

## Intestinal T-cell lymphomas

This page is about intestinal T-cell lymphomas, rare types of T-cell lymphoma that develop in the small bowel. They include enteropathy-associated T-cell lymphoma (EATL) and monomorphic epitheliotropic intestinal T-cell lymphoma (MEITL).

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## What is intestinal T-cell lymphoma?

Intestinal T-cell lymphoma is a rare type of **fast-growing (high-grade) non-Hodgkin lymphoma** that grows in your small bowel (gut or intestine). It develops from white blood cells called **T cells**, which is why it's called a **T-cell lymphoma**.

There are two types of intestinal T-cell lymphoma. They have complicated names based on their characteristics:

- Enteropathy-associated T-cell lymphoma (EATL) accounts for around 4 in every 5 cases of intestinal T-cell lymphoma. It used to be known as 'EATL type 1'. 'Enteropathy' means a 'disease of the intestines'.
- Monomorphic epitheliotropic intestinal T-cell lymphoma (MEITL) accounts for around 1 in every 5 cases of intestinal T-cell lymphoma. It used to be known as 'EATL type 2'. It is called MEITL because the abnormal T cells all look the same (they are 'monomorphic') and it grows in the lining of your intestine (your intestinal 'epithelium').

# Who gets intestinal T-cell lymphoma?

Intestinal T-cell lymphomas usually affect older people, often in their 60s. The different types of intestinal T-cell lymphoma affect different groups of people:

- EATL tends to affect people of northern European origin. It is strongly linked to an **autoimmune condition** called coeliac disease. Coeliac disease develops when your **immune system** reacts to gluten in the food you eat by making antibodies. These antibodies mistakenly attack your small bowel, so you can't absorb food properly. Coeliac disease is usually controlled by following a gluten-free diet. EATL only develops in people who have coeliac disease, although some people might not know they have coeliac disease until they are diagnosed with EATL. It is important to know that most people who have coeliac disease do **not** develop EATL.
- MEITL is not linked to coeliac disease. Scientists don't know what causes it. It is more common in people of Asian or Hispanic origin.

The [NHS website](#) and [Coeliac UK](#) have more information about coeliac disease.

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## Symptoms of intestinal T-cell lymphoma

The most common **symptoms** of intestinal T-cell lymphoma are bowel and stomach problems. You might have:

- **tummy (abdominal) pain**
- **weight loss**
- diarrhoea, which might have blood in it
- **fatigue**
- an itchy **rash**.

You might be malnourished (not getting enough nutrients) if you are not absorbing food properly. You might also have '**B symptoms**' (fevers, night sweats and unexplained weight loss). Very occasionally, you might have a blockage in your bowel or a burst bowel ('perforated' or 'ruptured' intestine). This is very serious.

Intestinal T-cell lymphoma can be difficult to diagnose because the symptoms are similar to lots of other conditions that affect the bowel, including coeliac disease itself. The bowel can also be difficult to see on standard **scans**.

You usually need an endoscopy to diagnose intestinal T-cell lymphoma. This is an examination of your bowel using a thin tube that is inserted into your body through your mouth or bottom (anus). Tools can be passed through the tube to take samples of your small bowel (a small bowel **biopsy**) to be examined under a microscope. The **NHS website** has more information about endoscopy.

Intestinal T-cell lymphoma might be present in several different places in the small bowel but it doesn't usually spread to other parts of the body. Although it is usually diagnosed at an **early stage** (stage 1 or 2), many people who have intestinal T-cell lymphoma become very ill.

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## Treatment of intestinal T-cell lymphoma

Intestinal T-cell lymphoma is difficult to treat, partly because most people are very unwell by the time they are diagnosed. It is also rare, which makes it difficult to determine which treatments gives the best outcome. Lymphoma specialists (haematologists or oncologists) and bowel specialists (gastroenterologists) work together to plan the most appropriate treatment for each individual person with intestinal T-cell lymphoma.

Several **targeted drugs** are being tested in intestinal T-cell lymphoma in **clinical trials**. Your specialist 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 intestinal T-cell 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 likely to be treated with **chemotherapy**. You might have surgery to remove the affected parts of your bowel first.

There are no standard treatment protocols for intestinal T-cell lymphoma. The most common **chemotherapy regimens** used for intestinal T-cell lymphoma have been:

- CHOP: cyclophosphamide, doxorubicin (or hydroxydaunorubicin), vincristine (also known as **Oncovin**®) and **prednisolone**
- CHEOP: CHOP plus **etoposide**.

In recent years, a treatment protocol called the Newcastle regimen appears to have improved outcomes in people fit enough to tolerate it. It involves a regimen called IVE/MTX-ASCT: ifosfamide, etoposide (also known as **Vepicid**®) and **epirubicin** with **methotrexate** and **autologous stem cell transplantation**.

However, your medical team might recommend a different chemotherapy regimen.

You are also likely to need support to make sure you get enough nutrients. You might have a feeding tube so that you can be fed liquid food or you might be fed through a tube into a vein (intravenously). If you have coeliac disease, it is important to follow a gluten-free diet to help to avoid further damage to your small bowel.

If you respond to chemotherapy and you are well enough, your medical team are likely to recommend that you have a **self (autologous) stem cell transplant**. This gives you a better chance of staying in remission (no evidence of lymphoma) after treatment.

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## Relapsed or refractory intestinal T-cell lymphoma

It is common for intestinal T-cell lymphoma to come back (**relapse**) after being in remission. Sometimes the lymphoma doesn't respond to treatment (**refractory lymphoma**). In these cases, your doctor might consider:

- a different **chemotherapy regimen**, such as DHAP (**dexamethasone, high-dose cytarabine** [also known as **Ara-C**] and cisplatin [also known as **Platinol®**]), ICE (ifosfamide, carboplatin and etoposide), bendamustine or gemcitabine
  - a **donor (allogeneic) stem cell transplant** if your lymphoma responds to more chemotherapy and you are well enough
  - a targeted drug, usually through a clinical trial.
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## Research and targeted treatments

Many **targeted drugs** are being tested to see if they can help people with T-cell lymphoma. Intestinal T-cell lymphomas are rare so there are few clinical trials looking specifically at this type of lymphoma. Suitable clinical trials are likely to be recruiting people with several types of T-cell lymphoma. Drugs that are being tested in T-cell lymphomas include:

- **antibody–drug conjugates** such as **brentuximab vedotin**
- **cell signal blockers** such as avelumab
- **proteasome inhibitors** such as bortezomib or carfilzomib
- **HDAC inhibitors** such as vorinostat, romidepsin or belinostat.

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 search for a trial that might be suitable for you, visit [Lymphoma TrialsLink](#).

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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).

## 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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✓	Evidence-based
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