Optimal care pathway for people with Waldenström's macroglobulinaemia

FIRST EDITION

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Endorsed by

ALLG Australasian Leukaemia & Lymphoma Group

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Australian Government
Cancer Australia

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Lymphoma Australia

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/>, from the Cancer Council website <www.cancer.org.au/OCP> and from Lymphoma Australia <www.lymphoma.org.au>

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Enquiries about this publication can be sent to <bloodcancerpartnerships@leukaemia.org.au>.

Contents

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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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 Waldenstrom's macroglobulinaemia (WM)*.

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

Step 1: Prevention and early detection

Prevention

The cause of WM is not fully understood, and there are currently no effective prevention strategies. At present, there is no evidence linking lifestyle, environmental or behavioural factors to prevention of WM.

Risk factors

The risk factors for developing WM include:

- age (occurs mainly in people over 60)
- family or personal history of WM, multiple myeloma and Non-Hodgkins lymphoma and autoimmune disease
- Caucasian males are more likely to get WM
- autoimmune diseases both organ specific and systemic
- infective disorders such as Hepatitis C.

Early detection

There is no established benefit regarding early detection of asymptomatic WM.

Screening recommendations

Routine screening for WM is not currently recommended in either the general population or in asymptomatic relatives of people with WM. Patients with WM are also at increased risk (almost 2-fold) of other second malignancies.

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

WM is an indolent lymphoma; therefore, it often develops slowly. Patients with WM may present with subtle signs and symptoms, however approximately half of all newly diagnosed WM patients are asymptomatic and 30% are identified through abnormal blood tests.

Symptoms, when present can include:

- fatigue due to anaemia and/or hyperviscosity. Hyperviscosity syndrome is a medical emergency (see treatment section).
- B symptoms such as night sweats, fevers, unexplained weight loss >10% of body weight within the past 6 months
- headache, blurred vision, confusion, epistaxis, shortness of breath, other bleeding symptoms.
- neuropathy such as numbness, weakness, balance difficulties, falls and pain
- easy bruising or bleeding
- dyspnoea
- muscle cramps.

Initial investigations by the GP should include:

- history and physical examination of the skin, all lymph node groups, abdomen, neurological and cardiorespiratory examination
- full blood count and imaging (ultrasound, chest x-ray, CT)
- urea, electrolytes, creatinine, lactate dehydrogenase (LDH)
- liver function tests iron studies
- beta-2 microglobulin
- serum EPG and immunofixation to confirm underlying monoclonal protein.

Checklist

- ❑ Symptoms indicative of hyperviscosity require urgent clinical investigation
- ❑ Signs and symptoms recorded
- ❑ Patient notified of support services such as Cancer Council 13 11 20, Leukaemia Foundation 1800 620 420 or Lymphoma Australia 1800 953 081
- ❑ Referral options discussed with the patient and/or carer including cost implications

Timeframe

All investigations should be completed, and a path of action decided, **within 4 weeks**.
For patients with symptoms suggestive of hyperviscosity, the blood tests should be performed **within 1 day** of presentation.

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

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 and Lymphoma Australia 1800 359 081.

Timeframe continued

In most cases referral to a specialist **within 2 weeks** is appropriate. Urgent referral should occur **within 72 hours** if the patient presents with severe B symptoms, anaemia (Hb <80g/L), or **within 24 hours** if symptoms of hyperviscosity are observed.

Step 3: Diagnosis, staging and treatment planning

WM is diagnosed by blood tests demonstrating an IgM paraprotein in the peripheral blood. Additionally, either a bone marrow biopsy to detect infiltration of clonal B cells, plasmacytoid lymphocytes and plasma cells in the bone marrow or lymph node biopsy to demonstrate involvement with clonal plasmacytoid lymphocytes is required to differentiate WM from other low-grade lymphomas or from IgM MGUS.

Other routine investigations include:

- evaluation of relevant organ function (creatinine, uric acid, bilirubin, lactate dehydrogenase, haptoglobin, transaminases, alkaline phosphatase, ß2-microglobulin)
- iron studies
- chest radiograph (unless computed tomography [CT] has been performed for other reasons)
- viral serology (hepatitis B, hepatitis C, HIV, Epstein-Barr virus and cytomegalovirus)
- if there are clinical suspicions, or laboratory notification, of cryoglobulinemia, the serum sample should be collected and transported to the laboratory at 37 degrees.

Under certain circumstances the following investigations may be undertaken:

- CT scan and other imaging
- nerve conduction studies and laboratory evaluations (and a referral to a neurologist considered)
- when Bing Neel syndrome is suspected neurologist review and MRI brain, and spine should be undertaken and lumbar puncture considered.
- molecular tests.

Genetic testing

Currently there are no genetic tests applicable to predict family risk of WM.

Treatment planning

The multidisciplinary team should discuss patients with WM 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 WM.

Checklist

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

Timeframe

Patients presenting with WM and symptoms of hyperviscosity should be assessed urgently **within 1-2 days** for potential plasmapheresis.

The timing of diagnostic work-up should be guided by the severity of anaemia, level of paraprotein and symptoms and in general should be completed **within four weeks** following assessment by a haematologist.

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

Step 3: Diagnosis, staging and treatment planning continued

Communication

The lead clinician's[1] responsibilities include:

- discussing a timeframe for diagnosis and treatment options with the patient and/or carer
- explaining the role of the multidisciplinary team in treatment planning and ongoing care
- encouraging discussion about the diagnosis, prognosis, advance care planning and palliative care while clarifying the patient's wishes, needs, beliefs and expectations, and their ability to comprehend the communication
- providing appropriate information and referral to support services as required
- communicating with the patient's GP about the diagnosis, treatment plan and recommendations from multidisciplinary meetings.

Timeframe continued

Patients suspected to have only IgM MGUS can be reasonably reviewed, with their results **in 6 months**. If their IgM remains stable, it is appropriate they return to their GP for **6 monthly review** of symptoms and IgM levels thereafter **annually** if stable, and re-referral to the haematologist in accordance with the criteria above.

Step 4: Treatment

Establish intent of treatment

WM is a highly treatable low grade lymphoma. While incurable with current therapies, many patients with WM have such a prolonged survival that they may have a 'functional cure' of their WM.

The stages of WM are as follows:

- smouldering WM – asymptomatic phase, IgM paraprotein, no cytopenias, hyperviscosity or organomegaly. Regular surveillance, 'watch and wait' is indicated and may last for years
- symptomatic WM – treatment is indicated, most commonly due to symptoms of bone marrow failure or autoimmune manifestations.

Accurate classification is important as treatment is only indicated for symptomatic WM.

Watchful waiting

Many patients with newly diagnosed WM have asymptomatic smouldering disease which does not warrant treatment. Many people diagnosed with WM will not start treatment immediately, but instead have regular check-ups for symptoms. This is known as 'active surveillance', or 'watch and wait'. As WM is often a slow growing disease, it is a safe strategy which means people diagnosed with asymptomatic WM can avoid the side-effects that treatment can bring.

The most adopted, currently available approach for systemic treatment of WM in Australia is rituximab-based chemotherapy, either in combination with dexamethasone and cyclophosphamide (DRC), or bendamustine (BR). For patients considered ineligible for rituximab-chemotherapy, single agent zanubrutinib BTK inhibitor is PBS-funded for patients with a CIRS score ≥6.

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 and nutritional 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

Watchful waiting is usually recommended for newly diagnosed WM with asymptomatic smouldering disease which does not warrant treatment. Watchful waiting may also be recommended for mildly symptomatic patients if symptoms are not severe enough to warrant urgent treatment. Determining the tempo of disease progression during the period of watchful waiting is of value for prognostication.

When systemic therapy is indicated, timing should align with patient preferences but not be delayed to the point where the condition worsens.

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

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

Step 5: Care after initial treatment and recovery

Provide a treatment and follow-up summary to the patient, carer and GP outlining:

- the diagnosis, including tests performed and results
- treatment received (types and date)
- current toxicities (severity, management and expected outcomes)
- interventions and treatment plans from other health professionals
- potential long-term and late effects of treatment and care of these
- supportive care services provided
- a follow-up schedule, including tests required and timing
- contact information for key healthcare providers who can offer support for lifestyle modification
- a process for rapid re-entry to medical services for any issues arising.

Communication

The lead clinician's responsibilities include:

- explaining the treatment summary and immediate and long term 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

Treatment will depend on the features of disease, previous management, duration of response and the patient's preferences. Initial relapses/progression of WM in need of treatment is usually very responsive to treatment (for years).

Advance care planning

Advance care planning is important for all patients but especially those with multiply relapsed and refractory disease. It allows them to plan for their future health and personal care by thinking about their values and preferences. This can guide future treatment if the patient is unable to speak for themselves.

Survivorship and palliative care

Survivorship and palliative care should be addressed and offered early. Early referral to palliative care can improve quality of life. Referral should be based on need, not prognosis.

Communication

The lead clinician and team's responsibilities include:

- explaining the treatment intent, likely outcomes and side effects to the patient 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.

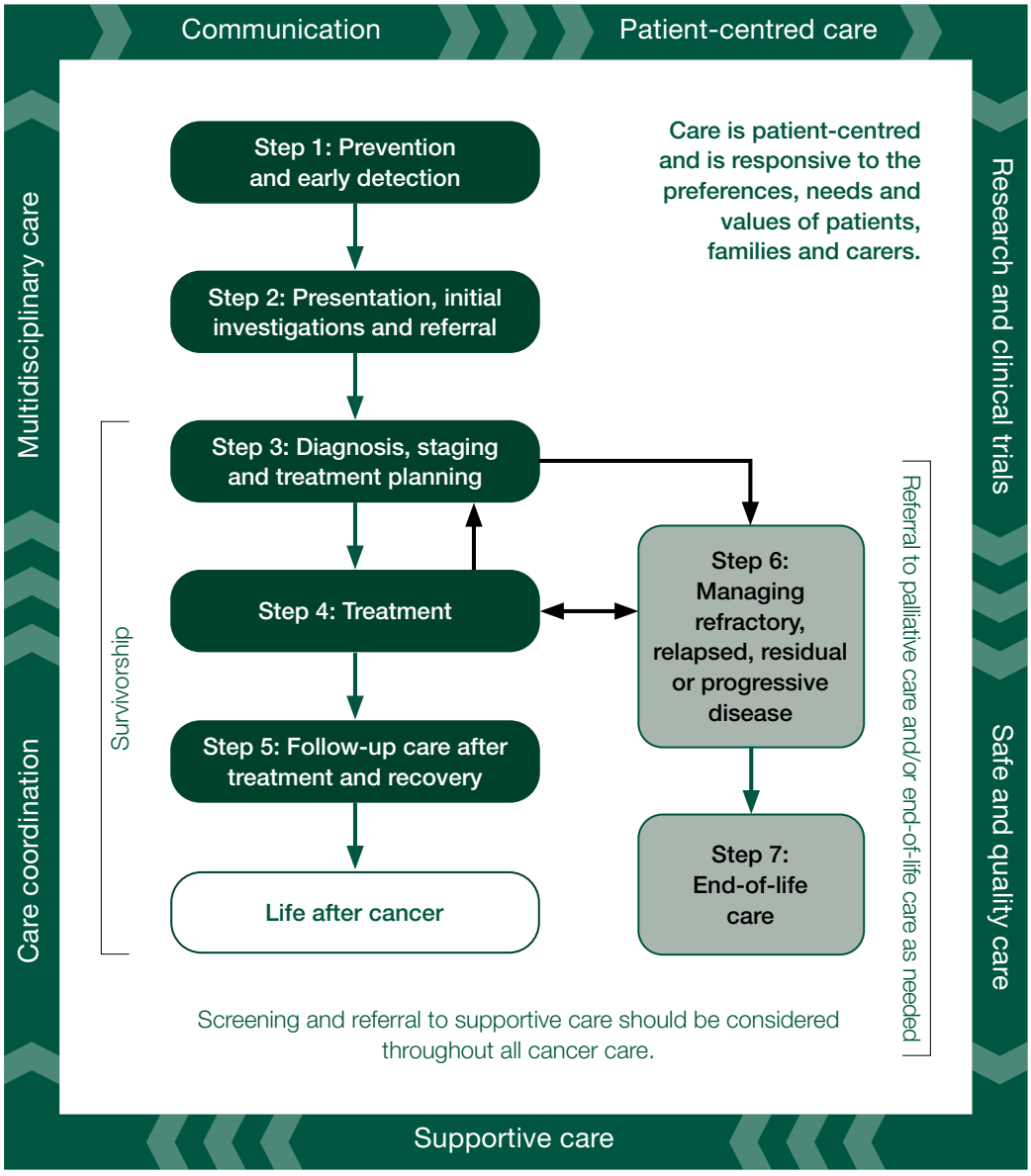
Intent of the optimal care pathways

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

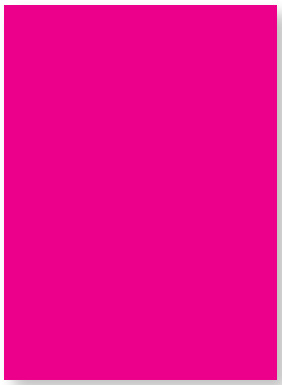
The quick reference guides are for health professionals and health services. They provide a summary of each optimal care pathway for health professionals and patients.

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 with cancer

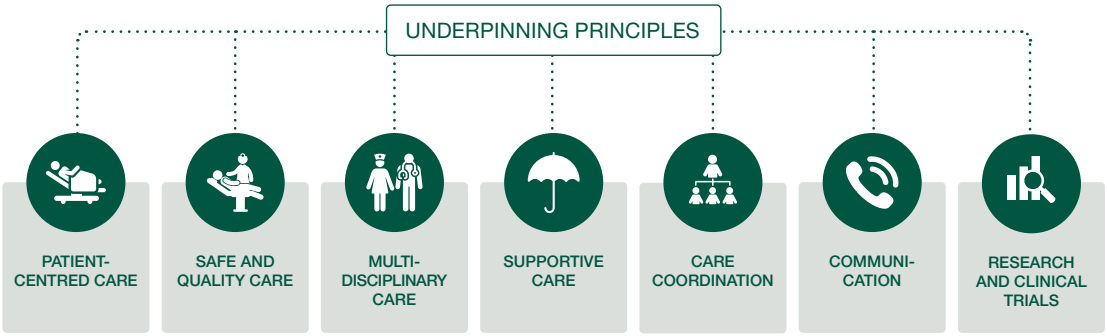
The *Optimal care pathway for Aboriginal and Torres Strait Islander people with cancer* provides a tool to help reduce disparities and improve outcomes and experiences for Aboriginal and Torres Strait Islander people with cancer. This resource can be used in conjunction with the optimal care pathway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

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 | Cancer Australia <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 Waldenström's Macroglobulinaemia Working Group.

Figure 3: Timeframes for care of Waldenström's macroglobulinaemia

Step in pathway	Care point	Timeframe
Presentation, initial investigations and referral	Signs and symptoms	Presenting symptoms should be triaged by a general practitioner.
	Initial investigations initiated by GP	For asymptomatic patients who do not need a prompt referral to a specialist, all investigations should be completed, and a path of action decided, **within 4 weeks** of first presentation. For patients with symptoms that suggest hyperviscosity (headache, blurred vision, confusion and epistaxis) **urgent same-day** assessment is required.
	Referral to specialist	In most cases referral to a specialist **within 2 weeks** is appropriate, but refer **within 72 hours** if severe B symptoms, anaemia (Hb <80 g/L), or symptoms of hyperviscosity. An urgent referral (**review within 24 hours**) should be made for any patient with symptoms of marked hyperviscosity such as epistaxis or new onset of confusion. A low-level IgM paraprotein (<10 g/L) without anaemia or symptoms suggestive of WM (or another lymphoproliferative disease) may be appropriately monitored by the general practitioner.
Diagnosis, staging and treatment planning	Diagnosis and staging	The timing of diagnostic work-up should be guided by the severity of anaemia, level of paraprotein and symptoms and in general for a patient with symptomatic WM should be completed **within 4 weeks** following assessment by a haematologist. Patients presenting with WM and symptoms of hyperviscosity should be assessed urgently **within 2 days** for potential plasmapheresis. Patients suspected to have only IgM MGUS can be reasonably reviewed, with their result **in 6 months**. If their IgM remains stable, it is appropriate they return to their general practitioner for **6–12 monthly review** of symptoms and IgM levels and re-referral to the haematologist in accordance with the criteria above.
	Multidisciplinary team meeting and treatment planning	Multidisciplinary meeting and treatment planning should occur before starting any disease-directed therapy. There needs to be a clear indication for starting therapy such as symptomatic disease, Hb <100 g/L or IgM rising >60 g/L.

Figure 3: Timeframes for care of Waldenström's macroglobulinaemia (continued)

Step in pathway	Care point	Timeframe
Treatment	Watchful waiting	Watchful waiting is usually recommended for newly diagnosed WM with asymptomatic smouldering disease that does not warrant treatment. Watchful waiting may also be recommended for mildly symptomatic patients if symptoms are not severe enough to warrant urgent treatment. Determining the tempo of disease progression during the period of watchful waiting is of value for prognostication.
	Systemic therapy	When systemic therapy is indicated, timing should align with patient preferences but not be delayed to the point where the condition worsens.
	Radiation therapy	Timeframes for treatment should reflect the overall goals of care and severity of symptoms.

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

Waldenström's macroglobulinaemia (WM) is a rare disorder, accounting for 1–2% of non-Hodgkin's lymphomas with a male preponderance and a median age of presentation of 63–75 years (Kastritis et al. 2018).

In the United States, the reported age-adjusted incidence is 3.4 per million (male) and 1.7 per million (female), while in Europe and the United Kingdom the reported incidences are 7.3 and 4.2 per million for males and females, respectively (Kastritis et al. 2018).

Accurate incidence is unclear in Australia. However, it is likely to be similar to European/UK data. The Australian Institute of Health and Welfare (AIHW) does not provide collated information relating to incidence for this particular disease.

Step 1: Presentation, initial investigations and referral

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

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 cause of WM is not fully understood, and there are currently no clear prevention strategies.

1.2 Risk factors

The risk factors for developing WM include:

- age – the median age at time of diagnosis for WM is between 63 and 75 years of age
- family history – a family history of WM, multiple myeloma and non-Hodgkin's lymphoma are risk factors for WM
- race/ethnicity – WM is more prevalent among Caucasian populations than African Americans or people of Asian descent
- gender – Caucasian males are more likely to get WM
- autoimmune diseases, both organ-specific (e.g. Graves' disease, haemolytic anaemia thrombocytopenic purpura, multiple sclerosis and others) and systemic (e.g. Sjögren's syndrome, rheumatoid arthritis, SLE, systemic sclerosis and others) (see 'Epidemiology' section for further details)
- infective disorders (e.g. hepatitis C).

No clear environmental causes have been identified that contribute to developing WM. There is currently no evidence linking lifestyle or behavioural factors to preventing WM.

There is an increased risk of WM when there is a personal history of:

- autoimmune disorders with associated increased risk ranging from 2.2–2.7-fold. These include organ-specific diseases such as thyroiditis, Graves' disease, haemolytic anaemia, thrombocytopenic purpura and multiple sclerosis. Systemic autoimmune diseases are also implicated including Sjögren's syndrome, rheumatoid arthritis, SLE, ankylosing spondylitis, systemic sclerosis, coeliac disease, polymyalgia rheumatic, giant cell arteritis and rheumatic fever (Royer et al. 2010)
- infective disorders (hepatitis C associated with 2.7-fold risk) (Sergentanis et al. 2015).

A family history of monoclonal gammopathy of undetermined significance (MGUS) is associated with a four-fold increased risk of WM. A family history of WM is associated with up to 20-fold increased risk in first-degree relatives (Kristinsson et al. 2010).

1.3 Risk reduction

While there is no evidence linking lifestyle changes to reduced risk of WM, 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

A thorough family history review for autoimmune disease, WM, non-Hodgkin Lymphoma and multiple myeloma is recommended.

While guidelines generally do not recommend screening of asymptomatic family members, the presence within a family of WM and either another B-cell lymphoproliferative disorders or multiple myeloma may prompt discussion regarding the merits of screening for symptoms in family members by the treating specialist.

No specific genetic tests are recommended in asymptomatic individuals.

1.4 Early detection

There is no established benefit regarding early detection of asymptomatic WM.

1.4.1 Screening recommendations

Routine screening for WM is not currently recommended in either the general population or in asymptomatic relatives of people with WM. Screening of asymptomatic members may be warranted under conditions outlined in section 1.3.1 'Genetic family history screening'.

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

Approximately half of all newly diagnosed WM patients are asymptomatic with no signs or symptoms, 30% of whom are identified through abnormal blood tests (The Rory Morrison Registry 2021).

WM is an indolent lymphoma, so it often develops slowly. Some people are asymptomatic when they are first diagnosed. The symptoms that do develop are related to the two defining aspects of the disease – infiltration of lymphoplasmacytic cells into the bone marrow (leading to cytopenias, in particular, symptomatic anaemia) and other tissues such as the spleen and nerves, and the presence of IgM paraprotein in the blood (leading to the hyperviscosity syndrome) (Gertz 2022).

The following are features of WM.

Common symptoms:

- fatigue due to anaemia and/or hyperviscosity
- B symptoms – night sweats, fevers, unexplained weight loss of more than 10% body weight
- hyperviscosity symptoms – headache, blurred vision, confusion, epistaxis, shortness of breath, other bleeding symptoms (hyperviscosity syndrome is a medical emergency – see treatment section)
- neuropathy (e.g. numbness, weakness, balance difficulties, falls and pain) is present in about 20% of patients with WM at diagnosis (Treon et al, 2010)
- easy bruising or bleeding due to low platelets or hyperviscosity syndrome
- dyspnoea due to anaemia or hyperviscosity syndrome
- muscle cramps.

Rare symptoms:

- organomegaly and lymphadenopathy
- skin abnormality – rash, cryoglobulinaemia, ulcers
- recurrent infections.

Other rare manifestations:

- symptoms of amyloidosis – nephrotic syndrome, weight loss, organ failure (especially renal), hepatic and cardiac impairment, peripheral oedema, macroglossia
- central nervous system (CNS) symptoms (Bing-Neel syndrome) due to infiltration of WM into the CNS including the brain and cerebrospinal fluid causing variable symptoms of confusion, weakness, headache and psychiatric disturbance
- cryoglobulinaemia – IgM clumps together and leads to impaired circulation in small blood vessels; this manifests as skin ulcers, reduced blood flow to the hands and feet, renal impairment and arthropathy

- cold-agglutinin haemolytic anaemia (IgM autoantibodies against red cell antigens are most active at low temperatures 3 to 4 degrees Celsius – this causes red cells to 'agglutinate' (stick together), the immune system is activated, and red cells are prematurely destroyed (extravascular haemolysis); this often occurs in the cooler parts of the body including the fingers, feet, nose and ears (Gertz 2022).
- Schnitzler's syndrome – erythematous urticarial skin vasculitis is a skin reaction from IgM deposition reacting with epidermal basement membrane, presenting as chronic hives, fever and arthralgias (Gertz 2022).

2.1.1 Timeframe for general practitioner consultation

Presenting symptoms should be triaged by a general practitioner. The presenting symptoms of WM are often non-specific. For instance, in patients presenting with B symptoms, there remains a broad differential diagnosis and these patients should be assessed **within two weeks** for further diagnostic assessment. Symptoms of headache, blurred vision, confusion and epistaxis can indicate hyperviscosity and require **urgent same-day assessment**.

2.2 Assessments by the general practitioner

Patients with WM may present with subtle signs and symptoms. The presence of unexplained B symptoms or unexplained anaemia should prompt consideration of a haematological malignancy, with further assessment including (but not limited to):

- a thorough history and physical examination of the skin, all lymph node groups, abdominals, neurological and cardiorespiratory systems
- presentation with headache, confusion and blurred vision may represent symptoms of hyperviscosity, a potentially urgent clinical scenario
- blood tests to assess organ dysfunction including:
  - full blood count
  - urea
  - electrolytes
  - creatinine
  - liver function tests
  - lactate dehydrogenase (LDH)
  - iron studies
  - other tests may include beta-2 microglobulin
- raised total protein and/or globulins can be a clue to the presence of an underlying paraprotein and may direct the general practitioner to also request serum EPG and immunofixation
- imaging of the affected area including ultrasound, chest radiography and computed tomography (CT) scan as appropriate.

2.2.1 Timeframe for completing investigations

For patients who do not need a prompt referral to a specialist, all investigations should be completed, and a path of action decided, **within four weeks** of first presentation. For patients with symptoms suggestive of hyperviscosity (see above), the blood tests above should be performed **within one day** of presentation.

2.3 Initial referral

If the cancer diagnosis is confirmed or the results are inconsistent or indeterminate, the general practitioner must refer the patient to a haematologist or medical oncologist with professional expertise in WM to make the diagnosis.

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 WM 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.

If WM is suspected, the following are indicators of concern that should lead to **prompt referral** to a haematologist:

- symptoms of hyperviscosity, in particular:
  - recent changes in vision may require **immediate referral** to an emergency department
  - an urgent referral – review **within 24 hours** should be made for any patient with symptoms of marked hyperviscosity – for example, epistaxis, new onset of confusion
- an IgM level over 60 g/L or a lower level associated with profound anaemia requires urgent referral to haematologist **within two days**
- symptomatic anaemia requiring transfusion
- neurological symptoms – acute confusion and visual change require prompt referral to an emergency department; peripheral neuropathy requires referral to neurologist
- marked B symptoms (weight loss >10%, persistent fevers >38°C, or persistent drenching night sweats) require prompt referral to haematologist **within one week**.

A low-level IgM paraprotein (<10 g/L) without anaemia or symptoms suggestive of WM (or another lymphoproliferative disease) may be appropriately monitored by the general practitioner, with repeat measurements after **six months** and then **annually** if stable.

If an IgM paraprotein is detected in a symptomatic patient, they should be referred to a specialist with the associated blood test results and clinical information.

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

In most cases referral to a specialist **within two weeks** is appropriate. Urgent referral should occur **within 72 hours** if severe B symptoms or anaemia (Hb <80 g/L) are present, or **within 24 hours** if symptoms of hyperviscosity are observed.

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 WM supportive care needs may arise for patients at this time:

- assistance for dealing with the emotional distress and/or anger of dealing with the uncertainty of a cancer diagnosis or a potential cancer diagnosis
- support to deal with the fear of the unknown, including anxiety/depression, interpersonal problems and adjustment difficulties (this should include an assessment of existing coping strategies and abilities to identify the needs for the patient, their carer and family)
- consider using the assistance, guidance and support of an allied health professional or bulk-billed / gap payment psychologist
- management of physical symptoms including pain, nausea and fatigue
- encouragement and support to increase levels of exercise (Cormie et al. 2018; Hayes et al. 2019)
- assessment of health literacy for self-care early in the diagnosis
- identifying impairments and providing targeted interventions to improve function level (Silver et al. 2013)
- an integrated and structured approach to self-care enquiry – identifying Social Health, Exercise, Education, Diet and Sleep Hygiene (SEEDS) is one such practical tool (Arden 2015)
- psycho-oncology support to improve their knowledge and motivation towards healthy self-care behaviours and to emotionally regulate throughout the course of the disease and treatments (this is particularly the case in patients on a 'watch and wait' approach, which can be associated with significant stress and anxiety)

- reviewing the patient's medication to ensure optimisation and to improve adherence to medicine used for comorbid conditions
- nutritional assessment and support
- management of iron deficiency (see section 4.2.3 'Supportive therapy')
- management of infections
- in advanced WM, hypoalbuminanemia is common, and dietetic support may be required.

Chronic disease management plans can be used to plan and coordinate the healthcare needs of the patient through multidisciplinary care.

Support groups, such as WMozzies, are available for guidance on WM. For patients, they reduce social isolation, inform and improve health literacy by providing treatment information via newsletters, online forums and links to other useful support and information. The WMozzies website also has links to the WhiMSICAL patient-derived data registry capturing treatment outcomes, quality-of-life information and COVID-19 data.

See: 'Chronic disease management plan'.

For more information refer to the National Institute for Health and Care Excellence 2015 guidelines, *Suspected cancer: recognition and referral* <www.nice.org.uk/guidance/ng12/chapter/Recommendations-on-patient-support-safety-netting-and-the-diagnostic-process>.

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 is being obtained).

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

The treatment team, after taking a thorough medical history and making a thorough medical examination of the patient, should undertake the following investigations under the guidance of a specialist.

WM is diagnosed by blood tests demonstrating an IgM paraprotein in the peripheral blood (serum immunoglobulin levels and serum protein electrophoresis). Additionally, either a bone marrow biopsy to detect infiltration of clonal B cells, plasmacytoid lymphocytes and plasma cells in the bone marrow or a lymph node biopsy to demonstrate involvement with clonal plasmacytoid lymphocytes is required to differentiate WM from other low-grade lymphomas or from IgM MGUS. Bone marrow biopsies are rarely performed for asymptomatic patients with a low-level IgM paraprotein (<10 g/L), normal full blood count and no concerning symptoms.

Other investigations include:

- evaluation of relevant organ function (creatinine, uric acid, bilirubin, lactate dehydrogenase, haptoglobin, transaminases, alkaline phosphatase, beta-2-microglobulin)
- haemolysis screen (reticulocytes, haptoglobin, unconjugated bilirubin, LDH, direct antiglobulin test)
- iron studies
- chest radiograph (unless CT has been performed for other reasons)
- viral serology (hepatitis B, hepatitis C, HIV, Epstein-Barr virus and cytomegalovirus) (Hallek et al. 2018)
- if there are clinical suspicions, or laboratory notification, of cryoglobulinemia, the serum sample should be collected and transported to the laboratory at 37 °C.

WM often follows the precursor asymptomatic IgM MGUS phase. IgM MGUS is defined by an IgM paraprotein of or under 30 g/L, associated with a clone of lymphoplasmacytic cells, without cytopenias, organomegaly, lymphadenopathy or bone marrow infiltration and an absence of symptoms (Gertz 2022).

The following investigations are only recommended under certain circumstances:

- CT scans and other imaging – CT scans are not recommended for asymptomatic patients or during routine evaluation. They are useful in patients with clinical lymphadenopathy, before starting treatment (Talaulikar et al. 2022). Guidelines for WM do not recommend the routine use of MRI, PET scan or ultrasound in WM outside of clinical trials. PET scans may be useful for patients with confirmed or suspected Richter's transformation.
- If symptoms and signs of a peripheral neuropathy are present, nerve conduction studies and laboratory evaluations for alternate causes should be undertaken (HbA1c, vitamin B12, HIV, ANCA, ANA) and referral to a neurologist considered.
- When Bing-Neel syndrome is suspected, neurologist review and an MRI brain and spine (with gadolinium) should be undertaken, and a lumbar puncture considered.
- Lymph node biopsy is not indicated for all patients with suspected WM. However, either core or excision biopsy may be considered in patients who have lymphadenopathy, in particular those with rapidly enlarging lymph nodes, to exclude transformation to diffuse large B-cell lymphoma.

Molecular genetics tests

- The MYD88 L265P mutation can be performed on biopsy tissue to assist in confirming the diagnosis of WM on the judgement of the pathologist/haematologist. It is present in the clonal lymphocytes of 90–95% of patients with WM. However, it non-specific, being present in more than 60% of patients with IgM MGUS and in some cases of other B-lymphoproliferative disorders including marginal zone lymphoma.
- CXCR4 mutations are present in more than 30% of patients with WM and can give prognostic information. Nonsense mutations are associated with more extensive bone marrow involvement and higher IgM levels.
- Mutations in MYD88 and CXCR4 are associated with response to therapy, but at present mutation status does not generally influence treatment choice.

Mutations in TP53 are uncommon in WM, tend to occur in MYD88(WT), are more common in relapsed disease and may be associated with poorer prognosis (Gustine et al. 2019; Poulain et al. 2017).

3.1.1 Timeframe for completing investigations

Patients presenting with WM and symptoms of hyperviscosity should be assessed for potential plasmapheresis **within two days**.

The timing of diagnostic work-up should be guided by the severity of anaemia, level of paraprotein and symptoms and, in general, should be completed **within four weeks** following assessment by a haematologist.

Patients suspected to have only IgM MGUS can be reasonably reviewed, with their results, in **six months.** If their IgM remains stable, it is appropriate they return to their general practitioner for **six-monthly review** of symptoms and IgM levels thereafter annually if stable, and re-referral to the haematologist in accordance with the criteria above.

3.1.2 Genetic testing (family risk)

Currently there are no genetic tests applicable to predict family risk of WM, but in patients with WM approximately 7% have a family history of WM or IgM MGUS, and the investigation of other family members is best done only after consultation with a haematologist.

3.1.3 Pharmocogenetics

Pharmacogenetics describes how individual genetic differences can lead to differences in the way certain medicines interact with the body. These interactions can affect the effectiveness of medications and any side effects. Applying pharmacogenetics to treatment planning may help patients to be prescribed the most appropriate treatment at the optimal dose from the beginning of treatment (NHMRC, 2013).

3.2 Staging, prognostic assessment and risk stratification

Staging is a critical element in treatment planning and should be clearly documented in the patient's medical record. Staging with CT is indicated before starting treatment or to investigate focal symptoms. PET imaging may be valuable if there is suspicion of large cell transformation.

The stages of WM are as follows:

- smouldering WM – asymptomatic phase, IgM paraprotein, no cytopenias, hyperviscosity or organomegaly; regular surveillance 'watch and wait' is indicated and may last for years
- symptomatic WM – treatment is indicated , most commonly due to symptoms of bone marrow failure or autoimmune manifestations.

Accurate classification is important because treatment is only indicated for symptomatic WM (Pratt et al. 2022). Clonal lymphoplasmacytic cells are present in the bone marrow in both smouldering and symptomatic WM.

Making prognostic estimates for WM should be very cautiously approached because newer treatments, generally given after relapse, result in prolonged responses and longer term survival. It is important to convey to the patient that a prognosis is an 'educated guess' only and based on average survival rates, which are influenced by other factors such as age and other comorbidities. Many patients die with, rather than from (or because of), their WM. The International Prognostic Scoring System for Waldenström's Macroglobulinemia (IPSSWM) has traditionally been used to estimate five-year overall survival in three risk categories (0–1 criteria, low risk, 2 criteria or age >65 intermediate risk, >2 criteria high risk), comprising the assessment of five adverse prognostic features. However, estimates of overall survival using the IPSSWM may not be applicable in the modern era.

The IPSSWM score correlates with symptoms of WM; however, it is not used to guide treatment choices – age, comorbidities and the goals of therapy guide treatment choices.

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.

Performance status should be recorded both at time of presentation and pre-morbid assessment. Consider using a validated geriatric assessment tool when determining fitness and evaluating intensity of treatment and potential impact of treatment on performance status.

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 WM.

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 WM. 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

Cancer prehabilitation uses a multidisciplinary approach combining exercise, nutrition and psychological strategies to prepare patients for the challenges of cancer treatment such as systemic therapy and radiation 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 cancer care 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 with WM have a higher risk of secondary malignancies, (OR 1.7). Those 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 WM, 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 et al, 2013).
- reviewing the patient's medication to ensure optimisation and to improve adherence to medicine used for comorbid conditions.

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

3.6.2 Fertility preservation and contraception

Cancer and cancer treatment may cause fertility problems. This will depend on the age of the patient, the type of cancer 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.

More information

See the Cancer Council website <https://www.cancervic.org.au/cancer-information/treatments/common-side-effects/fertility/fertility-overview.html> for more information.

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 WM as well as to relevant websites and support groups such as:
  - WMozzies the national affiliate of the International Waldenström Macroglobulinemia Foundation with excellent educational material specific to WM, as well as the Leukaemia Foundation and Lymphoma Australia,

- 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 carers
- 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 WM, 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 WM. For detailed clinical information on treatment options refer to these resources:

- Updated WM guidelines (v2) are available on the Myeloma Australia website <https://myeloma.org.au/wp-content/uploads/2022/06/MSAG-waldenstrom-guidelines-jun22.pdf>
- Guide to lab diagnosis: a practical guide to laboratory investigations at diagnosis and follow up in Waldenström macroglobulinaemia: recommendations from the Medical and Scientific Advisory Group, Myeloma Australia, the Pathology Sub-committee of the Lymphoma and Related Diseases Registry and the Australasian Association of Clinical Biochemists Monoclonal Gammopathy Working Group – PubMed (nih.gov) <https://pubmed.ncbi.nlm.nih.gov/31902622/>.

4.1 Treatment intent

WM is an incurable cancer. The goals of treatment can be defined as one or more of the following, which are not exclusive:

- improvement in quality of life and longevity with close observation and initiation of local or systemic therapy as indicated
- symptom management.

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.

The potential benefits need to be balanced against the morbidity and mortality risks of treatment and the likelihood of sequencing of therapies during the patient's lifespan.

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

The current recommended therapeutic approach is based on the 2022 Myeloma Australia MSAG guidelines (Teh et al. 2022).

Watchful waiting

Many patients with newly diagnosed WM have asymptomatic smouldering disease, which does not warrant treatment.

Many people diagnosed with WM will not start treatment immediately but instead have regular check-ups for symptoms. This is known as 'active surveillance' or 'watch and wait'. As WM is often a slow growing disease, it is a safe strategy which means people diagnosed with asymptomatic WM can avoid the side-effects that treatment can bring. Even for people who have symptoms of WM, 'watch and wait' might still be a suitable strategy if those symptoms are not severe enough to warrant urgent treatment.

Watchful waiting can cause significant anxiety in some patients. Appropriate education and support should be offered (see section 4.2.3. 'Supportive therapy').

Consensus-defined indications for considering therapy are (MSAG guidelines (Teh et al. 2022):

- IgM paraprotein >60 g/L independent of symptoms, especially when inexorably rising
- symptomatic hyperviscosity
- Hb <100 g/L
- neutrophils <1.0×10^9/L
- platelets <100×10^9/L
- symptomatic organomegaly
- bulky (>5 cm) and/or symptomatic lymphadenopathy
- symptomatic infiltration of other tissues
- immune haemolytic anaemia or thrombocytopenia
- fever, night sweats, weight loss or fatigue attributed to WM
- peripheral neuropathy
- nephrotic syndrome
- amyloidosis
- symptomatic cryoglobulinaemia.

IgM-related neuropathy is an accepted indication for WM therapy.

4.2.1 Systemic therapy

The most commonly adopted, currently available approach for treating WM in Australia is rituximab-based chemotherapy, either in combination with dexamethasone and cyclophosphamide (DRC), or bendamustine (BR). For patients considered ineligible for rituximab-chemotherapy, single-agent zanubrutinib BTK inhibitor is PBS-funded for patients with a CIRS score ≥6 (Castillo et al. 2020).

Decisions about first-line treatment need to consider the tempo of the disease (WM can take several years of indolent progression to reaching a need for therapy), patient comorbidities and wishes, and PBS criteria, as well as the likely expectation of sequencing of available therapies in the future. For example, rituximab-based chemotherapy and bendamustine are currently only available in first-line treatment of WM, and not for patients with relapsed disease. Rituximab-based chemotherapy is a fixed duration first-line approach, whereas zanubrutinib requires continuous oral therapy until disease progression or intolerance (Tam et al. 2020; Trotman et al. 2020).

Unlike the other low-grade lymphoproliferative diseases, there is no data to support a role for rituximab-based chemotherapy maintenance after first-line therapy for WM, with no PFS advantage to this approach (Rummel et al. 2016).

Most patients with WM respond well to their first-line treatment, with 70% of patients having a partial response or better (more than 50% reduction in IgM levels). Incremental responses can continue to improve over months – years after starting treatment (Castillo et al. 2020, Rummel et al. 2013). There can be discordance between IgM level and clinical response. Even patients without a major (>50%) reduction in IgM may have an excellent symptomatic and haemoglobin response to therapy and should not be considered a treatment failure.

Monitoring pre- and post-treatment and response assessment:

- Patients with WM require monitoring of IgM during the asymptomatic phase and post-treatment for response assessment. It is important to note that (unlike myeloma) classification of response is based on the IgM level, not the paraprotein measurement. It is crucial not to regard an only minor decline in IgM as a marker of therapy failure in isolation from other indications of therapeutic response such as improved haemoglobin levels and/or resolution of symptoms. Ideally, patients should be monitored at the same laboratory to ensure consistency of results.
- BMAT for monitoring is not incorporated in routine care.
- Neutropenia and mild thrombocytopenia is a common consequence of both immunochemotherapy and zanubrutinib and should be closely monitored. G-CSF treatment is effective at reversing neutropenia.

Treatment for Bing-Neel

This rare CNS manifestation of WM requires CNS-penetrating therapies. R-DC and R-bendamustine are not appropriate therapies for patients presenting with Bing-Neel syndrome. Ibrutinib has demonstrated efficacy in Bing-Neel syndrome, although it is not currently funded in Australia. Seeking therapy on compassionate grounds may be useful in this context. Zanubrutinib crosses the blood–brain barrier and is likely to be similarly effective in this condition. Systemic and intrathecal therapies including cytarabine and methotrexate can be considered.

4.2.2 Timeframe for starting treatment

Rate of disease progression is usually gradual and treatment initiation is rarely urgent. Timing should be discussed to align with the patient's preferences but not delayed to the point where progressive fatigue, impaired performance status, or compromised organ function occurs.

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)
- coordination for combined therapy with radiation therapy, especially where facilities are not co-located.

4.2.3 Supportive therapy

Plasmapheresis

- Patients with symptomatic hyperviscosity should be referred urgently to a centre with capabilities for plasmapheresis.
- In asymptomatic patients with IgM levels over 60 g/L, plasmapheresis is recommended if starting therapy with rituximab-based chemotherapy due to the risk of IgM flare, which may take two to four months to resolve (Castillo et al. 2020). Alternatively, to reduce the risk of IgM flare, the chemotherapy component may be administered without rituximab-based chemotherapy for the initial two cycles when treating patients with IgM levels >40 g/L.
- Avoidance of red cell transfusions is recommended; if necessary, this should be timed to follow plasmapheresis.
- Careful fluid management to prevent exacerbation of hyperviscosity and acute pulmonary oedema is required.

Pausing BTK inhibitor therapies and managing withdrawal symptoms

- BTKi may require a three- to seven-day pause before and after surgery depending on the surgery and patient's bleeding risk.
- BTKi withdrawal symptoms, characterised by fever, body ache, night sweats, arthralgia, headache and fatigue (seen in 10–20% of patients when pausing BTKi for surgery), should be managed with a short course of oral prednisone (Castillo et al. 2016).
- An IgM rebound is commonly seen when stopping or interrupting BTKi.

Prevention and management of infection

- Routine and additional vaccinations should ideally be given before immunosuppressive therapy. In addition to the standard Australian adult vaccination schedule, people who are immunocompromised are advised to receive:
  - a yearly influenza vaccine
  - one extra pneumococcal vaccine 'booster dose' in addition to the general population
  - additional doses of meningococcal, HPV and hepatitis B vaccines can be considered on a case-by-case basis
  - recombinant zoster vaccination is recommended but the high cost may mean it is not accessible for all patients (see note below re: the live attenuated herpes zoster vaccine).
- COVID-19 disease – patients with WM should be encouraged to be up to date with COVID-19 vaccination including the maximum recommended booster vaccines as per government guidelines. Ideally, COVID-19 vaccinations should be given weeks before immunosuppressive therapy, particularly if B-cell depletion is being prescribed (e.g. rituximab, BTKi). Patients on BTKis, or who have recently received or are currently receiving rituximab-chemotherapy, should be considered for tixagevimab and cilgavimab (Evusheld). The efficacy of Evusheld against new and evolving SARS-COVID variants is unclear.
- Live attenuated vaccines are contraindicated in people who are immunocompromised, and this includes patients with WM on certain therapies. Live attenuated vaccines include BCG, Japanese encephalitis, MMR, rotavirus, oral typhoid, varicella, yellow fever and zoster vaccination. Consultation with the treating haematologist and/or immunologist is advised if considering giving these vaccines.

- Manage neutropenia – granulocyte colony-stimulating factor support may be required for patients receiving chemotherapy or oral therapies (especially zanubrutinib) due to the onset of neutropenia, to reduce the risk of febrile neutropenia illness.
- Prevent infection – prophylactic antimicrobials should be used according to clinical and institutional guidelines, incorporating risk factors such as prior infection and prior line(s) of treatment.
- Manage hypogammaglobulinaemia and recurrent infections – intravenous or subcutaneous immunoglobulin therapy should be considered in patients with severe hypogammaglobulinaemia and recurrent infections according to version 3 of Ig criteria.

Nephropathy management

- The presence of nephropathy often requires careful diuresis and fluid management due to extravascular fluid in consultation with a renal physician.

Neuropathy management

- The sensorimotor neuropathy of WM can be partially reversed with rituximab-based therapy, and with BTKi use in some patients. Management of neuropathy should be undertaken in conjunction with a neurologist.
- The use of proteasome inhibitors should be avoided in patients with neuropathy, and given such neuropathy is often occult, careful screening during the administration of proteasome inhibitors in the relapsed/refractory setting is important.
- Occupational therapy and physiotherapy support should be used to preserve or enhance function. Consideration of multimodal pain management tools is recommended where pain is present.

Iron deficiency

- Patients with WM have excessive production of hepcidin, impairing oral iron absorption, and leading to functional iron deficiency. Intravenous iron supplements may be necessary to maintain a ferritin over 200 ug/L and transferrin saturation over 15%.

Information about vaccinations for patients with WM

Patients with WM should be up to date with the Australian adult vaccination schedule.

Vaccination – routine and additional vaccinations

If required, any vaccinations to complete this should ideally be given before immunosuppressive therapy. In addition to the standard Australian adult vaccination schedule, people who are immunocompromised are advised to receive:

- a yearly influenza vaccine
- one extra pneumococcal vaccine ‘booster dose’ in addition to the general population
- additional doses of meningococcal, HPV and hepatitis B vaccines can be considered on a case-by-case basis
- recombinant zoster vaccination (Shingrix) is recommended, but the high cost may mean it is not accessible for all patients (see note below re: the live attenuated herpes zoster vaccine).

Vaccination COVID-19

People with haematological malignancies and those on immunosuppressive therapies are at high risk of severe COVID-19 disease and also have a less efficient responses to vaccination. Patients with WM should be encouraged to be up to date with COVID-19 vaccination including the maximum recommended booster vaccines as per government guidelines. Ideally, COVID-19 vaccinations should be given before immunosuppressive therapy, particularly if B-cell depletion is being prescribed (e.g. rituximab, BTKi). Patients who have recently received or are currently receiving immunosuppressive therapy should be considered for tixagevimab and cilgavimab (Evusheld) or other TGA-approved passive antibody therapies if current COVID-19 variants are sensitive.

Vaccination: Live attenuated vaccines

Live attenuated vaccines are contraindicated in people who are immunocompromised, and this includes patients with WM on certain therapies. Live attenuated vaccines include BCG, Japanese encephalitis, MMR, rotavirus, oral typhoid, varicella, yellow fever and zoster vaccines. Consultation with the treating haematologist and/or immunologist is advised if considering giving these vaccines.

4.2.4 Radiation therapy

Radiation therapy has a role in the palliative care of patients with significant symptomatic nodal disease despite multiple systemic therapies for WM. When used for symptom control it may improve quality of life.

Timeframes for treatment should reflect the overall goals of care and severity of symptoms.

Training and experience required of the appropriate specialists

There are no specific sub-specialty skill needs required for the delivery of palliative radiation in WM beyond those generally required for registration and practice as a radiation oncologist.

The training and experience of the radiation oncologist should be documented.

Health service unit characteristics

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

- linear accelerator (LINAC) capable of image-guided radiotherapy (IGRT)
- dedicated CT planning
- access to MRI and PET imaging
- automatic record-verify of all radiation treatments delivered
- a treatment planning system
- medical physicists, radiation therapists and nurses with radiation therapy experience
- coordination for combined therapy with systemic therapy, especially where facilities are not co-located
- participation in Australian Clinical Dosimetry Service audits
- an incident management system linked with a quality management system.

4.2.5 Emerging therapy

Future trends for treating relapsed refractory WM include BCL2 antagonists (e.g. venetoclax) or multiagent regimens with a proteasome inhibitor backbone (bortezomib, ixazomib, carfilzomib). Agents targeting CXCR4 or P13K, bi-specific antibodies and CD19 CAR-T therapies are in ongoing clinical trials, as are other later generation reversible covalent BTKi (acalabrutinib), emerging reversible non-covalent BTKi (tirabrutinib, orelabrutinib, vecabrutinib) and the promising noncovalent BTKi pirtobrutinib (Castillo et al. 2022).

Venetoclax and ibrutinib should not be used in combination due to reports of fatal ventricular arrhythmia.

4.2.6 Target therapies and immunotherapy

The BTI inhibitor zanubrutinib is the only PBS-funded targeted therapy for WM and is generally recommended as a second-line therapy unless the patient is unsuitable for rituximab chemotherapy.

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 WM. 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
- management of physical symptoms such as nausea, diarrhoea, fatigue or other specific treatment-related adverse events that will vary by agent
- 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 cancer 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 and WMozzies <www.wmozzies.com.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 cancer, all team members should recommend that people with cancer 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 cancer 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 cancer-related fatigue, improving physical endurance, achieving independence in daily tasks, optimising nutritional intake, returning to work and ongoing adjustment to cancer 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.

Cancer survivors may experience inferior quality of life and cancer-related symptoms for up to five years after their diagnosis (Jefford et al. 2017). Distress, fear of cancer recurrence, fatigue, obesity and sedentary lifestyle are common symptoms reported by cancer survivors (Vardy et al. 2019).

Due to an ageing population and improvements in treatments and supportive care, the number of people surviving cancer is increasing. International research shows there is an important need to focus on helping cancer survivors cope with life beyond their acute treatment. Cancer survivors often face issues that are different from those experienced during active treatment for cancer 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 cancer recurrence, cancer-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 active treatment

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 cancer 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

Responsibility for follow-up care should be agreed between the lead clinician, the general practitioner, relevant members of the multidisciplinary team and the patient. This is based on guideline recommendations for follow up care, as well as the patient's current and anticipated physical and emotional needs and preferences.

WM is a highly treatable low-grade lymphoma. While incurable with current therapies, many patients with WM have such a prolonged survival that they may have a 'functional cure' of their WM. Patients with WM may need multiple courses of therapy over their lifetime, given the relapsing-remitting nature of the lymphoma. Monitoring for late effects and second malignancies is required, and primary prevention and risk minimisation strategies are encouraged. Responsibility for follow-up care should be shared between the haematologist, general practitioner, relevant members of the multidisciplinary team and the patient.

Monitoring for disease progression (assessing symptoms, IgM and FBC) is an integral part of care and occurs during regular clinic visits. Symptomatic disease progression will trigger reassessment and consideration of the introduction of therapy.

Patient-reported outcomes

Patients often keep records of their symptoms and IgM levels, prompted by their increased WM-specific health literacy through engagement with WMozzies / the IWMF and participation in the WhiMSICAL registry, empowering patients to be true partners in their care.

Specific monitoring for complications of therapy should include:

- after rituximab-chemotherapy – history, physical examination and blood tests every three months for at least two years, then clinician-directed follow-up intervals indefinitely (Kastritis et al. 2018)
- patients on zanubrutinib need to be seen every three months for FBC and IgM levels and clinical review; in stable patients this may be conducted alternately with the haematologist and general practitioner, who is well placed to monitor and initiate investigations for secondary malignancies
- supportive care for WM-related fatigue
- monitoring for second malignancies, including age-appropriate malignancy screening, skin cancers, bowel cancers, prostate cancers and secondary myeloid neoplasms such as the myelodysplastic syndrome (related to prior alkylating agent therapy).

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 (surveillance for recurrence or secondary and metachronous cancers, 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 cancer and cancer 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 cancer 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 <https://www.mycareplan.org.au/>
- South Australian Cancer Service – Statewide Survivorship Framework resources
- American Society of Clinical Oncology – guidelines.

5.2.1 Preventing progression

People frequently ask if there is anything else they can do to reduce the risk of WM progression. 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 WM 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 <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 cancer 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.

5.4.2 Supportive therapies

Patients with WM may experience multiple lines of treatment over many years and therefore benefit from reassessment of their supportive care needs on an ongoing basis, but particularly:

- at diagnosis
- before each subsequent line of therapy
- when experiencing more serious side effects and health issues
- when nearing end of life or palliative care.

The ongoing needs of survivorship for patients with WM should be routinely addressed. Using a structured framework such as the Australian Survivorship Care Plans may be useful for clinicians.

Support groups such as WMozzies are available for guidance. For patients, they reduce social isolation, inform and improve health literacy by providing treatment information via newsletters, online forums and links to other useful support and information. The WMozzies website also has links to the WhiMSICAL patient-derived data registry capturing treatment outcomes, quality-of-life information and COVID-19 data.

5.4.3 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 cancer-related fatigue, coping with cognitive changes, improving physical endurance, achieving independence in daily tasks, returning to study or work and ongoing adjustment to cancer and its consequences.

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

5.4.4 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 immediate and long term 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.

5.4.5 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 refractory, relapsed or progressive disease

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

6.1 Signs and symptoms of relapsed or progressive disease

Patients may suspect relapsed or progressive disease if they have recurrent symptoms, or via FBC and IgM surveillance. The spectrum of possible symptoms is similar to those at presentation. In addition, the potential for transformation to large cell lymphoma should be considered in the uncommon situation of rapid onset of B symptoms or rapidly enlarging lymphadenopathy.

6.2 Managing refractory, relapsed or progressive disease

Managing refractory, relapsed or progressive disease is complex, particularly after a second progression, 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 WM 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 cancer 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

Treatment will depend on the tempo of disease progression, previous therapy, toxicity from prior treatment and the patient's preferences. Asymptomatic relapse or progression can be managed with observation.

In managing people with relapsed or progressive WM, considerations include:

- the choice of second and subsequent lines of treatment – this is strongly influenced by the prior treatment regimens and duration of response
- with additional immunosuppressive effects of therapy, the risk of opportunistic infection rises. Prophylaxis is important with antimicrobials and/or immunoglobulin replacement as appropriate. Similarly, consider G-CSF as a primary or secondary prophylaxis for periods of neutropenia.

Second-line and subsequent therapies

The following should be considered:

- non-chemotherapy regimens with a BTKi such as zanubrutinib (for patients who have not had prior BTKi exposure)
- rituximab-chemotherapy regimens (R-DC or R-bendamustine if not used previously) – this may include repeating the first-line rituximab-chemotherapy (especially R-DC) if well tolerated and the remission duration was for several years (note that R-bendamustine is TGA-approved but not PBS-funded in this setting at the time of publication)
- other combinations used in the multiply relapsed setting that are not PBS-funded include:
  - proteasome inhibitor combination regimens, covalent BTKi and BCL 2 inhibitors (note that these are not TGA-approved or PBS-funded at the time of publication)
  - R-CHOP regimen for WM in the context of histological transformation – vincristine should be used with care and avoided in patients with neuropathy (Castillo et al. 2020).
  - purine nucleoside analogue therapies (e.g. fludarabine, cladribine) used historically to treat WM are contraindicated due to profound immunosuppression. Also, these therapies are associated with an increased risk of secondary malignancies such as MDS or AML, and are associated with stem cell toxicity for potential transplant-eligible patients.

Autologous and allogeneic transplantation should be considered only in highly selected young, fit patients with at least partial response, aggressive disease kinetics and lack of other therapeutic options (Pratt et al. 2022).

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 cancer diagnosis but especially those with repeated progressions and reduced response duration signalling 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 cancer-related fatigue, improving physical endurance, achieving independence in daily tasks, returning to work and ongoing adjustment to cancer and its consequences.

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

A conversation about elective cessation of continuous oral therapies is advised, but a history of any BTKi withdrawal symptoms should be taken into consideration and managed.

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
- Canteen for children of parents with cancer <www.canteen.org.au/young-people/my-parent-has-cancer>.

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>
- 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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Our thanks also to the Blood Cancer Taskforce, which recommended the development of optimal care pathways for all of the major blood cancer subtypes as part of the *National strategic action plan for blood cancer* (2020). The national action plan was commissioned by the federal government and developed by the Blood Cancer Taskforce, with support from the Leukaemia Foundation.

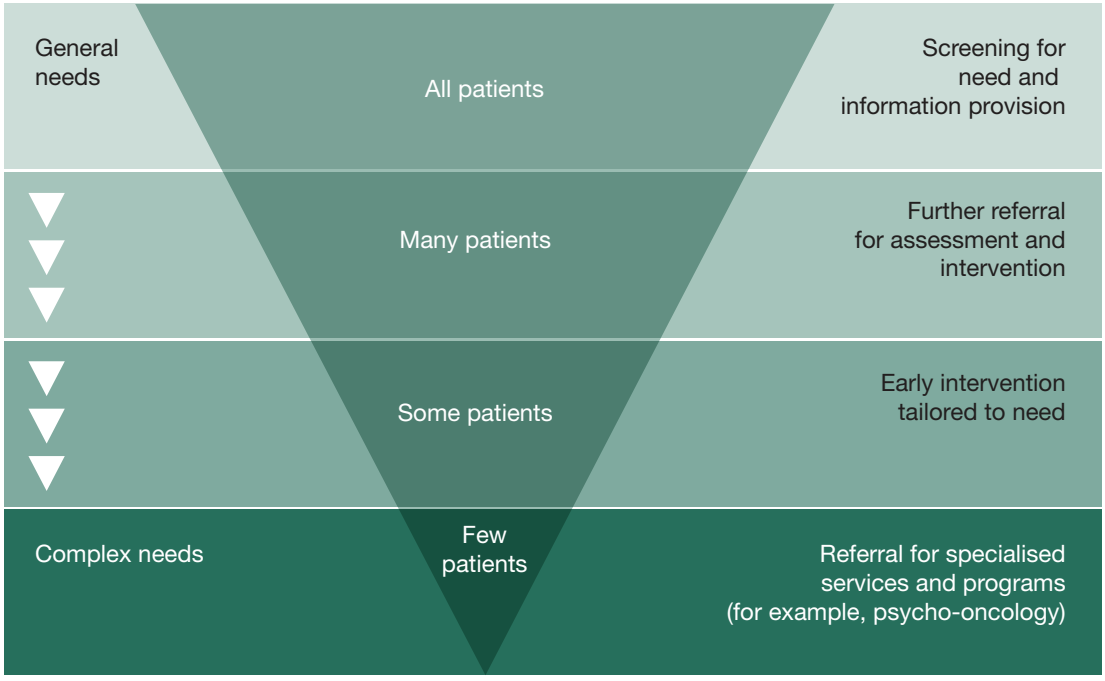
Appendix A: Supportive care domains

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 per cent 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 WM

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 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>

WMozzies

- Website <https://wmozzies.com.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.

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 in 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.

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.

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.

relative survival rate – the probability of being alive for a given amount of time after diagnosis compared with the general population.

risk stratification – a systematic process to target and identify select patients who are at risk of poorer health outcomes, and who are expected to benefit most from a particular intervention or interventions.

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.

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

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