Primary CNS lymphoma

A primary CNS lymphoma is a rare type of lymphoma (cancer of the lymphatic system) that starts in the central nervous system (CNS).

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This information describes primary CNS lymphomas, their symptoms, tests, and possible treatments. It should ideally be read with our general information about brain tumours and non-Hodgkin lymphoma which go into more detail about tests, treatments and their side effects. We also have information on spinal cord tumours.

We hope this information answers your questions. If you have any further questions, you can ask your doctor or nurse at the hospital where you are having treatment.

Primary CNS lymphoma

The central nervous system (CNS) is made up of the brain and spinal cord. The brain controls different functions of the body, including how we think, feel, learn and move. The spinal cord is made up of nerves that run down the middle of the back (spine). Messages between the brain and other parts of the body travel through the spinal cord.

A lymphoma is a cancer of the lymphatic system, which is part of the body's immune system and helps fight infection. Cancer cells can form in the lymphatic tissue in the brain. Lymphomas that start in the CNS are called primary CNS lymphomas and are usually fast growing. They are rare and are most likely to affect older people.

The most common site for a primary CNS lymphoma is the cerebrum (see the diagram below). Sometimes there's more than one tumour. Less commonly, they may start in other parts of the CNS, including the lining of the brain (meninges), the eye (primary intraocular lymphoma) or the spinal cord.

Some CNS lymphomas have spread from lymphoma in another part of the body. This is called a secondary lymphoma. It's important to know if lymphoma in the CNS is a primary or a secondary lymphoma because they are treated differently.

This information is about lymphoma that starts in the CNS.
A side view of the brain

**Causes of primary CNS lymphoma**

Usually the cause is not known. Most people who develop this type of cancer have a normal immune system. However, primary CNS lymphoma may affect people whose immune system is not working properly – for example, in people with HIV (human immunodeficiency virus) or after organ transplants.

**Symptoms**

Some possible symptoms are:

**Raised intracranial pressure**

The symptoms may be due to the tumour causing an increase in pressure within the skull (raised intracranial pressure). The pressure can increase because of a blockage in the fluid-filled space in the brain (the ventricles). This leads to a build up of the fluid that surrounds and protects the brain and spinal cord, called cerebral spinal fluid (CSF). The increased pressure may also be caused by the tumour itself.

Raised intracranial pressure can cause headaches, sickness (vomiting) and problems with vision.

**Changes in behaviour and personality**

It’s not unusual for there to be changes in behaviour and personality and you may be confused at times.

**Seizures (fits)**

Some people may have seizures with this type of tumour. Our booklet on brain tumours has more information on this.

**Changes in balance/movement**

Sometimes balance and coordination are affected. This may make walking more difficult.

**Numbness or weakness**

Some people may have numbness or weakness on one side of the body.

**Problems with vision**

If the lymphoma starts in the the eye, or is close to it, you may have changes in vision.
Rarely, if the lymphoma starts in the spinal cord, this can cause tingling and weakness in the arms or legs.

**Tests**

Your doctors need to find out as much as possible about the position and size of the lymphoma so they can plan the best treatment for you. You will have tests, which your doctor or nurse will explain to you.

The doctor will physically examine you and do checks on your nervous system. This includes checking your reflexes and the power and feeling in your arms and legs. They also shine a light at the back of your eye to check if the optic nerve is swollen, which can be a sign of raised pressure in the brain. Your doctor will ask some questions to check your reasoning and memory.

You will have blood tests taken to check the number of cells in your blood (full blood count) and to see how well your kidneys and liver are working. Your doctor or nurse may talk to you about having a test for HIV.

You will have a CT or MRI scan to find out the exact position and size of the tumour.

**CT (computerised tomography) scan**

A CT scan takes a series of x-rays that build up a three-dimensional picture of the inside of the body. You will have a full body scan. The scan is painless and takes 10–30 minutes. CT scans use small amounts of radiation, which will be very unlikely to harm you or anyone you come into contact with.

You may be given an injection of a dye, which allows particular areas to be seen more clearly. This may make you feel hot all over for a few minutes. If you’re allergic to iodine or have asthma you could have a more serious reaction to the injection, so it’s important to let your doctor know beforehand.

**MRI (magnetic resonance imaging) scan**

This test is similar to a CT scan but uses magnetism instead of x-rays to build up a detailed picture of areas of your body. Before the scan you may be asked to complete and sign a checklist. This is to make sure it’s safe for you to have an MRI scan.

Before having the scan, you'll be asked to remove any metal belongings, including jewellery. Some people are given an injection of dye into a vein in the arm. This is called a contrast medium and can help the images from the scan to show up more clearly. During the test you will be asked to lie very still on a couch inside a long cylinder (tube) for about 30 minutes. It’s painless but can be slightly uncomfortable, and some people feel a bit claustrophobic during the scan. It’s also noisy, but you’ll be given earplugs or headphones.

**Lumbar puncture**

A lumbar puncture is carried out to see if there are any tumour cells present in the CSF.

Your doctor uses a local anaesthetic to numb the lower part of your back and then passes a needle gently into the spine. They then take a small sample of CSF to be checked for lymphoma cells.

A lumbar puncture is usually done as an outpatient and it only takes a few minutes.

**Bone marrow sample**

You may have this done to make certain the lymphoma has not started outside the brain. A small sample of bone marrow is taken from the back of the hip bone (pelvis) or occasionally the breast bone (sternum). It’s
sent to a laboratory to check for abnormal white blood cells. You’ll be given a local anaesthetic injection to numb the area. You may also be offered a short-acting sedative to reduce any pain or discomfort during the test.

The doctor or nurse passes a needle through the skin into the bone. They then draw a sample of liquid and a small core of bone marrow from inside the bone into a syringe. It can feel uncomfortable for a few seconds when the liquid and marrow are being drawn into the syringe. The test takes about 15-20 minutes but removing the bone marrow sample only takes a few minutes.

**Biopsy**

To diagnose primary CNS lymphoma you will usually need to have a small sample of tissue removed from the tumour (biopsy). You need an operation under general anaesthetic to have this done. A neurosurgeon (brain surgeon) makes a small hole in the skull and passes a fine needle into the tumour to remove a small sample from it. A CT scan is done at the same time to help guide the surgeon to the exact area.

The sample is examined under a microscope by a doctor called a pathologist to find out what type of cells are present.

**Treatment**

The treatment you have will depend on your individual situation and your general health. The main treatment for primary CNS lymphoma is usually chemotherapy. Some people may have radiotherapy after chemotherapy or it may be given on its own. Drugs called **steroids** are also used.

A team of specialists will plan your treatment. This will usually include a cancer doctor (oncologist), doctors who specialise in conditions of the brain (a neurologist or neurosurgeon) and a specialist nurse.

They will explain the aims of your treatment, what it involves, and the benefits and disadvantages. Deciding on the treatments that are right for you is a decision you make in partnership with your doctor. Make sure you have enough information and time to help you make any treatment decisions.

**Chemotherapy**

**Chemotherapy** is the use of anti-cancer (cytotoxic) drugs to destroy cancer cells. It is often the first treatment for primary CNS lymphoma. A nurse will usually give you the chemotherapy drugs into a vein (intravenously). This is usually through a fine tube that goes under the skin of your chest and into a vein close by (central line). Or it can be given into a fine tube that is put into a vein in your arm and goes up into a vein in your chest (PICC line).

Chemotherapy can sometimes be given into the fluid surrounding the brain by a lumbar puncture, called **intrathecal chemotherapy**. It’s done in a similar way to a lumbar puncture. Before removing the needle, the doctor puts a small amount of liquid chemotherapy into the spinal fluid.

Another way of giving chemotherapy is to inject the drugs into a special flat plastic bubble called an ommaya reservoir. A surgeon places it under your scalp during an operation. It holds the chemotherapy drugs as they flow through a small tube into the ventricles (fluid-filled cavities) of the brain.

Doctors use chemotherapy drugs that are able to pass into the brain and spinal cord. Part of the brain’s natural protection stops certain drugs getting through (called the blood-brain barrier). The most common drug doctors use to treat primary CNS lymphoma is **methotrexate**, which is given in high doses. It can be given on its own or in combination with other chemotherapy drugs and/or steroids.

**Side effects of chemotherapy**

Your doctor or specialist nurse will tell you what to expect. They can prescribe medicines to prevent or reduce side effects and give you advice on managing them.

One of the main side effects is risk of infection. Chemotherapy temporarily reduces the number of white blood cells, which help fight infection. You may be given drugs to reduce your risk of getting an infection.

Chemotherapy can also affect red blood cells, which carry oxygen around the body, and platelets which help the blood to clot. Other side effects can include feeling sick, sore mouth and hair loss. Always let your doctor or nurse know about any side effects you have. We have more detailed information on some possible side effects of chemotherapy.
Radiotherapy

Radiotherapy treatment uses high energy rays to destroy the cancer cells. Radiotherapy for primary CNS lymphoma is given to the whole brain. Some people have radiotherapy after chemotherapy but not everyone will need it, particularly if you are older. If you are not well enough to have chemotherapy you may be treated with radiotherapy on its own. When lymphoma starts in the eye, radiotherapy can be given directly to the eye.

Side effects of radiotherapy

Your doctor and specialist nurse will talk to you about the immediate side effects and the risk of long-term effects. Newer ways of giving radiotherapy aim to give a higher dose of radiotherapy to the tumour without damaging the surrounding normal brain.

You will be very tired after radiotherapy and this can carry on for weeks or longer after it finishes. Try to get plenty of rest. The skin on your scalp may become itchy and red or darker and you will lose the hair on in the area being treated. This usually grows back again after 2-3 months. You will be given advice on looking after the skin on your scalp and coping with hair loss.

Steroids

Steroids are used to reduce swelling around the lymphoma and may also shrink the lymphoma. They improve symptoms and help you to feel better. If you have raised pressure in the brain you will be treated with steroids straightaway. They are also used if a primary spinal lymphoma is causing pressure on the spine.

You usually have steroids as tablets, but they may also be given as an injection into a vein (intravenously). Possible side effects of steroids include indigestion, weight gain, restlessness, agitation and sleep disturbance. Taking steroids with food can help reduce indigestion. Your doctor may also prescribe medication to prevent it. It is very important to take them exactly as they have been prescribed.

HIV treatment

If you have HIV, you will be advised to start highly active antiretroviral therapy (HAART). This improves survival in people with HIV-related lymphomas.

Surgery

Surgery is rarely used to treat primary CNS lymphoma because there are often several tumours or the tumour is deep in the brain.

Medicines for seizures

If you have seizures (fits), you may be given a medicine called an anticonvulsant to help prevent them.

Help with your recovery

It may take some time for treatment to improve your symptoms. Some people may need support to help them to recover. This may be from a physiotherapist to help improve your balance, walking or strength. Occupational therapists can provide equipment and help you be more independent. Other services such as speech therapy or psychological support services are also available.

Your feelings

Being diagnosed with a brain tumour can feel frightening at times. You may have many different emotions, including anxiety, anger, fear or feeling low or depressed. Many people go through these in coping with their illness. It’s important to get the support you need.

You may find it helpful to talk things over with family and close friends and with your doctor or nurse. You can also talk to one of our cancer support specialists. Sometimes people need more help to cope with difficult feelings of anxiety or depression. Your doctor can refer you to a counsellor or psychologist for more support.

Other information
Driving

You may not be allowed to drive for a period of time, depending on the treatment you have had and if you have had any seizures (fits). Although this can be upsetting, it’s important to follow the advice you are given. Your doctor will ask you to contact the Driver & Vehicle Licensing Agency (DVLA). It is your responsibility to contact the DVLA. Your doctor or nurse will explain what you need to do.

Useful organisations

Driver and Vehicle Licensing Association (DVLA)

The DVLA advises GPs and other members of the medical profession on the medical standards of fitness to drive. Patients should seek advice from their doctors.

The lymphoma Association

The Lymphoma Association gives emotional support, advice and information on all aspects of lymphoma. It has a national network of people with lymphoma and local groups.

References and thanks

This information has been compiled using a number of reliable sources, including:


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Thanks to people like you

Thank you to all of the people affected by cancer who reviewed what you’re reading and have helped our information to grow.

You could help us too when you join our Cancer Voices Network - [find out more](#).

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