Primary Mediastinal B-Cell Lymphoma (PMBCL)

Introduction

Primary Mediastinal B-Cell Lymphoma (PMBCL) is a subtype of non-Hodgkin Lymphoma (NHL). It is most common in people aged between 25 and 40 years, but can affect people of any age. If you have been diagnosed with PMBCL, you have a fast-growing blood cancer that affects a type of white blood cell called B-cell Lymphocytes (B-cells). Many people with PMBCL can be cured with the right treatment.

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.

B-cells are made in your bone marrow (the spongy part in the middle of your bones), but live in your spleen and lymph nodes. Some can even live in a gland called your thymus gland - a butterfly shaped organ that sits in the front of your chest, just behind your breastbone.
Some of the symptoms you get with PMBCL will be related to the structures within your mediastinal area. These may include:

- Cough
- Pain or aching in your chest
- Changes to your voice
- Feeling out of breath
- Swelling in your neck, arms and/or face

Other symptoms that are common for most types of lymphoma include:

- feeling unusually tired (fatigued)
- feeling out of breath
- losing your appetite – not feeling like eating
- bruising or bleeding more easily than usual
- infections that don’t go away, or keep coming back (recurrent)
- frequent high temperatures (fever) or chills
- sweating at night more than usual – drenching your bedding and clothing
- losing weight without trying
- a new lump in your chest, neck, under your arms, your groin, or other areas of your body. These lumps may or may not hurt, depending on where they are.
- B-symptoms

**B-Symptoms**

B-symptoms are a group of three distinct symptoms that some people with lymphoma/CLL can get. They often occur together and may indicate that your lymphoma is more advanced.

- Drenching night sweats – where your clothes and bedding become saturated.
- Losing weight without trying, and without other reason.
- A high fever of 37.5° or more that keeps coming back or does not go away even when you don’t have an infection. You may even get chills.
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Diagnosis and Staging

If your doctor suspects you may have lymphoma, you will need a biopsy. A biopsy is a procedure to remove part, or all of an affected lymph node or a sample of your bone marrow. The biopsy is then checked by scientists in a laboratory to see if there are changes that help the doctor diagnose PMBCL. You may have a local or general anaesthetic to make you more comfortable during the biopsy. This will depend on what part of your body the biopsy is taken from.

Some people may need “keyhole surgery” called a “mediastinoscopy” so the doctor can get enough of affected tissue to sample. If you do have a mediastinoscopy, you will have a general anaesthetic and the surgeon will make a small cut over your breastbone. This is usually done as an outpatient procedure, so you will not need to stay in hospital overnight, but you will need someone to take you home and stay with you overnight.

From the biopsy and location of your disease, your doctor will be able to tell you if you have PMBCL.

If you have PMBCL, your doctor will organise more tests to check if it has spread to other parts of your body. These tests are called “staging”.

Because lymphoma cells can travel to any part of your body, your PMBCL stage will depend on how many areas of your body are affected and where those areas are located. You might have Stage 1, 2, 3 or 4.

Staging of Lymphoma

Stage 1 Stage 2 Stage 3 Stage 4

Staging tests may include:

- Blood tests
- Positron emission tomography (PET) scan
- Computed tomography (CT) scan
- 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, or to make sure your heart is not being affected by your disease).

As well as the above tests, you may have some extra tests to check how well your body’s organs are working, including your lungs, liver, kidneys and heart. This is done to make sure it is safe for you to have treatment, and as a “baseline” so your
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The doctor can make sure that your treatment does not cause too much damage to them.

Please make sure you ask your doctor to explain your test results.

**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 PMBCL, even if they're not available in Australia?
8. What are the main, and the worst side effects of this treatment?
9. What clinical trials am I eligible for?
10. Will I need to have time off work? If yes, can I see a social worker to help me arrange finances?
11. 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?

**If you are parent with dependant children**

12. What support is available to my child so they don't fall behind in school during treatment?
13. What support is available to my other children to help them cope during their siblings' treatment?
14. If you are a parent and you are having treatment for PMBCL you may like to ask What support is available to help my children understand and cope with my diagnosis and treatment?

**Additional Questions if you live in rural Australia**

15. Can I have my appointments and treatment close to home?
16. Is telehealth an option for me?
17. How long will I need to be away from home for treatment?
18. How often will I need to come to the city (or be away from home)?
19. What financial, accommodation and travel assistance is available to me?
20. Is there a social worker who can help me arrange these things?

If your doctor recommends having radiation treatment you will need to go to appointments every day. If you 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.

If you are a parent with dependant children:

12. What support is available to my child so they don't fall behind in school during treatment?
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Treatment options

If you have PMBCL, you will start treatment soon after you are diagnosed. This treatment may have a combination of different medicines, including chemotherapies, a monoclonal antibody, and some people may even have radiotherapy.

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 is usually given over 3 to 5 weeks. During this time, you will need to go to the radiation centre everyday (Monday-Friday) for treatment.

Chemotherapy (chemo) – These medicines might be a tablet or given as a drip (infusion) in a cancer clinic or hospital. Chemo kills fast growing cells, so it 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 lymphoma cells and attract other disease fighting white blood cells and proteins to the cancer. This helps your own immune system to fight the PMBCL.

Immune checkpoint inhibitors (ICIs) – Given as an infusion at a cancer centre or hospital. ICIs work to improve your own immune system, so that your own body can fight the cancer. They do this by blocking some of the protective barriers lymphoma cells put up, that make them invisible to your immune system. Once the barriers are removed, your immune system can see and fight the cancer.

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. First-line treatment lasts for several months and you will have the medicines at regular times. Each time you have the treatment it is called a “cycle”. Once you finish your first-line treatment, you may be cured and not need treatment again.

For some people though, the PMBCL may come back in the future. If this happens it is called a “relapse”. In other people, PMBCL may not go away with your first-line treatment. If this happens it is called “refractory”.

If you have relapsed or refractory PMBCL, you may need a different type of treatment. This new treatment will be called “second line treatment.”
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First-line treatment

Some of the more common treatments you may be offered for your first-line treatment include:

R-CHOP – This includes a monoclonal antibody called rituximab (or a medication similar to rituximab called a biosimilar), and 3 different types of chemotherapy called doxorubicin, vincristine and cyclophosphamide. You will also get a steroid called prednisolone and anti-sickness medicines.

R-EPOCH – This includes the same medications as R-CHOP but has an extra chemotherapy called etoposide (This may also include DA-EPOCH-R. The DA in this means dose adjusted so your doctor may adjust the dose to manage your side effects and symptoms better).

Methotrexate – This is chemotherapy, and may be given to you if you have, or are at risk of getting PMBCL in your brain or spinal cord. The doctor will give you a needle in your back (under sterile conditions), and inject the methotrexate into the fluid surrounding your spinal cord. You may have this with either R-CHOP or R-EPOCH.

Radiotherapy

Clinical trial – These may include targeted therapies and other treatments. Ask your doctor if you are eligible for any clinical trials.

Second-line treatment

If you have relapsed or refractory PMBCL, your doctor will want to give you a different type of treatment, which may work better than the first-line treatment you had. These may include:

- a combination of different chemotherapies and/or monoclonal antibodies or immune checkpoint inhibitors.
- High dose chemotherapy followed by a stem-cell transplant – to learn more about stem cell transplants please see our website [here](#).
- 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 [here](#).
- Clinical trial – These may include targeted therapies and other treatments. Ask your doctor if you are eligible for any clinical trials.

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 for people with PMBCL in the future. They may 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 to find a clinical trial.

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 PMBCL coming back, and for side-effects you may have from your treatment.

Resources and support

Lymphoma Australia offers a wide range of resources and support for people living with lymphoma or CLL, and their carers. How to access our resources:

- **Visit** our website [www.lymphoma.org.au](http://www.lymphoma.org.au) for more information.
- **Phone** our Lymphoma Care Nurse Hotline on 1800 953 081.
- **Email** our Lymphoma Care Nurses nurse@lymphoma.org.au
- **Booklet:** Understanding Non-Hodgkin Lymphoma (NHL)

- **Factsheets:** Visit our [website](http://www.lymphoma.org.au), or give us a call if you would like some factsheets on a variety topics related to lymphoma
- **Join** our Facebook page [Lymphoma Down Under](http://www.facebook.com/LymphomaDownUnder) (make sure you complete all the membership questions when you join).

Cancer Council offers a range of services, including free counselling, to support people affected by cancer, including patients, families and friends. Services may be different depending on where you live. You can contact them at [www.cancer.org.au](http://www.cancer.org.au) or by phone on 13 11 20.

**Medicare Australia:** Check with your GP if you are eligible for a Mental Health Treatment Plan (MHTP). This plan is funded by Medicare and can provide you with up to 10 sessions with a registered psychologist. More information can be found [here](http://www.medicare.gov.au).

**WeCan** is an Australian supportive care website to help find the information, resources and support services you may need following a diagnosis of cancer. You can visit their website at [www.wecan.org.au](http://www.wecan.org.au).

**Canteen** provides support for young people aged 12-25 years who have cancer, or, who have a parent with cancer. Find out more at their website [here](http://www.canteen.org.au).