Enteropathy-associated T-cell lymphoma (EATL)

This information page is about enteropathy-associated T-cell lymphoma (EATL), a rare type of T-cell lymphoma that is most often linked to coeliac disease.

On this page

What is EATL and who gets it?

Symptoms

Treatment

Relapsed or refractory EATL

Research and targeted treatments

Follow-up

What is EATL and who gets it?

Enteropathy-associated T-cell lymphoma (EATL) is a rare type of non-Hodgkin lymphoma (NHL) that develops from a T lymphocyte (T cell). You can read more about what T-cell lymphoma is on our overview page.

EATL grows in the small bowel (part of your intestines). ‘Enteropathy’ means a ‘disease of the intestines’. There are 2 types of EATL:

- **Type 1, classical EATL**, accounts for around 8–9 in every 10 cases of EATL in the UK.
• **Type 2, monomorphic EATL**, accounts for around 1–2 in every 10 cases of EATL in the UK. It is also called monomorphic epitheliotropic intestinal T-cell lymphoma.

Type 1 develops in people with coeliac disease and is most common in Caucasian (white) people.

Coeliac disease is an autoimmune condition, which is a condition where your body attacks itself. In coeliac disease, your immune system mistakes substances inside gluten (found in grains like wheat, barley and rye) as a threat to your body. Your immune system attacks these substances, causing damage to your small bowel. People with coeliac disease should follow a gluten-free diet to avoid damage to the small bowel.

**NHS Choices** have more information on coeliac disease. **Coeliac UK** also have extensive information about coeliac disease and a gluten-free diet.

Some people are only diagnosed with coeliac disease when they are diagnosed with EATL. **It is important to remember that the vast majority of people with coeliac disease do not go on to develop EATL.**

Very rarely, people have refractory coeliac disease where symptoms don’t get better despite following a gluten-free diet. These people have a higher risk of developing EATL. If this applies to you, your doctor will monitor you closely for signs of EATL.

Most people with type 2 EATL do not have coeliac disease. It is not known what causes type 2 EATL.

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**Symptoms**

The most common symptoms of EATL are bowel and stomach problems. You might have:

- abdominal (tummy) pain
- weight loss
- diarrhoea
ulcers in your bowel and occasionally bowel perforation (a hole through the wall of your bowel).

You might be malnourished (not getting enough nutrients) if you are not absorbing food properly.

You might also have enlarged (swollen) lymph nodes (glands) in other parts of your body.

’B symptoms’ (fevers, night sweats and weight loss) sometimes develop in people with EATL. These symptoms often appear together.

EATL is normally only in the small bowel when it is diagnosed although it can spread to other parts of the body.

EATL can be difficult to diagnose because the symptoms are similar to those of many more common diseases of the bowel. The bowel is also difficult to see on standard scans. For these reasons, most people are very unwell by the time they are diagnosed because of the digestive problems caused by the lymphoma and coeliac disease.

To have biopsies of your small bowel, you are likely to have an endoscopy. A thin tube with a camera is inserted into your body through your mouth or anus (via your bottom). Tools can also be passed through the tube to take biopsies.

NHS Choices have more information about endoscopy.

Treatment

It is important that you follow a gluten-free diet if you are diagnosed with coeliac disease. This helps to avoid further damage to your small bowel.

T-cell lymphomas are rare, so it is difficult to determine which treatment gives the best outcome. You might be asked if you would like to take part in a clinical trial to help test new and current treatments.

EATL is difficult to treat successfully, partly because most people are very unwell by the time they are diagnosed.
The vast majority of people with EATL are treated with chemotherapy. Some people have surgery to remove the affected parts of their bowel before starting chemotherapy.

The most common chemotherapy regimen (combination of drugs) used for EATL is CHOP, which is made up of:

- cyclophosphamide
- doxorubicin (hydroxydaunorubicin)
- vincristine (Oncovin®)
- prednisolone (a steroid).

Your doctor might suggest a more intensive chemotherapy regimen if you are well enough. A targeted drug might be available, usually through a clinical trial.

Chemotherapy is given in cycles. The drugs are given on certain days, which are followed by a break in treatment for your body to recover before the next cycle begins.

Your medical team should monitor you closely and you are 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 intravenously (into a vein).

If you are well enough and your lymphoma has been reduced by chemotherapy, your doctor is likely to recommend you have more chemotherapy together with a stem cell transplant. This is to give you a better chance of staying in remission (no evidence of lymphoma) for a long time. ICE chemotherapy (ifosfamide, carboplatin and etoposide) is often used before a stem cell transplant, but your doctor might suggest a different regimen.

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**Relapsed or refractory EATL**

It is common for EATL to relapse (come back) after being in remission. Sometimes the lymphoma is refractory (doesn’t respond) to treatment. In these cases, your doctor might consider:
• a different chemotherapy regimen, eg GEM-P (gemcitabine, cisplatin and methylprednisolone)

• an allogeneic (donor) stem cell transplant if your lymphoma is reduced by chemotherapy and you are well enough

• a targeted drug, usually through a clinical trial.

Research and targeted treatments

There are several targeted drugs that can be effective in treating EATL, either on their own or together with chemotherapy. These are usually only available if you are taking part in a clinical trial because they are still being investigated.

You may not be suitable for a clinical trial, even if it is recruiting people with the type of lymphoma you have. EATL is rare so it is difficult to recruit enough people to run a clinical trial in EATL alone. Suitable clinical trials are likely to be recruiting people with several types of T-cell lymphoma. Your general health is also an important factor in whether or not you can enter a trial. Clinical trials only recruit people who are well enough to tolerate the study treatment. You can find out more about clinical trials and search for a trial that might be suitable for you at Lymphoma TrialsLink.

Drugs being tested in T-cell lymphoma but not yet approved in the UK include:

• cell signal blockers disrupt signals that keep cancer cells alive or allow them to grow. They act on different pathways in cells
  
  ○ HDAC inhibitors like belinostat (Beleodaq®) or romidepsin (Istodax®) are approved in the US for use in people with relapsed or refractory T-cell lymphom
  
  ○ PI3-K inhibitors like duvelisib
  
  ○ PD1 or PD-L1 inhibitors like nivolumab and avelumab

• immunomodulatory drugs like lenalidomide (Revlimid®) and thalidomide. These affect the activity of the immune system in several different ways.
They can help the immune system to attack the lymphoma cells and prevent the lymphoma from growing.

- **proteasome inhibitors** like bortezomib (Bortezomib Accord or Velcade®) and carfilozomib. These disrupt the balance of proteins in cells, causing them to die.

New drugs are being developed and tested all the time. This is not intended to be a comprehensive list of drugs in development.

## Follow-up

When your treatment is finished, you have regular **follow-up appointments** at the hospital. You are usually seen every 1–3 months at first, then every 6 months after a year or so. Your follow-up usually stops after around 3 years if you remain in remission.

You should contact your medical team or GP if you develop any symptoms of lymphoma or have other concerns between your appointments. Your specialist might bring your appointment forward if they think they need to see you sooner.

## 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

- Overview of T-cell lymphomas
- Chemotherapy
- Stem cell transplants
- Glossary

Acknowledgements

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Tell us what you think and help us to improve our resources for people affected by lymphoma. If you have any feedback, please visit www.lymphoma-action.org.uk/feedback or email publications@lymphoma-action.org.uk.

All our information is available without charge. If you have found it useful and would like to make a donation to support our work you can do so on our website www.lymphoma-action.org.uk/donate. Our information could not be produced without support from people like you. Thank you.

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