MALT lymphoma (extranodal marginal zone lymphoma)

This information page is about MALT lymphoma – a slow-growing type of non-Hodgkin lymphoma. It most commonly develops in the stomach (gastric MALT lymphoma) but can develop in other parts of the body (non-gastric MALT lymphoma).

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What is MALT lymphoma?

Lymphoma is a type of cancer. It happens when a growth of a lymphocyte (white blood cell) population goes out of control.

Mucosa-associated lymphoid tissue (MALT) lymphoma is a type of low-grade (slow-growing) non-Hodgkin lymphoma (NHL).

MALT lymphoma belongs to a group of marginal zone lymphomas. Marginal zone lymphomas develop from B lymphocytes (B cells) that are normally found in the marginal zone. The marginal zone is at the edge of areas of lymphoid tissue and is where B cells are found. Lymphoid tissue (where lymphocytes collect) is part of the immune system, for example the lymph nodes or spleen.

MALT lymphomas are uncommon – around 1 in 13 cases of NHL are MALT lymphomas.

What is mucosa-associated lymphoid tissue?

Mucosal tissue is the soft, moist, protective tissue that lines many parts of your body, e.g. your mouth, gut, breathing passages and other internal organs. Lymphoid tissue is normally found in mucosal tissue of the gut – mucosa-associated lymphoid tissue (MALT).

MALT helps protect the body from infections and toxins in the gut. In areas outside the gut, similar lymphoid tissue can form when normal lymphocytes collect in tissue outside lymph nodes in response to an infection or inflammation (a reaction to injury, irritation or infection). This is a normal process. However, MALT lymphomas can develop when abnormal lymphocytes collect in this lymphoid tissue.

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

- gastric (stomach) MALT lymphoma – about a third of MALT lymphomas
- non-gastric MALT lymphoma, which most commonly affects the salivary glands, thyroid, lungs, skin, bowel or tissues around the eye.
Who gets MALT lymphoma and what causes it?

MALT lymphomas can affect people of any age but they are most common in people in their 50s and 60s.

What causes MALT lymphoma?

MALT lymphoma develops in areas where MALT tissue has formed in response to:

- **inflammation** caused by a chronic (long-lasting) infection
- **an autoimmune condition** (a condition in which the body’s immune system reacts against its own tissue instead of protecting it).

It is a normal process for MALT tissue to form in these cases. MALT tissue rarely becomes a lymphoma.

Gastric MALT lymphoma has been strongly linked to infection by *Helicobacter pylori*, a type of bacteria. This is a very common infection that doesn’t usually cause serious problems but is also linked to stomach ulcers and indigestion.

Some research suggests that some conditions could be linked to a higher risk of developing MALT lymphoma in certain areas of the body:

- **Sjögren syndrome**, an autoimmune condition that causes dry mouth and dry eyes, may be linked to MALT lymphoma in the salivary glands and lungs.
- **Chlamydia psittici infection**, which can be caught from birds, including those kept as pets (e.g., parrots), may be linked to MALT lymphoma in the tissues surrounding the eye and lacrimal (tear) glands.
- **Hashimoto’s thyroiditis**, an autoimmune inflammation of the thyroid gland, may be linked to MALT lymphoma in the thyroid.
- **Borrelia burgdorferi infection**, which causes Lyme disease and is carried by infected ticks, may be linked to MALT lymphoma of the skin.
• *Campylobacter jejuni* infection, a common cause of food poisoning, may be linked to MALT lymphoma of the small bowel.

• *Rheumatoid arthritis* is linked to MALT lymphoma of the lungs.

Most people with any of these conditions never develop lymphoma.

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**Symptoms**

The symptoms of MALT lymphoma depend on where the lymphoma develops. Unlike many other types of lymphoma, MALT lymphomas develop in areas outside the lymph nodes (glands), which are called ‘extranodal’ sites.

**Symptoms of gastric MALT lymphoma**

Most people with gastric MALT lymphoma have persistent indigestion. This is likely to be related to the *H. pylori* infection rather than to the lymphoma. Indigestion often improves when the infection is treated, regardless of whether the lymphoma decreases. Indigestion is usually the only symptom.

Other symptoms of gastric MALT lymphoma can include:

• abdominal (tummy) pain
• nausea and vomiting
• weight loss.

Some people have symptoms of anaemia (for example, tiredness or shortness of breath) caused by bleeding in the stomach. Rarely, severe abdominal (tummy) pain or a mass (lump) in the abdomen develops.

Gastric MALT lymphoma is normally diagnosed when people have an endoscopy (thin tube with a camera passed through the mouth) to investigate these symptoms. However, there are many more common causes of such symptoms and the vast majority of cases of *H. pylori* do not lead to lymphoma.
Symptoms of non-gastric MALT lymphoma

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

Some people have symptoms. The symptoms depend on where the lymphoma develops, for example:

- **salivary glands**: lump in a salivary gland, eg in front of the ear or the jaw
- **tissues surrounding the eye or in the lacrimal (tear) glands**: redness of the eye, dislike of bright light, lumps in the corners of the eye
- **thyroid**: lump in the neck, problems with swallowing
- **lung**: cough, shortness of breath, coughing up blood, fevers, weight loss; it often causes no symptoms and is picked up on a chest X-ray
- **skin**: one or more red-brown lumps or flat, discoloured areas on the back or arms and legs
- **bowel**: often involves general symptoms, such as weight loss, fever and night sweats; you might have changes in your bowel habits, eg constipation.

Other areas of your body can be affected.

Around 1 in 3 to 1 in 2 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

You need a biopsy to confirm your diagnosis if lymphoma is suspected. A sample of tissue that is affected by lymphoma is removed during a small operation. The sample is then examined by an expert lymphoma pathologist. The pathologist does tests on the tissue to find out what type of lymphoma you have.

If the abnormal tissue is in your stomach or bowel, the biopsy is likely to be removed by endoscopy. In this test, a flexible tube with a light and a tiny camera in its tip is used. To look at your stomach, the tube is passed down through your mouth (gastroscopy). To look at your bowel, the tube is passed through your back passage (colonoscopy). You might also have an
ultrasound scan, which uses high-frequency sound waves, at the same time as an endoscopy. This type of scan can be used to see if the lymphoma has spread through the wall of your stomach or bowel.

If the abnormal tissue is in your lungs, you might have a bronchoscopy, where a tube is passed through your nose or mouth into your lungs.

You have other tests to 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 and your blood cell counts
- **X-rays and scans**, usually a CT scan but sometimes an MRI, eg if the lymphoma is around your eye. A PET scan might be done if your specialist thinks it would be helpful in planning your treatment.

What other tests you need depends on where the lymphoma is:

- You might have a **bone marrow biopsy** to see if the lymphoma is affecting your bone marrow. This is most likely if you have lymphoma in your lungs, bowel or eye socket.
- If the lymphoma is in your stomach, you might have a stool sample taken or other tests might be done to confirm *H. pylori* infection.
- If the lymphoma is in your bowel, you might have X-rays with a barium enema (fluid passed into your bowel through your back passage) or meal. Barium is a chemical that allows these parts of your body to be seen on X-rays.

**What does ‘stage’ mean?**

**Stage** describes how much of your body is affected by lymphoma.

- Stage 1 and 2 lymphomas are described as ‘early’ stage.
- Stage 3 and 4 lymphomas are described as ‘advanced’ stage.

If lymphoma is extranodal (outside the lymph nodes), an ‘E’ is usually added to the stage.
MALT lymphomas are extranodal. They grow slowly so most people have early-stage (stage 1E) lymphoma when they are diagnosed. If MALT lymphoma has spread outside of the area it started from, it might be described as ‘disseminated’. The outlook is similar for both early-stage and advanced-stage (disseminated) MALT lymphoma.

**Outlook**

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

However, MALT lymphomas develop slowly. The majority of people with MALT lymphomas are treated successfully even if the lymphoma is in several places in the body when they are first diagnosed.

**Treatment**

Your treatment depends on where the lymphoma is and whether you have an infection linked to the lymphoma.

- If your lymphoma is linked to an infection, clearing the infection clears the lymphoma in most people.
- If your lymphoma is not linked to an infection but is localised (in 1 or few areas), you may be given radiotherapy.
- Some people are given no treatment to start with if the lymphoma is not causing problems and they can’t have radiotherapy – this is called active monitoring or ‘watch and wait’.
- Some people need chemo-immunotherapy (chemotherapy with the antibody treatment rituximab), particularly people with widespread MALT lymphoma or MALT lymphoma that does not clear after treating any related infection.

Surgery is rarely used for MALT lymphoma as most cases can be treated successfully with less invasive methods.
Treating infections linked to MALT lymphoma

The first treatment for gastric MALT lymphoma is usually to treat the *H. pylori* infection. Treating the infection clears the lymphoma in most people.

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. In some cases, treating the infection also clears the lymphoma. However, most people with non-gastric MALT lymphoma need radiotherapy or chemo-immunotherapy.

**Treating *H. pylori***

You are normally given a combination of antibiotics and acid-reducing medication. If you have gastric MALT lymphoma without evidence of *H. pylori*, you may still be given this treatment as *H. pylori* can be difficult to detect.

You usually have a test (a breath test or stool test) to check you are clear of the infection about 6–8 weeks after you have finished antibiotic treatment.

About 3–6 months after the treatment has finished you have an endoscopy. You have a biopsy during the endoscopy. The biopsy is tested to make sure that the infection has cleared and the lymphoma is decreasing.

Some people need more than a single course of treatment to clear the infection.

If the lymphoma is reduced by treating the infection but there is still some lymphoma left, you may be monitored to see if it clears completely over time. This is often known as active monitoring or ‘watch and wait’. It can take many months, sometimes more than a year, to clear the infection and the lymphoma for some people. Other types of treatment after antibiotics are not always needed, even if the infection takes some time to clear completely.

A few people have residual lymphoma after treatment for *H. pylori* or the treatment does not reduce the lymphoma. These people need other
treatments, like chemo-immunotherapy or sometimes radiotherapy. You are most likely to need these types of treatment if:

- you don’t have a *H. pylori* infection
- the lymphoma has spread deeply through your stomach wall
- the lymphoma has spread to your lymph nodes
- you have certain genetic changes in your lymphoma cells – your doctor can tell you if you have such genetic changes.

### Radiotherapy

If your lymphoma is not linked to an infection, and treating the infection doesn’t clear it, you may be treated with radiotherapy. Radiotherapy is very effective if the lymphoma is in an area suitable for this type of treatment and is in a single area or just a couple of areas.

### Active monitoring or ‘watch and wait’

If your lymphoma is not getting worse or causing any symptoms, you may be monitored without treatment for a time. This is called active monitoring or ‘watch and wait’.

MALT lymphoma is slow-growing so treatment can usually be safely delayed until it is clear that the lymphoma is getting worse. This approach allows you to avoid the side effects of treatment for as long as possible. Delaying treatment has no effect on how well it works when you do need treating.

### Chemo-immunotherapy

Most people whose lymphoma is not linked to an infection, or which doesn’t clear after treating the infection, have chemo-immunotherapy. Chemo-immunotherapy for MALT lymphoma is chemotherapy in combination with the antibody treatment rituximab.

The chemotherapy most often used for MALT lymphoma is:

- chlorambucil tablets, or
- CVP (a combination of the intravenous chemotherapy drugs cyclophosphamide and vincristine together with tablets of the steroid prednisolone).

**Side effects of treatment**

Treatments affect people differently. Each type of treatment or drug has a different set of possible side effects. The treatments used for MALT lymphoma are generally well tolerated by most people. It is common to feel sick. Your medical team should give you more information about any side effects associated with your treatment. Ask for more information if you are worried about potential side effects. Your medical team can also give advice and treatment if you experience troublesome side effects during your treatment.

**Transformation**

MALT lymphoma can transform (change) into a faster-growing or more aggressive type of lymphoma. Overall, transformation happens in around 1 in 10 people with MALT lymphoma over the course of their disease. Transformed MALT lymphoma is treated like a high-grade (fast-growing) non-Hodgkin lymphoma. The most common treatment is the chemo-immunotherapy regimen (combination of drugs), R-CHOP, which includes:

- the antibody treatment rituximab
- 3 chemotherapy drugs: cyclophosphamide, doxorubicin (hydroxydaunorubicin), and vincristine (Oncovin®)
- the steroid prednisolone.

**Follow-up**

When you are in remission (no evidence of lymphoma) after your treatment, you have regular follow-up appointments in the clinic for several years. Your doctor makes sure you are recovering well and that your lymphoma is still in remission.
If you had gastric MALT lymphoma, you usually have an endoscopy every 3–6 months for at least 2 years. After a couple of years, appointments usually become less frequent if you remain well. You may have biopsies taken during the endoscopies.

You do not normally have scans at follow-up appointments unless you have developed new symptoms.

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**Relapsed and refractory MALT lymphoma**

Treatment for MALT lymphoma is successful for most people. MALT lymphoma can *relapse* (come back), but more treatment is usually successful at putting it back in remission. Non-gastric MALT lymphoma is more likely to come back than gastric MALT lymphoma.

If the *H. pylori* infection has come back, causing the lymphoma to come back, more antibiotic-based treatment usually clears both the infection and the lymphoma.

Some people with only a small amount of lymphoma might not need further treatment. These people may be monitored (*‘watch and wait’*) to see if the lymphoma clears on its own.

A few people whose lymphoma has relapsed need other treatments as well as antibiotic-based treatment to put their lymphoma back into remission. *Chemo-immunotherapy* and/or *radiotherapy* are very effective treatments for relapsed MALT lymphoma.

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**Research and targeted treatments**

Researchers carry out clinical trials to test different ways of treating MALT lymphoma. They aim to find out which treatments give the best outcomes with the least possible side effects. Clinical trials might test:

- which treatment works best for what people
whether different ways of giving treatments can improve outcomes, eg giving maintenance therapy to stop the lymphoma coming back after successful treatment

- the safety and effectiveness of newer drugs for MALT lymphoma, particularly for people whose lymphoma has relapsed or didn’t respond to initial treatments (refractory lymphoma).

Your doctor can advise whether there is a clinical trial that is suitable for you. You can also search for suitable clinical trials at Lymphoma TrialsLink.

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**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.


Further reading

- Glossary
- Chemotherapy
- Rituximab
- Active monitoring (‘watch and wait’)
- What is lymphoma?

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