

Optimal care pathway for people with multiple myeloma

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 multiple myeloma*.

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

Step 1: Prevention and early detection

Prevention

The causes of multiple myeloma (MM) are not fully understood, and there is currently no clear prevention strategy.

Risk factors

The risk factors for developing MM include:

- age (occurs mainly in people aged over 60)
- clinically defined monoclonal gammopathy of undetermined significance (MGUS)
- having a first-degree relative with MM, however the absolute risk for anyone with a relative with MM is low
- people of black African, Maori or Pacific Islander descent

- gender (males have a slightly higher risk)
- a high body mass index
- exposure to radiation and chemicals, but published evidence has not been consistent.

Early detection

There is currently no established benefit to early detection of MGUS, the pre-malignant phase of myeloma.

Screening recommendations

There is no indication for screening for MM in the general population, but patients with MGUS or smouldering myeloma require regular monitoring to assess for possible progression to MM.

General health checklist

- Recent weight changes discussed and recorded
- Alcohol intake and smoking status discussed and support offered if appropriate
- Physical activity recorded
- Referral to a dietitian, physiotherapist or exercise physiologist considered
- Sun smart advice

Step 2: Presentation, initial investigations and referral

Signs and symptoms to investigate

MM can present with many otherwise unexplained, non-specific symptoms and/or blood test abnormalities, as well as end-organ complications or related organ tissue injury.

End-organ complications that are often associated with MM include: bone loss, renal failure, hypercalcaemia, immune suppression or anaemia.

Signs and symptoms include:

- fatigue and general weakness
- bone pain or a broken bone with minimal trauma
- frequent infections
- unintentional weight loss
- symptoms of hypercalcaemia including mental fogging or confusion, new-onset constipation or abdominal pain and increased thirst

- new-onset back pain, particularly with neurological symptoms such as leg weakness, loss of bladder or bowel control, or loss of sensation
- hyperviscosity (easy bruising, bleeding gums, cloudy vision), but this is rare.

Blood test abnormalities that may suggest MM include:

- anaemia
- high levels of creatinine
- hypercalcaemia
- elevated ESR without a known infection or inflammation
- increase protein–albumin gap without an infection or inflammation
- a paraprotein on serum protein electrophoresis
- elevated serum kappa or lambda light chains with an abnormal kappa–lambda light chain ratio.

Checklist

- Signs and symptoms recorded
- Investigations completed
- Supportive care needs assessed, and referrals to allied health services actioned as required
- Patient notified of support services such as Cancer Council 13 11 20, Leukaemia Foundation 1800 620 420 and Myeloma Australia 1800 693 566
- Referral options discussed with the patient and/or carer including cost implications

Step 2: Presentation, initial investigations and referral continued

Initial investigations

If there is high suspicion of MM after clinical assessment, the GP should promptly refer the patient to a haematologist or specialist centre to confirm the diagnosis.

In cases where clinical suspicion is lower, preliminary investigations include the following:

- blood test to identify paraproteins or elevated free light chains that may indicate underlying MM
- blood and urine tests that may indicate end-organ dysfunction
- x-ray or CT imaging of painful areas to assess for fractures, lytic lesions and/or soft tissue plasmacytomas.

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, Leukaemia Foundation and Myeloma Australia.

Timeframe

Presenting symptoms should be promptly assessed. If moderate or severe symptoms are identified, the GP should review promptly (**within 2 days**) to avoid missing a medical emergency.

In non-urgent cases the GP should have results, review the patient and finalise a path of action **within 4 weeks**.

Patients with a paraprotein and/or elevated light chain and end-organ damage should be seen by a specialist **as soon as possible**, ideally **not more than 1 week**.

If there is no end-organ damage, a specialist consult should occur **within 4 weeks**.

Step 3: Diagnosis, staging and treatment planning

Diagnosis and staging

Tests that are always indicated include:

Blood and urine tests to assess for myeloma and myeloma-defining events:

- full blood count, differential and blood film
- urea and electrolytes, calcium, phosphate, magnesium, urate
- liver function test, albumin
- beta-2 microglobulin, LDH, C-reactive protein
- serum protein electrophoresis and immunofixation
- serum free light chain
- 24-hour urine collection: protein excretion, creatinine clearance, Bence Jones protein.

Bone marrow aspirate and trephine with morphology, immunohistochemistry, cytogenetics, FISH and flow cytometry.

Other tests to look for end-organ damage include a whole-body low-dose CT skeletal survey and, in selected cases, whole-body or whole-spine and pelvis MRI or PET-CT.

The Revised International Staging System (R-ISS) staging criteria for MM is based on: beta-2 microglobulin, albumin, LDH and cytogenetics/FISH assessment.

Treatment planning

The multidisciplinary team should ideally discuss all newly diagnosed patients with

multiple myeloma prior to treatment implementation.

In some cases, treatment may be required sooner.

Research and clinical trials

See the OCP resources appendix and relevant steps for a list of clinical trial resources relevant to MM.

Communication

The lead clinician's¹ 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.

Checklist

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

Timeframe

Investigations should be completed **within 2 weeks** of the first consult, or sooner depending on clinical urgency. Prospective review at a multidisciplinary meeting should ideally occur for all cases.

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 4: Treatment

MM is complex and treatment aims may change throughout the disease course. It's important to note that MM is an incurable cancer, and nearly all patients will eventually relapse after each line of therapy.

Establish intent of treatment

For MM:

- to obtain deep remission for durable disease control
- to improve quality of life and/or longevity without expectation of deep remission
- symptom palliation.

For solitary plasmacytoma:

- potential cure.

Systemic therapy is administered in virtually all patients treated for MM and may include immunomodulatory drugs, proteasome inhibitors, chemotherapy, monoclonal antibodies and corticosteroids.

Autologous stem cell transplant (ASCT) is usually included as part of frontline therapy for transplant-eligible patients.

Radiation therapy can provide rapid local control of myeloma or plasmacytoma that is causing pain or acute organ compromise such as spinal cord compression. In patients with solitary bone plasmacytoma or solitary extramedullary plasmacytoma, radiation therapy alone can offer durable control, and potentially cure.

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.

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

Systemic therapy should begin **within 2 weeks** of diagnosis and staging. In cases with critical organ compromise or rapid clinical progression, treatment may need to start **within 24 hours** of diagnosis.

Radiation therapy should start **within 24 hours** of referral where possible for patients with acute critical organ compromise, with a maximum acceptable waiting time of 48 hours.

For symptomatic tumours, radiation therapy should start **within 48 hours** of referral, with a maximum acceptable waiting time of 14 days.

For solitary bone plasmacytoma or extramedullary plasmacytoma, radiation therapy should begin **within 14 days** of referral, with a maximum acceptable waiting time of 28 days.

Step 5: Care after initial treatment

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 the care of these
- supportive care services provided
- a follow-up schedule, including tests required and timing, as well as vaccinations where appropriate

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

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

Detection

Most relapsed or progressive disease will be detected via routine follow-up blood tests or by the presence of symptoms.

Treatment

Treatment will depend on patient factors (age and frailty), disease factors (tempo of relapse, risk-group stratification), prior treatment-related factors (responsiveness and side effects to prior treatment type) and the patient's preferences.

When managing people with relapsed or refractory MM, consider a clinical trial. If no clinical trial is available, a different treatment regimen than was previously used can be given.

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 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:

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>

Myeloma Australia <www.myeloma.org.au>