



Hodgkin Lymphoma (HL)

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 which are a type of white blood cell. When lymphocytes gain DNA mutations they divide and grow uncontrollably resulting in lymphoma.

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

There are over 80 different subtypes of lymphoma, that are classified according to its clinical behaviour. "Aggressive" (high grade or fast growing) lymphomas are those that grow quickly, usually weeks to months and need treatment immediately. "Indolent" (low grade or slow growing) lymphomas usually develop over 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.

HL affects both men and women and is most common in 15 – 29 year olds. There is a second peak later in life (over 70 years), although it can occur at any age. HL is characterised by the presence of very large cells called Reed-Sternberg (RS) cells, although other abnormal cell types may be present.

HL is potentially an aggressive (or fast growing) lymphoma. Usually the first sign of HL is a painless, rapidly growing lump in the neck, arm pit or groin. The swelling may be painful if the lymph node is pressing on a sensitive area (nerve or blood vessel). Other common symptoms may include sweats, fever, unexplained weight loss, generalised all over body itch and people may also notice fatigue, loss of appetite, shortness of breath or pain in lymph nodes after alcohol.

TYPES OF HL

There are a number of different types of HL which may influence what treatment a person receives and how well they respond to this treatment. HL is divided into two main classifications, Classical HL which accounts for 90 - 95 percent of cases and the remainder are Nodular Lymphocyte Predominant HL.

CLASSICAL HL

Nodular Sclerosis Classical HL is the most common subtype accounting for 60 – 80 percent of all HL cases and is more common in women than men. This subtype receives its name from its appearance under the microscope with **nodular** referring to 'nodule-like' pattern in the tissue and **sclerosis** referring to 'scar tissue' as there is typically a mixture of Reed-Sternberg cells, normal white blood cells and scar tissue.

Mixed Cellularity Classical HL accounts for 15 - 30 percent of all HL cases. This subtype is more common in men than women and typically affects older adults.

Lymphocyte Depleted Classical HL accounts for less than 1 percent of all HL cases and is rarely diagnosed.

Lymphocyte Rich Classical HL accounts for less than 5 percent of all HL cases. This subtype of HL is typically diagnosed at an earlier stage than other subtypes of HL and has a very low relapse rate.

NODULAR LYMPHOCYTE PREDOMINANT HL

Nodular Lymphocyte Predominant HL accounts for 5 to 10 percent of all HL cases. This subtype is more common in men than women and is typically slower growing than the other more aggressive subtypes of HL. This subtype of HL resembles other slow growing non-Hodgkin lymphoma subtypes that are chronic in nature with late relapses that are highly treatable. It is treated differently from 'classical' HL and is not discussed further here.

DIAGNOSIS AND STAGING

A biopsy is always required for a diagnosis of HL. 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.

FACT SHEET

The biopsy can be done under local or general anaesthetic depending on what part of the body is being biopsied.

Once a diagnosis of classical HL 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 as staging (stage I-IV). Staging tests may include:

- Positron emission tomography (PET) /CT scan
- Computed tomography (CT) scan
- Bone marrow biopsy (in some patients but not all)
- Baseline tests prior to treatment starting to check organ function: heart scan, kidney scan, respiratory tests & blood tests

TREATMENT OPTIONS

Since classical HL is a rapidly growing lymphoma, treatment may need to start within a few weeks after diagnosis is made.

The standard treatment for patients with classical HL is a combination of drugs (chemotherapy), with some patients also receiving radiotherapy. There are two chemotherapy regimen options, depending on various factors, that your doctor may recommend.

- ABVD (doxorubicin, bleomycin, vinblastine and dacarbazine) – given over one day on Day 1 and Day 15 of a 28-day cycle
- Escalated BEACOPP (bleomycin, etoposide, doxorubicin, cyclophosphamide, vincristine, procarbazine and prednisolone) – multiple days over a 21-day cycle

A high percentage of patients achieve remission with standard first line treatment. For some people, the lymphoma returns (relapses) or in the rare cases does not respond to initial treatment (refractory) and for these patients there are other treatments that can be successful. These can include:

- High dose combination chemotherapy followed by an autologous stem cell transplant (minority may receive an allogeneic stem cell transplant from a donor)
- Combination chemotherapy
- Immunotherapy: such as brentuximab or pembrolizumab
- Radiotherapy
- Clinical trial

TREATMENTS UNDER INVESTIGATION

Many new treatments and combination medicines are currently being tested in clinical trials around the world for both newly diagnosed and relapsed/refractory classical HL including:

- Bendamustine (Ribomustin™)

- Gemcitabine (Gemzar™)
- CAR T-cell therapy
- LAG 3 inhibitor with pembrolizumab (Keytruda™)

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. Go to the ClinTrial Refer website www.clintrial.org.au/ to search available clinical trials
3. See 'Understanding Clinical Trials' fact sheet, www.lymphoma.org.au/

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

Lymphoma Australia offers a wide variety of resources and support for people with lymphoma and their carers. Please visit our website www.lymphoma.org.au/ for further information.

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 type of HL?
- 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 HL has relapsed, will you do another tissue biopsy to confirm this?

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