Burkitt lymphoma

This information page is about Burkitt lymphoma, a very fast-growing type of high-grade non-Hodgkin lymphoma (NHL). This page is mainly about the sporadic form, which is the most common form of Burkitt lymphoma in the UK.

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Quick overview of Burkitt lymphoma

What is it?

Burkitt lymphoma is a cancer of the lymphatic system. It develops from B lymphocytes (white blood cells that fight infection) and usually grows very quickly. It often responds well to treatment and, in many cases, can be cured with intensive chemo-immunotherapy, which is chemotherapy with antibody therapy.

What are the symptoms and how is it diagnosed?

Symptoms often develop quickly, over just a few days or weeks. Burkitt lymphoma usually causes lots of lymph nodes to enlarge in many different parts of the body, and often involves your abdomen (tummy) and bowel. Other organs like your spleen and liver may be affected. The lymphoma may also be in the bone marrow and can spread to your central nervous system (CNS; brain and spinal cord).
How is it treated?

Treatment usually begins very soon after diagnosis. If you are fit enough, this is a combination of strong chemotherapy drugs and the antibody treatment rituximab. Treatment is usually given through a central line and most people stay in hospital for all or most of their treatment course, which can take several months.

If you have lower-risk Burkitt lymphoma or you can’t have stronger treatment, you may be offered less intensive treatment.

Drugs that reach your CNS, including intrathecal chemotherapy (chemotherapy given by lumbar puncture into the fluid around your spine), are usually given as part of your treatment.

What is Burkitt lymphoma?

Lymphomas can develop when a lymphocyte grows out of control. There are two main groups of lymphomas: Hodgkin lymphomas and non-Hodgkin lymphomas (NHL). Non-Hodgkin lymphomas are further grouped into:

- low-grade (slow-growing) or high-grade (fast-growing)
- T-cell lymphoma (develops from abnormal T lymphocytes or T cells) or B-cell lymphoma (develops from abnormal B lymphocytes or B cells).

Burkitt lymphoma is a very fast-growing type of high-grade NHL. It develops from abnormal B cells.

There are different types of Burkitt lymphoma, including:

- an endemic (found regularly in a certain area) form that occurs mainly in children in Africa
- a sporadic (found occasionally) form, which is the most common form of Burkitt lymphoma in the UK
- immunodeficiency-associated Burkitt lymphoma, which usually develops in people with HIV or who have had an organ transplant.

This page is mainly about the sporadic form of Burkitt lymphoma. We have other information about lymphoma in people with HIV or who have had an organ transplant.

Occasionally, a lymphoma has features of Burkitt lymphoma and another type of lymphoma. This might be called a ‘high-grade B-cell non-Hodgkin lymphoma, not otherwise specified’. If this is the case for you, your hospital specialist can tell you if your lymphoma will be treated like Burkitt lymphoma or a different type of lymphoma.
Who gets Burkitt lymphoma and what causes it?

Burkitt lymphoma is uncommon – about 210 people are diagnosed with this type of lymphoma every year in the UK. Although it is the most common type of NHL in children, it can occur at any age.

Burkitt lymphoma affects about three times more men than women.

The endemic form is linked to malaria and to the Epstein-Barr virus (EBV), a common virus that also causes glandular fever.

The cause is less clear for the sporadic form, although this is sometimes linked with EBV infection. It is important to note that the vast majority of people who have EBV do not go on to develop lymphoma.

People with HIV are more likely to develop Burkitt lymphoma than people without HIV, but most people with Burkitt lymphoma do not have HIV.

What are the symptoms of Burkitt lymphoma?

Symptoms of Burkitt lymphoma usually develop quickly, over just a few days or weeks.

The most common symptom is one or more lumps, which often develop in several parts of your body. These are caused by lymphoma cells building up in your lymph nodes (glands), causing them to enlarge (swell).

You need a biopsy and other tests like blood tests and scans to diagnose Burkitt lymphoma.

I discovered a lump under my arm. Looking back, I wasn’t too anxious about the lump. It felt about the size of a finger nail and I thought it was just a boil that would go away perfectly normally. But within 3 weeks the lump had grown to the size of an orange and was getting bigger by the day. Jennie, diagnosed at 27

Burkitt lymphoma is often found in the abdomen (tummy) and bowel. The lymphoma can cause a variety of symptoms depending where it is growing. For example, if the lymphoma is growing in your abdomen or bowel, you might have:

- abdominal (tummy) or back pain
- nausea and diarrhoea
- swelling of your abdomen because of fluid collecting
- bleeding from your bowel or pain from bowel obstruction (blockage).

The lymphoma can affect other parts of your body, for example:
- organs like your spleen, liver, ovaries, kidneys and breasts
- your bone marrow (the spongy centre of some of our bones)
- your central nervous system (CNS; brain and spinal cord).

Burkitt lymphoma found outside of the lymph nodes is called ‘extranodal’.

Burkitt lymphoma in the bone marrow can cause low blood counts as the lymphoma cells take up the space of normal cells. You might develop:

- anaemia (low red blood cells), which can cause tiredness and shortness of breath
- thrombocytopenia (low platelets), which makes you more likely to bruise and bleed.

People with Burkitt lymphoma often have bulky disease (large lumps) as this type of lymphoma grows very quickly.

Some people have other common symptoms of lymphoma, which include weight loss, night sweats and fevers. These three symptoms often occur together and are called ‘B symptoms’.

**How is Burkitt lymphoma diagnosed?**

Burkitt lymphoma is diagnosed with a small operation called a biopsy. A sample of tissue that is affected by lymphoma, such as a swollen lymph node, is removed, usually under local anaesthetic. The sample is examined by an expert lymphoma pathologist. The pathologist then does tests on the tissue to find out what type of lymphoma it is.

Occasionally, Burkitt lymphoma cells are found first in the bone marrow or in tissue removed for other reasons.

You have other tests to find out more about your general health. Tests are also needed to find out which parts of your body are affected by lymphoma – this is called ‘staging’. These tests usually include:

- a physical examination
- blood tests to look at your general health, including your blood cell counts
- a scan – usually a combined PET/CT scan
- bone marrow tests
- a lumbar puncture to check if the lymphoma has spread to your CNS.

Although waiting for the results of your tests can be difficult, your specialist is collecting important information during this time. It is important that your specialist knows exactly what type of lymphoma you have so they can give you the most appropriate treatment.
What does ‘stage’ mean?

The tests you have are part of ‘staging’ the lymphoma – working out how far it has spread and how much of your body is affected. Different staging systems are used for adults and children with Burkitt lymphoma. For both, there are 4 stages, ranging from stage 1 (lymphoma in one area) to stage 4 (the stage at which lymphoma is most widespread).

Staging is important because it helps your specialist plan the best treatment for you.

What do ‘low-risk’ and ‘high-risk’ mean?

Burkitt lymphoma is normally divided into ‘low-risk’ and ‘high-risk’ based on stage and other factors, for example:

- levels of a chemical called ‘lactate dehydrogenase’ (LDH) in your blood
- how the lymphoma is affecting your day-to-day life.

Most people with Burkitt lymphoma have a high-risk form. This might sound alarming but there are very effective treatments for Burkitt lymphoma. People with low-risk Burkitt lymphoma might need slightly less treatment.

What is the outlook for people with Burkitt lymphoma?

Treatment for Burkitt lymphoma is intensive, but many people can be cured.

Most children are cured with current standard treatment and the outcome is often very good for younger adults. Burkitt lymphoma can be more difficult to treat in older adults who are generally less able to tolerate intensive treatment.

Survival statistics can be confusing as 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. 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 consider other ‘risk factors’, like your age, symptoms, and other conditions you have. Using these, they can predict how likely you are to respond to a particular treatment.

How is Burkitt lymphoma treated?

Burkitt lymphoma is fast-growing and treatment normally starts as soon as possible. Chemotherapy with the antibody treatment rituximab is recommended for Burkitt lymphoma, even if the disease is localised (found in only one place) as it grows so quickly. However, some people might also have surgery to remove large tumours or a bowel obstruction.
In chemotherapy, a regimen (combination of drugs) is used – each drug kills lymphoma cells in a different way. The treatment is given in cycles with drugs given on certain days followed by a rest period before the next cycle begins.

Treatments for Burkitt lymphoma are intensive. You are likely to need to stay in hospital for several months while you have your treatment. There are several reasons for this:

- each treatment takes several days to give
- you need treatments to support your body while you are having treatment, such as fluids to keep your kidneys working well
- you need lots of tests after treatment to check your organs are working well and to check your blood counts.

Some people may be able to go home between treatments. Discuss your treatment plan and what it involves with your medical team.

You are likely to have a central line fitted – this is a tube that stays in your vein throughout your treatment. Your treatment can be given and blood samples can be taken through the central line, avoiding the need for repeated needles.

As Burkitt lymphoma can spread to your CNS, most people also have chemotherapy that can reach their CNS. This might involve intrathecal chemotherapy, which is chemotherapy given by lumbar puncture into the fluid around your spinal cord. Only certain drugs can be given in this way, for example methotrexate and cytarabine. Sometimes, drugs that reach your CNS can be given intravenously (into a vein).

### Chemotherapy for Burkitt lymphoma

Your specialist chooses a chemotherapy regimen based on the results of your tests and your individual circumstances.

- If you have low-risk Burkitt lymphoma, a common regimen is 3 cycles of R-CODOX-M (rituximab, cyclophosphamide, vincristine, doxorubicin, methotrexate).
- If you have high-risk Burkitt lymphoma, a common regimen is 2 cycles of R-CODOX-M alternating with 2 cycles of R-IVAC (rituximab, ifosfamide, etoposide, cytarabine).

Your specialist might suggest a different regimen.

If you are older or have other health conditions, the treatment might need to be modified to make it safer for you. Clinical trials in recent years have tested whether the intensity of chemotherapy for Burkitt lymphoma can be reduced, particularly for people unable to tolerate high-intensity regimens. Lower intensity regimens may be more effective for some people as they may be more likely to complete the treatment course, for example DA-EPOCH-R, which is DA-EPOCH chemotherapy with rituximab.
Children and adolescents have different chemotherapy regimens, which are usually very effective. Treatment usually consists of several courses of chemotherapy, including:

- induction treatment to put the lymphoma into remission (no evidence of lymphoma)
- consolidation treatment to keep the lymphoma in remission.

Rituximab is likely to be added to the chemotherapy.

It was explained to me that the chemotherapy used for Burkitt lymphoma is stronger than many other regimens, so I would be spending quite a lot of the time in hospital. In fact I was in hospital for 4 months and was given CODOX-M and R-IVAC. The first part of the chemotherapy was awful but scans showed that the chemotherapy was working and I knew I had to keep going. Ian, diagnosed at 47

Side effects and supportive care

All treatment has a risk of side effects (unwanted effects). Your medical team can give you more information about the typical side effects of the treatment they recommend for you.

Chemotherapy is often most effective against fast-growing cells, like Burkitt lymphoma cells. For this reason, treatment usually kills a large number of lymphoma cells very quickly. This can cause a problem called ‘tumour lysis syndrome’. Dying cells release chemicals as they break down. If a lot of cells break down at once, your body may struggle to get rid of the extra chemicals. This can cause serious problems for your kidneys and heart.

There are effective treatments for tumour lysis syndrome and you are monitored closely for any signs of it developing. If your specialist thinks you are at risk of tumour lysis syndrome, you can have drugs like allopurinol and rasburicase (Fasturtec®) to prevent it.

You have other supportive treatments to control side effects during your treatment. These might include:

- antiemetics, which help reduce nausea (feeling sick) and vomiting (being sick)
- prophylactic (preventive) antibiotics and antifungals, to reduce the risks of infection
- growth factors (G-CSF), to help your bone marrow recover quickly by boosting your blood counts
- blood transfusions
- other drugs that reduce some of the side effects of the chemotherapy drugs used in these regimens.

These drugs might include mesna, which protects the bladder lining from irritation, and folinic acid, which can reduce the side effects of methotrexate.
Our section on living with lymphoma has more information on what you can do to support yourself during your treatment, for example information about diet, exercise and the emotional impact of a lymphoma diagnosis.

**What does follow-up involve?**

You have a scan at the end of treatment to see how you have responded. This is usually a PET/CT scan. Sometimes it is hard to tell whether any lumps in your body after treatment are scar tissue or lymphoma. A PET/CT scan shows areas of active (growing) lymphoma.

You might need other tests after treatment finishes. Your specialist can use the results of the scan and other tests, if needed, to see if you are remission (no evidence of lymphoma) or if you need further treatment.

You have regular follow-up appointments when you are in remission. These are to check that:

- you are recovering well from treatment
- you have no signs of relapse
- you are not developing any late effects (side effects that develop months or years after treatment).

You might have blood tests. You are unlikely to have a scan unless you have troubling symptoms.

At each appointment, your specialist examines you and asks if you have any concerns or symptoms.

The schedule for follow-up appointments depends on your individual circumstances and varies between hospitals. Usually, you have frequent appointments at first, sometimes monthly.

Burkitt lymphoma usually stays in remission if treatment is successful. Relapse becomes less likely as time goes on. The frequency of appointments gradually decreases if you remain well. Follow-up usually continues for around 2 years after the end of treatment but some people are followed-up for longer.

**What happens if Burkitt lymphoma comes back or doesn’t respond to treatment?**

If your lymphoma doesn’t respond well to your first treatment, it is called ‘refractory’.
Relapsed or refractory Burkitt lymphoma can be difficult to treat. In both situations, your specialist might offer you a stronger treatment or an experimental treatment as part of a clinical trial. If you decide against further treatment, you can have palliative care to help control your symptoms. Your specialist can discuss all the options with you.

**Treatment for relapsed or refractory Burkitt lymphoma**

If you are fit enough, you might be offered a different type of chemotherapy. This is sometimes called ‘salvage chemotherapy’. If you respond to chemotherapy, your specialist might suggest high-dose chemotherapy and a stem cell transplant. This intensive form of treatment can offer a better chance of a long-lasting remission.

An experimental treatment might be available through a clinical trial. Read more about clinical trials in our dedicated section, Lymphoma TrialsLink, and search for a trial that might be suitable for you.

**Further information and support**

If you would like further information on anything you have read on this page or would like to talk about any aspect of your lymphoma, please call our confidential Freephone helpline on **0808 808 5555**, visit our website at www.lymphomas.org.uk, or email **information@lymphomas.org.uk**. We also have support groups and forums that can offer support for people with lymphoma. We might be able to put you in touch with a Buddy – someone who has been in a similar situation to you and can offer support.

**Sources used**

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@lymphomas.org.uk or phone on 01296 619409 if you would like a copy.


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We continually strive to improve our resources for people affected by lymphoma and we would be interested in any feedback you might have about this information. Please visit www.lymphomas.org.uk/feedback or email publications@lymphomas.org.uk if you have any comments. Alternatively please phone our helpline on 0808 808 5555.

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