Optimal care pathway for people with AL amyloidosis

FIRST EDITION

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Endorsed by

Australian Amyloidosis Network

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Statement of acknowledgement

We acknowledge the Traditional Owners of Country throughout Australia and their continuing connection to the land, sea and community. We pay our respects to them and their cultures and to Elders past, present and emerging.

This work is available from the Leukaemia Foundation website <www.leukaemia.org.au/blood-cancer/journey/active-treatment/optimal-care-pathways/ocps-for-healthcare-professionals/> and from the Cancer Council website <www.cancer.org.au/OCP>.

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Contents

Welcome and introduction i

Summary 1

Intent of the optimal care pathways 5

Optimal care pathway resources 6

Principles of the optimal care pathway 8

Principle 1: Patient-centred care 8

Principle 2: Safe and quality care 11

Principle 3: Multidisciplinary care 12

Principle 4: Supportive care 12

Principle 5: Care coordination 14

Principle 6: Communication 15

Principle 7: Research and clinical trials 17

Summary – optimal timeframes 18

Optimal care pathway 19

Step 1: Prevention and early detection 20

Step 2: Presentation, initial investigations and referral 22

Step 3: Diagnosis, staging and treatment planning 26

Step 4: Treatment 34

Step 5: Care after initial treatment and recovery 42

Step 6: Managing relapsed, residual or progressive disease 48

Step 7: End-of-life care 52

Contributors and reviewers 55

Appendix A: Supportive care domains 57

Appendix B: Psychological needs 58

Appendix C: Special population groups 59

Appendix D: Complementary therapies 65

Appendix E: Members of the multidisciplinary team for AL amyloidosis 66

Resource list 67

Glossary 71

References 73

Welcome and introduction

As the current Steering Committee Chair of the Australian Haematology Optimal Care Pathways, I am honoured to introduce our latest initiative aimed at improving care for patients with haematological conditions. This is the third edition of the Haematology OCPs, with the first and revised editions led by Professor Robert Thomas OAM and the second by Professor Peter Mollee.

With the support of the federal government, our latest optimal care pathways have expanded the suite of guides to include a broader range of haematological conditions.

Optimal Care Pathways are clear, evidence-based guides that outline high-quality principles and best practice opportunities for all those involved in cancer care. These guidance documents serve as a national standard for providing the best possible care for patients, their families, and carers, regardless of their geographical or personal circumstances. By outlining the key steps in diagnosis, treatment, and ongoing management, we aim to reduce variations in management and improve outcomes for patients.

We recommend the optimal care pathways to people living with haematological conditions and their carers. These resources can guide discussions with a patient's healthcare team and support individuals in making informed decisions about their care. Specific optimal care pathways are available for Aboriginal and Torres Strait Islander people, and the Guides to the best cancer care for patients are available in various languages.

The current edition of Haematology Optimal Care Pathways cover a range of haematological disease groups, including Acute Lymphoblastic Leukaemia, Myeloproliferative Neoplasms, Cutaneous T-cell non Hodgkin Lymphoma, Waldenstrom Macroglobulinaemia, and AL Amyloidosis. We believe these pathways will be a valuable tool for clinicians, patients, and their families, and we are committed to ensuring that they continue to evolve and improve over time.

These optimal care pathways are endorsed by the federal government through Cancer Australia and by all states and territories, with Australia-wide clinical acceptance and government support.

As Steering Committee Chair of this project, I express my sincerest appreciation for the individual OCP Chairs, Co-Chairs, and all content contributors for each pathway, as well as the individuals and organizations who contributed to the review of these guidelines. We also acknowledge the support of the Federal, State, and Territory governments.

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Vice President 2021-23, HSANZ
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Summary

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 AL Amyloidosis*.

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

Step 1: Prevention and early detection

Prevention

The cause of AL amyloidosis is unknown, and there are currently no effective prevention strategies.

Risk factors

- age (occurs mainly in people aged over 40)
- gender (slightly more common in males)
- a pre-existing monoclonal gammopathy (small proportion go onto to develop AL amyloidosis).

Of those with a monoclonal gammopathy, structural changes to immunoglobulin light chains make them more likely to misfold and form amyloid fibrils.

Early detection

Routine screening for AL amyloidosis is not currently recommended in the general population.

For patients with a known diagnosis of a monoclonal gammopathy (MGUS, smouldering myeloma, multiple myeloma) a high index of clinical suspicion and careful assessment can facilitate early diagnosis of AL amyloidosis.

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

AL amyloidosis is a multi-organ disease and can present with many otherwise unexplained, non-specific symptoms and/or blood test abnormalities, as well as end-organ complications. As the symptoms are variable and also observed in other more common conditions such as diabetes mellitus, the diagnosis of AL amyloidosis can easily be overlooked which contributes to delayed diagnosis.

Symptoms and signs can include heart failure, fatigue, near-syncope, postural hypotension, oedema, raccoon eyes, macroglossia, carpal tunnel syndrome, peripheral neuropathy, dyspnoea, weight loss, indigestion, renal impairment, proteinuria, hepatomegaly, jaw claudication and gastrointestinal bleeding. The symptoms and signs reflect the presence and extent of organ involvement by amyloidosis.

Common clinical presentations, particularly if they occur concurrently, that raise the suspicion of amyloidosis include:

- heart failure with preserved ejection fraction
- nephrotic syndrome
- peripheral neuropathy, especially rapidly progressive neuropathy and/or autonomic neuropathy
- hepatomegaly with normal imaging appearance.

In most patients more than one organ is affected (most frequently the heart, kidneys, gastrointestinal tract, nervous system and liver), however, some patients may only have one affected organ. Soft tissue involvement such as macroglossia and periorbital purpura are highly suggestive of AL amyloidosis but are uncommon (occurring in only 10%).

Checklist

- ❑ Unexplained, non-specific symptoms, such as fatigue and weight loss, and/or blood abnormalities, as well as end-organ complications.
- ❑ Signs and symptoms recorded
- ❑ Patient notified of support services such as Cancer Council 13 11 20, Leukaemia Foundation 1800 620 420, Australian Amyloidosis Network <www.AAN.org.au> and Myeloma Australia 1800 693 566
- ❑ Referral options discussed with the patient and/or carer including cost implications

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

Initial investigations by the GP should include:

The role of the general practitioner is to suspect the patient has a disease that requires a detailed history and examination that may be followed by basic investigations to help identify the organ system/s that require specialist assessment.

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, Leukaemia Foundation 1800 620 420, Australian Amyloidosis Network <www.AAN.org.au> and Myeloma Australia 1800 693 566.

Timeframe

The timing for specialist referral is guided by the severity and type of organ dysfunction. Heart failure, nephrotic syndrome or deteriorating kidney function should be seen by the appropriate specialist **within 1 week**.

For less severe cases to be seen by a specialist physician, the appropriate time frame is **4 weeks**.

Step 3: Diagnosis, staging and treatment planning

If AL amyloidosis is suspected, the following initial investigations are indicated. If these tests are all negative, a diagnosis of AL amyloidosis is extremely unlikely:

- serum and urine protein electrophoresis and immunofixation
- serum free light chains.

A diagnosis of systemic AL amyloidosis requires a tissue biopsy (with Congo Red stain or electron microscopy if available).

- targeted biopsy of an affected organ has the highest sensitivity for diagnosing amyloidosis.
- the biopsy can be at a peripheral 'distant' screening tissue (abdominal fat aspirate, rectum, bone marrow, gingiva, salivary gland, etc) but the sensitivity of these biopsies is significantly lower. If there is a high clinical suspicion of amyloidosis and the initial distant site screening biopsies are negative, then the clinically affected organ should be biopsied.

Once AL amyloidosis is suspected or confirmed by tissue biopsy, prompt referral to a haematologist or specialist centre is required for AL subtype confirmation, completion of staging investigations and treatment planning.

Treatment planning

Treatment will depend on several factors, including age, stage, comorbidities, frailty and patient choice. The multidisciplinary team should discuss patients with AL amyloidosis 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 AL Amyloidosis.

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.

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, Australian Amyloidosis Network and Myeloma Australia)
- ❑ Treatment costs discussed with the patient and/or carer

Timeframe

If AL amyloidosis is suspected, diagnostic and staging investigations should be completed **within 4 weeks** of the first consultation by the haematologist or specialist treating centre, or sooner depending on clinical urgency.

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 4: Treatment

Establish intent of treatment

The natural history of AL amyloidosis is complex, and treatment aims may change throughout the disease course. Effective treatments can result in removal of amyloid deposition and recovery of organ function. It's important to note that the monoclonal gammopathy that underlies AL amyloidosis is generally not curable, and many patients will eventually relapse after each line of therapy.

The intent of treatment can include:

- to obtain deep remission with the aim of reversing amyloid deposition and improving the function of organs affected by the amyloidosis
- to improve quality of life and/or longevity
- symptom palliation.

The treatment intent should be established in a multidisciplinary setting, documented in the patient's medical record and conveyed to the patient and carer as appropriate.

Systemic therapy will be suitable for most patients diagnosed with AL amyloidosis with the aim of preventing early death, improving organ function and prolonging survival. Prompt initiation of anti-plasma cell therapy is essential.

Induction therapy can include a combination of proteasome inhibitors, chemotherapy, monoclonal antibodies or corticosteroids and aims to rapidly reduce the amyloidogenic free light chain by directly targeting the underlying plasma cell clone. Patients with IgM-related AL Amyloidosis require therapy directed against the underlying lymphoid clone, most commonly a lymphoplasmacytic clone with treatment adapted from Waldenström macroglobulinemia (WM).

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

Timeframe

Systemic therapy should start **within 4 weeks** of diagnosis particularly if there is cardiac and/or renal disease.

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 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.

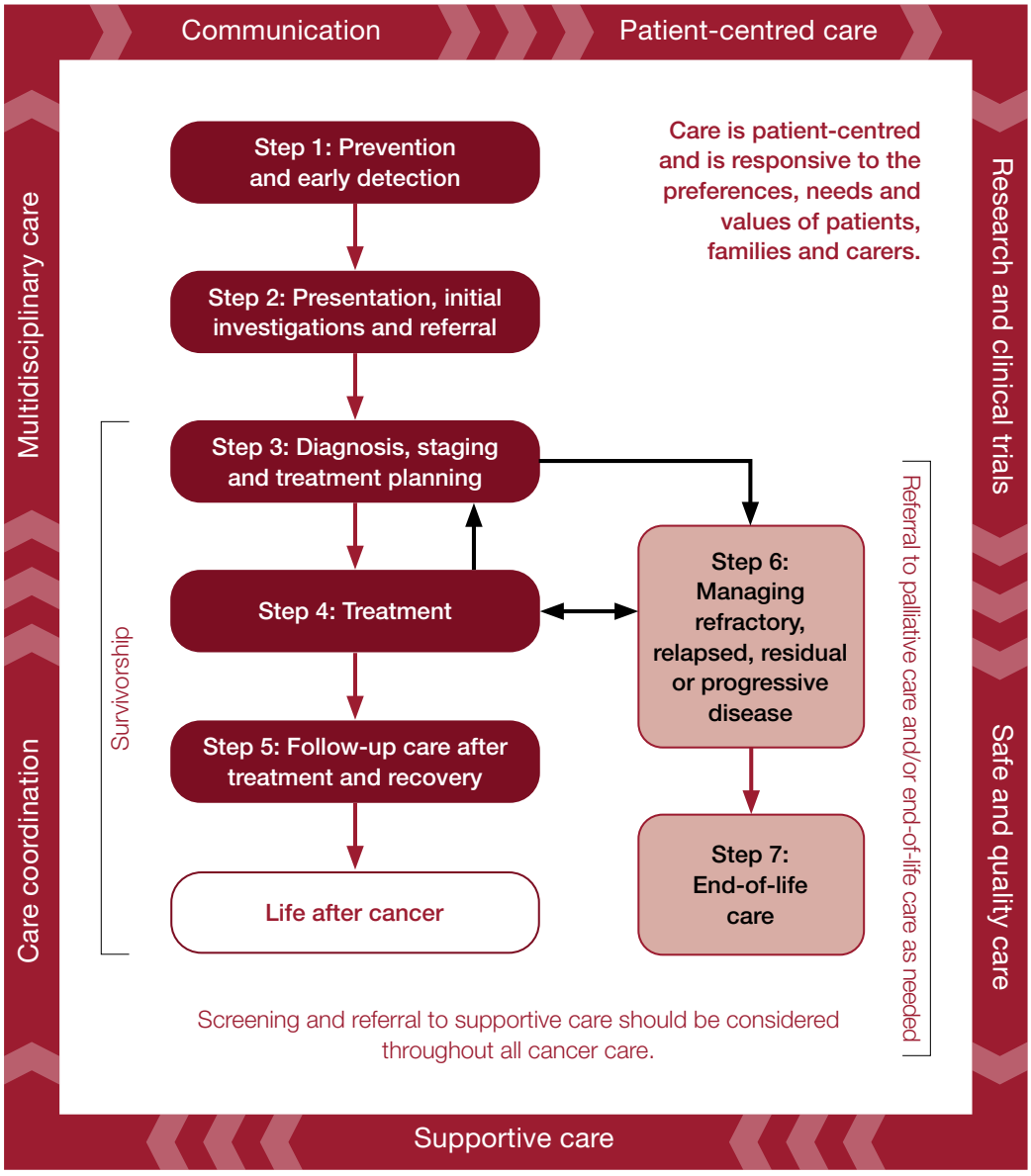
Intent of the optimal care pathways

Optimal care pathways map seven key steps in cancer care. Each of these steps outlines nationally agreed best practice for the best level of care. While the seven steps appear in a linear model, in practice, patient care does not always occur in this way but depends on the particular situation (e.g. the type of cancer, when and how the cancer is diagnosed, prognosis, management, the patient's decisions and their physiological response to treatment).

The principles underpinning optimal care pathways always put patients at the centre of care throughout their experience and prompt the healthcare system to deliver coordinated care.

The optimal care pathways do not constitute medical advice or replace clinical judgement, and they refer to clinical guidelines and other resources where appropriate.

Figure 1: The optimal care pathway



Optimal care pathway resources

There are three resources for each pathway: an optimal care pathway, a quick reference guide for health professionals and a guide to best cancer care for patients, carers and families.

Optimal care pathways

This optimal care pathway is designed for health professionals and health services. However, patients and carers may find useful information in this version to help understand the processes their treating health professionals are following.

This resource aims to:

- assist health professionals to provide optimal care and support to patients with cancer, their families and carers
- provide optimal timeframes for delivering evidence-based care
- emphasise the importance of communication and collaboration between health providers and people affected by cancer
- assist and inform new health professionals or trainees who are entering the cancer care workforce
- provide value to health systems to identify gaps in current cancer services, bring about quality improvement initiatives and improve how services are planned and coordinated. Adherence to the pathways should be measured wherever possible.

Visit the Cancer Council website <www.cancer.org.au/OCP> to view the optimal care pathways.

Quick reference guides

The quick reference guides are for health professionals and health services. They provide a summary of each optimal care pathway for health professionals and patients.

The quick reference guides include:

- optimal timeframes within which tests or procedures should be completed
- checklists with indicators related to recommendations in the optimal care pathway.

Visit the Cancer Council website <www.cancer.org.au/OCP> to view the quick reference guide for this optimal care pathway.

Guides to best cancer care

The guides to best cancer care are consumer resources that help patients understand the optimal cancer care that should be provided at each step. Carers, family and friends may also find the guides helpful.

The guides to best cancer care:

- include optimal timeframes within which tests or procedures should be completed
- include prompt questions to support patients to understand what might happen at each step of their cancer journey and to consider what questions to ask
- provide information to help patients and carers communicate with health professionals
- are available in eight languages.

Visit the Cancer Council's website <www.cancercareguides.org.au> to view the guides to best cancer care.

Optimal care pathway for Aboriginal and Torres Strait Islander people with cancer

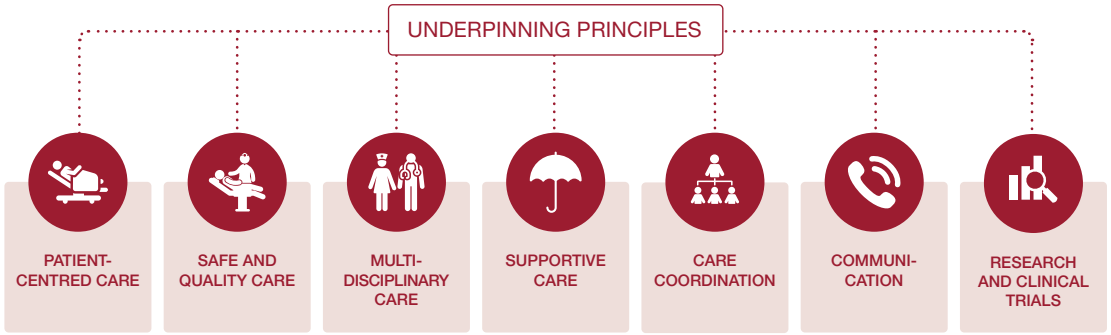
The *Optimal care pathway for Aboriginal and Torres Strait Islander people with cancer* provides a tool to help reduce disparities and improve outcomes and experiences for Aboriginal and Torres Strait Islander people with cancer. This resource can be used in conjunction with the optimal care pathway for each cancer type.

Visit the Cancer Australia website <https://www.canceraustralia.gov.au/publications-and-resources/cancer-australia-publications/optimal-care-pathway-aboriginal-and-torres-strait-islander-people-cancer> to view the optimal care pathway for Aboriginal and Torres Strait Islander people with cancer.

Principles of the optimal care pathway

The seven principles of care define appropriate and supportive cancer care that is the right of all patients and the right of those caring for and connected with them.

Figure 2: The seven principles underpinning the optimal care pathway



Principle 1: Patient-centred care

Patient-centred care informs and involves patients in their care and respects and responds to the preferences, needs and values of patients, families and carers.

A patient-centred focus increases the experience and satisfaction of patients, their families and carers, and staff, as well as safety and cost-effectiveness (ACSQHC 2019a).

Patient-centred care means:

- patients are informed and involved in decisions about their cancer and the treatment, post-treatment and recovery program ahead
- patients, their families and carers are provided with access to appropriate and accessible health information
- respect for the cultural and religious beliefs of patients and their families is demonstrated when discussing the diagnosis of cancer
- active communication is used to engage patients, their families and carers in the care process – an essential step for patients to be informed
- care processes are mutually beneficial for patients and providers
- special needs are addressed – for example, the needs of people with disabilities or mental health issues.

Informed choice and consent

An informed patient has greater confidence and competence to manage their cancer journey.

Health professionals are responsible for enabling patients to make informed choices according to their preferences, needs and values. Patients should be provided with:

- individualised and timely information and guidance about their treatment
- details of their care, including the advantages and disadvantages of each treatment, the associated potential side effects, the likely outcomes on their performance status (how well a patient is able to carry out activities of daily life) and subsequently their quality of life and any financial implications, at each stage of the pathway (ACSQHC 2020).

Health professionals have a legal responsibility to obtain consent for all procedures from either the patient or their substitute decision-maker if they are not deemed competent.

Referral choices and informed financial consent

Patients have the right to receive the information they need to be able to make an informed decision on where to be referred for treatment. Treating specialists and practitioners should clearly explain the costs or how to find out the costs of services, tests and treatment options upfront to avoid consumers experiencing 'bill shock'.

At the time of referral, the patient's general practitioner or other referring doctor should discuss the different 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. Referral decisions influence the care patients receive along the pathway and the direct and indirect costs they and their carers may incur. Different referrals have different costs:

- referral to a public hospital, which may involve some costs
- initial referral to a private specialist with associated costs, with the option of ongoing treatment in a public hospital at any time
- referral to a patient's choice of practitioner for immediate and ongoing private hospital management with associated costs.

Patients should be made aware that even though public hospital health care is 'free' to all Australian citizens and most permanent residents of Australia, there are still associated direct costs such as:

- over-the-counter medication and prescriptions
- wound dressings
- travel costs
- parking fees
- tests that are not covered by Medicare.

A cancer diagnosis and treatment may affect a patient's or carer's income. This is an indirect cost associated with cancer. Social work support is essential to help patients and their families deal with this issue. Patients should be advised not to undergo private care with significant out-of-pocket expenses if financially constrained. Specialists in private practice need to explain costs at the start of each new treatment to acknowledge the cumulative out-of-pocket expenses that patients can incur.

Patients and carers should be made aware of other forms of potential financial support that may be available, including whether the diagnosis or treatment triggers any insurance or access to superannuation, patient-assisted travel schemes, Centrelink or other forms of social security.

For more information on informed financial consent see Cancer Council's Standard for informed financial consent <https://www.cancer.org.au/health-professionals/resources/informed-financial-consent>.

Financial counselling services can provide advice on dealing with financial difficulties. These services can be accessed publicly (via social workers at hospitals, financial counsellors at neighbourhood houses or rural financial aid), privately or through cancer support services such as local charity groups or social work services.

For practical and financial assistance, patients may consider Cancer Council's financial services <www.cancer.org.au/support-and-services/practical-and-financial-assistance.>

Shared care

Shared care between a cancer specialist and primary care health professional is delivered in two or more settings by two or more professionals. The primary care provider is usually a general practitioner but can include nurses and allied health practitioners. Shared care can be delivered throughout the care pathway including during treatment, follow-up care, survivorship care and end-of-life care.

Shared care offers several advantages to patients, including the potential for treatment closer to home and more efficient care with less duplication and greater coordination. Evidence comparing shared care and specialised care indicates equivalence in outcomes including recurrence rate, cancer survival and quality of life (Cancer Research in Primary Care 2016).

Telehealth can enable efficient shared care and should be explored for all patients. Patients in some rural or remote locations may access specialists via Medicare Benefit Scheme funded telehealth consultations. General practitioners working in rural or remote locations should be aware of specialist multidisciplinary teams with facilities to reduce the travel burden and costs for patients.

Principle 2: Safe and quality care

Hospitals and health professionals are responsible for providing safe and quality care.

Health professionals need to have appropriate training and experience to undertake treatment for AL amyloidosis. Patients should be referred to an individual practitioner or service with appropriate expertise.

Safe and high-quality care is care provided by appropriately trained and credentialed health professionals who undertake regular quality reviews of their performance, contribute to regular audits of their care and are actively involved in continuing professional development. Hospitals and clinics must have the equipment, staff numbers, policies and procedures in place to support safe and high-quality care for cancer patients. Patients should be offered the safest options for care, which may include using telehealth (Cancer Australia 2020).

Hospital quality committees should ensure all health care is informed by evidence, and health professionals and health service managers (including executives) have a responsibility to evaluate and monitor their practice. Optimal care pathways provide a framework to help evaluate and monitor practice over time. Services should be routinely collecting relevant minimum datasets to support benchmarking, quality care and service improvement. Hospital committees and health professional peak bodies should be auditing this process (ACSQHC 2017; 2020).

The Australian Council on Health Standards <https://www.achs.org.au/> has created a set of indicators that helps hospitals conform to appropriate standards.

All new diagnoses should be reported, as appropriate, to the relevant state or territory cancer registry.

Patient-reported experience and outcome measures

Patient-reported experience measures (PREMs) and patient-reported outcome measures (PROMs) should be incorporated into routine cancer care.

PREMs are used to obtain patients' views and observations on aspects of healthcare services they have received (AIHW 2018). Patient experience data is collected for specific services and then relayed to service providers to instigate improvements in patient services (ACSQHC 2019b).

The Australian Hospital Patient Experience Question Set (AHPEQS) is a tool used to assess patient experiences of treatment and care in a private or public hospital. AHPEQS helps to improve the safety and quality of health care by allowing organisations to understand the patient's perspective (AIHW 2018; ACSQHC 2019b).

PROMs measure aspects of a person's health status such as symptoms, quality of life and needs and are collected directly from patients either online, via a smartphone or through paper-based means.

Collecting PROMs, and then instigating an appropriate clinical response, has been shown to prolong survival, reduce health system use and improve patients' quality of life. While there are many sets of PROMs questions that are relevant to any cancer patient, specific questions can be tailored to particular cancer types, populations or different phases of cancer care.

Principle 3: Multidisciplinary care

Multidisciplinary care is an integrated team approach that involves all relevant health professionals discussing all relevant treatment options and making joint recommendations about treatment and supportive care plans, taking into account the personal preferences of patients.

Multidisciplinary care improves patient outcomes. Cancer Australia's 'Principles of multidisciplinary care' provides a flexible definition, allowing services to vary implementation according to cancer type and the service location. The principles stipulate:

- a team approach that involves core disciplines that are integral to providing good care, including general practice, with input from other specialties as required
- communication among team members about treatment planning and plans for follow-up
- access to the full therapeutic range for all patients, regardless of geographical remoteness or size of institution
- care delivery in accordance with nationally agreed standards
- patient involvement in decisions about their care (Cancer Australia 2019a).

In addition to these principles, treatment teams should consider clinical trial participation for all eligible patients.

Multidisciplinary meetings, often called MDMs, should be based on the principles outlined above.

For more information on the principles of multidisciplinary care and the benefits of adopting a multidisciplinary approach, see All about multidisciplinary care on the Cancer Australia website <https://www.canceraustralia.gov.au/clinicians-hub/multidisciplinary-care/all-about-multidisciplinary-care>.

Principle 4: Supportive care

Supportive care is a vital part of any cancer treatment program. Supportive care deals with issues that emerge for patients, families and carers from the effects of the cancer diagnosis and its treatment. It is made up of all the services, information and resources patients may need to meet their physical, psychological, social, information and spiritual needs from the time of diagnosis.

Supportive care may be 'patient-defined' and based on unmet needs. It is a core component of evidence-based clinical care and its benefits are well established. All cancer patients and their carers should be formally supported and have access to understandable, relevant information about the medical, practical and emotional aspects of the cancer and its treatment (Fitch 2008). The wishes and needs of the patient, their family and their carers should determine the level of support provided. Supportive care is a standard or routine aspect of cancer care and the treatment team should make patients aware of this.

Supportive care should begin from the time of diagnosis and continue throughout the cancer pathway.

For health professionals, supportive care involves:

- screening and assessing patients and families for their supportive care needs
- providing patients with access to a range of multidisciplinary support services, groups and therapies designed to assist them to live with cancer and its treatment and optimise recovery
- optimising referral pathways to community support organisations (cancer-related non-government, not-for-profit and charities) that provide services to cancer survivors – these address many of the care-navigation, psychosocial and information needs of cancer survivors and those affected by cancer (Australian Cancer Survivorship Centre 2019)
- being aware of and delivering culturally appropriate care.

All members of the multidisciplinary team have a role in providing supportive care along the care pathway, with special attention at transition points.

Supportive care involves routinely and systematically assessing patients to determine their needs. Health professionals can use a variety of validated screening tools for this task (see box below). Clinical review and individual assessment are still required to ensure all patient concerns are identified.

More information

Visit the WeCan website <www.wecan.org.au> for information and resources on supportive care.

Validated screening tools

- National Comprehensive Cancer Network Distress Thermometer and Problem Checklist <www.nccn.org/patients/guidelines/content/PDF/distress-patient.pdf>
- Supportive Care Needs Assessment Tool for Indigenous People (SCNAT-IP) <www.scnatip.org>.

Key review points

The treatment team should assess patients for supportive care needs at these key stages:

- initial presentation or diagnosis (first three months)
- the beginning of treatment or a new phase of treatment
- change in prognosis
- if a patient is found to have a germline genetic mutation predisposing to cancer
- end of treatment
- throughout survivorship
- diagnosis of recurrence
- change in or development of new symptoms
- palliative care
- end-of-life care
- other time points based on clinical judgement.

The team also needs to decide whether the patient requires ongoing referral to supportive care services. Access to services can be through general practice–led chronic disease management plans, team care arrangements and mental health plans. Community support services also have a role to play.

See Appendices A, B and C for more information on supportive care and the specific needs of people that may arise.

Principle 5: Care coordination

Care coordination is the responsibility of every professional, both clinical and non-clinical, who works with patients, their families and carers.

Seamless care coordination is essential for patients to successfully navigate the complex health system. Care coordination is a comprehensive approach to achieving continuity of care for patients. It aims to ensure care is delivered in a systematic, connected and timely way that promotes efficiency and reduces the risk of duplication and over-servicing to meet the medical and personal needs of patients.

Care coordination includes:

- proactive and timely communication with patients, their families and carers
- treatment plans, survivorship care plans and/or advance care directives
- coordinated appointments to ensure timely diagnosis, treatment and survivorship care
- appropriate tests and results being available to the treating team so treatment decisions can be made
- medical records being available to all members of the treating team and at scheduled appointments
- translation or interpreter services arranged if the patient/carer is from a non-English-speaking background or has difficulty communicating due to a physical disability
- practical support such as transport, accommodation, advance care planning and financial support
- referral and access to supportive care
- access to clinical trials
- access to telehealth for people in rural and remote areas and for managing vulnerable patients.

Care coordination brings together different health professionals, teams and health services. It also encompasses MDMs, multidisciplinary assessment clinics, supportive care screening and assessment, referral practices, data collection, common protocols, information for patients and individual clinical treatment.

Care coordination should cross the acute and primary care interface and should aim to achieve consistency of care through clear communication, linkages and collaborative integrated care planning.

Care coordination can be facilitated through electronic health record management such as My Health Record. My Health Record is a secure online database that helps with data collection and care coordination (My Health Record 2019).

Formal care coordination through appointed care coordinators plays an important role in managing and supporting patients through the health system. The availability of dedicated care coordinators varies across states and territories according to the complexity of care required and local service capacity and resourcing.

Principle 6: Communication

Everyone employed in the healthcare system is responsible for ensuring the communication needs of patients, their families and carers are met.

Good and open communication is a key principle of care for cancer patients. This includes communication between oncology and primary care health professionals and with patients. General practitioners should be involved in care from the point of diagnosis, and patients should be encouraged to maintain a relationship with their general practitioner through all stages of cancer care. Communication should be regular and timely.

Attendance of a family member or carer at clinical appointments is beneficial for many patients, as the family member or carer can provide informational and emotional support. General practitioners and clinicians should encourage and support the involvement of family members and carers by providing an inclusive and supportive consultation environment (Laidsaar-Powell et al; 2018a). Laidsaar-Powell et al. provide evidence-based guidance on how to support family member or carer involvement in consultations (Laidsaar-Powell et al. 2018a, 2018b).

Every person with cancer will have different communication needs, including cultural and language differences. When anyone involved in treatment communicates with patients, they should be truthful and transparent but aware of cultural and psychological sensitivities. In communicating with patients, healthcare providers should undertake to:

- empower patients to be active in treatment discussions
- use professionally trained interpreters if required – for example, when communicating with people from culturally diverse backgrounds whose primary spoken language is not English and for people with a hearing impairment (visit the Translating and Interpreting Services website <www.tisnational.gov.au> for more information on interpreter and language services)
- use culturally sensitive and appropriate forms of communication for people from culturally diverse backgrounds and Aboriginal and Torres Strait Islander people, as appropriate
- provide appropriate information for people from culturally diverse backgrounds
- provide information on community-based supportive care services and resources to patients and their families and carers
- identify the patient's substitute treatment decision-maker to ensure they are involved in relevant discussions
- ensure patients, their families or their carers have the opportunity to ask questions
- seek consent before conveying information between health professionals or healthcare teams or with family and carers
- be respectful if a patient seeks a second opinion from another health professional
- ensure patients do not have to convey information between areas of care (it is the provider's and healthcare system's responsibility to transfer information between areas of care)
- communicate in plain language (avoiding complex medical terms and jargon)
- ensure information is communicated at a level relevant to the patient's health literacy and that of their families and carers (ACSQHC 2020)
- use tools, diagrams and aids as appropriate (Gilligan et al. 2017)
- ensure the patient is aware of how to access electronic patient information, where appropriate

- allow enough time for communication, especially when conveying complex or sensitive information such as an initial diagnosis
- check the patient's and/or their family or carer's understanding by asking the patient and/or their family or carer to say in their own words what has been conveyed.

Healthcare providers should also consider offering patients a question prompt list before a consultation and recordings or written summaries of their consultations afterwards. Question prompt lists are effective in improving communication and the psychological and cognitive outcomes of cancer patients. Recordings or summaries of key consultations improve patients' recall of information and satisfaction (Hack et al. 2012). Written care plans, treatment summaries, survivorship care plans and advance care directives are effective records and communication tools.

Communication skills training programs that use role-play to develop skills and observe patient interactions to provide feedback, should be available to health professionals at every level of practice (Gilligan et al. 2017).

Communication skills training programs and resources can be found on the following websites:

- Australian Commission on Safety and Quality in Healthcare, Communicating for safety resource portal <c4sportal.safetyandquality.gov.au>
- state and territory Cancer Councils <www.cancer.org.au/about-us/state-and-territory-councils> for the relevant council
- eviQ <education.eviq.org.au>
- VITAL talk <www.vitaltalk.org>.

Telehealth has become an increasingly acceptable alternative to face-to-face consultations. When using telehealth, the team must consider what is best for the patient, including the patient's preferences. A face-to-face consultation should be the first option, if it is safe, when delivering critical diagnosis information, a change in therapy or prescribing intensive treatment. If this is not an option, a video consultation should be considered, and the patient should be encouraged to have a support person with them to assist (Cancer Australia 2020).

Principle 7: Research and clinical trials

Research and clinical trials play an important role in establishing the efficacy and safety of diagnostic, prognostic and therapeutic interventions, as well as establishing the role of psychological, supportive care and palliative care interventions (Sjoquist et al. 2013).

Clinical trials are the foundation for improved cancer outcomes, allowing new treatments to be tested and offering patients access to potentially more effective therapies than otherwise available to them.

Clinical trials are available for multiple types of cancer and may be a valuable option for people with rare, difficult-to-treat conditions for which there may be limited evidence about how the condition is best treated or managed (Australian Clinical Trials 2015).

Treating specialists and multidisciplinary teams should be aware of or search for clinical trials that may be suitable for their patients. Specialists are encouraged to refer appropriate patients to other treating centres to participate in research or clinical trials at any stage of the care pathway and be willing to discuss the importance of informed consent and the pros and cons of participating in such trials. Any member of the multidisciplinary team can encourage cross-referral between clinical trials centres. Possible ineligibility to participate in a clinical trial should be discussed with the patient. Acknowledge disappointment and offer support in this instance.

Health services should strive to implement policies and procedures that facilitate equitable access to clinical trials for all patients, including culturally diverse patients, regional patients and those from Aboriginal or Torres Strait Islander communities.

The use of telehealth technology, such as the Australasian Tele-trial Model, hopes to improve access to trials for patients being treated in rural and regional areas (COSA 2016). The principles outlined in the Australasian Tele-trial Model are consistent with the National Teletrials Compendium (Australian Government Department of Health 2021b), which provides guidance on the national approach to teletrials that has been agreed by all states and territories. Clinical trials must adhere to the Good Clinical Practice quality standards, which provides assurance that the data and reported results are credible and accurate and that the rights, integrity and confidentiality of clinical trial participants are protected (Australian Government Department of Health 2021b).

Australian Cancer Trials is a national clinical trials database. It provides information on the latest clinical trials in cancer care, including trials that are recruiting new participants. Search for a trial <www.australiancancertrials.gov.au> via its website.

You can also search the Australian New Zealand Clinical Trials Registry <www.anzctr.org.au>, the Australasian Leukaemia and Lymphoma Group trials website <www.allg.org.au/clinical-trials-research/current-clinical-trials>, ClinTrial Refer <www.clintrialrefer.org.au> or ClinicalTrials.gov <www.clinicaltrials.gov> for international studies.

Education and training

Research and clinical trials provide an opportunity to educate health professionals who are in training. Cancer centres may be affiliated with teaching hospitals, universities or research groups to promote higher education or to develop the academic workforce, leading to more sustainable practice. Specialists should be encouraged to take up and retain active membership to professional societies and organisations that can assist with professional development opportunities.

Summary – optimal timeframes

Evidence-based guidelines, where they exist, should inform timeframes. Treatment teams need to recognise that shorter timeframes for appropriate consultations and treatment can promote a better experience for patients. Three steps in the pathway specify timeframes for care (Figure 3). They are designed to help patients understand the timeframes in which they can expect to be assessed and treated, and to help health services plan care delivery in accordance with expert-informed time parameters to meet the expectation of patients. These timeframes are based on expert advice from the AL amyloidosis Working Group.

Figure 3: Timeframes for care of AL amyloidosis

Step in pathway	Care point	Timeframe
Presentation, initial investigations and referral	Signs and symptoms	Presenting symptoms should be promptly assessed. If there are no severe symptoms or severe blood test abnormalities, work-up can be initiated in general practice.
	Initial investigations initiated by GP	Where a patient has moderate or severe symptoms, investigations should be completed **within 1 week**. In non-urgent cases the general practitioner should complete the investigations, review the patient and finalise the path of action **within 4 weeks**.
	Referral to specialist	The timing for specialist referral is guided by the severity and type of organ dysfunction. Heart failure, nephrotic syndrome or patients with deteriorating kidney function should be seen by the appropriate specialist **within 1 week**. For less severe cases the appropriate timeframe is **4 weeks**.
Diagnosis, staging and treatment planning	Diagnosis and staging	Diagnostic testing to type the amyloidosis should be initiated once amyloid has been identified on biopsy. Most investigations to characterise the haematopoietic clone and organ staging should be done **4 weeks before** starting treatment.
	Multidisciplinary team meeting and treatment planning	Consultation with an amyloidosis centre and/or multidisciplinary meeting and treatment planning should occur **before starting** any disease-directed therapy.
Treatment	Systemic therapy	Systemic therapy should start **within 4 weeks** of diagnosis, particularly if there is cardiac and/or renal disease.

Optimal care pathway

Seven steps of the optimal care pathway

Step 1: Prevention and early detection

Step 2: Presentation, initial investigations and referral

Step 3: Diagnosis, staging and treatment planning

Step 4: Treatment

Step 5: Care after initial treatment and recovery

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

Step 7: End-of-life care

AL amyloidosis arises from a clonal expansion of bone marrow plasma cells that produce abnormal immunoglobulin light chains. These light chains misfold and convert from their soluble states into highly organised fibrillar aggregates that deposit in tissues. AL amyloidosis most commonly affects the heart (~74%), kidney (~65%), peripheral nervous system (~20%), liver (~17%), gastrointestinal tract (~8%) and carpal tunnels, with variable involvement of other organs. Progressive infiltration leads to organ dysfunction and end-stage complications including restrictive cardiomyopathy and nephrotic syndrome.

The underlying plasma cell clone is typically subtle, with only a slightly increased number of bone marrow plasma cells. However, 8% of newly diagnosed patients with AL amyloidosis have symptomatic myeloma and 38% have more than 10% bone marrow plasmacytosis (levels seen with smouldering myeloma), while 54% have less than 10% bone marrow plasmacytosis (levels akin to monoclonal gammopathy of undetermined significance) (Kourelis et al. 2013). Occasionally the precursor protein may be produced by a low-grade B cell lymphoproliferative disorder.

AL amyloidosis is the second most common form of systemic amyloidosis (wild type ATTR amyloidosis is the most common type), with the incidence in Australia estimated to be approximately 10 cases per million persons per year. Most cases are diagnosed over the age of 40, and there is a slight male predominance (Wisniowski et al. 2019).

Is AL amyloidosis a blood cancer?

People commonly ask whether AL amyloidosis is a blood cancer. The amyloid deposits in AL amyloidosis are not cancerous, but the amyloid-forming protein is produced by an underlying bone marrow neoplasm that is usually benign (akin to monoclonal gammopathy of undetermined significance) but which can also be malignant (e.g. symptomatic myeloma). As such, while AL amyloidosis is not typically considered to be a cancer, it can be considered a blood cancer related disorder. Throughout this booklet the word ‘cancer’ can generally be taken to refer to the condition of AL amyloidosis.

The terminology seen in association with amyloidosis can cause confusion. Nomenclature recommendations from the International Society of Amyloidosis (Benson et al. 2020) have been included in the glossary of this OCP.

Step 1: Prevention and early detection

This step outlines recommendations for the prevention and early detection of AL amyloidosis.

Evidence shows that not smoking, avoiding or limiting alcohol intake, eating a healthy diet, maintaining a healthy body weight, being physically active, being sun smart and avoiding exposure to oncoviruses or carcinogens may help reduce cancer risk (Cancer Council Australia 2018).

1.1 Prevention

The causative factors of AL amyloidosis are not yet known, and there are currently no effective prevention strategies.

1.2 Risk factors

In order to develop AL amyloidosis patients must have:

- a monoclonal immunoglobulin light chain
- capacity for that monoclonal light chain to misfold and form amyloid fibrils. The risk factors for these two processes are outlined below.

1. A pre-existing monoclonal gammopathy:

The risk factors for developing the pre-existing monoclonal gammopathy that underlies AL amyloidosis are less well studied than in myeloma and MGUS (see myeloma OCP); however, they do include the following:

- **Age**: The risk of amyloidosis increases as a person gets older. For AL amyloidosis, most people diagnosed are older than 40, with a median age of 66 years.
- **Family history**: Very rare families with a genetic predisposition to AL amyloidosis have been described. Otherwise, an underlying genetic cause has not been identified, and there is no available screening test for genetic predisposition.
- **Gender**: AL amyloidosis is slightly more common in males than females (Wisniowski et al. 2019)
- Only a small proportion of patients with a monoclonal gammopathy develop AL amyloidosis (1% of MGUS followed for up to 50 years developed AL amyloidosis in one series; up to 15% of patients with symptomatic myeloma have overt, coexisting AL amyloidosis).

2. Structural changes to immunoglobulin light chains that make them more likely to misfold and form amyloid fibrils:

- Lambda light chains more commonly form amyloid (Perfetti et al. 2012), being the constituent amyloid protein in 80% of AL cases (Merlini et al. 2013).
- Somatic mutations (Oberti et al. 2017) acquired during clonal selection lead to alterations in the amino acid sequence of light chains and this, coupled with other poorly understood modifications, such as N-glycosylation (Kumar et al. 2019) affect the light chain molecular kinetics, making them more prone to form amyloid fibrils.

1.3 Risk reduction

While there is no evidence linking lifestyle changes to reduced risk of AL amyloidosis, it is important to encourage people to reduce modifiable risk factors for other types of cancer and health conditions. This includes providing advice on regular screening, skin checks, sun-safe behaviours (Kleinstern et al. 2016; 2020), preventing or reducing obesity and support to quit smoking.

1.3.1 Genetic family history screening

There is no evidence to support that either genetic testing or medical screening is warranted for family members of people with AL amyloidosis.

1.4 Early detection

Early detection of AL amyloidosis is difficult, with overt signs and symptoms often appearing at the end stages of disease.

However, for patients with a known diagnosis of a monoclonal gammopathy (MGUS, smouldering myeloma, multiple myeloma), a high index of clinical suspicion and careful assessment can facilitate early diagnosis of AL amyloidosis. Such assessments should include:

- clinical history and examination for symptoms and signs of cardiac, renal, hepatic, peripheral and autonomic nerve dysfunction or soft tissue involvement.
- non-invasive organ screening with creatinine, urea and electrolytes, urinary protein assessment, liver function tests, and a low threshold for checking the cardiac biomarker NT-ProBNP or BNP (Merlini et al. 2012).

1.4.1 Screening recommendations

There is no indication for screening for AL amyloidosis in the general population.

Step 2: Presentation, initial investigations and referral

This step outlines the process for the general practitioner to initiate the right investigations and refer to the appropriate specialist in a timely manner. The types of investigations the general practitioner undertakes will depend on many factors, including access to diagnostic tests, the availability of medical specialists and patient preferences.

2.1 Signs and symptoms

AL amyloidosis can present with many otherwise unexplained, non-specific symptoms (e.g. fatigue and weight loss) and/or blood test abnormalities, as well as end-organ complications. The symptoms and signs typically reflect the presence and extent of organ involvement by amyloidosis.

The most frequently affected organs are the heart, kidneys, gastrointestinal tract, nervous system and liver. Soft tissue involvement such as macroglossia and periorbital purpura are highly suggestive of AL amyloidosis. However, these latter signs are uncommon (representing approximately 10% of symptoms) and, more importantly, often appear late in the course of the disease when organ damage is advanced (Merlini et al. 2018).

In most patients more than one organ is affected; however, some patients may only have one affected organ. As the symptoms are variable and also observed in other more common conditions such as diabetes mellitus, the diagnosis of AL amyloidosis can easily be overlooked, which contributes to delayed diagnosis.

The following signs and symptoms or abnormalities on investigations can more specifically suggest AL amyloidosis:

- Cardiac:
  - ECG: low voltages in limb leads, poor R wave progression in chest lead, rhythm disturbances electrocardiography
  - echocardiography: a thick-walled heart (interventricular septum ≥ 12 mm) with preserved ejection fraction, restrictive cardiomyopathy pattern, enlarged left and right atria
  - persistently raised troponin.
- Renal:
  - proteinuria without other identified causes
  - however, all proteinuria needs to be monitored and, if the proteinuria is increasing, this needs to be investigated including in diabetics with proteinuria that appears worse than expected for their stage of diabetes.
- Neurological:
  - sensory or sensorimotor peripheral neuropathy, especially a rapidly progressive neuropathy
  - autonomic neuropathy characterised by:
    - > postural hypotension
    - > erectile dysfunction
    - > alternating diarrhoea and constipation, faecal incontinence
    - > gastroparesis – early fullness, bloating, regurgitation, vomiting
    - > nocturia, urinary retention, incontinence
  - carpal tunnel syndrome, especially if bilateral.

- Liver:
  - hepatomegaly with a normal appearance on ultrasound or CT imaging without other causes (e.g. fatty liver).
- Gastrointestinal:
  - malabsorption, weight loss, early satiety with upper intestinal involvement
  - diarrhoea, colic and bleeding with large bowel involvement.
- Soft tissue:
  - macroglossia, periorbital purpura, amyloid nail dystrophy.

2.1.1 Timeframe for general practitioner consultation

Presenting symptoms should be promptly assessed. If there are no severe symptoms or blood test abnormalities, work-up can be initiated in general practice.

2.2 Assessments by the general practitioner

The role of general practitioners is to suspect the patient has a disease that requires a detailed history and examination that may be followed by basic investigations to help identify the organ system(s) that require specialist assessment.

General practitioners and other primary care providers should have the following systems embedded in their daily practice:

- awareness that rare diseases (more than 10,000 individual diseases) affect 8% of Australians and may make up 10% of a general practitioner's workload (Elliott & Zurynski 2015). This is equivalent to diabetes or asthma. A higher index of suspicion for a rare disease is warranted, especially for symptoms that are not clear or cannot be fully explained by a provisional diagnosis
- awareness that AL amyloidosis is a multi-organ disease and can present with any of a long list of symptoms and signs including fatigue, near-syncope, postural hypotension, oedema, raccoon eyes, macroglossia, carpal tunnel syndrome, peripheral neuropathy, dyspnoea, weight loss, indigestion, renal impairment, proteinuria, hepatomegaly, jaw claudication and gastrointestinal bleeding (Vaxman et al. 2020).

Thus, the role of general practitioners in diagnosing AL amyloidosis is to:

- suspect that the patient has a serious disease
- take a detailed history
- conduct a comprehensive physical examination including routine urine testing strip (e.g. Labstix, Multistix)
- initiate basic investigations to identify organ involvement requiring specialist assessment.

Depending on the organ impairment, basic investigations might include:

- creatinine, urea and electrolytes and urine assessment for albuminuria and/or proteinuria
- liver function tests
- ECG and echocardiogram.

2.2.1 Timeframe for completing investigations

Where a patient has moderate or severe symptoms, investigations should be completed **within one week.**

In non-urgent cases the general practitioner should complete the investigations, review the patient and finalise the path of action **within four weeks**.

2.3 Initial referral

To further the diagnostic process, the general practitioner should refer the patient to an appropriate specialist (e.g. haematologist for a monoclonal gammopathy, cardiologist for heart failure, nephrologist for proteinuria, neurologist for peripheral neuropathy or a health service with the relevant specialty services).

Patients should be enabled to make informed decisions about their choice of specialist and health service. General practitioners should make referrals in consultation with the patient after considering the clinical care needed, cost implications (see referral options and informed financial consent on page 9), waiting periods, location and facilities, including discussing the patient's preference for health care through the public or the private system.

Referral for suspected or diagnosed AL amyloidosis should include the following essential information to accurately triage and categorise the level of clinical urgency:

- important psychosocial history and relevant medical history
- family history, current symptoms, medications and allergies
- results of current clinical investigations (imaging and pathology reports)
- results of all prior relevant investigations
- notification if an interpreter service is required.

Many services will reject incomplete referrals, so it is important that referrals comply with all relevant health service criteria.

If access is via online referral, a lack of a hard copy should not delay referral.

The specialist should provide timely communication to the general practitioner about the consultation and should notify the general practitioner if the patient does not attend appointments.

Aboriginal and Torres Strait Islander patients will need a culturally appropriate referral. To view the optimal care pathway for Aboriginal and Torres Strait Islander people with cancer and the corresponding quick reference guide, visit the Cancer Australia website <https://www.canceraustralia.gov.au/publications-and-resources/cancer-australia-publications/optimal-care-pathway-aboriginal-and-torres-strait-islander-people-cancer> and <https://www.canceraustralia.gov.au/publications-and-resources/cancer-australia-publications/optimal-care-pathway-aboriginal-and-torres-strait-islander-people-cancer-quick-reference-guide>.

Download the consumer resources – *Checking for cancer* and *Cancer* from the Cancer Australia website <https://www.canceraustralia.gov.au/publications-and-resources/cancer-australia-publications/checking-cancer-what-expect> and <https://www.canceraustralia.gov.au/publications-and-resources/cancer-australia-publications/cancer-what-expect>.

2.3.1 Timeframe for referring to a specialist

The timing for specialist referral is guided by the severity and type of organ dysfunction.

- Heart failure, nephrotic syndrome or deteriorating kidney function should be seen by the appropriate specialist **within one week**.
- For less severe cases to be seen by a specialist physician, the appropriate timeframe is **four weeks**.

2.4 Support and communication

2.4.1 Supportive care

The patient's general practitioner should consider an individualised supportive care assessment where appropriate to identify the needs of an individual, their carer and family. Refer to appropriate support services as required. See validated screening tools mentioned in Principle 4 'Supportive care'.

A number of specific needs may arise for patients at this time:

- From diagnosis patients should be offered psychosocial support to help them cope with any psychological distress (anxiety/depression, interpersonal problems and adjustment difficulties) that may arise from dealing with a potentially life-threatening diagnosis and organ failure. These issues can be magnified due to delayed diagnosis, with patients seeing a number of specialists over many months.
- Encourage and support patients to increase exercise levels (Cormie et al. 2018; Hayes et al. 2019).

For more information refer to the National Institute for Health and Care Excellence 2015 guidelines, *Suspected cancer: recognition and referral* < https://www.nice.org.uk/guidance/ng12>.

For additional information on supportive care and needs that may arise for different population groups, see Appendices A, B and C.

2.4.2 Communication with patients, carers and families

The general practitioner is responsible for:

- providing patients with information that clearly describes to whom they are being referred, the reason for referral and the expected timeframes for appointments
- requesting that patients notify them if the specialist has not been in contact within the expected timeframe
- considering referral options for patients living rurally or remotely
- supporting the patient while waiting for the specialist appointment (Cancer Council 13 11 20 and Leukaemia Foundation 1800 620 420 are available to act as a point of information and reassurance during the anxious period while further diagnostic information).

More information

Refer to Principle 6 'Communication' for communication skills training programs and resources.

Step 3: Diagnosis, staging and treatment planning

Step 3 outlines the process for confirming the diagnosis and stage of cancer and for planning subsequent treatment. The guiding principle is that interaction between appropriate multidisciplinary team members should determine the treatment plan.

3.1 Specialist diagnostic work-up

Suspecting an AL amyloidosis diagnosis

Generally, it will be a specialist who suspects a diagnosis of AL amyloidosis when investigating organ impairment.

Common clinical presentations, particularly if they occur concurrently, that raise the suspicion of amyloidosis include:

- heart failure with preserved ejection fraction
- nephrotic syndrome
- peripheral neuropathy, especially rapidly progressive neuropathy and/or autonomic neuropathy
- hepatomegaly with normal imaging appearance.

If AL amyloidosis is suspected, then a full monoclonal gammopathy screen with the following is indicated as, if these tests are all negative, a diagnosis of AL amyloidosis becomes extremely unlikely:

- serum protein electrophoresis and immunofixation
- urine electrophoresis and immunofixation
- serum free light chains.

A diagnosis of systemic AL amyloidosis requires a tissue biopsy:

- The gold standard for histological detection of amyloid deposits is the Congo red stain. In circumstances when Congo red staining is not definitive, alternate methods such as electron microscopy to detect typical non-branching, 8–12 nm diameter fibrils may be diagnostic.
- Biopsy of the involved organ has the highest sensitivity for diagnosing amyloidosis and so should be the biopsy site of choice. However, the site of biopsy can be a peripheral ‘distant’ screening tissue (abdominal fat aspirate, rectum, bone marrow, gingiva, salivary gland, etc.) but the sensitivity of these biopsies is significantly lower. If there is a high clinical suspicion of amyloidosis and the initial distant site screening biopsies are negative, then the clinically affected organ should be biopsied.

Once AL amyloidosis is suspected or confirmed by tissue biopsy, prompt referral to a haematologist or specialist centre is required to confirm that the amyloidosis is of the AL subtype, completion of staging investigations and treatment planning.

Confirming the AL subtype

- Every patient must have their amyloid precursor protein identified with a high level of confidence; correctly identifying the AL subtype is critical for management.
- Sometimes a second biopsy may be necessary to obtain enough material to confirm the AL subtype.
- Immunohistochemistry (IHC) at a centre with a high level of expertise in this area is recommended to confirm the amyloid constituent protein for all cases of systemic amyloidosis. Because of the limitations of immunohistochemical typing, biopsy review at an Australian Amyloidosis Network (aan.org.au) service is encouraged, especially in cases when the amyloid typed according to IHC does not match the clinical phenotype, or if there is significant residual doubt as to the interpretation of the IHC.
- Proteomic analysis of amyloid deposits in formalin-fixed paraffin embedded tissue biopsy samples by laser capture microdissection and tandem mass spectrometry is the gold standard test for amyloid subtyping.
- Genetic studies are sometimes needed to diagnose or exclude hereditary cases.
- The presence of amyloid deposition in a tissue biopsy with a monoclonal gammopathy in serum or urine testing is inadequate to confirm a diagnosis of the AL type of amyloidosis.
- The pattern of organ involvement is not diagnostic of the type of amyloidosis in itself because the various types of systemic amyloidosis often have overlapping clinical features.

Evaluating the plasma cell clone

- The underlying plasma cell clone (or less commonly, lymphoid clone) should be fully assessed at baseline via:
  - serum protein electrophoresis and immunofixation
  - serum kappa and lambda free light chains
  - urine protein electrophoresis and immunofixation
  - bone marrow aspirate and trephine including plasma cell enumeration, plasma cell and B cell clonality assessment and plasma cell fluorescence in situ hybridisation.
- Assessment for the presence of symptomatic myeloma (see the *Optimal care pathway for people with multiple myeloma)*.

Evaluating the extent and severity of organ involvement

Tests always indicated:

- Cardiac:
  - NT-ProBNP (or BNP)
  - hs-TnT or hsTnI (or cTnI or cTnT)
  - ECG
  - transthoracic echocardiogram with global longitudinal strain assessment.
- Renal:
  - 24-hour urine protein quantitation with a urine protein:creatinine ratio
  - serum urea, electrolytes & creatinine (UEC) with eGFR, and serum albumin.

- Hepatic:
  - liver span (clinically or imaging)
  - LFTs, especially ALP.
- Neuropathy:
  - clinical assessment
  - postural blood pressure.
- Endocrinopathy:
  - TFTs and fasting BSL.
- Coagulopathy:
  - APTT, PT/INR, +/- factor X.

Tests that are indicated in select cases:

- cardiac MRI
- cardiac amyloid bone scintigraphy (with Tc-99m PYP, DPD or HMDP)
- 24-hour Holter monitor
- nerve conduction studies
- respiratory function tests
- CT chest, abdomen and pelvis
- sex and/or adrenal hormone assessment.

3.1.1 Timeframe for completing investigations

If AL amyloidosis is suspected, diagnostic and staging investigations should be completed **within four weeks** of the first consultation by the haematologist or specialist treating centre, or sooner depending on clinical urgency.

3.2 Staging and prognostic assessment

Staging and prognostic assessment are critical elements in treatment planning. The key prognostic determinants in patients with AL amyloidosis are the presence and severity of cardiac involvement and the clonal plasma cell burden, measured by the difference between the involved and uninvolved serum free light chains. The baseline plasma cell burden, cardiac stage and presence and extent of organ involvement should be clearly documented in the patient's medical record.

There are three cardiac staging systems currently in use – two are based on cardiac biomarkers alone:

- European modification of the Mayo 2004 system (Wechalekar et al. 2013)
- Boston University 2019 system where the BNP is used instead of NT-ProBNP (Lilleness et al. 2019)
- a staging system based on both cardiac biomarkers and the Freelite serum free light chain assay (Kumar et al. 2012).

A renal staging system based on eGFR and 24-hour proteinuria can predict dialysis-free survival and is also recommended (Palladini et al. 2014).

3.3 Performance status

Patient performance status is a central factor in cancer care and should be clearly documented in the patient's medical record.

Performance status should be measured and recorded using an established scale such as the Karnofsky scale or the Eastern Cooperative Oncology Group (ECOG) scale.

3.4 Treatment planning

3.4.1 Key considerations beyond treatment recommendations

A number of factors should be considered at this stage:

- the patient's overall condition, life expectancy, personal preferences and decision-making capacity
- discussing the multidisciplinary team approach to care with the patient
- appropriate and timely referral to an MDM
- pregnancy and fertility (if applicable)
- support with travel and accommodation
- teleconferencing or videoconferencing as required.

3.4.2 Timing for multidisciplinary team planning

The multidisciplinary team should meet to discuss patients before initiating treatment so a treatment plan can be recommended and there can be early preparation for the post-treatment phase. The level of discussion may vary, depending on the patient's clinical and supportive care factors. Some patients with non-complex cancers may not be discussed by a multidisciplinary team; instead, the team may have treatment plan protocols that will be applied if the patient's case (cancer) meets the criteria. If patients are not discussed at an MDM, they should at least be named on the agenda for noting. The proposed treatment must be recorded in the patient's medical record and should be recorded in an MDM database where one exists.

Teams may agree on standard treatment protocols for non-complex care, facilitating patient review (by exception) and associated data capture.

Results of all relevant tests and access to images should be available for the MDM. Information about the patient's concerns, preferences and social and cultural circumstances should also be available.

3.4.3 Responsibilities of the multidisciplinary team

The multidisciplinary team requires administrative support in developing the agenda for the meeting, for collating patient information and to ensure appropriate expertise around the table to create an effective treatment plan for the patient. The MDM has a chair and multiple lead clinicians. Each patient case will be presented by a lead clinician (usually someone who has seen the patient before the MDM). In public hospital settings, the registrar or clinical fellow may take this role. A member of the team records the outcomes of the discussion and treatment plan in the patient history and ensures these details are communicated to the patient's general practitioner. The team should consider the patient's values, beliefs and cultural needs as appropriate to ensure the treatment plan is in line with these.

3.4.4 Members of the multidisciplinary team

The multidisciplinary team should be composed of the core disciplines that are integral to providing good care. Team membership should reflect both clinical and supportive aspects of care.

See Appendix E for a list of team members who may be included in the multidisciplinary team for AL amyloidosis.

Core members of the multidisciplinary team are expected to attend most MDMs either in person or remotely via virtual mechanisms. Additional expertise or specialist services may be required for some patients. An Aboriginal and Torres Strait Islander cultural expert should be considered for all patients who identify as Aboriginal or Torres Strait Islander

3.4.5 Responsibilities of individual team members

The general practitioner who made the referral is responsible for the patient until care is passed to another practitioner who is directly involved in planning the patient's care.

The general practitioner may play a number of roles in all stages of the cancer pathway including diagnosis, referral, treatment, shared follow-up care, post-treatment surveillance, coordination and continuity of care, as well as managing existing health issues and providing information and support to the patient, their family and carer.

A nominated contact person from the multidisciplinary team may be assigned responsibility for coordinating care in this phase. Care coordinators are responsible for ensuring there is continuity throughout the care process and coordination of all necessary care for a particular phase (COSA 2015). The care coordinator may change over the course of the pathway.

The lead clinician is responsible for overseeing the activity of the team and for implementing treatment within the multidisciplinary setting.

3.5 Research and clinical trials

Participation in clinical trials, patient registries and tissue banking, where available, is encouraged for patients with AL amyloidosis. Cross-referral between clinical trials centres should be encouraged to facilitate participation.

For more information visit:

- Cancer Australia <www.australiancancertrials.gov.au>
- Australian New Zealand Clinical Trials Registry <www.anzctr.org.au>
- Australasian Leukaemia and Lymphoma Group <https://www.allg.org.au/clinical-trials-research/current-clinical-trials>
- ClinTrial Refer <www.clintrialrefer.org.au>
- ClinicalTrials.gov <www.clinicaltrials.gov> for an international view.

3.6 Support and communication

3.6.1 Prehabilitation

Prehabilitation uses a multidisciplinary approach combining exercise, nutrition, and psychological strategies to prepare patients for the challenges of AL amyloidosis treatment such as systemic therapy. Team members may include anaesthetists, oncologists, surgeons, haematologists, clinical psychologists, exercise physiologists, physiotherapists, and dietitians, among others.

Patient performance status is a central factor in the care of patients with AL amyloidosis and should be frequently assessed. All patients should be screened for malnutrition using a validated tool, such as the Malnutrition Screening Tool (MST). The lead clinician may refer obese or malnourished patients to a dietitian preoperatively or before other treatments begin.

Patients who currently smoke should be encouraged to stop smoking before receiving treatment. This should include an offer of referral to Quitline in addition to smoking cessation pharmacotherapy if clinically appropriate.

Evidence indicates that patients who respond well to prehabilitation may have fewer complications after treatment. For example, those who were exercising before diagnosis and patients who use prehabilitation before starting treatment may improve their physical or psychological outcomes, or both, and this helps patients to function at a higher level throughout their cancer treatment (Cormie et al. 2017; Silver 2015).

For patients with AL amyloidosis, the multidisciplinary team should consider these specific prehabilitation assessments and interventions for treatment-related complications or major side effects:

- conducting a physical and psychological assessment to establish a baseline function level
- identifying impairments and providing targeted interventions to improve the patient's function level (Silver & Baima 2013)
- reviewing the patient's medication to ensure optimisation and to improve adherence to medicine used for comorbid conditions.

Following completion of primary treatment, rehabilitation programs have considerable potential to enhance physical function.

3.6.2 Fertility preservation and contraception

AL amyloidosis and its treatment may cause fertility problems. This will depend on the age of the patient, the organs involved and the treatment received. Infertility can range from difficulty having a child to the inability to have a child. Infertility after treatment may be temporary, lasting months to years, or permanent (AYA Cancer Fertility Preservation Guidance Working Group 2014).

Patients need to be advised about and potentially referred for discussion about fertility preservation **before** starting treatment and need advice about contraception **before, during and after** treatment. Patients and their family should be aware of the ongoing costs involved in optimising fertility. Fertility management may apply in both men and women. Fertility preservation options are different for men and women and the need for ongoing contraception applies to both men and women.

The potential for impaired fertility should be discussed and reinforced at different time points as appropriate throughout the diagnosis, treatment, surveillance and survivorship phases of care. These ongoing discussions will enable the patient and, if applicable, the family to make informed decisions. All discussions should be documented in the patient's medical record.

3.6.3 Supportive care

See validated screening tools mentioned in Principle 4 ‘Supportive care’.

A number of specific challenges and needs may arise for patients at this time:

- assistance for dealing with psychological and emotional distress while adjusting to the diagnosis and any initial period of observation; treatment phobias; existential concerns; stress; difficulties making treatment decisions; anxiety or depression or both; psychosexual issues such as potential loss of fertility and premature menopause; history of sexual abuse; and interpersonal problems
- diaries, reminders or other tools to aid with oral medication adherence
- management of physical symptoms such as pain and fatigue (Australian Adult Cancer Pain Management Guideline Working Party 2019)
- malnutrition or undernutrition, identified using a validated nutrition screening tool such as the MST (note that many patients with a high BMI [obese patients] may also be malnourished [WHO 2018])
- support for families or carers who are distressed with the patient’s cancer diagnosis
- specific spiritual needs that may benefit from the involvement of pastoral/spiritual care.

Additionally, palliative care may be required at this stage.

3.6.4 Supportive therapies

A number of specific challenges and needs may arise for patients at this time. Consider:

- optimising dental hygiene and ensuring restorative dental work is up to date
- support to cease smoking
- discussing fertility issues if relevant (see section 3.6.2)
- vaccinations for seasonal influenza, pneumococcal disease and COVID-19, ideally before starting any immunosuppressive therapy because subsequent responses are greatly impaired (Herishanu et al. 2021)
- correcting vitamin D deficiency (Molica et al. 2012; Shanafelt et al. 2011)
- immunoglobulin replacement therapy for patients with hypogammaglobulinemia and frequent infections. The National Blood Authority has information on the eligibility criteria for immunoglobulins <www.criteria.blood.gov.au/checkeligibility>.

For more information on supportive care and needs that may arise for different population groups, see Appendices A, B and C.

3.6.5 Communication with patients, carers and families

In discussion with the patient, the lead clinician should undertake the following:

- establish if the patient has a regular or preferred general practitioner and if the patient does not have one, then encourage them to find one
- provide written information appropriate to the health literacy of the patient about the diagnosis and treatment to the patient and carer and refer the patient to the Guide to best cancer care (consumer optimal care pathway) for AL amyloidosis, as well as to relevant websites and support groups such as:
  - Australian Amyloidosis Network <www.aan.org.au>
  - Myeloma Australia <www.myeloma.org.au>
  - Leukaemia Foundation <www.leukaemia.org.au>

- provide a treatment care plan including contact details for the treating team and information on when to call the hospital
- discuss a timeframe for diagnosis and treatment with the patient and carer
- discuss the benefits of multidisciplinary care and gain the patient's consent before presenting their case at an MDM
- provide brief advice and refer to Quitline (13 7848) for behavioural intervention if the patient currently smokes (or has recently quit), and prescribe smoking cessation pharmacotherapy, if clinically appropriate
- recommend an 'integrated approach' throughout treatment regarding nutrition, exercise and minimal or no alcohol consumption among other considerations
- communicate the benefits of continued engagement with primary care during treatment for managing comorbid disease, health promotion, care coordination and holistic care
- where appropriate, review fertility needs with the patient and refer for specialist fertility management (including fertility preservation, contraception, management during pregnancy and of future pregnancies)
- be open to and encourage discussion about the diagnosis, prognosis (if the patient wishes to know) and survivorship and palliative care while clarifying the patient's preferences and needs, personal and cultural beliefs and expectations, and their ability to comprehend the communication
- encourage the patient to participate in advance care planning including considering appointing one or more substitute decision-makers and completing an advance care directive to clearly document their treatment preferences. Each state and territory has different terminology and legislation surrounding advance care directives and substitute decision-makers.

3.6.6 Communication with the general practitioner

The lead clinician has these communication responsibilities:

- involving the general practitioner from the point of diagnosis
- ensuring regular and timely communication with the general practitioner about the diagnosis, treatment plan, any specific preventative measures and recommendations from MDMs and inviting them to participate in MDMs (consider using virtual mechanisms)
- supporting the role of general practice both during and after treatment
- discussing shared or team care arrangements with general practitioners or regional cancer specialists, or both, together with the patient.

More information

Refer to Principle 6 'Communication' for communication skills training programs and resources.

Step 4: Treatment

Step 4 describes the optimal treatments for AL amyloidosis, the training and experience required of the treating clinicians and the health service characteristics required for optimal cancer care.

All health services must have clinical governance systems that meet the following integral requirements:

- identifying safety and quality measures
- monitoring and reporting on performance and outcomes
- identifying areas for improvement in safety and quality (ACSQHC 2020).

Step 4 outlines the treatment options for AL amyloidosis. For detailed clinical information on treatment options refer to:

- Australian Guidelines <https://myeloma.org.au/wp-content/uploads/2021/11/MSAG-Clinical-Practice-Guidelines_Managemen-of-Systemic-Amyloidosis-Oct19.pdf>.

4.1 Treatment intent

The natural history of AL amyloidosis is complex, and treatment aims may change throughout the disease course. Effective treatments can result in removing amyloid deposition and recovering organ function. It's also important to note that the monoclonal gammopathy that underlies AL amyloidosis is generally not curable, and many patients will eventually relapse after each line of therapy.

The intent of treatment can be defined as one of the following:

- to obtain deep remission with the aim of reversing amyloid deposition and improving the function of organs affected by the amyloidosis
- to improve quality of life and/or longevity
- symptom palliation.

The treatment intent should be established in a multidisciplinary setting, documented in the patient's medical records and conveyed to the patient and carer as appropriate. The potential benefits need to be balanced against the morbidity and risks of treatment.

The lead clinician should discuss the advantages and disadvantages of each treatment and associated potential side effects with the patient and their carer or family before treatment consent is obtained and begins so the patient can make an informed decision. Supportive care services should also be considered during this decision-making process. Patients should be asked about their use of (current or intended) complementary therapies (see Appendix D).

Timeframes for starting treatment should be informed by evidence-based guidelines where they exist. The treatment team should recognise that shorter timeframes for appropriate consultations and treatment can promote a better experience for patients.

Initiate advance care planning discussions with patients before treatment begins (this could include appointing a substitute decision-maker and completing an advance care directive). Formally involving a palliative care team/service may benefit any patient, so it is important to know and respect each person's preference (Australian Government Department of Health 2021a).

4.2 Treatment options

Treatment will depend on several factors including age, stage, comorbidities, frailty and patient choice. This should be decided in conjunction with a multidisciplinary team. Prompt initiation of anti-plasma cell therapy is essential to preserve organ function and prolong life.

The aims of therapy are to:

- Reduce the pathogenic monoclonal light chains as rapidly and deeply as possible.
  - Absolute reductions in involved free light chain levels have been shown to correlate with improved survival, regardless of treatment strategy (Palladini et al. 2012).
  - Response is assessed by standardised criteria developed by the International Society for Amyloidosis. The optimal haematological endpoint after chemotherapy is complete response. A very good partial response (reduction in difference between the involved and uninvolved free light chain to < 40 mg/L) is also associated with improved likelihood of an organ response and improved overall survival. Because of the biological and analytical variability of the free light chain assay, care should be taken with decisions to change therapy based on haematological response when the baseline free light chain is low.
  - It should be noted that there is more than one assay available to measure free light chain levels and the results are not harmonised or interchangeable. The same free light chain assay should be used when comparing baseline and post-treatment free light chain measurements.
- Tailor therapy to the individual patient, anticipating toxicities of the various agents, taking into account the extent and degree of organ involvement.
  - An important caveat in managing AL amyloidosis is that patients are frailer and experience significantly higher treatment-related toxicity and mortality than patients with myeloma. Patients with AL amyloidosis commonly present with multi-organ dysfunction, impaired nutrition and limited physiological reserve that can make delivering effective chemoimmunotherapy extremely difficult. Therefore, treatment decisions should be made by carefully assessing patient-specific risks and benefits for each therapeutic strategy.
- Deliver response adapted therapy – that is, treatment adapted according to the haematological and organ response.
  - Monitoring of both haematological response and organ response is critical during therapy of AL amyloidosis.
  - Rather than a defined treatment course for all patients, therapy may need to be altered: early (after two or three cycles) in the event of suboptimal haematological response, or later if organ response is suboptimal despite a haematological response.
- Organ-specific supportive care (see section below).

4.2.1 Systemic therapy

Most patients diagnosed with AL amyloidosis will be suitable to receive systemic therapy.

Therapy can prevent an early death, improve organ function and prolong survival.

Induction therapy aims to rapidly reduce the amyloidogenic free light chain and is most commonly a regimen directed against an underlying plasma cell clone. Patients with IgM-related AL amyloidosis require therapy directed against a lymphoid clone, more akin to treating Waldenström macroglobulinemia (WM).

Induction therapy can include a combination of:

- proteasome inhibitors (PIs)
- chemotherapy
- monoclonal antibodies (mAbs)
- corticosteroids.

Consider clinical trials for eligible patients.

Timeframes for starting treatment

- Once the diagnosis of AL has been confirmed, treatment should be started as soon as possible. The timeframe for treatment depends on the organ involved. Treatment should start within two weeks for those with cardiac disease.

Training and experience required of the appropriate specialists

Haematologists/medical oncologists must have training and experience of this standard:

- Fellow of the Royal Australian College of Physicians (or equivalent)
- adequate training and experience that enables institutional credentialing and agreed scope of practice within this area (ACSQHC 2015).

Cancer nurses should have accredited training in these areas:

- anti-cancer treatment administration
- specialised nursing care for patients undergoing cancer treatments, including side effects and symptom management
- the handling and disposal of cytotoxic waste (ACSQHC 2020).

Systemic therapy should be prepared by a pharmacist whose background includes this experience:

- adequate training in systemic therapy medication, including dosing calculations according to protocols, formulations and/or preparations, such as those provided by eviQ <https://www.eviq.org.au/>.

If no haematologist/medical oncologist is locally available (e.g. regional or remote areas), some components of less complex therapies may be delivered by a general practitioner, general physician or nurse with training and experience that enables credentialing and agreed scope of practice within this area. This should be in line with a detailed treatment plan or agreed protocol, and with communication as agreed with the primary managing specialist or as clinically required.

The training and experience of the appropriate specialist should be documented.

Health service characteristics

To provide safe and quality care for patients having systemic therapy, health services should have these features:

- a clearly defined path to emergency care and advice after hours
- access to diagnostic pathology including basic haematology and biochemistry, and imaging
- access to appropriate molecular pathology (not required on site and can be through central specialist laboratory)
- access to cytotoxic drugs prepared in a pharmacy with appropriate facilities
- occupational health and safety guidelines regarding handling of cytotoxic drugs, including preparation, waste procedures and spill kits (eviQ 2019)
- guidelines and protocols to deliver treatment safely (including dealing with extravasation of drugs).

4.2.2 Supportive therapy

Patients with AL amyloidosis commonly present with multi-organ dysfunction, impaired nutrition and limited physiological reserve. Optimising organ support with specialist assistance is critical, particularly when embarking on chemotherapy. Patients should be treated as part of a multidisciplinary team. Attention should be paid to the symptoms that patients identify as having the greatest morbidity and symptomatic concern. The following should be considered:

Cardiac involvement

- Patients with cardiac involvement should be under the care of a cardiologist. Experience in managing cardiac amyloidosis is preferable.
- Patients with cardiac amyloidosis have a restrictive cardiomyopathy and, as such, management differs from common approaches to heart failure with reduced ejection fraction. Careful attention to fluid balance is required. Symptomatic cardiac failure may need to be managed with fluid restriction, a low-salt diet, loop diuretics and potassium-sparing diuretics. Some heart failure medications are usually not appropriate: ACE inhibitors and calcium-channel blockers should be avoided, particularly in patients with autonomic neuropathy, impaired renal function and baseline hypotension; digoxin is contraindicated for atrial fibrillation control; and beta-blockers are poorly tolerated, and if required for arrhythmia management, the lowest possible dose should be used.
- Arrhythmia management can be complex, and involvement of a cardiologist is required.

Renal involvement

- Patients with renal involvement should be under the care of a nephrologist. Experience in managing renal amyloidosis is preferable.
- Nephrotic syndrome should be managed supportively with diuretic therapy, salt and fluid restriction and, because of the ongoing protein loss, protein intake should not be restricted. Intravenous albumin replacement therapy may be considered. ACE inhibitors should be limited to patients who do not have significant cardiac disease or autonomic nervous system involvement. The risks and benefits of prophylactic anticoagulation in patients with nephrotic syndrome are complex and should be considered on an individual basis. This should be discussed with the treating haematologist.
- Renal replacement therapy should be considered in patients with end-stage kidney disease, taking into account age, the severity of other organ involvement and fitness for chemotherapy. Renal transplantation may be considered on an individual basis.

Peripheral and autonomic neuropathy

- It is preferable that patients with peripheral or autonomic neuropathy be monitored under the care of a neurologist.
- Peripheral neuropathy may require reduction or cessation of neuropathic chemotherapies. Neuropathic pain should be treated with medications including pregabalin, amitriptyline, gabapentin or duloxetine. Referral to a pain/palliative care team may be useful for pain management.
- Orthostatic hypotension may be managed by midodrine, fludrocortisone and conservative measures. Conservative measures include support stockings, bed elevation and fluid and salt optimisation. Patients may need to be screened for adrenal insufficiency.

Gastrointestinal involvement

- The assessment and optimisation of nutritional status is recommended in all patients. Referral to a dietitian to optimise supplementation could be considered for some patients.
- Domperidone can be particularly effective in patients with significant early satiety and nausea.
- Antimotility agents (e.g. loperamide or codeine) may be required for patients with significant diarrhoea.

Haematological system supportive care

- Venous thromboembolism prophylaxis should be considered on an individual patient basis.
- Patients with haemostatic abnormalities should be managed by the treating haematologist.
- Infection prophylaxis should be considered where indicated, including:
  - immunoglobulin replacement therapy for patients with frequent infections – see the National Blood Authority website <http://www.nba.gov.au> for criteria for the clinical use of intravenous immunoglobulin in Australia.
  - pharmaceutical prophylaxis including that against varicella zoster reactivation, and Pneumocystis *jirovecii* should follow institutional guidelines
  - vaccinations against hepatitis B, pneumococcus, influenza, COVID-19 and other pathogens that are deemed necessary because of epidemiological prevalence (live vaccines should be avoided).

4.2.3 Emerging therapy

AL therapies can be divided into those directed against the plasma cell clone, and those directed against the fibrils. The goal is to develop therapies that can target both the plasma cell clone and the fibrils. A range of biological, targeted and novel immunotherapeutic approaches are being used to treat AL amyloidosis. Efforts should be made to identify patients who may be eligible for clinical trials, particularly when PBS options are exhausted or unavailable.

Some emerging therapies include:

- anti-fibril therapies directed at clearing the amyloid deposits
- bispecific antibodies (T-cell engagers)
- antibody–drug conjugates
- novel monoclonal antibodies
- BCL2 inhibitors (these are particularly effective for patients with the cytogenetic lesion of t(11;14)).

4.3 Palliative care

Palliative care is a multidisciplinary approach to symptom management, psychosocial support and assistance in identifying care goals for patients with serious illness and their families.

Early referral to palliative care can improve the quality of life for people with cancer, improve caregiver outcomes and, in some cases may have survival benefits. (Haines 2011; Temel et al. 2010; Zimmermann et al. 2014). This is particularly true for cancers with poor prognosis.

The lead clinician should ensure patients receive timely and appropriate referral to palliative care services. Referral should be based on clinical need rather than prognosis. Emphasise the value of palliative care in improving symptom management, and quality of life to patients and their carers.

The '*Dying to Talk'* resource <www.health.gov.au/contacts/dying-to-talk> may help health professionals when initiating discussions with patients about future care needs (see 'More information'). Ensure that carers and families receive information, support and guidance about their role in palliative care (Palliative Care Australia 2018).

Patients, with support from their family or carer and treating team, should be encouraged to consider appointing a substitute decision-maker and to complete an advance care directive.

Refer to step 6 for a more detailed description of managing patients with relapsed, or progressive disease.

More information

These online resources are useful:

- Advance Care Planning Australia <www.advancecareplanning.org.au>
- Care Search <www.caresearch.com.au/Caresearch/>
- Dying to Talk <www.dyingtotalk.org.au>
- the Palliative Care resource kit <www.health.gov.au/health-topics/palliative-care>
- Palliative Care Australia (for patients and carers) <www.palliativecare.org.au>.

4.4 Research and clinical trials

Participation in clinical trials, patient registries and tissue banking, where available, is encouraged for patients with AL Amyloidosis. Many emerging treatments are only available on clinical trials that may require referral to certain trial centres.

For more information visit:

- Cancer Australia <www.australiancancertrials.gov.au>
- Australian New Zealand Clinical Trials Registry <www.anzctr.org.au>
- Australasian Leukaemia and Lymphoma Group <https://www.allg.org.au/clinical-trials-research/current-clinical-trial/>
- ClinTrial Refer <www.clintrialrefer.org.au>
- ClinicalTrials.gov <www.clinicaltrials.gov> for an international view

4.5 Support and communication

4.5.1 Supportive care

See validated screening tools mentioned in Principle 4 ‘Supportive care’.

A number of specific challenges and needs may arise for patients at this time:

- assistance for dealing with emotional and psychological issues, including body image concerns, fatigue, quitting smoking, traumatic experiences, existential anxiety, treatment phobias, anxiety/depression, interpersonal problems and sexuality concerns
- potential isolation from normal support networks, particularly for rural patients who are staying away from home for treatment
- decline in mobility or functional status as a result of treatment
- assistance with beginning or resuming regular exercise with referral to an exercise physiologist or physiotherapist (COSA 2018; Hayes et al. 2019).

Early involvement of general practitioners may lead to improved survivorship care following acute treatment. General practitioners can address many supportive care needs through good communication and clear guidance from the specialist team (Emery 2014).

Patients, carers and families may have these additional issues and needs:

- financial issues related to loss of income (through reduced capacity to work or loss of work) and additional expenses as a result of illness or treatment
- advance care planning, which may involve appointing a substitute decision-maker and completing an advance care directive
- legal issues (completing a will, care of dependent children) or making an insurance, superannuation or social security claim on the basis of terminal illness or permanent disability.

Cancer Council 13 11 20, Leukaemia Foundation 1800 620 420, Myeloma Australia 1800 693 566 and Australian Amyloidosis Network via website <www.aan.org.au> information and support lines can assist with information and referral to local support services.

4.5.2 Rehabilitation

Rehabilitation may be required at any point of the care pathway. If it is required before treatment, it is referred to as prehabilitation (see section 3.6.1).

All members of the multidisciplinary team have an important role in promoting rehabilitation. Team members may include occupational therapists, speech pathologists, dietitians, social workers, psychologists, physiotherapists, exercise physiologists and rehabilitation specialists.

To maximise the safety and therapeutic effect of exercise for people with AL amyloidosis, all team members should recommend that people with AL amyloidosis work towards achieving, and then maintaining, recommended levels of exercise and physical activity as per relevant guidelines. Exercise should be prescribed and delivered under the direction of an accredited exercise physiologist or physiotherapist with experience in AL amyloidosis care (Vardy et al. 2019). The focus of intervention from these health professionals is tailoring evidence-based exercise recommendations to the individual patient’s needs and abilities, with a focus on the patient transitioning to ongoing self-managed exercise.

Other issues that may need to be dealt with include managing amyloidosis-related fatigue, improving physical endurance, achieving independence in daily tasks, optimising nutritional intake, returning to work and ongoing adjustment to AL amyloidosis and its consequences. Referrals to dietitians, psychosocial support, return-to-work programs and community support organisations can help in managing these issues.

4.5.3 Communication with patients, carers and families

The lead or nominated clinician should take responsibility for these tasks:

- discussing treatment options with patients and carers, including the treatment intent, and expected outcomes, and providing a written version of the plan and any referrals
- providing patients and carers with information about the possible side effects of treatment, managing symptoms between active treatments, how to access care, self-management strategies and emergency contacts
- encouraging patients to use question prompt lists and audio recordings, and to have a support person present to aid informed decision making
- initiating a discussion about advance care planning and involving carers or family if the patient wishes.

4.5.4 Communication with the general practitioner

The general practitioner plays an important role in coordinating care for patients, including helping to manage side effects and other comorbidities, and offering support when patients have questions or worries. For most patients, simultaneous care provided by their general practitioner is very important.

The lead clinician, in discussion with the patient's general practitioner, should consider these points:

- the general practitioner's role in symptom management, supportive care and referral to local services
- using a chronic disease management plan and mental health care management plan
- how to ensure regular and timely two-way communication about:
  - the treatment plan, including intent and potential side effects
  - supportive and palliative care requirements
  - the patient's prognosis and their understanding of this
  - enrolment in research or clinical trials
  - changes in treatment or medications
  - the presence of an advance care directive or appointment of a substitute decision-maker
  - recommendations from the multidisciplinary team.

More information

Refer to Principle 6 'Communication' for communication skills training programs and resources.

Step 5: Care after initial treatment and recovery

The term 'cancer survivor' describes a person living with cancer, from the point of diagnosis until the end of life. Survivorship care in Australia has traditionally been provided to patients who have completed active treatment and are in the post-treatment phase. But there is now a shift to provide survivorship care and services from the point of diagnosis to improve cancer-related outcomes.

Survivors of AL amyloidosis may experience inferior quality of life and disease-related symptoms for up to five years after their diagnosis (Jefford et al. 2017). Survivors commonly report symptoms of distress, fear of recurrence, fatigue, obesity and sedentary lifestyle (Vardy et al. 2019).

Due to an ageing population and improvements in treatments and supportive care, the number of people surviving AL amyloidosis is increasing. International research shows there is an important need to focus on helping cancer survivors cope with life beyond their acute treatment. Survivors often face issues that are different from those experienced during active treatment for AL amyloidosis and may include a range of issues, as well as unmet needs that affect their quality of life (Lisy et al. 2019; Tan et al. 2019).

Physical, emotional and psychological issues include fear of amyloidosis recurrence, amyloidosis-related fatigue, pain, distress, anxiety, depression, cognitive changes and sleep issues (Lisy et al. 2019). Late effects may occur months or years later and depend on the type of cancer treatment. Survivors and their carers may experience impacted relationships and practical issues including difficulties with return to work or study and financial hardship. They may also experience changes to sex and intimacy. Fertility, contraception and pregnancy care after treatment may require specialist input.

The Institute of Medicine, in its report *From cancer patient to cancer survivor: Lost in transition*, describes the essential components of survivorship care listed in the paragraph above, including interventions and surveillance mechanisms to manage the issues a cancer survivor may face (Hewitt et al. 2006). Access to a range of health professions may be required including physiotherapy, occupational therapy, social work, dietetics, clinical psychology, fertility and palliative care. Coordinating care between all providers is essential to ensure the patient's needs are met.

Cancer survivors are more likely than the general population to have and/or develop comorbidities (Vijayvergia & Denlinger 2015). Health professionals should support survivors to self-manage their own health needs and to make informed decisions about lifestyle behaviours that promote wellness and improve their quality of life (Australian Cancer Survivorship Centre 2010; Cancer Australia 2017).

5.1 Transition from initial treatment from AL amyloidosis

The transition from active treatment to post-treatment care is critical to long-term health. In some cases, people will need ongoing hospital-based care, and in other cases a shared follow-up care arrangement with their general practitioner may be appropriate. This will vary depending on the type and stage of AL amyloidosis and needs to be planned.

Shared follow-up care involves the joint participation of specialists and general practitioners in the planned delivery of follow-up and survivorship care. A shared care plan is developed that outlines the responsibilities of members of the care team, the follow-up schedule, triggers for review, plans for rapid access into each setting and agreement regarding format, frequency and triggers for

communication.

After completing initial treatment, a designated member of the multidisciplinary team (most commonly nursing or medical staff involved in the patient's care) should provide the patient with a needs assessment and treatment summary and develop a survivorship care plan in conjunction with the patient. This should include a comprehensive list of issues identified by all members of the multidisciplinary team involved in the patient's care and by the patient. These documents are key resources for the patient and their healthcare providers and can be used to improve communication and care coordination.

The treatment summary should cover, but is not limited to:

- the diagnostic tests performed and results
- diagnosis including stage, prognostic or severity score
- disease characteristics
- treatment received (types and dates)
- current toxicities (severity, management and expected outcomes)
- interventions and treatment plans from other health providers
- potential long-term and late effects of treatment
- supportive care services provided
- follow-up schedule
- contact information for key healthcare providers.

5.2 Follow-up care

Because AL amyloidosis is an ongoing condition, typically with relapse after each line of therapy, patients are rarely discharged permanently from specialist or MDM care. Follow-up care focuses on supportive care and disease surveillance.

After treatment, patients are often left with ongoing symptoms from organ failure with only a proportion going on to organ recovery from endogenous macrophage clearance of the amyloid deposits. This is a slow process. Patients may also have ongoing symptoms from treatment toxicity such as neuropathy from bortezomib.

Responsibility for follow-up care should be discussed, agreed upon, shared and well communicated between the patient, their lead clinician, other specialists and the general practitioner. The patient should be informed and guided with how their care is managed in an MDM. The patient should be empowered to guide their care plan.

Disease surveillance consists of regular testing of the monoclonal gammopathy and non-invasive testing for organ response and progression.

Evidence comparing shared follow-up care and specialised care indicates equivalence in outcomes including recurrence rate, cancer survival and quality of life (Cancer Research in Primary Care 2016).

Ongoing communication between healthcare providers involved in care and a clear understanding of roles and responsibilities is key to effective survivorship care.

In particular circumstances, other models of post-treatment care can be safely and effectively provided such as nurse-led models of care (Monterosso et al. 2019). Other models of post-treatment care can be provided in these locations or by these health professionals:

- in a shared care setting
- in a general practice setting
- by non-medical staff
- by allied health or nurses
- in a non-face-to-face setting (e.g. by telehealth).

A designated member of the team should document the agreed survivorship care plan. The survivorship care plan should support wellness and have a strong emphasis on healthy lifestyle changes such as a balanced diet, a non-sedentary lifestyle, weight management and a mix of aerobic and resistance exercise (COSA 2018; Hayes et al. 2019).

This survivorship care plan should also cover, but is not limited to:

- what medical follow-up is required (screening and assessment for medical and psychosocial effects)
- model of post-treatment care, the health professional providing care and where it will be delivered
- care plans from other health providers to manage the consequences of AL amyloidosis and AL amyloidosis treatment
- wellbeing, primary and secondary prevention health recommendations that align with chronic disease management principles
- rehabilitation recommendations
- available support services
- a process for rapid re-entry to specialist medical services for suspected recurrence
- recommended vaccination schedule.

Survivors generally need regular follow-up, often indefinitely after treatment finishes. The survivorship care plan therefore may need to be updated to reflect changes in the patient's clinical and psychosocial status and needs.

Processes for rapid re-entry to hospital care should be documented and communicated to the patient and relevant stakeholders.

Care in the post-treatment phase is driven by predicted risks (e.g. the risk of recurrence, developing late effects of treatment and psychological issues) as well as individual clinical and supportive care needs. It is important that post-treatment care is based on evidence and is consistent with guidelines. Not all people will require ongoing tests or clinical review and may be discharged to general practice follow-up.

The lead clinician should discuss (and general practitioner reinforce) options for follow-up at the start and end of treatment. It is critical for optimal aftercare that the designated member of the treatment team educates the patient about the symptoms of recurrence.

General practitioners (including nurses) can:

- connect patients to local community services and programs
- manage long-term and late effects
- manage comorbidities
- provide wellbeing information and advice to promote self-management
- screen for cancer and non-cancerous conditions
- deliver recommended vaccinations.

More information

Templates and other resources to help with developing treatment summaries and survivorship care plans are available from these organisations:

- Australian Cancer Survivorship Centre
- Cancer Australia – Principles of cancer survivorship
- Cancer Council Australia and states and territories
- Clinical Oncology Society of Australia – Model of survivorship care
- eviQ – Cancer survivorship: introductory course
- MyCarePlan.org.au
- South Australian Cancer Service – Statewide Survivorship Framework resources
- American Society of Clinical Oncology – guidelines.

5.2.1 Preventing recurrence

People frequently ask if there is anything else they can do to reduce the risk of AL amyloidosis. Not smoking, eating a healthy diet, being sun smart, avoiding or limiting alcohol intake, being physically active and maintaining a healthy body weight may help reduce the risk of a second primary cancer. However, none of these factors have been shown to affect the risk of AL amyloidosis recurrence or progression.

Encourage and support all cancer survivors to reduce modifiable risk factors for other cancers and chronic diseases. Ongoing coordination of care between providers should also deal with any comorbidities, particularly ongoing complex and life-threatening comorbid conditions.

5.3 Research and clinical trials

Support cancer survivors to participate in research or clinical trials where they are available and appropriate. These might include studies to understand survivors' issues, to better manage treatment side effects, improve long-term immune function or to improve models of care and quality of life.

For more information visit:

- Cancer Australia <www.australiancancertrials.gov.au>
- Australian New Zealand Clinical Trials Registry <www.anzctr.org.au>
- Australasian Leukaemia and Lymphoma Group <https://www.allg.org.au/clinical-trials-research/current-clinical-trials>
- ClinTrial Refer <www.clintrialrefer.org.au>
- ClinicalTrials.gov <www.clinicaltrials.gov> for an international view.

5.4 Support and communication

5.4.1 Supportive care

See validated screening tools mentioned in Principle 4 'Supportive care'. Additionally, the 'Cancer Survivors Unmet Needs (CaSun)' is another validated screening tool that may help health professionals to identify the unmet needs of patients during survivorship <www.headwayhealth.com.au/wp-content/uploads/2014/01/Manual-for-the-CASUN-and-CaSPUN-questionnaires.pdf>

A number of specific challenges and needs may arise for survivors:

- financial and employment issues (e.g. loss of income and assistance with returning to work, and the cost of treatment, travel and accommodation)
- appointing a substitute decision-maker and completing an advance care directive
- legal issues such as completing a will.

Being a rare disease, it can be difficult to access appropriate supportive care. Psychological help and education can help the patient and caregiver to cope with the demands of this disease and its treatment. Resources such the AAN, Myeloma Australia and the Leukaemia Foundation can be useful agencies as well as the general practitioner, specialised nursing support and psychology and counselling services (Taylor et al. 2021).

5.4.2 Rehabilitation and recovery

Rehabilitation may be required at any point of the care pathway from the pre-treatment phase through to disease-free survival and palliative care (Cormie et al. 2017).

Issues that may need to be dealt with include managing amyloidosis-related fatigue, coping with cognitive changes, improving physical endurance, achieving independence in daily tasks, returning to study or work and ongoing adjustment to AL amyloidosis and its consequences.

Exercise is a safe and effective intervention that improves the physical and emotional health and wellbeing of AL amyloidosis patients. Exercise should be embedded as part of standard practice in amyloidosis care and be viewed as an adjunct therapy that helps counteract the adverse effects of AL amyloidosis and its treatment.

Cancer survivors may find referral to specific cancer rehabilitation, optimisation programs or community-based rehabilitation appropriate and beneficial. Other options include referral to allied health supports through team care arrangements and mental health plans. Some community support organisations (cancer-related non-government, not-for-profit and charities) provide services to cancer survivors.

5.4.3 Communication with patients, carers and families

The lead clinician (themselves or by delegation) should take responsibility for these tasks:

- explaining the model of post-treatment care and the roles of health professionals involved in post-treatment care including the role of general practice
- explaining the treatment summary and follow-up care plan
- discussing the development of a shared follow-up and survivorship care plan where a model of shared follow-up care has been agreed
- discussing how to manage any of the physical, psychological or emotional issues identified
- providing information on the signs and symptoms of recurrent disease
- providing a survivorship care plan with information on secondary prevention and healthy living
- providing contact details of the care team involved
- providing clear information about the role and benefits of palliative care and advance care planning.

5.4.4 Communication with the general practitioner

The lead clinician should ensure regular, timely, two-way communication with the general practitioner about:

- the patient's progress
- the follow-up care plan including recommended vaccinations
- potential late effects
- supportive and palliative care requirements
- any shared care arrangements
- clarification of various roles in patient care
- a process for rapid re-entry to medical services for patients with suspected recurrence or if there are other concerns.

More information

Refer to Principle 6 'Communication' for communication skills training programs and resources.

Step 6: Managing relapsed, residual or progressive disease

Patients who present with progressive disease should be managed by a multidisciplinary team and offered timely referral to appropriate physical, practical and emotional support.

Step 6 is concerned with managing relapsed, residual or progressive disease.

The time to relapse after initial treatment is variable and so it is important to continue to monitor patients.

6.1 Signs and symptoms of relapsed or progressive disease

Most patients with AL amyloidosis will relapse after responding to initial treatment. Access to the best available therapies, including clinical trials and treatment with a multidisciplinary team, are crucial to achieving the best outcomes for relapsed disease.

Signs and symptoms will depend on whether there is progressive organ dysfunction resulting from recurrent light chain amyloid production and deposition. Patients may be asymptomatic or have no worsening of their symptoms when there is free light chain progression but without worsening of their organ involvement by amyloidosis.

Retreatment is timed to prevent further amyloid organ deposition and so this is started when there is enough production of amyloidogenic light chain to place a patient at risk of further amyloid production and organ injury. A switch in treatment regimen is also indicated if a suboptimal haematological response is observed – that is, the amyloidogenic light chain has not been reduced enough to result in improvement in organ function.

Hence, the decision to retreat or switch treatment is based on the following haematological or serum free light chain parameters rather than waiting for new symptoms or signs of organ damage:

- progressive rise in serum free light chains or paraprotein (or so-called haematological relapse/ progression)
- suboptimal haematological response.

Unfortunately, sometimes new or worsening organ dysfunction can be the first indicator of relapse or progression, particularly because haematological progression can sometimes be difficult to assess (e.g. in renal failure). It is therefore important to survey for both haematological as well as organ progression with clinical and pathology parameters.

For specific criteria of suboptimal response, haematological and organ progression, surveillance recommendations see the MSAG website <https://myeloma.org.au/health-professional-resources/>.

6.2 Managing relapsed or progressive disease

Managing relapsed or progressive disease is complex and should therefore involve all the appropriate specialties in a multidisciplinary team including palliative care where appropriate. From the time of diagnosis, the team should offer patients appropriate psychosocial care, supportive care, advance care planning and symptom-related interventions as part of their routine care. The approach should be personalised to meet the patient's individual needs, values and preferences. The full complement of supportive care measures as described throughout the optimal care pathway and in Appendices A, B and C should be offered to assist patients and their families and carers to cope. These measures should be updated as the patient's circumstances change.

Survivorship care should be considered and offered at an early stage. Many people live with advanced AL amyloidosis for many years. As survival is improving in many patients, survivorship issues should be considered as part of routine care. Health professionals should therefore be ready to change and adapt treatment strategies according to disease status, prior treatment tolerance and toxicities and the patient's quality of life, in addition to the patient's priorities and life plans.

6.3 Multidisciplinary team

If there is an indication that a patient's AL amyloidosis has returned, care should be provided under the guidance of a treating specialist. Each patient should be evaluated to determine if referral to the original multidisciplinary team is necessary. Access to the best available therapies, including clinical trials, as well as treatment overseen by a multidisciplinary team, are crucial to achieving the best outcomes for anyone with relapsed or progressive disease. The multidisciplinary team may include new members such as palliative care specialists.

6.4 Treatment

Managing relapsed or progressive AL amyloidosis is complex and should therefore involve all the appropriate specialties in a multidisciplinary team including palliative care where appropriate.

From the time of diagnosis, the team should offer patients appropriate psychosocial care, supportive care, advance care planning and symptom-related interventions as part of their routine care. The approach should be personalised to meet the patient's individual needs, values and preferences. The full complement of supportive care measures as described throughout the optimal care pathway should be offered to patients, their families and carers. These measures should be updated as the patient's circumstances change.

Treatment options

There are other therapeutic regimens to treat relapsed or progressive AL amyloidosis or for those who require a better response. In relapsed disease, treatment should be initiated when there is haematological progression or increasing production of amyloidogenic light chain prior to developing further organ damage by amyloid deposition. The choice of salvage regimen should consider patient factors (age and frailty), disease factors (tempo of relapse, presence and degree of organ involvement), prior treatment-related factors (responsiveness and side effects to prior treatment regimens) and patient preference.

In managing people with relapsed AL amyloidosis, enrolment into a clinical trial is the first preference. If no clinical trial is available, treatment may include:

- using a different drug regimen that contains drugs of a different class or a different drug of the same class
- retreatment with a previous regimen
- autologous stem cell transplantation in selected patients
- supportive and/or palliative care.

Refer to the updated MSAG *Clinical practice guideline: AL amyloidosis* <https://myeloma.org.au/wp-content/uploads/2017/10/AL-MSAG-guidelines.pdf> for managing relapsed AL amyloidosis.

The potential goals of treatment should be discussed, respecting the patient's cultural values. Wherever possible, written information should be provided.

Encourage early referral to clinical trials or accepting an invitation to participate in research.

6.5 Advance care planning

Advance care planning is important for all patients with a diagnosis of AL amyloidosis but especially those with advanced disease. Patients should be encouraged to think and talk about their healthcare values and preferences with family or carers, appoint a substitute decision-maker and consider developing an advance care directive to convey their preferences for future health care in the event they become unable to communicate their wishes (Australian Government Department of Health 2021a).

More information

Refer to section 4.3 ‘More information’ for links to resources.

Refer patients and carers to Advance Care Planning Australia <www.advancecareplanning.org.au> or to the Advance Care Planning National Phone Advisory Service on 1300 208 582.

6.6 Palliative care

Early referral to palliative care can improve the quality of life for people with cancer and in some cases may be associated with survival benefits (Haines 2011; Temel et al. 2010; Zimmermann et al. 2014). The treatment team should emphasise the value of palliative care in improving symptom management and quality of life to patients and their carers. Refer to section 4.3 for more detailed information.

The lead clinician should ensure timely and appropriate referral to palliative care services. Referral to palliative care services should be based on the patient’s need and potential for benefit, not prognosis.

More information

Refer to the end of section 4.3 ‘Palliative care’ for links to resources.

6.7 Research and clinical trials

The treatment team should support the patient to take part in research and clinical trials where available and appropriate.

For more information visit:

- Cancer Australia <www.australiancancertrials.gov.au>
- Australian New Zealand Clinical Trials Registry <www.anzctr.org.au>
- Australasian Leukaemia and Lymphoma Group <www.allg.org.au/clinical-trials-research/current-clinical-trials>
- ClinTrial Refer <www.clintrialrefer.org.au>
- ClinicalTrials.gov <www.clinicaltrials.gov> for an international view.

6.8 Support and communication

6.8.1 Supportive care

See validated screening tools mentioned in Principle 4 ‘Supportive care’.

A number of specific challenges and needs may arise at this time for patients:

- assistance for dealing with emotional and psychological distress resulting from fear of death or dying, existential concerns, anticipatory grief, communicating wishes to loved ones, interpersonal problems and sexuality concerns
- potential isolation from normal support networks, particularly for rural patients who are staying away from home for treatment
- cognitive changes as a result of treatment and disease progression such as altered memory, attention and concentration (a patient may appoint someone to make medical, financial and legal decisions on their behalf – a substitute decision-maker – before and in case they experience cognitive decline)
- management of physical symptoms (please see section above 5.4.2 Supportive therapies for more information)
- decline in mobility or functional status as a result of recurrent disease and treatments (referral to physiotherapy or occupational therapy may be required)
- coping with hair loss and changes in physical appearance (refer to the Look Good, Feel Better program – see ‘Resource list’
- appointing a substitute decision-maker and completing an advance care directive
- financial issues as a result of disease recurrence such as gaining early access to superannuation and insurance
- legal issues (completing a will, care of dependent children) and making an insurance, superannuation or social security claim on the basis of terminal illness or permanent disability.

6.8.2 Rehabilitation

Rehabilitation may be required at any point of the care pathway, from preparing for treatment through to palliative care. Issues that may need to be dealt with include managing amyloidosis-related fatigue, improving physical endurance, achieving independence in daily tasks, returning to work and ongoing adjustment to AL amyloidosis and its consequences.

Exercise is a safe and effective intervention that improves the physical and emotional health and wellbeing of amyloidosis patients. Exercise should be embedded as part of standard practice in amyloidosis care and be viewed as an adjunct therapy that helps counteract the adverse effects of AL amyloidosis and its treatment.

6.8.3 Communication with patients, carers and families

The lead clinician should ensure there is adequate discussion with patients and carers about the diagnosis and recommended treatment, including treatment intent and possible outcomes, likely adverse effects and the supportive care options available.

More information

Refer to Principle 6 ‘Communication’ for communication skills training programs and resources.

Step 7: End-of-life care

Step 7 is concerned with maintaining the patient's quality of life and meeting their health and supportive care needs as they approach the end of life, as well as the needs of their family and carers.

Some patients with advanced AL amyloidosis will reach a time when active treatment is no longer appropriate. The team needs to share the principles of a palliative approach to care when making decisions with the patient and their family or carer. End-of-life care is appropriate when the patient's symptoms are increasing, and functional status is declining.

7.1 Multidisciplinary palliative care

If the treatment team does not include a palliative care member, the lead clinician should consider referring the patient to palliative care services, with the general practitioner's engagement. This may include inpatient palliative unit access (as required).

The multidisciplinary team may consider seeking additional expertise from these professionals:

- clinical psychologist
- clinical nurse specialist or practitioner
- social worker
- palliative medicine specialist
- pain specialist
- pastoral or spiritual carer
- bereavement counsellor
- music therapist
- art therapist
- cultural expert.

The team might also recommend that patients access these services:

- home- and community-based care
- specialist community palliative care workers
- community nursing.

If the patient does not already have an advance care directive in place, a designated member of the treatment team should encourage them to develop one in collaboration with their family or carer (Australian Government Department of Health 2021a).

It is essential for the treatment team to consider the appropriate place of care, the patient's preferred place of death and the support needed for the patient, their family and carers.

The treatment team should also ensure that carers and families receive the information, support and guidance about their role according to their needs and wishes (Palliative Care Australia 2018).

More information

The treatment team can refer patients and carers to these resources:

- Palliative Care Australia <www.palliativecare.org.au>
- Advance Care Planning Australia <www.advancecareplanning.org.au>
- or to Advance Care Planning Australia's National Advisory Service on 1300 208 582.

7.2 Research and clinical trials

Clinical trials may help improve palliative care and in managing a patient's symptoms of advanced cancer (Cancer Council Victoria 2019). The treatment team should support the patient to participate in research and clinical trials where available and appropriate.

For more information visit:

- Cancer Australia <www.australiancancertrials.gov.au>
- Australian New Zealand Clinical Trials Registry <www.anzctr.org.au>
- Australasian Leukaemia and Lymphoma Group <www.allg.org.au/clinical-trials-research/current-clinical-trials>
- ClinTrial Refer <www.clintrialrefer.org.au>
- ClinicalTrials.gov <www.clinicaltrials.gov> for an international view.

7.3 Support and communication

7.3.1 Supportive care

See validated screening tools mentioned in Principle 4 'Supportive care'.

A number of specific challenges and needs may arise for patients at this time:

- assistance for dealing with emotional and psychological distress from anticipatory grief, fear of death or dying, anxiety/depression and interpersonal problems
- management of physical symptoms including (please see section above 5.4.2 Supportive therapies for more information)
- decline in mobility or functional status, affecting the patient's discharge destination (a referral to physiotherapy, exercise physiology, occupational therapy or social work may be needed)
- appointing a substitute decision-maker and completing an advance care directive
- legal issues (completing a will, care of dependent children) and making an insurance, superannuation or social security claim on the basis of terminal illness or permanent disability
- specific support for families where a parent is dying and will leave behind bereaved children or adolescents, creating special family needs
- arranging a funeral.

These services and resources can help:

- referral to 13 11 20 for Cancer Council Australia's Pro Bono Program for free legal, financial, small business accounting and workplace assistance (subject to a means test)
- *Sad news sorry business* (Queensland Health 2015) for the specific needs of Aboriginal and Torres Strait Islander people.

For more information on supportive care and needs that may arise for different population groups, see Appendices A, B and C.

7.3.2 Communication with patients, carers and families

The lead clinician is responsible for:

- being open to and encouraging discussion with the patient about the expected disease course, considering the patient's personal and cultural beliefs and expectations
- discussing palliative care options, including inpatient and community-based services as well as dying at home and subsequent arrangements
- providing the patient and carer with the contact details of a palliative care service
- referring the patient to palliative care in the community according to the carer's wishes.

7.3.3 Communication with the general practitioner

The lead clinician should discuss end-of-life care planning to ensure the patient's needs and goals are met in the appropriate environment. The patient's general practitioner should be kept fully informed and involved in major developments in the patient's illness path.

More information

For support with communication skills and training programs, see these sources:

- *Sad news sorry business* <https://www.health.qld.gov.au/__data/assets/pdf_file/0023/151736/sorry_business.pdf>
- Principle 6 'Communication'.

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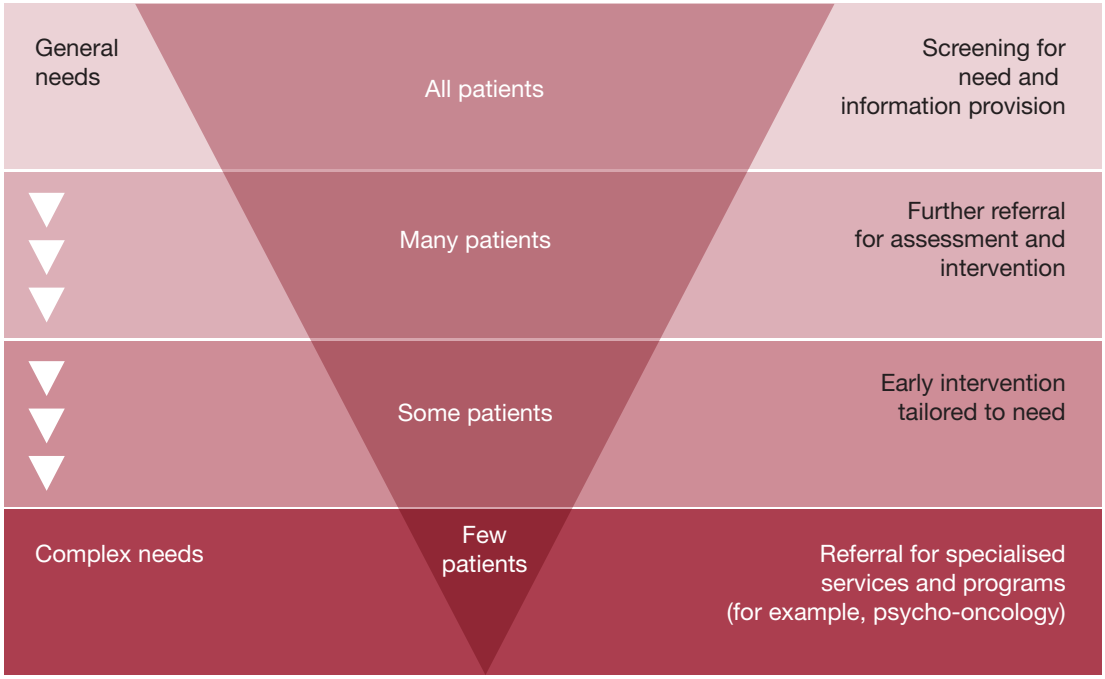
Appendix A: Supportive care domains

Supportive care in cancer refers to the following five domains:

- the physical domain, which includes a wide range of physical symptoms that may be acute, relatively short lived or ongoing, requiring continuing interventions or rehabilitation
- the psychological domain, which includes a range of issues related to the patient's mental health and wellbeing and personal relationships
- the social domain, which includes a range of social and practical issues that will affect the patient, carer and family such as the need for emotional support, maintaining social networks and financial concerns
- the information domain, which includes access to information about cancer and its treatment, recovery and survivorship support services and the health system overall
- the spiritual domain, which focuses on the patient's changing sense of self and challenges to their underlying beliefs and existential concerns (Palliative Care Victoria 2019).

Fitch's (2000) model of supportive care recognises the variety and level of intervention required at each critical point as well as the need to be specific to the individual patient (Figure A1). The model targets the type and level of intervention required to meet patients' supportive care needs.

Figure A1: Fitch's tiered approach to supportive care



Appendix B: Psychological needs

Consider a referral to a psychologist, psychiatrist, pastoral/spiritual care practitioner, social worker, specialist nurse or a relevant community-based program if the patient has these issues:

- displaying emotional cues such as tearfulness, distress that requires specialist intervention, avoidance or withdrawal
- being preoccupied with or dwelling on thoughts about cancer and death
- displaying fears about the treatment process or the changed goals of their treatment
- displaying excessive fears about cancer progression or recurrence
- worrying about loss associated with their daily function, dependence on others and loss of dignity
- becoming isolated from family and friends and withdrawing from company and activities that they previously enjoyed
- feeling hopeless and helpless about the effect that cancer is having on their life and the disruption to their life plans
- struggling to communicate with family and loved ones about the implications of their cancer diagnosis and treatment
- experiencing changes in sexual intimacy, libido and function
- struggling with the diagnosis of relapsed, refractory or advanced disease
- having difficulties quitting smoking (refer to Quitline on 13 7848) or with other drug and alcohol use
- having difficulties transitioning to palliative care.

Additional considerations that may arise for the multidisciplinary team include:

- support for the carer – encourage referrals to psychosocial support from a social worker, psychologist or general practitioner
- referral to an exercise physiologist or physiotherapist as a therapeutic approach to prevent and manage psychological health
- referral to wellness-after-cancer programs to provide support, information and offer strategies.

Appendix C: Special population groups

The burden of cancer is not evenly spread across Australia. People experiencing socioeconomic disadvantage, Aboriginal and Torres Strait Islander communities, culturally diverse communities, people living with a disability, people with chronic mental health or psychiatric concerns and those who live in regional and rural areas of Australia have poorer cancer outcomes.

Aboriginal and Torres Strait Islander people

Cancer is the third leading cause of burden of disease for Aboriginal and Torres Strait Islander people. While Australia's cancer survival rates are among the best in the world, Aboriginal and Torres Strait Islander people continue to experience a different pattern of cancer incidence and significant disparities in cancer outcomes compared with non-Indigenous Australians.

For Aboriginal and Torres Strait Islander people, health and connection to land, culture, community and identity are intrinsically linked. Health encompasses a whole-of-life view and includes a cyclical concept of life–death–life.

The distinct epidemiology of cancer among Aboriginal and Torres Strait Islander people, and unique connection to culture, highlight the need for a specific optimal care pathway for Aboriginal and Torres Strait Islander people with cancer. Ensuring this pathway is culturally safe and supportive is vital to tackling the disparities for Aboriginal and Torres Strait Islander people.

Published in 2018, the *Optimal care pathway for Aboriginal and Torres Strait Islander people with cancer* provides guidance to health practitioners and service planners on optimal care for Aboriginal and Torres Strait Islander people with cancer across the cancer continuum.

- In addition to the key principles underpinning cancer-specific pathways, these are the key concepts that are fundamental to Aboriginal and Torres Strait Islander health:
- providing a holistic approach to health and wellbeing
- providing a culturally appropriate and culturally safe service
- acknowledging the diversity of Aboriginal and Torres Strait Islander peoples
- understanding the social determinants and cultural determinants of health (Cancer Australia 2015).

To view the optimal care pathway for Aboriginal and Torres Strait Islander people with cancer and the corresponding quick reference guide, visit the Cancer Australia website <https://www.canceraustralia.gov.au/publications-and-resources/cancer-australia-publications/optimal-care-pathway-aboriginal-and-torres-strait-islander-people-cancer> and <https://www.canceraustralia.gov.au/publications-and-resources/cancer-australia-publications/optimal-care-pathway-aboriginal-and-torres-strait-islander-people-cancer-quick-reference-guide>.

To view the consumer resources – Checking for cancer and Cancer from the Cancer Australia website <https://www.canceraustralia.gov.au/publications-and-resources/cancer-australia-publications/checking-cancer-what-expect> and <https://www.canceraustralia.gov.au/publications-and-resources/cancer-australia-publications/cancer-what-expect>.

Culturally diverse communities

For people from culturally diverse backgrounds in Australia, a cancer diagnosis can come with additional complexities, particularly when English proficiency is poor. In many languages there is not a direct translation of the word 'cancer', which can make communicating vital information difficult. Perceptions of cancer and related issues can differ greatly in people from culturally diverse backgrounds, and this can affect their understanding and decision making after a cancer diagnosis. In addition to different cultural beliefs, when English language is limited, there is potential for miscommunication of important information and advice, which can lead to increased stress and anxiety for patients.

A professionally trained interpreter (not a family member or friend) should be made available when communicating with people with limited English proficiency. Navigation of the Australian healthcare system can pose problems for those with a non-Anglo culture, and members of the treatment teams should pay particular attention to supporting these patients.

The Australian Cancer Survivorship Centre has developed a glossary of more than 700 cancer terms in nine different languages. The multilingual glossary has been designed as a resource for professional translators, interpreters and bilingual health professionals working in the cancer field. The glossary is a unique tool that enables language professionals with access to accurate, consistent and culturally appropriate terminology.

Visit the Peter Mac website <www.petermac.org/multilingualglossary> to see the glossary.

People with disabilities

Disability, which can be physical, intellectual or psychological, may have existed before the cancer diagnosis or may be new in onset (occurring due to the cancer treatment or incidentally). Adjusting to life with a disability adds another challenge to cancer care and survivorship.

Several barriers prevent people with disabilities from accessing timely and effective health care (AIHW 2017):

- physical limitations
- competing health needs
- the trauma of undergoing invasive procedures
- potential barriers associated with obtaining informed consent
- failure to provide assistance with communication
- lack of information
- discriminatory attitudes among healthcare staff.

In caring for people with disabilities and a cancer diagnosis, the Australian Institute of Health and Welfare disability flag should be used at the point of admittance to correctly identify and meet the additional requirements of a person with disability. Facilities should actively consider access requirements, and health practitioners should make reasonable adjustments where required.

Patients aged between seven and 65 years who have a permanent or significant disability may be eligible for support or funding through the National Disability Insurance Scheme (National Disability Insurance Agency 2018). More information can be found on the NDIS website <www.ndis.gov.au>.

Patients aged 65 years or older (50 years or older for Aboriginal or Torres Strait Islander people) may be eligible for subsidised support and services through aged care services. An application to determine eligibility can be completed online or over the phone. More information can be found at the My Aged Care website <www.myagedcare.gov.au>.

More information

Talking end of life is a resource that shows how to teach people with intellectual disability about end of life. It is designed for disability support workers but is also helpful for others including families, health professionals and educators.

To view the resource, visit the Talking End of Life website <www.caresearch.com.au/tel/tabid/4881/Default.aspx>.

Older people with cancer

Planning and delivering appropriate cancer care for older people can present a number of challenges. This could also be true for frail people or those experiencing comorbidities. Effective communication between oncology and geriatrics departments will help facilitate best practice care, which takes into account physiological age, complex comorbidities, risk of adverse events and drug interactions, as well as the implications of cognitive impairment on suitability of treatment and consent (Steer et al. 2009).

At a national interdisciplinary workshop convened by the Clinical Oncology Society of Australia, it was recommended that people over the age of 70 undergo some form of geriatric assessment, in line with international guidelines (COSA 2013; palliAGED 2018). Screening tools can be used to identify those patients in need of a comprehensive geriatric assessment (Decoster et al. 2015).

This assessment can be used to help determine life expectancy and treatment tolerance and guide appropriate referral for multidisciplinary intervention that may improve outcomes (Wildiers et al. 2014).

Frailty is not captured through traditional measures of performance status (e.g. ECOG) and includes assessment in the domains of:

- function
- comorbidity
- presence of geriatric syndromes
- nutrition
- polypharmacy
- cognition
- emotional status
- social supports.

Adolescents and young adults

In recent years, adolescent and young adult oncology has emerged as a distinct field due to lack of progress in survival and quality-of-life outcomes (Ferrari et al. 2010; Smith et al. 2013). The significant developmental change that occurs during this life stage complicates a diagnosis of cancer, often leading to unique physical, social and emotional effects for young people at the time of diagnosis and throughout the cancer journey (Smith et al. 2012).

In caring for young people with cancer, akin to the comorbidities that require specific care in the older cancer population, the treatment team needs to pay careful attention to promoting normal development (COSA 2014). This requires personalised assessments and management involving a multidisciplinary, disease-specific, developmentally targeted approach that adheres to the following principles:

- understanding the developmental stages of adolescence and supporting normal adolescent health and development alongside cancer management
- understanding and supporting the rights of young people
- communication skills and information delivery that are appropriate to the young person
- meeting the needs of all involved, including the young person, their carers and their family
- working with educational institutions and workplaces
- considering survivorship and palliative care needs.

An oncology team caring for an adolescent or young adult with cancer should be able to demonstrate these specific areas of expertise:

- be able to ensure access to expert adolescent and young adult health providers who have knowledge specific to the biomedical and psychosocial needs of the population
- understand the biology and current management of the disease in the adolescent and young adult age group
- consider participating in research and clinical trials for each patient
- engage in proactive discussion and management of fertility preservation, late effects of treatment, ongoing need for contraception, and psychosocial and psychosexual needs
- provide treatment in an environment that is friendly to adolescents and young adults.

People experiencing socioeconomic disadvantage

In general, people from lower socioeconomic groups are at greater risk of poor health, have higher rates of illness, disability and death, and live shorter lives than those from higher socioeconomic groups (AIHW 2016). People experiencing socioeconomic disadvantage are less likely to participate in screening programs, more likely to be obese, less likely to exercise and much more likely to smoke, which are all risk factors for cancer. In 2010–2014 age-standardised cancer incidence rates were higher in the lowest socioeconomic areas compared with the highest socioeconomic areas for all cancers combined (Cancer Australia 2019b).

Socioeconomic status and low health literacy are closely correlated. Therefore, effective communication with patients and carers is particularly important given the prevalence of low health literacy in Australia (estimated at 60% of Australian adults) (ACSQHC 2014).

Consideration should be taken for cancer patients experiencing socioeconomic disadvantage to reduce their risk of being underserved for health care.

People with chronic mental health or psychiatric concerns

A diagnosis of cancer may present additional challenges to people who have pre-existing chronic mental health or psychiatric concerns, resulting in exacerbation of their mental health symptoms. This may include heightened anxiety, worsening depression or thoughts of self-harm.

As poor adjustment and coping can affect treatment decisions, people who are known to have a mental health diagnosis need psychosocial assessment in the oncology setting to formulate a plan for ongoing support throughout treatment.

Psychosocial support can assist with challenges in communicating with health professionals, enhance understanding of the treatment journey, ensure capacity for consent to treatment options and improve compliance with treatment requests. A referral for psychosocial support from a health professional to the psycho-oncology team can ensure these patients are provided with targeted interventions or referrals to community-based services that may mitigate problems associated with the impacts of social isolation that frequently accompany chronic mental ill-health.

Many patients with chronic mental health problems may be well known to external service providers. Psycho-oncology health professionals can form meaningful partnerships with existing service providers to optimise patient care throughout treatment and beyond.

Drug use disorders fall within the area of mental health conditions. People who are opiate dependent may have specific and individual requirements regarding pain management and their own preference for type of opiate prescribed or used.

Sexually and gender diverse groups

People who identify as sexually or gender diverse may have unique needs following a cancer diagnosis. Sexually or gender diverse identities include (but are not limited to) people who identify as lesbian, gay, bisexual or transgender. There is no universally agreed upon initialism to describe this community, with other terms such as queer/questioning (Q), intersex (I), asexual (A) and pansexual (P) often included, as well as a plus symbol (+) indicating inclusivity of other identities not explicitly mentioned. For the purposes of this document, this community is referred to as LGBTQI+.

Sexual orientation and gender identity are relevant across the entire spectrum of cancer care, from prevention to survivorship and end-of-life care. LGBTQI+ people are less likely to participate in cancer screening, and some segments of the LGBTQI+ community exhibit elevated rates of specific cancer risk factors – for example, higher rates of smoking and alcohol use. Regarding treatment, there may be unique factors relevant to LGBTQI+ people that may affect decision making. Additionally, the LGBTQI+ population experiences higher rates of anxiety, depression and stressful life circumstances, and may be at risk of inferior psychosocial outcomes following a cancer diagnosis. LGBTQI+ people are also more likely to be estranged from their families of origin, and for older people, less likely to have adult children who may provide support and care.

Barriers to care for LGBTQI+ people include past negative interactions with healthcare systems, experiences or fear of discrimination and harassment in healthcare settings, assumptions of cisgender/heterosexual identity, lack of recognition or exclusion of same-sex partners from care, and a lack of relevant supportive care and information resources.

To provide safe and appropriate care for LGBTQI+ people with cancer, healthcare providers should:

- display environmental cues to show an inclusive and safe setting for LGBTQI+ patients
- avoid assumptions about the sexual orientation or gender identity of patients and their partners
- facilitate positive disclosure of sexual orientation or gender identity
- include same sex/gender partners and families of choice in care
- be aware of relevant supportive care and information resources
- provide non-judgemental, patient-centred care.

Appendix D: Complementary therapies

Complementary therapies may be used together with conventional medical treatments to support and enhance quality of life and wellbeing. They do not aim to cure the patient's cancer. Instead, they are used to help control symptoms such as pain and fatigue (Cancer Council Australia 2019).

The lead clinician or health professional involved in the patient's care should discuss the patient's use (or intended use) of complementary therapies not prescribed by the multidisciplinary team to assess safety and efficacy and to identify any potential toxicity or drug interactions.

The lead clinician should seek a comprehensive list of all complementary and alternative medicines being taken and explore the patient's reason for using these therapies and the evidence base.

A transparent and honest discussion that is free from judgement should be encouraged.

While some complementary therapies are supported by strong evidence, others are not. For such therapies, the lead clinician should discuss their potential benefits and use them alongside conventional therapies (NHMRC 2014).

If the patient expresses an interest in using complementary therapies, the lead clinician should consider referring patients to health providers within the multidisciplinary team who have expertise in the field of complementary and alternative therapies (e.g. a clinical pharmacist, dietitian or psychologist) to assist them to reach an informed decision. Costs of such approaches should be part of the discussion with the patient and considered in the context of evidence of benefit.

The lead clinician should assure patients who use complementary therapies that they can still access a multidisciplinary team review and encourage full disclosure about therapies being used.

More information

See the Clinical Oncological Society of Australia's position statement Use of complementary and alternative medicine by cancer patients <https://www.cosa.org.au/media/1133/cosa_cam-position-statement_final_new-logo.pdf>.

Appendix E: Members of the multidisciplinary team for AL amyloidosis

The multidisciplinary team may include the following members:

- clinical haematologist / haemato-pathologist*
- specialist nurses*
- Aboriginal health practitioner, Indigenous liaison officer or remote general practitioner
- fertility specialist
- general practitioner
- allied health representative (e.g. exercise physiologist)
- occupational therapist, physiotherapist or social worker
- psychologist
- spiritual/pastoral care.

* Denotes core members. Core members of the multidisciplinary team are expected to attend most multidisciplinary team meetings either in person or remotely.

Resource list

For patients, families and carers

Advance Care Planning Australia

Advance Care Planning Australia provides national advance care planning resources for individuals, families, health professionals and service providers. Resources include a national advisory service, information resources, a legal forms hub and education modules.

- Telephone: 1300 208 582
- Website <www.advancecareplanning.org.au>

Australian Amyloidosis Network

The AAN is an amalgamation of four multi-disciplinary medical amyloidosis centres, dedicated to the diagnosis and management of Australian patients with all types of amyloidosis. This website has been developed by the AAN as a practical resource for Australian patients, carers and their doctors, to enhance awareness and ensure the timely and correct diagnosis of their type of amyloidosis.

- Website <www.aan.org.au>

Australian Cancer Survivorship Centre

The Australian Cancer Survivorship Centre has developed information resources and events to help people move from initial treatment to post treatment and beyond, including those receiving maintenance treatments. While they do not provide clinical advice, they connect with a range of providers to enable improved care.

- Telephone: (03) 8559 6220
- Website <www.petermac.org/cancersurvivorship>

Australian Commission on Safety and Quality in Health Care

The Australian Commission on Safety and Quality in Health Care has developed a resource for patients and carers explaining the coordination of care that patients should receive from their health service during cancer treatment. The resource is called *What to expect when receiving medication for cancer care* <https://www.safetyandquality.gov.au/publications-and-resources/resource-library/what-expect-when-receiving-medication-cancer-care>.

Beyond Blue

Beyond Blue provides information about depression, anxiety and related disorders, as well as about available treatment and support services.

- Telephone: 1300 22 4636
- Website <www.beyondblue.org.au>

Cancer Australia

Cancer Australia is a specialist agency within the Australian Government's Health portfolio, providing national leadership in cancer control across all cancers, for all Australians.

Cancer Australia's purpose is to minimise the impact of cancer, address disparities, and improve the health outcomes of people affected by cancer in Australia by providing national leadership in cancer control.

Cancer Australia achieves this by developing and promoting evidence-based best practice cancer care; providing consumer and health professional cancer information; funding priority cancer research; and strengthening national cancer data capacity.

Cancer Australia provides accessible, evidence-based information about cancer for people affected by cancer, carers and their families through the Cancer Australia websites, resource library and video content.

- Website <www.canceraustralia.gov.au>

Cancer Council's Cancer Information and Support Service

Cancer Council 13 11 20 is a confidential telephone support service available to anyone affected by cancer. This service acts as a gateway to evidence-based documented, practical and emotional support available through Cancer Council services and other community organisations. Calls will be answered by a nurse or other oncology professional who can provide information relevant to a patient's or carer's situation. Health professionals can also access this service.

- Telephone: 13 11 20 – Monday to Friday, 9.00am to 5.00pm (some states have extended hours)
- Website <www.cancer.org.au/about-us/state-and-territory-councils/>

Cancer Council's Cancer Connect

Cancer Connect is a free and confidential telephone peer support service that connects someone who has cancer with a specially trained volunteer who has had a similar cancer experience.

A Connect volunteer can listen with understanding and share their experiences and ways of coping. They can provide practical information, emotional support and hope. Many people newly diagnosed with cancer find this one-to-one support very beneficial.

For more information on Cancer Connect call Cancer Council 13 11 20.

Canteen

Canteen helps adolescents, young adults and parents to cope with cancer in their family. Canteen offers individual support services, peer support services and a youth cancer service, as well as books, resources and useful links.

- Telephone: 1800 835 932 to talk to a health professional about information and support for young people or 1800 226 833 for other enquiries
- Website <www.canteen.org.au/>

Clinical trial information

For a collection of clinical trials available in Australia and internationally see the following information:

- Cancer Australia <www.australiancancertrials.gov.au>
- Australian New Zealand Clinical Trials Registry <www.anzctr.org.au>
- Australasian Leukaemia and Lymphoma Group <https://www.allg.org.au/clinical-trials-research/current-clinical-trials>
- ClinTrial Refer <www.clintrialrefer.org.au>
- ClinicalTrials.gov <www.clinicaltrials.gov> for an international view.

CanEAT pathway

A guide to optimal cancer nutrition for people with cancer, carers and health professionals.

- Education website <https://education.eviq.org.au/courses/supportive-care/malnutrition-in-cancer>
- Patient website <https://patients.cancer.nsw.gov.au/coping-with-cancer/physical-wellbeing/eating-well>

Guides to best cancer care

The short guides help patients, carers and families understand the optimal cancer care that should be provided at each step. They include optimal timeframes within which tests or procedures should be completed, prompt lists to support patients to understand what might happen at each step of their cancer journey and to consider what questions to ask, and provide information to help patients and carers communicate with health professionals.

The guides are located on an interactive web portal, with downloadable PDFs available in multiple languages.

- Website < www.cancercareguides.org.au>

Leukaemia Foundation

The Leukaemia Foundation provides specialist support, funds leading-edge research and advocates for Australians diagnosed with blood cancer. The foundation guides patients and their loved ones through the emotional, physical and psychosocial challenges of a blood cancer diagnosis, treatment and survivorship.

The foundation's team of qualified health professionals can answer questions, talk through concerns and connect patients to blood cancer support groups. The team can also help with practical concerns such as accommodation close to treatment, transport to appointments and financial assistance.

- Telephone: 1800 620 420
- Website <www.leukaemia.org.au>

Look Good, Feel Better

A free national community service program, run by the Cancer Patients Foundation, dedicated to teaching cancer patients how to manage the appearance-related side effects caused by treatment for any type of cancer.

- Telephone: 1800 650 960
- Website <www.lgfb.org.au>

MDS Foundation

The MDS Foundation is a global non-profit advocacy organisation that for more than 25 years has supported patients and their families as well as healthcare providers in the fields of MDS and its related diseases. The foundation supports and educates patients, their communities and healthcare providers, and contributes to innovative research in the fields of MDS and its related continuum of diseases to better diagnose, control and ultimately cure these diseases.

- Website <https://www.mds-foundation.org>
- Email <patientliaison@mds-foundation.org>

Myeloma Australia

Myeloma Australia is the only Australian myeloma specific not-for-profit organisation. They support, educate, inform, empower and bring hope to people who are living with myeloma, and their loved ones. They also raise community awareness and understanding of myeloma; educate health professionals involved in the care and treatment of those living with myeloma; advocate for improved patient access to the latest treatments at affordable prices; and facilitate myeloma research in Australia.

Myeloma Australia provides up-to-date information and caring support via its specialist myeloma nurses. All services are provided free of charge to ensure equitable access.

- Telephone: 1800 693 566
- Website <https://myeloma.org.au>

For any myeloma-related queries, email a Myeloma Support Nurse <nurses@myeloma.org.au>.

Quitline

Quitline is a confidential, evidence-based telephone counselling service. Highly trained Quitline counsellors use behaviour change techniques and motivational interviewing over multiple calls to help people plan, make and sustain a quit attempt.

Quitline is a culturally inclusive service for all, and Aboriginal counsellors are also available. Health professionals can refer patients to Quitline online.

- Telephone: 13 7848
- Website <www.quit.org.au> or the relevant website in your state or territory

For health providers

Australian Cancer Survivorship Centre

The Australian Cancer Survivorship Centre provides expertise in survivorship care, information, support and education. Its purpose is to support and enable optimal survivorship care.

- Telephone: (03) 8559 6220
- Website <www.petermac.org/cancersurvivorship>

Australian Commission on Safety and Quality in Health Care

The Australian Commission on Safety and Quality in Health Care has developed a guide for clinicians containing evidence-based strategies to support clinicians to understand and fulfil their responsibilities to cancer patients.

This guide is particularly relevant to Steps 3 to 6 of the optimal care pathway. The guide is titled *NSQHS Standards user guide for medication management in cancer care for clinicians* <www.safetyandquality.gov.au/publications-and-resources/resource-library/nsqhs-standards-user-guide-medication-management-cancer-care-clinicians>.

Cancer Australia

Cancer Australia provides evidence-based information for health professionals including guidance, cancer learnings, cancer guides, reports, resources, videos, posters and pamphlets.

- Website <www.canceraustralia.gov.au>

Cancer Council Australia

Information on prevention, research, treatment and support provided by Australia's peak independent cancer authority.

- Website <www.cancer.org.au>

CanEAT pathway

A guide to optimal cancer nutrition for people with cancer, carers and health professionals.

- Education website <https://education.eviq.org.au/courses/supportive-care/malnutrition-in-cancer>
- Patient website <https://patients.cancer.nsw.gov.au/coping-with-cancer/physical-wellbeing/eating-well>.

eviQ

A clinical information resource providing health professionals with current evidence-based, peer-maintained, best practice cancer treatment protocols and information relevant to the Australian clinical environment.

- Website <www.eviq.org.au>

National Aboriginal Community Controlled Health Organisation

The National Aboriginal Community Controlled Health Organisation (NACCHO) is the national leadership body for Aboriginal and Torres Strait Islander health in Australia. NACCHO provides advice and guidance to the Australian Government on policy and budget matters and advocates for community-developed solutions that contribute to the quality of life and improved health outcomes for Aboriginal and Torres Strait Islander people.

- Website <www.naccho.org.au/about>

National Health and Medical Research Council

Information on clinical practice guidelines, cancer prevention and treatment.

- Website <www.nhmrc.gov.au>

Glossary

advance care directive – voluntary person-led document that focus on an individual's values and preferences for future health and medical treatment decisions, preferred outcomes and care. They are completed and signed by a competent person. They are recognised by specific legislation (statutory) or common law (non-statutory). Advance care directives can also appoint the substitute decision-maker(s) who can make decisions about health or personal care on the individual's behalf if they are no longer able to make decisions themselves. Advance care directives focus on the future health care of a person, not on the management of his or her assets. They come into effect when an individual loses decision-making capacity.

advance care planning – the process of planning for future health and personal care, where the person's values, beliefs and preferences are made known so they can guide decision making at a future time when that person cannot make or communicate their decisions.

alternative therapies – treatments used in place of conventional medical treatment.

amyloid – is the term for the protein fibril.

amyloidosis – is the clinical disease resulting from deposition of amyloid fibrils where fibril deposition is known to be the cause of the disease rather than a bystander effect.

amyloid forming proteins – at least 36 proteins are known to cause amyloidosis in humans and, of these, 34 are endogenous human proteins.

care coordinator – the health provider nominated by the multidisciplinary team to coordinate patient care. The care coordinator may change over time depending on the patient's stage on the care pathway and the location in which care is being delivered.

complementary therapies – supportive treatment used in conjunction with conventional medical treatment. These treatments may improve wellbeing and quality of life and help people deal with the side effects of cancer.

end-of-life care – includes physical, spiritual and psychosocial assessment, and care and treatment, delivered by health professionals and ancillary staff. It also includes support of families and carers and care of the patient's body after their death.

immunotherapy – a type of cancer treatment that helps the body's immune system to fight cancer. Immunotherapy can boost the immune system to work better against cancer or remove barriers to the immune system attacking the cancer.

indicator – a documentable or measurable piece of information regarding a recommendation in the optimal care pathway.

informed financial consent – the provision of cost information to patients, including notification of likely out-of-pocket expenses (gaps), by all relevant service providers, preferably in writing, before admission to hospital or treatment (Australian Government Department of Health 2017).

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

localised vs systemic AL amyloidosis – in systemic amyloidosis, amyloidogenic proteins are produced at a distant site, such as the bone marrow in AL amyloidosis, and deposit in organs that are anatomically separate, such as the heart. In localised amyloidosis, deposits occur only at the site of light chain production. This OCP only deals with systemic AL amyloidosis.

multidisciplinary care – an integrated team approach to health care in which medical and allied health providers consider all relevant treatment options and collaboratively develop an individual treatment plan for each patient.

multidisciplinary team – comprises the core disciplines that are integral to providing good care. The team is flexible in approach, reflects the patient's clinical and psychosocial needs and has processes to facilitate good communication.

multidisciplinary team meeting – a meeting of health professionals from one or more clinical disciplines who together make decisions about recommended treatment of patients.

nomenclature – all amyloidosis is named by an abbreviation of fibril protein followed by the term 'amyloidosis'. In the case of AL amyloidosis, the fibril is AL ('A' = amyloid, 'L' = light chain) and the disease is AL amyloidosis. Another acceptable term is immunoglobulin light chain amyloidosis. The term 'primary amyloidosis' should no longer be used.

optimal care pathway – the key principles and practices required at each stage of the care pathway to guide the delivery of consistent, safe, high-quality and evidence-based care for all people affected by cancer.

performance status – an objective measure of how well a patient can carry out activities of daily life.

primary care health professional – in most cases this is a general practitioner but may also include general practice nurses, community nurses, nurse practitioners, allied health professionals, midwives, pharmacists, dentists and Aboriginal health workers.

prognostic assessment – evaluation of clinical features (e.g. pathological, biochemical, molecular, genetic, simple clinical measurements) to predict a patient's likelihood of responding to treatment, developing disease or experiencing a medical event.

spiritual care – the aspect of humanity that refers to the way individuals seek and express meaning and purpose and the way they experience their connectedness to the moment, to self, to others, to nature, and to the significant or sacred.

substitute decision-maker – a person permitted under the law to make decisions on behalf of someone who does not have competence or capacity.

supportive care – care and support that aims to improve the quality of life of people living with cancer, cancer survivors and their family and carers and particular forms of care that supplement clinical treatment modalities.

survivorship – an individual is considered a cancer survivor from the time of diagnosis, and throughout their life; the term includes individuals receiving initial or maintenance treatment, in recovery or in the post-treatment phase.

survivorship care plan – a formal, written document that provides details of a person's cancer diagnosis and treatment, potential late and long-term effects arising from the cancer and its treatment, recommended follow-up, surveillance, and strategies to remain well.

therapy – a medicine that blocks the growth and spread of cancer by interfering with specific molecules.

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