

## Hepatosplenic T-cell lymphoma

This page is about hepatosplenic T-cell lymphoma, a rare type of lymphoma that affects your liver and spleen.

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## What is hepatosplenic T-cell lymphoma?

Hepatosplenic T-cell lymphoma is a rare type of **fast-growing (high-grade) non-Hodgkin lymphoma** that grows in the liver and spleen. It is a type of **T-cell lymphoma** – a lymphoma that develops from white blood cells called **T cells**.

'Hepato' means 'relating to the liver' and 'splenic' means 'relating to the **spleen**'.

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## Who gets hepatosplenic T-cell lymphoma?

Hepatosplenic T-cell lymphoma is a very rare condition. It usually affects younger adults, typically people in their mid-30s. It is more common in men than women.

Hepatosplenic T-cell lymphoma is linked with low function of the **immune system**. It is more likely to develop in people who are taking medicines that suppress the immune system – for example, after a transplant or for inflammatory bowel diseases such as Crohn's disease. However, it can develop spontaneously in people who don't have pre-existing conditions and aren't taking medication.

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## Symptoms of hepatosplenic T-cell lymphoma

The most common **symptoms** of hepatosplenic T-cell lymphoma are:

- a **swollen liver**, which might make you feel bloated, cause fluid to build-up in your tummy, or make your skin or the whites of your eyes look yellow (jaundice)
- a **swollen spleen**, which might cause pain behind your ribs on the left side, or make you feel full very quickly when you're eating
- **anaemia** (low red blood cell count), which can make you tired and short of breath
- **thrombocytopenia** (low platelet level), which can make you bruise or bleed more easily than normal
- **neutropenia** (low neutrophil level), which makes you more prone to infections than usual
- fever, night sweats and weight loss ('**B symptoms**').

Unlike most other lymphomas, hepatosplenic T-cell lymphoma doesn't usually cause swollen lymph nodes. It can be difficult to diagnose because the lymphoma cells are scattered around your body rather than in lumps.

Most people with hepatosplenic T-cell lymphoma have **advanced stage** lymphoma (stage 3 or 4) when they are diagnosed.

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## Treatment of hepatosplenic T-cell lymphoma

Hepatosplenic T-cell lymphoma is difficult to treat and it is common for the lymphoma to come back (relapse). Because it is rare, no treatments have become established as internationally recognised standards of care for hepatosplenic T-cell lymphoma. **Clinical trials** in this rare condition are unusual. However, some trials of experimental agents do allow people with hepatosplenic T-cell lymphoma to take part, particularly if their lymphoma has relapsed. Several **targeted drugs** are being tested. 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 hepatosplenic 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**.

The most common **chemotherapy regimens** used for hepatosplenic T-cell lymphoma are:

- CHOP: cyclophosphamide, doxorubicin (or **hydroxydaunorubicin**), vincristine (also known as **Oncovin®**) and **prednisolone**
- CHEOP: CHOP plus **etoposide**
- ICE: **ifosfamide**, **carboplatin** and **etoposide**
- IVAC: **ifosfamide**, **etoposide** (also known as **VP-16**) and **cytarabine** (also known as **Ara-C**).

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

If you respond to chemotherapy and you are well enough, your doctor might recommend that you have a **stem cell transplant**. This could be a **donor (allogeneic) stem cell transplant** or a **self (autologous) stem cell transplant**. A stem cell transplant might give you a better chance of staying in **remission** (no evidence of lymphoma).

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## Relapsed or refractory hepatosplenic T-cell lymphoma

It is common for hepatosplenic T-cell lymphoma to come back (**relapse**) after being in remission. Sometimes, it 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®**])
  - a **donor (allogeneic) stem cell transplant** if you have not already had one, 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. Drugs that are being tested in hepatosplenic T-cell lymphoma include:

- **antibody treatments** such as **alemtuzumab**, which binds to a protein called CD52 on T cells
- **proteasome inhibitors** such as **carfilzomib**
- **HDAC inhibitors** such as **romidepsin**.

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 find 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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