Cutaneous (Skin) Lymphomas

Lymphoma Australia Nurse
Support Line 1800 953 081
nurse@lymphoma.org.au

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.**

Overview

Cutaneous lymphoma is a type of cancer that often looks like patches, papules, plaques or tumours on your skin. However, unlike other skin cancers such as melanoma, basal cell carcinoma and squamous cell carcinoma that begin in skin cells, cutaneous lymphomas start in a type of blood cell called a lymphocyte, which travels to and lives in the layers of your skin.

We have two main types of lymphocytes called T-cell lymphocytes and B-cell lymphocytes. They are types of white blood cells that fight infection and disease. T-cells directly attack germs and cells that are developing abnormally, while B-cells make antibodies that attack the germs and damaged cells. T-cells also help other immune cells to work more effectively.

Cutaneous lymphoma can start or develop in your T-cell or B-cell lymphocytes. These subtypes may be indolent (slow developing) or aggressive (fast developing).

Indolent Cutaneous Lymphoma

Indolent cutaneous lymphomas may not need any treatment, as they do not usually spread to other parts of your body. You may live many years without even knowing you have lymphoma. However, if they become itchy or are causing you concern, treatment may be offered to manage symptoms.

Indolent lymphomas are typically long-term conditions that many people live with throughout their lives. While they are generally not considered curable, they grow slowly and are not immediately life-threatening. Many people with indolent cutaneous lymphoma continue to lead full, active, and meaningful lives.

In rare cases though, indolent lymphomas can begin to behave more aggressively and can "transform" into an aggressive type of lymphoma which will need treatment.

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Subtypes of Indolent Cutaneous T-cell Lymphoma (CTCL)

Mycosis Fungoides (MF)

Mycosis Fungoides (MF) is the most common subtype of CTCL, making up more than half of all people diagnosed with cutaneous lymphoma. It is usually indolent and may develop over many months or years. Uncommonly MF can change or transform to a more aggressive form called large cell transformation, and can spread to your lymph nodes or internal organs such as your lungs or liver.

Primary Cutaneous Anaplastic Large-Cell Lymphoma (pcALCL)

Primary cutaneous anaplastic large cell lymphoma (pcALCL) is a rare subtype of CTCL. It usually only affects your skin, but in very rare cases it may spread to your internal organs such as your lymph nodes, liver or spleen. You may only have one raised skin lesion or you may have more than one. It is indolent and should not be confused with another type of anaplastic large cell lymphoma that is aggressive. pcALCL can affect people of all ages including children, but is more common in those 45-60 years of age.

Lymphomatoid Papulosis (LyP)

Lymphomatoid Papulosis is considered pre-cancerous. If you have LyP, you have a slightly higher chance of developing CTCL than the general population. Some specialists may manage it in the same way they would manage an early stage CTCL. LP is more common in children and adults up to middle age.

Subcutaneous Panniculitis-like T-cell Lymphoma (SPTCL)

This is also a rare subtype of CTCL. If you have SPTCL you may have lumps in the fatty (subcutaneous) tissue just under your skin. SPTCL is more common in young adults, and in people with autoimmune diseases such as lupus erythematosus.

Subtypes of Indolent Cutaneous B-cell Lymphoma (CBCL)

Primary Cutaneous Follicle Centre Lymphoma

This is the most common subtype of CBCL. It is usually indolent and develops over months, or even years. It usually appears as bumpy reddish or brownish lesions or tumours on the skin of your head, neck, chest, or abdomen. This subtype usually affects people around the age of 60 years, and is more common in Western countries.

Primary Cutaneous Marginal Zone B-cell Lymphoma

This is the second most common subtype of CBCL. It is also indolent and can occur in people of all ages, including children, but is more common around the age of 55 years.

People who have had Lyme disease are at increased risk of Primary Cutaneous Marginal Zone B-cell Lymphoma. It also affects men twice as often as women.

EBV Positive Mucocutaneous Ulcers

This is a very rare subtype of CBCL found in patients who are immune compromised and have had Epstein-Barr virus (EBV)- the virus that causes glandular fever.

You likely will only have one ulcer on your skin, or in your gastrointestinal tract or mouth. Most people do not need treatment for this subtype of CBCL.

However, if you are taking immunosuppressive medications your doctor may review and decrease the dose to allow your immune system to recover a little. In rare cases you may need treatment with a monoclonal antibody such as Rituximab, or low-dose radiotherapy for persistent ulcers that do not improve or cause severe pain.

Aggressive Cutaneous Lymphoma

Aggressive Cutaneous Lymphomas will usually need treatment soon after you are diagnosed. Aggressive T-cell Cutaneous Lymphoma includes a subtype called Sezary Syndrome, while aggressive B-cell cutaneous lymphoma includes a subtype called Primary Cutaneous Diffuse Large B-cell Lymphoma. These aggressive lymphomas can spread to your lymph nodes, blood, bone marrow and other parts of your body.

Subtype of Aggressive Cutaneous T-cell Lymphoma (CTCL)

Sezary Syndrome (SS)

Sezary Syndrome (SS) is a rare subtype of CTCL that affects both your skin and blood, meaning that you will have cancerous T-cells in the layers of your skin causing a rash, but also have cancerous T-cells in your blood. Most people with SS will have a rash that covers most of their skin (about 80% of your body), which is usually very itchy. You will receive treatments with the aim to reduce the number of abnormal T-cells in your blood and skin, and to reduce your symptoms.

Subtype of Aggressive Cutaneous B-cell Lymphoma (CBCL)

Primary Cutaneous Diffuse Large B-cell Lymphoma

This is less common than other subtypes of CBCLs. It is more common in women than men and tends to be aggressive or fast growing. It can develop over weeks to months, and it usually affects older people around the age of 75 years. It often starts on your legs (leg type) as one or more lesions or tumours, but can also grow on your arms and torso (chest and abdomen). Some lesions can ulcerate and spread to other internal parts of your body.

Symptoms of Cutaneous Lymphoma

A rash is a common symptom of cutaneous lymphoma, but may appear similar to, or mimic, other skin conditions such as dermatitis, eczema, or psoriasis

The rash due to lymphoma may include:

- Patches, plaques, papules or tumours
- Erythroderma or redness over most of your body
- Thickened skin on hands, feet or over joints
- Fragile, cracked skin that may bleed
- Itchiness

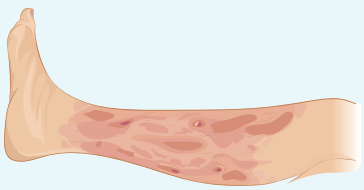
Patches - flat areas of skin that that may be smooth or scaly.

Papules - small, solid raised areas of skin, and may look like a hard pimple

Plaques - hardened areas of skin that are slightly raised, scaly and thickened.

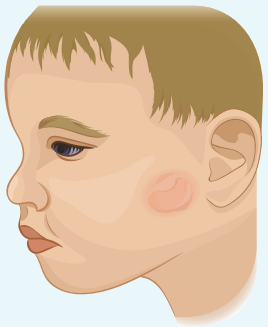
Tumours - raised bumps, lumps or nodules that can sometimes become sores that do not heal

DLBCL ON LEG

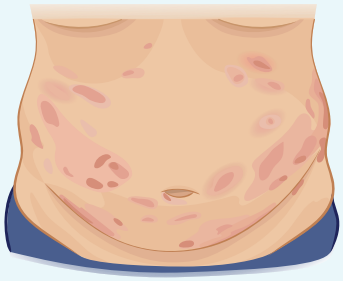


LUMP ON FOREHEAD

LUMP ON JAWLINE



ABDOMEN RASH



If your rash does not go away within two weeks, gets bigger, or spreads to other parts of your body, visit your doctor.

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Diagnosis and Staging of Cutaneous Lymphoma

Diagnosis

You will need a biopsy to check if your skin lesions or lumps are lymphoma. A biopsy is when your doctor removes a piece of the lesion so it can be checked in pathology. There are different ways the biopsy can be done, and the one you have will depend on your individual situation.

Your doctor will consider the size and location of your lesions and talk to you about which type of biopsy would give you the most accurate diagnosis.

The types of biopsies you may have include:

Skin biopsy of your rash or lesion, which is when a small sample of the affected skin is removed and sent to pathology to be checked.

Lymph node biopsies may be done in the following ways:

- Core needle incisional biopsy
- Excisional node biopsy

In some cases a fine needle biopsy may be used to take a sample of your lymph node, though this is not the preferred way to check for lymphoma.

Staging

If your biopsy results confirm you have lymphoma, you will need to have more tests to find out what subtype it is, and stage (or how far the disease has spread). These can include:

- Physical examination of the skin over your whole body - your doctor may ask your permission to take photos, to compare your progress later.
- Positron emission tomography (PET scan) - a scan of your whole body.
- Computed tomography (CT scan) – a 3D scan of certain parts of your body
- Blood tests.
- Bone marrow biopsy – this may be used if you have an aggressive subtype of cutaneous lymphoma.

TNM staging system

The staging of cutaneous lymphoma uses a system called TNM. If you have MF or SS there will be an extra letter added - TNMB.

T = size of **T**umour – or how much of your body is affected by the lymphoma.

N = lymph **N**odes involved – has the lymphoma gone to your lymph nodes, and how many lymph nodes have lymphoma in them.

M = **M**etastasis – checks if, and how far the lymphoma has spread inside your body.

B = **B**lood – is in your blood and bone marrow.

Staging of Cutaneous Lymphoma

	Cutaneous Lymphoma	Mycosis fungoides (MF) or Sezary Syndrome (SS) only
T Tumour or skin affected	**T1** - you only have one lesion. **T2** - you have more than one skin lesion but the lesions are in one area, or two areas close together on your body. **T3** - you have lesions across many areas of your body.	**T1** – less than 10% of your skin is affected. **T2** - more than 10 % of your skin is affected. **T3** - you have one or more tumours bigger than 1cm. **T4** - you have erythema (redness) covering more than 80% of your body.
N Lymph Nodes	**N0** – your lymph nodes appear normal. **N1** – one group of lymph nodes are involved. **N2** – two or more groups of lymph nodes are affected in your in your neck, above your clavicle, underarms, groin or knees. **N3** – lymph nodes in, or near your chest, lungs and airways, major blood vessels (aortic) or hips are involved.	**N0** – Your lymph nodes appear normal. **N1** – you have abnormal lymph nodes with low grade changes. **N2** – You have abnormal lymph nodes with high grade changes. **Nx** – you have abnormal lymph nodes, but the grade is not known.
M	**M0** – none of your lymph nodes are affected. **M1** – lymphoma has spread to your lymph nodes outside of your skin.	**M0** – none of your internal organs are involved, such as lungs, liver, kidneys, brain. **M1** – lymphoma has spread to one or more of your internal organs are involved.

	Cutaneous Lymphoma	Mycosis fungoides (MF) or Sezary Syndrome (SS) only
B Blood	**N/A**	**B0** – less than 5% (5 out of every 100) cancerous lymphocytes in your blood. These cancerous cells in your blood are called Sezary cells. **B1** – More than 5% of the lymphocytes in your blood are Sezary cells. **B2** – More than 1000 Sezary cells in a very small amount (1 microliter) of your blood.
Your doctor may use other letters such as "a" or "b" to further describe your lymphoma cells. These may refer to the size of your lymphoma, the way the cells look, and whether they have all come from one abnormal cell (clones) or more than one abnormal cell. Please ask your doctor to explain your individual stage, and what it means for your treatment.		

Treatment

Many people with indolent cutaneous lymphomas will not need treatment, however some will. If you need treatment, you may have it with a doctor who is a skin specialist (dermatologist), cancer or blood specialist (oncologist or haematologist), radiation oncologist or surgeon.

If you have an indolent lymphoma, your treatment will be given with the goal to lessen any symptoms you get and make you more comfortable.

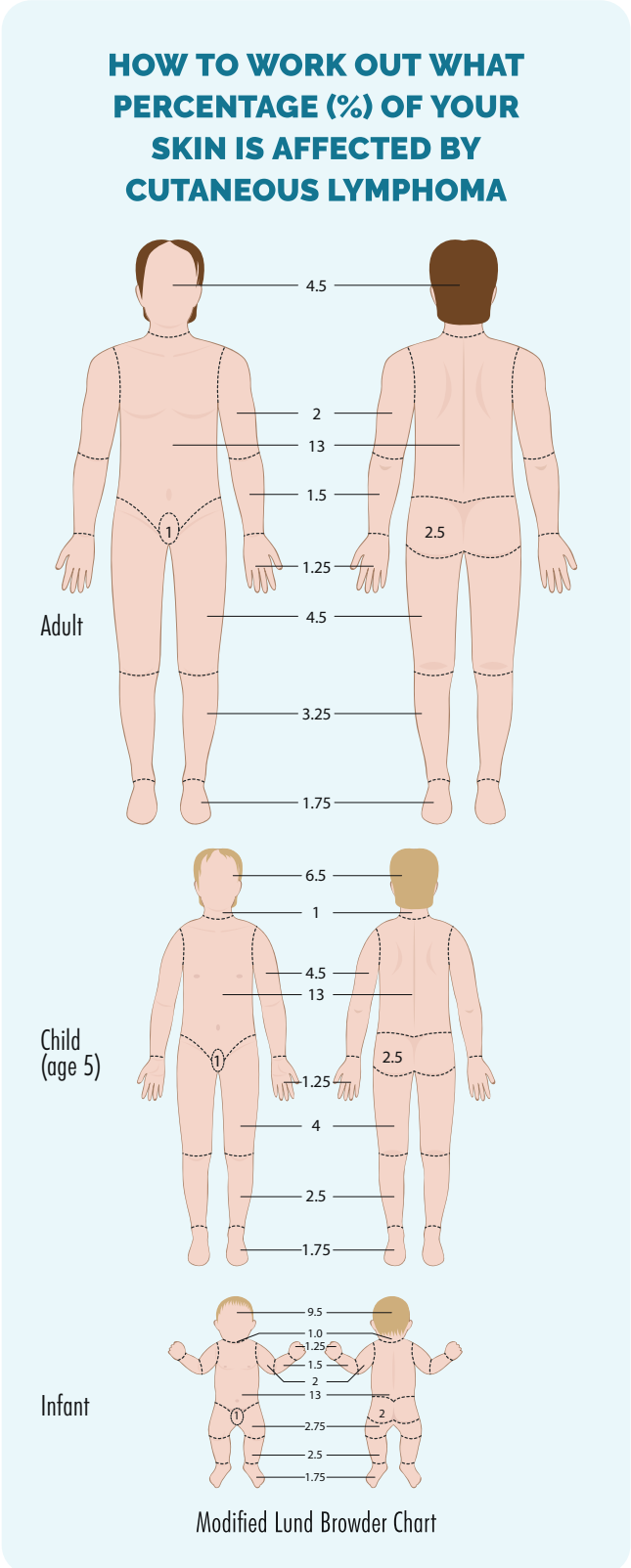
Local or Skin Directed Treatment

Topical treatments
Topical treatments are creams that you rub into the area of lymphoma. There are different types of creams that can be used, but can include some of the following:

Corticosteroids – are toxic for lymphoma cells and help to destroy them. They can also reduce inflammation and help improve symptoms such as itching.

Retinoids – are medications very similar to vitamin A. They can help reduce inflammation, and regulate the growth of cells on the skin. They are used less commonly, but are helpful in some particular types of skin lymphoma.

Topical Chemotherapy – gels such as Ledaga (containing chlormethine) which attach to the DNA in the abnormal cells to stop them from growing and destroy the lymphoma.



Surgery

In some cases you may have surgery, either under a local or general anaesthetic, to remove the whole area of skin affected by lymphoma. This is more likely if you have a single lesion or several smaller lesions. It is more commonly used as part of the process to diagnose your lymphoma, rather than as a treatment.

Phototherapy

Phototherapy is a type of treatment that uses specialised lights (often UV) on areas of your skin affected by lymphoma. The UV interferes with the cells' growing process. By damaging the growing process, the lymphoma is destroyed.

Radiotherapy

The X-rays cause damage to the cell's DNA (the genetic material of the cell) which makes it impossible for the lymphoma to repair itself. This causes the cell to die. It usually takes a few days or even weeks after radiation treatment begins for the cells to die. This effect can last for several months, meaning that cancerous lymphoma cells in the treated area/s can continue to be destroyed months after treatment has finished.

Systemic Treatment

Systemic treatment is used to treat more areas of your body, so is more likely to be used if your lymphoma is widespread across your skin, or has spread to other parts inside your body such as your blood, bone marrow, lymph nodes or organs. It may include tablets or medications given through a drip into your vein.

Chemotherapy

Chemotherapy is a type of treatment that directly attacks fast growing cells, so it can be effective at destroying fast growing lymphomas. But it cannot tell the difference between healthy and cancerous fast-growing cells, so it can cause some unwanted side-effects such as hair loss, nausea and vomiting, or diarrhea or constipation.

Immunotherapies

Immunotherapies can help your immune system find and fight the lymphoma more effectively. They can do this in several ways. Monoclonal antibodies, attach to the lymphoma to help your immune system "see" the lymphoma to recognise and destroy it. Some immunotherapies are also able to affect the structure of the lymphoma cell wall, causing them to die. Examples of immunotherapies used in skin lymphomas are Mogamulizumab and Brentuximab Vedotin.

Others, such as interleukins and interferons, are special proteins that naturally occur in our bodies but can also be taken as medicine. They work by boosting your immune system, helping it to wake up other immune cells, and by telling your body to make more immune cells to fight the lymphoma.

Targeted therapies

Targeted therapies are medications made that target something specific to the lymphoma cell, so they often have fewer side effects than other treatments. These medications work by interrupting the signals the lymphoma cells need to survive. When they do not get these signals, the lymphoma cells stop growing because they do not get the nutrients they need to survive. Examples of these are Vorinostat and Romidepsin.

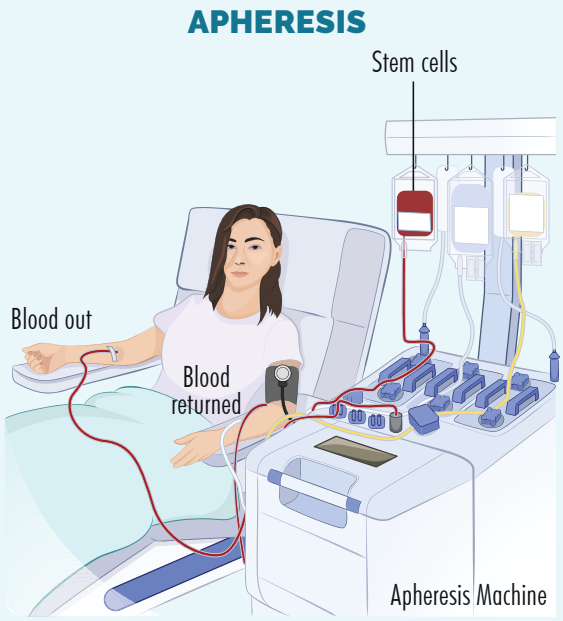
Stem cell transplant

Stem cell transplants are only used if your lymphoma does not respond to other treatments (is refractory), or comes back after a time of remission (relapses). It is a multi-step treatment where your own, or a donor's stem cells (very immature blood cells) are removed through a procedure called apheresis, and then given to you at a later time, after you have had high dose chemotherapy.

With cutaneous lymphoma, it is more common for you to receive stem cells from a donor rather than your own. This type of stem cell transplant is called an Allogeneic Stem Cell transplant.

Extracorporeal photopheresis (ECP)

Extracorporeal photopheresis is a treatment used predominantly for advanced MF and SS. ECP is also performed using an apheresis machine, Your white blood cells are removed and mixed with a medicine called psoralen.



Step 1: Blood is removed from an IV line in your arm, or from your central line.

Step 2: Blood is processed in the apheresis machine and the stem cells are separated and removed from your blood.

Step 3: Blood is returned to you (minus the targeted cells).

This makes them highly sensitive to ultraviolet (UV) light. The white blood cells are then treated with UV light inside the machine, and returned to you. Your immune cells are then more reactive to the lymphoma and cause the lymphoma cells to be killed.

Supportive care

Proper skincare is vital for those with cutaneous lymphoma. This can help provide significant relief from irritated skin, and prevent skin infections.

Some general strategies to help care for your skin include:

- Wearing loose-fitting cotton or cotton-blend clothing. Avoid irritating fabrics like wool and synthetics.
- When washing clothes, it is important to use unscented laundry detergents. Avoid harsh fabric softeners, dryer sheets, and starch.
- Standard soaps are alkaline and strip the skin's protective lipids. Use pH-balanced bars, and fragrance-free, gentle cleansers.
- Limit showers and baths to just 5 minutes in lukewarm (not hot) water to prevent your skin from drying out.
- Apply a moisturiser immediately after bathing while your skin is still damp - this will help seal in the moisture.

Specific products and care when you have cutaneous lymphoma is highly individualised. Speak to your medical team about which types of supportive skin care would best suit your needs.

Follow up

Some cutaneous lymphomas are lymphomas you will live with for the rest of your life, and you may need treatment at different times throughout your lifetime to keep it under control. In between treatments you will still have regular check-ups with your specialist. The time in between treatments, or when you finish 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.

Your doctor will also want to make sure you're not having any side effects from your treatment. The longer you are in remission, the less often you will need to be seen, but ask your doctor how often you will need to see them. If you have any concerns or worries please contact your healthcare team.

Summary

Cutaneous lymphoma is a subtype of Non-Hodgkin Lymphoma resulting from cancerous blood cells called lymphocytes, travelling to and living in the layers of your skin.

Indolent Cutaneous Lymphomas may not need treatment, however this may be recommended to manage symptoms if they make you uncomfortable.

Aggressive Cutaneous Lymphomas need treatment soon after you are diagnosed.

There are several different specialist doctors that may manage your care, and this will depend on your individual circumstances.

If your lymphoma is affecting your mental health or mood you can ask your doctor for a referral to a psychologist to help you cope.

Resources and Support

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 for more information.
- **Phone** our Lymphoma Care Nurse Support Line on 1800 953 081.
- **Email** our Lymphoma Care Nurses nurse@lymphoma.org.au
- **Visit** our Factsheets and Booklets page or give us a call if you would like to download more information on a variety topics related to lymphoma.
- **Join** our closed Facebook page Lymphoma Down Under (make sure you complete all the membership questions when you join).

Cutaneous Lymphoma Foundation offers a range of resources and support people affected by cutaneous lymphoma. Visit their website here: www.clfoundation.org.

Cancer Council offers a range of services 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)

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: www.wecan.org.au.

Canteen provides support for young people aged 12-25 years who have cancer, or, who have a parent with cancer. Visit 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. visit their website here: www.healthtranslations.vic.gov.au.

Useful links

Types of Lymphoma: Cutaneous (skin) Lymphoma

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Symptoms of Lymphoma

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Questions to Ask Your Doctor

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Health Translations

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Definitions

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Stem Cell Transplants

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