Freephone helpline 0808 808 5555
information@lymphoma-action.org.uk
www.lymphoma-action.org.uk

Hepatosplenic T-cell lymphoma

This information page is about hepatosplenic T-cell lymphoma.

On this page

What is hepatosplenic T-cell lymphoma and who gets it?

Hepatosplenic T-cell lymphoma is a very rare type of **non-Hodgkin lymphoma** (NHL) that develops from T lymphocytes (T cells). Our **overview page** explains what T-cell lymphoma is.

Hepatosplenic T-cell lymphoma causes swelling of the liver and spleen (an organ of the **immune system**):

- 'hepato' refers to the liver
- 'splenic' refers to the spleen.

Hepatosplenic T-cell lymphoma usually occurs in younger adults, with an average age in the mid-30s at the time of diagnosis. It can occur in children.

It affects more men than women.

Hepatosplenic T-cell lymphoma is sometimes linked with **immunosuppression** (lowered immunity), for example, it can occur after organ transplant or after treatment for inflammatory bowel conditions like Crohn's disease. However, it is very rare for people with these conditions to develop hepatosplenic T-cell lymphoma.

Symptoms

Hepatosplenic T-cell lymphoma causes a range of **symptoms**. Most people have:

- enlarged (swollen) spleen and liver
- **anaemia** (low red blood counts), which can make you tired and short of breath
- **thrombocytopenia** (low platelets), which can make you more likely to bruise and bleed; low platelets are particularly noticeable in this type of lymphoma
- **neutropenia** (low neutrophils), which can increase your risk of infection
- '**B symptoms**' (fevers, night sweats and weight loss), which often appear together.

You might also have enlarged lymph nodes (glands), but enlarged liver and spleen are more common.

Abnormal T cells are usually found in the bone marrow (the spongy tissue in the centre of bones where blood cells are made) and sometimes in the blood.

The vast majority of people have **advanced-stage disease** when they are diagnosed.

Treatment

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.

Hepatosplenic T-cell lymphoma is usually difficult to treat. It is common for the lymphoma to relapse (come back).

You are most likely to be treated with a **chemotherapy regimen** (combination of drugs), for example 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 chemotherapy drug called pentostatin is sometimes used.

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.

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**. If a suitable donor is available, this might be an **allogeneic (donor) stem cell transplant**. An allogeneic stem cell transplant is a very intensive form of treatment but can sometimes lead to a long-term remission (no evidence of lymphoma).

Relapsed or refractory hepatosplenic T-cell lymphoma

It is common for hepatosplenic T-cell lymphoma to **relapse** 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**
- 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.

Hepatosplenic T-cell lymphoma is very rare, so 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. Your doctor can advise you whether there is a trial suitable for you. You can find out more about clinical trials and search for a trial that might be suitable for you at **Lymphoma TrialsLink**.

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. Your appointments then become less frequent if you are well over time. 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.

- British Committee for Standards in Haematology (BCSH). Guidelines for the management of mature T-cell and NK-cell neoplasms (excluding cutaneous T-cell lymphoma). August 2013. Available at: **www.bit.do/bsh-guidelines-tnhl** (Accessed August 2016).

- Kircher SM et al. Chapter 14: T-cell non-Hodgkin lymphoma. In Marcus R et al. Lymphoma: pathology, diagnosis and treatment. 2nd edition. Cambridge University Press. Published 2014.
- Armitage JO. The aggressive peripheral T-cell lymphomas: 2015. *American Journal of Hematology*. 2015; 90: 665-73. Available at: **www.bit.do/aggressive-tcell** (Accessed August 2016).
- Sims MC, et al. Long-term outcomes of autologous stem cell transplantation for peripheral T-cell lymphomas across the Thames Valley (1997–2012). *British Journal of Haematology*, 2015. 169: 452–455. Available at: **www.bit.do/sct-tcell** (Accessed August 2016).
- Swerdlow SH, et al. The 2016 revision of the World Health Organization classification of lymphoid neoplasms. *Blood*, 2016. 127: 2375–2390. Available at: **www.bit.do/tcell-classification** (Accessed August 2016).

Further reading

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

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