

## MALT lymphoma

This information is about MALT lymphoma (also known as extranodal marginal zone lymphoma). This is a slow-growing type of non-Hodgkin lymphoma. It most commonly develops in the stomach (called gastric MALT lymphoma) but it can develop in other parts of the body (called non-gastric MALT lymphoma).

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We have separate information about the topics in **bold font**. Please get in touch if you'd like to request copies or if you would like further information about any aspect of lymphoma. Phone 0808 808 5555 or email [information@lymphoma-action.org.uk](mailto:information@lymphoma-action.org.uk).

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# What is MALT 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**, helping fight infections. There are two types of lymphocyte: **B lymphocytes (B cells)** and **T lymphocytes (T cells)**. Lymphomas can be grouped as **Hodgkin lymphomas** or **non-Hodgkin lymphomas**, depending on the types of cell they contain.

MALT lymphoma is a slow-growing (low-grade) non-Hodgkin lymphoma that develops from B cells. It is a type of marginal zone lymphoma. Marginal zone lymphomas develop in a particular region at the edge of lymphoid tissues (collections of lymphocytes) called the marginal zone.

There are three types of marginal zone lymphoma:

- MALT lymphoma (extranodal marginal zone lymphoma of **mucosa-associated lymphoid tissue**), which develops in lymphoid tissue outside lymph nodes
- **nodal marginal zone lymphoma**, which develops in lymph nodes
- **splenic marginal zone lymphoma**, which develops in the spleen.

MALT lymphoma is the most common type of marginal zone lymphoma, but it is still rare. Only around 8 in every 100 people diagnosed with non-Hodgkin lymphoma have MALT lymphoma.

MALT lymphoma can develop almost anywhere in the body, but it most often develops in the stomach. MALT lymphomas are normally divided into:

- **gastric (stomach) MALT lymphoma**, which makes up about a third of all MALT lymphomas
- **non-gastric MALT lymphoma**, which most commonly affects the salivary glands, thyroid, lungs, skin, bowel (gut) or tissues around the eye.

## What is 'mucosa-associated lymphoid tissue' (MALT)?

'Mucosa' is the soft, moist, protective layer of cells that lines many parts of your body, such as your mouth, gut, airways and some internal organs. 'Lymphoid tissue' means a collection of lymphocytes. Mucosa-associated lymphoid tissue (MALT) refers to collections of lymphocytes in your mucosa. It includes your tonsils at the back of your throat, as well as lymphoid tissue that might be scattered around the rest of your body, such as your gut, lungs, skin, **thyroid gland** and salivary (spit) glands.

MALT can form part of your body's normal response to infection. However, MALT lymphomas can develop if abnormal lymphocytes collect in this lymphoid tissue.

## Who gets MALT lymphoma?

MALT lymphoma can affect people of any age but it typically develops in people over the age of 60.

MALT lymphoma is more common in people who have, or have had, an infection. Scientists think this is due to long-term inflammation, which can prompt MALT to form.

Bacterial infections are linked with MALT lymphoma developing in different parts of the body.

- Gastric MALT lymphoma is strongly linked to infection with a bacteria called *Helicobacter pylori* (*H. pylori*). This is a very common infection that can cause stomach ulcers and indigestion. It doesn't usually cause serious problems. Around 9 out of every 10 people with gastric MALT lymphoma have *H. pylori* infection. However, most people who have *H. pylori* infection do **not** go on to develop gastric MALT lymphoma.
- MALT lymphoma affecting the tear ducts and other tissues around the eye might be linked to infection with a bacteria called *Chlamydophila psittaci*, which is spread by birds and can cause lung infections.
- MALT lymphoma affecting the skin might be linked to infection with a bacteria called *Borrelia burgdorferi*, which is spread by ticks and causes **Lyme disease**.
- MALT lymphoma affecting the small bowel might be linked to infection with a bacteria called *Campylobacter jejuni*, a common cause of food poisoning.
- MALT lymphoma affecting the lungs might be linked to infection with a bacteria called *Achromobacter xylosoxidans*, which can cause lung infections, usually in people with lowered immune systems.

MALT lymphoma affecting some parts of the body are more common in people who have certain **autoimmune conditions**.

- MALT lymphoma affecting the salivary (spit) glands might be linked to an autoimmune condition called **Sjögren's syndrome**.
- MALT lymphoma affecting the thyroid gland might be linked to an autoimmune condition called **Hashimoto's thyroiditis**.

Like other marginal zone lymphomas, MALT lymphoma is also more common in people who have hepatitis C virus infection.

However, the vast majority of people who have these infections or autoimmune conditions do **not** go on to develop lymphoma.

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## Symptoms of MALT lymphoma

The **symptoms** of MALT lymphoma depend on where the lymphoma develops. Unlike many other types of lymphoma, MALT lymphoma develops outside the lymph nodes. It does not usually cause **swollen lymph nodes**. People with MALT lymphoma very rarely experience the classic 'B symptoms' often associated with lymphoma (a combination of **unexplained weight loss, night sweats** and/or **fever**).

### Symptoms of gastric MALT lymphoma

Gastric MALT lymphoma involves the stomach. It might not cause any symptoms at all – it could be found during tests for something else. However, it can cause symptoms such as:

- persistent indigestion (this is often the only symptom)
- tummy pain
- feeling sick (nausea) or being sick (vomiting)
- weight loss.

Some people have symptoms of **anaemia** (for example, tiredness or shortness of breath) caused by bleeding in the stomach.

### Symptoms of non-gastric MALT lymphoma

Most people with MALT lymphoma that develops outside the stomach (non-gastric MALT) have no symptoms at all and the lymphoma is found when they have a test to investigate something else.

Some people might have symptoms. These depend on where the lymphoma develops.

- MALT lymphoma affecting your gut (bowel) can make it difficult for you to absorb enough food. You might have diarrhoea, bloating or tummy pain. You might feel very tired and lose weight without trying to.
- MALT lymphoma affecting your tear ducts or tissues around your eye might cause symptoms such as redness of the eye, dark pink lumps inside your eyelid or the corners of your eye, double vision, a droopy eyelid or a bulging eye. Symptoms usually develop on one side but in around 1 in 10 cases, they can affect both eyes.
- MALT lymphoma affecting your lungs might cause a cough, shortness of breath, coughing up blood, or chest pain.

- MALT lymphoma affecting your salivary (spit) glands might cause a lump in front of your ear or in your mouth or jaw.
- MALT lymphoma affecting your skin might cause pink, red or purplish patches or lumps to develop on your skin, most often on your arms, chest or back. They can develop in one place or in several places and are very slow-growing.
- MALT lymphoma affecting your thyroid gland might cause a lump at the front of your neck, a hoarse voice, shortness of breath or difficulty swallowing. Some people might have symptoms of an underactive thyroid (for example, feeling tired a lot of the time, being sensitive to the cold and putting on weight).

Other areas of your body can be affected.

Up to half of people with MALT lymphoma have lymphoma in several places in their body. If this is the case for you, you might have a mixture of symptoms.

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## Diagnosis and staging of MALT lymphoma

MALT lymphoma is usually diagnosed by removing a sample of tissue and looking at it under a microscope. This involves a small procedure called a **biopsy**, which is usually done under a local anaesthetic. The biopsy sample is examined by an expert lymphoma **pathologist**. The pathologist also tests the sample for particular proteins that are found on the surface of lymphoma cells. This can help your medical team decide on the most appropriate treatment for you.

The type of biopsy you need, and how it is carried out, depends on where your lymphoma has developed. If your lymphoma is in your stomach, bowel or lungs, the biopsy is likely to be taken during a test called an **endoscopy**. This uses a thin, flexible tube with a tiny camera to look inside your body. You might have a sedative (a drug to relax you) first. Small surgical tools can be threaded through the tube so your doctor can take biopsy samples.

- If the abnormal tissue is in your stomach, the tube is passed through your mouth down your gullet and into your stomach (gastroscopy).
- If the abnormal tissue is in your bowel, the tube is passed through your back passage into your gut (colonoscopy)
- If the abnormal tissue is in your lungs, the tube is passed through your nose or mouth into the airways of your lungs (bronchoscopy).

You will also have other tests to help your medical team work out the best treatment for you.

If your lymphoma involves your stomach or bowel, you are likely to have a breath test to check for *H. pylori* in your stomach as *H.pylori* infection is a treatable condition linked to the development of some MALT lymphomas. You are given a capsule or drink containing a harmless, tasteless chemical. If you have *H. pylori* infection, it breaks the chemical down into carbon dioxide. About 30 minutes later, you have to blow into a tube or bag that tests the level of carbon dioxide in your breath. You might also have to provide a sample of your stool (faecal sample) to test for *H. pylori* infection.

You will also have **blood tests** to look at your general health, check your **blood cell counts**, make sure your kidneys and liver are working well and test for infections such as *H. pylori* and hepatitis C virus.

You will have other tests to find out which areas of your body are affected by lymphoma. This is called **staging**. It usually involves having a **CT scan**. You might also have other staging tests depending on where the lymphoma is growing:

- an **ultrasound scan** to look for lymphoma in your salivary (spit) glands, thyroid gland or the wall of your stomach or gut
- an **MRI scan** if you have lymphoma around your eye or brain
- a chest **X-ray** if you have lymphoma affecting your lungs
- a **PET scan** if your specialist thinks it would be helpful in planning your treatment, although this is less common for marginal zone lymphomas than other types of lymphoma
- a sample of your bone marrow cells collected (a **bone marrow biopsy**) to check if you have lymphoma cells in your bone marrow, although this is rare in MALT lymphomas.

You usually have your tests done as an outpatient. It takes a few weeks to get all the results. **Waiting for test results** can be a worrying time, but it is important for your doctor to gather all of this information in order to plan the best treatment for you.

MALT lymphomas grow slowly so most people have **early-stage** (stage 1 or 2) lymphoma when they are diagnosed.

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## Outlook for MALT lymphoma

MALT lymphoma develops slowly and treatment is usually successful. Although the lymphoma often comes back (**relapses**), it can be treated again to keep it under control. Most people live with this type of lymphoma for many years. You might have periods when you feel well and don't need treatment, and other periods when your symptoms get worse and you need more treatment.

Your doctor is best placed to advise you on your outlook based on your individual circumstances.

### Transformation

Occasionally, MALT lymphoma can change (**transform**) into a faster-growing type of lymphoma. This happens in less than 1 in every 10 people with MALT lymphoma.

If your doctor thinks your lymphoma might have transformed, you will usually have a **biopsy** to check for faster-growing lymphoma cells, and a **PET/CT scan** to see which areas of your body are affected. Transformed MALT lymphoma is treated in the same way as fast-growing (high-grade) non-Hodgkin lymphoma such as **diffuse large B-cell lymphoma (DLBCL)**.

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## Treatment of MALT lymphoma

Treatment for MALT lymphoma depends on where the lymphoma is, what symptoms you have and whether you have an infection linked to the lymphoma. The main treatment options are:

- **treating infections that might be linked to the lymphoma**
- **active monitoring (watch and wait)**
- **radiotherapy**
- **chemo-immunotherapy**.

Surgery is used very occasionally if the lymphoma is only affecting one part of your body and is in an area where it can be safely removed.

### Treating infections linked to MALT lymphoma

If you have gastric or bowel MALT lymphoma, the first treatment is usually to treat *H. pylori* infection. You might have this treatment even if your tests haven't found evidence of *H. pylori* infection because it can be difficult to detect.

*H. pylori* treatment usually involves a 7 to 14-day course of antibiotics as well as acid-reducing medication. You have a breath test or stool (faecal) sample test 6 to 8 weeks after your antibiotic treatment to check if the infection has gone. You might need another course of antibiotics if the infection hasn't cleared.

Treating *H. pylori* infection clears gastric MALT lymphoma in most people but it can take 6 to 12 months to respond to the treatment. You have a repeat endoscopy to check if the lymphoma has responded to treatment. If you still have some lymphoma left after antibiotic treatment for *H. pylori*, you might have active monitoring, radiotherapy or chemo-immunotherapy, depending on how much lymphoma there is and how it is affecting you.

If you have other forms of non-gastric MALT lymphoma and your doctor thinks your lymphoma might be linked to an infection, they might suggest antibiotics as a first treatment. However, when treatment is required, most people with non-gastric MALT lymphoma have radiotherapy or chemo-immunotherapy.

## Active monitoring

If your lymphoma is not causing symptoms, you might not need treatment straightaway (apart from antibiotics, if your lymphoma is linked to an infection). Instead, your doctor might suggest monitoring you closely – including regular endoscopies if you have gastric MALT – and saving treatment until it is needed. This is called **active monitoring or 'watch and wait'**. This approach allows you to avoid the side effects of treatment for as long as possible. Delaying treatment in this way does not affect how well it works when you do need it, or how long you might live.

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**I had a meeting with a very helpful nurse specialist who said I could contact her if I ever needed to between appointments, which were roughly every three months. Feelings of anxiety arose as I felt for lumps and bumps before each appointment, but much of the time I was able to forget about the diagnosis altogether**

Pat, who was diagnosed with non-gastric MALT lymphoma with a period of active monitoring

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Read more about **Pat's story** on our website.

## Radiotherapy

If your lymphoma is causing symptoms and it is not linked to an infection, or treating the infection doesn't clear it, you might be treated with **radiotherapy**. Radiotherapy can be very effective if your lymphoma is only affecting one or a few places close together in your body. However, you might need chemo-immunotherapy if:

- your lymphoma is too widespread for radiotherapy
- your lymphoma is close to important organs that might be damaged by radiotherapy
- your doctor thinks you have a high risk of your lymphoma getting worse or coming back.

## Chemo-immunotherapy

Most people with widespread MALT lymphoma that is not linked to an infection, or which doesn't clear after treating the infection, have **antibody therapy** (such as **rituximab**), either on its own, or combined with **chemotherapy**. When used together this is called 'chemo-immunotherapy'.

Combinations of chemo-immunotherapy drugs that might be used to treat MALT lymphoma include:

- rituximab plus bendamustine
- rituximab plus chlorambucil
- rituximab plus **CVP** (R-CVP)
- rituximab plus **CHOP** (R-CHOP).

If you have had successful chemo-immunotherapy, your doctor may recommend rituximab as a **maintenance therapy**. Maintenance rituximab is usually given by a subcutaneous (under the skin) injection every 2 to 3 months for up to 2 years. Maintenance therapy may prolong the length of your remission. Your doctor will discuss the potential risks and benefits of maintenance rituximab if this is an option for you.

MALT lymphoma is rare. This makes it difficult to determine exactly which treatment gives the best outcome. Your doctor might recommend a different regimen (combination of drugs), or they might ask you if you'd like to take part in a **clinical trial**.

## Side effects of treatment

Treatments affect people differently. Each type of treatment or drug has a different set of possible **side effects**. Your medical team should give you information about the side effects associated with any treatment they recommend for you. Ask for more information if you are worried. Your medical team can offer advice or prescribe medicines if you experience troublesome side effects during your lymphoma treatment.

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## Follow-up of MALT lymphoma

When you are in **remission** (disappearance or significant shrinkage of lymphoma) after your treatment, or during a period of active monitoring ('watch and wait'), you have regular **follow-up appointments** in the clinic.

Your follow-up appointments are to check that:

- you are recovering well from any treatment
- you have no signs of the lymphoma coming back (relapsing) or getting worse
- you are not 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. If you had gastric MALT lymphoma, you might have follow-up endoscopies every few months in the first 2 years after your treatment, followed by endoscopies every 12 to 18 months to monitor for late complications. You might have other tests depending on where your lymphoma affected you.

You are unlikely to have a scan unless you have troubling symptoms.

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## Relapsed and refractory MALT lymphoma

MALT lymphoma may **come back (relapse)** after successful treatment. This can be several years later. If MALT lymphoma comes back, it often affects the same organ as before but it can come back in other places. If you had MALT lymphoma affecting one eye, salivary (spit) gland or lung or on one side of your thyroid gland, it might come back on the opposite side.

You can usually have more treatment to give another period of remission. This approach can often control the lymphoma for many years. The treatment you have for relapsed or refractory MALT lymphoma depends on where the lymphoma is, what treatment you've already had and how long you responded to it.

If you have gastric MALT lymphoma and *H. pylori* infection has come back, more **antibiotic-based treatment** usually clears both the infection and the lymphoma.

If you have non-gastric MALT or gastric MALT lymphoma without *H. pylori* infection, you are likely to need other treatments.

Several **targeted drugs** are being tested in **clinical trials**. Your doctor might ask you if you would like to take part in a clinical trial to help test new treatments and to find out what the best treatment is for MALT lymphoma that has come back (relapsed) or has not responded to previous treatment (refractory lymphoma).

If you don't want to take part in a clinical trial, or if there isn't one that is suitable for you, you are most likely to be treated with **radiotherapy** or with **rituximab** combined with **chemotherapy**.

Very occasionally, your medical team might recommend that you have a **stem cell transplant** using your own stem cells (an '**autologous stem cell transplant**'). Stem cell transplants are usually only considered for MALT lymphoma if it relapses very soon after previous treatment. Stem cell transplants are a very intense form of treatment and you have to be fit enough to have one. A stem cell transplant allows you to have very high doses of chemotherapy. It might give you a better chance of having a long-lasting remission than standard chemotherapy regimens.

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## Research and targeted treatments

MALT lymphoma is uncommon so most clinical trials also include people with other types of low-grade lymphoma, including other marginal zone lymphomas.

There are several types of treatment being tested in clinical trials for marginal zone lymphomas, including drugs already approved for other types of lymphoma. These include:

- **B-cell receptor pathway inhibitors**, such as BTK inhibitors, which block signals involved in the growth and survival of lymphoma cells
- **proteasome inhibitors** which disrupt the balance of proteins in lymphoma cells
- **BCL-2 inhibitors** which activate the process of natural cell death
- **immunomodulators** which change how your immune system responds to lymphoma cells
- **antibody treatments** which bind to proteins on the surface of lymphoma cells to help your own immune system destroy them
- **CAR T-cell therapy** which uses modified cells from your own immune system to fight lymphoma.

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](#).

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## References

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

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