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Nodular lymphocyte-predominant Hodgkin lymphoma

This information is about nodular lymphocyte-predominant Hodgkin lymphoma (NLPHL), a rare type of Hodgkin lymphoma.

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What is NLPHL?

Nodular lymphocyte-predominant Hodgkin lymphoma (NLPHL) is a type of **Hodgkin** lymphoma.

Lymphoma is a type of blood cancer that develops when white blood cells
called lymphocytes grow out of control. This means that they divide in an
abnormal way or do not die when they should.

• Lymphomas can be grouped as **Hodgkin lymphomas** or **non-Hodgkin lymphomas**, depending on how the abnormal cells look under a microscope.

NLPHL develops from white blood cells called B lymphocytes.

- Lymphocytes are a type of cell in your immune system. They travel around your body in your lymphatic system and your blood, helping you fight infections.
- There are two types of lymphocytes (B cells) and T lymphocytes (T cells).

Nodular lymphocyte-predominant Hodgkin lymphoma is a slow-growing type of lymphoma. It gets its name because of where it grows and how it looks under a microscope:

- It is called 'nodular' because it grows in lymph nodes.
- It is called 'lymphocyte-predominant' after the abnormal cells that a pathologist can see under a microscope. These cells are sometimes called 'popcorn cells' because they look a bit like pieces of popcorn.

Who gets NLPHL?

NLPHL is rare. Around 200 people are diagnosed with it in the UK every year. It can develop at any age, although it's most common in people in their 30s to 50s. It can also affect children and young adults.

NLPHL is much more common in males than females, but scientists don't know why.

3 in 4 people with NLPHL are male

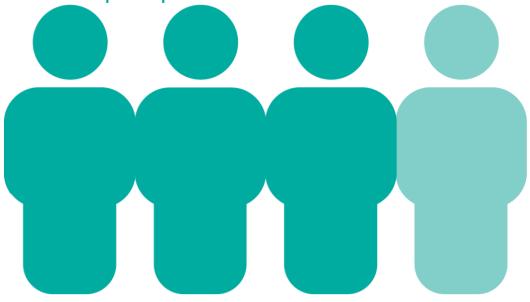


Figure: 3 in 4 people with NLPHL are male

There is no known cause for most cases of NLPHL.

Symptoms

The most common symptom of NLPHL is a lump or lumps. These typically develop in your neck, armpits or groin. They can develop in other places, too. The lumps are caused by lymphoma cells building up in your lymph nodes, which makes the lymph nodes swell. The swollen lymph nodes are usually painless.

I had raised lymph nodes in my neck, and could feel small lumps which felt very small and very mobile. At the time I thought it was just my lymphatic system doing its job.

Surindar, diagnosed with NLPHL in 2019

Swollen lymph nodes are often the only symptom. Sometimes, though, people have other symptoms, such as:

- losing weight without trying to
- **fevers** (temperature above 38°C)
- drenching sweats, especially at night.

These three symptoms are called 'B symptoms'.

Diagnosis and staging

NLPHL is diagnosed by a small procedure called a **biopsy**. A surgeon removes a swollen lymph node (or part of one) and sends it to a lab. In the lab, a specialist lymphoma pathologist looks at it under a microscope to check for lymphoma cells. They might also run specialised tests to help them find out the exact type of lymphoma it is.

You also have **scans** to find out if you have lymphoma in any other parts of your body. This is called **staging**. Staging usually involves having a **CT scan** or a **PET/CT scan**. Children might have an **MRI scan** instead.

Depending on the results of your other tests, you might have a **bone marrow biopsy** to check for lymphoma cells in the spongy tissue in the centre of your bones where blood cells are made. This involves having a sample of cells taken from your hip bone or, occasionally, your breastbone.

You also have **blood tests** to look at your general health, check your **blood cell counts**, make sure your kidneys and liver are working well and rule out infections that could flare up if you need treatment.

You usually have your tests done as an outpatient. It can take a few weeks to get all the results back. Waiting for the results of your tests can be difficult. It is important for your medical team to find out exactly what type of lymphoma you have and where it is. This helps them plan the most appropriate treatment for you.

Outlook

Treatment for NLPHL is very effective. Most people either go into complete remission (no sign of lymphoma on tests and scans), or have a long period without any symptoms. Although NLPHL can come back (relapse), treatment is usually successful. For most people, having NLPHL will have little or no effect on how long you live.

Your medical team are best placed to advise you on your outlook based on your individual circumstances. They can use the results of your tests and other factors (for example, your age and physical fitness) to help choose the best treatment for you.

Transformation

Occasionally, NLPHL can change (transform) into a faster growing type of lymphoma. This is uncommon. It happens in around 1 in 100 people with NLPHL each year.

Transformed lymphoma needs stronger treatment than NLPHL. It is usually treated like a type of fast-growing (high-grade) non-Hodgkin lymphoma called **diffuse large B-cell lymphoma (DLBCL)**. Transformed NLPHL often responds well to treatment and many people go into complete remission (no sign of lymphoma on tests or scans).

Treatment

The treatment your medical team will recommend for you depends on several factors, including:

- the stage of your lymphoma
- your symptoms
- your age and general health
- your feelings about the treatment options
- whether or not you plan to have children in the future.

Your medical team also take into account the possible **side effects** or **late effects** (health problems that develop months or years after treatment) of the different treatment options. They should tell you about these.

If appropriate, your consultant can refer you to a **fertility** specialist before you start treatment.

Treatment of early stage NLPHL

Most people with NLPHL have **early stage** lymphoma (**stage 1** or **stage 2** without any **B symptoms**) when they are diagnosed. This means the lymphoma is only growing in one place, or a few places close together. If you have early stage NLPHL, you might only need treatment to the area affected by lymphoma.

If your lymphoma is in a place where it can be removed, you might be offered surgery. If you have no signs of lymphoma left after surgery, you might not need any more treatment. Instead, your medical team might recommend that you have regular check-ups to make sure the lymphoma stays under control. This is called **active monitoring** ('watch and wait'). If there are lymphoma cells left behind after your surgery, your medical team are likely to recommend a course of **radiotherapy**, too.

If you have early stage NLPHL that can't be removed, you might have radiotherapy to the part of your body affected by lymphoma. This is usually very successful.

Your consultant might ask you if you'd like to take part in a **clinical trial**, if there is one suitable for you. If you don't want to take part in a clinical trial, or if there isn't one suitable for you, there are other effective options.

If these options aren't suitable for you, your team are likely to suggest the same treatment options used for people with advanced-stage NLPHL.

Treatment of advanced stage NLPHL

Around 1 in 4 people have advanced stage NLPHL (stage 2 with B symptoms, stage 3 or stage 4) when they are diagnosed. Advanced NLPHL still responds very well to treatment. The main difference is that you have treatment to your whole body rather than just one area.

If you have advanced-stage NLPHL but your symptoms aren't interfering with your day-to-day life, you might not need treatment straightaway. Instead, your medical team might recommend **active monitoring** ('watch and wait'). This involves having regular check-ups to monitor how the lymphoma is affecting you.

Active monitoring means that you avoid the side effects of treatment for as long as possible. Treatment is still available if your lymphoma starts to cause problems or if you develop symptoms that are hard to cope with. Treatment is just as successful in people who save it until they need it as in people who have treatment straightaway.

If your lymphoma needs treatment, you are most likely to have **chemotherapy** combined with the **antibody** treatment **rituximab** (R). The combination of chemotherapy drugs you might have includes:

- R-ABVD
- R-CHOP
- R-CVP.

People who are not well enough to have chemotherapy might have rituximab on its own.

Follow-up

When you finish treatment, you have a scan to check how well your lymphoma has responded. This is usually a CT or PET/CT scan, but children might have an ultrasound or MRI scan instead.

You have regular **follow-up appointments** with your specialist after you finish your treatment. These are to check that:

- you are recovering well from treatment
- you have no signs of the lymphoma coming back (relapsing)
- you are not developing any late effects (side effects that can develop months
 or years after treatment).

At first, your follow-up appointments are every 3 months or so. If you stay well, they gradually become less frequent. Your medical team should explain how follow-up works at your hospital.

Relapsed or refractory NLPHL

Treatment for NLPHL is usually effective. However, it can sometimes come back (relapse) and need more treatment. Occasionally, it doesn't respond well to initial treatment (refractory lymphoma). Relapsed or refractory lymphoma are usually treated in a similar way.

If NLPHL comes back, you might have the same symptoms you had before or you might have different symptoms. If your medical team suspects your lymphoma has come back, they are likely to recommend that you have another **biopsy** to make sure it hasn't changed (**transformed**) to a faster-growing type of lymphoma. You might also have more scans.

Relapsed or refractory NLPHL usually responds very well to treatment.

If your lymphoma comes back but it isn't causing problems, you might not need treatment straightaway. Instead, you might have a period of **active monitoring** ('watch and wait').

If you need treatment, your medical team might recommend:

radiotherapy if the lymphoma is only in one part of your body

- chemotherapy, usually with different drugs if you had chemotherapy before, although your consultant might consider using the same drugs again if they worked well for you
- rituximab, either on its own or with chemotherapy.

If your lymphoma comes back very soon after treatment, or comes back several times, your medical team might suggest a **stem cell transplant**. This is a very intensive treatment and you have to be fit enough to have it. Most people with NLPHL do not need a stem cell transplant.

Research in NLPHL

NLPHL is rare. There are not many clinical trials looking at new treatment options. However, existing treatment is usually very successful. Most research is looking at ways of using treatments more effectively to make sure **side effects** and **late effects** are as low as possible.

Your consultant might offer you the chance to take part in a clinical trial if there is one suitable for you. You can find out more about clinical trials and search for a trial that might be suitable for you at Lymphoma TrialsLink.

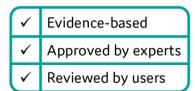
References

The full list of references for this page is available on our website. Alternatively, email publications@lymphoma-action.org.uk or call 01296 619409 if you would like a copy.

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