Lymphoma case based learning resource

Overview of the lymphoma case study: Arthur's story

This case study recounts the experience of Arthur, a 78-year-old male diagnosed with non-Hodgkin's Lymphoma (NHL).

The case study contains five sections:

- 1. Reduce risk.
- 2. Find the condition early.
- 3. Have the best treatment and support during active treatment.
- 4. Have the best treatment and support between and after active treatment.
- 5. Have the best care at the end of life.

It is recommended that you complete the sections and their related activities in order. This is because each section and each activity includes information that will help you complete the sections and activities that follow.

Learning activities

At times, you will have learning activities to complete. Click on the learning activities button and a list of questions will pop up. The questions will relate to the content you've just read or the video you've just watched.

Videos

There is a video component to this case study that is presented in seven parts. You can watch the video clips when prompted throughout this case study or at any time by clicking on the video icon in the right-side menu. Learning activities throughout the case study will discuss the video and ask questions about it.

Resource Links

Resource links are included throughout the case study. These links lead to interesting articles or websites, and are designed to encourage you to explore other available resources.

PDF of lymphoma module

You can download a PDF version of the lymphoma module.

Suggested citation:

Cancer Australia. (2018) EdCaN module: Lymphoma case based learning resource, version 2.2.

Aim of the lymphoma case study

This case study aims to facilitate the development of competencies that reflect the role of the Specialist Cancer Nurse (SCN) in providing a coordinated approach to care planning, implementation and evaluation for people affected by NHL across the cancer journey.

Rationale

Non-Hodgkin lymphoma was the most common cancer affecting the blood and lymphatic system in Australia in 2011. Overall, it was the sixth most commonly diagnosed cancer and the seventh most common cause of cancer death. ⁴

Lymphoma is more common in the older person. Older people diagnosed with cancer can have unique support needs, associated with co-morbidities, and changes in general health, functional status, and social circumstances.

This case study focuses on the SCN's role in the provision and coordination of care for people with NHL, as well as exploring issues associated with diagnosis of cancer in the older person.

There are many points along the cancer journey when SCNs can improve outcomes for people at risk or affected by NHL. These include:

Section 1: Reduce risk

The causes of most lymphomas, including NHL, are unknown, although identified risk factors include an underlying immunodeficiency condition, presence or history of specific infections, occupational risks and medical / co-morbidity risks.⁵

While many of the identified risks are difficult to avoid and there is limited practical benefit for prevention programs, the SCN can promote avoidance of risk factors and community education about these risks.

Section 2: Find the condition early

Many of the symptoms associated with lymphomas including NHL replicate common symptoms of influenza and common viruses which are frequently seen in primary health care settings.⁶

The SCN has an important role in educating primary care colleagues about the signs and symptoms of NHL to promote efficient referral to specialist services and timely commencement of treatment.

Section 3: Have the best treatment and support during active treatment

Treatment for NHL depends on a range of factors associated with the type and grade of the disease. The SCN can assist people affected by NHL to understand the rationale for treatments, and support them to make decisions congruent with their needs and wishes.

Antineoplastic agents are commonly used in the treatment of NHL,⁷ and this may be used in combination with radiotherapy and/or monoclonal antibodies. These treatments can be associated with significant effects and toxicities which require evidence based management.

Care of the older person with cancer raises unique challenges for the multidisciplinary team (MDT).⁸ The SCN has a role in ensuring that any barriers to optimal care are addressed, and that treatment and care of the older person with cancer is optimal.

Section 4: Have the best treatment and support between and after active treatment

While prognosis following treatment for NHL has improved, completion of treatment is often associated with significant fears about relapse as well as managing adverse effects of treatment. The care coordinator is a key member of the MDT, providing support for the person affected by lymphoma throughout the cancer journey.

Section 5: Have the best care at the end of life

Advance care planning is integral to the management of older people with life limiting conditions such as cancer. The SCN supports people affected by NHL to voice their wishes regarding individual care choices at end of life.

An evidence based multidisciplinary approach is required for management of symptoms associated with progressive disease. ¹⁰ The SCN acts as a resource to colleagues in other settings regarding strategies for effective symptom management.

Carers of people with cancer face a range of competing demands, and caregiving can negatively impact on the carer's health. 11 SCNs play an important role in supporting carers and facilitating access to carer support services.

Section 1: Reduce risk

Objectives

On completion of this section, you should be able to:

- 1. Interpret key epidemiological trends in the incidence, mortality and survival from lymphoma.
- 2. Explain current evidence regarding risk factors associated with the development of lymphoma.

Non-Hodgkin Lymphoma in Australia

In 2013, there were 5,589 new cases of non-Hodgkin lymphoma in Australia (3,151 new cases in men and 2,438 new cases in women). In 2013, the age standardised incidence rate was 22 cases per 100,000 persons.⁵¹

The incidence of NHL has increased steadily from 1982 (1,918 cases) to 2013 (5,589 cases).51

Incidence of NHL increases with age, with the average age at diagnosis from males at 64.3 and females 66.8.4 Up to 85 years, males have a greater risk of developing NHL (1 in 34) compared to females (1 in 50).4

In 2014, there were 1,598 deaths from NHL, with lymphoma the 9th leading cause of cancer death in Australia.⁵¹ There have been significant survival gains for non-Hodgkin lymphoma. Five-year survival increased significantly between 1982–1986 and 2007–2011, from 44.8% to 72.1%.⁴ Increases in survival from non-Hodgkin lymphoma have been attributed to more effective treatment, particularly the introduction of antibody therapies.¹³

At the end of 2012, there were 5,097 people living who had been diagnosed with lymphoma that year, 20,200 people who had been diagnosed with lymphoma in the previous 5 years (from 2008 to 2012) and 50,750 people who had been diagnosed with lymphoma in the previous 31 years (from 1982 to 2012). 51

Learning activity		
Completed	Activity	
	Access <u>Cancer in Australia: an overview. 2014</u> and <u>Cancer incidence</u> <u>projections, Australia 2011 to 2020</u> ¹⁴ , and:	
	 Review the cancer incidence projections for NHL for 2011-2020 and discuss the projected trends for the incidence of NHL among both males and females. 	
	 Discuss the implications of the above trends and changes in demography (i.e. an increased ageing population), for the incidence of NHL in Australia. 	

Risk factors

While the causes of most lymphomas are unknown, risk factors associated with developing lymphoma have been identified. The following risk factors for NHL have been reported:5, 15

- immunodeficiency (post-transplantation immunosuppression; HIV/AIDS; congenital immune deficiency; acquired autoimmune disease).
- infectious organisms (Epstein Barr Virus; Helicobacter pylori for gastric lymphoma; Human T-lymphotrophic virus types I (HTLV-I); Human herpes virus- (HHV8) infection for primary effusion lymphoma).
- occupational exposures (to pesticides or herbicides; farming).
- medical and co-morbid factors (childhood appendectomy; skin cancer; diabetes; tuberculosis).
- lifestyle factors (cigarette smoking).

Systematic reviews have identified individual risk factors including wood preserving chemicals ¹⁶ and hepatitis C. ¹⁷

Learning activity	
Completed	Activity Access the following resources to respond to these learning activities: • Clinical practice guidelines for the diagnosis and management of lymphoma ¹⁵ • Pentachlorophenol and cancer risk: focusing the lens on specific chlorophenols and contaminants ¹⁶ Risk of NHL and lymphoproliferative precursor diseases in US Veterans with hepatitis C virus ¹⁷
	 1 Appraise the strength of evidence in relation to the listed risk factors for NHL: HIV/AIDS infection. Epstein-Barr virus. Hepatitis C. cigarette smoking. pentachlorophenol exposure.
	2. Using an evidence based approach, discuss how you would support and advise an individual's relative who approached you seeking guidance in relation to their concerns of developing lymphoma after being diagnosed with hepatitis C.

Section 2: Find the condition early

Objectives

On completion of this section, you should be able to:

- 1. Describe common symptoms associated with NHL.
- 2. Identify and discuss investigations that may be performed when NHL is suspected.
- 3. Describe common concerns and reactions of people with symptoms which may be associated with NHI
- 4. Implement strategies to provide information, education and support to people undergoing investigation of NHL symptoms.

Early detection

Despite a number of classical symptoms that occur on presentation of NHL, it can be a difficult condition to diagnose. NHL has been reported in most sites of the body, and many of the symptoms are similar to common symptoms associated with non-serious conditions such as influenza and infections.⁶

Low grade or indolent NHL usually presents with painless swelling of the lymph nodes and a history of intermittent lymphadenopathy. Bone marrow involvement is common and may be associated with cytopenias. Splenomegaly is seen in approximately 30% to 40% of cases.

In aggressive (intermediate) grade NHL, fatigue, weakness, and lymphadenopathy are also common. Approximately 30% to 40% of individuals also have B symptoms, which may be associated with more advanced or aggressive disease. These include: 18, 19

- unexplained fever
- unintentional loss of greater than 10% of body weight in the six months before diagnosis
- drenching night sweats.

Individuals with aggressive B-cell lymphomas present with large abdominal or mediastinal masses. Other less common symptoms include:⁶

- malaise
- itching
- nausea and vomiting
- coughing
- headaches
- susceptibility to infections.

There are no specific screening tests for lymphoma. For individuals at risk of immunodeficiency-associated lymphoma, surveillance is recommended.²⁰

Meet Arthur

Case study: meet Arthur

Arthur is a 78-year-old-male who has recently been diagnosed with NHL.

Watch Arthur's first video and then work through the learning activities. Use your notebook at the top of the screen to answer the learning activity questions and record your thoughts.

Arthur's Story 1: Meet Arthur



Learning activities			
Completed		Activities	
		Discuss strategies and resources that may improve the awareness of the symptoms of lymphoma among primary care colleagues caring for Arthur.	
		2 Identify symptoms Arthur described which may relate to a diagnosis of NHL.	
		3 Describe the pathophysiological basis to Arthur's symptoms in relation to NHL.	
		4 Discuss other potential causes for Arthur's symptoms.	

Responding to symptoms

An accurate assessment of individuals with suspected lymphoma requires a range of investigations depending on the presenting symptoms.

A <u>guide for general practitioners</u> has been developed to support diagnosis and management in primary health care settings.²⁰

Investigations for suspected NHL include:3, 20

- full medical history, with particular examination of risk factors
- physical examination
- FBE, Serum LDH, ESR
- chest x-ray
- CT or PET-CT (optional)
- lymph node biopsy (as per indications)
- endoscopic ultrasound (gastric MALT lymphoma)
- head CT or brain MRI and lumbar puncture to analyse cerebrospinal fluid (MCL and DLBCL).

Less than 1% of individuals presenting with peripheral lymphadenopathy will actually have a malignancy. Enlarged intra-abdominal or retroperitoneal nodes however are usually malignant.¹⁵ Initial investigations of a full blood count and chest X-ray should be performed prior to biopsy. Other malignancies should be considered such as a head and neck cancer or breast cancer in the case of axillary lumps in women.¹⁵

Current guidelines outline the following indications to guide the use of lymph node biopsy:15

- age 40 years or older
- supraclavicular location
- node diameter greater than 2.25 cm
- firm hard texture
- lack of tenderness
- present more than a few weeks.

Fine needle aspiration (FNA) is the biopsy investigation of choice in the initial triage for individuals presenting with peripheral lymphadenopathy where other diagnoses have been ruled out. This should be followed by flow cytometry studies. ¹⁵ if the FNA outcome is reported as lymphoma then excisional lymph node biopsy is required for a definitive diagnosis, sub-typing and clinical management. ¹⁵ Current NCCN guidelines recommend incisional or excisional lymph node biopsy to establish a diagnosis of NHL. ³

While some lymphomas can be diagnosed with morphology outcomes alone, most require exploration of further variables to ensure an accurate diagnosis. It is essential that although not all tests are required in every case, advanced laboratory services, specifically immunophenotyping and molecular techniques, should be available and accessible to support the diagnosis of lymphoma.⁷

Learning activities		
Completed	Activities .	
	Access the Clinical practice guidelines for the diagnosis and management of lymphoma, 15 and discuss the investigations and actions that are recommended for a person presenting with:	
	peripheral lymphadenopathy	
	intracavity presentation	
	 constitutional (B) symptoms (weight loss and fever). 	
	2 Identify the potential concerns of a person being assessed for symptoms indicative of lymphoma.	

Follow up diagnostic investigations

Additional investigations may be required if presenting symptoms are suggestive of specific organ involvement (e.g. CT scan, chest X-ray). Best practice indicates referral to a clinical haematologist or medical oncologist. A pathologist must be involved in the complex diagnostic process associated with lymphoma.¹⁵

Bone marrow aspiration and trephine (BMAT) provides reporting of the extent and the pattern of marrow involvement, along with the cell type. BMAT is indicated for staging at initial diagnosis and in rare circumstances for the primary diagnosis and sub-typing of lymphoma in individuals with no other accessible disease.^{3, 15, 18}

BMAT is not recommended for the primary diagnosis of lymphoma because of frequent histological discordance between marrow and other sites. It is recommended that the procedure be carried out by haematologists (trained or in training), or other medical practitioners specifically trained in this technique.¹⁵

Learning act	Learning activity		
Completed		Activity	
		Outline the information and resources you would provide for a person to prepare them for the following tests investigating a haematological malignancy and the associated nursing considerations:	
		Fine Needle Aspiration	
		 excisional lymph node biopsy 	
		Bone Marrow Aspiration and Trephine	
		PET scan.	

Section 3: Have the best treatment and support during active treatment

Objectives

On completion of this section, you should be able to:

- 1. Define the features of the World Health Organization (WHO) lymphoma classification system.
- 2. Discuss the implications of NHL staging and histopathology for a person's cancer journey.
- 3. Discuss current treatment approaches for the person affected by NHL.
- 4. Discuss key supportive care needs of people diagnosed with and undergoing treatment for NHL.
- 5. Implement evidence based interventions to respond to the supportive care needs of people affected by NHL.
- 6. Discuss the specific challenges associated with cancer control efforts for the older person.
- 7. Explain the SCN's role in ensuring the older person with cancer receives optimal care.
- 8. Using evidence based approaches, support the person affected by NHL to participate in decisions about their treatment and care, according to preferences.

Types of lymphoma

Lymphoma includes more than 20 lymphoproliferative malignant diseases that originate from T and B cells in the lymphatic system, with the majority (70-80%) arising from lymph nodes, and the remainder extranodal. Lymphoma can present anywhere normal lymphocytes are found.²¹

Hodgkin's Lymphoma is a form of lymphoma distinguished histopathologically by the presence of Hodgkin or Reed Sternberg cells. It is uncommon; accounting for approximately 0.5% of all newly diagnosed cancers, and differs to NHL in that it predominantly affects young adults. With recommended treatments it is generally curable. HL accounts for approximately 10% of all lymphomas.²¹

There are over 30 specific subtypes of NHL. B-cell lymphomas represent more than 85% of cases, and less than 15% of cases are T-cell lymphomas.²¹

Common subtypes of NHL include:3

Mature B-cell lymphomas:

- Chronic lymphocytic leukaemia (CLL) / Small lymphocytic lymphoma (SLL)
- Hairy cell leukemia (HCL)
- Follicular lymphoma (FL)
- Diffuse large B-cell lymphoma (DLBCL)
- Burkitt lymphoma (BL)
- AIDS-related B-cell lymphoma
- Primary Cutaneous B-cell lymphomas
- Marginal Zone lymphomas (MZL):
 - o Extranodal MZL of mucosa associated lymphoid tissue (MALT lymphoma):
 - Gastric MALT lymphoma
 - Non-gastric MALT lymphoma
 - Nodal MZL
 - Splenic MZL
- Mantle cell lymphoma (MCL).

Mature T-cell and NK-cell lymphomas:

- Peripheral T-cell lymphoma (PTCL)
- Mycosis fungoides (MF) and Sezary syndrome (SS)
- Adult T-cell leukaemi a / lymphoma (ATLL)
- Extranodal NK/T-cell lymphomas, nasal type (ENKL)
- T-cell prolymphocyticleukaemia (T-PLL)
- Primary Cutaneous CD30+ T-Cell lymphoproliferative disorders
- T-cell Large Granular Lymphocytic Leukaemia

Overview of the WHO classification system for lymphoma

The WHO classification is now recognised as the recommended international classification scheme and is used in most national guidance documents, including the Australian clinical practice guidelines for the diagnosis and management of lymphoma. The WHO classification system identifies the following four variables which are used to determine an NHL subtype:⁷

- cell morphology
- immunophenotype
- genetic features
- clinical features.

In 2008, the WHO classifications were updated to add new diseases and subtypes that have been able to be recognised due to advances in diagnostic procedures. Genetic features, detected by cytogenetics or fluorescence in-situ hybridization (FISH) have been identified as important in defining specific NHL subtypes.³

In the treatment of lymphoma, however, the various WHO categories fall into the distinct clinical groups of low grade, aggressive (intermediate) and high grade lymphomas. The Ann Arbor staging system is typically used to characterise HL which more typically spreads in an orderly way to more adjacent node groups but is also applied to other lymphomas.¹⁵

This system is based on the:19

- distribution and number of involved sites
- presence or absence of extranodal sites
- presence or absence of constitutional symptoms (B symptoms).

Accurate diagnosis and classification is fundamental to the effective management of lymphoma. It is vital therefore that diagnostic principles and classification of the disease are consistent and do not compete.⁹

Learning activities			
Completed		Activities	
		1 Access a recent physiology resource and:	
		 explain the structures and functions of the lymphatic system 	
		review the maturation sequence of the lymphocyte	
		distinguish between B lymphocytes and T lymphocytes.	
		2 Access the NCCN Guidelines – Non-Hodgkin's Lymphomas ³ or the National Cancer Institute site, and outline the WHO classification system for lymphoma.	
		3 Discuss how the MDT uses the information provided by the WHO classification system.	

Prognostic indicators

The person's prognosis and treatment plan are determined by their specific disease and health status: 19

- pathologic sub-classification
- stage and disease bulk
- age
- ECOG performance status
- molecular and biological markers.

The International Prognostic Factor Index (IPI) has been shown to predict the outcome of individuals, predominantly with B-cell lymphomas.^{15, 22} Factors included in the IPI include age, stage, number of extranodal sites, LDH and performance status. An age-adjusted model for individuals under the age of 60 and a model for follicular lymphoma have also been developed.¹⁹

Bone marrow involvement has been associated with significantly shorter survival in individuals with intermediate or high-grade lymphoma.³

Learning activity		
Completed		Activity
		Access the <u>NCCN Guidelines – Non-Hodgkin's Lymphomas</u> ³ and for follicular lymphoma and diffuse large B cell lymphoma:
		outline the independent predictors of survival outcomes
		identify the impact of these risk factors on five year survival
		 identify specific prognostic markers for diffuse large B cell lymphoma.

Supportive care needs at diagnosis

People with lymphoma experience a range of treatment and support needs that require a coordinated approach to the planning and delivery of care. These needs vary in severity and complexity depending on disease type, stage and treatment required:^{23, 24}

- physical needs
- symptoms related to the location and pathology of the lymphoma
- psychological needs
- anxiety associated with a malignant diagnosis
- uncertainty relating to the future
- loss of control of day-to-day life
- social/practical needs
- impact on financial circumstances
- changes in relationships
- changes in social roles (husband, father, friend)
- information needs
- specific lymphoma resources
- treatment options and impact of treatment on self and others
- prognosis
- access to support groups
- access and information regarding complementary therapies.

Learning activities		
Completed	Activi	ities
	1	Describe key principles for ensuring the needs of a person newly diagnosed with NHL are identified.
	2	Identify a list of potential care providers and services that may be involved in meeting the supportive care needs of a person newly diagnosed with NHL.

Health history	
Patients name:	Arthur
Sex:	M ☑ F □
Age:	78
Diagnosis:	Stage III Diffuse Large B-cell lymphoma
Social History:	Retired train driver.
	Financially stable drawing on pension. Lives with wife Phoebe, who has moderate phase dementia. Arthur is Phoebe's primary carer. They live in the family home of 45 years. It is a three bedroom, two story dwelling on a large double block.
	Their daughter Joan lives a 15-minute drive away with her husband and two young children.
	Arthur has limited contact with old work colleagues due to Phoebe's increasing care requirements, but he enjoys playing cards with his neighbour once or twice a week.

Learning activity	
Completed	Activity 1 Discuss possible supportive care needs Arthur may have that relate to his role as a carer, and suggest SCN responses.

The older person with cancer

It was estimated that in 2015, 65.5 per cent of new cancer cases in Australian men will be diagnosed in men aged 65 years and over, and 55 per cent of new cases in Australian women will be diagnosed in women aged 65 years and over. Psychosocial changes associated with ageing and multiple comorbidities in older people can influence the type of treatment and support required. The unique treatment and care needs of older people with cancer have been described as a new specialty termed 'geriatric oncology'. So

Studies suggest that changes associated with ageing can sometimes lead to older people receiving suboptimal treatment, or treatment that is not consistent with their personal preferences.^{25, 26} Some studies report that older people, compared to younger people, with aggressive lymphoma are less likely to be treated for cure and are less likely to survive for five years due to limited access to clinical trials.²⁷

Beneficent ageism refers to a neglect of an older person's wishes. The older person's social role is distorted and health professionals assume an ability to represent their best interests without endorsement from them.²⁸

Additional complications can arise in the older person due to co-morbidity factors. Comprehensive geriatric assessments may help to support decision making in relation to treatment plans for older people. Aging is highly individualised and often poorly reflected in chronologic age. The management of the older person with cancer should thus be based on assessment of physiologic rather than chronologic age. The following factors should be taken into account when assessing the older person with cancer: 30-32

- functional status
- co-morbidity
- cognition
- social support
- psychological state
- concomitant medications
- nutrition
- continence
- carer support.

Age-related changes are also associated with disease-specific issues. In NHL, the implication for the older person is decreased duration of complete response possibly secondary to increased circulating levels of interleukin-6.³²

Resource link

Cancer Forum. November 2013. Vol 37 Issue No 3. Geriatric oncology

Learning act	Learning activities		
Completed		Activities	
		Provide an example of how each of the age-related factors may impact an older person affected by NHL.	
		Access the article <u>Tools for assessing elderly cancer patients</u> , 30 and <u>NCCN clinical practice guidelines for oncology. Senior adult oncology.</u> <u>Version 2.2014</u> (a free resource, but you must register and then click 'Remember me' to bypass the login page in future), and:	
		 discuss benefits or limitations associated with using a geriatric assessment tool in an oncology setting 	
		 appraise current tools used for geriatric assessment in light of the essential domains of a comprehensive geriatric assessment 	
		 describe actions that could be taken to ensure the older person's treatment preferences are considered 	
		 explain how you would respond to concerns of a colleague that a person's treatment plan should be modified due to their age. 	

Treatment approaches for NHL

Treatment approaches for malignant lymphomas are complex. In this module, we examine treatment approaches for NHL only.

A treatment plan needs to be developed by a MDT in conjunction with the individual affected, who is informed of their options and the aims of treatment. The aims may be defined in terms of potentially curative treatment or palliative management plan. The effects of the disease and its management, the need for long-term follow up, and the potential for late complications of treatment need to be discussed.¹⁵

Treatment plans for NHL can include:7

- watch and wait
- systemic combined antineoplastic agent regimens with or without immunotherapy
- radiotherapy
- Radioimmunotherapy
- Haematopoietic stem cell transplantation (HSCT).

Treatment approaches vary depending on the type and stage of NHL, the person's overall clinical presentation and their response to treatment. The principles associated with treating the different grades of NHL are outlined below.

Low grade lymphoma:6, 15

- the highest priority of treatment is to maximise the individual's overall survival, maintain quality of life and avoid treatment-related morbidity
- treatment recommendations are discussed using 'surrogate endpoints' including overall response rates, complete remission rates, and 'molecular' complete remission rates
- treatment options may include radiotherapy, antineoplastic agents, monoclonal antibody therapy, watch and wait approach and HSCT.

Aggressive (Intermediate)grade lymphoma:15

- the treatment intent, where feasible, is cure, as these lymphomas are very chemosensitive
- the main treatment modality is antineoplastic agents and monoclonal antibody therapy
- in some individuals radiation therapy provides additional benefit, particularly where there is bulky local disease
- surgery has little role in this disease, other than in diagnosis.

High grade lymphoma:15

- due to the complexities associated with the disease and its management, individuals with newly diagnosed high grade lymphoma should ideally be managed in specialist units experienced in treating these disorders
- treatment approaches involve intensive antineoplastic agent regimens and include central nervous system (CNS) prophylaxis
- radiotherapy has little role in this disease.

Learning activities		
Completed	Activities	
	Access the NCCN Guidelines – Non-Hodgkin's Lymphomas ³ (a free resource, but you must register and then click 'Remember me' to bypass the login page in future), and outline the current treatment options recommended for DLBCL.	
	2 Summarise the evidence for and against dose reduction of antineoplastic agents in the older person diagnosed with DLBCL.	

Adjuvant treatment approaches for NHL

Immunotherapy

The application of immunotherapy, a therapy which boosts the immune system, in the management of NHL has focused on the use of Interferon. Typically used as a maintenance therapy, Interferon is used to treat low grade lymphomas in those who can tolerate it. The use of Interferon in Australia may be considered on an individual basis.¹⁵

Further use of immunotherapy in the treatment of NHL is under investigation with the implementation of clinical trials in which a vaccine derived from healthy B cells is administered.⁷

Radioimmunotherapy

The addition of radioisotopes to anti-CD20 antibodies is also known as systemic targeted radiation or STAR. This therapy inflicts collateral damage to adjacent tumour cells as a result of the cytotoxicity of the beta particles emitted by the radionucleotide which is attached to the antibody.³³ Normal tissues are also potentially damaged by the emissions of the radioisotope; thus, care is taken when determining appropriate radioisotopes to use in radioimmunotherapy.³³

Two radioimmunotherapy agents used in the treatment of NHL are iodine 131 or tositumomab (Bexxar) and ibritumomabtiuxetan (Zevalin). The agents are administered as an outpatient usually one week apart. The most common and major toxicity associated with both agents is myelosuppression.⁷

Proteasome inhibitors

A new class of drugs for the treatment of NHL is proteasome inhibitors. Proteasomes are responsible for maintaining protein concentrations in cells and are able to interrupt an abnormal accumulation of disease related proteins. Bortezomib (Velcade) has been reported as the most commonly used proteasome inhibitor.⁷

Haematopoietic stem cell transplantation

Options for transplantation in individuals with NHL vary according to sub-type and are only usually considered once standard therapy has failed. In Australia an allogeneic stem cell transplant is recommended as a treatment option for immunodeficiency associated lymphomas. However, its application in other sub-types is recommended as a last resort for those who have failed all other treatments and do not present with a high risk of transplant related complications. 15

Learning activities		
Completed		Activities
		Outline the information and supportive care which would be provided to an individual receiving treatment with radioimmunotherapy as an outpatient.
		2 Access a current text and / or research and summarise the role of proteasome inhibitors in the management of NHL.
		Access a current text and / or research and summarise the role of haematopoietic stem cell transplantation in the management of NHL.

Case study

Arthur's story 2: Arthur's treatment



Learning activities		
Completed	Activities	
	Access the following resources to respond to the activities below:	
	<u>NCCN Guidelines – Non-Hodgkin's Lymphomas</u> ³	
	NCCN Clinical practice guidelines for oncology - senior adult oncology ³²	
П	Note: both NCCN resources are free to access, but you must register and then click 'Remember me' to bypass the login page in future.	
_	1 Develop a care plan to address Arthur's information needs at this time.	
	2 Identify factors you would encourage Arthur to consider when making his decision about treatment.	
	Plan in detail how you would support Arthur in the process of making treatment decisions.	
	4 Identify issues the MDT may discuss to ensure Arthur receives the optimal approach to treatment.	

Early treatment and disease effects

There are a number of early treatment and disease effects which may arise in the person affected by NHL which require prompt assessment and interventions.

Acute tumour lysis syndrome (TLS) is characterised by metabolic changes resulting from the body's inability to process the large volume of intracellular components released with rapid tumour cell destruction.³ The metabolic changes can result in cardiac arrhythmias, seizures, loss of muscle control, acute renal failure and even death.³

The risk is highest in individuals where the following are present:^{34, 35}

- tumours with high growth fractions
- dehydration/ poor urinary output prior to cytotoxic treatment
- bulky abdominal disease (greater than 8-10cm)
- extensive lymph node involvement on diagnosis and prior to treatment
- high white blood cell count
- elevated potassium, phosphorus and uric acid prior to treatment
- elevated lactate dehydrogenase prior to treatment.

The development of TLS is frequently associated with commencement of treatment and is usually observed within 12-72 hours.³ It may also be precipitated by corticosteroids, ionizing radiation, hormonal therapy, and biological response modifiers. Management of TLS includes prevention and prompt correction of fluid and electrolyte imbalances.^{34, 35}

Large mediastinal masses may cause severe airways obstruction at presentation. 15

Cardiac tamponade requires prompt initiation of specific therapy together with pericardial paracentesis.¹⁵

Superior vena cava obstruction, although not uncommon in individuals with NHL, is not usually life threatening. ¹⁵

Massive abdominal involvement which is commonly associated with ascites, is usually due to Burkitt's lymphoma, and may be responsible for:¹⁵

- perforation and/or obstruction of bowel (including intussusception)
- gastrointestinal haemorrhage
- obstruction of ureters, inferior vena cava and lymphatics.

Neurological emergencies associated with NHL may include paraplegia, cranial nerve palsies, meningeal disease and intracerebral tumour. Primary central nervous system lymphoma (PCNSL) is a rare, extranodal form of NHL. Individuals may present with neurological changes. Treatment includes high dose methotrexate requiring specific nursing interventions and evaluation to ensure safety and decrease drug toxicity.³⁶

Resource link

Access <u>Oncological Emergencies and Paraneoplastic Syndromes</u> (you will need to set up a free account) for further information about:

Tumour lysis syndrome (page 6)

Spinal cord compression (page 3)

Superior vena cava obstruction (page 1).

•

Learning activities			
Completed		Activities	
		For each of the following conditions, explain the pathophysiology of the condition in relation to NHL and the signs and symptoms which may be associated with their occurrence:	
		• TLS	
		cardiac tamponade	
		bowel perforation	
		• paraplegia.	
		Outline nursing and medical assessment, monitoring and interventions associated with administration of high dose methotrexate to prevent and manage toxicities.	

Health history

Patients name: Arthur

Sex: M ☑ F □

uric acid

lactate dehydrogenase (LDH)

Age: 78

Diagnosis: Stage III Diffuse Large B-cell lymphoma

Enlarged mediastinal mass present on chest x-ray

PET scan indicates positive supraclavicular and mediastinal lymph nodes

Bone marrow aspirate and trephine reveals 74% blasts.

Treatment protocol:

CHOP-R	Frequency: every 3 weeks
	No. of cycles: 6 to 8 (restaging will determine the number of cycles)
Vincristine	1.4 mg/m2 IVI up to a MAXIMUM of 2mg Day 1
Doxorubicin	50 mg/m2 IVI Day 1
Cyclophosphamide	750 mg/m2 IVI Day 1
Prednisolone	100 mg orally Days 1 to 5, total dose 500mg over 5 days
Rituximab	375 mg/m2 by IV infusion Day 1
Significant blood pathology 8 hours pos	st commencement of therapy includes:
Hb	95g/L
WCC	1.5 x 109/L
neutrophils	15%
platelets	82 × 109/L
sodium	138 mEq/L
Potassium	6.3 mEq/L
bicarbonate	26 mmol/l
phosphorus	10.5mg/dL
serum calcium	1.92 mmol/L
creatinine	2.4mg/dL

Arthur reports ongoing fatigue, reduced appetite, nausea, and shortness of breath on exertion.

25.2mg/dL

5090 U/L

Learning activities		
Completed	Activities	
	1 Identify common side effects of CHOP-R regimen, and for each drug:	
	 outline a plan for prevention and management of each of these effects 	
	 discuss nursing considerations associated with the drug administration. 	
	2 Discuss the pre-treatment information and support that Arthur should receive.	
	 Provide examples of a range of evidence based strategies for delivering, tailoring and reinforcing information and providing support. 	
	Outline key information to be conveyed.	
	 Identify resources which may facilitate information and supportive care provision. 	
	Outline the prognostic and management implications of the pathology results in Arthur's health history.	
	4 Discuss significant results which may indicate TLS in Arthur's case.	
	 Summarise the likely medical management associated with increased risk of TLS in Arthur's case. 	
	 Outline the nursing assessment required and implications associated with caring for Arthur when at risk of and affected by TLS. 	
	Outline the nursing interventions to manage current symptoms Arthur is experiencing:	
	• fatigue	
	reduced appetite	
	• nausea	
	shortness of breath on exertion.	

Supportive care needs during treatment for NHL

A range of supportive care needs result from the effects of the disease and the treatment approaches used in the management of NHL.

Myelosuppression is a common treatment-related effect, and the risk is significantly increased in individuals by age 65.³² It is associated with administration of:⁷

- alkylating agents
- anthracyclines
- purine analogs
- radioimmunotherapy agents.

Early interventions are crucial to prevent viral reactivation and infections and include:^{3,7,37}

- administration of myeloid growth factors
- administration of antiviral prophylaxis or pre-emptive therapy
- transfusion support
- maintaining neutropaenic precautions
- effective hand-washing techniques
- education.

Studies report that the risk of myelosuppression in individuals treated for lymphoma is decreased by 50% when using growth factors. The effective management of myelosuppression is crucial, as evidenced by reported mortality rates of 5% to 30% in individuals aged 70 and over with neutropaenic infections.³²

Prophylactic use of granulocyte colony-stimulating factor (G-CSF) has been shown to reduce the incidence, length and severity of chemotherapy-related neutropaenia, reduce the risk of febrile neutropaenia, and improve relative dose intensity of the chemotherapy delivered.³⁷

The indication for prophylactic G-CSF use depends on the risk of febrile neutropaenia or other neutropaenic events which may compromise treatment.³⁷ The main toxicity associated with G-CSF therapy is mild to moderate bone pain which is usually controlled by non-narcotic analgesics.³⁷

Anaemia, associated with lymphoid malignancies and their treatment, is a debilitating adverse effect, contributing to fatigue, reduced quality of life and even poor treatment outcomes and reduced survival. Red blood cell transfusion may be indicated for individuals with Hb in the range of 70-100g/L and is likely when Hb is less than 70g/L.³⁸ Platelets may be also indicated for individuals with thrombocytopaenia. Clinical practice guidelines outline the appropriate prophylactic and therapeutic use of platelets. ³⁹

Resource links

The following resources are recommended for further information on the use of blood components.

- <u>Guidelines for the administration of blood components.</u> ³⁹ Australian and New Zealand Society of Blood Transfusion. 2011
- Australian and New Zealand Society of blood transfusion website
- <u>BloodSafe e-learning Australia website</u>

Common gastrointestinal effects include nausea, vomiting and diarrhoea. While these may be managed with pharmacological approaches, the SCN has a key role in assessing individuals to monitor for potential dehydration, electrolyte abnormalities and nutritional deficiencies.⁷

Alopecia may be caused by administration of alkylating agents and anthracyclines or radiation therapy.⁷ It has been reported that both men and women find hair loss related to cancer treatment a source of distress, depression and loss of confidence.⁴⁰ The SCN has a key role in meeting the supportive care needs of people affected by alopecia and making referrals to support services in this area.

Mucositis may be associated with antineoplastic agents and radiotherapy. The severity and incidence of mucositis depends on the treatment regimen.⁷

Learning activities	3
Completed	Activities
	Define 'dose intensity' and discuss why it is significant in the management of NHL.
	2 Discuss evidence based interventions to promote early detection of neutropaenic sepsis in a person receiving treatment for NHL.
	3 Discuss the evidence based nursing responses to a person demonstrating signs of neutropaenic sepsis.
	4 Outline the current evidence regarding the following aspects of neutropaenic precautions and compare this evidence to current practice within your health care facility:
	isolation precaution
	dietary restrictions.
	5 Explain the mechanisms of action of colony stimulating factors and their role in supportive care for the person with NHL.
	7 Discuss nursing considerations associated with administering colony stimulating factors for the person with NHL.
	8 Outline the indications for transfusion support in the person receiving treatment for NHL.
	9 Explain nursing considerations in administering blood and blood products for the person with NHL.

Section 4: Have the best treatment and support between and after active treatment

Objectives

On completion of this section, you should be able to:

- 1. Implement evidence based interventions to respond to the ongoing supportive care needs of a person following treatment for lymphoma.
- 2. Critique the role of the care coordinator in care of people following treatment for lymphoma.
- 3. Tailor supportive care interventions for a person following treatment for lymphoma to their personal and social circumstances.
- 4. Identify the specific support strategies to assist the carer of the person with lymphoma.

Follow up treatment and care

There are no clear guidelines regarding follow up care for the person affected by lymphoma. It is recommended that an individualised management plan should be organised for surveillance with the individual, their family, the GP and the specialist.²⁰ Follow up should aim to detect recurrent or relapsed disease, long term side effects of therapy and secondary cancers.

More than 50% of people affected by aggressive lymphoma have either primary refractory disease or, more often, relapse after a complete response to their initial treatment.¹⁵ For individuals with relapsed or progressive disease that is non-responsive to therapy, symptom control and palliative measures would be appropriate.²⁰ It is recommended that active treatments such as single antineoplastic agent, corticosteroids, monoclonal antibodies and radiotherapy are considered, preferably within a clinical trial.²⁰

The care coordinator may help to facilitate the extended members of the MDT to implement an efficient, timely and comprehensive assessment of the person's needs.

Arthur's story 3: Promoting continuity of care

Learning activities		
Completed	Activities	
	1 Discuss the role of the care coordinator in Arthur's care.	
	2 Discuss how the care 'record' held by Arthur may support his cancer journey. Refer to current evidence in your response.	

Case study

Arthur's story 4: Arthur's supportive care needs



Learning activities		
Completed		Activities
		Discuss how Arthur and his family may benefit from the input of the care coordinator.
		Discuss possible implications of Arthur's statement regarding him not needing the aged care facility ('there's a place there for me too if I need it, not on your nelly, I'm alright').
		3 Describe how this statement might be used as a cue for further discussion.

Carer needs

Research relating to the demands of care giving by family members identifies a range of concerns which commonly become a source of stress for the carer. These include:¹¹

- uncertainty about treatment
- lack of knowledge about care
- role changes within the family
- lack of transportation for treatment
- strained financial resources
- physical restrictions
- lack of social support
- fears of being alone.

Disruptions and emotional strains associated with care giving are common experiences for families of people with cancer. Feelings of tiredness, difficulty getting enough sleep, and feelings of resentment and isolation are among the most commonly reported disruptions and emotional strains and among the most difficult coping challenges.¹¹

Carers of the older person with cancer are often older themselves. They may be a spouse or an adult child with many competing demands.⁴¹

Adequate support is not a reality for many carers, who face physical, emotional and financial costs to their wellbeing.⁴²

A carer must be recognised as both a key partner in the care team and a recipient of care in accordance with the palliative care service provision model.⁴²

A carer both provides and needs support, yet the needs of carers are often overlooked. Carers frequently subordinate their own emotional and health needs beneath those they are caring for and these needs are frequently overlooked by health and care workers who are not trained to recognise them.¹¹

The largest survey of Australian carers' health and wellbeing conducted to date found that carers have the lowest level of wellbeing of any population group.⁴³

Learning activity		
Completed	Activity	
	Access <u>An exploratory study of GP awareness of carer emotional needs</u> <u>in Western Australia</u> ⁴⁴ and which explores the needs of carers.	
	 Identify and discuss the key challenges experienced by carers of people diagnosed with lymphoma. 	
	 Discuss specific challenges that may be experienced by people of different age groups. 	

Case study

Nine months following the completion of treatment, Arthur relapses. As his condition deteriorates he is transferred to the aged care facility where Phoebe lives.

Arthur's story 5: Arthur relapses



Learning activities		
Completed	Activities	
	Discuss the practical strategies that the team can implement to support Joan with her concerns.	
	Outline strategies to ensure continuity of care between Arthur's care providers.	
	3 Access the <u>NCCN Guidelines – Non-Hodgkin's Lymphomas</u> ³ (a free resource, but you must register then click 'Remember me' to bypass the login page in future), and outline the treatment options which may be offered to Arthur with relapsed DLBCL.	

Section 5: Have the best care at the end of life

Objectives

On completion of this section, you should be able to:

- 1. Analyse the role of the SCN in facilitating the transition to palliative care for the older person with lymphoma.
- 2. Describe the importance of advance care directives and identify how they should be applied in practice.
- 3. Identify and analyse the principles of a multidisciplinary approach to symptom management.
- 4. Analyse different approaches to bereavement risk assessment and support.

A palliative approach

Palliative care occurs in a range of care settings, including inpatient settings, and in the person's home. For people with cancer whose care needs are likely to be prolonged, and where these needs are unable to be met with available personal, community and health care resources, admission to a residential aged care facility may be considered. Such decisions are likely to be associated with significant fears and concerns.

Three forms of palliative care have been described to support the delivery of palliative care in residential aged care:⁹

- a palliative approach
- specialised palliative care provision
- end of life care.

Learning activities		
Completed		Activities
		Discuss the challenges associated with providing palliative care in a residential aged care facility.
		2 Discuss ways in which the SCN can collaborate with staff in residential aged care facilities to ensure optimal palliative care.

Palliative care in residential care facilities

There is a need to ensure resources are available to facilitate effective palliative care in residential care and in particular in relation to the preparation of a good death.⁴⁵ Three principles have been identified to support optimal care at end of life. These are:⁹

- individualised care by consistent care providers
- effective care teams that include the family
- comprehensive advance care planning.

Advance care planning is an area that is integral to the establishment and management of palliative care within residential aged care facilities and includes practices built on communication with residents and their family about their wishes, beliefs and values about future care.⁹

Advance care statements or living wills are used to express a person's view and preferences about their end of life care. Advanced care directives are a subset of advanced care statements which record advance refusals of particular types of life prolonging treatment.⁴⁶

There may be additional barriers to managing symptoms in older people with cancer, due to concerns about the effect of treatments, and co-morbidities, and communication difficulties. The implementation of carefully determined procedures for pain identification and assessment and an evidence based multidisciplinary approach to pain management are essential for successful pain management in aged care facilities.¹⁰

Family meetings can be an important mechanism for facilitating communication and planning care. Family meetings between the person, their family and health care professionals can be used to:⁴⁷

- share information and concerns
- clarify goals of care
- discuss diagnosis, treatment, and prognosis
- develop a plan of care for the person and their family carers.

Learning activities		
Completed	Activities	
	Access Pain in Residential Aged Care facilities' Management Strategies, ¹⁰ and identify strategies that may be useful in the management pain for the person with NHL in a residential aged care facility.	
	 Access Family meetings in palliative care: Multidisciplinary clinical practice guidelines, 47 and identify: situations when you would consider organising a family meeting 	
	 how you would determine who would be involved in the family meeting 	
	 principles for conducting an effective family meeting 	
	 skills you would need to develop to conduct effective family meetings. 	

Case study

Arthur's story 6: multidisciplinary team meeting



Learning activities				
Completed		Activities		
		1 Discuss the purpose of this MDT meeting in Arthur's case.		
		2 Discuss how you would go about preparing for such a meeting.		
		Critique the meeting process, identifying factors which reflect an effective MDT and those areas which could be improved.		
		4 Discuss how this MDT meeting may be similar or different to an MDT held in an acute treatment facility.		
		Describe how information discussed in the MDT may be communicated to other health care providers caring for Arthur and provide examples of how this information may improve Arthur's care.		

Bereavement support

How people die will be in the lasting memories of carers and families, and it is important that health professionals provide the best care possible at the end of life.⁴⁸

Risk assessment of bereavement needs is recommended as best practice and should be performed by a member of the MDT prior to the person's death. Individual's identified as high risk should be referred to a resourced and accessible specialist bereavement service.²⁴

Evidence suggests that Australian palliative care services provide a high level of bereavement support including risk assessment. However, due to a lack of designated bereavement workers those responsible for providing bereavement support are often those who are responsible for the other aspects of the person's care. ⁴⁹

Resource links

Bereavement and Grief. 2013, CareSearch

Learning activity				
Completed	Activity			
	 Access Survey of bereavement support provided by Australian Palliative Care Services⁴⁹ and Guidelines for the assessment of complicated bereavement risk in family members of people receiving palliative care, ⁵⁰ and: analyse and discuss the benefits and risks of using a MDT opinion to determine bereavement risks as opposed to a formal risk assessment tool discuss how bereavement support might be enhanced in your own clinical area. 			

Case study

<u>Arthur's story 7: Supporting Arthur's daughter</u>



Learning activities				
Completed	Activities			
	Discuss the bereavement care you would recommend for Joan at this time, providing a rationale for your approach.			
	2 Discuss how you would determine the need for ongoing bereavement support for Joan.			

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