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 of the Optimal care pathway for people with acute myeloid leukaemia.

Step 1: Prevention and early detection

Prevention

The causes of AML are not fully understood and there is currently no clear prevention strategy.

Risk factors

Most people have no identifiable risk factors. It is possible for AML to run in families but is uncommon. Known risk factors include:

- advanced age
- prior chemotherapy, radiation therapy or high-dose radiation exposure
- known previous haematological disorder with a risk of leukaemic transformation
- known predisposing genetic disorders with a risk of leukaemic presentation
- obesity
- tobacco smoking

- having a first-degree relative with AML
- environmental exposure to industrial chemicals such as benzene.

Early detection

In patients with pre-existing pre-leukaemic disorders (e.g. myelodysplasia, other myeloid neoplasms) and predisposing genetic disorders, routine care of these should include full blood counts and bone marrow biopsies at appropriate clinical intervals.

There are no screening programs for AML.

Step 2: Presentation, initial investigations and referral

Signs and symptoms

Symptoms at presentation are usually non-specific and may include:

- fatigue, pallor or other symptoms of anaemia
- symptoms of serious infection, such as tachycardia, high fevers, rigors
- unresolving or unusual infection/fever
- abnormal bleeding or bruising
- sore gums or mouth ulcers
- unexplained bone pain
- unintentional weight loss
- unexplained fevers.

The following signs and symptoms require consultation as a medical emergency:

- sepsis
- symptomatic anaemia
- severe thrombocytopenia < 20 × 10^9/L
- major laboratory abnormalities
- very high white cell count (> 50 × 10^9/L) or signs of hyperviscosity
- spontaneous/uncontrolled bleeding
- coagulopathy.

Checklist

- Recent weight changes discussed and the patient’s weight recorded
- Alcohol intake discussed and recorded and support for reducing alcohol consumption offered if appropriate
- Smoking status discussed and recorded and brief smoking cessation advice offered to smokers
- Physical activity recorded
- Referral to a dietician considered
- Referral to a physiotherapist or exercise physiologist considered
- Education on being sun smart considered
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

Initial investigations
If a serious blood disorder is suspected a focused medical history and thorough clinical assessment should be undertaken. Full blood count and film should be performed immediately. If the patient is clinically unwell (presents with symptomatic anaemia, spontaneous bleeding, sepsis or has symptoms of hyperviscosity), immediate referral to an emergency facility is recommended without waiting for blood results. Morphologic evidence of APL, disseminated intravascular coagulation, severe thrombocytopenia and any organ dysfunction (renal/liver failure) should be considered a medical emergency.

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, if there are likely to be 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 on 13 11 20.

Timeframe
The GP should start investigations immediately if AML is suspected. Laboratory results should be actively followed up and progressed on the same day. Patients with sepsis, bleeding or severe symptoms should be regarded as a medical emergency and be referred immediately to an appropriate emergency facility without necessarily waiting for results of laboratory tests (same day). Patients with a laboratory diagnosis of possible AML should be referred for an urgent assessment by a haematologist at an appropriate facility within 24 hours (unless advised otherwise by a haematologist).

Step 3: Diagnosis, staging and treatment planning

Diagnosis
Diagnostic evaluation is required first to establish a precise diagnosis, according to the most recent classification system, and second to assess the presence and management of comorbidities and the patient’s fitness because these affect both response to treatment and toxicity from treatment.

The treatment team should:
- take a thorough medical history and perform a thorough physical examination, including assessing for the presence of extramedullary disease
- undertake the following investigations under guidance of a specialist:
  - peripheral blood tests
  - bone marrow aspirate
  - trephine biopsy +/- lumbar puncture, imaging or tissue biopsy when extramedullary disease is suspected.

Specialist testing (including genetic testing): Every patient being considered for AML therapy should have samples taken for morphological assessment, cytogenetics, flow cytometry and molecular pathology.

Most genetic abnormalities in AML only occur in abnormal blood cells and are not related to genetic abnormalities that affect the whole body and/or are inherited. However, heritable genetic abnormalities may be identified in a small number of patients.

Other pre-treatment investigations:
Careful clinical and haematological assessment is required to identify patients in whom the start of chemotherapy could or should be delayed. The presence of an active infection at diagnosis is important to identify.

Treatment planning: Because of the urgency and complexity of treatment, every clinical haematology unit should have predefined peer-reviewed treatment models of care that have been endorsed by the multidisciplinary team. Induction treatment is often required before a full MDM ratifies details of the ongoing management plan (which should include full details of the response assessment).

Checklist
- Diagnosis confirmed
- Full histology obtained
- Samples taken for morphological assessment, cytogenetics, flow cytometry and molecular pathology
- Performance status and comorbidities measured and recorded
- Patient discussed at an MDM and decisions provided to the patient and/or carer
- Clinical trial, registry and tissue-banking of samples (if available) enrolment considered
- Supportive care needs assessment completed and recorded and referrals to allied health services actioned as required
- Patient referred to support services (such as Cancer Council) as required
- Treatment costs discussed with the patient and/or carer
Step 4: Treatment

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

Establish intent of treatment
- Curative
- Anti-leukaemia therapy to improve quality of life and/or longevity without expectation of cure
- Symptom palliation including active supportive care

Treatment options to induce remission

Patients fit for intensive chemotherapy: Induction chemotherapy should ideally only be started when all diagnostic criteria have been satisfied. Once patients are in remission, consolidation therapy is always indicated when cure is the intention.

Patients not fit for intensive chemotherapy: Referral to a clinical trial should be a priority. Available treatment options include low-dose chemotherapy, hypomethylating agents for select patients, or palliative/supportive care to control symptoms.

Allogeneic stem cell transplant: Should be considered for select patients (refer to the AML optimal care pathway).

Radiation therapy: May be used for symptom control and occasionally for treating extramedullary disease.

Other treatment options

Acute promyelocytic leukaemia
Rapid initiation of APL-specific therapy is essential and, in some cases, may precede formal confirmation of the diagnosis.

Refractory disease
- Allogeneic stem cell transplant for select patients.
- Palliative systemic treatment is often a reasonable option.
- Clinical trials and experimental therapy should be considered.

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, visit the Palliative Care Australia website (<www.palliativecare.org.au>.

Communication
- 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 of treatment established
- Risks and benefits of treatments discussed with the patient and/or carer
- Early referral for transplant considered in select patients
- Treatment plan discussed with the patient and/or carer and provided to the patient’s GP
- Treating specialist has adequate qualifications, experience and expertise
- Supportive care needs assessment completed and recorded, and referrals to allied health services actioned as required
- Early referral to palliative care considered
- Advance care planning discussed with the patient and/or carer

Timeframe
- Induction therapy should begin promptly after diagnosis and confirmation of a treatment plan. Consolidation therapy should begin within six weeks of induction chemotherapy starting. Donor searches should begin as soon as the patient’s risk status is known.

Step 3: Diagnosis, staging and treatment planning continued

Research and clinical trials:
Participation in clinical trials, registries and tissue banking is considered a standard of care for patients with AML.

Communication

The lead clinician’s 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

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

Timeframe
Morphological assessment to identify APL should be conducted immediately and the result conveyed to the treating physician as soon as possible. For all patients with AML, other results necessary for immediate management decisions should be available within 72 hours of the patient presenting.

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 suspected recurrence.

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 assessment completed and recorded and referrals to allied health services actioned as required
- Patient-reported outcome measures recorded

Step 6: Managing relapsed or refractory disease

Detection

Relapse occurs in more than 50% of patients. Most cases of relapsed AML are diagnosed through 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 location and extent of disease, previous management and the patient’s preferences. Supportive care is integral to the care of all patients with relapsed or refractory AML.

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 assessment completed and recorded 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 assessment completed and recorded, and referrals to allied health services actioned as required
- Patient referred to palliative care
- Advance care directive in place