Chronic Eosinophilic Leukaemia – Not Otherwise Specified (CEL-NOS)

A Guide for Patients

Leukaemia Care
YOUR Blood Cancer Charity

Introduction

Being diagnosed with chronic eosinophilic leukaemia – not otherwise specified (CEL-NOS) can be a shock, particularly when you may have never heard of it. If you have questions about CEL-NOS – 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. For more personalised information, talk to your haematologist, clinical nurse specialist or hospital pharmacist.

This booklet was written and updated by our Patient Information Writer, Isabelle Leach and peer reviewed by Professor Claire Harrison and Professor Mary Frances McMullin. We are also grateful to Brian Penfold for their contribution as a patient reviewer.

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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In this booklet

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 8:30am – 5:00pm Monday - Friday and 7:00pm – 10:00pm on Thursdays and Fridays. If you need someone to talk to, call **08088 010 444**.

Alternatively, you can send a message via WhatsApp on **07500068065** on weekdays 9:00am – 5:00pm.

Nurse service

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

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.leukaemiacare.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.leukaemiacare.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@leukaemiacare.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.leukaemiacare.org.uk**.

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@leukaemiacare.org.uk**

Patient magazine

Our magazine includes inspirational patient and carer stories as well as informative articles by medical professionals: **www.leukaemiacare.org.uk/communication-preferences/**

What is chronic eosinophilic leukaemia – not otherwise specified (CEL-NOS)?

Chronic eosinophilic leukaemia (CEL) is a rare myeloproliferative neoplasm (MPN). MPNs are chronic disorders where the myeloid stem cells in the bone marrow make too many abnormal red blood cells, white blood cells, or platelets which do not function properly. In the case of CEL, too many eosinophils are being made. Because the increase in the number of eosinophils is unexplained, it is actually known as chronic eosinophilic leukaemia – not otherwise specified (CEL-NOS). In CEL-NOS, there is no evidence of secondary causes for the increase in the number of eosinophils such as parasitic infection, allergy or cancer.

Hyper-eosinophilia is the main feature of CEL-NOS. The normal level of eosinophils in the blood is less than 500 x 10^6/l. If the level is between 500 x 10^6/l and 1500 x 10^6/l, the patient is said to have eosinophilia. If the levels are greater than 1500 x 10^6/l, the patient is said to have hyper-eosinophilia.

CEL-NOS is also characterised by either:

- An increased number of myeloblasts (immature myeloid cells) in the blood and bone marrow, but less than 20% of the white blood cells
- Eosinophils which are clonal (genetically identical) or both

If the eosinophils cannot be demonstrated as being clonal or there is no increase in myeloblasts in the bone marrow, then the diagnosis for these patients with hyper-eosinophilia should be idiopathic hyper-eosinophilic syndrome (iHES) rather than CEL-NOS. The term idiopathic describes any disease for which the cause is unknown.

Who is affected by CEL-NOS?

There is no firm data on the incidence of CEL-NOS due to the rarity of the disorder and the difficulty in distinguishing CEL-NOS from iHES. The current 2016 World Health Organisation guidelines provide guidance on how to distinguish CEL-NOS from

iHES; however, this can still be difficult.

Nevertheless, it has been reported that the incidence rate of all hyper-eosinophilic syndromes (HESs), including CEL-NOS, is said to be around 0.036 per 100,000 persons.

CEL-NOS is usually diagnosed between the ages of 20 and 50, but it has also been described in children and adults older than 60 years of age. It is also more common in men than women (male-to-female ratio of 1.47).

What causes CEL-NOS?

The exact cause of CEL-NOS is unknown. It has not been linked with any specific chromosome abnormality or gene mutation. Rarely, CEL-NOS may be due to a genetic mutation resulting from environmental factors, smoking or chemical/radiation exposure. However, for most patients, no specific cause can be found. Patients with CEL-NOS do not have the Philadelphia chromosome BCR-ABL fusion gene or other genetically defined entities such as PDGFRα (Platelet-derived growth factor receptors-alpha), PDGFRβ (Platelet-derived growth factor receptors-beta), or FGFR1 (Fibroblast growth factor receptor 1) abnormalities.

At Leukaemia Care we have a freephone helpline service available for anyone who has been affected by blood cancer. We can provide emotional and practical support as well as offer medical advice. The telephone number is **08088 010 444.**

What are the symptoms of CEL-NOS?

In around 10% of patients who do not have any signs or symptoms, CEL-NOS is diagnosed by chance during a routine blood test. However, other patients can experience severe symptoms and signs of cardiovascular or neurological complications due to organ damage brought about by the high eosinophil levels.

Patients with CEL-NOS may experience the following symptoms:

- Fever
- Itching
- Diarrhoea
- Night sweats
- Unexplained weight loss
- Unexplained shortness of breath
- Cough
- Swollen lymph nodes (small swellings in the lymphatic system where lymph is filtered, and lymphocytes are formed. They are part of the immune system)
- Muscle pains
- Anaemia (low level of red blood cells and haemoglobin which is carried by the red blood cells)
- Decreased level of platelets, which are small blood cells that help the body form clots to stop bleeding
- Mucosal ulceration
- Fibrosing of the inside lining of the heart
- Enlarged spleen

The most severe cases of CEL-NOS are mainly due to either tissue damage, particularly thickening and scarring of the heart, or transformation to acute myeloid leukaemia (AML) in patients who have a high level of myeloblast cells.

How is CEL-NOS diagnosed?

In patients with hyper-eosinophilia, the first step is to exclude any secondary causes of eosinophilia, such as allergies, infections, medications, auto-immune disorders and/or cancers.

Diagnostic tests

Blood and bone marrow samples are examined to reach a diagnosis of CEL-NOS according to the 2016 WHO classification criteria. The following diagnostic tests are required:

- **Blood sample:** Blood is obtained to measure the complete blood cell count (number and quality of white blood cells, red blood cells and platelets), as well as white blood cell differentiation to show which white blood cells are increased.
- **Bone marrow biopsy:** A sample of bone marrow is taken and the cells from the blood and bone marrow are examined under the microscope by specialists.
- **Chromosome and gene analysis:** These are used to exclude bone marrow and blood disorders which also have hyper-eosinophilia as a feature.

Investigating a patient with hyper-eosinophilia is a priority, as it can speed up the potential diagnosis of CEL-NOS. This enables early initiation of treatment before any tissue damage becomes established, particularly heart fibrosis.

To determine if any damage to the patient's organs has occurred, the following tests may be carried out:

- Blood chemistry tests to check that the liver, kidneys and spleen are working properly.
- Other tests to look out for changes in cardiovascular and pulmonary (heart and lung) systems. These include:
  - Chest X-rays
  - Echocardiography (ultrasound to create an image of the heart)
  - Pulmonary function test (a series of tests to determine the severity of pulmonary impairment)
  - Cardiac troponin T test (troponin T is a protein

How is CEL-NOS diagnosed? (cont.)

found in the cardiac muscles, which is released into the bloodstream when the heart is damaged)

2016 World Health Organisation diagnosis of CEL-NOS

Because patients with CEL-NOS and iHES both have hyper-eosinophilia and associated organ damage, the 2016 World Health Organisation (WHO) stipulates the following characteristics should be present for a diagnosis of CEL-NOS:

- Eosinophil count is greater than 1500 x 10^6/l in the peripheral blood
- The patient does not meet any of the WHO criteria for the following illnesses:
  - Chronic myeloid leukaemia (CML) with presence of BCR-ABL1 gene
  - Atypical BCR-ABL1-negative CML
  - Polycythaemia vera (excess of red blood cells)
  - Essential thrombocythaemia (excess of platelets with abnormal blood clotting)
  - Chronic neutrophilic leukaemia (excess of neutrophil white blood cells)
  - Chronic myelomonocytic leukaemia (excess of monocyte white blood cells)
  - Primary myelofibrosis (build-up of scar tissue in the bone marrow)
- No mutations in any of the following genes: PDGFRα, PDGFRβ and FGFR1 (Fibroblast growth factor receptor 1) and no PCM1 JAK2, ETV6-JAK2 or BCR-JAK2 fusion genes
- Myeloblast cells in the peripheral blood and the bone marrow make up less than 20% of white blood cells, and there are no diagnostic features of AML, including the chromosome mutations inv(16) (p13.1q22) and t(16;16)(p13;q22).
- There is a clonal component to chromosome or gene

abnormality or myeloblast cells are more than 2% in the peripheral blood or more than 5% in the bone marrow blood

FIP1L1 PDGFRα gene

Recently, it was discovered that patients with iHES have an abnormality of chromosome 4, which causes the fusion of the FIP1L1 gene (Factor interacting with Papola and CPSF1) to the PDGFRα gene, resulting in the FIP1L1 PDGFRα fusion gene rearrangement. These patients with iHES and the FIP1L1 PDGFRα fusion gene have been reclassified as having CEL-NOS because the FIP1L1 PDGFRα gene is considered a strong indicator of a clonal disease, a feature of CEL-NOS.

More importantly, patients with the FIP1L1-PDGFRα gene are known to respond extremely well to the tyrosine kinase inhibitor imatinib, which prevents the actions of the mutated BCR-ABL, c-KIT and PDGFRα genes. Tyrosine kinases are important components of the signalling and role determination in cell growth and cell differentiation.

Differential diagnosis with iHES

The diagnosis of CEL-NOS is based on the presence of a clonal increase of eosinophilic cells and the exclusion of other bone marrow cancers and blood disorders in which hyper-eosinophilia is a feature. This will differentiate it from iHES.

iHES is defined as unexplained hyper-eosinophilia for at least six months with evidence of organ dysfunction directly attributable to the hyper-eosinophilia itself. On occasion, it is not possible to confirm clonality, in which case a diagnosis of iHES is made, providing there is no increase of myeloblast cells in the blood (more than 2%) or bone marrow (more than 5%).

In addition, transformation of some patients with iHES to AML also provides evidence that the disorder was likely from the start to be a clonal CEL-NOS.

What is the treatment for CEL-NOS?

The foundation of treatment of CEL-NOS is interferon, hydroxycarbamide and tyrosine kinase inhibitors such as imatinib.

Because CEL-NOS is so rare, there is no standard treatment. The course of CEL-NOS is different depending on the patient's circumstances. Additionally, in some patients, CEL-NOS can be stable for many years and then transform into AML. Your haematologist will create a treatment plan suitable for you.

Chemotherapy

Chemotherapy involves using drugs to prevent cancer cells from growing and dividing, leading to the cancer cells being destroyed over time. Targeted chemotherapy is a chemotherapy treatment that targets the specific genes or proteins of leukaemia cells.

Patients with FIP1L1-PDGFRα gene or PDGFRα gene mutations

The targeted chemotherapy, imatinib, is a tyrosine kinase inhibitor which prevents the kinase activity of the FIP1L1-PDGFRα gene and stops the production of the abnormal eosinophils. CEL-NOS patients who have the FIP1L1-PDGFRα gene respond very well to treatment with low-dose imatinib. In selected cases, where no response is seen, the dose of imatinib may be increased.

Case reports of patients with PDGFRα gene mutations, or variants of PDGFRα other than FIP1L1-PDGFRα, have shown that imatinib can produce durable remissions in these patients. Patients with a PDGFRα mutation who received imatinib for a median duration of 6.6 years had a response rate of 96%, and their ten-year overall rate of survival was 90%. All the patients who achieved remission continued to respond to imatinib, and none of the patients showed any disease progression.

Chemotherapy options for other patients

It is estimated that approximately 10% to 20% of patients with CEL NOS have the FIP1L1-PDGFRα

gene. The goal of therapy in these patients is to prevent organ damage caused by the eosinophilia.

Hydroxycarbamide

Although not a cure, hydroxycarbamide is an effective chemotherapy for controlling hyper-eosinophilia. It can also be used in combination with steroids to improve the response rate.

Chemotherapy drugs such as vincristine, chlorambucil, cyclophosphamide, etoposide, cyclosporine, and 2-chlorodeoxyadenosine can be used as second-line drugs, if hydroxycarbamide chemotherapy is not effective.

High dose imatinib

If the therapies above have not produced any results, higher doses of imatinib for patients without the mutated PDGFRα and PDGFRβ genes may eventually produce a response, if only partial. For many patients, even if they do not have these mutations, imatinib can improve blood counts and symptoms for many years, if the drug is taken on a regular basis.

Immunotherapy

Interferon-alpha (IFN-α) is a drug of purified derivative fractions of white blood cells. This immunotherapy helps boost the body's natural immune system to fight the leukaemia. The use of IFN-α in CEL-NOS is partly guided by its known efficacy in CML, polycythaemia vera and essential thrombocythaemia.

IFN-α is often used in CEL-NOS patients who do not respond to other therapies including steroids (prednisone) and hydroxycarbamide. It has shown reductions in white cell counts and reversed organ injury in patients with CEL-NOS.

Stem cell transplant

Allogeneic stem cell transplants, where blood-forming stem cells are donated from a genetically similar donor, are used in patients with aggressive CEL-NOS. While a stem cell transplant may be a treatment option for some patients, most people with CEL-

What is the treatment for CEL-NOS? (cont.)

NOS are older, so they may not be able to benefit from a stem cell transplant.

Survival following a stem cell transplant ranges from eight months to five years. Although success has been described in several cases, the role of stem cell transplants as a treatment for CEL-NOS is not well recognised.

Supportive care

Supportive or palliative care is medical care that relieves symptoms without dealing with the cause of the condition. Examples of supportive care for CEL-NOS include:

- **Leukapheresis** - Leukapheresis is a procedure in which the excess white blood cells are separated out from the blood to help reduce the large numbers of eosinophils and reduce the thickening of the blood. This is achieved using an electrophoresis machine. However, it does not represent an effective maintenance therapy.
- **Blood-thinners** - Anticoagulants (which thin the blood) and anti-platelet agents (which prevent blood clotting) help patients avoid getting clots and embolisms caused by their eosinophilia.
- **Splenectomy** - A splenectomy is an operation to remove the spleen, which makes lymphocyte white blood cells, and it may be recommended for some patients. As the spleen becomes enlarged with the high number of eosinophils, it can cause patients severe abdominal pain. Splenectomy is not a standard treatment, but it can be a part of a palliative care treatment plan. A surgical oncologist is a doctor specialising in cancer surgery and will usually perform this procedure.
- **Cardiac surgery** - Cardiac surgery may prolong survival in patients with cardiac heart disease. Heart valve replacements or surgery on scarred heart muscle can help recover heart function.

Future therapies

Interleukin-5 (IL-5) is an immune substance which is vital for eosinophils to mature and proliferate. Antibodies against IL-5 (mepolizumab) and the IL-5 receptor (benralizumab) have been developed with the aim of decreasing eosinophil levels. Both types of antibodies prevent IL-5's action. Mepolizumab is successful for the treatment of patients with severe eosinophilic asthma, but is still in the early research stages for iHES.

An FGFR1-inhibitor (pemigatinib) is currently being evaluated in patients with myeloid or lymphoid cancers with encouraging results. A clinical trial that was started in 2016 is estimated to be completed in December 2020 (trial identifier NCT03011372 at **https://clinicaltrials.gov/**).

Ruxolitinib is a JAK1/JAK2 inhibitor whose efficacy for decreasing eosinophil levels counts in patients with hyper-eosinophilia, including those with CEL-NOS, is being evaluated in a clinical trial. The trial is currently enrolling patients and is estimated to be completed by June 2024 (trial identifier NCT03801434 at **https://clinicaltrials.gov/**).

Follow-up

Follow-up after treatment is an important part of cancer care. Follow-up for chronic eosinophilic leukaemia is often shared among the cancer specialists (oncologists) or blood specialists (haematologists) and your GP. Your healthcare team will work with you to decide on follow-up care to meet your needs.

What is the prognosis of CEL-NOS?

Risk factors which will influence the prognosis of patients with CEL-NOS are evaluated by experienced haematologists. Your haematologist is the best person to advise you on your prognosis, based on your individual circumstances.

Determining what severity and duration of hyper-eosinophilia will cause tissue damage, and in

What is the treatment for CEL-NOS? (cont.)

which patients, is challenging. Moreover, because CEL-NOS is very rare, there is no evidence from clinical trials on the level of hyper-eosinophilia at which treatment should be started in the patients who do not have tissue or organ disease.

Evidence from case reports suggests that CEL-NOS usually progresses slowly, remaining the same for many years. However, in some patients, it may change quickly or transform into AML.

In general, the outcome of patients with CEL-NOS as defined by the WHO criteria is modest. CEL-NOS is often unresponsive to conventional treatment and has a risk of transformation to AML. A series of 10 case reports of patients with CEL-NOS showed a median survival of 22.2 months (range: 2.2–186.2). In a study which included 17 CEL-NOS patients and 51 iHES patients, CEL-NOS patients showed disease-specific survival of 14.4 months ranging from 1.0 to 120.1 months. Disease-specific survival is where deaths from other causes are not included.

Given the rarity of CEL-NOS and the difficulties distinguishing it from iHES, prognoses, in terms of survival and transformation rates to AML, must be viewed with caution.

Living with CEL-NOS

After a diagnosis of CEL-NOS, you may find that it affects you both physically and emotionally. This chapter will talk about both of these aspects.

Emotional impact of CEL-NOS

Being told you have cancer can be very upsetting. Some of the symptoms of CEL-NOS can be hard to cope with and, because of this, you may need emotional, as well as practical, support. Being diagnosed with a rare disease can affect the whole of you, not just your body, and can impact you emotionally at any point of your journey. It is likely that you will experience a range of complex thoughts and emotions, some of which may feel strange or unfamiliar to you. It is important to know that these feelings are all valid and a normal response to your illness.

Looking after you

Following a diagnosis of CEL-NOS, you may want to make changes to your lifestyle to try to stay as well as possible after your diagnosis and during treatment. Don't try to change too much at once. Adopting a healthy way of living is about making small, manageable changes to your lifestyle.

A healthy lifestyle includes having a well-balanced diet and being physically active. With some of the side effects you may be experiencing, the idea of getting out and being active may be the last thing you want to do, but it is important to try and stay as active as possible to make you feel better and reduce some of the symptoms or side effects you may be experiencing.

One of the most commonly reported side effects of the treatment of CEL-NOS is fatigue. This isn't normal tiredness and doesn't improve with sleep.

Some general tips on how to deal with fatigue include:

- Have a regular lifestyle – try going to bed and waking up at approximately the same time every day and try to avoid lying in.
- Take part in regular, gentle

Living with CEL-NOS (cont.)

exercise to maintain your fitness levels as much as possible.

- Reserve your energy for what you find important and build rest periods around those times.
- Before going to bed avoid stimulants such as alcohol, coffee, tea or chocolate, or using laptops, tablets or mobile phones.
- Keep your bedroom quiet and at a comfortable temperature.
- Talk about your worries with family, friends or your doctor or nurse, or patient support groups.
- Discuss your fatigue with your doctor or nurse.

You can find more information about living well with leukaemia on our website at: **www.leukaemiacare.org.uk.**

Practical support

Work and finances

Being diagnosed with CEL-NOS can sometimes lead to difficulties relating to your work life. Your diagnosis may lead to temporary sick leave or a reduction in working hours, but it can also mean that you have to stop work altogether. You may need to make an arrangement with your employer for times when you may need to go into hospital or for those times when you may not be well enough to go into work.

Your consultant or your GP can arrange letters to confirm your diagnosis and the effects it may have on your work life to your employer. It is often worth taking time to explain CEL-NOS to your employer, as it is likely they will never have heard of the disease.

It is important for you to know that people with any form of cancer are covered by law by the Equality Act. This means that legally your employer cannot discriminate against you and must make reasonable

arrangements and adjustments relating to your disease.

If you would like advice about some of the financial help available to you, then you can speak to our Patient Advocacy team on **08088 010 444**. Alternatively, Macmillan has published a booklet about financial support following a diagnosis of cancer that might be useful to you. They can also give you personal advice over the phone via their helpline at **0808 808 0000** and you can discuss which benefits you are eligible for. Some Macmillan centres can arrange face-to-face meetings with a benefits advisor. They can also provide financial assistance in the form of grants – ask your nurse in the hospital how to apply.

As CEL-NOS is a cancer, you will also be entitled to apply for a medical exemption certificate which means that you are entitled to free NHS prescriptions. Your GP or specialist nurse at the hospital can provide you with the details of how to apply for this. If you are undergoing chemotherapy you may qualify for a Blue Badge to help with hospital car parking.

To apply for a badge, contact your local council. You can find more information on practical issues, including a toolkit on applying for a Blue Badge, on our website: **www.leukaemiacare.org.uk.**

Talking about CEL-NOS

Talking to your haematologist

CEL-NOS is a rare condition. It is important for you to develop a good working relationship with your haematologist, so you are given the best treatment possible for you.

The following gives advice on working well with your haematologist:

- If it's an initial consultation, take along a list of your current medications and doses, and a list of any allergies you may have.
- If you have a complicated medical history, take a list of diagnoses, previous procedures and/or complications.
- Make a list of questions to take to your appointment. This will help the discussion with your haematologist.
- It can be useful to repeat back what you have heard so that you can be sure that you fully understood.
- Note information down to help you remember what was said.
- Be open when you discuss your symptoms and how you are coping. Good patient-doctor communication tends to improve outcomes for patients.
- Bring someone else along to your appointment – they can provide support, ask questions and take notes.
- Don't be afraid to ask for a second opinion – most haematologists are happy for you to ask.

You need to tell your haematologist if...

You're having any medical treatment or taking any products such as prescribed medicines, over the counter treatments or vitamins. It is important to understand that treatments, including complementary therapies, which are perfectly safe for most people, may not be safe if you are being treated for CEL-NOS.

Remember, if you choose to start any form of complementary

therapy outside of your medical treatment, discuss this with your haematology consultant or clinical nurse specialist, prior to beginning it. It is important to understand the difference between complementary therapies, used alongside standard treatment, and alternative therapies, used instead of standard treatment. There is no evidence that any form of alternative therapy can treat CEL-NOS.

Talking to other people

Telling people you have a rare condition like CEL-NOS can be hard to explain. You might find it useful to let your close family and friends, as well as your employer know about your health condition. It might be easier to provide people with basic information and give them information leaflets or booklets like this one about CEL-NOS if they want to know more in-depth details.

It is probably best to focus conversations on the symptoms that you are experiencing, how the condition affects you and how you feel about it. Often people misunderstand and, unfortunately, it will mostly fall to you to educate them as best as you can. Where possible, it's advisable to let people know what you find helpful and unhelpful, in terms of what others say and do. Often people make assumptions and do what they think helps. For example, saying you look well, recounting stories of others they know with a similar diagnosis, encouraging you to look ahead and stay positive isn't always what people really want to hear. In many ways, the more you communicate with them the better.

These points may help you:

- Explain that you have a condition that means your bone marrow does not function properly, and that this affects the number of blood cells it produces
- Explain your symptoms (maybe you are tired, or have a lot of pain)
- Explain what you need (maybe

Talking about CEL-NOS (cont.)

more help day-to-day, or someone to talk to)

You could also consider the following when telling people about your diagnosis:

- **Find out more** - Try to find out as much as you can about your condition, from reliable internet sources, charitable organisations or your consultant haematologist. The more you know, the more you can share.
- **Have a print-out to hand** - It may help to have some information to hand to share with family and friends. This will take the pressure off you having to remember everything they may want to know.
- **Explain your needs** - Try and be clear about what your needs may be. Perhaps you need help with the weekly food shop, help with cooking dinner, or someone to drive you to and from appointments. You may find that friends and family are pleased that they can do something to help you.
- **Be open about how you feel** - Don't be afraid of opening up about how you feel, as people who care will want to help you as best they can. Talk as and when you feel comfortable, so those around you will know when you need them most.

Glossary

Acute Myeloid Leukaemia (AML)

A rapid and aggressive cancer of the myeloid cells in the bone marrow.

Anaemia

A condition where the number of red blood cells are reduced. Red blood cells contain haemoglobin and transport oxygen to body cells. This may be due to a lack of iron, leukaemia, or sickle cell disease.

Blast Cells (blasts)

Immature cells found in the bone marrow which are not fully developed. Up to 5% of the cells found in the bone marrow are blast cells. Patients with leukaemia have a much higher number of immature, abnormal cells called blasts cells.

Blood Cancer

Cancer of the blood cells from the bone marrow or lymphatic system. There are three main types of blood cancer:

- Leukaemia begins in the bone marrow and is classified according to the type of blood cell it affects (either myeloid or lymphoid) and whether it grows quickly (acute) or slowly (chronic).
- Lymphoma starts in the lymphocyte white blood cells within the lymphatic system.
- Myeloma is a cancer of the plasma cells and starts in the bone marrow. Plasma cells are a type of white blood cell that makes antibodies.

Bone Marrow

The soft blood-forming tissue that fills the cavities of bones and contains fat, immature and mature blood cells, including white blood cells, red blood cells and platelets.

Bone Marrow Aspirate

Bone marrow aspirates consist of taking a sample of the liquid part of the soft tissue bone marrow inside your bones using a syringe. They are crucial to establish a diagnosis of leukaemia and may be performed at stages during treatment to monitor progress.

Bone Marrow Biopsy

A bone marrow biopsy involves the collection of a sample of bone marrow from the hip bone, generally under local anaesthesia. A bone marrow surgical instrument with a cylindrical blade, called trephine, is used to remove a one or two-centimetre core of bone marrow in one piece.

Glossary (cont.)

Chemotherapy

Drugs that work in different ways to stop the growth of cancer cells, either by killing the cells or by stopping them from dividing.

Chromosomes

Thread-like structures which carry the genes, and are located in the nuclei of every cell in the body. There are 46 chromosomes (23 pairs) in humans.

Chronic Leukaemia

Leukaemias which progress slowly and are less aggressive than acute leukaemia. There are two main types: chronic lymphocytic leukaemia and chronic myeloid leukaemia.

Chronic Lymphocytic Leukaemia (CLL)

A leukaemia in which the B-lymphocytes (B-cells) in the bone marrow start multiplying excessively leading to large numbers of small, mature lymphocyte cells, which are unable to fight infection, and their presence prevents the bone marrow from producing healthy blood cells of all types.

Chronic Myeloid Leukaemia (CML)

A leukaemia in which the myeloid cells start multiplying in the bone marrow leading to large numbers of abnormal, immature myeloid cells called blasts, which prevent the bone marrow from producing enough healthy blood cells of all types.

Clinical Trial

A trial designed and planned to determine a specific answer or aim; for example, whether treatment A is better than treatment B. The study will be conducted in patients who meet particular inclusion criteria, and the results are collected and analysed to provide an answer.

Clonal

This refers to an organism descended from, and genetically identical to, a single common ancestor.

DNA (deoxyribonucleic acid)

Thread-like chain of amino acids found in the nucleus of each cell in the body which carries genetic instructions used in the growth, development and functioning of the individual's cells.

Eosinophil

A type of white blood cell which has a protective immunity role against parasites and allergens.

Genes

Genes are made up of DNA which stores the genetic information required to make human proteins.

Granulocytes

A group of white blood cells, which have granular bodies in their cytoplasm. They include the neutrophils, eosinophils and basophils white blood cells, all of which protect the body from bacteria, allergens and inflammation. For more information, see White blood cells in this glossary.

Haemoglobin

A red protein contained within the red blood cells and responsible for transporting oxygen to the tissues of the body.

Lymph Nodes

A component of the lymphatic system (part of the body's immune system) that contains lymphocytes which produce antibodies and macrophage cells which digest dead cells. Lymph nodes are swollen with cell fragments in the event of infection or cancer. They are located mainly in the spleen but also in the neck, armpit and groin.

Lymphocytes

Lymphocytes are a type of white blood cell that are vitally important to the immune response. There are three types of lymphocytes: B-cells, T-cells and natural killer (NK)-cells.

Monocyte

A white blood cell that attacks invading organisms and helps combat infections

Myeloblasts (or myeloid blasts)

Name given to blast cells in the myeloid cell line. These cells originate in the bone marrow and eventually become the following white blood cells: neutrophils, monocytes, macrophages, basophils, and eosinophils. Myeloid cells also give to the red blood cells and platelets.

Philadelphia chromosome, also called Breakpoint Cluster Region-Abelson Murine Leukaemia Viral proto-oncogene 1 (BCR-ABL1)

An abnormal chromosome fusion gene due to a swapping over and fusion of sections of DNA between chromosomes 9 (ABL1) and 22 (BCR), resulting in a new fusion gene BCR-ABL1. This gene causes overproduction of myeloid cells. It is found in all patients with chronic myeloid leukaemia and some patients with acute lymphoblastic leukaemia.

Glossary (cont.)

Red blood cells

Small blood cells that contain haemoglobin and carry oxygen and other substances to all tissues of the body.

Remission

Remission has occurred when:

- Blood cell counts have returned to normal
- Less than 5% of abnormal, leukaemia cells are still present in the bone marrow

Spleen

The largest organ of the lymphatic system whose function is to help clear the body of toxins, waste and other unwanted materials. The spleen is located under the ribs on the left of the abdomen.

Stem Cell

The most basic cell in the body that has the ability to develop into any of the body's specialised cell types, from muscle cells to brain cells.

Stem Cell Transplant

Transplant of stem cells derived from part of the same individual or a donor.

Targeted Therapy

Drugs that specifically interrupt the leukaemia cells from growing in the body. However, these drugs do not also harm the body's healthy cells the way conventional drugs do.

Tyrosine Kinase Inhibitors (TKIs)

Drugs that inhibit the tyrosine kinase enzyme which controls the function of a cell. Tyrosine kinase inhibitors can switch 'off' tyrosine kinase enzymes that are permanently active due to a mutation.

White Blood cells

White blood cells create an immune response against both infectious disease and foreign invaders. Granulocyte white blood cells include the neutrophils (protect against bacterial infections and inflammation), eosinophils (protect against parasites and allergens) and basophils (create the inflammatory reactions during an immune response). Others include the lymphocytes (recognise bacteria, viruses and toxins, to which they produce antibodies) and monocytes (clear infection products from the body).

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.leukaemiacare.org.uk
support@leukaemiacare.org.uk

Blood Cancer UK

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

0808 2080 888
www.bloodcancer.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

Leukaemia Care,
One Birch Court,
Blackpole East,
Worcester,
WR3 8SG

Leukaemia Care is registered as a charity in England and Wales (no.1183890) and Scotland (no. SC049802).
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Registered office address: One Birch Court, Blackpole East, Worcester, WR3 8SG

Leukaemia Care
YOUR Blood Cancer Charity