Helpline (freephone) 0808 808 5555
information@lymphoma-action.org.uk
www.lymphoma-action.org.uk

Burkitt lymphoma

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

On this page

What is Burkitt lymphoma?

Lymphoma is a type of blood cancer that develops when white blood cells called **lymphocytes** grow out of control. Lymphocytes are part of your **immune system**. They travel around your body in your **lymphatic system**, helping you fight infections. There are two types of lymphocyte: **T lymphocytes (T cells)** and **B lymphocytes (B cells)**.

There are lots of different **types of lymphoma**. Burkitt lymphoma is type of **non-Hodgkin lymphoma** that develops from B cells. It is a **fast-growing (high-grade) lymphoma**.

Who gets Burkitt lymphoma?

Burkitt lymphoma is uncommon. Only around 250 people are diagnosed with it in the UK each year. It affects three to four times more men than women.

There are three subtypes of Burkitt lymphoma, which tend to affect different types of people:

- **Sporadic Burkitt lymphoma** is the most common subtype in the UK. 'Sporadic' means it occurs occasionally and irregularly. Sporadic Burkitt lymphoma is sometimes linked to infection with Epstein-Barr virus (EBV – the virus that causes glandular fever). However, most people with EBV do **not** develop lymphoma. It can affect people of any age, but it typically develops in children and young adults.
- **Immunodeficiency-associated Burkitt lymphoma** sometimes develops in **people who have HIV** or in **people who have had an organ transplant**.
- **Endemic Burkitt lymphoma** is more common in areas where malaria is widespread (endemic), such as equatorial Africa, Brazil and Papua New Guinea. It is linked to infection with malaria and EBV. It usually develops in young children.

This information focuses on the sporadic form of Burkitt lymphoma.

Occasionally, a high-grade lymphoma has features of Burkitt lymphoma and another type of lymphoma. This is called '**high-grade B-cell non-Hodgkin lymphoma, not otherwise specified**'.

If you are under 18, or are a parent or carer of someone under 18 who has Burkitt lymphoma, our section on **lymphoma in children** has more information for this age group. Young people (up to 24) with Burkitt lymphoma might find our section on **lymphoma in young people** helpful.

Symptoms

Burkitt lymphoma grows very rapidly, which means that **symptoms** 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 **swollen lymph nodes**.

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

Because it grows quickly, people with Burkitt lymphoma often develop large lumps of lymphoma (known as 'bulky disease'). The exact symptoms you experience depend on where the lymphoma is growing.

Sporadic Burkitt lymphoma often grows in the tummy (abdomen) and bowel, which might cause:

- tummy or back pain
- feeling sick or being sick
- diarrhoea
- swelling of your tummy
- bleeding from your bowel
- pain from a blockage in your bowel.

It is common for adults with Burkitt lymphoma to experience fevers, night sweats and weight loss. These three symptoms often occur together and are known as 'B symptoms'.

Around 1 in 3 people with Burkitt lymphoma have lymphoma cells in their bone marrow (the spongy centre of some of your larger bones). Here, the lymphoma cells take up space that is normally used to make healthy blood cells. This might lead to:

- **anaemia** (shortage of red blood cells), which can cause tiredness and shortness of breath
- **thrombocytopenia** (shortage of platelets), which makes you more likely to bruise and bleed
- **neutropenia** (shortage of white blood cells), which makes you more likely to get infections.

Burkitt lymphoma might also develop in your head and neck, or other parts of your body, such as inside your chest, your breasts, spleen or central nervous system (CNS; brain and spinal cord).

Endemic Burkitt lymphoma usually develops in the jaw or face, or in the bowel or urinary tract. Immunodeficiency-associated Burkitt lymphoma often affects the small bowel.

Diagnosis and staging

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. An expert **lymphoma pathologist** examines the sample under a microscope and does specialist tests on the tissue to find out what type of lymphoma it is.

You have other tests to find out more about your general health and to find out which parts of your body are affected by lymphoma. This is called '**staging**'. These tests usually include:

- a **CT scan**, often combined with a **PET scan**, to look at where the lymphoma is growing in your body
- a **bone marrow biopsy** to check for lymphoma cells in your bone marrow
- a **lumbar puncture** to look for lymphoma cells in the fluid around your brain and spinal cord
- **blood tests** to check your blood cell counts and test how well your liver and kidneys are working.

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. This helps them plan the most appropriate treatment for you.

> **When I went in to receive my diagnosis, I don't think anything could have shocked me. Frankly, I was past caring about the diagnosis. I was just anxious to start doing something about whatever was wrong.**
> **I was diagnosed with Burkitt lymphoma, a fast growing high-grade non-Hodgkin lymphoma. I had barely heard of lymphoma, much less Burkitt lymphoma, so wasn't surprised when my doctor told me they didn't often see people with Burkitt lymphoma.**
>
> Ian, diagnosed with Burkitt lymphoma in 2009

Staging of Burkitt lymphoma

Different staging systems are used for **adults** and **children** with Burkitt lymphoma. Both systems have 4 stages, ranging from stage 1 (lymphoma in one area) to stage 4 (the stage at which lymphoma is most widespread).

Staging helps your specialist plan the most appropriate treatment for you.

Low-risk and high-risk Burkitt lymphoma

Burkitt lymphoma is normally classed as ‘low-risk’ or ‘high-risk’ based on the results of your tests and scans, and how the lymphoma is affecting your day-to-day life.

Because it grows quickly, most people have high-risk Burkitt lymphoma when they are diagnosed. This might sound alarming but there are very effective treatments for Burkitt lymphoma. People who have high-risk Burkitt lymphoma usually need more intensive treatment than people who have low-risk Burkitt lymphoma.

Outlook

Burkitt lymphoma is usually treated with the aim of curing it, and many people go into **complete remission** (no evidence of lymphoma). Most children who have treatment for Burkitt lymphoma are cured. The outcome is also very good for younger adults. In older adults, Burkitt lymphoma can be more difficult to treat because older people are generally less able to tolerate intensive treatments.

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 individual factors (for example, your age and how fit you are) to help them judge how likely you are to respond to a particular treatment.

Remember that treatments are improving all the time and survival statistics are usually measured over 5 or 10 years after treatment. This means that statistics only tell you how people did in the past. Those people might not have received the same treatment as you. 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. Because of this variability, many people do not find survival statistics helpful. However, if you do want to know more about survival statistics for Burkitt lymphoma, **Cancer Research UK** have some information that you might find useful.

Treatment

Burkitt lymphoma grows very quickly so you usually start treatment straightaway.

The treatment your medical team recommends for you depends on whether you have **high-risk** or **low-risk** Burkitt lymphoma. Your team also takes into account your age, your general health and fitness, your feelings about treatment and factors that could be important to you in the future, such as having a family. They also consider any potential **side effects**, or long-term or **late effects** (health problems that might develop months or years after treatment). Your medical team should explain the possible side effects and late effects of your planned treatment.

Most people with Burkitt lymphoma have **chemotherapy** combined with the antibody treatment **rituximab**. Your medical team might consider several different **chemotherapy regimens** (combinations of drugs). They will recommend a chemotherapy regimen based on the results of your tests and your individual circumstances.

If you have high-risk Burkitt lymphoma and you are fit enough, you are likely to be treated with one of the following treatment regimens:

- **R-CODOX-M/R-IVAC**: **r**ituximab plus **c**yclophosphamide, vincristine (**O**ncovin), **dox**orubicin and **m**ethotrexate alternating with **r**ituximab plus **i**fosfamide, etoposide (**V**P-16) and cytarabine (**A**ra-**C**).
- **DA-EPOCH-R**: **d**ose-**a**djusted **e**toposide, **p**rednisolone, vincristine (also known as **O**ncovin), **c**yclophosphamide and doxorubicin (or **h**ydroxydaunorubicin) plus **r**ituximab. Six cycles of treatment are usually given for high-risk disease.
- **R-HyperCVAD (HDMTX)**: **r**ituximab plus **c**yclophosphamide, **v**incristine, doxorubicin (**A**driamycin) and **d**examethasone, and **h**igh-**d**ose **m**ethot**r**e**x**ate; '**hyper**' is short for 'hyperfractionated', which means that you have the same drug more than once in a day.
- **R-BFM**: **r**ituximab plus the '**B**erlin, **F**rankfurt, **M**ünster' protocol. This is a regimen developed in Germany that uses different chemotherapy drugs at different times during treatment. Treatment is given in phases: induction therapy, to put the lymphoma into remission; consolidation therapy, to keep the lymphoma in remission; reinduction therapy, to get rid of any lymphoma that is left after induction therapy; and finally maintenance therapy, to help prevent a relapse.

- **R-LMB**: **r**ituximab plus the '**l**ymphome **m**alin de **B**urkitt' protocol. This is a regimen developed in France that uses different chemotherapy drugs at different times during treatment. Treatment is given in phases: cytoreductive therapy, to reduce the number of cancer cells; induction therapy, to put the lymphoma into remission; consolidation therapy, to keep the lymphoma in remission; and finally maintenance therapy, to help prevent a relapse.

If you have low-risk Burkitt lymphoma, or if you are not fit enough to tolerate intensive chemotherapy, you might have a less intensive regimen such as:

- **DA-EPOCH-R**: **d**ose-**a**djusted **e**toposide, **p**rednisolone, vincristine (also known as **O**ncovin), **c**yclophosphamide and doxorubicin (or **h**ydroxydaunorubicin) plus **r**ituximab. Three cycles of treatment are usually given for low-risk disease.
- **R-CHOP**: **r**ituximab plus **c**yclophosmamide, doxorubicin (or **h**ydroxydaunorubicin), vincristine (**O**ncovin) and **p**rednisolone.
- **R-CHEOP**: **r**ituximab plus **c**yclophosmamide, doxorubicin (or **h**ydroxydaunorubicin), **e**toposide, vincristine (**O**ncovin) and **p**rednisolone.

Most people also have chemotherapy to prevent the lymphoma spreading to their CNS. This is called **CNS prophylaxis**. It 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 through a drip into a vein (**intravenously**).

There are lots of other chemotherapy regimens and your specialist might suggest a different one for you, based on your particular circumstances.

Most treatments for Burkitt lymphoma are very intensive. You are likely to need to stay in hospital for some of your treatment so your medical team can monitor you and give you medicines to support your body while you recover. Depending on the type of chemotherapy you are receiving, at some hospitals you might have the option of having some intravenous chemotherapy at home. This is called 'ambulatory chemotherapy'. You go to the hospital every day for check-ups and to have your treatment set up in a special pump. You wear the pump in a backpack so you don't have to stay in hospital while you have the treatment. You can contact the hospital at any time if you have any problems.

Most people have a **central line** fitted. This is a tube that stays in your vein throughout your treatment. You have your treatment through the central line, and you can also have blood samples taken from the line, avoiding the need for repeated needles.

> **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 with Burkitt lymphoma in 2009

Tumour lysis syndrome

Chemotherapy is often very effective against fast-growing cells, like Burkitt lymphoma cells. It can kill a large number of lymphoma cells very quickly. These cells release chemicals as they break down. If a lot of cells break down at once, your body might struggle to get rid of the extra chemicals. This can cause serious problems for your kidneys and heart. This is called ‘tumour lysis syndrome’.

If your specialist thinks you are at risk of developing tumour lysis syndrome, they will prescribe medicines to help prevent it. During your treatment, your medical team monitors you closely for any signs that it might have developed. They can give you treatment for it if you need it.

Follow-up

You have a **scan** at the end of treatment to see how you have responded. This is usually a PET/CT scan. You might also have other tests. Your specialist can use the results of the scan and other tests, if needed, to see if you are in remission (no evidence of lymphoma) or if you need further treatment.

While my treatment had finished, my recovery had only just begun. The question I had to face was how to move on from feeling 'institutionalised' to returning to the 'real world' and adapting to a 'new normality'. To help with this, I received support from specialist nurses and had regular check-ups with my consultant over 5 years.

Kathleen, diagnosed with Burkitt lymphoma in 2006

When you are in remission after treatment, you have **follow-up appointments** to check that:

- you are recovering well from treatment
- you have no signs of the lymphoma coming back (relapsing)
- you are not developing any **late effects**.

At these appointments, your doctor examines you and asks if you have any concerns or symptoms. You might have blood tests. You are unlikely to have a scan unless you have troubling symptoms.

Depending on the usual practice at your hospital, you might have regular follow-up appointments, or you might be given guidance on **booking your own appointments** as-and-when you need them. Most hospital teams offer follow-up in their clinic for at least 1 year after you finish treatment. It is rare to experience a relapse if you've been in remission for more than a year.

After your planned hospital follow-up period ends, and providing you remain in remission, your GP usually becomes your main point of contact if you have any health concerns or notice anything unusual. Your GP should have a record of your diagnosis and all the treatment you've had.

If you are ever concerned about your lymphoma, however, you can contact your hospital team at any time. Don't wait for an appointment if you are worried.

Relapsed or refractory Burkitt lymphoma

Burkitt lymphoma usually stays in remission after successful treatment. However, sometimes it does not respond well to treatment. This is called 'refractory' lymphoma. Sometimes, it comes back (**relapses**) after successful initial treatment. Relapse is most likely to happen within 2 years of the end of your first treatment. As time goes on, lymphoma is less likely to relapse.

Relapsed or refractory Burkitt lymphoma can be difficult to treat. Your specialist might ask you if you'd like to take part in a **clinical trial** testing a new treatment. They might suggest more chemotherapy. This is sometimes known as 'salvage' treatment. The most commonly used regimens include:

- **R-IVAC**: **r**ituximab plus **i**fosfamide, etoposide (**V**P-16) and cytarabine (**A**ra-**C**)
- **R-GDP**: **r**ituximab plus **g**emcitabine, **d**examethasone and cisplatin (**P**latinol)
- **R-DHAP**: **r**ituximab plus **d**examethasone, **h**igh-dose cytarabine (**A**ra-C) and cisplatin (**P**latinol)
- **R-ICE**: **r**ituximab plus **i**fosfamide, **c**arboplatin and **e**toposide.

If you respond to more chemotherapy and you are fit enough, your specialist might suggest high-dose chemotherapy and a **stem cell transplant**. This aims to make your remission last as long as possible. A stem cell transplant works best if the lymphoma responds at least partially to salvage therapy.

If you decide against further treatment, you can have **palliative care** to help control your symptoms. Your specialist can discuss all of your options with you.

Research and targeted treatments

Treatment for Burkitt lymphoma is usually successful but doctors continue to research treatments that are effective with as few **side effects** and **late effects** as possible. Current research is focusing particularly on targeted drugs, including:

- new **antibody therapies**, including 'bispecific' antibodies that stick to a protein on lymphoma cells and another protein on your own immune cells, boosting your body's immune response to lymphoma
- targeted drugs such as **HDAC inhibitors**, **BTK inhibitors**, **PI3K inhibitors** and **proteasome inhibitors**
- **antibody-drug conjugates**
- **cell signal blockers**
- **CAR T-cell therapy**.

Your medical team might ask if you would like to take part in a clinical trial. Find out more about clinical trials, and search for a trial that might be suitable for you, on **Lymphoma TrialsLink**.

References

The full list of references for this page is available on our website. Alternatively, email **publications@lymphoma-action.org.uk** or call 01296 619400 if you would like a copy.

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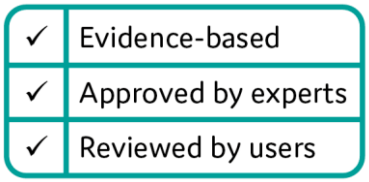

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