
Large Granular Lymphocytic Leukaemia (LGLL)

**A Guide for
Patients**

Leukaemia Care
YOUR Blood Cancer Charity

Introduction

Being diagnosed with Large Granular Lymphocytic Leukaemia (LGLL) can be a shock, particularly when you have never heard of it. If you have any questions about LGLL, including what causes it, who it affects, how it affects your body, what symptoms to expect and likely treatments - this booklet covers the basics for you.

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If you would like any information on the sources used for this booklet, please email communications@leukaemiacare.org.uk for a list of references.

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About Leukaemia Care

Leukaemia Care is a national charity dedicated to ensuring that people affected by blood cancer have access to the right information, advice and support.

Our services

Helpline

Our helpline is available 9.00am - 10.00pm on weekdays and 9.30am - 12.30pm on Saturdays. If you need someone to talk to, call **08088 010 444**

Nurse service

We have two trained nurses on hand to answer your questions and offer advice and support, whether it be through emailing **nurse@leukaemicare.org.uk**, over the phone on **08088 010 444** or via LiveChat.

Patient Information Booklets

We have a number of patient information booklets like this available to anyone who has been affected by a blood cancer. A full list of titles – both disease specific and general information titles – can be

found on our website at **www.leukaemicare.org.uk/support-and-information/help-and-resources/information-booklets/**

Support Groups

Our nationwide support groups are a chance to meet and talk to other people who are going through a similar experience. For more information about a support group local to your area, go to **www.leukaemicare.org.uk/support-and-information/support-for-you/find-a-support-group/**

Buddy Support

We offer one-to-one phone support with volunteers who have had blood cancer themselves or been affected by it in some way. You can speak to someone who knows what you are going through. For more information on how to get a buddy call

08088 010 444 or email
support@leukaemicare.org.uk

Online Forum

Our online forum, **www.healthunlocked.com/leukaemia-care**, is a place for people to ask questions anonymously or to join in the discussion with other people in a similar situation.

Patient and carer conferences

Our nationwide conferences provide an opportunity to ask questions and listen to patient speakers and medical professionals who can provide valuable information and support.

Website

You can access up-to-date information on our website, **www.leukaemicare.org.uk**, as well as speak to one of our care advisers on our online support

service, LiveChat (9am-5pm weekdays).

Campaigning and Advocacy

Leukaemia Care is involved in campaigning for patient well-being, NHS funding and drug and treatment availability. If you would like an update on any of the work we are currently doing or want to know how to get involved, email **advocacy@leukaemicare.org.uk**

Patient magazine

Our quarterly magazine includes inspirational patient and carer stories as well as informative articles by medical professionals. To subscribe go to **www.leukaemicare.org.uk/communication-preferences/**

What is LGLL?

Large granular lymphocytic leukaemia (LGLL) is characterised by an excessive production of certain white blood cells - large granular T cell or natural killer (NK) lymphocytes - which infiltrate the bone marrow, spleen and liver.

Lymphocytes are a type of white blood cell involved in the body's immune response. There are two main types of lymphocytes, called B-cells and T-cells:

- 1.** B-cells (formed in the bone marrow) produce antibodies that seek out and immobilise bacteria, viruses, and toxins which invade the body.
- 2.** T-cells (formed in the thymus gland, behind the sternum) destroy the invading organisms that have been tagged by the B-cells as well as any cells that have become cancerous.

NK cells are also a type of lymphocyte best known for killing virally infected cells, and detecting and controlling early signs of cancer. Unlike B-cells and T-cells, they are part of the innate immune system and can be activated immediately once a pathogen attacks.

LGLL accounts for 2% to 5% of chronic lymphoid cancers in Europe and North America, and 5% to 6% in Asia. LGLL is commonly seen in elderly patients and has a median age at diagnosis of 66.5 years. Although, up to 15% of patients are younger than 50-years-old when diagnosed with LGLL. The incidence of LGLL is the same in men and women.

There are three categories of mature T-cell and NK-cell cancers:

- 1.** T-cell LGLL (85% of LGLL cases)
- 2.** Chronic lymphoproliferative disorder of NK-cells (10% of LGLL cases)
- 3.** Aggressive NK-cell LGLL (5% of LGLL cases)

T-cell LGLL and chronic lymphoproliferative disorder of NK-cells share the same symptoms and diagnostic features, as well as treatment options. Patients mainly present with cytopenias (reduction in the number of mature blood cells), most commonly neutropenia (as a result of low white blood cell counts) with or without anaemia (low red blood cell count). Thrombocytopenia (low platelet count) is less common.

Symptoms include recurrent infections as a result of the neutropenia or fatigue. They can also have associated autoimmune destruction of red cells or platelets.

Aggressive NK-cell LGLL is associated with Epstein-Barr virus infection (a virus linked with glandular fever). It is seen mainly in young patients of Asian descent, and has a very poor prognosis as it does not generally respond to chemotherapy.

What causes LGLL?

The actual cause of LGLL is not completely understood, but recent studies have shown that the STAT3 (Signal Transducer and Activator of Transcription 3) gene has frequently undergone mutation in patients with LGLL. Both T-cell LGLL and chronic NK-cell lymphocytosis have the same genetic mutation of STAT3. STAT3 genes provide instructions for making the proteins involved in chemical signalling pathways within cells.

LGLL is commonly associated with autoimmune diseases, particularly rheumatoid arthritis. The association of LGLL with

autoimmune disease is reinforced by the presence of rheumatoid factor, antinuclear antibodies and antineutrophil antibodies. These are all proteins which attack healthy cells in the body and are made by the immune system.

Chronic lymphocytic leukaemia, follicular lymphoma, and mantle cell lymphoma which are all B-cell associated diseases have also been reported in patients with LGLL. The association of B-cell abnormalities in LGLL, which is essentially a T-cell and NK-cell cancer, suggests that either a common antigen (toxin or other foreign substance which induces an immune response in the body) is causing both B-cell and T-cell cancers, or that LGLL is due to a humoral immune response (involving antibodies in body fluids rather than in body cells), where an overactive anticancer response is the reason for the overproduction of lymphocytes.

Symptoms of LGLL

Chronic LGLL

The chronic T-cell and NK-cell variants of LGLL generally appear and progress slowly; therefore, there may be few symptoms at the onset. The most common features of LGLL are summarised below:

- Mild cytopenias (neutropenia with or without anaemia), often picked up on routine blood tests
- Presence of autoimmune diseases (e.g. rheumatoid arthritis)
- Elevated numbers of large granular lymphocyte (LGL) T-cell or NK-cells in the blood
- Splenomegaly (enlarged spleen)

Other symptoms can include:

- Fatigue
- Night sweats
- Oral sores
- Recurring infections

Aggressive LGLL

This variant presents suddenly, and progresses rapidly. The most common features are:

- Wide range of cytopenias
- Severe B symptoms: Unexplained weight loss >10% of body weight in the previous six months, severe fatigue (unable to work or perform usual activities), fevers >38°C (100.5°F) for at least two weeks without evidence of infection, and drenching night sweats (soaking the bed sheets) for more than a month without evidence of infection
- Hepatosplenomegaly (enlarged liver and spleen)
- Peripheral lymphadenopathy (enlarged lymph nodes around the body)
- Presence of Epstein-Barr virus infection



Diagnosis of LGLL

Unlike B-cell lymphoid cancers, LGLLs are not clearly defined by alterations in the patient's genes, and their classification relies on a combination of changes in the form of the cells and the patient's characteristics.

A definite diagnosis of LGLL is reached when the criteria set out in the revised WHO 2017 classification of haematopoietic and lymphoid cancers are met.

The diagnosis of LGLL, which requires evidence of an increase in T-cell or NK-cell LGLs together with supporting clinical symptoms, is based on the following tests:

Morphology

A peripheral blood smear (examining blood samples under a microscope) will reveal the structure of the cells in the blood.

- Evidence of increased numbers of circulating LGLs greater than $0.5 \times 10^9/L$ (normal range is $0.25 \times 10^9/L$) in peripheral blood usually lasting for more than six months will provide the diagnosis. The increase in numbers of circulating LGLs is important because while LGL cells are easily distinguished,

there is no visual difference between the LGL cancer cells and normal LGL cells. LGLs are large lymphocytes characterised by mononuclear nuclei (meaning 'round nucleus' as opposed to 'multi-lobed nucleus') and azurophilic granules in their cytoplasm which surrounds the nucleus. Azurophilic means the granules are readily seen as blue-purple spots in the cells with a Romanowsky stain.

- Normal LGLs account for 10% to 15% of the peripheral blood mononuclear cells. Most of the normal LGLs in the peripheral blood are NK-cells or T-cell lymphocytes.

Bone marrow aspiration or biopsy can also be used to confirm the diagnosis.

Flow cytometry and immunophenotyping

Immunophenotyping is a process that uses antibodies to identify cells based on the types of antigens or markers on the surface of the cells. This process is used to diagnose specific types of leukaemia and lymphoma by comparing the

cancer cells to normal cells of the immune system. Flow cytometry is a common technique used for immunophenotyping. It can separate different types of blood or bone marrow cells, and then identify and count them. Importantly it can distinguish different types of lymphocytes and determine if the LGL cells are T-cells or NK-cells.

LGLL is typically characterised by distinct immunophenotypic cell populations:

- T-cell cancer LGLs are usually positive for CD3+/CD8+/CD57+/CD16+, which are characteristic of cytotoxic T-cells
- NK-cell cancer LGLL are commonly positive for CD2+/CD16+/CD56+, but negative for CD3.

T-cell clonality

Genetic testing for evidence of T-cell clonality (genetically identical cancer cells) is needed to distinguish clonal cancer T-LGL leukaemia cells from normal T-LGL cells. This test is not useful for NK-LGL leukaemia which can make the diagnosis challenging in this situation.

Treatment options for LGLL

Given the chronic nature of T-cell LGLL, treatment is not always essential. However, the vast majority of patients will eventually need treatment at some point during the course of the disease. Indications for treatment include:

- Severe neutropenia (absolute neutrophil count $<0.5 \times 10^9/L$)
- Moderate neutropenia (absolute neutrophil count $0.5 \times 10^9/L$ to $1 \times 10^9/L$) associated with recurrent infections
- Symptomatic or transfusion-dependent anaemia
- Associated autoimmune conditions requiring therapy

There are no definite treatment regimens established for LGLL so far, because it is such a rare disorder. Treatment recommendations come mainly from experience of treating patients in small retrospective studies and case-series.

Treatment regimens for T-cell LGLL and NK-LGLL are virtually the same, although the more aggressive forms of T-cell LGLL and NK-LGLL tend not to respond

to chemotherapy.

Treatment consists mainly of immunosuppressive therapies. Specific targeted therapies involving inhibitors of the JAK/STAT pathway or cytokine inhibitors are currently being investigated. Cytokines are small proteins secreted by the immune system which are involved in cell signalling.

First-Line Therapy

Immunosuppressive therapies

Immunosuppressive therapies remains the primary treatment option for patients with LGLL. The result of a ground-breaking study, which showed a very good overall response rate with oral low-dose methotrexate, has established this treatment as the best option in patients with LGLL. More recently, a study of patients from a French LGLL registry showed that cyclophosphamide was also an efficient alternative, especially for neutropenic patients and for those who failed a response with methotrexate.

At present, first-line therapy consists of a single immunosuppressive oral agent

such as low dose methotrexate, cyclophosphamide, or cyclosporin A. Treatment should be continued for at least four months prior to evaluating patients' responses.

If you also have anaemia, cyclosporin A may be a better option for first- or second-line therapy, particularly in patients with pure red cell aplasia.

Second-Line Therapy

Chemotherapy and chemotherapy combinations

For patients with LGLL who do not respond to first-line therapies, a number of drugs such as fludarabine, cladribine, and bendamustine have shown promise. However, they have only been tested in a small number of patients.

Combinations of chemotherapies such as cyclophosphamide, doxorubicin, vincristine, and prednisone, or chemotherapy regimens which include cytosine arabinoside, have not been effective in patients with chronic LGLL, and also have severe side effects. However, for patients with aggressive forms of LGLL, these

drugs may be of benefit.

Immunotherapy

In patients with refractory LGLL, immunotherapy (therapy that uses the body's own immune system to fight the cancer) may be helpful. Refractory indicates a cancer that does not result in remission or that gets worse. Alemtuzumab, which is a monoclonal antibody (anti-CD52) immunotherapy drug, can be considered when combined chemotherapies have not resulted in a response. Although Alemtuzumab has been reported to produce a response, the risk of opportunistic infections for patients needs to be considered. An opportunistic infection is an infection that takes advantage of a host with a weakened immune system, or an environment that is beneficial to the infecting organism.

The monoclonal antibody (anti-CD20), rituximab, has been used for patients where LGLL and rheumatoid arthritis co-exist. However, it is not generally considered an adequate treatment for LGLL.

Treatment options for LGLL (cont.)

Splenectomy

A splenectomy (the removal of the spleen) is normally considered in patients with splenomegaly which is causing symptoms, or in patients with severe refractory cytopenias.

Treating an aggressive form of T-cell LGLL

Some patients with T-cell LGLL suffer from a rare aggressive form of the disease which has a very poor prognosis. In aggressive and refractory LGLL, chemotherapy combinations have not shown to be successful, which has suggested the possibility of haematopoietic stem cell transplantation, given its success in other leukaemias.

Treating an aggressive NK-cell LGLL

For patients with aggressive NK-cell LGLL, initial treatment with L asparaginase, either alone or in combination with other chemotherapies, followed by an allogeneic stem cell transplantation is considered to be the best option. L-asparaginase

is an enzyme which breaks down asparagine in the body, an amino acid used by the body to make proteins. Normal cells can make asparagine for themselves, but cancer cells cannot and therefore they cannot survive.

New treatments

Research has identified the involvement of the STAT, JAK and RAS genes in patients with LGLL and the connection of LGLL with autoimmune disorders. Different specific inhibitors of the pathways involved have been tested to discover new treatments.

OPB-31121, a STAT inhibitor, had a significant anticancer effect on human leukaemia cells in the laboratory by inhibiting STAT3 and STAT5. Phase 1 trials with OPB-31121 in human volunteers are underway.

Tofacitinib citrate is a specific JAK3 inhibitor which has shown good results for patients with refractory rheumatoid arthritis. In nine patients with refractory LGLL and rheumatoid arthritis, tofacitinib achieved haematologic

response in six out of nine cases, and improved neutropenia in five out of seven patients.

Tipifarnib, a farnesyltransferase inhibitor, has shown improvement in marrow haematopoiesis in the laboratory, but has not achieved any clinical responses in small trials of patients with myelodysplastic syndromes or case report of a patient with NK-cell LGLL. The exact mechanism of action of this drug is currently unknown but it is recognised that it inhibits activation of the RAS gene.

BNZ-1, a selective inhibitor of the cytokines, interleukin-2 (IL2) and IL15, was shown to safely decrease T regulatory cells and NK-cells while not affecting white blood cells in a recent phase 1 trial. It is currently being trialled in patients with LGLL in a phase I/II trial (NCT03239392).

Prognosis

Given the chronic nature of T-cell LGLL and chronic NK-cell lymphocytosis LGLL, these cancers are associated with a median overall survival of nine

to ten years. Deaths, which occur in less than 10% of patients, are mainly due to severe infections.

The aggressive variant of T-cell LGLL has a poor prognosis even with treatment because of its aggressive nature. Patients with mild symptoms may have a better prognosis than those with severe symptoms at diagnosis. Patients who get an early diagnosis and rapid treatment generally fare better than those with a late diagnosis and delayed treatment.

Aggressive NK-LGLL has a very poor prognosis because patients are shown to not respond to any of the treatments currently available.

Seeing your doctor

Your symptoms

Whatever symptoms you have, make sure you write a list of all of them to share with your doctor as they may be important to your treatment.

Your appointment

Arranging an appointment with your GP will be one of the first things you will need to do when you start to notice symptoms. Pick a time convenient for you that you know you will be able to attend.

Your preparation

It is important to know exactly what you would like to ask your doctor. Make a list of your questions and leave spaces for the answers so you can write them down when you see the doctor. This way you can go into the meeting ready and prepared.

Examples of questions to ask the doctor:

- What tests will I need to have?
- What will the tests show?
- How long will it take to get the

results back?

- How common is my condition?
- What sort of treatment will I need?
- How long will my treatment last?
- How will I know if my treatment has worked?
- What will the side effects be?
- Are there any food or medications I need to avoid?
- Will I be able to go back to work?
- Where can I get help with claiming benefits and grants?
- Where can I get help dealing with my feelings?

Talking to your doctor

Be honest with your doctor; there is no need to feel embarrassed about anything. If you saw your healthcare team before seeing your doctor, be sure to share with your doctor everything your healthcare team told you about your condition, the blood tests you had performed, and the next steps in your LGLL journey. Ask also

if you will receive any intensive treatment or palliative care.

Your support

If it helps, take a family member or friend in with you for support. Some people take a pen and paper in to make notes, and repeat back to their doctor everything they have been told to ensure that they are both on the same page and that nothing has been missed or forgotten.

The next steps

Always ensure that you leave the GP surgery, or the hospital, having shared everything you know about your condition, with all of your questions answered, and knowing exactly what the next steps are, whether it is more tests, further treatment or palliative care. You can ask for a summary letter of the consultation to have everything in writing. Your doctor will generally send a letter like this to your GP.

Furthermore, be sure to access all of the other support available to you as this may be able to help

you with your feelings towards your diagnosis and treatment.

Telling your family

Planning who to tell

Telling your family and friends that you have been diagnosed with LGLL can be difficult.

You may want to create a list of people you want to tell, starting with close family and friends, and then extending it beyond, from your colleagues at work to friends in your neighbourhood.

Planning what to say

It is important to know what you want to say and exactly how much you want people to know. Being clear in your mind about that before speaking to anyone will make this a much smoother experience. Know your story that you want to tell, the diagnosis, the prognosis, the next treatment steps, and what you expect to be going through physically and emotionally. Be sure to speak to people in an environment where both of you can hear each other clearly and where there are likely to be no interruptions.

How to say it

Using a conciliatory tone will

help keep both yourself and the other person calm. Deliver what you have to say slowly, calmly, concisely, and sentence by sentence to allow the other person time to take in the information. Be sincere, and hold their hands if you need to.

You can use the following sentences to help you articulate what you need to say:

- "This is going to be difficult, but I need to tell you something."
- "I've had some bad news but there's a good chance that everything will be okay after I've had treatment."
- "You know I've been feeling unwell for a while. I've had some tests and they've found out what's wrong."

How to respond

Naturally, people will feel sad and concerned for you. Everyone deals with this type of news in their own way, from shock and silence, to questions and support. Invariably, people respond positively, which in turn means you will respond back positively.

Accepting help

Sometimes people feel guilty for getting cancer, that they weren't strong enough, and that they will be a burden on those around them. This is where your loved ones come in, so make sure you do ask for and accept offers to help and support you. Do not try to cope on your own. If they offer to help, tell them that you will get in touch when you need them.

Repeating yourself to different people can become burdensome. Your network of family and friends can help you out by telling those beyond them about your current situation.

You can receive help from us on how to deal with your family and friends during your blood cancer journey. You can visit www.leukaemiacare.org.uk, or call **08088 010 444**, to find out more.

Managing your emotions

Being told that you have cancer may be difficult for you to deal with.

Indeed, you may have a positive demeanour, which will obviously be helpful to you during the next steps in the management of your condition. However, you may experience a range of emotions, including uncertainty, isolation, anxiety, anger, sadness and depression. Understanding each emotion and developing ways that help you deal with them will help you move forward with your life.

Uncertainty

You may think "What happens next?" and you may be unsure about your health and what the future holds for you. You may or may not have had meetings with your healthcare team to discuss the next steps. Once you have a clear path set out in front of you, you will be able to develop a clearer picture of where you are headed. Gaining a sensible balance between being vigilant about your symptoms and carrying on with your life will help ease any anxieties. Help,

care, kindness and support will be available to you from your healthcare team, and you will have access to counsellors and therapists when you need it.

Isolation

If you have received a diagnosis of LGLL, you may feel alone.

Alternatively, you may feel dealing with your cancer allows you to be around those closest to you. Being around those closest to you, such as your family and friends, can be positive and negative.

Let them know what you do and don't want to do, how you do and don't wish to be treated, and what you do and don't feel comfortable talking about. Sometimes, it is difficult for your family, friends and colleagues to understand what you are feeling and going through. Being clear will help create the kind of positive, supportive, and caring environment that will help as you move forward with your life.

Anxiety

Being fearful of the unknown, especially when we are feeling

threatened, is natural. You may experience an increased heart rate, rapid breathing, and muscle tension. These things help us to face a danger or run away. These changes in you are part of the 'fight or flight' response. Any feeling of discomfort, pain or even another disappointment with your healthcare team may elicit such responses, and give you sleepless nights or feelings of worry. This is completely natural.

Such reflexes and responses will ease over time with the building of daily routines and planning things for the future, which will help you to cope with the physical effects of anxiety. Cognitive behavioural therapy can help you deal with your worrying thoughts.

Anger

Feeling angry after your diagnosis is natural and normal. You may be angry with yourself, your body, with the healthcare team or with family and friends. You may display your anger as impatience, irritability and frustration with people and things that would not normally bother you.

Understanding exactly what is making you angry will help you deal with your feelings effectively. In addition, setting yourself achievable goals that stretch you will help reduce the anger and impatience you feel, especially with each passing success. Don't forget to congratulate yourself for each successfully completed task, however small.

Physical exercise is a great way to release your anger and frustrations, and channel your energy positively with no negative impact on your body. Talking about your feelings, and letting them out, will also help stop you lashing out at people and keep you calm.

Sadness and depression

You may feel a sense of loss of the person you used to be, and how safe you felt. You may also feel that your illness is a heavy burden on those around you. You might be feeling low, which is a natural effect of your illness, treatment and recovery. However, if this low mood persists for more

Managing emotions (cont.)

than several weeks, and you feel hopeless, and lose interest and pleasure with things in life, then you may have depression.

Your first steps should be to speak to your loved ones around you about your mood and state of mind, and then contact your GP. You may lift the way you feel by engaging in activities that you were enjoying before diagnosis, to connect back with your life. Only do as much as you can and try and talk about your thoughts and feelings. This will help lighten your burden and put things into perspective. If you have made any acquaintances or friends in the same position as you, talk to them over coffee as they will understand what you are facing.

Self-confidence

Being forced to adjust from your daily routine during the visits to the hospital for treatment can take its toll. This interruption of your life, along with your lack of energy because of your condition and the effects of your treatment, can impact on how you feel about your appearance and how you

feel emotionally. In turn, this can knock your self-confidence and self-esteem. Your feelings of relief, hope and optimism have just been replaced with their polar opposites.

You can gradually build your self-confidence and self-esteem back up by engaging in the activities you did before your diagnosis, and socialising with family, friends, and fellow patients. This will help create a supportive atmosphere to get you back to your old self.

Mindfulness and relaxation

Simple practices from mindfulness and relaxation techniques can help you calm the mind, release tension and ease any pain in your muscles:

- Put yourself in a relaxing environment, sat or lying down comfortably.
- Loosen your clothing so you can move more freely.
- Calmly breathe in through your nose, and out through your mouth, developing a steady

natural rhythm, focusing on your chest and abdomen as you do so.

- Visualise that you are inhaling positivity and exhaling negativity.

By taking some time out of your day to do these exercises, you can help quieten your mind and remove the stress of coming to terms with your diagnosis, so you feel more calm and relaxed.

Survivorship

Someone who is living with or is beyond a cancer diagnosis can be considered a cancer survivor.

Survivorship can be defined as:

"...cover[ing] the physical, psychosocial and economic issues of cancer, from diagnosis until the end of life. It focuses on the health and life of a person with cancer beyond the diagnosis and treatment phases. Survivorship includes issues related to the ability to get health care and follow-up treatment, late effects of treatment, secondary cancers and quality of life. Family members, friends and caregivers are also part of the survivorship experience."

When living with cancer, you will face new challenges to cope with from physical to psychological and social ones. Survivorship aims to provide personalised care based on your need to improve your health, wellbeing, quality of life, and your confidence and motivation, to help you manage. Survivorship also focuses on your health and life with cancer after the end of treatment until the end

of life. At this point, your routine of meeting frequently with your healthcare professionals also ends, so you may feel a mixture of emotions from relief to fear, anxiety and uncertainty about the future. You may wonder how you will slot back into your life after coming through the treatment period.

Your survivorship pathway began at the point when you were diagnosed with LGLL. By this point, you will have been starting to receive support for work, finance, and personal relationships, through to managing pain, fatigue and making positive lifestyle changes, such as starting a healthy diet and gentle exercising.

Your individual needs as a patient will be identified and addressed, including:

- Dealing with the emotional impact of receiving a LGLL diagnosis which may have created feelings of uncertainty, fears of recurrence and difficulties in planning for the future. These will be discussed with you to develop your

individualised care plan with support from social care staff and therapists, as you need it.

- Improving your quality of life through efficient and co-ordinated care during your treatment, with effective communication within the treatment team, and a positive attitude.
- Taking care of any comorbidities – that is, other medical conditions and diseases – and offering you cancer rehabilitation based on your clinical needs as assessed by informed professionals, and ensuring compliance with the National Cancer Rehabilitation Pathways and Rehabilitation Peer Review requirements.
- Providing you with a treatment summary from the diagnosis of your condition to the end of your treatment. This would include any ongoing medication and note possible symptoms that may occur in the future. You would also be provided details of who to contact in addition to your GP for any concerns you may have.
- Preparing you fully for the impact of your LGLL and treatment, the physical and physiological side effects of treatments and the psychological impact of LGLL in general. You will be provided physical equipment, and taught about various coping strategies to adapt to your new situation.
- Supporting you with advice for social and financial difficulties, including caring responsibilities, your inability to participate in social activities, any debt and financial worries from not being able to work, and perhaps the need to return to work before you feel ready.
- Receiving health and nutrition advice from a nutritionist on following a healthy and balanced diet to help improve your general health and wellbeing. The World Cancer Research Fund published a report for cancer survivors which suggests that even small dietary and lifestyle changes can produce large health benefits.

Palliative care

Palliative care in LGLL

If you have an aggressive form of LGLL and your treatment has not worked, your healthcare team may feel that further treatment will not help and you will be offered palliative care. Palliative care, also known as supportive care, involves a holistic or "whole person" approach, which includes the management of your pain and symptoms as well as psychological, social and spiritual support for you and your loved ones.

Rather than looking for a cure using intensive treatment, palliative care aims to reduce your symptoms, control your LGLL, extend your survival, and give you and your loved ones the best quality of life possible. Your doctor will discuss the options with you in detail before you decide the next steps.

Who provides palliative care?

Your palliative care will be provided by a team of health and social care professionals trained

in palliative medicine who will coordinate your care.

These professionals can include your GP, hospital doctors and nurses, community nurses, hospice staff and counsellors, social care staff, physiotherapists, occupational therapists, complementary therapists, and religious leaders, if you would like this. Your palliative care services may be provided by the NHS, local council or a charity. You may receive day-to-day care at your home and at the hospital.

What is the clinical course?

You will have a number of treatments, and be prone to frequent infections because of the LGLL and the impact of your treatments. Your therapy may continue because of potential remission and/or useful palliation.

You may experience various pains and other clinical complications such as:

- **Bone pain:** Radiotherapy and/

or oral steroids, and sometimes non-steroidal anti inflammatory drugs (NSAIDs) may be used, although these are used with caution because they can interfere with your immune system and kidney function.

- **Bone marrow failure:** Blood and platelet transfusions are provided to prevent and fight recurrent infections and bleeding episodes.
- **Oral problems:** Analgesic mouth washes and topical ointments may help with ulceration. Chewing gum and mouth washes have been shown to help with dry mouth, tooth decay and oral thrush.
- **Night sweats and fever:** These can place a heavy burden on carers because of so many changes of night clothes and bedding.
- **Pathological fractures:** Orthopaedic intervention and subsequent radiotherapy, with consideration given to prophylactic pinning of long bones and/or radiotherapy

to prevent fracture will be performed. This will reduce the likelihood of complex pain syndromes developing.

- **Spinal cord compression:** Immediate high single daily dose oral steroids will be given.
- **Back pain from wedge and crush fractures of the vertebrae of the spinal column:** Treatments can include analgesics, antidepressants and/or anticonvulsant medication used in tandem with opioids.
- **Hypercalcaemia:** Treatment is usually with intravenous hydration and intravenous bisphosphonates.
- **Loss of appetite:** Low-dose steroids may temporarily boost the appetite, while small, frequent and appetising meals and supplement drinks will also help.

End of life care

When does end of life care begin?

If your treatment hasn't worked and you are going through palliative care, you will be offered end of life care. End of life care begins when you need it and may last a few days, months or years.

What does end of life care involve?

End of life care is support for people who are in the last few months or years of their life. The aim is to help you enjoy a good quality of life until you die, and to die with dignity. The professionals looking after you will ask you about your wishes and preferences on how to be cared for and put these into action. They will also provide support to your family, carers and loved ones. You will be able to decide where you will receive end of life care, be it at home or in a care home, hospice or hospital. The same will be true of where you would like to die. Wherever you are, you will receive high quality end of life care.

Who provides end of life care?

A team of health and social care professionals may be involved in your end of life care, including hospital doctors and nurses, your GP, community nurses, hospice staff and counsellors, social care staff, physiotherapists, occupational therapists or complementary therapists, and religious leaders, if you would like this. If you are being cared for at home or in a care home, your GP will have overall responsibility for your care with the support from community nurses, along with your family and friends.

What choices do I have in terms of end of life care?

Deciding where you want to die can be a difficult choice to make. Working out what you and your loved ones want, together with seeing what services are available to you, can help to make the decision a little easier.

- **Staying at home:** A place of

familiarity, surrounded by your loved ones, may be something that you will find reassuring. External care professionals will be able to visit you at home to make sure your symptoms are looked after.

- **Hospices:** These are specialised in looking after those with life-limiting illnesses and those who are coming to the end of their life. Hospices are staffed with care professionals who are able to keep an eye on you, make sure that your symptoms are controlled and offer you a number of services to make your stay as comfortable as possible. For more information on the care that they can provide, go to <https://www.hospiceuk.org/>
- **Residential care/nursing homes:** If you think that your stay may be a few months or more, then a nursing home may be more suitable than a hospice. These can be private or run by a charity or the local council so be sure to check if there are any fees.
- **Hospitals:** Although you may be used to staying in a hospital

ward, the care routine cannot always be tailored to your specific needs. Pressures on the NHS mean that your stay will only be as long as strictly required. As soon as the condition you were admitted for has been resolved, you will need to go back to your home or nursing home. However, a number of specialists will be available to help look after you for specific problems, and a number of hospitals also have a designated palliative care team for patients who require them.

Whatever your choice, speak with your GP or healthcare team who will be able to help you put everything into place.

Glossary (cont.)

Chemotherapy

A form of cancer treatment that uses one or more anticancer drugs as part of a standardised chemotherapy regime.

Immunophenotyping

A set of tests to indicate the number of leukaemia cells in blood and bone marrow samples.

Large Granular Lymphocytic Leukaemia (LGLL)

The reproduction of cytotoxic cells, which causes neutropenia, anaemia and/or thrombocytopenia. This condition is often associated with autoimmune disorders, especially rheumatoid arthritis, and other lymphoproliferative disorders.

Palliative care

Also known as supportive care, this is a type of care that focusses on improving the quality of life for a patient with a life-threatening illness and their loved ones.

Remission

When all tests indicate the absence of an illness.

Spleen

The largest organ in the lymphatic system. Similar in structure to a large lymph node it acts primarily as a blood filter and defends the body against infection.

Survivorship

A focus on the life of someone with cancer after diagnosis and treatment.

Tell us what you think!

If you would like to give us some feedback about this patient information booklet, please hover over the code to the right using your phone or tablet's camera. Click the link as it appears and this will take you to a short web form to fill in.

Suitable for Android, iPhone 7 and above.



Useful contacts and further support

There are a number of helpful sources to support you during your diagnosis, treatment and beyond, including:

- Your haematologist and healthcare team
- Your family and friends
- Your psychologist (ask your haematologist or CNS for a referral)
- Reliable online sources, such as Leukaemia Care
- Charitable organisations

There are a number of organisations, including ourselves, who provide expert advice and information.

Leukaemia Care

We are a charity dedicated to supporting anyone affected by the diagnosis of any blood cancer.

We provide emotional support through a range of support services including a helpline, patient and carer conferences, support group, informative website, one-to-one buddy service and high-quality patient information. We also have a nurse on our help line for any medical queries relating to your diagnosis.

Helpline: **08088 010 444**
www.leukaemicare.org.uk
support@leukaemicare.org.uk

Bloodwise

Bloodwise is the leading charity into the research of blood cancers. They offer support to patients, their family and friends through patient services.

020 7504 2200
www.bloodwise.org.uk

Cancer Research UK

Cancer Research UK is a leading charity dedicated to cancer research.

0808 800 4040
www.cancerresearchuk.org

Macmillan

Macmillan provides free practical, medical and financial support for people facing cancer.

0808 808 0000
www.macmillan.org.uk

Maggie's Centres

Maggie's offers free practical, emotional and social support to people with cancer and their families and friends.

0300 123 1801
www.maggiescentres.org

Citizens Advice Bureau (CAB)

Offers advice on benefits and financial assistance.

08444 111 444
www.adviceguide.org.uk

Leukaemia Care is a national charity dedicated to providing information, advice and support to anyone affected by a blood cancer.

Around 34,000 new cases of blood cancer are diagnosed in the UK each year. We are here to support you, whether you're a patient, carer or family member.

Want to talk?

Helpline: **08088 010 444**

(free from landlines and all major mobile networks)

Office Line: **01905 755977**

www.leukaemiacare.org.uk

support@leukaemiacare.org.uk

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Registered charity
259483 and SC039207

Leukaemia Care
YOUR Blood Cancer Charity

