Optimal care pathway for people with cutaneous T-Cell lymphoma (CTCL)

Quick reference guide

Support: Assess supportive care needs at every step of the pathway and refer to appropriate health professionals or organisations.

The optimal care pathways describe the standard of care that should be available to all cancer patients treated in Australia. The pathways support patients and carers, health systems, health professionals and services, and encourage consistent optimal treatment and supportive care at each stage of a patient's journey. Seven key principles underpin the guidance provided in the pathways: patient-centred care; safe and quality care; multidisciplinary care; supportive care; care coordination; communication; and research and clinical trials. This quick reference guide provides a summary for clinicians of the *Optimal care pathway for people with cutaneous T-cell lymphoma.*

Please note that not all patients will follow every step of the pathway.

Step 1: Prevention and early detection

Prevention

At present, the causes of most CTCL cases are not fully understood, and there are currently no clear prevention strategies.

Risk factors

The risk factors for developing CTCL include:

- older age
- sex (CTCL is more common in males than females)
- immune status (immunocompromised patients are at a higher risk of developing CTCL)
- environmental exposure.

Early detection

CTCL is often challenging to diagnose, particularly in the early or erythrodermic stages. In many cases, it can take multiple skin biopsies over a period of years to establish a definitive diagnosis.

In the early stages of MF (the patch of plaque stage or stage IA-IIA), it may be difficult to distinguish the disease from benign inflammatory skin conditions, as neoplastic cells can constitute a minority in the infiltration of the skin, and clinical and histological findings may overlap.

CTCL may be similar to, or have features that overlap with, other skin conditions. Diagnosis is reliant on careful clinical pathological correlations (i.e., the integration of patient's symptoms and signs with findings on skin biopsy, coupled with other diagnostic testing). Specific histopathology is required prior to diagnosis in order to develop a holistic view.

Screening recommendations

Routine screening for CTCL is not currently recommended in either the general population or in relatives of people with CTCL.

General health checklist

- ❑ Recent weight changes discussed and the patient's weight recorded
- ❑ Alcohol intake and smoking status discussed and support offered if appropriate
- ❑ Physical activity recorded
- ❑ Referral to a dietitian considered

Step 2: Presentation, initial investigations and referral

The following signs and symptoms should be investigated:

- itch, pain, and/or a burning sensation on the skin
- skin changes or rash (commonly occurring in sun-protected areas), which may include redness, changes in pigmentation, scaly patches, thick lesions (plaques), or bumps (tumours) covering a variable amount of skin
- enlarged lymph glands (groin, underarms, and/or neck).

It should be noted that skin-limited signs may be the only disease manifestations.

Sepsis stemming from altered skin integrity and bacteremia may be a mode of presentation in some patients.

Initial investigations by the GP should include a complete physical examination with a particular focus on underarms, buttocks, thighs, and the sides of breasts, specifically examining the skin and lymph nodes, and:

- skin biopsy or biopsies (punch or incisional biopsy)
- blood tests (FBE, blood film, UEC, LFT, LDH, calcium).

Checklist

- ❑ Multiple skin biopsies completed. CTCL is often challenging to diagnose, and in many cases, it can take multiple skin biopsies over a period of years to establish a definitive diagnosis
- ❑ Signs and symptoms recorded
- ❑ Patient notified of support services such as Cancer Council 13 11 20, Leukaemia Foundation 1800 620 420

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Step 2: Presentation, initial investigations and referral continued

When an itchy patch or rash on a patient's skin does not improve and continues to increase in size, this should prompt the GP to send the patient to a dermatologist for further investigation.

Referral options

At the referral stage, the patient's GP or other referring doctor should advise the patient about their options for referral, waiting periods, expertise, potential out-of-pocket costs and the range of services available. This will enable patients to make an informed choice of specialist and health service.

Communication

The GP's responsibilities include:

- explaining to the patient and/or carer who they are being referred to and why
- supporting the patient and/or carer while waiting for specialist appointments
- informing the patient and/or carer that they can contact Cancer Council 13 11 20 and Leukaemia Foundation 1800 620 420.

Checklist continued

- ❑ Referral options discussed with the patient and/or carer including cost implications.

Timeframe

Ideal timeframes depend on the presumptive diagnosis and/or disease kinetics. All initial investigations should be completed **within 1 month** of presentation. If blood test results are significantly abnormal, prompt referral to a haematologist is advised.

More advanced stages of disease should be seen **within 1 month** (e.g., presumed Sézary syndrome or MF with tumour development).

Step 3: Diagnosis, staging and treatment planning

Diagnosis and staging should include:

- a review of historical biopsies by anatomical pathologists who hold expertise in the diagnosis of CTCL.

Tests that are always indicated include:

- a routine evaluation:
  - medical examination including estimation of skin tumour burden using a modified severity-weighted assessment tool (mSWAT: https://www.clfoundation.org/modified-severity-weighted-assessment-tool-mswat), measuring the total body surface area (BSA) by using the patient's palm and fingers to represent 1% BSA
  - blood tests including assessment for blood involvement by immunophenotyping (refer to OCP Step 3 for details)
  - skin biopsy (for histology, immunophenotyping, and T cell receptor gene rearrangement studies or specific gene panel testing performed at a CTCL referral centre)
- medical photography:
  - specialist medical photography for skin mapping at baseline and throughout treatment for monitoring disease progression and response to treatment.

Tests that may be required based on clinical scenario include:

- PET/CT or CT of chest/abdomen/pelvis, assessing for lymph node enlargement
- lymph node biopsy.

Genetic testing

Currently there are no genetic tests applicable to predict family risk of CTCL.

Treatment planning

The multidisciplinary team should discuss patients with CTCL before starting any disease-directed therapy.

Research and clinical trials

Consider enrolment where available and appropriate. See the OCP resources appendix and relevant steps for clinical trial resources relevant to CTCL.

Checklist

- ❑ Diagnosis has been confirmed
- ❑ Performance status and comorbidities measured and recorded
- ❑ Patient discussed at multidisciplinary meetings and decisions provided to the patient and/or carer
- ❑ Clinical trial considered
- ❑ Supportive care needs assessed and referrals to allied health services actioned as required
- ❑ Referral to support services (such as Cancer Council, Leukaemia Foundation, Lymphoma Australia)
- ❑ Treatment costs discussed with the patient and/or carer

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Step 3: Diagnosis, staging and treatment planning continued

Communication

The lead clinician's[1] responsibilities include:

- discussing a timeframe for diagnosis and treatment options with the patient and/or carer
- explaining the role of the multidisciplinary team in treatment planning and ongoing care
- encouraging discussion about the diagnosis, prognosis, advance care planning and palliative care while clarifying the patient's wishes, needs, beliefs and expectations, and their ability to comprehend the communication
- providing appropriate information and referral to support services as required
- communicating with the patient's GP about the diagnosis, treatment plan and recommendations from multidisciplinary meetings.

Timeframe

Investigations should be completed **within 8 weeks**. The timeframe is dependent on results of existing tests and clinical urgency based on results of baseline investigations.

Step 4: Treatment

Establish intent of treatment

- curative
- anti-cancer therapy to improve quality of life and/or longevity without expectation of cure
- symptom palliation.

The aims of treatment of CTCL are improvement in symptom control, quality of life and cosmesis.

Early-stage disease

Early-stage MF has a favourable prognosis, and those patients who are asymptomatic have a 10% risk of progression within 10 years with a near-normal life expectancy. For these patients, skin-directed therapies are the main treatment, including corticosteroids, light therapy (including phototherapy), localised radiotherapy, and total skin electron therapy.

Advanced stage disease

For patients with advanced-stage disease (Stage IIB or greater) or patient's refractory to skin-directed therapies, a combination of systemic and skin-directed therapies are recommended. Typical systemic therapies include methotrexate, pegylated interferon, retinoids, histone deacetylase inhibitors (i.e. vorinostat or romidepsin), extracorporeal photopheresis, and Brentuximab vedotin.

Chemotherapy is generally reserved for treatment in refractory or rapidly progressive advanced mycosis fungoides.

Palliative care

Early referral to palliative care can improve quality of life and in some cases survival. Referral should be based on need, not prognosis. For more information, visit the Palliative Care Australia website <www.palliativecare.org.au>.

Communication

The lead clinician and team's responsibilities include:

- discussing treatment options with the patient and/or carer including the intent of treatment as well as risks and benefits
- discussing advance care planning with the patient and/or carer where appropriate
- communicating the treatment plan to the patient's GP
- helping patients to find appropriate support for exercise programs where appropriate to improve treatment outcomes.

Checklist

- ❑ Intent, risk and benefits of treatment discussed with the patient and/or carer
- ❑ Treatment plan discussed with the patient and/or carer and provided to GP
- ❑ Supportive care needs assessed and referrals to allied health services actioned as required
- ❑ Early referral to palliative care considered and advance care planning discussed with the patient and/or carer

1 Lead clinician – the clinician who is responsible for managing patient care.
The lead clinician may change over time depending on the stage of the care pathway and where care is being provided.

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Step 5: Care after initial treatment and recovery

Provide a treatment and follow-up summary to the patient, carer and GP outlining:

- the diagnosis, including tests performed and results
- treatment received (types and date)
- current toxicities (severity, management and expected outcomes)
- interventions and treatment plans from other health professionals
- potential long-term and late effects of treatment and care of these
- supportive care services provided
- a follow-up schedule, including tests required and timing
- contact information for key healthcare providers who can offer support for lifestyle modification
- a process for rapid re-entry to medical services for any issues arising.

Communication

The lead clinician's responsibilities include:

- explaining the treatment summary and follow-up care plan to the patient and/or carer
- informing the patient and/or carer about secondary prevention and healthy living
- discussing the follow-up care plan with the patient's GP.

Checklist

- ❑ Treatment and follow-up summary provided to the patient and/or carer and the patient's GP
- ❑ Supportive care needs assessed and referrals to allied health services actioned as required
- ❑ Patient-reported outcome measures recorded

Step 6: Managing relapsed or progressive disease

Detection

Most relapsed or progressive disease will be detected via routine follow-up or by the patient presenting with symptoms.

Treatment

Evaluate each patient for whether referral to the original multidisciplinary team is appropriate. Treatment will depend on the features of disease, previous management and the patient's preferences.

Advance care planning

Advance care planning is important for all patients but especially those with advanced disease. It allows them to plan for their future health and personal care by thinking about their values and preferences. This can guide future treatment if the patient is unable to speak for themselves.

Survivorship and palliative care

Survivorship and palliative care should be addressed and offered early. Early referral to palliative care can improve quality of life and in some cases survival. Referral should be based on need, not prognosis.

Communication

The lead clinician and team's responsibilities include:

- explaining the treatment intent, likely outcomes and side effects to the patient and/or carer and the patient's GP.

Checklist

- ❑ Treatment intent, likely outcomes and side effects explained to the patient and/or carer and the patient's GP
- ❑ Supportive care needs assessed and referrals to allied health services actioned as required
- ❑ Advance care planning discussed with the patient and/or carer
- ❑ Patient referred to palliative care if appropriate
- ❑ Routine follow-up visits scheduled

Step 7: End-of-life care

Palliative care

Consider a referral to palliative care. Ensure an advance care directive is in place.

Communication

The lead clinician's responsibilities include:

- being open about the prognosis and discussing palliative care options with the patient
- establishing transition plans to ensure the patient's needs and goals are considered in the appropriate environment.

Checklist

- ❑ Supportive care needs assessed and referrals to allied health services actioned as required
- ❑ Patient referred to palliative care
- ❑ Advance care directive in place

Visit our guides to best cancer care webpage <www.cancercareguides.org.au> for consumer guides. Visit our OCP webpage <www.cancer.org.au/OCP> for the optimal care pathway and instructions on how to import these guides into your GP software.

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