Cutaneous T-cell lymphoma

Your guide to best cancer care

About this guide

Being told you have or could have cutaneous T-cell lymphoma (CTCL) can be overwhelming, especially if there has been a long journey to diagnosis. The two most common types of CTCL include mycosis fungoides and Sézary syndrome. A lot can happen quickly, and you might have many questions. CTCL is often a slow progressing disease and generally has an excellent prognosis – especially if identified in the early stages. This resource can help to guide you, your family, and friends through this experience.

Other international resources are available online but may discuss approaches to testing and treatment that are not standard of care in Australia.

Information and support

For more information about CTCL, visit the Leukaemia Foundation website **www.leukaemia.org.au/blood-cancer/lymphoma/non-hodgkin-lymphoma/skin-lymphomas/**.

Cancer Council: for information and support, call **13 11 20** to talk to an experienced healthcare professional or visit **www.cancer.org.au**.

Leukaemia Foundation: To speak with an experienced healthcare professional who can provide you with disease-specific information, answer your questions, talk through your concerns, and connect you to blood cancer support groups, call **1800 620 420** or visit **www.leukaemia.org.au**. The team can also help with practical concerns such as accommodation close to treatment, transport to appointments, and financial assistance.

Lymphoma Australia: Lymphoma Australia nurses can help patients and their families understand their diagnosis and treatment options, navigate the lymphoma journey, and connect patients with others and appropriate support networks. Call **1800 953 081** or visit **www.lymphoma.org.au** for education, support, and awareness for lymphoma and CTCL.

More information is available in the 'Resources' section of the optimal care pathway for cutaneous T-cell lymphoma. **www.cancer.org.au/OCP**.

Information for carers: Information, resources and support for carers and loved ones are available on the Leukaemia Foundation website **www.leukaemia.org.au/blood-cancer/carers-and-friends/** and on the Cancer Council website **www.cancercouncil.com.au/cancer-information/carers/**.

Translating and Interpreting Service (TIS): If you need a translator, call TIS on **13 14 50** or visit **www.tisnational.gov.au**.

Initial tests and referrals

Symptoms

Your general practitioner (GP), other primary care physician, or dermatologist will do a check-up to see if they can find out what is making you unwell. They will ask you about any symptoms you might be having such as an itch, pain and/or burning sensation on the skin, skin conditions that may present similar to dermatitis, psoriasis, tinea or fungal skin infections, fever, night sweats, and weight loss. Symptoms limited to skin may be the only signs of CTCL. You may feel anxious or concerned after the diagnosis is made, but these concerns often ease when you are reviewed by a specialist or specialist team.

Australian Government Cancer Australia
Leukaemia Foundation

Initial tests you may have

Physical exam. Specifically examining the skin and lymph nodes paying particular focus on the underarms, buttocks, thighs, and sides of breasts.

Skin biopsy can be done by a punch or incisional biopsy. For a punch biopsy, a round tipped tool is used to remove a sample of skin and its deeper layers for testing. An incisional biopsy will make a small cut (incision) to remove part of a skin lesion to be sent for testing. These procedures are usually performed under local anaesthetic, to lessen pain.

Blood tests. A sample of your blood is collected and sent for a full blood examination and may include a liver function test.

Referrals

If your GP thinks you may have CTCL, they'll refer you to a haematologist or dermatologist at a public hospital or in private practice for more tests. Haematologists are highly trained in conditions that affect blood and bone marrow. You may also be referred to a radiation oncologist (a specialist medical doctor specialising in the use of radiation therapy) if radiotherapy is included in your treatment plan.

It can help to have a family member, carer, or friend attend appointments with you.

Let your doctor know what is most important to you in a specialist (e.g., that they specialise in CTCL, cost, location, bedside manner, expertise). It is important that the specialist you see is familiar with and has expertise in CTCL.

Timeframes

The timing of your specialist appointment will depend on your initial results of tests performed by your GP or doctor. For a small number of patients, an urgent appointment or hospital admission may be required, but for most people with no urgency, an appointment **within four weeks** is appropriate.

Questions you might want to ask your GP

- Are there any symptoms that I should watch out for while I am waiting for my appointment to see a specialist?
- Can I choose the specialist I see?
- What emotional and mental health support services are available and how do I access them?
- Can I choose whether I go to a public or private hospital?
- What are the differences of being treated in the public versus private system?

It's a good idea to keep a written diary or digital record of treatment details and appointments with your GP and specialists. You can ask permission to record your appointments. It's also helpful to bring a list of questions and ask for a written treatment plan.

Don't ignore new signs and symptoms. Alert your GP or specialist. Trust yourself. It's OKAY to be persistent.

Travel to access tests, treatment and care

You will likely have to travel to see a specialist, undergo tests, or access treatment, especially if you live in a rural or regional area. In some cases, you may have to leave home for an extended period of time. A treatment plan can be set at a major hospital with specialists in CTCL, with much of the plan able to be carried out in your home city or state/territory. Accommodation and transportation support services are available, and it is encouraged to have family support with you whilst you undergo treatment. Ask your doctor about travel subsidy schemes available in your state/territory, more information is available on the Leukaemia Foundation website **www.leukaemia.org.au/how-we-can-help/transport-services/** and on the Cancer Council website **www.cancer.org.au/support-and-services/transport-to-treatment**.

Mental health and emotional wellbeing

A blood cancer diagnosis can affect your mental and emotional wellbeing. Patient support organisations, your GP, or another health professional such as a psychologist or social worker can help you develop strategies to cope. They can help you access a mental health treatment plan if required. There may also be community support groups available in your state/territory for CTCL or non-Hodgkin lymphomas.

Diagnosis, staging and treatment planning

The haematologist/specialist will do more tests to see if you have CTCL, to check how advanced it is, and to help determine the best treatment for you. It is important to note that staging for CTCL, especially in Sézary Syndrome, can fluctuate greatly overtime. The staging of CTCL does not directly link to that of other cancers and interpretation should rely on specialist advice.

You may have one or a combination of these tests:

- **Review of previous skin biopsies** by specialists in the diagnosis of CTCL.
- **Blood tests.** Full blood examination and other tests to determine if markers of CTCL are present in the blood.
- **Imaging tests** such as CT scans of the chest, abdomen, and pelvis to check lymph nodes and other imaging tests including PET scans may occur depending on stage of disease.
- **Skin biopsy** for further examination of the rash by specialists.
- **Lymph node biopsy.** A doctor will put a thin needle into your lymph node to remove a small sample of cells which will then be sent to a laboratory for examination.
- **Medical photography** for skin mapping.
- **Bone marrow biopsy.** This may be part of the diagnostic work-up depending on the stage of your disease. This procedure can be used to assess the extent of lymphoma spread or can be helpful in differentiating CTCL from other lymphomas. A doctor will put a needle into your hip bone to take a sample of your bone marrow. The sample of bone marrow tissue will be sent to a laboratory for examination. This is usually done with local anaesthetic and mild sedation or inhaled pain relief.

Timeframes

You should have most results back **within eight weeks**.

Questions you might want to ask

- What is CTCL and where can I find more information about it?
- What tests will I need and why?
- Will my follow-up appointments be face-to-face or by tele/video health or a combination?
- What's my prognosis?
- How much will appointments and tests cost and how much will I have to pay myself?
- Can I choose where I have treatment?
- Will I need to start treatment straight away?
- How long will the treatment last?
- How will my disease be monitored?
- Is there information that I can share with my family or friends?
- What support services are available to me?

Treatment

Your specialist will talk to you about your treatment options for CTCL. You may need more than one treatment type to get the best results, and you may require ongoing treatment to manage your CTCL for the rest of your life.

You may have one or a combination of these treatments:

- **Corticosteroids.** Drugs that help change your immune response by reducing inflammation and are applied directly onto skin patches and plaques.
- **Light therapy.** UV light is used to decrease inflammation in the skin.
- **Radiotherapy** uses localised x-rays to kill cancer cells and stop the cancer growing.
- **Total skin electron therapy** treats the skin surface and reduces patches and plaques caused by CTCL.
- **Extracorporeal photopheresis.** A procedure in which blood is drawn from a vein and the white blood cells in the sample are treated with UV light (PUVA phototherapy), and then re-infused into the person's vein. The UV light treatment helps to target the cancerous T cells and triggers the immune system to destroy the cells. This is most effective for specific types of more advanced CTCL.
- **Interferon.** Drugs that stimulate the immune system to specifically suppress the CTCL cells. This is given as an injection under the skin.

- **Systemic chemotherapy.** Drugs that are used to kill cancer cells to stop the cancer growing. Generally reserved for treatment in refractory or rapidly progressive and advanced CTCL. Chemotherapy for CTCL can be delivered intravenously (through IV) or orally in pill form. Some subtypes of CTCL are more likely to respond to chemotherapy than others.

Other more novel or directed treatments may be appropriate for you in discussion with your treating team. Clinical trials may also be considered during your treatment journey.

For more information visit **www.cancer.org.au/cancer-information/treatment**.

Supportive care (treatment or services that support you through a cancer experience) are also available, often delivered through a multidisciplinary team that may include nurses, pharmacists, psychologists, physiotherapists, and dietitians.

Timeframes

By evaluating your symptoms and test results, your specialist will determine when your treatment should start. For most people past early-stage disease, radiation therapy is rarely urgent and can be commenced **within two to four weeks** of the decision being made. There may be instances where your treatment could be delayed beyond this timeframe. If this occurs your specialist will discuss this with you.

Clinical trials

Your specialist may recommend participating in a clinical trial (or you can ask if you are eligible for any clinical trials). Clinical trials often provide access to promising new treatments that are not yet available to the general public. Many people with cancer are now living longer, with a better quality of life, because of clinical trials.

For more information visit **www.australiancancertrials.gov.au** or **www.anzchog.org/clinical-trials-research/clinical-trial-initiatives/**, or search the Australian New Zealand Clinical Trials Register **www.anzctr.org.au**.

Complementary therapies and other medications

Speak to your healthcare team about any dietary supplements, vitamins, herbal remedies, and other medications that you use or would like to use, including prescription and over-the-counter medicines (e.g., paracetamol or ibuprofen). Some medicines, complementary therapies, vitamins, and foods might not work well with your treatment.

Fertility and sexual health

Blood cancer and blood cancer treatment may cause fertility problems for both men and women. This will depend on age, the type of blood cancer, and the treatment received. If this is relevant, get advice from your treating team about contraception before, during, and after treatment. Discuss having children whilst undergoing treatment and the need for referral to a fertility specialist and options for fertility preservation before starting treatment. Diagnosis and treatment may affect your sex life or overall sexual health. For support and, if necessary, referral to counselling services, contact the Leukaemia Foundation on **1800 620 420** or visit **www.leukaemia.org.au** and type 'relationships and sex' in the search bar.

You can ask for more time to think about your treatment options.

You can ask your GP for a referral to another specialist for a second opinion.

Questions you might want to ask

- What treatment do you recommend and why?
- Are there alternatives?
- What will happen if I don't have treatment?
- How long will I need to be on treatment?
- What will treatment cost and how much of the cost will I have to pay myself?
- What activities/exercise will help me during and after treatment?
- Is there any specific diet I should follow or foods or drink to avoid during treatment?
- How will the treatment affect my day-to-day life, and can I still work?
- Who is in my treatment team and who is the main contact person if I have any questions or feel unwell?
- What side effects could I have from treatment and how should I manage them?
- Will treatment affect my ability to have children, and if yes, what options should I consider?

Decisions about cost

You may have to pay for some appointments, tests, medications, accommodation, travel, or parking.

Speak with your GP, specialist, or private health insurer (if you have one) to understand what is covered and what your out-of-pocket costs may be.

If you have concerns about costs, talk to your healthcare team or a social worker about:

- being bulk-billed or being treated in the public system
- help with accommodation and/or transport during treatment
- the possible financial impact of your treatment and financial support schemes you may be able to access
- how your treatment might impact your ability to work.

For more information about financial and practical support call the Cancer Council on **13 11 20** or visit **www.cancer.org.au/support-and-services/practical-and-financial-assistance**, or contact the Leukaemia Foundation on **1800 620 420** or visit **www.leukaemia.org.au/how-we-can-help/practical-support-services/**.

Care during treatment

Lifelong treatment for your CTCL may be required, and this can affect your physical, psychological, nutritional, and general wellbeing. Due to the skin complications experienced with CTCL, such as patches, plaques, and broken skin, the risk for infection and sepsis is high and should be managed with your treating doctor. Patients are urged to follow their prescribed skincare plan as intact skin provides protection from infection and sepsis. Comprehensive support and side effect management should be part of your routine ongoing care.

A number of specific needs may arise for patients at this time:

- general skin care including regular use of bland or non-perfumed moisturisers and/or topical steroids
- measures to reduce skin bacterial load such as bleach baths
- clothing that does not irritate the skin
- assistance to cope with the emotional distress and/or anger of dealing with a potential cancer diagnosis, anxiety/depression, interpersonal problems, and adjustment difficulties
- management of physical symptoms including fatigue and higher likelihood of infection
- encouragement and support to increase levels of exercise.

Ongoing care plan

Current therapies mean that most people with CTCL will go on to live long lives. You may need regular ongoing follow-up appointments and care, so your specialist and healthcare team will work with you to make a care plan for you and your GP. Your care plan may need to be updated over time if your needs change. This plan will explain:

- who your main contact person is after treatment
- how often you should have check-ups and what tests this will include
- understanding and dealing with potential side effects of treatment
- how to get help quickly if you have an urgent problem
- how to look after your overall health and wellbeing
- what healthcare and community support services are available to you and how to access them.

Your specialist and healthcare team will talk to you about your needs and can refer you to other health professionals and community support services. Other information you get might be about:

- the side effects of treatment and the specialists you may need to see
- how to make healthy lifestyle choices to give you the best chance of staying well.

For more information visit **www.cancer.org.au/cancer-information/after-a-diagnosis/after-cancer-treatment**.

Questions you might want to ask

- Who should I contact if I'm feeling unwell?
- What can I do to be as healthy as possible?
- Where can I get more help?

Living with relapsed or progressive disease

Most patients with CTCL will have a recurrence of their disease at some stage in their treatment journey, although the timeframe at which this happens is uncertain. While relapse or disease progression is common with CTCL, this does not always mean that the disease is behaving more aggressively or is likely to be life-limiting. Access to the best available therapies, including clinical trials and treatment with a multidisciplinary team, are crucial to achieving the best outcomes for recurrence or progression of the disease.

Treatment will depend on the features presented in the recurrence of the disease, previous treatment, and the patient's preferences.

In managing people with CTCL, treatment may include:

- biological therapies
- immunomodulators
- monoclonal antibodies
- chemotherapy
- stem cell transplant
- participation in clinical trials.

Questions you might want to ask

- What is the phase of my disease?
- What are my treatment options?
- What are the chances that the treatment will work this time?
- Can I go back to a previous treatment?
- Is there a clinical trial available?
- What financial, practical, or emotional support is available?

Advance care planning

Your GP or healthcare team may talk with you, your family, and carer about future treatment and medical needs.

Advance care directive

Sometimes known as a living will, an advance care directive is a legally binding document that you prepare to let your family and medical team know about the treatment and care you might want or not want in case you become too unwell to make those decisions yourself. For more information visit **www.advancecareplanning.org.au**.

Symptom and palliative care management

Palliative care can help you to live as well as you can including managing pain and symptoms, psychosocial support, and assistance in identifying care goals. This care may be at home, in a hospital, or at another location you choose. Your specialist may refer you to palliative care services, but this doesn't always mean end-of-life care. Today people can be referred to these services much earlier if they're living with blood cancer or if their blood cancer returns. Speak to your GP or haematologist/specialist or visit **www.palliativecare.org.au**.

Making treatment decisions

You may decide not to have treatment at all, or to only have some treatment to reduce pain and discomfort. You can always change your mind about your treatment. Just talk to your specialist. For more information visit **www.cancer.org.au/cancer-information/treatment/advanced-cancer-treatment**.

Questions you might want to ask

- What can be done to reduce my symptoms?
- What extra support can I get if my family and friends care for me at home?
- Can you help me to talk to my family about what is happening?
- What support is available for my family or carer?
- Can I be referred to a community support service?
- What support services are available to me to help manage the ongoing nature of my blood cancer?

Published in May 2024.

This resource is based on information from the optimal care pathway for people with cutaneous T-cell lymphoma (1st edition), available at **www.cancer.org.au/OCP**.