Optimal care pathway for people with acute lymphoblastic leukaemia (ALL)

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 acute lymphoblastic leukaemia*.

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

Step 1: Prevention and early detection

Prevention

The cause of ALL is not fully understood and there are currently no clear prevention strategies.

Risk factors

Currently known risk factors for developing ALL include the following:

- radiation exposure
- certain chemical exposures (chemotherapy drugs/benzene)
- some genetic syndromes (Down syndrome, Fanconi anaemia, Bloom syndrome, Neurofibromatosis, Li-Fraumeni syndrome, Ataxia-telangiectasia)
- age (adults over the age of 50 and children have an increased risk of ALL)
- gender (males are at a slightly higher risk of developing ALL).

Early detection

There are no specific tests that allow for the early detection of ALL. Patients identified at an increased risk of ALL should be closely observed by their care providers for arising symptoms.

Screening recommendations

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

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 most common presenting symptoms are those related to:

- anaemia
- thrombocytopenia
- neutropenia.

Other signs and symptoms include enlargement of the liver, spleen, lymph nodes or testicles, pain in the bone or joints, cranial nerve palsies, headaches, visual or auditory symptoms, seizures, dizziness, nausea and transient ischemic attack.

Most initial referrals for ALL will be to an Emergency Department. The practitioner at the point of referral must expedite assessments for referred patients.

If ALL is suspected initial investigations by the GP should include:

- a focussed medical history and thorough clinical assessment
- full blood examination and film.

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.

Checklist

- ❑ Symptoms related to anaemia, thrombocytopenia and/or neutropenia
- ❑ Signs and symptoms recorded
- ❑ Patient notified of support services such as Cancer Council 13 11 20 and Leukaemia Foundation 1800 620 420
- ❑ Referral options discussed with the patient and/or carer including cost implications

FIRST EDITION
This quick reference guide was published in June 2023

Australian Government
Cancer Australia

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

Step 2: Presentation, initial investigations and referral continued

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.

Timeframe

If there is suspicion of ALL, the GP should begin investigations **immediately**. If the patient is clinically unwell (e.g., they present with sepsis, bleeding or severe symptoms) they should be regarded as a medical emergency and referred to an emergency facility.

Patients with suspected ALL who present to an emergency department should be triaged as a medical emergency initially and **immediately** discussed with a clinical haematology service and/or transferred to a specialist centre.

Step 3: Diagnosis, staging and treatment planning

ALL is diagnosed based on full blood count, bone marrow investigation and a thorough medical history and physical examination.

Laboratory studies include:

- complete blood count with differential, chemistry profile, liver function tests, and coagulation studies.

The following imaging studies should also be undertaken:

- CT scan of the neck, chest, abdomen and pelvis with intravenous contrast
- CT or MRI of the head
- scrotal ultrasound.

Other diagnostic tests include:

- bone marrow aspirate
- lymph node biopsy to be used in cases where there is lymphadenopathy
- lumbar puncture to determine CNS involvement
- cytogenetics, molecular studies and flow cytometry to determine whether the case is T-ALL or B-ALL.

Genetic testing

NA

Treatment planning

The multidisciplinary team should discuss patients with acute lymphoblastic leukaemia 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 acute lymphoblastic leukaemia.

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

Timeframe

Initial diagnostic workup including bone marrow aspirate, blood work up and imaging should all be performed **within 48 hours** of patient presentation with results received **within 24 hours** of the procedure.

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

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 continued

Lymph node biopsies and lumbar puncture should be performed **within 72 hours** of patient presentation.

The results of cytogenetics, FISH and excisional or core biopsies should be available **within 72 hours**.

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.

All patients treated for ALL will receive **systemic therapy**. Induction therapy is the first phase of initial therapy and may include a combination of: high-dose corticosteroid, anthracycline, vincristine, asparaginase and TKIs (in PH positive disease). Treatment for newly diagnosed ALL generally consists of 9-12 months of intensive chemotherapy followed by up to 3 years of maintenance chemotherapy.

Allogeneic stem cell transplant should be considered in patients with high-risk disease who are minimal residual positive or unable to complete their standard therapy.

CNS prophylaxis should be given throughout the entire course of therapy to prevent ALL from spreading to the CNS.

Radiation therapy can be considered as an emergency treatment in the following situations:

- radiation to the brain area can be considered for patients with leukaemia in the CNS at diagnosis
- radiation to the testes may be considered in patients with testicular disease at diagnosis that remains following induction therapy
- cranial irradiation can be administered to prevent ALL from spreading to the brain
- total body irradiation may be administered in preparation for a bone marrow transplant.

Targeted therapies and immunotherapy should be considered for patients with CD20+ disease or Philadelphia positive disease. The following therapies should be considered:

- Rituximab for patients with CD20+ disease
- a tyrosine kinase inhibitor such as dasatinib, imatinib or ponatinib for patients with Philadelphia positive disease
- Blinatumomab for patients with minimal residual disease and B-lineage disease.

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'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

- ❑ 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

Timeframe

Induction therapy should commence **as soon as possible**. Consolidation therapy should commence **immediately following induction therapy**.

Radiation should be commenced **within 72 hours** of recognition of the issue where organ preservation is the goal. When applied with palliative goals, timing is guided by severity of the relevant symptoms.

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.

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

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 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'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.

Endorsed by:

ALLG <www.allg.org.au> ANZTCT <www.anztct.org.au> Cancer Council <www.cancer.org.au> HSANZ <www.hsanz.org.au> Leukaemia Foundation <www.leukaemia.org.au>