

# Central nervous system (CNS) lymphoma

CNS lymphoma is lymphoma which affects the central nervous system (the brain, spinal cord and eyes). This information explains the different types of CNS lymphoma, the symptoms, diagnosis and how it can be treated.

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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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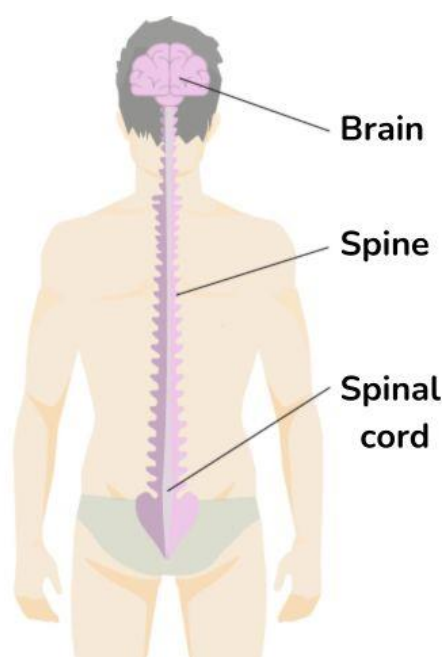
## What is central nervous system lymphoma?

Central nervous system lymphoma is lymphoma that is in your brain, spinal cord or eyes (your central nervous system, or CNS). CNS lymphomas are rare forms of **non-Hodgkin lymphoma**.

### What is the CNS?

The CNS is the part of your body that controls all your body's functions. It includes:

- your **brain**, which is your body's control centre
- your **spinal cord**, which runs down your back inside your spinal bones and carries signals between your brain and your nerves
- your **meninges**: protective layers of tissue that cover your brain and spinal cord (this is what becomes inflamed if you have 'meningitis')
- liquid called **cerebrospinal fluid (CSF)**, which surrounds your brain and spinal cord
- your **eyes**.



The brain and spinal cord

Your CNS is protected from harmful chemicals and infections by a layer of cells and blood vessels called the 'blood–brain barrier'. This separates your brain from your bloodstream and only lets certain substances through. Your blood–brain barrier is important to protect your brain. However, it can make it difficult for drugs to get into your CNS.

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## Types of CNS lymphoma

Lymphoma in the CNS is usually a fast-growing (high-grade) **non-Hodgkin lymphoma**. It is most often a form of **diffuse large B-cell lymphoma**. Rarely, **Burkitt lymphoma**, **T-cell lymphoma** or **low-grade non-Hodgkin lymphomas** can affect your CNS.

CNS lymphoma can be either 'primary' or 'secondary'.

- **Primary CNS lymphoma** is lymphoma that starts in your central nervous system and isn't growing anywhere else when you are diagnosed. It usually develops in your brain but it can also affect your eyes (this is called intraocular lymphoma).
  - **Secondary CNS lymphoma** is lymphoma that starts somewhere else in your body and spreads to your CNS. This includes lymphoma that comes back (**relapses**) in your CNS following treatment. Secondary CNS lymphoma usually develops in the brain but it can sometimes grow in the protective tissues that cover your brain and spinal cord (the meninges).
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## Who gets CNS lymphoma?

**Primary CNS lymphoma** is rare. Only around 160 people are diagnosed with it each year in the UK. It tends to affect people over 60. It is slightly more common in men than women.

Primary CNS lymphoma is slightly more likely to affect people who have a lowered immune system, such as:

- people who are taking medicines to dampen their immune system (immunosuppressants)
- people who have human immunodeficiency virus (**HIV**)
- people who have had an **organ transplant**.

However, most people with these conditions do **not** develop CNS lymphoma.

**Secondary CNS lymphoma** is lymphoma that has spread to the CNS from elsewhere in the body. This can occasionally occur with fast-growing types of lymphoma, such as **Burkitt lymphoma**, **lymphoblastic lymphoma** or **diffuse large B-cell lymphoma** (DLBCL). However, even if you have one of these types of lymphoma, your risk of developing secondary CNS lymphoma is low.

Scientists aren't sure why lymphoma sometimes comes back (**relapses**) in the CNS. They think it might be because most of the **chemotherapy** drugs used to treat lymphoma elsewhere in the body do not reach the CNS. This means that when lymphoma is treated successfully in other places in your body, some lymphoma cells might survive in your CNS. This could cause the lymphoma to come back in your CNS.

People who have a higher risk of developing secondary CNS lymphoma are sometimes offered treatment to try to reduce the risk. This is called **CNS prophylaxis**.

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## Symptoms of CNS lymphoma

The **symptoms** of CNS lymphoma depend on where exactly the lymphoma is. They can include:

- confusion, problems with your memory or difficulty concentrating
- increased irritability or other changes in your personality
- problems with your speech (for example, difficulty finding the right word)
- muscle weakness or loss of feeling in part of your body (for example, in one arm or leg)
- fits (seizures)
- headaches or drowsiness
- feeling or being sick
- problems with your balance.

If you have CNS lymphoma involving your eye, you might get blurred vision or notice small dots or shapes that seem to float quickly across your vision ('floaters'). Floaters are very common and are not usually a sign of lymphoma.

If the lymphoma is pressing on your spinal cord, you might experience:

- back pain, which might travel down your legs
- numbness or tingling in your legs
- weakness and, in some cases, paralysis, most often affecting the legs but sometimes also the arms or tummy.

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## Diagnosis of CNS lymphoma

CNS lymphoma is diagnosed with a procedure called a **biopsy**. A sample of tissue is removed and examined under a microscope by an expert **lymphoma pathologist**. The pathologist does specialist tests on the tissue to find out what type of lymphoma it is.

If the lymphoma is only in your brain, a specialist brain surgeon (a neurosurgeon) will perform the biopsy, usually under a general anaesthetic. They use a CT scan to make sure they take the biopsy sample from the best place. If you have lymphoma in other parts of your body as well as your CNS, you are likely to have the tissue sample taken from one of the other places where the lymphoma is growing (for example, a lymph node).

You have other tests to find out which parts of your body are affected by lymphoma and to check your general health. These tests might include:

- an **MRI scan**, which is good at showing lymphoma in the brain and other parts of the CNS
- a **PET/CT scan** to check for lymphoma in other parts of your body
- a **lumbar puncture** to check for lymphoma cells in the fluid around your brain and spinal cord
- a specialist eye examination to check for lymphoma in your eyes, possibly followed by a minor procedure to take a sample of the jelly inside your eyes (a '**vitreous biopsy**')
- for men, an **ultrasound scan** of your testicles
- a **bone marrow biopsy** to check for lymphoma cells in your bone marrow (although this is not usually needed if you've had a PET scan)
- **blood tests** to check your blood cell counts and test how well your liver and kidneys are working.

Not everyone needs all of these tests. Your **medical team** decide what tests you need based on your individual circumstances.

Your medical team should be able to give you an idea of when to expect your test results. **Waiting for the results** of your tests can be difficult. However, it's important for your medical team to know exactly what type of lymphoma you have and where it is. This helps them plan the most appropriate treatment for you.

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## **Outlook**

The outlook for CNS lymphoma has improved a lot over recent years. However, CNS lymphomas can still be difficult to treat. Some treatments have a risk of causing long-term side effects. Your medical team balance the chance of successful treatment against the risk of long-term problems when deciding on the most appropriate treatment for you.

Your medical team are best placed to advise you on your outlook based on your individual circumstances. They can use the results of your tests and other factors (for example, your age and physical fitness) to help them judge how likely you are to respond to a particular treatment.

If you choose to research survival statistics, it is important to remember that they don't tell you what your individual outlook is – they only tell you how a group of people with the same diagnosis did over a period of time. They are usually measured 5 to 10 years after treatment, so they only tell you how people did in the past. Those people might not have received the same treatment as you. Many people do not find survival statistics helpful because of this variability.

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## **Treatment for CNS lymphoma**

Your medical team recommend treatments based on several factors, including:

- what type of CNS lymphoma you have
- where the lymphoma is growing
- how it is affecting you
- your age and your general health.

## Treatment for primary CNS lymphoma

Treatment for primary CNS lymphoma is usually given in two phases:

- ‘Remission induction therapy’ aims to target all the lymphoma that can be seen on scans to achieve a remission.
- ‘Consolidation therapy’ aims to destroy any lymphoma cells left behind, to stop the lymphoma coming back and to help keep the lymphoma under control for as long as possible.

You might have treatment with **steroids** first, to help shrink the lymphoma and reduce any swelling around it. This helps relieve your symptoms while you are having tests and before you start your main treatment. However, doctors try not to give you steroids until after you have had a biopsy because they can affect the result.

### Remission induction therapy

Your consultant might ask you if you’d like to take part in a **clinical trial** to give you access to a newer drug, if there is one suitable for you.

If there is not a clinical trial that is suitable for you, or if you don’t want to take part in a clinical trial, your medical team are likely to suggest a course of **chemotherapy** that includes drugs that can cross the blood–brain barrier. You usually have this combined with an **antibody therapy** such as **rituximab**. The exact combination of drugs (**chemotherapy regimen**) your team recommends depends on the results of your tests and your individual circumstances. The most common regimen for primary CNS lymphoma is **MATRix**:

- high-dose **methotrexate**
- cytarabine (also known as **Ara-C**)
- **thiotepa**
- **rituximab**.

You have all these drugs through a drip into a vein. It is an intensive treatment and you have to be fit enough to have it. You stay in hospital for some of your treatment so your medical team can monitor you. You often require other treatments to help support your body during your chemotherapy, such as **growth factors** to boost your blood counts, antibiotics to treat infections, and sometimes blood transfusions.

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I found chemo very hard. At the end of each cycle I would become neutropenic, pick up an infection and become extremely ill. The doctors would hit me with a strong dose of antibiotics and I would recover enough to go home and put some weight on, ready to face the next cycle.

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Richard, diagnosed with primary CNS lymphoma

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If you are not well enough to have strong chemotherapy, or you have other health conditions that mean MATRix is not suitable for you, your medical team might recommend:

- a gentler chemotherapy or lower doses of treatment
- **radiotherapy** to your brain (and to your eyes, if they are affected by the lymphoma)
- **steroids**.

### **Consolidation therapy**

If the lymphoma responds to high-dose chemotherapy and you are fit enough, your medical team is likely to recommend high-dose chemotherapy with an **autologous stem cell transplant**. This aims to keep your lymphoma under control for as long as possible. Stem cell transplants are a very intense form of treatment and you have to be fit enough to have one.

If a stem cell transplant isn't suitable for you, you might be offered:

- **radiotherapy** to your brain
- radiotherapy to your eyes, if they are affected by the lymphoma.

Radiotherapy to your brain can cause long-term problems with your memory, concentration and thinking processes (**cognitive impairment**). Your medical team balance the risk of long-term side effects with the benefits the treatment might have for you when deciding on the most appropriate treatment for you.

### **Treatment for secondary CNS lymphoma**

Secondary CNS lymphoma is rare so there is no standard treatment. Your medical team will recommend a treatment plan based on your individual circumstances.



As with primary CNS, you might have treatment with **steroids** first, to help shrink the lymphoma and reduce any swelling around it.

Your consultant might ask you if you'd like to take part in a **clinical trial**, if there is one suitable for you. Alternatively, they might suggest a course of high-dose **chemotherapy** that includes drugs that are able to cross the blood–brain barrier. A common regimen for secondary CNS lymphoma is known as MARIETTA:

- MATRix (high-dose **methotrexate**, cytarabine [also known as **Ara-C**], **thiotepa** and **rituximab**)
- followed by, or alternating with, cycles of R-ICE (**rituximab**, ifosfamide, carboplatin and **etoposide**)
- with **intrathecal chemotherapy**.

You may be offered a dose-reduced version of MARIETTA or an alternative regimen based on your specific circumstances.

If high-dose chemotherapy controls the lymphoma, you might be able to have a **stem cell transplant** to reduce the risk of the lymphoma coming back.

**Radiotherapy** may be considered if lymphoma remains in the CNS following other treatment or if stem cell transplant is not a possible consolidation treatment option.

If you are not fit enough for intensive treatment options, or if your lymphoma doesn't respond to it, you might be offered a different chemotherapy regimen, or radiotherapy to help ease your symptoms.

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## Follow-up

When you finish treatment for CNS lymphoma, you usually have an MRI scan to check how well your lymphoma has responded. Your doctor might recommend follow-up MRI scans every 3 to 4 months for 2 years, to make sure the lymphoma has not come back (relapsed).

You will also have regular **follow-up appointments** to check:

- how well you are recovering from the lymphoma and the treatment
- how you are coping with any long-term side effects or **late effects** (side effects that develop months or years after treatment) of your treatment
- for any signs that the lymphoma might be coming back.

Sometimes, the symptoms of CNS lymphoma get better quite quickly after treatment. However, nerve tissues grow very slowly and it can sometimes take a long time for symptoms to improve. Some people find their symptoms never go away completely, although they often get better over time.

Your medical team can refer you to specialists who can help support your physical, practical and emotional recovery. This might include physiotherapists, occupational therapists and psychologists.

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**After 4 rounds of MATRix chemo, a stem cell transplant and 20 sessions of radiotherapy I came home. I had a private carer to stay with me for the first 3 weeks. I would have really struggled without her help. Since then my recovery has been slower than I thought it would be, but talking to others on Lymphoma Action, I now accept that this is not uncommon. My GP arranged a set of 6 physio classes which helped. My balance is still not great but it's improving. I use a mobility scooter to do my food shopping and get to the GP surgery. I'm gradually able to walk further and now take short walks without my walking frame or walking stick.**

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Penny, diagnosed with CNS lymphoma

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**If you are ever concerned about your lymphoma, contact your hospital team. Don't wait for an appointment if you are worried.**

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## **Relapsed or refractory CNS lymphoma**

If CNS lymphoma comes back (relapses) or doesn't respond (refractory) after treatment, it can be difficult to treat. Your consultant might ask you if you'd like to take part in a **clinical trial** to give you access to a newer treatment, if there is one suitable for you. If there isn't, or if you don't want to take part in a trial, other treatments might be available. These depend on how fit you are at the time, what treatment you had before and how the lymphoma is affecting you. Your specialist can talk through your options. Possible treatments include:

- high-dose chemotherapy, possibly followed by a stem cell transplant if you haven't already had one

- radiotherapy to your brain
  - gentler chemotherapy treatments
  - palliative treatments given with the aim of easing your symptoms
  - **CAR-T cell therapy** for secondary CNS lymphoma in certain circumstances.
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## Research

Ongoing research is exploring new treatment options for CNS lymphoma and how to optimise the use of current therapies. Current research is focusing on **targeted treatments**, including:

- **CAR-T cell therapy**, which involves having your own T cells collected and genetically modified (changed) in a laboratory to help them recognise and kill lymphoma cells
- Immunomodulatory drugs, such as **lenalidomide**, that affect the activity of your immune system
- **BTK inhibitors** (and related drugs), which block signals that B cells send to help them divide or stay alive
- **Checkpoint inhibitors**, which block specific proteins, allowing the immune system to recognise and attack lymphoma cells.

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 to search for a trial that might be suitable for you, visit **Lymphoma TrialsLink**.

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## Further support

Illnesses that affect the brain and nerves can be difficult to live with. It is very important that you find the right help and support if you have CNS lymphoma, or if you are caring for someone with this type of lymphoma.

We offer a range of support services to help you live your life with and after CNS lymphoma:

- Our Freephone helpline offers information and support about any aspect of lymphoma. Call us on **0808 808 5555**, email **information@lymphoma-action.org.uk** or LiveChat via our website.

- Our [Live your Life workshops](#) provide practical guidance and support to help you live with and beyond lymphoma.
- Find out about our [online services and support](#), including webinars, videos and online Support Groups.
- Our [Buddy Service](#) can connect you with someone who has a personal experience of lymphoma and understands what you're going through.
- Read the stories of others affected by CNS lymphoma, including [Claire](#), [Imtiaz](#) and [Richard](#).

We also have information for people [caring for someone with lymphoma](#).

Our [list of organisations](#) that offer support and information for people with lymphoma might also be useful. The following organisations offer support and information that is particularly relevant for people with CNS lymphoma:

- [The Brain Tumour Charity](#)
- [Brainstrust](#)
- [Brain and Spine Foundation](#)
- [Headway](#).

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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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