

## Extranodal NK/T-cell lymphoma, nasal type

This information is about extranodal NK/T-cell lymphoma, nasal type – a rare type of T-cell lymphoma associated with the Epstein-Barr virus (EBV) infection. It usually develops in the nose.

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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).

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## What is extranodal NK/T-cell lymphoma?

Extranodal NK/T-cell lymphoma, nasal type, is a rare **fast-growing (high-grade) non-Hodgkin lymphoma**. It grows outside the lymphatic system ('extranodal'), usually in the nose ('nasal'). The cells of origin are thought to be two different kinds of **lymphocyte** (white blood cell):

- **natural killer (NK) cells**
  - **cytotoxic T cells.**
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## Who gets extranodal NK/T-cell lymphoma?

Extranodal NK/T-cell lymphoma, nasal type, is more common in people from Asia, Central America and South America. It is very rare in the western world and only around 20 people are diagnosed with it in the UK each year. It usually develops in people around 50 to 60 years old. It is more common in men than women.

Extranodal NK/T-cell lymphoma, nasal type, is strongly linked to infection with a virus called Epstein-Barr virus (EBV). EBV is a very common virus, especially in young people, that can cause glandular fever. After you've been infected with it, EBV stays in your body but your **immune system** normally keeps it under control. Most people who have EBV infection do **not** develop lymphoma. However, sometimes it can cause genetic changes in your lymphocytes that might encourage lymphoma to develop.

Scientists are still working hard to understand why some people who have been infected with EBV go on to develop extranodal NK/T-cell lymphoma.

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## Symptoms of extranodal NK/T-cell lymphoma

People who have extranodal NK/T-cell lymphoma, nasal type, usually develop a fast-growing lump inside their nose or in the sinuses (air-filled spaces) around the nose. You might have **symptoms** that affect your nose, eyes or face, such as:

- a blocked nose
- nosebleeds, or nasal crusting
- swelling or pain in your face
- weepy eyes (blocked tear ducts) or eye discomfort.

The lymphoma might get bigger and grow into the roof of your mouth, or into your throat and eye sockets.

It is also common to have general symptoms, such as:

- **swollen lymph nodes**
- fever, night sweats and weight loss ('**B symptoms**')
- **fatigue**.

If the lymphoma affects your skin, you might have a rash or raised, red lumps that can break down (ulcerate) and scab over.

For the other types of NKT cell lymphoma that develop outside of the nose, it can spread to your lymph nodes, skin, testicles or gut.

Extranodal NK/T-cell lymphoma, nasal type, is usually diagnosed when it is at an **early stage** (stage 1E or 2E). The 'E' stands for 'extranodal'. If it develops outside the nose, it is often more advanced when it is diagnosed (stage 3 or 4).

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## Treatment of extranodal NK/T-cell lymphoma

Extranodal NK/T-cell lymphoma, nasal type, is rare. This makes it more difficult to determine the most suitable treatment approach for the best outcome as many centres report their results using different treatment approaches. Clinical trials in this type of lymphoma are unusual because it is such a rare condition, particularly in the western world.

For localised NK/T-cell lymphoma of the nose, **radiotherapy** is the most common and most important treatment for early staged extranodal NK/T-cell lymphoma, nasal type. You have it as part of your initial treatment, usually with **chemotherapy**. You might have your course of chemotherapy at the same time as you are having radiotherapy or after your radiotherapy.

If your lymphoma is more advanced (stage 3 or 4), you are likely to have chemotherapy without radiotherapy.

Chemotherapy used to treat extranodal NK/T-cell lymphoma, nasal type, includes **regimens** (combination of drugs) such as:

- DDGP: dexamethasone, cisplatin, gemcitabine and pegaspargase
- SMILE: dexamethasone (a **steroid**), **methotrexate**, **ifosfamide**, **L-asparaginase** and **etoposide**.

However, your medical team might recommend a different chemotherapy regimen. If you are not fit enough to have standard chemotherapy, you might be treated with radiotherapy on its own, lower doses of chemotherapy, or **L-asparaginase** on its own.

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

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## Relapsed or refractory extranodal NK/T-cell lymphoma

Extranodal NK/T-cell lymphoma, nasal type, is a very aggressive form of lymphoma. It is common for it to come back (**relapse**) after treatment. Sometimes, extranodal NK/T-cell lymphoma, nasal type, doesn't respond to treatment (refractory lymphoma). In these cases, your doctor might consider:

- a **targeted drug** usually through a clinical trial
  - a different chemotherapy regimen – examples include GELOX (**gemcitabine**, **L-asparaginase** and **oxaliplatin**) but your doctor might suggest another regimen
  - a **donor (allogeneic) stem cell transplant** if your lymphoma responds to more chemotherapy and you are well enough.
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## Research and targeted treatments

Many new treatments are being tested to see if they can help people with T-cell lymphoma. Drugs that are being tested in extranodal NK/T-cell lymphoma, nasal type, include:

- **antibody treatments** such as alemtuzumab and sipilizumab
- **antibody–drug conjugates** such as **brentuximab vedotin**
- **cell signal blockers** such as alisertib
- **immune checkpoint inhibitors** such as PD1 blockade
- **proteasome inhibitors** such as bortezomib
- **HDAC inhibitors** such as 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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## 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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