

MALT lymphoma (gastric and non-gastric)

This page 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 (when it is called gastric MALT lymphoma) but it can develop in other parts of the body (which is 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.

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 you 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: a lymphoma that develops 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, gut (bowel) or tissues around the eye.

What is 'mucosa-associated lymphoid tissue'?

'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 may be scattered around the rest of your body, such as your gut, lungs, skin, **thyroid gland** and salivary (spit) glands.

MALT can form as part of your body's normal response to inflammation. It helps protect you from infections. 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 in their mid-60s.

MALT lymphoma is more common in people who have been infected with certain viruses or who have particular **autoimmune conditions**. Scientists think this is due to long-term inflammation, which can prompt MALT tissue to form.

- Gastric MALT lymphoma is strongly linked to infection with 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 an infection called *Chlamydia psittaci*, which is spread by birds and can cause lung infections.
- MALT lymphoma of the skin might be linked to infection with *Borrelia burgdorferi*, which is spread by ticks and causes **Lyme disease**.
- MALT lymphoma affecting the small bowel might be linked to infection with *Campylobacter jejuni*, a common cause of food poisoning.
- MALT lymphoma affecting the lungs might be linked to infection with *Achromobacter xylosoxidans*, which can cause blood or lung infections, usually in people with lowered immune systems.
- 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.

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 'B symptoms' (a combination of **unexplained weight loss, night sweats** or **fever**).

Symptoms of gastric MALT lymphoma

Gastric MALT lymphoma might not cause any symptoms at all, and 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 often doesn't cause any symptoms but it can cause a cough, shortness of breath, coughing up blood, or chest pain.
- MALT lymphoma affecting your spit (salivary) 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 1 in 2 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.

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 operation 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. For example:

- If the abnormal tissue is in your stomach or bowel, the biopsy is likely to be taken during a test called an **endoscopy**. This uses a thin, flexible tube with a tiny camera in its tip to look inside your body. You might have a sedative (a drug to relax you) first. To look at your stomach, the tube is passed through your mouth down your gullet and into your stomach (gastroscopy). To look at your bowel, the tube is passed through your back passage into your gut (colonoscopy). Small surgical tools can be threaded through the tube so your doctor can take biopsy samples. They might take samples from a few different places in your stomach or bowel.
- If the abnormal tissue is in your lungs, you might have a type of endoscopy called a bronchoscopy, where the tube is passed through your nose or mouth into the airways of your lungs.

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

You are likely to have a breath test to check for *H. pylori* in your stomach. 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 poo (faecal sample) to be tested for *H. pylori* infection.

You also have **blood tests** to check for *H. pylori* infection, 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 hepatitis C virus.

You 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 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. You might have 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. Other staging tests depend on where the lymphoma is growing. You might have:

- an **ultrasound scan** to look for lymphoma in your spit glands, thyroid gland or the wall of your stomach or gut
- an **MRI scan** if you have lymphoma around your eye
- a chest **X-ray** if you have lymphoma affecting your lungs.

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.

Outlook for MALT lymphoma

MALT lymphoma develops slowly and treatment is usually successful. Although the lymphoma often **relapses** (comes back), 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 might have a **biopsy** to check for faster-growing lymphoma cells, or 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)**.

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

The first treatment for gastric MALT lymphoma is usually to treat *H. pylori* infection. You might be given 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-day course of antibiotics and acid-reducing medication. You have a breath 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 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, most people with non-gastric MALT lymphoma need radiotherapy or chemo-immunotherapy.

Active monitoring

If your lymphoma is not causing troublesome symptoms, you might not need treatment straightaway (apart from antibiotics, if it 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.

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 a few places in your body. However, you might need chemo-immunotherapy instead if:

- your lymphoma is more widespread
- 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 on the opposite side of your body.

Chemo-immunotherapy

Most people with MALT lymphoma that is not linked to an infection, or which doesn't clear after treating the infection, have **antibody therapy** (such as **rituximab**) combined with **chemotherapy**. This is sometimes called 'chemo-immunotherapy'.

MALT lymphoma is rare. This makes it difficult to determine exactly which treatment gives the best outcome.

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)
- rituximab plus fludarabine (rarely used nowadays).

However, 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 about potential side effects. Your medical team can offer advice or prescribe medicines if you experience troublesome side effects during your lymphoma treatment.

Follow-up of MALT lymphoma

When you are in **remission** (no evidence 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 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 an endoscopy every 3 to 6 months for the first 2 years after your treatment. You might have other tests depending on where your lymphoma affected you.

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

Relapsed and refractory MALT lymphoma

MALT lymphoma often **comes back (relapses)** after successful treatment, often 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, 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 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 (no evidence of lymphoma) than standard chemotherapy regimens.

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 targeted treatments being tested in clinical trials for marginal zone lymphomas, including drugs already approved for other types of lymphoma. These include:

- BTK inhibitors such as **ibrutinib**, acalabrutinib and zanubrutinib, which block signals that B cells send to help them stay alive and divide
- PI3K inhibitors such as **idelalisib**, umbralisib and copanlisib, which block a protein involved in the growth and survival of lymphoma cells
- immunomodulators such as **lenalidomide**, which change how your immune system works
- **proteasome inhibitors** such as bortezomib, which disrupt the balance of proteins in lymphoma cells, interfering with chemical signals between lymphoma cells and leading to cell death
- new **antibody treatments** such as **obinutuzumab**, ublituximab or varlilumab, which bind to proteins on the surface of lymphoma cells to help your own immune system destroy them.

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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