

Optimal care pathway for people with Hodgkin and diffuse large B-cell lymphomas

Quick reference guide



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

The optimal care pathways describe the standard of care that should be available to all cancer patients treated in Australia. The pathways support patients and carers, health systems, health professionals and services, and encourage consistent optimal treatment and supportive care at each stage of a patient's journey. Seven key principles underpin the guidance provided in the pathways: patient-centred care; safe and quality care; multidisciplinary care; supportive care; care coordination; communication; and research and clinical trials.

This quick reference guide provides a summary of the *Optimal care pathway for people with Hodgkin and diffuse large B-cell lymphomas*.

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

Step 1: Prevention and early detection

Prevention

The causes of Hodgkin lymphoma and diffuse large B-cell lymphomas (DLBCL) are not fully understood, and there is currently no clear prevention strategy.

Risk factors

All ages are at risk for Hodgkin lymphoma and DLBCL. However, DLBCL is most common in middle-aged to older adults. There is a 'peak' of higher incidence of Hodgkin lymphoma in adolescents and young adults, as well as older adults. Other risk factors include:

- intrinsic immunosuppression – patients who have received an organ transplant, are HIV-positive or are being therapeutically treated with immunosuppressants

- Epstein-Barr virus infection with immune deficiency
- family history of lymphoproliferative disorder.

Risk factors specific to DLBCL include gender (males have a slightly higher risk), a high BMI, B-cell activating autoimmune diseases, prolonged hair dye use, obesity and personal or family history of a lymphoproliferative disorder.

Early detection

There is no established role for screening in Hodgkin lymphoma or DLBCL. People at risk of developing immunodeficiency-associated lymphoma should be made aware of this increased risk.

Checklist

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

Step 2: Presentation, initial investigations and referral

The following signs and symptoms are common:

- any abnormal lump or mass in any organ
- lymphadenopathy:
 - persistent beyond 2 weeks
 - associated with systemic symptoms (see next bullet point)
 - that does not resolve despite appropriate treatment of infection
 - associated with pain in the lymph nodes following alcohol consumption

- one or more of these symptoms even without lymphadenopathy:
 - persistent unexplained fever, drenching sweats, unintentional weight loss, persistent severe itch and frequent infections.

Checklist

- Signs and symptoms recorded
- Investigations completed
- Supportive care needs assessment completed and recorded, and referrals to allied health services actioned as required
- Patient notified of support services such as Cancer Council 13 11 20
- Referral options discussed with the patient and/or carer including cost implications

Step 2: Presentation, initial investigations and referral continued

Initial investigations

If there is a high likelihood of a malignant process, prompt referral to a specialist centre for a tissue diagnosis is appropriate.

For others, further investigations should be completed and may include:

- full blood examination
- imaging of the affected areas using ultrasound, x-ray and CT, as appropriate
- biopsy, as appropriate
- a period of observation of **up to 4 weeks** for patients without significant or progressive symptoms.

Referral options

At the referral stage, the patient's GP or other referring doctor should advise the patient about their options for referral, waiting periods, expertise, if there are likely to be out-of-pocket costs and

the range of services available. This will enable patients to make an informed choice of specialist and health service.

All patients with suspected lymphoma should be evaluated and cared for by a lymphoma-specific multidisciplinary team. Healthcare providers should provide clear routes of rapid access to specialist evaluation.

Communication

The GP's responsibilities include:

- explaining to the patient and/or carer who they are being referred to and why
- supporting the patient and/or carer while waiting for specialist appointments
- informing the patient and/or carer that they can contact Cancer Council on 13 11 20.

Timeframe

For patients who don't need a prompt referral, all investigations should be completed, and a path of action decided, **within 4 weeks** of their first presentation.

Patients should be referred to a specialist:

- **within 72 hours** if the presence of lymphoma is highly likely
- **within 4 weeks** if indicators of concern are absent.

If under observation, the patient should be reviewed by their GP **within 6 weeks** of initial presentation.

Step 3: Diagnosis, staging and treatment planning

Diagnosis

A tissue diagnosis is required before initiating definitive treatment, although one may have been performed before referral. Fresh and fixed tissue samples should be collected from the tissue biopsy for anatomical pathology, flow cytometry, cytogenetics and gene mutation testing.

Staging

The disease stage, including evaluation of bone marrow status if full blood test results are significantly abnormal, should be determined. This should include an FDG-PET/CT scan.

Genetic testing

Currently there are no genetic abnormalities known to predispose lymphoma. Any referral made to a familial cancer service should be based on multidisciplinary team recommendations. There are currently no known mutations that require routine testing in these disorders.

Treatment planning

A multidisciplinary meeting (MDM) should be conducted **before implementing treatment**. A meeting may not be possible if treatment is urgent, but the treatment plan should still be ratified.

Research and clinical trials

Consider enrolment where available and appropriate. Search for a trial <www.australiancancertrials.gov.au>.

Communication

The lead clinician's¹ responsibilities include:

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

Checklist

- Diagnosis confirmed
- Full histology obtained
- Performance status and comorbidities measured and recorded
- Patient discussed at an MDM and decisions provided to the patient and/or carer
- Clinical trial enrolment considered
- Supportive care needs assessment completed and recorded and referrals to allied health services actioned as required
- Patient referred to support services (such as Cancer Council) as required
- Treatment costs discussed with the patient and/or carer

Timeframe

Timing of diagnostic investigations should be guided by the initial severity of symptoms. Staging should be completed **within 2 weeks**.

¹ Lead clinician – the clinician who is responsible for managing patient care.

The lead clinician may change over time depending on the stage of the care pathway and where care is being provided.

Step 4: Treatment

Establish intent of treatment

- Curative
- Anti-cancer therapy to improve quality of life and/or longevity without expectation of cure
- Symptom palliation

Systemic therapy: The vast majority of patients with Hodgkin lymphoma or DLBCL are likely to benefit from systemic therapy. A range of biological and targeted therapies are increasingly being used to treat patients with these lymphomas.

Radiation therapy should be considered for suitable patients with localised disease or those with more advanced disease with a dominant bulky lesion. Radiation therapy is usually used in conjunction with chemotherapy.

Palliative care:

Early referral to palliative care can improve quality of life and in some cases survival. Referral should be based on need, not prognosis. For more, visit the Palliative Care Australia website <www.palliativecare.org.au>.

Communication

The lead clinician and team's responsibilities include:

- discussing treatment options with the patient and/or carer including the intent of treatment as well as risks and benefits
- discussing advance care planning with the patient and/or carer where appropriate
- communicating the treatment plan to the patient's GP
- helping patients to find appropriate support for exercise programs where appropriate to improve treatment outcomes.

Checklist

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

Timeframe

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

Step 5: Care after initial treatment and recovery

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

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

- a process for rapid re-entry to medical services for suspected recurrence.

Potential late effects of therapy that may require specific screening and monitoring will be determined by the primary treatment used.

Communication

The lead clinician's responsibilities include:

- explaining the treatment summary and follow-up care plan to the patient and/or carer
- informing the patient and/or carer about secondary prevention and healthy living
- discussing the follow-up care plan with the patient's GP.

Checklist

- Treatment and follow-up summary provided to the patient and/or carer and the patient's GP
- Supportive care needs assessment completed and recorded and referrals to allied health services actioned as required
- Patient-reported outcome measures recorded

Step 6: Managing recurrent or residual disease

Detection

Most cases of recurrent Hodgkin lymphoma or DLBCL are identified through routine follow-up or by the patient presenting with symptoms, or with abnormal 'non-specific' laboratory tests such as serum LDH.

Treatment

Evaluate each patient for whether referral to the original multidisciplinary team is appropriate. Treatment will depend on the location and extent of disease, previous management and the patient's preferences. Combination chemotherapy regimens with autologous stem cell transplantation is considered in first relapse. In subsequent relapses or where autologous stem cell transplantation is not appropriate, clinical trials should be considered. Novel biologic agents may be indicated, in the case of Hodgkin lymphoma.

Advance care planning

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

Survivorship and palliative care

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

Communication

The lead clinician and team's responsibilities include:

- explaining the treatment intent, likely outcomes and side effects to the patient and/or carer and the patient's GP.

Checklist

- Treatment intent, likely outcomes and side effects explained to the patient and/or carer and the patient's GP
- Supportive care needs assessment completed and recorded and referrals to allied health services actioned as required
- Advance care planning discussed with the patient and/or carer
- Patient referred to palliative care if appropriate
- Routine follow-up visits scheduled

Step 7: End-of-life care

Palliative care

Consider a referral to palliative care. Ensure an advance care directive is in place.

Communication

The lead clinician's responsibilities include:

- being open about the prognosis and discussing palliative care options with the patient
- establishing transition plans to ensure the patient's needs and goals are considered in the appropriate environment.

Checklist

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

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.