Nodular lymphocyte-predominant Hodgkin lymphoma

This information is about nodular lymphocyte-predominant Hodgkin lymphoma (NLPHL), which is an uncommon type of Hodgkin lymphoma.

On this page

Quick overview of NLPHL

What is NLPHL?

Who gets NLPHL and why?

Symptoms

Diagnosis and staging

Outlook

Treatment

Relapse

Transformation

Research
Quick overview of NLPHL

This section is an overview of the information on this page. There is more detail in the sections below.

What is it?

NLPHL is a rare, slow-growing type of lymphoma. It is a type of cancer where the abnormal cells are lymphocytes (a type of white blood cell).

What are the symptoms and how is it diagnosed?

The only symptom for most people with NLPHL is one or more lumps. These are enlarged lymph nodes (swollen glands). They are often in only one place in the body. A few people have other general symptoms of lymphoma, like night sweats, weight loss and fevers.

NLPHL is diagnosed by biopsy. You have other tests to find out how widespread it is, for example blood tests and scans.

How is it treated?

Treatment for NLPHL is usually very successful. If NLPHL is in only one place or the lumps are close together and you are well, you might only need surgery to remove the lumps. Some people then have radiotherapy to the affected area. Others are monitored to look out for the lymphoma coming back. More widespread NLPHL might be monitored until it is causing problems. If you have symptoms, you are likely to have chemotherapy. If NLPHL comes back, treatment is usually successful again.

What is NLPHL?

Nodular lymphocyte-predominant Hodgkin lymphoma (NLPHL) is a type of lymphoma that develops from abnormal B lymphocytes (B cells, which are a type of white blood cell).
NLPHL is not very common – around 1 in 20 cases (5%) of Hodgkin lymphoma are NLPHL. There are about 200 cases of NLPHL diagnosed in the UK each year.

Other types of Hodgkin lymphoma are grouped as classical Hodgkin lymphoma and treated in the same way. NLPHL tends to grow more slowly than classical Hodgkin lymphoma, and is often diagnosed earlier. It can be treated differently from classical Hodgkin lymphoma.

The name ‘NLPHL’ describes the abnormal cells and where they are usually found:

- ‘nodular’ because it occurs in the lymph nodes (glands)
- ‘lymphocyte-predominant’ are particular cells from this type of lymphoma that can be seen under a microscope; you might hear them called ‘popcorn cells’ because they look a bit like pieces of popcorn.

**Who gets NLPHL and why?**

NLPHL affects more males than females.

3 in 4 people with NLPHL are male
It most often affects people in their 30s–50s. It can also occur in older people and in children.

There is no known cause for most cases of NLPHL. Brothers, sisters and parents of someone with NLPHL have a very slightly higher risk of getting it themselves, but this risk is still very low.

## Symptoms

Most people with NLPHL develop lumps they can see or feel, often in only one area of the body. These are enlarged (swollen) lymph nodes. The lumps tend to grow quite slowly and are not usually painful. They are usually the only symptom of NLPHL.

Only a small number of people with NLPHL have other symptoms. Possible symptoms might include ‘B symptoms’: weight loss, night sweats and a high temperature (fever) that comes and goes. These symptoms are uncommon in people with NLPHL, though they can develop in many other types of lymphoma.

## Diagnosis and staging

NLPHL is diagnosed by tests on a sample of the abnormal cells – a biopsy. These tests help to distinguish NLPHL from other types of lymphoma.

You have other tests to find out which areas of your body the lymphoma is growing in; this is called ‘staging’.

Your tests are likely to include:

- **Blood tests**, which are done to check your blood cell counts, your general health and to rule out infections, such as hepatitis, that could flare up when you have treatment.

- A **scan**, most likely a CT scan or a PET/CT scan, which looks for any signs of lymphoma in internal lymph nodes and body organs such as your liver and spleen. Some people have an MRI scan instead, particularly children.
A few people might have a bone marrow biopsy to check for lymphoma in the bone marrow (the spongy tissue in the centre of your bones where blood cells are made). Very few people with NLPHL have lymphoma in their bone marrow.

You usually have your tests done as an outpatient. It can take a few weeks to get all the results back. Waiting for test results can be a worrying time. It is important that your consultant knows exactly what type of lymphoma you have and how it is affecting you so you can have the best treatment.

Most people with NLPHL have ‘early stage’ or ‘localised’ lymphoma (stage 1 or 2) when they are diagnosed. This is lymphoma 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.

Some people have more widespread NLPHL when they are diagnosed – ‘advanced stage’ lymphoma (stage 3 or 4).

1 in 4 people have advanced NLPHL

Advanced NLPHL still responds very well to treatment. The main difference is that you have treatment to your whole body rather than to one area only.
Outlook

The outlook for NLPHL is generally very good. More than 9 in 10 people are either cured or have a long time with no sign of the disease (known as a remission), often 10 years or longer. There are several reasons for this:

- it is usually diagnosed at an early stage, when it is in only one or two groups of lymph nodes
- it tends to grow in lymph nodes that can be easily felt, rather than deep inside the body
- it grows very slowly
- it usually responds very well to treatment.

Although NLPHL can come back (relapse), treatment is often successful and most people are cured or have another long remission.

Your consultant is best placed to advise you on your outlook based on your individual circumstances. They can use the results of your tests and consider other factors, like your age, symptoms, and other conditions you might have to predict how likely you are to respond to a particular treatment.

Treatment

Treatment for NLPHL is usually very successful. Most people live for many years, even if the lymphoma comes back and needs more treatment.

Your treatment plan depends on several factors including:

- the stage of your lymphoma
- whether you have symptoms
- your age and general health
- your stage of life – for example, whether you plan to have children.

Your consultant thinks about the long-term or late effects (side effects that develop long after treatment) of treatment when making a treatment plan for you.
Your medical team should give you information about the possible side effects and late effects of your planned treatment.

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

This section describes the usual treatment in adults. We have a section dedicated to lymphoma in children and young people. You might find this useful if you are the parent of a child with NLPHL or you are a young person with NLPHL.

**Treatment of early stage NLPHL without B symptoms**

If you have early-stage NLPHL with no other symptoms, you are most likely to have:

- surgery to remove all the lymph nodes that contain lymphoma, followed by radiotherapy to the affected area
- surgery followed by active monitoring (‘watch and wait’)
- radiotherapy alone.

If you are having surgery, your medical team should give you more information about what’s involved, as this depends where the lymph nodes are. Ask how long you need to stay in hospital and what to expect from your recovery.

If you have no signs of lymphoma after your surgery, your consultant might suggest you have active monitoring (‘watch and wait’). You don’t have any more treatment but instead you have regular check-ups to make sure the lymphoma is still under control. Active monitoring allows you to avoid the side effects of radiotherapy. There is a chance that the lymphoma will come back in the future, but if it does, you can have more treatment. This is nearly always successful. Research suggests there is no difference in survival rates between people who have active monitoring and those who have radiotherapy after their surgery.

If there is lymphoma left behind after your surgery, you are likely to have radiotherapy. If your lymphoma is in more than one place, you might have
radiotherapy on its own, without having surgery. If you can’t have radiotherapy, you are treated as described for advanced-stage NLPHL.

**Treatment of early stage NLPHL with B symptoms**

If you have early stage NLPHL with B symptoms, you may need stronger treatment. You are likely to be treated in the same way as someone with advanced NLPHL and symptoms. Most people in this situation have chemotherapy.

**Treatment of advanced stage NLPHL**

If you have advanced-stage (stage 3 or 4) NLPHL but are well, you may have active monitoring (‘watch and wait’ – check-ups to make sure the lymphoma is not getting worse). Treatment can be safely reserved until the lymphoma is causing problems.

If your lymphoma is causing symptoms, you are most likely to have treatment with a combination of chemotherapy drugs (a regimen). Regimens that might be considered include:

- **ABVD**, which is the standard treatment for classical Hodgkin lymphoma
- **CHOP**, which is a common regimen for people with many types of lymphoma
- **CVP**, which is often used for children with NLPHL.

Many people are given the antibody treatment rituximab with their chemotherapy.

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

**Follow-up**

You have regular follow-up appointments with your specialist after you finish your treatment. At first, these are every three months or so. If all remains well, they gradually become less frequent.
Relapse

Like other types of low-grade (slow-growing) lymphoma, NLPHL can sometimes relapse (come back). It is very important to keep all your follow-up appointments as relapse can happen many years after you were first treated.

Relapsed NLPHL usually responds very well to treatment.

The lymphoma is most likely to come back in the same place as it was before. It rarely becomes more widespread.

If you relapse, you need a biopsy to make sure the NLPHL has not transformed (changed to a faster-growing lymphoma). Transformed lymphoma needs different treatment.

If NLPHL relapses, your consultant considers the same factors as before when deciding how best to treat you, as well as:

- what treatment you had before and how you coped with it
- how long it was before your NLPHL came back.

You may have a period of active monitoring (‘watch and wait’) if the lymphoma is not causing problems.

Treatments your consultant might consider include:

- **radiotherapy**, particularly if you did not have it as part of your previous treatment
- **chemotherapy** – usually with different drugs if you had chemotherapy before but 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 several times and you are fit enough, your consultant might consider high-dose chemotherapy and a **stem cell transplant**. People with NLPHL do not usually need this intensive form of treatment.
It can be upsetting when your lymphoma comes back. Remember that treatment for relapsed NLPHL is usually very successful. You are likely to get back into long-term remission (no sign of lymphoma).

**Transformation**

NLPHL can sometimes transform (change) into a faster growing type of lymphoma. Transformation is uncommon. It is difficult to say what proportion of people’s lymphoma transforms because NLPHL is rare. Recent reports suggest fewer than 1 in 10 people experience transformation.

Transformed lymphoma often needs stronger treatment than NLPHL to cure it. It is usually treated like diffuse large B-cell lymphoma (DLBCL). Many people with transformed NLPHL still have a good outcome and can be cured.

**Research**

NLPHL treatment is often very successful so most research for this type of lymphoma focuses on reducing side effects, particularly late effects. Other clinical trials are looking at treatment for NLPHL that has relapsed or hasn’t responded very well to treatment (refractory). These trials test different chemotherapy regimens or newer targeted drugs.

Your consultant may 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**

Find our key references at the top, followed by the webpages and journal articles used to write this page.

- Key references:
  McKay P, et al. Guidelines for the investigation and management of


- Webpages:

- Journal articles:


- Akhtar S, et al. High dose chemotherapy and autologous stem cell transplantation in nodular lymphocyte-predominant Hodgkin lymphoma: A retrospective study by the European society for blood and marrow


Further reading

- Living with lymphoma
- Active monitoring
- Chemotherapy
- Radiotherapy
- Rituximab
- Transformation of lymphoma
- Hodgkin lymphoma overview page
- Glossary
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