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Burkitt lymphoma

This information is about Burkitt lymphoma, a fast-growing type of high-grade non-Hodgkin lymphoma. The 'sporadic' form of Burkitt lymphoma is the most common form of Burkitt lymphoma in the UK.

On this page

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.

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 times more males than females.

> **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.

Read **Ian's story** on our website.

There are three main types of Burkitt lymphoma, which tend to affect different types of people.

- **Sporadic Burkitt lymphoma** is the most common type 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 linked to infection with malaria and EBV. It is more common in areas where malaria is widespread (endemic), such as equatorial Africa, Brazil and Papua New Guinea. It usually develops in young children.

This information focuses mainly 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

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 **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 fingernail 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

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's 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 your chest, breasts, spleen or central nervous system (brain and spinal cord).

Endemic Burkitt lymphoma usually develops in the jaw or face, or in the bowel or urinary tract.

Diagnosis and staging

Burkitt lymphoma is diagnosed with a small procedure called a **biopsy**. A sample of tissue, 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 can 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.

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.

Most people have high-risk Burkitt lymphoma when they are diagnosed. This might sound alarming but there are very effective treatments available. 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** (disappearance or significant shrinkage of lymphoma).

> **The consultant told me I had Burkitt lymphoma, and although it was rare and aggressive, it was also treatable.**
>
> Jean, diagnosed with Burkitt lymphoma

Read **Jean's story** on our website.

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 use the results of your tests and other individual factors (for example, your age and how fit you are) to help judge how likely you are to respond to a particular treatment.

Treatment

You usually start treatment for Burkitt lymphoma straightaway.

The treatment your medical team recommends for you depends on:

- whether you have high-risk or low-risk Burkitt lymphoma
- where in your body your lymphoma is (**stage**)
- your age

- your general health and fitness
- factors that could be important to you in the future, such as having a family
- potential **side effects** or **late effects** (health problems that might develop months or years after treatment)
- your preferences and feelings about treatment.

Most people with Burkitt lymphoma have **chemotherapy** combined with the antibody treatment **rituximab**. This is called chemo-immunotherapy. 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 / **r**ituximab plus **i**fosfamide, etoposide (**V**P-16) and cytarabine (**A**ra-**C**). Two alternating cycles of each regimen is given to give four treatment cycles in total.
- **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.

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

- **R-CODOX-M**: **r**ituximab plus **c**yclophosphamide, vincristine (**O**ncovin), **dox**orubicin and **m**ethotrexate
- **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.

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

Most people also have chemotherapy to prevent the lymphoma spreading to their central nervous system (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. Sometimes, drugs that reach your CNS can be given through a drip into a vein (**intravenously**).

Most treatments for Burkitt lymphoma are very intensive and can last several months. You might 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.

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.

Depending on the type of chemotherapy you are receiving, at some hospitals you might have the option of having some 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.

Marc talks about his experience of treatment for Burkitt lymphoma.

https://www.youtube.com/watch?v=iMLKLKV0LSw&feature=emb_logo

Tumour lysis syndrome

A rare but potentially serious side effect of chemotherapy is known as '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 tests and scans to see if you are in **remission** (disappearance or significant shrinkage of lymphoma) or if you need further treatment.

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

- are recovering well from treatment
- have no signs of the lymphoma coming back (relapsing)
- 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.

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 I have aches, pains or unexplained lumps, I do not hesitate to see my doctor. No, I am not turning into a hypochondriac, but I am only too well aware that cancer generally needs to be identified quickly and I do not propose to take any chances.

Kathleen, diagnosed with Burkitt lymphoma

Read **Kathleen's story** on our website.

If you are ever concerned about your lymphoma 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 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

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 BTK inhibitors, PI3K inhibitors, cell signal blockers and proteasome inhibitors
- **antibody-drug conjugates**
- **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 619409 if you would like a copy.

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