

# Peripheral T-cell lymphoma not otherwise specified (PTCL-NOS)

When a T-cell lymphoma doesn't fit into any particular category of T-cell lymphoma, it is called peripheral T-cell lymphoma not otherwise specified (PTCL-NOS). This page is about PTCL-NOS.

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## What is PTCL-NOS?

PTCL-NOS is a type of **T-cell lymphoma** – a **non-Hodgkin lymphoma** that develops from white blood cells called **T cells**. PTCL-NOS includes all T-cell lymphomas that don't fit into any other category. The lymphomas within this group can be quite different from each other. However, they share some similarities, such as the symptoms they cause. When more evidence becomes available in the future, PTCL-NOS is likely to be split into several subtypes of T-cell lymphoma. At the moment, it is usually discussed as a single type of lymphoma.

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## Who gets PTCL-NOS?

PTCL-NOS is the most common T-cell lymphoma, accounting for around 1 in 3 cases. It can occur at any age, but it is more common in people in their 60s. It tends to affect more men than women. Most people have **advanced stage** lymphoma when they are diagnosed.

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## Symptoms of PTCL-NOS

People with PTCL-NOS may have **swollen lymph nodes**. These can develop anywhere in the body but the most common places are the neck, armpit or groin. PTCL-NOS is more commonly found outside of the lymph nodes (extranodal areas).

- The **bone marrow** (the spongy tissue in the centre of bones where blood cells are made) is often affected, which can lead to **anaemia** (low red blood cells) and **thrombocytopenia** (low platelet levels).
- The liver and spleen might be enlarged, causing **bloating or tummy (abdominal) pain**. Lymphoma elsewhere in your gut might cause pain, sickness, diarrhoea or vomiting.
- The skin can be affected by itchy red patches.
- If you have lymphoma in your chest, you might feel short of breath, develop a cough or have pain or a feeling of pressure in your chest.

Fevers, night sweats and unexplained weight loss (known as '**B symptoms**') are also common in people with PTCL-NOS.

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## Treatment of PTCL-NOS

Although it is the most common T-cell lymphoma, PTCL-NOS is still rare. This makes it difficult to determine which treatment will give you the best outcome.

There are no standard treatments for PTCL-NOS. Several **targeted drugs** are being tested for PTCL-NOS and different **chemotherapy regimens** (combinations of drugs) are being compared in **clinical trials**. Your doctor might ask you if you would like to take part in a clinical trial to test new treatments and help find out what the best treatment is for PTCL-NOS.

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 regimen used for PTCL-NOS is CHOP:

- **cyclophosphamide**
- **doxorubicin** (or **hydroxydaunorubicin**)
- **vincristine** (also known as **Oncovin®**)
- **prednisolone** (a steroid).

You might have a chemotherapy drug called **etoposide** in addition to CHOP (CHEOP), or you might have a different chemotherapy regimen altogether. If you are not fit enough to have CHOP or CHEOP, you might be treated with less intensive chemotherapy, such as gemcitabine or bendamustine.

If you respond to chemotherapy and you are well enough, your doctor is 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 PTCL-NOS

It is common for PTCL-NOS to come back (**relapse**) after being in remission. Sometimes, PTCL-NOS 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®**]) or ICE (**ifosfamide, carboplatin and etoposide**)
  - 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. Drugs that have been or are being tested in PTCL-NOS include:

- **antibody treatments** such as alemtuzumab, which binds to a protein called CD52 on T cells
- **antibody–drug conjugates** such as **brentuximab vedotin**
- **immune checkpoint inhibitors** such as avelumab and pembrolizumab
- **proteasome inhibitors** such as bortezomib or carfilzomib
- **HDAC inhibitors** such as vorinostat, romidepsin or belinostat
- a chemotherapy drug called **pralatrexate**.

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.

## Acknowledgements

- With thanks to Prof Tim Illidge, Professor of Targeted Therapy and Oncology and Honorary Consultant Oncologist, University of Manchester, the Christie NHS Foundation Trust, for reviewing this information.
- We would like to thank the members of our Reader Panel who gave their time to review this information.

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Content last reviewed: August 2019

Next planned review: August 2022

LYMweb0208PTCL-NOS2019v2

✓	Evidence-based
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