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Support for anyone affected by CLL and SLL.

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CLL SUPPORT

Chronic Lymphocytic Leukaemia (CLL)

Small Lymphocytic Lymphoma (SLL)

A guide for patients and others affected by CLL or SLL

CLL SUPPORT

Introduction

Being diagnosed with Chronic Lymphocytic Leukaemia or Small Lymphocytic Lymphoma can be a shock. You are bound to have lots of questions. This booklet aims to answer some of them for you. It covers the basics and points you to further information if you want to find out more. There is a lot to learn, so we suggest you take your time. You could start with what is most relevant to you now and refer to other sections as the need arises.

Contents

All the information contained in this guide is up to date at the time of publication. However, rapid changes in CLL research may mean that some details are superseded by new information.

About CLL Support

Our charity was founded in 2004 by patients with Chronic Lymphocytic Leukaemia (CLL) and their partners. It continues to be run by volunteers, all of whom have been personally affected by CLL or Small Lymphocytic Lymphoma (SLL). CLL and SLL are slightly different forms of the same disease and are managed in the same way. CLL is used to refer to both forms of the disease. Currently we have more than 3,000 members.

Our aim is to inform, support and empower CLL patients and their partners and families. We hope that this booklet will do just that for you.

We consulted our members for advice on what to include in this booklet. Then we submitted the draft to national UK CLL experts to check it for accuracy and to make sure our information was up to date. We also shared the draft with a panel of nurses, patients and their partners for comment, before finalising the text. **The guide is not intended to replace advice from your haematologist.**

Becoming a member of CLL Support

If you are not already a member of CLL Support, we urge you to join. Membership is free and will give immediate access to our regular newsletter. It will also enable you to register for our free and very popular webinars and conferences across the UK.

To join go to:

cllsupport.org.uk

You'll find out about what we are doing, informative articles, videos of presentations given at our conferences by leading CLL experts, and much more, on our website: **cllsupport.org.uk**

What we offer

- Free, friendly and informative conferences for members.
- A very active on-line forum: **healthunlocked.com**
- A helpline: **0800 977 4396**. Please note that we operate an answerphone message system and aim to call you back within 48 hours. Our helpline is staffed by volunteers, all of whom have personal experience of CLL either as patients or as partners of patients. If you feel you need medical advice, please contact your medical team.
- Campaigning and advocacy.
- Regular newsletters.
- Signposting to other sources of help.

What is CLL?

Chronic Lymphocytic Leukaemia (CLL) is a type of cancer that affects white blood cells.

Under normal circumstances healthy white blood cells help our bodies fight infection and disease. In CLL, the bone marrow produces too many white blood cells, called lymphocytes, which aren't fully developed and which don't work properly. Over time, these cells accumulate in the blood and outnumber the normal cells. As well as the blood and bone marrow, white blood cells are also found in large numbers in the lymphatic system, the spleen and other body tissues. This may give rise to enlarged lymph nodes and spleen. See the diagram and explanation on page 13.

When the abnormal cells accumulate mainly in the lymph nodes rather than the blood, it is called Small Lymphocytic Lymphoma (SLL).

CLL can behave very differently in different people. The term 'chronic' means that in most cases this type of cancer develops or progresses very slowly over months or years. Around a third of CLL patients may never need treatment. However, in some cases the disease progresses more rapidly and the options for treatment will need to be considered.

How common is it?

CLL is the most common form of leukaemia in adults in Western countries.

Approximately 4,220 adults are diagnosed with CLL in the UK each year. It is more common in white people and is rarely seen in children.

What causes CLL?

The exact causes for CLL are not known, but research is ongoing to find out more. It is not thought to be caused by lifestyle.

Factors that can increase the risk of CLL are:

- Age

The risk of developing CLL increases with age. Only about 10% of CLL patients are under 55 years of age.

- Gender

Men are about twice as likely as women to develop CLL.

- Family History

5% of patients who have CLL also have a relative with CLL. Over 20 genes have been identified which predispose people to developing CLL, however it is not considered a hereditary disease.

Indicates where more information is available on our website.

Signs and symptoms

CLL usually develops slowly and more than half of all patients don't have any symptoms at the early stages of the disease.

Because people with CLL don't always feel unwell at first, it's often found when they have a routine blood test for some other medical reason. As the disease develops, the abnormal cells (known as B-lymphocytes) grow steadily and accumulate in the bone marrow, lymph nodes and blood. This means that the bone marrow may not be able to produce the normal numbers of healthy blood cells as it becomes swamped.

Over time, CLL patients may experience increasing symptoms which can include:

- Tiredness, sometimes increasing to extreme fatigue.
- Swollen lymph nodes, especially in the neck, armpits or groin. This might lead to discomfort as the nodes increase.
- Frequent or more severe infections.
- Breathlessness, tiredness and headaches caused by anaemia (lack of red blood cells).
- Bruising or bleeding easily, for example bleeding from the gums.
- Swollen abdomen due to spleen enlargement or enlarged nodes.

Other symptoms can include: frequent, severe night sweats, unexplained weight loss, and/or fevers (high temperatures) in the absence of any infections.

Tell your doctors about any new symptoms, or any that seem to be getting worse.

It might be worth recording these in a diary. This will allow you to look at the pattern of these symptoms, which you can then show your doctor.

"Being told I had CLL was such a shock. My mind just went blank."

Diagnosis

If CLL is suspected, a number of tests will be carried out.

The Full Blood Count (FBC)

FBC is one of the key tests and is usually the first step.

Blood contains three types of cells:

- Red blood cells carry oxygen to all tissues in the body. Muscles and other tissues need oxygen to produce energy from food.
- White blood cells fight and prevent infection. White blood cells consist mostly of neutrophils and lymphocytes. There are small numbers of three other types of cells.
- Platelets. These stick together and stop bleeding, for example at a cut or bruise.

For an FBC a sample of blood is taken and examined in the laboratory. The number and appearance of red cells, white cells and platelets are recorded and compared with normal blood test results.

A chart of normal blood counts is given on page 10.

However, an FBC on its own does not confirm CLL and further tests will be required.

The most common tests include:

- Immunophenotyping. This is a laboratory technique to find out if there are abnormal blood cells and if so how many. It's a very sensitive test and can spot even small numbers of CLL cells.
- Peripheral blood film. This is a look at the blood cells under a microscope, which can help identify any abnormal cells.
- Lymph node biopsy. Most patients won't need this, as usually blood tests are sufficient for a diagnosis. However, it may be necessary if your nodes are swollen, but the blood tests show as normal. This can be associated with Small Lymphocytic Lymphoma (SLL).

A lymph node biopsy is a minor surgical procedure where a small sample is taken from a lymph node, then examined under a microscope to help confirm the diagnosis.

Further tests are carried out when the time comes to consider treatment. See page 16 for further information.

Staging

Once diagnosed, your consultant will 'stage' your CLL. Staging is used to describe where the CLL is located and the extent to which the CLL is affecting the blood count and the number and size of lymph nodes. Staging CLL in this way helps your consultant to predict how quickly the cancer may grow and to keep track of it.

There are two main systems to stage CLL. Most doctors in the UK use the Binet system. In the USA, doctors commonly use the Rai system.

The Binet staging system

This is a three-step staging system (A, B and C) based on the number of swollen lymph node areas and blood cell counts.

Stage A

- No anaemia and a normal platelet count.
- Fewer than three areas of lymph node enlargement.

Stage B

- No anaemia and a normal platelet count.
- Three or more areas of lymph node enlargement.

Stage C

- Anaemia and/or low platelet count, regardless of the number of areas of lymph node enlargement.

What's the outlook?

It is possible to have CLL and still have a good quality of life.

Many patients have no symptoms and carry on with life as normal. For some, their CLL may be so slow developing that treatment is not necessary. Others can manage their condition with the appropriate treatment.

New treatments introduced in the last few years mean that, generally speaking, patients now have a better prognosis (outlook) than before.

For more information on treatments, see page 16.

For more information on prognosis see page 29.

What do my blood results mean?

As a CLL patient, you will need to have regular blood tests to monitor the level of CLL cells in your blood and also to look at your general health.

You will find it useful to keep a copy of your blood results to help you see how you are progressing.

One of the main tests your medical team is interested in is the Full Blood Count (FBC). This measures how many blood cells of each kind there are in your blood. The FBC is important as it shows how the CLL, or treatment, is affecting you. The following chart shows the main blood counts that your medical team will be focusing on. It is useful to know some of the medical terms that may be used in discussions with your medical team.

	White cells	Red cells	Platelets
Medical name:	Neutrophils Lymphocytes	Erythrocytes	Platelets/ Thrombocytes
What they do:	Fight infection	Carry oxygen	Stop bleeding
Low counts are called:	Neutropenia	Anaemia	Thrombocytopenia
Symptoms:	Infections	Tiredness, breathlessness	Bruising, bleeding

"I used the diagnosis to live life more, do more things."

How many of each kind of blood cell should you have?

In a healthy person, the blood counts usually stay the same, with slight variations (both up and down) over time or because of infections. The table below shows how many of each kind of blood cell a healthy person will have. If your counts are outside the ranges shown, your doctor will monitor you carefully to see if there is a trend that could in the future lead to treatment.

	White Blood Count (WBC)	Haemoglobin (HGB)	Platelets	Absolute Lymphs	Absolute Neuts
RANGE Men	4.0-11.0	130-180	150-440	0.85-4.1	2.0-7.5
RANGE Women	4.0-11.0	115-165	150-440	0.85-4.1	2.0-7.5

Note that these counts have to be assessed by a doctor. They will be different for patients of African – Caribbean descent. Please check our website for further information.

*Haemoglobin is a protein used by red blood cells to distribute oxygen to other tissues and cells in the body. For a fuller explanation of terms please refer to our website.

There is a chart at the end of this booklet for you to record your test results.

With CLL, total white cell counts tends to go up, often well above normal, while the other cell counts tend to go down. This is because the white cells crowd out the other cells or prevent them being manufactured. However, remember:

- Your blood tests must be looked at as a trend over time. A single or even a pair of results can be misleading. Many factors, other than CLL, can cause the blood counts, including the lymphocyte count, to go up or down – infection, trauma, operations, other illnesses, medication etc. So, any changes must be viewed in the broader context.
- Tests can be reported as 'normal' even though CLL may be present.
- Tests may be reported as 'abnormal' even though CLL is not present.
- Other conditions may mimic CLL.
- Often follow-up tests are needed to clarify the results of tests.

Rule of thumb

As a general guide, it is important to be aware that the white cell count is most useful at the time of diagnosis to identify CLL. However, after diagnosis, changes in the CLL cells (i.e. the white cells increasing) are not as useful in monitoring CLL as changes in the normal cells (i.e. falling haemoglobin and/or platelets).

The immune system and CLL

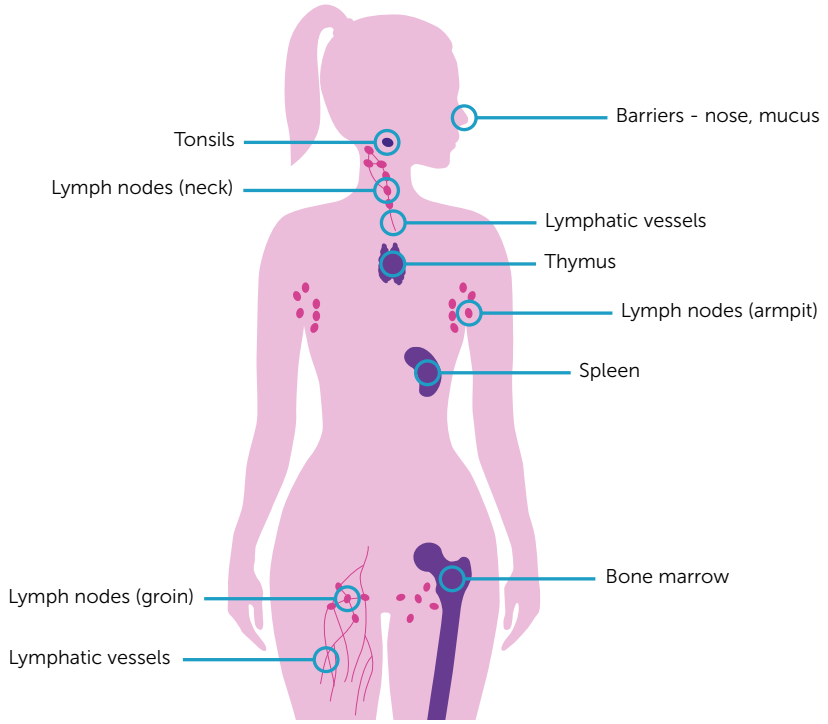
Normally, your immune system helps your body to fight infections. However, in CLL patients, the immune system can be weakened, and there is an increased risk of more severe infections. Your doctor or nurse may want to talk to you about this, and it will help you to understand some of the terms that might be used.

Your immune system is a network of cells, tissues and organs, which protect your body against infection. Lymphocytes play an important part in this. There are lots of different types of lymphocyte, but the important ones to know about are T cells and B cells. These are types of white blood cells which fight infection. CLL affects B cells by allowing them to grow out of control, because they don't 'switch off' and die when they should. They then crowd out other cells, like T cells, from the bone marrow. They may swamp your lymph nodes and spleen, making them enlarged. When this happens, your body can't produce enough T cells, and the B cells that are produced are immature and don't work properly.

One of the key measures of how your CLL may be progressing is to look at the number of white cells in your blood. This is usually done by examining the Absolute Lymphocyte Count (ALC), and you may hear this mentioned by your consultant.

Your lymphatic system

The lymphatic system is a network of thin tubes called lymph vessels, that run around your body. Connected to the vessels are lymph nodes or lymph glands. In CLL, these can become swollen as the abnormal lymphocytes build up there. The ones in your neck, armpits and groin are usually most affected and you may also experience swelling of your spleen.



How it works

The lymphatic system is similar to the blood circulation. The lymph vessels branch through all parts of the body like the arteries and veins that carry blood. But the lymphatic system tubes are much finer and carry a colourless liquid called lymph.

The lymph contains a high number of a type of white blood cells called lymphocytes. These cells fight infection and destroy damaged or abnormal cells.

As the blood circulates around the body, fluid leaks out from the blood vessels into the body tissues. This fluid carries food to the cells and bathes the body tissues to form tissue fluid. The fluid then collects waste products, bacteria, and damaged cells. It also collects any cancer cells if these are present. This fluid then drains into the lymph vessels.

Vaccinations

If you have CLL, you are more likely to pick up infections. This is because CLL weakens your immune system. It is important, therefore, to make sure you are vaccinated for a range of infections as soon after diagnosis as possible. You may also need regular booster vaccinations. Your doctor will advise you about this.

You must NOT receive vaccines, which contain live or attenuated (weakened) viruses.

These include: yellow fever, oral polio, measles, smallpox, MMR and shingles.

Also note that babies who have received the oral polio vaccine should be avoided for at least a week, as they can pass on the live virus. Similarly, avoid children who have been recently vaccinated using a live virus including the nasal flu vaccine.

It is hoped that a new non-live shingles vaccination (Shingrix) will be available in the future.

What vaccinations should I have?

Your healthcare team will advise you, but the most useful vaccinations you should consider are:

Flu vaccine. This may not work for you as well as for people without CLL, but it should offer at least some protection. You should have this vaccine annually. Your close family should be vaccinated too as this will protect them from getting flu and from passing it on to you.

Pneumonia. For pneumococcus, modern practice for CLL patients is to give two vaccines some time apart. These are known as Prevenar-13 (child vaccine) and the usual Pneumovax-23. You should talk to your CLL consultant about having these. Your GP may not be aware of this. It is not necessary to have this vaccine annually. Your doctor will advise when you may need to renew it.

For Haemophilus influenza (HiB), another cause of pneumonia, vaccination is also recommended. The simplest way to protect against HiB is to have one dose of Menitorix (conjugate of HiB and MenC) which can be given at the same time as Prevenar-13. This also protects against Meningococcus (MenC), a cause of meningitis.

Meningitis. See above for HiB.

For more information on vaccinations for CLL patients, see the video of the presentation given by Dr Helen Parry at the CLL Support Birmingham Conference in 2018.

Intravenous Immunoglobulin Therapy (IVIg)

If you are getting lots of infections and your antibodies (infection-fighting proteins in your blood) are low, you may be offered IVIg as an infusion every four weeks or so to help boost your immune system. Immunoglobulin can also be offered to some patients as self-administered injections under the skin at home. This is usually given every 7 days.

Blood transfusion

If you become significantly anaemic, either because of your CLL or during treatment, you may require one or more blood transfusions. **If you've been treated with certain chemotherapy drugs, such as fludarabine, bendamustine and alemtuzumab (Campath) and need a transfusion, you must receive irradiated blood products. Note that this applies to platelet transfusions also.** For this reason, if you fall into this category, it is advisable to wear a wristband identifying you as an at-risk leukaemia patient who needs irradiated blood products. Your consultant or Clinical Nurse Specialist should give you a warning card that you should always carry with you.

Some simple tips to avoid infection

For CLL patients infection can be a real risk. However, you can reduce this by:

- Avoiding people with an infection.
- Avoiding crowded places where there is a risk of infection.
- Washing your hands regularly.
- Trying to keep good personal hygiene.
- Taking care to keep your mouth clean. Consider using an alcohol-free mouthwash.
- Avoiding foods that are not fresh or may be contaminated.
- Drinking plenty of fluids.

Seek advice from your consultant or GP regarding a prescription for a course of antibiotics for you to have to hand in the event of getting an infection.

Treatment

In the majority of cases, CLL is not yet curable, but it is very treatable and it is usually possible to control the disease. You may well have a normal life span with a good quality of life after diagnosis.

The treatments available for CLL are improving and changing. They are determined by current NHS funding regulations, which might make you eligible or not eligible for different treatments. Whether you have had treatment before and how many lines of treatment will also be a consideration.

When to start treatment

Many people with CLL don't need to begin treatment immediately after being diagnosed. If you do need to start treatment, your treatment options may depend on your general health, including any other health problems, and also your wishes.

You will need to have regular blood checks to see whether your disease is progressing. This is often called 'watch and wait'. However, it may be more helpful to use the terms 'medical monitoring' or 'active monitoring'. It is important that you attend these appointments so that your consultant can track your CLL, and talk to you about how you are feeling.

The indications to start treatment may include:

- Enlarging lymph nodes, liver or spleen, pressing on organs, causing them not to function properly.
- Falling haemoglobin or platelet counts.
- Excessive fevers, weight loss or frequent, drenching night sweats.
- Your lymphocyte count doubling within six months.
- Extreme fatigue, which affects your ability to cope.

NOTE: A rising lymphocyte (white blood cell) count alone is not usually an indication that treatment is necessary.

The aim of starting treatment is to improve symptoms, improve blood counts and prolong survival with as good a quality of life as possible. Your consultant will not recommend treatment until it becomes necessary. Current clinical thinking is not to start treatment immediately after diagnosis as there is no evidence that this improves the outcome.

Tests to help determine treatment options

The following tests may be carried out:

Bone Marrow aspiration and biopsy

This involves extracting a small amount of fluid from the marrow space and also a sample of the more solid part of the marrow for analysis in the laboratory. This is called a bone marrow biopsy or trephine biopsy.

Imaging

Ultrasound and, more frequently, CT (Computed Tomography) scanning enables your consultant to more accurately assess enlarged lymph nodes and spleen.

FISH test, also known as Cytogenetic tests

FISH stands for Fluorescence In Situ Hybridisation. The test looks at the genetic make-up of your CLL cells, as this can influence your treatment options. If you have an abnormality of the chromosomes in your CLL cells, known as a 17p deletion, TP53 deletion or mutation, you are less likely to respond to chemotherapy and alternative treatments will be considered. Note: FISH does not test TP53 and another test will be required for this.

More information on tests available is given on page 29 in the section on prognostic factors.

What type of treatment will I have?

Treatments have improved enormously over the last 20 years, and even more in the last 10. It is now normal to use combinations of drugs and these have been increasingly successful.

Any treatment will be tailored to your particular health profile and the risk profile of the CLL itself. Your consultant will advise you on the best available for you.

The standard, first-line treatment for many younger patients is called chemo-immunotherapy. However, some patients may not be suitable for this and other treatments are now available.

Chemo-immunotherapy

Chemotherapy is the use of anti-cancer drugs to destroy cancer cells. In addition, you will probably receive other drugs called monoclonal antibodies. These are drugs that can bind to and kill specific cells, and the combination is called chemo-immunotherapy. The most common chemo-immunotherapy for CLL is called FCR.

FCR

FCR stands for the three drugs Fludarabine, Cyclophosphamide and Rituximab. Rituximab is an example of a monoclonal antibody drug (immunotherapy). Fludarabine and Cyclophosphamide are examples of anti-cancer drugs. If you are less than 65 years of age, fit, and have no other medical problems, you may be treated with FCR. However, there is a general move towards the more targeted treatments.

FCR is usually given in 'cycles'. Each cycle is 28 days long, and you'll normally have treatment each day for five days, then have a break for 23 days without treatment. You may have up to six cycles.

Fludarabine and Cyclophosphamide are tablets.

Rituximab is given as an infusion into a vein. Some terms you may hear for this are 'drip' and 'intravenous' or 'IV'. Usually only the first day of each 28-day cycle requires you to attend hospital (for the Rituximab infusion) and the remaining four days you finish the chemotherapy in tablet form at home.

Some other treatments:

Monoclonal antibodies

- **Rituximab** and the newer **obinutuzumab** are both monoclonal antibody drugs. As well as being part of FCR treatment they are also used in combination with drugs including some targeted treatments (page 19).

- **Bendamustine**

This is another chemotherapy drug, given as an infusion, in combination with rituximab.

- **Chlorambucil**

Also a chemotherapy drug taken in tablet form, along with a monoclonal antibody infusion.

Your healthcare team will discuss with you how many courses of treatment are recommended, and when.

" I was on 'watch and wait' for ten years. I used this time to learn as much about CLL as I could. "

Newer treatments becoming available:

These are some of the newer targeted therapies you may be offered.

- **Ibrutinib**

Ibrutinib is a targeted drug which works by blocking signals within cells that are important for their survival. This drug is particularly useful if you have certain genetic characteristics known as 17p deletion, 11q or TP53 mutation.

Ibrutinib is currently used mainly if a first treatment hasn't worked as well as expected, or if CLL has returned after previous treatment. Ibrutinib is taken in capsule form, usually once a day.

- **Acalabrutinib**

Acalabrutinib is new treatment which acts in a way similar to ibrutinib, but which may have fewer side effects in some patients. Both these drugs are very similar and block the same pathway in the CLL cells' metabolism.

- **Idelalisib**

Idelalisib blocks some of the proteins inside cancerous cells that encourage the cancer to grow. It may be used along with Rituximab. It may be used to treat you if you have not responded to other treatments. It is taken in tablet form.

- **Venetoclax**

Venetoclax is a new treatment which blocks the growth of CLL cells and promotes cell death. It is taken in capsule form once a day. Venetoclax is usually administered together with other drugs such as obinutuzumab or rituximab, in combination, to increase effectiveness.

- **Chimeric Antigen Receptor T cells (CAR-T cells)**

In CLL, the normal immune system has become very weak. In CAR-T cell therapy, a CLL patient's T cells are removed, manipulated in a laboratory to make them better able to kill CLL cells. They are then returned to the patient. At present CAR-T cell therapy is not available for CLL, and it is still at an experimental stage.

- **Clinical trials**

Your doctor may suggest that a clinical trial could be an option for you. Please see the separate section on page 21 on trials for more information.

Side effects from treatment

You may experience side effects from your treatment, although this varies greatly between patients and depends on the type of treatment you receive. Chemo-immunotherapy may cause the following, listed below. You are unlikely to have all of these and for most people the side effects aren't severe and stop when treatment stops. There are medicines you can take to alleviate these symptoms. Ask for advice from your medical team.

- Infections
- Aches
- Constipation
- Diarrhoea
- Tiredness
- Low blood pressure
- Low platelet count
- Low red cell count (anaemia)
- Mouth sores
- Nausea

You should report any side effects to your medical team who will be able to help.

There are medicines you can take to help with nausea, for example.

Newer treatments may have different side effects, which your doctor will be looking out for, and it is therefore important to keep appointments to review your progress.

Second malignancies (new cancers) are a long-term development for a significant minority of CLL patients following chemo-immunotherapy.

"What can I expect and what's the process?"

Clinical trials

You may have heard about clinical trials. When you need treatment, it may be something you wish to consider. This section explains what these are, and why they may be right for you.

What is a clinical trial?

Clinical trials are planned studies involving patients. In CLL, the studies are usually testing new drugs, typically comparing them with existing treatments in order to find better therapies. Trials are essential for evaluating new treatments.

What are the different types of clinical trials?

There are three types of clinical trial: phase 1, phase 2 and phase 3. Each new drug treatment must go through all phases.

Phase 1 trials are concerned with safety, optimum dosage and frequency, and side effects.

Phase 2 trials try to find out what measurable effect the new drug may have on the disease.

Phase 3 trials compare new treatments with the best currently available treatment (standard treatment).

What is a randomised clinical trial?

Most phase 3 trials are randomised. That is, patients who enter the trial are allocated to one of two or more planned treatments, usually by computer. Each group of patients in the trial will have a treatment which is effective against CLL and the object is to see how well each treatment performs against the others.

How are clinical trials planned?

Clinical trials are very closely controlled and doctors are required to write a detailed plan of how the trial will proceed. This is then reviewed by independent bodies of experts to ensure that the trial meets all of the strict criteria before it can go ahead.

Are there advantages in being in a clinical trial?

The main advantages of being in a trial are:

- You may receive a new treatment that is better than standard treatment but is not available outside the trial. Note that you may also be allocated to the group for standard treatment.
- You will be very closely monitored by your medical team, who want to assess the effectiveness of the treatment and will keep a close eye on how you respond.

Is a clinical trial right for me?

Your consultant will advise you if you are suitable to be considered for a trial. There are usually certain preconditions that mean that not all patients qualify. If you have other health problems apart from CLL you could be excluded, depending on their nature and severity. You may also be excluded because of age limitations, depending on the nature of the trial. However, there are usually several CLL trials running at any one time and you may well qualify for one of them. Before entering a trial, you would have a thorough check-up, which may include scans and heart checks.

How do I find a clinical trial?

Your CLL consultant will have all the information on the trials that are currently recruiting patients and will be able to advise if you would qualify. You can also find information from the NHS at bepartofresearch.nihr.ac.uk This will tell you if a trial is available in your local area.

> "You need to be informed as a patient to get the right advice."

Finding a CLL specialist

It is important that you are seen by a haematology (blood) consultant who specialises in CLL. This is because treatment options, including trials, are developing quickly and a CLL specialist will be in the best position to know about them.

Unfortunately, at present, there is no publicly available register of CLL specialists.

However, CLL specialists tend to work at a university (teaching) hospital. If you are being seen at a district general hospital (DGH) for your CLL, your consultant should be able to tell you the name of your regional teaching hospital. Your consultant will know the nearest specialist hospital for blood cancers and where CLL clinical trials are being run in your area. If you want this information, don't hesitate to ask your consultant for it. See also "How do I find a clinical trial?" advice on page 22 above.

If you would like a second opinion about your diagnosis or treatment options, you are entitled to ask the hospital you attend for your CLL or ask your GP. The NHS states that you have a 'right to be seen at a hospital / by a consultant of your choice'. As a general rule it is best for all doctors involved in your care to be kept informed.

"Shared care" is another option. This involves being seen at a regional specialist centre in conjunction with local follow-up. This approach can ensure specialist input and access to trials, while most of your care remains local. There should always be close communication between the specialist centre, local team, GP and you.

To summarise:

- As a patient, you can ask your GP for a referral to a hospital of your choice, but you may not get the outcome you would like.
- Once diagnosed, you can ask your consultant for a referral to a CLL specialist or regional centre. This is then down to the consultant, who may not agree to accept you. However, you could try other hospitals, or for shared care – an intermittent visit to the blood cancer centre, but more routine visits locally.

Your medical team

You are likely to see a number of different people at your CLL clinic. It can be confusing to know who does what.

The multidisciplinary team (MDT)

The team of medical specialists looking after you, known as an MDT, will vary in its membership and size, depending on where you go for your CLL care.

At diagnosis you will be seen by a Consultant Haematologist. When you go for check-ups you may see different members of the team. Later, if treatment starts, you will get to meet more members of the team.

Members of the MDT are required to attend a meeting every week or fortnight to discuss the diagnosis and treatment options for the patients assigned to them.

Patients may have their cases referred to the MDT at any time when significant changes occur and further treatment options need to be considered. The MDT will take into account your views and circumstances. The conclusions and recommendations of the MDT are shared with you at clinical appointments.

These are some of the members of an MDT:

- Consultant Haematologist
  A specialist in blood diseases, who instigates investigations to obtain a diagnosis and who is responsible for patient treatment.
- Haemato-oncologist
  A consultant who specialises in the treatment of blood cancers.
- Specialist Registrar (SpR)
  A doctor in training to be a consultant haematologist.
- Clinical Nurse Specialist (CNS)
  A nurse who assists the patient through staging and treatment, and with communications.
- Key Worker
  A member of the MDT, most often a CNS, allocated to a patient when treatment begins. The key worker liaises between the patient, MDT and other staff and helps the patient navigate the care system.
- Research Nurse
  This nurse ensures that patients who are eligible and might benefit from a clinical trial are discussed at the MDT meeting.
- Radiologist
  This doctor is a specialist in diagnosing and treating disease using medical imaging techniques such as x-ray, computed tomography (CT), magnetic resonance imaging (MRI).
- Histopathologist
  This doctor analyses tissue samples and returns a diagnosis.

Other specialists, such as a clinical psychologist, or a dietitian, may be available to provide advice to the MDT and additional care to patients.

"I have trust in my medical team. That's a real comfort."

Questions to ask

As you cope with your CLL and treatment, you need to have honest, open discussions with your doctor. You should feel comfortable asking about anything, no matter how small it might seem. Here are some questions you might want to ask. Nurses, social workers and other members of the treatment team may also be able to give you answers. Be sure to write down any questions you have.

Taking another person with you and/or recording your conversation, with the agreement of the person you are consulting, can be helpful.

When told you have CLL

For many people, a diagnosis of CLL is unexpected, even a shock. Most will need time for the news to sink in before they know what questions to ask. You can request an appointment at the CLL clinic soon after hearing your diagnosis to ask questions. Here are some questions you might like to ask:

- What is CLL?
- What stage (risk group) am I?
- How is CLL likely to affect my day-to-day life?
- Will I die from it?
- Can I have a copy of my test results? Will you explain them to me?
- What further tests will I need to have and when?
- When do you decide to treat? Why not start treatment straight away?
- What can I do to help myself?
- Where can I go for further information and support?

For many patients, treatment may not be considered necessary or advisable for you at the stage of diagnosis. You will be monitored to see how your CLL is progressing. This is often referred to as 'watch and wait', however 'active monitoring' or 'medical observation' may be better terms to use. It's up to you!

> **"My consultant welcomed my questions."**

When deciding on a treatment plan

This is a key decision for you and the team treating you. It's reasonable for the patient to need sufficient time to discuss the relevant issues with your consultant and CLL team. Here are some questions you may wish to ask at successive appointments:

- What tests will I have to help determine what the treatment options are in my case?
- What's involved in these tests and what do they tell you?
- What are my options for treatment?
- How do these treatments work?
- What experience do you and your team have of these treatments? (e.g. number of patients per annum, how well the treatment is tolerated, remission i.e. no sign of the CLL in a scan or other examination following treatment...)
- Am I eligible for any clinical trials? How do I find out more about them?
- What is the best place for me to be treated? Why's that?
- What treatment plan do you recommend for me and why?
- How do I go about getting a second opinion?
- Do I have a key worker? If so, name and contact details.

As you narrow down the options, you may want to come back for more detailed information:

- What are the possible side effects of this treatment, both in the short term and long term?
- What can be done to relieve these side effects?
- How might this treatment affect my life now and in the future (work, travel, sex life, fertility, life style)?
- How is the treatment administered and over how long a period?
- What evidence is there of the effectiveness of this treatment?
- How would you sum up the pros and cons of the treatments I am considering?
- Are there any support services I can contact about living with CLL?

When starting treatment

These are some of the concerns you may have when you are about to start treatment:

- Who will be part of my healthcare team and what does each member do?
- If I'm worried about the costs of medical care, who can help me?
- What support services are available to my family and me?
- Whom should I call with questions and problems?
- Can I have a copy of my care plan?

While you are on treatment

Good communication with the team treating you is vital, both to them and to you.

Be sure to raise anything which is bothering you, for instance:

- What side effects should I report immediately to you?
- Is there a direct emergency number to call if I am concerned? Is this a 24hr service?
- How about contacting you and your team? Who do I call, for what and when and on what number? How about email contact?
- What is the role of my key worker? And who is my key worker, their working hours and the contact number to call? For an explanation of "key worker' see page 24.
- What side effects can be expected? And how should I deal with them?
- How will you and I know if the treatment is working?
- What counselling support services are available to me if I start to feel overwhelmed or depressed?

After your treatment

Some key questions to ask at this stage are:

- What follow-up can I expect post treatment?
- When can I return to work?
- What is the chance of the CLL coming back? Should I watch for specific signs and symptoms?
- How do I get a treatment summary and survivorship care plan?

Your outlook: prognostic factors

Your prognosis means the likely course of your medical condition. CLL behaves very differently in different patients. Prognostic factors cannot predict time to treatment, infection rates, survival etc. in a given individual, but you may find it helpful to know what the possible outlook could be for you.

If you want to know the results of tests that can tell you and your doctor about your future outlook with CLL, you may need to ask them. Your consultant haematologist and health care team are the best people to ask. You'll find it useful to inform yourself first about prognostic factors. This will help you, and the doctor, determine the level of information you are seeking. Not everyone wants all the details.

Only you as the patient can ask about your prognosis; those close to you cannot get this information without your permission. Your prognosis will change if you have treatment and according to how well you respond to it.

CLL generally progresses very slowly and for some patients survival can be measured in decades.

About one-third of patients will never need treatment. A second third will eventually need treatment. The remaining third require treatment at or shortly after diagnosis.

Staging

Staging, described on page 8 in this booklet, will be a key indicator of how your CLL is progressing. A low stage usually does not require treatment, whereas a high stage usually does.

Lymphocyte doubling time

This is measured at every CLL appointment with your doctor. If your absolute lymphocyte count (ALC) takes more than 12 months to double, you have a better outlook than those whose count doubles in less than 12 months. But lymphocyte counts may fluctuate, sometimes dramatically, due to infection or stress, so caution is needed in interpreting this data.

Other tests

For newly diagnosed patients, a range of laboratory tests on a blood sample will give you a broad picture of how your CLL is likely to behave. Not all tests are routinely funded by the NHS. None of the tests are strictly essential at diagnosis, whereas they become essential if treatment is being considered.

When considering treatment, you should expect as a minimum FISH for 17p and sequencing for TP53. It can be useful for some patients to be tested for IgHV. Discuss this with your consultant. More information on these tests is given below.

Some CLL centres have the facility for tests on-site, and all hospitals have links to centres where tests can be performed.

FISH

This is a test for chromosomal abnormalities, which about three-quarters of CLL patients have in their CLL cells. These abnormalities frequently change the expression or function of molecules that result in increased cancer cell growth or survival.

Each chromosome has a short arm called 'p' and a long arm called 'q'. The four most common abnormalities in CLL are 17p-, 11q-, +12 and 13q-. '-' means a bit of the chromosome is missing (a deletion). '+' means there is an extra copy of the chromosome (a trisomy).

13q deletion is associated with a good prognosis. +12 is of neutral prognostic significance. 17p deletion may be associated with a shorter time to treatment and a less favourable prognosis. Deletion of parts of chromosome 17p result in loss of the TP53 gene, which is needed for the protective protein p53, is associated with a more rapid disease progression and a poorer response to chemotherapy. You will be offered, therefore, an alternative treatment. The negative significance of 11q is no longer seen with some of the current treatment options.

IgHV status

CLL patients with unmutated IgHV genes have a significantly less good prognosis with conventional chemoimmunotherapy than those with mutated IgHV genes.

For patients whose CLL has already progressed to requiring treatment the most useful prognostic tests are FISH analysis for 17p deletion and sequencing for TP53.

More details are available on the website.

Complementary therapies

It is important to distinguish between "alternative medicine", which is offered in place of orthodox medical treatment, and "complementary therapies", which do not replace orthodox treatment.

You should not use alternative therapies, which are often offered in place of proven medical care and can be dangerous.

Complementary therapies can have a place alongside orthodox medical treatment, especially for helping with relaxation and inner calmness.

Always discuss first with your GP, or most appropriate healthcare professional, any complementary therapies you are considering taking. If you do then start a complementary therapy, let the complementary therapy practitioner know about any medicines you are taking and any other complementary treatments you are having. You may wish to consider physical therapies, such as aromatherapy and reflexology. These therapies can help ease tension and anxiety.

You might wish to try a mind-body therapy, such as meditation, relaxation and visualisation, which aim to create a balance between mind and body. The goal here is to increase a feeling of inner calm and reduce stress.

An excellent place to start is our own Support ACT website here: **http://cllsupport-act.org.uk**

Ask at your CLL clinic if they can recommend a place to go for complementary therapy. Some hospitals offer free short courses on-site for their cancer patients. Or enquire at a Maggie's Centre. These centres specialise in giving free cancer support from therapists, nutritionists and benefits advisors, for example. Check for one near you. **maggiescentres.org.uk**

Macmillan has a comprehensive 86-page booklet "Cancer and Complementary Therapies", which you can download from their website or order a print copy. They also do a 44-page booklet: "Healthy Eating and Cancer". **macmillan.org.uk**

> **"Watch and wait has become easier with time. I use mindfulness."**

Living well with CLL

Having CLL often involves years on active monitoring (watch and wait) before treatment, and often no treatment is ever required. Whatever the prognosis you will benefit from medical observation and a healthy lifestyle. This will improve your well-being and lower your risk of getting other illnesses. A well-balanced diet, being physically active, reducing stress, and being involved in your healthcare are all key elements to living well with CLL.

Some of the feelings you may express are given below and are perfectly normal.

Shock

Shock is often the first reaction when you are told you have cancer. You might:

- Feel numb.
- Not believe what is happening.
- Be unable to express any emotion.
- Find that you can only take in small amounts of information.
- Need to have the same information repeated to you.

Your disbelief may be so strong that you find it difficult to talk about your illness with your family and friends. Or you may find that you need to talk about it over and over again to help the news sink in.

Denial

You might cope with the news of your cancer diagnosis by pretending it's not happening. This may not be a conscious decision but a gut reaction.

You might feel that you can't think about it and find that you:

- Don't want to know anything about your cancer or treatment.
- Prefer to talk about it as little as possible or not at all.

This is another completely natural reaction.

You can tell the people around you quite firmly that, for the time being, you don't want to talk about your condition if you feel this way.

> "My wife and family were the hardest hit, not me."

Total denial

In extreme cases, denial can be unhelpful. Some people convince themselves that either there is nothing wrong with them or that their illness isn't cancer.

You may need professional help from a psychologist or counsellor if this reaction starts to get in the way of your treatment or makes your overall situation even worse.

Other people being in denial

Sometimes you may find denial happens the other way around. You might need to talk about your cancer, but your family and friends may be the ones in denial. They might:

- Try to dismiss the fact that you have CLL.
- Seem to ignore the fact that you have cancer.
- Play down your anxieties and symptoms.
- Talk about other people they know who have cancer, even of a different type.
- Deliberately change the subject.

People can react in this way because they are frightened of cancer themselves. They may be embarrassed by talking about it.

Or they may be terrified that someone they love has a life-threatening condition. If they don't talk about it, they can try to pretend it isn't happening.

But if you want their support, and to share how you feel with them, this behaviour may hurt or upset you. If you feel like this, try to:

- Tell them how you feel.
- Reassure them that you know what is happening.
- Explain that talking to them about your CLL will help you.
- Suggest that they support you by attending your next consultation.

Talking about your cancer

Talking about your situation can help.

It might help to talk to a counsellor if you would like to share your feelings with someone, but don't feel you're able to talk to your friends and family.

See also page 38 "Linking up with others affected by CLL".

Diet

A normal, well-balanced diet should include:

- Plenty of fresh fruit and vegetables – aim to eat at least five portions a day.
- Foods high in fibre, such as beans and cereals.
- Plenty of water or other non-alcoholic drinks. (NHS choices recommends 1.2 litres a day).

You should reduce your intake of:

- Red meat, particularly processed meats, and animal fats.
- Alcohol.
- Salted, pickled, fried and smoked foods.

There is quite good evidence that Vitamin D rich foods and Vitamin D supplements help the immune function as well as emerging data on a more favourable prognosis in some lymphomas.

Weight

If you are overweight, take steps to manage what you eat and exercise more to move towards your ideal weight. Please discuss diet options with your medical team to ensure it is suitable for your condition.

Unexplained weight loss (in other words, weight loss that isn't a result of dieting) is often a direct result of many cancers, including CLL. It can lead to complications over time. Before starting treatment for your CLL, your doctor should perform a thorough medical check-up to search for other possible causes of weight loss.

Fitness and fatigue

Generally, doctors advise at least 30 minutes a day, five days a week, of moderately paced activity such as walking. This level of activity is helpful for people even during treatment. But everyone is different and exercise needs to be tailored to you, taking into account your overall fitness, diagnosis, and other factors that could affect safety.

How exercise can help

Exercising can improve your quality of life and help you feel better. Some studies show that it can help speed up recovery after cancer treatment. Regular exercise can reduce stress and give you more energy.

Exercise helps reduce the risk of heart disease, strokes, diabetes and bone thinning (osteoporosis). Physical activity doesn't have to be very strenuous. You can start gently and build up the amount you do.

Tiredness and weakness are finally being recognised as among the most common side effects from cancer treatment. It's encouraging that taking regular exercise can help combat this. More tips for coping with fatigue are given in our help sheet.

Vitamins/supplements

Talk to your doctor about anything you're thinking about using, whether it's a vitamin, a diet, or anything else. Some herbal treatments, for example, can affect the way any drugs you are taking work and may block the effectiveness of the medication.

Stop smoking

If you smoke, speak to your doctor or call a stop smoking helpline for further advice. They can tell you where your local stop smoking service is.

Reduce stress

How best to reduce stress varies from person to person. Some meditate or pray: others turn to a hobby. Some complementary therapies can be helpful.
See page 31.

See also our website devoted exclusively to stress reduction:
http://cllsupport-act.org.uk

Getting involved in your healthcare

Whether on active monitoring (watch and wait) or receiving treatment, be aware of symptoms, appointments and medication regimes. If you have any problems or notice any new symptoms between your appointments that concern you, let your doctor know as soon as possible. With CLL, secondary cancers and ailments are more likely, so the more quickly you report the easier the treatment. This can also reduce stress.

Understanding more about CLL and its treatment can also help you cope. It means you can discuss treatment, tests and check-ups with your doctors and nurses and be involved in making decisions. This can make you feel more confident and give you back a feeling of control.

If you are a CLL patient your immune system doesn't work as it should and this means that you may be more susceptible to other forms of cancer, especially skin cancer. It is best to be aware of this and take precautions in the sun by using a high factor sunscreen and wearing a hat. You should also mention to your consultant any changes in your general health so that tests can be carried out if necessary.

The CLL Support site has an excellent range of further information.

Recommended websites

HealthUnlocked CLL Support

https://healthunlocked.com/cllsupport
This forum offers information, support and empathy.

Our sister charities also have a wealth of expertise and information.

Leukaemia Care – Chronic Lymphocytic Leukaemia (CLL) A guide for patients (Living with CLL, page 25). Download or order at **www.leukaemiacare.org.uk/support-and-information/help-and-resources/information-booklets/**

Macmillan – Understanding Chronic Lymphocytic Leukaemia (CLL) (Wellbeing, page 65) Download or order at **https://be.macmillan.org.uk/be/s-667-cancer-type-l.aspx**

Blood Cancer UK – Information on blood cancers, including CLL: **www.bloodcancer.org.uk**

Lymphoma Action – Information on CLL and SLL and how to live well. **www.lymphoma-action.org.uk**

Cancer Research UK – Living with Chronic Lymphocytic leukaemia, view at **www.cancerresearchuk.org/about-cancer/chronic-lymphocytic-leukaemia-cll/living-with**

Maggie's Centres – Free, practical, emotional and social support to people with cancer and their families and friends. Find out if there is a Maggie's Centre near you: 0300 123 1801 **www.maggiescentres.org**

We also recommend:

The comprehensive guidelines for the treatment of CLL on the British Society for Haematology (BSH) website are very informative.

www.b-s-h.org.uk/guidelines/guidelines/treatment-of-chronic-lymphocytic-leukaemia/

Support Act:
http://cllsupport-act.org.uk

Linking up with others affected by CLL

Many find fellow CLL patients, as well as their partners and others close to them, a valuable source of information and support. Like you, they have first-hand experience of CLL. They can empathise. Often they can give you practical tips for living well with CLL. Above all, fellow patients and their partners can help you feel less alone in coping with CLL.

Although it can be beneficial to exchange experiences with others in a similar position to yourself, you should be aware that each CLL case is individual. What is right for one may not be right for another. Any medical issue you have should be referred to a medically qualified person for advice.

There are various ways of linking up with others affected by CLL:

- Online forums, such as HealthUnlocked.
- CLL Support conferences.
- The CLL Support/Leukaemia Care buddy scheme.
- Regional support groups.
- The under 60s' CLL club.

HealthUnlocked CLLSupport

This free, global, online forum is a very active community, hosted by CLL Support. It has over 13,000 global members, including 3,400 in the UK. You can connect with other members, share experiences and also learn about the latest developments in CLL research. It is a well moderated site. Members are very supportive and there is a daily newsletter. Find it here: **https://healthunlocked.com/cllsupport**

CLL Support conferences

These conferences are held throughout the UK and are free to CLL Support members. You will hear leading experts speak about the latest developments in CLL treatment and be able to ask them questions. You will also have an opportunity to meet with CLL patients and their supporters and discuss with them issues of importance to you. See our website for information on upcoming conferences: **www.cllsupport.org.uk**

The CLL Support/Leukaemia Care buddy scheme

If you would like to have a buddy - a fellow CLL patient or carer - for telephone support, you can ask our sister charity Leukaemia Care if they can find you one. Leukaemia Care trains buddies and the service is free to users. To be put in touch with a buddy, or to become one yourself, email the Buddy Coordinator at **support@leukaemiacare.org.uk** or call on **01905 755 977**.

Regional support groups

Regional support groups do exist, but they tend to be for all types of leukaemia or for all blood cancers, rather than just for CLL. Take a look at this website to see if there is one near you:

www.leukaemiacare.org.uk/support-and-information/support-for-you/find-a-support-group/

It's worth asking the hospital you attend for CLL if they have a support group or know of one. Ask for their help if you would like to start a support group. Contact CLL Support for advice: **coordinator@cllsupport.org.uk**

"The best thing I did was to go to a CLL Support conference. It made me realise I was not alone and talking to other patients was so helpful."

Travel insurance

Having CLL means that you need to plan to get the right insurance

Travel insurance can be more expensive because insurance companies take into account the risk that you are likely to make a claim. However, it is important that you do have medical cover as part of your insurance, and you may be asked for detailed information regarding your CLL. This can sometimes be frustrating and you may be refused cover by some companies. It is very rare, however, that you will not be able to find suitable cover, although it may be more expensive and may impose restrictions. Don't be tempted to hide anything or you may not be fully covered, or even covered at all.

You may find it more difficult to get insurance cover to go to certain countries where medical costs are high, for example USA, Canada, China, Hong Kong and the Caribbean, and also for countries with poor health systems such as parts of Africa.

You may be offered cover which excludes any pre-existing medical conditions, and you should be aware that you may not be covered for any illness that your CLL may be linked to. For example, your insurance company may argue that a heart condition could be a result of previous chemotherapy treatment.

If you have an existing travel insurance policy, you must tell your insurer that you have been diagnosed with CLL.

Often the experience of other CLL patients can be very helpful and you may find advice on which insurance companies people have used on the CLLSupport HealthUnlocked website.

See also our helpsheet: Travel insurance companies.

You can help CLL Support

As a small charity run by volunteers, we are very dependent on the help and donations we receive from our members and others for our continuing success.

These are some of the ways you can help:

- Become a member of our association - it's free!
- Become a champion and raise our profile in your CLL clinic.
- Tell others your CLL story.
- Join our lobbying campaigns.
- Help run small discussion groups at our conferences.
- Tell others about us and what we do on social media.
- Help us fundraise.
- Give us a donation.
- Be a regular donor.
- Remember us in your will.
- Apply to be a trustee.

Perhaps you have particular skills to offer? We'd love to hear from you. Contact our coordinator to find out more **coordinator@cllsupport.org.uk**

You can also find details on our website **cllsupport.org.uk**

A list of CLL Support helpsheets

The following helpsheets are in preparation. They supplement the information in this booklet. We will alert members via our e-bulletins as and when they become available. Please also tell us any information we haven't covered which you'd like. Contact our coordinator at **coordinator@cllsupport.org.uk**

A glossary of terms used in CLL

A grid for recording your blood and other results

Supporting someone who has CLL

Setting up and running a local support group

Managing fatigue

Psychological impact of living with CLL

Travel insurance companies

Will making

Blood test results chart

We suggest you enter your results after each blood test

	White Blood Count (WBC)	Haemoglobin (Hgb)	Platelets	Absolute Lymphs	Absolute Neuts	NOTES
RANGE Men	4.0-11.0	130-180	150-440	0.85-4.1	2.0-7.5	
RANGE Women	4.0-11.0	115-165	150-440	0.85-4.1	2.0-7.5	

NOTE: Different laboratories may express the numbers in different ways, for example for Hgb (also sometimes shown as Hb) the range may be shown as 13.0-18.0 and 11.5-16.5. These numbers should be treated as guides. Results may vary between hospitals and test centres.