FACT SHEET

Diffuse Large B Cell Lymphoma (DLBCL)

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 effects 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 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.

Diffuse large B-cell lymphoma (DLBCL) is the most common subtype of lymphoma, accounting for around 30% of lymphoma cases. DLBCL affects both men and women (although slightly higher in men) and over half of people are aged over 60. It can also affect children, adolescents and young adults.

DLBCL is an aggressive (fast-growing) B-cell lymphoma subtype. The most common first sign of DLBCL is a painless lump that is rapidly growing in the neck, armpit or groin that is caused by swelling of the lymph node due to lymphoma. For some patients, this swelling may be painful if the lymph node is pressing on an area of the body causing pain. Other symptoms may include night sweats, fever, unexplained weight loss, fatigue, loss of appetite or shortness of breath.

TYPES OF DLBCL

There are a number of different subtypes of DLBCL that may influence what treatment a person receives and how well they respond to this treatment.

DLBCL can further be divided into subtypes based on the type of B-cell it has grown from (termed "cell of origin"). DLBCL can be classified as either germinal centre B-cell (GCB) or activated B-cell (ABC). The doctor examining your lymph node biopsy can tell the difference between these by the staining for certain proteins on the lymphoma cells. At present, this information is not used to direct initial treatment. There is also a more aggressive type of DLBCL called 'double hit' DLBCL which has genetic abnormalities that may also affect how well patients respond to treatment and may be treated with different treatment regimens.

DIAGNOSIS AND STAGING

A biopsy is always required for a diagnosis of DLBCL. 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 in the laboratory to see what the cells look like. The biopsy can be done under local or general anaesthetic depending on what part of the body the biopsy is being performed on.

Once a diagnosis of DLBCL 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. Because DLBCL 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) scan.
- Computed tomography (CT) scan
- Bone marrow biopsy
- Lumbar puncture & Magnetic resonance imaging (MRI) - if lymphoma suspected in the brain or spinal cord

The results of staging tests are then used to diagnose what stage of DLBCL the patient has starting at Stage 1 and going through to Stage 4. Staging is required to choose an appropriate treatment regimen and patients with stage 4 are still curable

with standard treatments. 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 tests and blood tests.

TREATMENT

A combination of chemotherapy medicines and a monoclonal antibody are used to treat DLBCL with some patients also receiving radiotherapy.

DLBCL is a fast-growing lymphoma and needs to be treated quickly. Chemotherapy with a monoclonal antibody treatment, called immunochemotherapy, is the main treatment for DLBCL. The standard treatment regimen for DLBCL is R-CHOP.

- R-CHOP (combinations of rituximab, cyclophosphamide, doxorubicin, vincristine and prednisolone)
- R-EPOCH (combinations of etoposide, vincristine, cyclophosphamide, doxorubicin, vincristine and prednisolone)
- Methotrexate (for patients that are high risk or suspected of lymphoma in the brain or spinal cord)
- Radiotherapy may also be used
- A combination of different treatments
- Clinical trial

The details of your treatment will vary on the stage of your DLBCL and your general fitness. The side effects from treatment varies between the different types of treatment.

DLBCL usually responds well to immune-chemotherapy, but in some people the lymphoma comes back (relapses) or in the rare cases does not respond to initial treatment (refractory in 25-35% of people), there are other treatments that can be successful and further treatment is needed.

- A stem cell transplant (although this treatment is not suitable for everyone)
- Combination of other treatments
- Clinical trial
- CAR T-cell therapy (after 2 prior therapies) soon available in Australia. Until then the government funded medical overseas program (MTOP).

TREATMENTS UNDER INVESTIGATION

Many new individual and combination medicines are currently being tested in clinical trials around the world for both newly diagnosed and relapsed/refractory DLBCL including;

- Chimeric antigen receptor therapy (CAR-T cells)
- Bispecific T-cell engaging antibodies
- Macrophage checkpoint inhibitors
- Polatuzumab vedotin
- Combination therapies

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

RESOURCES AND SUPPORT

Lymphoma Australia offer 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 or contact the Lymphoma Nurse Support Line: 1800 953 081 or nurse@lymphoma.org.au

SOME QUESTIONS TO ASK YOUR DOCTOR

- What type of DLBCL do I have?
- Is there any additional testing that can be done to give you greater insight into how to treat my type of lymphoma?
- What are the treatment options for my type of DLBCL?
- 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 DLBCL has relapsed, will you do another tissue biopsy to confirm this?

This resource was last reviewed and updated in February 2020.