

Optimal care pathway for children, adolescents and young adults with acute leukaemia

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, families and/or 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 children, adolescents and young adults with acute leukaemia* (CAYA acute leukaemia). The pathway covers people aged 0-30 years.

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

Step 1: Prevention and early detection

Prevention

Although risk factors have been identified, the cause of CAYA acute leukemia is unknown. There is currently no evidence that lifestyle plays a role. It is important to ensure the patient and their family and/or carer are aware of this to avoid feeling responsible for the illness.

Risk factors

The risk factors for developing a CAYA acute leukaemia include:

- genetic predisposition to an increased likelihood of developing CAYA acute leukaemia in patients with Down syndrome, neurofibromatosis type-1,

ataxia telangiectasia and inherited bone marrow failure syndromes

- having a sibling with a CAYA acute leukaemia; however, the risk is so low there is no recommendation to routinely screen siblings
- treatment with certain chemotherapy agents for another cancer.

Early detection

There are no screening tools for early detection of newly diagnosed CAYA acute leukaemia. In patients with an identified cancer predisposition, a specialist may recommend screening full blood count examinations or bone marrow aspirates.

General health checklist

There are no specific prevention or early detection checks that are recommended unless the patient presents with signs or symptoms (refer to Step 2).

Step 2: Presentation, initial investigations and referral

Because CAYA acute leukaemia is rare it represents a major diagnostic challenge for primary healthcare professionals including GPs and emergency physicians. It is important to recognise parental concern and the need to escalate investigations, particularly after repeated visits to healthcare professionals.

The following **signs and symptoms** should be investigated and may warrant the consideration of a full blood count and peripheral blood film examination:

- persistent unexplained fever
- diffuse bone pain with no obvious trauma and/or refusal to walk in children
- generalised lymphadenopathy
- hepatosplenomegaly
- pallor

- unexplained bruising, unexplained bleeding or petechiae
- extreme fatigue
- recurrent infections.

Children can sometimes have only mild symptoms so the medical practitioner should be alert to the diagnosis, particularly when there is a constellation of the symptoms/signs above.

Initial investigations include a thorough clinical examination full blood count and blood film (performed immediately).

Referral options

A clinical suspicion or laboratory findings that suggest CAYA acute leukaemia warrants immediate telephone referral and presentation to the nearest specialist service. The patient and their family and/or

Checklist

- Signs and symptoms recorded
- Supportive care needs assessed and referrals to allied health services actioned as required
- Patient notified of support services such as Camp Quality 1300 662 267, Cancer Council 13 11 20, Canteen 1800 835 932, Leukaemia Foundation 1800 620 420 and Redkite 1800 733 548
- Referral options discussed with the patient, family and/or carer including cost implications

Step 2: Presentation, initial investigations and referral continued

carer should be warned they may need to transfer to a more specialised service after initial assessment.

At the referral stage, the patient's GP or other referring doctor should advise the patient and their family and/or carer about their options for referral, waiting periods, 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, family and/or carer who they are being referred to and why
- informing the patient, family and/or carer that they can contact Camp Quality 1300 662 267, Cancer Council 13 11 20, Canteen 1800 835 932, Leukaemia Foundation 1800 620 420 and Redkite 1800 733 548.

Timeframe

If there is suspicion of CAYA acute leukaemia, patients should be referred to a specialist **immediately**.

Test results should be provided to the patient, family and/or carer **immediately**.

Step 3: Diagnosis, staging and treatment planning

Diagnosis and staging

Perform pre-treatment medical investigations on the day of presentation to the specialist cancer service.

The diagnostic laboratory investigations should be performed as follows.

Urgent pathway

For urgent cases, if it is safe to do so, a diagnostic bone marrow aspirate and lumbar puncture should be performed on the day of presentation. Urgent patients include those who present with hyperleukocytosis, tumour lysis syndrome, mediastinal mass and coagulopathies, and those with suspected acute promyelocytic leukaemia.

If there is a suspicion of acute promyelocytic leukaemia, urgent treatment with all-trans retinoic acid should be instigated immediately.

Standard pathway

The diagnostic bone marrow aspirates and lumbar puncture should be performed by the next business day. Clinical trial requirements, as well as the level of institutional resources, should also guide timings.

Measurable/minimal residual disease

The importance of this test for measuring the patient's response to treatment cannot be overstated. (See the *Optimal care pathway for CAYA leukaemia* for principles for MRD in CAYA acute leukaemia subcategories.)

Genetic testing

Paired tumour/germline sequencing should be considered in some patients with a family history or clinical findings that suggest a possible cancer predisposition syndrome. Once a diagnosis is confirmed, a comprehensive family cancer history of at least three generations' pedigree will help further identify patients and families with potential cancer predisposition or inherited syndromes.

Treatment planning

Immediate treatment is often required before a full multidisciplinary meeting (MDM) ratifies the plan. Multidisciplinary input is likely after treatment begins.

Research and clinical trials

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

Checklist

- Diagnosis has been confirmed
- Performance status and comorbidities recorded
- Patient discussed at MDM and decisions provided to the patient, family and/or carer
- Clinical trial enrolment considered
- Supportive care needs assessed and referrals to allied health services actioned as required
- Consideration of future fertility consequences and referral to fertility specialist as required
- Patient referred to support services (such as Camp Quality, Cancer Council, Canteen, Leukaemia Foundation and Redkite) as required
- Treatment costs discussed with the patient, family and/or carer as appropriate

Step 3: Diagnosis, staging and treatment planning continued

Communication

The lead clinician's¹ responsibilities include:

- discussing a timeframe for diagnosis and treatment options with the patient, family 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
 - discussing the effect of leukaemia treatment on future fertility and referral to a fertility specialist as required
 - communicating with the patient's GP about the diagnosis, treatment plan and recommendations from MDMs.

Timeframe

Investigations should be completed immediately.

Prospective review at an MDM for all cases.

Step 4: Treatment

Intent of treatment for CAYA acute leukaemia is curative.

Treatment options

- Chemotherapy with or without immunotherapy is the key component for treating CAYA acute leukaemia.
- Radiation therapy has a role in some CAYA acute leukaemia patients with testicular disease or overt central nervous system disease. Radiation therapy is used as part of the conditioning regimen for patients undergoing haematopoietic stem cell transplantation. It can be useful in palliating symptomatic masses in advanced disease.
- haematopoietic stem cell transplantation is reserved for patients at greatest risk of relapse. It is used as a salvage where primary treatment has failed.

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, family and/or carer including the intent of treatment as well as risks and benefits
- discussing advance care planning with the patient, family 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
- Patient referred to support services (such as Camp Quality, Cancer Council, Canteen, Leukaemia Foundation and Redkite) as required
- Early referral to palliative care considered and advance care planning discussed with the patient and/or carer

Timeframe

As soon as possible after presentation.

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.

Step 5: Care after initial treatment and recovery

Provide a treatment and follow-up summary to the patient, family and/or 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 suspected recurrence.

Communication

The lead clinician's responsibilities include:

- explaining the treatment summary and follow-up care plan to the patient, family and/or carer
- informing the patient, family 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, family and/or carer and the patient's GP
- Supportive care needs assessed and referrals to allied health services actioned as required
- Patient referred to support services (such as Camp Quality, Cancer Council, Canteen, Leukaemia Foundation and Redkite) as required
- Patient-reported outcome measures recorded

Step 6: Managing refractory, relapsed, residual or progressive disease

Detection

Most residual or recurrent 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 extent 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. 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, family and/or carer and the patient's GP.

Checklist

- Treatment intent, likely outcomes and side effects explained to the patient, family and/or carer and the patient's GP
- Supportive care needs assessed and referrals to allied health and community support services as required
- Advance care planning discussed with the patient, family 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 and community support services as required
- Patient referred to palliative care
- Advance care directive in place

Visit the guides to best cancer care webpage <www.cancercareguides.org.au> for consumer guides. Visit the 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:

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