## **FACT SHEET**

# Follicular Lymphoma (FL)

## Lymphoma Australia

## **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.

Follicular Lymphoma (FL) is a common subtype of lymphoma and overall accounting for around 20% of lymphoma cases. FL affects men and women (although slightly more common in men) and typically affects people over the age of 50. There is also a paediatric subtype of FL that occurs in children, adolescents and young adults that is a very rare but can usually be cured.

FL is slow growing B-cell lymphoma. The most common first sign of FL is a painless lump that is slowly growing in the neck, arm pit or groin that is caused by the swelling of the lymph nodes due to the presence of the 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 and people may also notice fatigue, loss of appetite or shortness of breath. Often patients with FL have no other obvious symptoms at diagnosis.

### **TYPES OF FOLLICULAR LYMPHOMA**

There are two different types of FL and this fact sheet will refer to the standard type of FL. The other type of FL is Transformed Lymphoma which can occur over time in patients with standard FL. Transformed Lymphoma is often a more aggressive subtype of lymphoma and it usually requires more intensive types of treatment. For further information please refer to our Transformed Lymphoma Fact Sheet.

## **DIAGNOSIS AND STAGING**

A biopsy is always required for a diagnosis of FL. A biopsy is an operation to remove a 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 FL is made, further tests are needed to be performed to see where else in the body the lymphoma is located (or the "stage", Stage 1 - Stage 4). Staging is required to choose the appropriate treatment regimen. Examples of staging tests 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

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 OPTIONS**

Treatment options for FL are based on the patient's stage, grade, their symptoms and current physical and mental wellbeing.

Early stage 1-3 FL: Early stages (stage 1-2) of FL may be treated

## **FACT SHEET**

with curative intent with radiotherapy alone in a small number of patients. No or little symptoms at diagnosis may not require treatment straight away and may be monitored on 'watch and wait' approach (see 'Watch and Wait' fact sheet). Studies have shown that patients who are managed with this approach have similar survival outcomes to patients who are treated early in the course of their lymphoma. This also has the advantage of avoiding unwanted treatment side effects and having more treatment options in the future, when it comes time to treat the lymphoma. Active treatment can begin when there are more symptoms and progression of the lymphoma.

Advanced stage 3-4 FL: When patients have more symptoms or advanced stage at diagnosis will require combination chemotherapy and a monoclonal antibody with or without radiotherapy. Common combination chemo-immunotherapy regimens include:

- G-Bendamustine (obinutuzumab (Gazyva) and bendamustine)
- R-Bendamustine (rituximab and bendamustine)
- R-CHOP (rituximab, cyclophosphamide, vincristine, doxorubicin and prednisolone)
- R-CVP (rituximab, cyclophosphamide, vincristine and prednisolone)
- Maintenance treatment with a monoclonal antibody is also used for up to 2 years post initial treatment for FL to prolong the amount of time the FL stays in remission.

After treatment, the majority of patients can have a remission that lasts for years however FL should be considered a longterm condition.

For patients with relapsed FL, it depends on the time taken to relapse, stage of FL at relapse and the patient's physical status as to what treatment will be chosen for them at this time. Treatments for relapsed FL can include any of the above treatments, as well as radioimmunotherapy, radiotherapy, a targeted therapy such as idelalisib (Zydelig<sup>™</sup>) or stem cell transplantation.

## TREATMENTS UNDER INVESTIGATION

Many treatments are currently being tested in clinical trials around the world for patients with both newly diagnosed and relapsed FL including;

- Bortezomib (Velcade<sup>™</sup>)
- Chimeric antigen receptor therapy (CAR-T cells)
- Ibrutinib (Imbruvica<sup>™</sup>)
- Lenalidomide (Revlimid<sup>™</sup>)
- Ofatumumab (Arzerra™)
- Pembroluzumab (Keytruda<sup>™</sup>)

## **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:

- Speak to their specialist to see what options are available
  Go to the ClinTrial Refer website <u>www.clintrial.org.au</u> to search available clinical trial
- Download the ClinTrial Refer app from the Apple or Android stores for your smart phone or device. The ClinTrial Refer service was developed to connect patients, health professionals and clinical trial sites
- 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 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 carer's. Please visit our website <u>www.lymphoma.org.au</u> for further information.

## SOME QUESTIONS TO ASK YOUR DOCTOR

- What stage and grade of FL 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 FL?
- Are there any other treatment options that are better for my type of lymphoma but are yet to be funded by PBS in Australia?
- Are there any clinical trials currently available to me?
- If you think my FL has relapse, will you do another tissue biopsy to confirm this?

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