Large granular lymphocytic leukaemia (LGLL)

This fact sheet will help you to understand LGLL, its side effects, possible treatments and how to take care of yourself if you have LGLL.

What is large granular lymphocytic leukaemia (LGLL)?

Large granular lymphocytic leukaemia (LGLL) is a type of blood cancer and a rare form of leukaemia. People with LGLL have large abnormal white blood cells (lymphocytes), containing pink granules; this is how it gets its name.

How does LGLL happen?

Everyone has white blood cells called large granular lymphocytes in their blood. They're part of the normal immune system and are cells which fight viruses and protect us from infection. LGLL happens when the body makes too many abnormal large granular lymphocytes in the bone marrow and not enough of the other types of blood cells. There are three main types of lymphocytes: B-cells, T-cells and NK-cells. LGLL affects the T-cells or, less commonly, the NK-cells.

There are two main types of LGLL:

- slow growing (chronic) T-cell or NK-cell LGLL (this is the most common type)
- fast growing (aggressive) T-Cell and/or NK-cell LGLL (a rare type).

To find out more about how your blood, bone marrow and immune system works, go to **bloodwise.org.uk**

Who gets LGLL?

LGLL is very rare. Around 200 people in the UK are diagnosed each year. The causes of LGLL are not known but it's important to understand that LGLL is not a condition which can be caught from someone else (contagious) and it can't passed on from parent to child (inherited).

We don't know exactly what causes LGLL, but we do know that there are some things that can affect how likely you are to develop it:

- **Sex** – LGLL affects both men and women, but it's slightly more common in women.
- **Age** – LGLL mainly affects older people – the average age at diagnosis is 60. Children rarely get LGLL.
- **Abnormal cell pathways and gene mutations** – people with LGLL may have abnormalities in genes such as STAT3 and/or STAT5.
- **Other health conditions** – around 30% of people with LGLL have rheumatoid arthritis. Rheumatoid arthritis is the second most common form of arthritis in the UK and causes inflammation in the joints. This suggests that overactivity of the immune system may be one of the causes of LGLL.

Symptoms of LGLL

Everyone is different, so it's important to remember that not everyone will have the same symptoms.

Around two thirds of people with LGLL have symptoms when they're diagnosed. Common symptoms before diagnosis include:

- extreme tiredness (fatigue) which may be caused by a lack of red blood cells (anaemia)
- muscle, bone and joint pain
- infections
- chronic neutropenia (a low level of a certain type of blood cell called neutrophils which means that you're more at risk of infection)
- swelling of the lungs (pneumonia)
- an enlarged spleen
- sore throat and loss of voice
- night sweats and difficulty sleeping
- breathlessness
- a loss of appetite.

One third of people with LGLL don't experience any symptoms and their condition may be diagnosed by chance following a routine blood test or a health check for something else.

Aggressive T-cell LGLL and NK-cell LGLL symptoms

If you have a fast growing (aggressive) T-cell or NK-cell LGLL, you may have:

- an enlarged liver and spleen (hepatosplenomegaly)
- swollen glands (lymph nodes)
- fever
- weight loss
- night sweats.

How is LGLL diagnosed?

LGLL is most often diagnosed from a full blood count (FBC) which is a blood test that measures the number of each type of cell in your blood: red cells, white cells and platelets. A FBC will give an initial diagnosis of LGLL, because the doctor can see the large pink granules under a microscope. The blood count has to be repeated after a few weeks to confirm the diagnosis.

Don't be afraid to ask your doctor to explain anything that you don't understand about your diagnosis.

You might find it helpful to write things down, or ask the person you're with to take notes for you. You may also find it helpful to take this fact sheet with you to appointments if you want to explain your condition to your healthcare team.

As LGLL is difficult to diagnose, your doctors may have to do some more tests, including:

- a blood test called immunophenotyping, to detect the presence or absence of different white blood cells (this tells if you have T-cell or NK-cell LGLL)
- a bone marrow test
- scans to monitor the impact of LGLL on other organs in your body.

You'll have more blood tests during follow-up appointments to monitor your body's response to LGLL and to check if you need treatment.

Find out more about blood and bone marrow tests and scans at **bloodwise.org.uk**

Treatments for chronic LGLL

Everyone is different and may need a different treatment.

As LGLL is usually a slow developing (chronic) condition, many people may not notice symptoms for a while. Some people may be placed on 'watch and wait' until they do need treatment. This will mean you won't start treatment straight away – instead you'll be monitored with regular tests and only start treatment when you need to. However, as a result of frequent infections, the majority of people (60%) with LGLL will eventually need treatment.

Currently there's not one standard form of treatment for LGLL, so it's important to speak to your healthcare team to find out which treatment is best for you.

The treatments for LGLL are designed to lower the activity of your immune system. Some of these treatments include:

- a mild chemotherapy (cell-destroying) drug, taken as a tablet, such as cyclophosphamide
- non-chemotherapy drugs like cyclosporine A
- an immunosuppressive drug called methotrexate
- steroid drugs such as prednisolone, which can be used to temporarily to improve neutropenia and the pain caused by rheumatoid arthritis
- in rare cases, when other treatments haven't worked, you may have your spleen removed (splenectomy) which can improve blood counts, but has shown limited results.

Treatment for aggressive T-cell LGLL and NK-cell LGLL

Unfortunately, aggressive T-cell LGLL and NK-cell LGLL are resistant to therapy, which means they're very hard to treat or they're untreatable. There's currently limited research into which treatments work best, however, more intensive chemotherapy is generally used.

If you don't respond to any of these treatments or if the LGLL comes back (relapse), there are other treatments available. Speak to your healthcare team to find out which treatment is best for you.

Supportive care

Not everyone with LGLL will have all of the treatment described but most are likely to have supportive care. Supportive care means treatments which do not impact on the LGLL cells directly, but can make you feel better. These include:

- G-CSF (granulocyte-colony stimulating factor), a drug used to raise the level of neutrophils and lower your risk of infection
- taking antibiotics on a long-term basis to help prevent severe infections
- blood transfusions to treat anaemia (low red blood cells)
- erythropoietin (EPO), a drug which can stimulate the production of your red blood cells to treat anaemia and avoid the need for blood transfusions.

For more information on supportive care, download our booklet from **bloodwise.org.uk**

Clinical trials

You may also want to consider taking part in a clinical trial which might give you the opportunity to have the newest available treatment, which may not be offered outside of the trial.

Find a clinical trial for LGLL on the UK Clinical Trials Gateway. Go to ukctg.nihr.ac.uk or read our booklet **Your guide to clinical trials**.

Common side effects of treatment

Different treatments will have different side effects. You can always speak to your specialist or healthcare team about the possible side effects of a particular treatment. You may experience some of the following symptoms with treatment:

- an increased risk of infection
- tiredness
- breathlessness and shortness of breath
- bruising or bleeding
- weakness
- yellowing of the skin (jaundice)
- loss of appetite
- bowel and liver changes
- sickness and vomiting
- hair loss
- loss of fertility
- diarrhoea.

The drugs used for treating chronic LGLL are not aggressive cell-killing drugs. This means that the side effects are usually mild.

Living with LGLL

Finding out you have LGLL can be really upsetting and can come as a shock, especially if you were diagnosed by chance. You might feel a range

of emotions at different times. There can be a physical impact on your day-to-day life too. It's important to know that people with LGLL can have a good quality of life.

- **Looking after yourself emotionally** – it's important to look after your mental health and wellbeing when you have LGLL. If you were diagnosed by chance, it can come as a shock, and if you're on watch and wait you might feel very anxious. You can always speak to your friends, family and healthcare team for support.
- **Food safety and diet** – if you have neutropenia, which means you're at greater risk from infections, you may need to change your diet and follow some standard food safety and hygiene advice to reduce the chance of getting an infection from your food.
- **Crowds and hygiene** – if you have severe neutropenia you may need to avoid crowded places such as train stations and festivals as this can increase your risk of infection.

To find out how neutropenia may affect your diet, read our booklet **Eating well with neutropenia**.

Your doctor or healthcare team can give you advice on any lifestyle changes you might need to make to keep yourself safe.

What's the outlook for someone with LGLL?

Although it's not possible to cure LGLL at the moment, most people (87%) who do have treatment generally respond well and have a good quality of life.

People with LGLL have a higher risk of developing infections which in very rare cases can be fatal. Having treatment – when you need it – lowers this risk.

Finding out more

We offer patient information on many blood cancer types and topics, online and in free printed booklets. They cover everything from symptoms and diagnosis through to treatment and living with your condition.

For our patient information, go to **bloodwise.org.uk/information-and-support**

We also have an online community you may like to join **bloodwise.org.uk/our-community**

For more support contact the UK LGLL Facebook support group by joining Facebook and searching 'UK LGLL'.

Or you can call our support line on **0808 2080 888** (Mon–Fri 10am–4pm). This is a freephone number.

See our website for more details of cancer information specialists and support groups. Go to **bloodwise.org.uk/living/where-get-help-and-support**

About Bloodwise

We're the UK's specialist blood cancer charity. We've been working to beat blood cancer since 1960. We fund world-class research; provide practical and emotional support to patients and their loved ones; and raise awareness of blood cancer.

We'd like to thank Dr Samir Agrawal for his help and support in developing the content and Russell Patmore for checking for clinical accuracy. A list of references used in this factsheet is available on request. Please email **information@bloodwise.org.uk**

Our fact sheets contain general information. Always listen to the advice of your specialist about your individual condition – because every person is different.

Disclaimer
We make every effort to make sure that the information in this fact sheet is accurate, but you shouldn't rely on it instead of a fully trained clinician. It's important to always listen to your specialist and seek advice if you have any concerns or questions about your health.

Bloodwise can't accept any loss or damage resulting from any inaccuracy in this information, or from external information that we link to.

The information in this fact sheet is correct at the time it was printed (July 2017).

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