

Follicular Lymphoma (FL)

Lymphoma Australia Fact Sheet

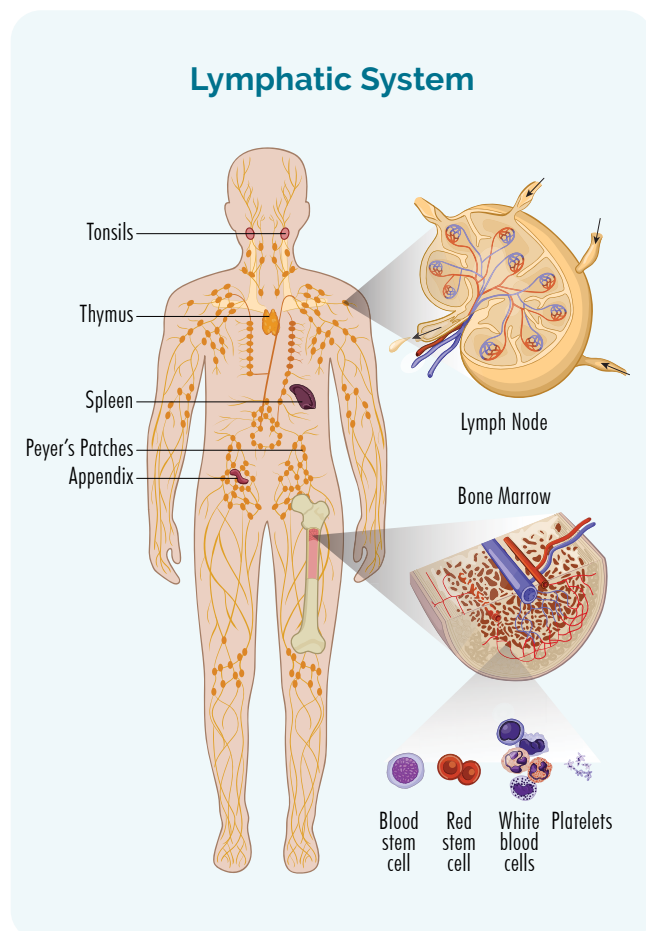
Introduction

Follicular Lymphoma is a subtype of non-Hodgkin Lymphoma (NHL). If you've been diagnosed with FL (FL), you have a type of slow growing blood cancer. This blood cancer affects a type of white blood cells called B-cell lymphocytes (B-cells).

B-cells fight infection and diseases to keep you healthy. They remember infections you had in the past, so if you get the same infection again, your immune system can fight it more effectively. When these cells don't grow or work properly, you can be diagnosed with lymphoma. This means you will have cancerous B-cells called lymphoma cells. Lymphoma cells can travel to any part of your body.

There are more than 80 different types of lymphoma, and FL is a fairly common subtype. About one out of every five people (20%) diagnosed with lymphoma will have FL. It is most common in people over 50 years old. However children, teenagers and young adults can also get FL.

If you have FL, you may not have any symptoms at first. Many people are diagnosed when they have a blood test, scans, or a physical exam for something else. If you do have symptoms, they might include:



- feeling unusually tired (fatigued)
- feeling out of breath
- bruising or bleeding more easily than usual
- infections that don't go away, or keep coming back (recurrent)
- sweating at night more than usual

- losing weight without trying
- a new lump in your neck, under your arms, your groin, or other areas of your body (this is caused by lymphoma cells gathering in the follicles of your lymph nodes and making it grow larger). These lumps may or may not hurt, depending on where they are located.

Diagnosis and staging

Your doctor may suspect you have lymphoma when they get your blood test results, X-ray or other scan results, or do a physical exam. But to diagnose FL, you will need a biopsy. A biopsy is a procedure to remove part, or all of an affected lymph node or bone marrow. Your bone marrow is the spongy part inside your bones where your blood cells are made. The biopsy is then checked by scientists in a laboratory to see if there are changes that help the doctor diagnose FL.

When you have a biopsy, you may have a local or general anaesthetic. This will depend on what part of your body the biopsy is taken from.

If you have FL, your doctor will organise more tests to check if the lymphoma has spread to other parts of your body. These tests are called "staging" and may include:

- Blood tests
 - Positron emission tomography (PET) scan
 - Computed tomography (CT) scan
- Bone marrow biopsy (Your doctor will use a needle to take a sample of marrow from the middle of your bone - usually hip, but sometimes the sample may be taken from a different bone. This will be done with local anaesthetic.
 - Lumbar puncture - (Your doctor will use a needle to take a sample of fluid from near your spine. This is done if the doctor needs to check if your lymphoma is in your brain or spinal cord)
 - Echocardiogram (this is an ultrasound to take a picture of your heart and see how well it is working. You may need this if you are going to have certain chemotherapy medications)
 - Cytogenetic tests – This is where your blood and tumour sample are checked for genetic variances that may be involved in your disease
 - Cytogenetics are tests on your blood and biopsies that look for changes in your chromosomes or genes. We usually have 23 pairs of chromosomes, and they are numbered according to their size. If you have FL, your chromosomes may look a little different.

On your chromosomes are many genes that tell the proteins and cells in your body how to look or act. If there is a change (variation) in these chromosomes or genes, your proteins and cells will not work properly. As a result you can develop different diseases, including lymphoma.

The three main changes that can happen with FL are called a deletion, a translocation and a mutation.

A deletion is when part of your chromosome is missing. If you have a deletion of part of the 6th chromosome it is called "del(6q)". The "q" tells the doctor it is the lower part of the chromosome that is missing. A "p" means the top part is missing. It is the same for other deletions.

If you have a variation in one of your genes called *BCL2* or *BCL6* it can result in changes to your chromosomes. Sometimes in FL, these genes can be rearranged by the cancer, that causes a change called a **"translocation"**. A translocation occurs when two different chromosomes swap a part of their chromosome. The two most common chromosomes involved in FL are the 14th and 18th chromosome. A translocation of these chromosomes is often written as t(14:18).

If you have a mutation in a gene called *EZH2* or a del(6q), it may mean one of your proteins called sestrin cannot work properly. Sestrin gives instruction to mTOR. mTOR is like a switch that tells your cells when to grow and when to stop growing. If you have a mutation affecting sestrin, then it cannot tell mTOR to switch off. This means mTOR is permanently on, so the cancerous cells keep growing. Some clinical trials are looking at medications that target *EZH2* and mTOR to help turn off the mTOR switch.

Your doctor will look at your cytogenetics and be able to work out the best treatment for you, based on the changes in your chromosomes and genes. There are other possible variations than the ones explained above. Please make sure you ask your doctor to explain your individual changes.

Questions for your doctor before you start treatment

1. What tests have been done? What tests still need to be done before treatment?
2. Do I have any genetic abnormalities in my blood or biopsies?
3. Can you explain these results to me?
4. What treatment will work best for me based on these results?
5. What is the intent of this treatment? (e.g., Will you be cured, or go into remission)
6. Do I have any other choices?
7. What are the best treatments that are available for my type of FL, even if they're not available in Australia?
8. What are the main, and the worst side effects of this treatment?
9. How long will treatment take on treatment days?
10. What clinical trials am I eligible for?
11. Will I need to have time off work? If yes, can I see a social worker to help me arrange finances?

12. What support services are available to help me cope with my diagnosis and learn strategies to avoid stress and talk to my family and friends about my diagnosis and treatment

Additional Questions if you live in rural Australia

13. Can I have my appointments and treatment close to home?
14. Are there better treatments for me in larger centres that are not available here?
15. Is telehealth an option for me?
16. How long will I need to be away from home for treatment?
17. How often will I need to come to the city (or be away from home)?
18. What financial, accommodation and travel assistance is available to me?
19. Is there a social worker who can help me arrange these things?
20. Are there any clinical trials I can join, and if I do, can I stay where I am or do I need to travel to another centre?

Paediatric (Childhood) type Follicular Lymphoma

Paediatric FL is rare, and mostly affects children. However, adults less than 40 years old can also have paediatric type FL. The difference between paediatric and standard FL is the cytogenetics. Paediatric FL will have a normal BCL2 and BCL6 gene, whereas standard FL will have a variation in these genes. Depending on which type you have, your doctor will recommend and talk with you about the best treatment option for you.

Treatment options

Because FL is generally a slow growing cancer, you may have it for years without any signs or symptoms. When you are diagnosed, you still may not need any treatment at all. Some people will only need to see their doctor and have a check-up (including some blood tests or scans) to watch if the lymphoma starts growing more quickly. This is called **'watch and wait'**.

Some patients call this "watch and worry", because it can be uncomfortable not doing anything to fight the cancer. But,

If you live a long way from the radiation centre and need help with a place to stay during treatment, please talk to your doctor or nurse about what help is available to you. You can also contact the Cancer Council or Leukemia Foundation in your state and see if they can help with somewhere to stay.

watch and wait is a great way to start. It means your own immune system is fighting the cancer and doing a good job keeping it under control. If your immune system is keeping it under control, you will not need extra help to fight the cancer. Extra medicine that can make you feel quite sick or cause long term side effects, are unlikely to help at this point. Research shows there is no benefit starting treatment early, when your type of cancer will not respond. Your health will not be improved, and you will not live longer by starting treatment earlier. If your FL starts to grow more, or you start to get symptoms from your disease, you might start treatment.

If you need treatment, talk to your doctor about having **genetic testing done before you start**. These tests can tell your doctor what treatment might work best for you. There are many choices available to treat FL, and more medicines are being developed and tested in clinical trials. Your doctor will consider several things when making choices about your treatment.

These include:

- your genetic changes
- how fast your lymphoma is growing
- how bad your symptoms are
- your age
- your overall health

Treatments can include:

Radiation/Radiotherapy – Radiation therapy is a cancer treatment that uses high doses of radiation to kill cancer cells and shrink tumours. Before having radiation, you will have a planning session. This session is important for the radiation therapists to plan how to target the radiation to the lymphoma, and avoid nearby healthy tissue. Radiation therapy usually lasts between 2-4 weeks. During this time, you will need to go to the radiation centre everyday (Monday-Friday) for treatment.

Chemotherapy (chemo) – These medications might be taken as a tablet or be given as a drip (infusion) in a cancer clinic or hospital. Chemo kills fast growing cells so can also affect some of your good cells that grow fast

Monoclonal Antibody (MAB) – Given as an infusion in a cancer clinic or hospital. MABs attach to the lymphoma cell and attract other disease fighting white blood cells and proteins to the cancer so your own immune system can fight the FL.

Targeted therapy – taken as a tablet either at home or in hospital. Targeted therapies attach to the lymphoma cell and block signals it needs to grow and produce more cells. This stops the cancer from growing and causes the lymphoma cells to die off.

Starting treatment

The first time you start treatment it's called first-line treatment. Once you finish your first-line treatment, you may not need treatment again for weeks, months or even years. Some people will not need treatment again at all.

If your FL gets to a point where you do need treatment again, it is called a "relapse". Treatment after a relapse is called second-line treatment (or third-line etc). But some people may not respond to their first line treatment. If your FL does not respond to the treatment, it is called "refractory" disease. In these cases, you may need to start a different type of treatment. If you have refractory disease, and start a new treatment, this is also called second-line treatment.

First-line treatment

If you have stage one or two lymphoma, and need treatment, you may be offered radiation treatment. Some people will only ever need to have one course of radiotherapy, and will not need any other treatment. Some people may have radiation treatment and chemotherapy. This is called chemo-radiation.

If you have stage 3 or 4 FL you may need more treatment. This can be a combination of treatments such as chemotherapy and a monoclonal antibody. You may or may not have this with radiation treatment.

Some of the combination treatments you may be offered are:

- Bendamustine (chemotherapy) and rituximab (or a medication similar to rituximab called a biosimilar) OR obinutuzumab (monoclonal antibody)
- CHOP (chemotherapy – including the chemo medications cyclophosphamide, doxorubicin and vincristine, with a steroid called prednisolone), with a monoclonal antibody - either rituximab (R-CHOP) or obinutuzumab (O-CHOP)
- CVP (chemotherapy – including chemo medications cyclophosphamide, vincristine with a steroid called prednisolone) with a monoclonal antibody either rituximab (R-CVP) or obinutuzumab (O-CVP)
- Chlorambucil (chemotherapy tablet) and rituximab (monoclonal antibody)
- Once chemotherapy ends, you may be offered "Maintenance treatment". This means you will continue to have treatment with the monoclonal antibody (either rituximab or obinutuzumab) for a further two years.
- You may also be eligible to participate in a clinical trial – Ask your doctor about this

First relapse, or refractory disease – Second line treatment

You may not need treatment again, but if you do, your doctor may offer you one of the above treatments or offer you:

- idelalisib (tablet) – idelalisib is a targeted therapy called a P13K kinase inhibitor. It targets proteins that give the lymphoma signals to grow and blocks these signals, preventing further growth and causing lymphoma cells to die off.
- Radiotherapy with or without chemo or monoclonal antibody
- Stem-cell transplant – to learn more about stem cell transplants please see out factsheets
- Transplants in Lymphoma
- Allogeneic stem cell transplants
- Autologous stem cell transplants
- Chimeric antigen receptor therapy (CAR T-cell therapy) – Please see our factsheet for more information on CAR T-cell therapy. All factsheets are available on our website. If you do not have access to a computer and would like a paper copy, please call us on 1800 359 081 or email us at enquiries@lymphoma.org.au
- You may also be eligible to participate in a clinical trial –ask your doctor about this

Further information on different treatment protocols can be viewed [here](#).

Clinical trials

Clinical trials are important to find new medicines, or combinations of medicines to improve treatment of FL in the future. They can also offer you a chance to try a new medicine, before it would otherwise be available. If you are interested in participating in a clinical trial, ask your doctor what clinical trials you are eligible for. You can also read our '[Understanding Clinical Trials](#)' fact sheet for websites to find a clinical trial. Some treatments being tested for FL in clinical trials include:

- Monoclonal antibodies not previously used in FL
- Immune checkpoint inhibitors (these medications block checkpoints that prevent your immune system from fighting the lymphoma)
- Targeted therapy (these medications target different parts of the lymphoma cell to stop it from growing - including EZH2 and mTOR discussed above)
- New medication that has not been put in a group yet and has not been used in people, or has only been given to very few people
- New techniques for radiation treatment
- Combination treatments including different types of medication

Transformed Lymphoma

Slow growing lymphomas like FL can sometimes transform into a faster growing lymphoma. If this happens to you, your treatment options will change. To learn more about transformed lymphoma please see our Transformed Lymphoma factsheet.

Follow up

You will still see your doctor for blood tests and scans after your treatment ends. Your doctor will check you for signs and symptoms of the FL coming back, and for side-effects you may have from your treatment.

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. Lymphoma Australia Fact sheets and booklets include:

- Booklet: What you need to know about lymphoma
- Lymphoma subtypes
- Understanding Clinical Trials
- Emotional impact of a lymphoma diagnosis and treatment
- Fear of cancer recurrence and anxiety

Visit the Lymphoma Australia [YouTube Channel](#) for presentations and interviews on a variety of topics about lymphoma subtypes, management and supportive care.

Contact the Lymphoma Nurse Support Line on 1800 953 081, email: nurse@lymphoma.org.au or join the private Facebook group: [Lymphoma Down Under](#).