have and where it is. This helps them plan the most appropriate treatment for you.

Your 'prognostic score'

Your doctor might use your test results to give you a score that can help predict your likely outcome after treatment. This is called a 'prognostic score'. One of the most common in the UK is the mantle cell prognostic index ('MIPI'), which gives you a score based on:

- your age
- your white blood cell count
- the level of a protein called **lactate dehydrogenase** (LDH) in your blood
- your ability to carry on with normal activities.

Some scoring systems also include how fast the lymphoma cells are dividing.

Your doctor might use your prognostic score to help decide on the most appropriate treatment for you.

Outlook

In around 1 in 10 people, mantle cell lymphoma grows slowly and causes few or no symptoms. Under a microscope, it has features of a low-grade lymphoma. These people might not need treatment for a long time, sometimes years.

In most people, mantle cell lymphoma is fast-growing and treatment needs to start straightaway. First-line treatment is usually successful at putting your lymphoma into remission (shrinking the lymphoma or getting rid of it completely) but the lymphoma almost always comes back within months or years, and needs more treatment.

In the last few years, several new treatments have become available that have improved the outlook for many people with mantle cell lymphoma.

Your medical team are best placed to advise you on your specific outlook based on your individual circumstances. They can use the results of your tests and other factors (for example, your age and physical fitness) to help choose the best treatment for you.

If you choose to research survival statistics, it is important to remember that they don't tell you what your individual outlook is – they only tell you how a group of people with the same diagnosis did over a period of time. They are usually measured 5 years or more after treatment, so they only tell you how people did in the past. Treatment options for mantle cell lymphoma have improved a lot in recent years and those people might not have received the same treatment as you. Many people do not find survival statistics helpful because of this variability.

Treatment

Treatment for mantle cell lymphoma depends on several factors including:

- the exact type of lymphoma you have, including how quickly it is growing
- your **prognostic score**
- what symptoms you have
- the stage of your lymphoma
- your age and general health
- your feelings about the treatment options

Slow-growing mantle cell lymphoma

If you have a slow-growing form of mantle cell lymphoma and you don't have troublesome symptoms, you might not need treatment straightaway. Instead, your doctor might recommend an approach called '**active monitoring**' (or 'watch and wait'). This involves having regular check-ups with your medical team to monitor your health and to see how the lymphoma is affecting you. You do not have treatment for the lymphoma unless it starts causing significant health problems. Active monitoring means that while you are well, you avoid the side effects of treatment for as long as possible. Treatment is still available when you need it, but this might not be for many months or, rarely, years.

If you develop troublesome symptoms, your lymph nodes become very large, or the lymphoma starts to affect your organs or blood counts, your medical team are likely to recommend treatment. The treatment options are usually the same as for fast-growing mantle cell lymphoma.

Fast-growing mantle cell lymphoma

The treatment your medical team recommends for you depends on the stage of your lymphoma and the signs and symptoms you have. **Stage 1** or **stage 2** mantle cell lymphoma is known as 'early-stage' lymphoma. **Stage 3** or **stage 4** mantle cell lymphoma is known as 'advanced-stage' lymphoma. Most people have advanced stage mantle cell lymphoma when they are diagnosed.

Early-stage mantle cell lymphoma

If you have early-stage mantle cell lymphoma, you might be offered **radiotherapy** to the affected area. This can be very successful, leading to long-lasting responses.

Advanced-stage mantle cell lymphoma

If you have advanced-stage mantle cell lymphoma that needs treatment, your consultant might ask you if you'd like to take part in a **clinical trial**. If there isn't a clinical trial suitable for you, or if you don't want to take part in a clinical trial, your medical team are likely to suggest a course of **chemotherapy** combined with an **antibody therapy** such as **rituximab**.

The exact combination of drugs (**chemotherapy regimen**) your team recommends depends on the results of your tests and your individual circumstances.

- People who are fit enough are usually offered stronger (more intensive) chemotherapy that includes a drug called cytarabine. This can be very effective in mantle cell lymphoma and might also help prevent the lymphoma spreading to your central nervous system. If you respond to this treatment, your medical team might suggest high-dose chemotherapy with a **stem cell transplant** using your own stem cells (an **autologous stem cell transplant**). This aims to keep your lymphoma under control as long as possible. High-dose chemotherapy is a very intense form of treatment and you have to be fit enough to have it. After this treatment, most people have **maintenance therapy** to help their **remission** (the time when your lymphoma has shrunk or gone completely) last as long as possible. Maintenance therapy involves having an injection of rituximab every 2 months for up to 3 years.

- If you are not fit enough to have intensive treatment, you might be offered a standard chemotherapy regimen combined with rituximab. The most common regimens are rituximab plus **CHOP** (R-CHOP), rituximab plus bendamustine or a variation of R-CHOP called VR-CAP, which includes a targeted drug called **bortezomib**. Your consultant will suggest the best option for you. If you respond to treatment, especially R-CHOP, you might be offered **maintenance therapy** to help keep your lymphoma under control for as long as possible. This involves having an injection of rituximab every 2 months for 2 to 3 years.
 - If you are not well enough to have standard chemotherapy, or you have other health conditions that mean it is not suitable for you, you might have a gentler (lower intensity) regimen such as rituximab plus chlorambucil (a chemotherapy drug that you take as a tablet) or rituximab plus **CVP**.
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Follow-up

You have a **scan** when you finish your treatment to see how you have responded. This is often a CT scan. If you are due to have a stem cell transplant, you usually have a PET/CT scan. If you had lymphoma in your bone marrow before you started treatment, you might have another bone marrow biopsy.

You might also have other tests. Your doctor uses the results of the scans and tests to see if you are in **remission** (disappearance or significant shrinkage of lymphoma) or if you need further treatment.

When you are in remission after treatment, you have regular **follow-up appointments**. These are to:

- check that you are recovering well from treatment
- give you your maintenance therapy (if you are having it)
- check for signs of the lymphoma coming back (relapsing)
- check whether you are developing any **late effects** (side effects that develop months or years after treatment).

At each appointment, your doctor examines you and asks if you have any concerns or symptoms. You might have blood tests. You are unlikely to have a scan unless you have new or worsening symptoms that could be a sign of your lymphoma coming back.

Relapsed and refractory mantle cell lymphoma

How long lymphoma stays under control (in remission) after successful treatment varies from person to person. At some point, mantle cell lymphoma usually **relapses**. 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 feelings about further treatment
- the treatments available (including **clinical trials**).

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

Occasionally, lymphoma doesn't respond well to your first treatment. This is called '**refractory**' lymphoma. Refractory lymphoma is usually treated in a similar way to relapsed lymphoma. Possible treatment options for relapsed or refractory mantle cell lymphoma include:

- A **targeted drug** such as **ibrutinib**. This is the most common second treatment for mantle cell lymphoma.
- Another targeted drug or experimental treatment as part of a **clinical trial**.
- A **chemotherapy regimen** you haven't had before, combined with **rituximab**.
- **CAR T-cell therapy** if you've already been treated with ibrutinib.
- A **donor (allogeneic) stem cell transplant**, if you've responded to further treatment and you are fit enough. This is a very intensive treatment and is usually only offered if you've already had an **autologous stem cell transplant**. However, some people with mantle cell lymphoma that doesn't respond to treatment (refractory lymphoma) might have an allogeneic transplant instead of autologous stem cell transplant.
- Supportive care (sometimes known as **palliative care**) to control symptoms.

Research

Scientists are testing many different treatment options for mantle cell lymphoma in **clinical trials**, including some treatments that are already approved for other types of lymphoma. Many of these are new targeted drugs that work by helping your own immune system get rid of the lymphoma.

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

New types of treatment that are being tested in people with mantle cell lymphoma include:

- New **antibody therapies**, including antibodies that bind to two different targets (one on lymphoma cells and one on T cells, which helps the T cells find and destroy the lymphoma cells). These are called 'bispecific' antibodies.
- **Cell signal blockers**, which block signals that cancerous B cells send to help them divide or stay alive. Cell signal blockers include targeted drugs such as BTK inhibitors (for example, **acalabrutinib** and **zanubrutinib**, which belong to the same drug family as **ibrutinib**) and BCL-2 inhibitors (for example, **venetoclax**).

Some of these might be available to you through a clinical trial. If you are interested in taking part in a clinical trial, ask your doctor if there is a trial that might be suitable for you. To find out more about clinical trials or to search for a trial that might be suitable for you, visit **Lymphoma TrialsLink**.

References

The full list of references for this page is available on our website. Alternatively, email publications@lymphoma-action.org.uk or call 01296 619409 if you would like a copy.

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