Burkitt Lymphoma (BL)

Lymphoma Australia
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Learning about your lymphoma can be like learning a new language. It takes time and practice. Please keep this document handy so you can refer back to it as often as you need to. **It will become easier to understand the more you read it.**

Introduction

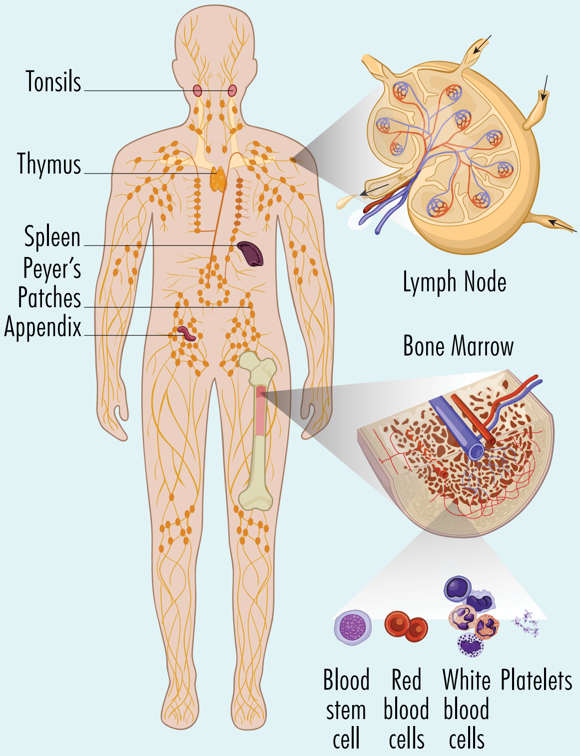
Burkitt lymphoma (BL) is a rare subtype of non-Hodgkin lymphoma that can affect children and adults. However, it is more common in children aged between 5 and 14 years.

Three out of every ten children with lymphoma have this subtype. It is much rarer in adults with only one or two adults in every 100 with lymphoma having BL.

If you've been diagnosed with BL, you have a type of fast-growing blood cancer. Fast growing cancers are sometimes called "aggressive". But even though this is an aggressive blood cancer, it usually responds well to treatment. Many people will even be cured with the right treatment.

Burkitt lymphoma affects a type of white blood cells called B-cell lymphocytes (B-cells). These cells are made in your bone marrow (the spongy middle part of your bones), before moving into your lymphatic system. They usually live in your lymph nodes or your spleen, and can also be found in your blood.

LYMPHATIC SYSTEM



B-cells fight infection and diseases to keep you healthy. They remember infections you had in the past, so if you get the same infection again, your immune system can fight it more effectively. When you have an infection, some of your B-cells move out of your spleen and lymph nodes, through the lymphatic system and to the area where your infection is. This means B-cells can be found in any part of your body.

When these cells don't grow or work properly, you can be diagnosed with lymphoma. You will have cancerous B-cells called lymphoma cells. The B-cells can travel all through your body, so your BL can also be found in any part of your body.

Signs & Symptoms

Because this is a fast-growing blood cancer, you may notice a lump that comes up quickly. These lumps are usually in your neck, armpit or groin. You may even get a lump in your chest or abdomen, but these may be harder to feel. Other symptoms you may have are:

- feeling unusually tired (fatigued)
- feeling out of breath
- bruising or bleeding more easily than usual
- infections that don't go away, or keep coming back (recurrent)
- sweating at night more than usual
- losing your appetite (not wanting to eat)
- losing weight without trying
- itchy skin
- B-symptoms (see picture on next page).

LUMP IN NECK

Types of Burkitt Lymphoma (BL)

There are three main types of BL.
They include:

Endemic BL: This subtype is more common in people with an African background, and is the most common type of lymphoma in children from Africa. It is more common in people who have had an infection with malaria or the Epstein-Barr virus (EBV). Endemic BL often starts in your jaw or other bones of your face, but it can also start in your abdomen (tummy area).

Sporadic BL: This subtype occurs all over the world and is believed to be more common in people who have had an infection with the Epstein-Barr virus (EBV). It usually starts in the lymph nodes in your abdomen, so it can cause swelling and pain there. This subtype of BL can spread to your brain and spinal cord, thyroid, tonsils, and bones in your face.

EBV is the virus that causes glandular fever or mononucleosis (mono). It's also sometimes called the "kissing disease" because it can be passed on through your saliva.

Immunodeficiency-associated BL: This subtype is more common for people who have a virus called human immunodeficiency virus (HIV), or who have developed acquired immunodeficiency syndrome (AIDS). However, you may also be at increased risk if you were born with immune deficiency, or if you take medication that weakens your immune system. These may include medications taken after an organ transplant, or for autoimmune diseases.

Diagnosis and Staging

If your doctor thinks you have BL you will need a biopsy. A biopsy is a procedure to remove part or all of an affected lymph node, or a sample of your bone marrow. The biopsy is checked by pathologists, to see if there are changes that help the doctor diagnose BL.

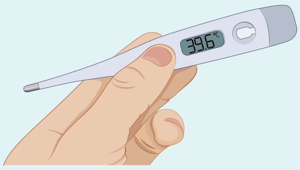
When you have a biopsy, you may have a local or general anaesthetic. This will depend on your age and what part of

B-SYMPTOMS

B-symptoms are a group of three distinct symptoms that some people with lymphoma can get. They often occur together and may indicate that your lymphoma is more advanced. **Contact your doctor as soon as possible if you get B-symptoms.**

Drenching night sweats – where your clothes and bedding become saturated.

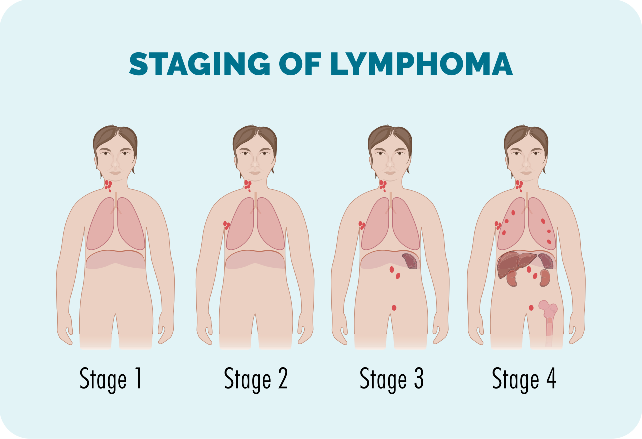
Losing weight without trying, and without other reason.



A high fever of 37.5° or more that keeps coming back or does not go away even when you don't have an infection. You may even get chills.

your body the biopsy is taken from. Your doctor will be able to tell you which subtype of BL you have when they get your biopsy results. Most children will have a general anaesthetic so they sleep through the procedure, avoiding distress and ensuring they remain very still during the biopsy.

If you have BL, your doctor will organise more tests to check if it has spread to other parts of your body. These tests are called "staging". Your stage will depend on where your lymphoma cells are, and how many parts of your body have the lymphoma. You might have Stage 1, 2, 3 or 4.


STAGING OF LYMPHOMA

Staging tests

Staging tests may include:

- Positron Emission Tomography (PET) scan
- Computed Tomography (CT) scan
- Bone Marrow Biopsy
- Lumbar Puncture

You can find more information on staging tests at our website. Just look down the left-hand side to find the test you want more information on: https://www.lymphoma.org.au/lymphoma/tests-diagnosis-and-staging/.

Stage I (1) and II (2) lymphoma are considered early-stage lymphomas, meaning that only one or two areas either above OR below your diaphragm have lymphoma. Stage III (3) lymphoma is considered an advanced stage and means that you have lymphoma on both sides of your diaphragm.

Stage IV (4) lymphoma is the most advanced stage, and means you have lymphoma on both sides of your diaphragm, and in one of your organs such as your liver, lungs, brain, bones or bone marrow.

Cytogenetic tests

Cytogenetics are tests on your blood and biopsies that look for changes in your chromosomes or genes. We usually have 23 pairs of chromosomes, and they are numbered according to their size. If you have BL, your chromosomes may look different.

On your chromosomes are many genes that tell the proteins and cells in your body how to look or act. If there is a change (mutation) in these chromosomes or genes, your proteins and cells will not work properly.

The mutation you will probably have if you have BL is called a translocation. This means, a gene called **"MYC gene"** which is usually on your 8th chromosome, will swap places with a gene on your 14th chromosome. Your doctor will be able to talk to you about any changes you have and explain these in more detail.

Please make sure you ask your doctor to explain your individual changes.

Double or Triple hit mutations

Rarely you may have a mutation called double hit or triple hit. If you do have this mutation, you have a rearrangement of your genes. This means that two or three of your genes have rearranged themselves and are no longer in the right place. This rearrangement means your genes are now unable to give proper instructions to your B-cells on how, and when to grow. If you have a double hit mutation, you have rearrangements in the MYC and, either your BCL2 or your BCL6 gene (only one of your BCL genes is rearranged in double hit, but you have two total rearrangements with MYC and BCL). If you have a triple hit mutation, you will have rearrangements in all three genes.

Rearrangements in Double hit and Triple hit lymphoma

	MYC	BCL2	BCL6
Double hit	Yes	Yes	No
Double hit	Yes	No	Yes
Triple hit	Yes	Yes	Yes

As well as the above tests, you may have other tests to check how well your body's organs are working, including your lungs, liver and kidneys. This is done to make sure it is safe for you to have treatment. It will also help your doctor to make sure that your treatment does not cause too much damage to them.

Before you start treatment

Before you start treatment you will also need to have some baseline tests done. These can include blood tests to check how well your liver and kidneys are working, scans on your heart, or lung function tests. These are to make sure that you are well enough to have treatment without it causing you to become too unwell. Throughout your treatment you will have regular blood tests which will be compared to these baseline tests. You may also have further lung tests and heart scans to make sure that the treatment has not caused any damage to your organs.

Fertility – some cancer treatments can make it harder to fall pregnant, or to get somebody pregnant. If you (or your child) are planning to have children later in life, talk to your doctor about how to preserve your fertility.

Questions for your doctor before you (or your child) start treatment

Starting treatment can feel overwhelming, and even knowing what questions to ask can be difficult. To help get you started, we have put together some questions you may like to consider asking. Click here to download our Questions to ask your Doctor" factsheet or scan the QR code at the end of this document.

Treatment options

Because BL is aggressive, you will start treatment soon after you are diagnosed. Depending on the results of your biopsies, you may have chemotherapy (chemo), with or without another medication called a monoclonal antibody (MAB). MABs help your own immune system fight the cancer more effectively, and chemo directly kills the lymphoma cells. However, the MAB only works if you have a certain marker on your lymphoma cells called CD20. So, if you don't have this marker, you will have chemo without the MAB.

Chemo and MABs are usually given to you as an infusion into your vein. If you have BL in your brain or spinal cord, or your doctor thinks there's a chance the lymphoma will spread there, you may also have an injection in your back where the doctor injects a little bit of chemo into the fluid that surrounds your spine. This is called intrathecal (or IT) chemo. Some MABs can be given as an injection into the fat in your abdomen (tummy area).

Treatment with chemo and MABs is given in cycles. This means you will have a period of treatment, then a break so your good healthy cells can recover, and then more treatment.

You may also need an operation to remove your tumour. Depending on the size and location of your tumour, your doctor may suggest you have the operation before chemo, or after a few cycles of treatment. If your tumour is very big, or too close to your spine, blood vessels or organs, they may want you to have some chemo first to shrink it. This makes the operation easier and protects your vital organs.

Treatment for children

Treatment for children with Burkitt lymphoma is often a little different from treatment for adults with Burkitt's. Many childhood cancers are treated with protocols based on clinical trials, so it is best to speak to your child's haematologist or treating team about what protocol they will have. However, many of the medications will be similar to those used to treat adults.

Treatment protocols

The most common chemotherapy protocols used for adults with BL include CODOX M alternating with IVAC, or DA-EPOCH

CODOX-M includes different types of chemo given over a 16-day period – but you will not have treatment every day. The chemotherapies you have are called doxorubicin, vincristine, cyclophosphamide, methotrexate and cytarabine. You will also get an injection (called filgrastim) in your tummy to help your white blood cells grow back more quickly to protect you from infection.

IVAC – after you have completed your cycle of CODOX-M and your blood counts have recovered, you will have a different protocol called IVAC. This protocol is given over 7 days and includes chemotherapies called cytarabine, etoposide and ifosfamide. You will also have some additional medications to protect your bladder, and to help with clearing the chemo from your body.

DA-EPOCH is a protocol that has several chemo medications given over a 6-day period then a break, before having the same protocol again 21 days after your first treatment day. The chemotherapies are called doxorubicin, vincristine, cyclophosphamide and etoposide. You will also have a steroid tablet and an injection in your tummy to help your infection fighting white blood cells grow back more quickly.

(The DA in DA-EPOCH/R stands for "dose adjusted", so the doctor can change the dose depending on your age, weight and overall health).

DA-EPOCH-R is the same as DA-EPOCH but has an extra medication called rituximab. This is a MAB, and you may receive this if your lymphoma cells have the CD20 marker on them.

Rituximab is not routinely given to children and teenagers. However, there are now clinical trials to see if DA-EPOCH-R can improve outcomes for children and teenagers, over DA-EPOCH alone. That said, children and teenagers with BL usually respond very well to treatment and have very good outcomes with high cure rates. New treatments in clinical trials therefore, are looking at how to minimise side effects and improve quality of life during and after treatment.

You will likely go into remission after treatment and have no signs of BL left. However, in rare cases the BL may not respond to treatment or may come back after a period of remission. If this happens to you, your doctor will offer you a different type of treatment with different medications or, may suggest you enroll in a clinical trial to get access to different treatments not otherwise available.

Find more information on treatment here.

Clinical Trials

Clinical trials are important because they help find new treatments. They can offer you the opportunity to try something new for free, that you would otherwise not have access to, or otherwise have to pay for. Some clinical trials may look at:

- how to treat BL
- safe doses to treat BL with less side-effects
- managing side-effects
- improving quality of life.

It is always worth asking your doctor what clinical trials you are eligible for, to see if any are of interest to you. You can also look online for clinical trials yourself at the ClinTrial Refer website. If you would like more information on clinical trials, please see our fact sheet Understanding Clinical Trials here.

Follow up

Finishing treatment can be a time of mixed emotions. You may feel relieved and excited, or you may feel worried and scared. You may even alternate between all of these emotions. This is very normal. However, you will not be alone. You will continue to see your specialist team on a regular basis, and be checked for any signs and symptoms of your lymphoma relapsing, and your doctor will also want to make sure you're not having any side effects from your treatment. Your doctor will let you know how often they want to see you, however the longer time you are in remission the less often they will need to see you.

If you have any concerns or worries please contact your healthcare team or contact our lymphoma care nurses on 1800 953 081. You can also email us on nurse@lymphoma.org.au.

Summary

- Burkitt lymphoma is an aggressive lymphoma that can affect children and adults.
- It usually responds well to treatment and can often be cured, or you may go into remission
- There are different subtypes of Burkitt Lymphoma but treatment is often the same.
- Some people with Burkitt lymphoma may have genetic mutations called either double or triple hit lymphoma.
- Report any concerns or B-symptoms or new or worsening symptms to your medical team.

Resources and Support

While many people look forward to finishing treatment, for some it can be a time of uncertainty and worry. It is normal to think about what happens next, to worry about if the cancer will come back, or to be frustrated if life does not go back to normal quickly enough.

There are a lot of resources and support to help you during this time. You can contact our lymphoma care nurses on 1800 953 081 or email nurse@lymphoma.org.au.

You can also find us on social media where you can connect with others who are living with, or who have beaten lymphoma.

Lymphoma Australia offers a wide range of resources and support for people living with lymphoma or CLL, and their carers. How to access our resources:

- **Visit** our website www.lymphoma.org.au for more information.
- **Phone** our Lymphoma Care Nurse Hotline on 1800 953 081.
- **Email** our Lymphoma Care Nurses nurse@lymphoma.org.au
- **Booklet:** Understanding Non-Hodgkin Lymphoma (NHL)
- **Downloadable information:** Visit our website, or give us a call if you would like some more information on a variety topics related to lymphoma
- **Join** our Facebook page Lymphoma Down Under (make sure you complete all the membership questions when you join).

Cancer Council offers a range of services, including free counselling, to support people affected by cancer, including patients, families and friends. Services may be different depending on where you live. You can contact them at www.cancer.org.au or by phone on 13 11 20.

Medicare Australia: Check with your GP if you are eligible for a Mental Health Treatment Plan (MHTP). This plan is funded by Medicare and can provide you with up to 10 sessions with a registered psychologist. More information can be found here.

WeCan is an Australian supportive care website to help find the information, resources and support services you may need following a diagnosis of cancer. You can visit their website at www.wecan.org.au.

Canteen provides support for young people aged 12-25 years who have cancer, or, who have a parent with cancer. Find out more at their website here www.canteen.org.au.

Health Translations: A collection of health related information collected by the Victorian Government with resources in different languages. You can visit their website at www.healthtranslations.vic.gov.au.

Useful links

Burkitt lymphoma webpage

SCAN ME

Questions to ask your doctor

SCAN ME

double/triple hit lymphoma

SCAN ME

Stem cell transplants

SCAN ME

CAR T-cell therapy

SCAN ME

Treatments

SCAN ME

eviQ treatment

SCAN ME

Health Translations

SCAN ME

Disclaimer: Lymphoma Australia has taken every precaution to make sure the information in this document is accurate and up-to-date. However, this information is intended for educational purposes only and does not substitute for medical advice. If you have any concerns about your health or wellbeing, please contact your treating team.

Notes

Notes