

Nodular Lymphocyte Predominant Hodgkin Lymphoma (NLPHL)



OVERVIEW

Lymphoma is the 6th most common cancer in Australia in adult men and women. It can affect people of all ages and is the most common blood cancer. Lymphoma is a cancer of the immune system and affects lymphocytes - a type of white blood cell. When lymphocytes gain genetic changes they divide and grow uncontrollably resulting in lymphoma.

There are two main types of lymphocytes - B-cells and T-cells. Lymphomas growing from B-cells are more common and account for around 85% of lymphoma cases; those caused by T-cells around 15%. The first lymphoma to be discovered was called "Hodgkin lymphoma" (HL - around 15% of all B-cell lymphomas), after Thomas Hodgkin, who described it. All subsequent lymphomas discovered were called "non-Hodgkin lymphoma" (NHL around 90% of all lymphomas, both B and T-cell lymphomas).

There are over 80 different subtypes of lymphoma, which can be classified according to how fast they grow. "Aggressive" (fast growing) lymphomas are those that grow quickly, usually weeks to months and need treatment immediately. "Indolent" (slow growing) lymphomas usually develop over months to years and often are not treated straight away but are monitored. It is important to know your subtype of lymphoma. Lymphoma cells can travel to any part of the body and be found in lymph nodes, the bone marrow, the spleen, blood, bone, skin and almost any organ or tissue.

Nodular lymphocyte predominant Hodgkin lymphoma (NLPHL) is a rare subtype of Hodgkin lymphoma which accounts for around 5% of HL cases. This subtype is more common in men than women, most often affects people between 30-50 years of age and is slower growing in nature to the more common subtypes of HL. NLPHL is different from other subtypes of HL in that there is an absence of the Reed-Sternberg cells and presence of abnormally large B-cells which are sometimes referred to as 'popcorn cells' which are distributed in a nodular pattern in the tissues. This disease more consistently expresses CD20 on the surface of lymphoma cells than other forms of Hodgkin lymphoma.

NLPHL resembles other slow growing B-cell non-Hodgkin lymphoma subtypes that are chronic in nature and multiple

relapses are expected and these are expected to be highly treatable. Usually the only sign of NLPHL is a painless lump in the neck, armpit or groin area that does not go away over time. Other symptoms that are not as common include sweats, fever, unexplained weight loss and fatigue otherwise known as B symptoms of lymphoma. NLPHL is treated differently than the more common forms of HL which is why we have created a separate fact sheet to discuss in further detail.

DIAGNOSIS AND STAGING

A biopsy is always required for a diagnosis of NLPHL. A biopsy is a surgical procedure to remove part of or all of an affected lymph node or other abnormal tissue to look at it under the microscope. The biopsy can be done under local or general anaesthetic depending on what part of the body is being biopsied.

Once a diagnosis of NLPHL is made there are further tests that need to be performed to see where else in the body the lymphoma may be and is referred to as staging. Because NLPHL is a blood cancer the lymphoma can travel all over the body, so it is important that a check of the entire body is done looking for the lymphoma. Staging tests may include:

- Positron emission tomography (PET) /CT scan
- Computed tomography (CT) scan
- Bone marrow biopsy

Patients will also undergo a number of baseline tests prior to any treatment commencing to check their organ function and these baseline tests may include a heart scan, kidney scan, breathing test and blood tests.

NLPHL is a highly treatable lymphoma with more than 9 out of 10 people being cured or achieving a long-term remission with no signs of the lymphoma often 10 years or longer at a time.

TREATMENT OPTIONS

NLPHL is treated according to what stage the lymphoma is at diagnosis, what symptoms you have, your age and general health. As this is a highly treatable lymphoma your treating team will be considering possible long-term side effects of treatment and given the age group who mainly present with this lymphoma the effect to people's fertility is addressed at this time also.

<p>Early Stage NLPHL without B symptoms</p> <ul style="list-style-type: none"> • Radiotherapy alone • Surgery followed by radiotherapy • Surgery alone
<p>Early Stage NLPHL with B symptoms</p> <ul style="list-style-type: none"> • Chemotherapy and monoclonal antibody *CVP-R x 2-3 cycles followed by radiotherapy • Chemotherapy and monoclonal antibody CVP-R x 6 cycles
<p>Advanced Stage NLPHL</p> <ul style="list-style-type: none"> • Chemotherapy and monoclonal antibody *CHOP-R x 6 cycles • Chemotherapy and monoclonal antibody *ABVD-R x 6 cycles • Chemotherapy and monoclonal antibody CVP-R x 6 cycles
<p>Relapsed NLPHL</p> <ul style="list-style-type: none"> • Chemotherapy and monoclonal antibody treatments as described above • Repeat of previous treatment if the remission period has been lengthy and swap to a different treatment if the remission period has been shorter • Consideration of autologous stem cell transplantation in certain cases of relapsed disease

*CVP-R – cyclophosphamide, vincristine, prednisolone and rituximab

*CHOP-R – cyclophosphamide, doxorubicin, vincristine, prednisolone and rituximab

*ABVD-R – doxorubicin, bleomycin, vinblastine, dacarbazine and rituximab

CLINICAL TRIALS

Clinical trials are essential in identifying effective medicines and determining optimal doses of these medicines for people diagnosed with lymphoma. People who are interested in participating in a clinical trial can find one using the following methods:

1. Speak to their specialist to see what options are available
2. See 'Understanding Clinical Trials' fact sheet on the website to find out more. <https://bit.ly/2FEeXX7>

FOLLOW UP

Once treatment is completed, people with lymphoma need to be followed up by their specialist with regular appointments to monitor:

- Evaluate the effectiveness of the treatment
- Ongoing treatment side effects

- Recovery from treatment
- Signs of lymphoma relapsing
- Potential late effects caused by treatment that can occur months or years later, that can be based on the duration and frequency of treatment, age, gender and overall health of each person.

RESOURCES AND SUPPORT

Organisation	How can they help?
Lymphoma Australia	<ul style="list-style-type: none"> • Lymphoma Australia offers a wide variety of resources and support for people with lymphoma or CLL and their carers. Please visit our website www.lymphoma.org.au for further information
	<ul style="list-style-type: none"> • Lymphoma Australia fact sheets and booklets (lymphoma.org.au/page/1218/fact-sheets) including: <ul style="list-style-type: none"> • Booklet: What you need to know about lymphoma • Lymphoma subtypes • Understanding clinical trials • Lymphoma Australia YouTube channel: Presentations on a variety of topics about subtypes, management and supportive care: youtube.com/user/LymphomaAustralia • Lymphoma Nurse Support Line: 1800 953 081 or email: nurse@lymphoma.org.au • Online private Facebook group: "Lymphoma Down Under": http://bit.ly/33tuwro

SOME QUESTIONS TO ASK YOUR DOCTOR

- What type of HL do I have?
- Is there any additional testing that can be done to give me greater insight into how to treat my type of lymphoma?
- What are the treatment options for my stage of NLPHL?
- Are there any treatment options that are better for my type of lymphoma but are yet to be funded by the PBS in Australia?
- Are there any clinical trials currently available to me?
- If you think my NLPHL has relapsed, will you do another tissue biopsy to confirm this?