Plasma Cell Leukaemia (PCL)

A Guide for Patients

Leukaemia Care
YOUR Blood Cancer Charity

Introduction

Being diagnosed with Plasma Cell Leukaemia (PCL) can be a shock, particularly when you may never have heard of it. If you have questions about PCL – 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 information, talk to your haematologist, clinical nurse specialist or hospital pharmacist.

Booklet compiled by our Patient Information Writer Isabelle Leach and peer reviewed by Dr Salim Shafeek.

Disclaimer: *As we are accredited by the Information Standard, all of our information has to adhere to a standardised process that ensures it is of the highest quality. Unfortunately, due to the rarity of PCL, we were unable to complete the production process which meant that this booklet cannot be formally accredited. However, we assure you that this information was created with the same values as that which is.*

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 – 5:30pm 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 – 5:30pm.

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

What is PCL?

Plasma cell leukaemia (PCL) is a rare and aggressive variant of multiple myeloma characterised by a very high number of plasma cells in the blood and bone marrow. The distinction between multiple myeloma and plasma cell leukaemia is based on the presence of a circulating peripheral blood plasma cell count of greater than 20% of white blood cells and/or more than $2x10^6$/ml of leukaemic plasma cells in the peripheral blood.

Approximately 60 to 70% of PCL cases are new blood cancers at diagnosis, known as primary PCL, while 30 to 40% occur as secondary transformations of multiple myeloma in patients with relapsed/refractory multiple myeloma, when they are known as secondary PCL.

Primary PCL is a distinct cancer with different molecular and chromosomal findings than secondary PCL and multiple myeloma. Primary PCL represents 2 to 4% of myelomas. Multiple myeloma is a relatively common cancer of plasma cells representing 10 to 15% of all blood cancers. Secondary PCL accounts for 1% of previously diagnosed multiple myeloma cases and is a more aggressive disease with a poorer prognosis.

Plasma cells develop from B-lymphocytes that have been activated, and they produce a single type of antibody which is specific to a particular antigen, for example the E coli bacteria.

Who is affected by plasma cell leukaemia?

Because PCL is such a rare condition, information about its incidence, disease characteristics and treatment have mainly been gathered retrospectively from case reports or series of case reports. There have not been many prospective trials of patients with PCL.

Retrospective studies have provided most of the information that is known about PCL. This is where data from studies that have already been completed are subsequently analysed to see which treatments achieved the best results for which patients.

A prospective trial is designed to determine a specific answer,

for example what are the disease characteristics of a condition, or whether treatment A is better than treatment B for the condition. The study is conducted in patients who meet particular inclusion criteria and following completion of the trial, the results are collected and analysed. Until recently, assessing treatments for PCL using prospective studies was not possible as it was difficult to recruit enough patients for a trial with meaningful results. The largest prospective trial to date included 29 patients with newly diagnosed primary PCL to investigate the efficacy of drug combinations which included bortezomib. See the treatment section for more details about this trial.

The risk of suffering from PCL is very low with an estimated incidence of 0.4 cases per million individuals per year. This incidence is from a recent retrospective study of case reports where 117 patients with primary PCL were treated between January 2006 and December 2016. A similar incidence for PCL was previously found in a retrospective study of the incidence of haematological cancers from cancer registries for the years 2000 to 2002, that included 92 cases of PCL.

Based on data from a series of patient case reports, the median age of patients with primary PCL at diagnosis is between 50 and 59 years. This is younger than for multiple myeloma patients where the median age at diagnosis is between 66 and 70 years, confirming that primary PCL is a different condition to multiple myeloma. PCL is twice as common in people of African descent compared with Caucasians, and it is slightly more common in men than women (60% to 40%); however, this data is based on case series. In a more recent retrospective study of 50 patients with primary PCL, the male to female ratio was 50% to 50%.

Secondary PCL occurs after the initial diagnosis of myeloma and gradual genetic progression and changes. This can be during the natural history of the myeloma progression through various treatments and new mutations at various time

What is PCL? (cont.)

points like a subsequent relapse. The likelihood for developing secondary PCL is very low at about 0.5 - 1%. There are no predictive factors or features towards secondary PCL, but the average age of diagnosis for secondary PCL is 66 years old.

What causes plasma cell leukaemia?

The causes of primary PCL are currently unclear. The control mechanisms that keep the plasma cells within the bone marrow, rather than letting them enter the blood stream as in PCL, are still unknown.

Primary PCL is a unique subgroup of the plasma cell myelomas, and it displays a different biological background and separate laboratory features. Despite having unique biological criteria, some of the genetic markers may overlap with multiple myeloma.

PCL has two types: **secretory** and **non-secretory**.

In the secretory type of PCL, M-protein is released.

- M-protein is an antibody, also called an immunoglobulin, which is secreted by cancerous plasma cells and can be detected in the blood and/or the urine of most myeloma patients.
- Multiple myeloma is classified depending on which immunoglobin antibody is identified. M-protein will normally be an immunoglobin G (IgG) or an immunoglobin A (IgA). Less commonly, IgD or IgE are seen. Secondary PCL occurs more frequently in IgE and IgD myelomas.
- These are areas of overlap between the secretory type of PCL and multiple myeloma.

In non-secretory type of PCL, no M-protein is secreted.

Immunophenotyping

The immunophenotyping process helps analyse the antibodies in patients based on the types of antigens or markers on the surface of the cancer cells. According to which antibodies are present, it is possible to identify the type of leukaemia.

In PCL, immunophenotyping

identifies CD20 antigens more commonly, and CD56 less frequently. CD56 is negative in about 80% of PCL patients, which helps differentiate between PCL and multiple myeloma.

Chromosome analysis

Patients with PCL show a higher incidence of high-risk genetic findings than patients with multiple myeloma. Chromosome analysis of primary PCL plasma cells shows abnormalities in at least 50% of patients. Mutations or translocations have been reported for t(11;14), t(14;16), t(11;14), t(14;20) and t(4;14), the KRAS genes, the tumour suppressor gene P53, as well as the cell regulator genes MYC and MAF, located on chromosomes 8 and 16, respectively. Translocation in genetics is the transfer of one part of a chromosome to another part of the same or a different chromosome, resulting in rearrangement of the genes.

The translocation of the t(11;14) chromosome mutation indicates a poor prognosis in PCL patients, whereas, in multiple myeloma, patients with this abnormality have a more favourable prognosis. MYC translocations are known to play a crucial role in progression of the PCL. The aggressive nature of primary PCL may partly be due to the high numbers of MAF and MYC translocations, TP53 and KRAS mutations and inactivation of TP53.

Symptoms of PCL

Due to the low incidence of PCL, details of its clinical presentation are relatively limited and mainly based on a series of case reports.

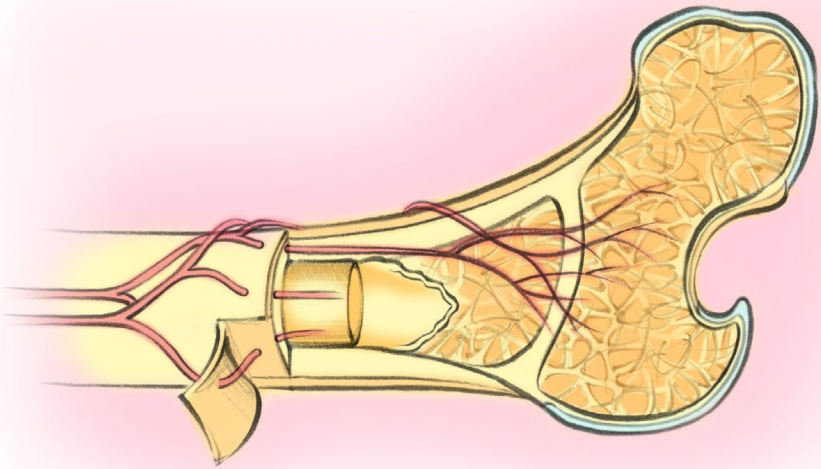
The clinical presentation in patients with PCL typically involves symptoms due to the organ damage. These symptoms include renal failure and bone pain associated with softened area within the bones (called lytic bone lesions) resulting from hypercalcemia (raised calcium levels in the blood).

Other symptoms reported from case series include:

- Pale appearance due to anaemia
- Pleural effusion (fluid on the lungs)
- Enlargement of the lymph nodes, spleen and liver

Rarely, the central nervous system is involved, where the leukaemia cells have spread to the brain and spinal cord. This involvement of organs outside the blood and bone marrow is common in primary PCL, particularly when compared with multiple myeloma.

Blood cell counts reveal low levels of platelets and red blood cells, but high levels of white blood cells, which are mainly plasma cells. Platelets are a type of blood cell that help to stop bleeding.



Diagnosis of PCL

PCL belongs to a unique subgroup of plasma cell abnormalities and has a different biological background, as well as distinct clinical and laboratory features, compared with multiple myeloma and the other plasma cell cancers.

Diagnosis of PCL requires more than 2000 circulating plasma cells per millilitre of peripheral blood and/or a number of plasma cells representing more than 20% of total white blood cells in the peripheral blood.

The diagnosis of primary PCL may be suggested by the presenting signs and symptoms which have a more rapid onset than in multiple myeloma, with a greater tendency of symptoms of increased metabolic rate such as fevers, sweating, weight loss, and fatigue. In addition, organ involvement, hypercalcaemia and renal involvement are common in primary PCL. Leukaemic plasma cells are regularly found in the liver, spleen and spinal fluid in patients with PCL. In addition, central nervous system involvement is associated with high-risk chromosome abnormalities which is also seen more frequently in primary PCL than with multiple myeloma.

Despite having unique biological criteria, some of the immunological and genetic markers may overlap with multiple myeloma. Therefore, the diagnosis of PCL is based on the combination of the symptoms experienced by the patient, microscopic examination of blood, lymph nodes or blood marrow, and the findings from immunophenotyping and chromosome analysis.

To confirm the diagnosis of a patient with suspected PCL, the following tests should be performed and reviewed carefully given the difficulty of diagnosis:

- Peripheral blood smear
- Blood laboratory tests: Complete blood count with differential, electrolyte levels, creatinine, liver enzymes, bilirubin, alkaline phosphatase, lactate dehydrogenase, uric acid, β2 microglobulin
- Bone marrow aspiration and

biopsy

- Serum protein electrophoresis with immunofixation. Electrophoresis is a diagnostic tool to visualise the fragments of protein molecules
- Protein electrophoresis of a sample from a 24-hour urine collection
- Magnetic resonance imaging (MRI) scan and cerebrospinal fluid examination in patients with neurological symptoms
- Immunophenotyping/ chromosome analysis

The diagnosis of PCL can easily be missed when using just light microscopy because the leukaemic plasma cells in the peripheral blood are very similar to circulating lymphocytes seen in conditions such as chronic lymphocytic leukaemia, hairy cell leukaemia, or marginal zone lymphoma. Consequently, immunophenotyping and chromosome analysis are generally required.

Immunophenotyping of peripheral blood can be used to confirm the presence of the circulating cancer plasma cells. Cancerous plasma cells negative for CD56 are generally considered indicative of secondary PCL.

Chromosome analysis of PCL plasma cells shows mutations or translocations of t(11;14), t(14;16), t(14;20) and t(4;14), KRAS genes, tumour suppressor gene P53 and regulator genes MYC and MAF.

Prognosis of PCL

Considering the low incidence of primary PCL, data on its prognosis are relatively limited. Overall survival of patients with primary PCL is known to be very poor when they are treated with conventional chemotherapy, with the majority dying within a year of the diagnosis.

PCL is a very aggressive blood cancer with no standard treatment at present. Nevertheless, induction with a bortezomib-based regimen followed by an allogeneic stem cell transplant or tandem stem cell transplant is achieving promising results. Making the diagnosis early is crucially important so as to be able to start bortezomib-based chemotherapy as soon as possible followed by an ASCT. This approach is known to prolong patient survival.

Prognostic factors

Patients with the t(11;14) chromosome abnormality in primary PCL are known to have a poor prognosis.

Negative prognostic factors for patients with PCL include elevated β2-microglobulin, hypercalcemia, elevated serum lactate dehydrogenase, low serum albumin, advanced age and poor renal function. Additionally, PCL patients with lower platelet counts and higher plasma cell counts are known to have a poor prognosis.

The way patients respond to treatment is also a good indication of prognosis in primary PCL. If the PCL is resistant to induction treatment, then their prognosis is likely to be very poor. Similarly, the failure of blood plasma cells to decline by 50% within 10 days, or to be cleared within four weeks, is thought indicative of unresponsive disease and a negative prognostic factor.

In view of the poor prognosis for patients with primary PCL, ground-breaking treatment regimens in addition to bortezomib, that can be followed by tandem ASCTs, are required to improve the prognosis for patients.

Treating PCL

The ideal outcome of treatment for patients with PCL is achieving complete remission with induction chemotherapy followed by a stem cell transplantation (SCT).

Complete remission is said to have occurred when the following conditions have been met:

- Blood cell counts return to normal
- Less than 5% of cancerous plasma cells are present in the bone marrow
- There are no cancerous plasma cells anywhere else in the body

Induction therapy with intensive chemotherapy and bortezomib-based regimens should be started as soon as possible followed by an autologous stem cell transplantation (ASCT).

In most cases of PCL, the goal of treatment is to reduce the large number of plasma cells in the blood and bone marrow, and prevent or reverse the complications in the liver, spleen and lungs. Successful treatment in these terms will help prolong survival.

Several chemotherapy regimens have been tried with varying results as complete remission is difficult to achieve in many patients with PCL. Conventional chemotherapy only achieves an overall survival of between four and 15 months, therefore new drugs and drug combinations are being tested. These include the following treatments.

Bortezomib

Bortezomib is a proteasome inhibitor which is used as a first-line treatment for newly diagnosed multiple myeloma patients and for the treatment of relapsed/refractory multiple myeloma and Mantle cell lymphoma.

In the prospective trial of bortezomib-based regimens in 29 patients with newly diagnosed primary PCL, bortezomib-based regimens were found to be very effective and achieved an overall survival of 18 months.

A more recent retrospective study of new drugs in 50 patients

with primary PCL also showed that drug combinations which included bortezomib showed efficacy, decreased the rate of early mortality and also achieved an overall survival of 18 months.

Finally, a retrospective analysis of 10 patients with newly diagnosed primary PCL who were treated with a combination of bortezomib, cyclophosphamide and dexamethasone followed by an ASCT, showed a response rate of 100% after induction therapy. Following the ASCT, after a 25-month follow up, survival rate was 70%.

Immunomodulators

Results with immunomodulators such as lenalidomide or thalidomide for the treatment of PCL have been mixed. In a prospective Phase 2 study of 23 newly diagnosed primary PCL patients, lenalidomide combined with dexamethasone resulted in an overall survival rate of 63% after a median follow-up of 15 months.

Autologous stem cell transplantation

Given the poor prognosis of patients with primary PCL, an ASCT is nearly always used as a fundamental part of treatment for patients. Allogeneic stem cell transplantation may be useful in younger patients.

In an ASCT, the patients' own healthy stem cells are collected from the bone marrow before they receive the high-dose chemotherapy. Following the chemotherapy treatment to kill the cancerous plasma cells, the healthy stem cells are transplanted back into the patient. This prevents the stem cells being damaged by the high-dose chemotherapy.

Evidence of the benefits of ASCT was seen in a retrospective analysis of a case series of patients with PCL, which showed an increased median overall survival of 34 months in patients who received an ASCT, compared with 11 months for patients who did not.

The Centre for International Blood

Treating PCL (cont.)

and Marrow Transplant Research in the United States analysed data collected between 1995 and 2006 for 97 patients with primary PCL who received an ASCT as part of their treatment. Overall survival rate at three years was 64%. For patients who received a single ASCT, overall survival rate was 56% but for patients who received a tandem ASCT, overall survival rate rose to 84%. A tandem ASCT means that two ASCTs are performed within a period of no more than six months apart.

Allogeneic stem cell transplantation

In an allogeneic SCT, healthy stem cells from a matching donor, which must at least be a partial match, are transplanted into the patient. A sibling, parent or child are likely to be good matches. Donors who are not relations may be found through national bone marrow registries. Allogeneic transplantations are generally considered for younger patients.

In the study mentioned by the Centre for International Blood and Marrow Transplant Research, data was also analysed for 50 patients who received a single allogeneic SCT. The three-year overall survival rate for these patients was 39%.

Tandem stem cell transplantation

The tandem SCT treatment approach leads to a significant improvement in survival for patients. Results with an allogeneic SCT are poor compared with tandem SCTs because they have a higher mortality rate following relapse. Therefore, an allogeneic SCT should only be performed if options are limited.

A large, recent, retrospective study of 117 patients with primary PCL confirmed the treatment rationale of induction with a bortezomib-based regimen followed by an SCT with an overall survival of 35 months for patients who underwent an SCT and 13 months for patients who did not.

For patients not eligible for SCT-based options, induction with a bortezomib-based regimen should

be started and then followed with maintenance therapy to keep the PCL under control.

Despite a limited amount of evidence from prospective trials or retrospective studies, maintenance treatment following SCTs is increasingly recommended because of the high tendency for early relapse. Maintenance regimens may include either bortezomib or lenalidomide, or both, in combination-based regimens.

Seeing your doctor

Your symptoms

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

Your appointment

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

Your preparation

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

Examples of questions to ask the doctor:

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

Talking to your doctor

Be honest with your doctors; there is no need to feel embarrassed about anything. If you saw your healthcare team before seeing your doctor, be sure to share with your doctor everything your healthcare team told you about your condition, the blood tests

you had performed, and the next steps in your PCL journey. Ask also if you will receive any intensive treatment or palliative care.

Your support

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

The next steps

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

Furthermore, be sure to access all of the other support available to you as this may be able to help you with your feelings towards your diagnosis and treatment.

Telling your family

Planning who to tell

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

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

Planning what to say

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

How to say it

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

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

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

How to respond

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

Accepting help

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

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

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

Managing your emotions

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

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

Uncertainty

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

Isolation

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

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

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

Anxiety

Being fearful of the unknown, especially when we are feeling

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

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

Anger

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

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

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

Sadness and depression

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

Managing your emotions (cont.)

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

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

Self-confidence

Being forced to adjust from your daily routine during the visits to the hospital for treatment can take its toll. This interruption of your life, along with your lack of energy because of your condition and the effects of your treatment, can impact on how you feel about your appearance and how you feel emotionally. In turn, this can knock your self-confidence and self-esteem. Your feelings of relief, hope, and optimism have just been replaced with their polar opposites.

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

Mindfulness and relaxation

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

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

mouth, developing a steady natural rhythm, focusing on your chest and abdomen as you do so.

- Visualise that you are inhaling positivity and exhaling negativity.

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

Survivorship

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

Survivorship can be defined as:

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

When living with cancer, you will face new challenges to cope with from physical to psychological and social ones. Survivorship aims to provide personalised care based on your need to improve your health, wellbeing, quality of life, and your confidence and motivation, to help you manage. Survivorship also focuses on your health and life with cancer after the end of treatment until the end of life. At this point, your routine of meeting frequently with your healthcare professionals also ends, so you may feel a mixture of emotions from relief to fear, anxiety and uncertainty about the future. You may wonder how you will slot back into your life after coming through the treatment period.

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

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

- Dealing with the emotional impact of receiving a PCL diagnosis which may have created feelings of uncertainty, fears of recurrence and difficulties in planning for the future. These will be discussed with you to develop your individualised care plan with support from social care staff and therapists, as you need it.
- Improving your quality of life through efficient and co-

ordinated care during your treatment, with effective communication within the treatment team, and a positive attitude.

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

Palliative care

Palliative care, also known as supportive care, involves a holistic or "whole person" approach, which includes the management of your pain and symptoms as well as psychological, social and spiritual support for you and your loved ones.

Palliative care aims to reduce your symptoms, control your PCL, extend your survival, and give you and your loved ones the best quality of life possible. Your doctor will discuss the options with you in detail before you decide the next steps.

Who provides palliative care?

Your palliative care will be provided by a team of health and social care professionals trained in palliative medicine who will coordinate your care.

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

What is the clinical course?

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

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

- Bone pain: Radiotherapy and/or oral steroids, and sometimes non-steroidal anti-inflammatory drugs (NSAIDs), may be used, although these are used with caution because they can interfere with your immune system and kidney function.
- Bone marrow failure: Blood and platelet transfusions are provided to prevent and

fight recurrent infections and bleeding episodes.

- Oral problems: Analgesic mouth washes and topical ointments may help with ulceration. Chewing gum, and mouth washes, have been shown to help with dry mouth, tooth decay and oral thrush.
- Night sweats and fever: These can place a heavy burden on carers because of so many changes of night clothes and bedding.
- Pathological fractures: Orthopaedic intervention and subsequent radiotherapy, with consideration given to prophylactic pinning of long bones and/or radiotherapy to prevent fractures will be performed. This will reduce the likelihood of complex pain syndromes developing.
- Spinal cord compression: Immediate high single daily dose oral steroids will be given.
- Back pain from wedge and crush fractures of the vertebrae of the spinal column: Treatments can include analgesics, antidepressants and/or anticonvulsant medication used in tandem with opioids.
- Hypercalcaemia: Treatment is usually with intravenous hydration and intravenous bisphosphonates.
- Loss of appetite: Low-dose steroids may temporarily boost the appetite, while small, frequent and appetising meals and supplement drinks will also help.

End of life care

When does end of life care begin?

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

What does end of life care involve?

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

Who provides end of life care?

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

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

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

- **Staying at home:** A place of familiarity, surrounded by your

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

- **Hospices:** These are specialised in looking after those with life-limiting illnesses and those who are coming to the end of their life. Hospices are staffed with care professionals who are able to keep an eye on you, make sure that your symptoms are controlled and offer you a number of services to make your stay as comfortable as possible. For more information on the care that they can provide, go to **https://www.hospiceuk.org/**
- **Residential care/nursing homes:** If you think that your stay may be a few months or more, then a nursing home may be more suitable than a hospice. These can be private or run by a charity or the local council so be sure to check if there are any fees.
- **Hospitals:** Although you may be used to staying in a hospital ward, the care routine cannot always be tailored to your specific needs. Pressures on the NHS mean that your stay will only be as long as strictly required. As soon as the condition you were admitted for has been resolved, you will need to go back to your home or nursing home. However, a number of specialists will be available to help look after you for specific problems, and a number of hospitals also have a designated palliative care team for patients who require them.

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

Glossary

Albumin

Albumin is a protein made by the liver which accounts for up to 60% of the total protein in the blood. Albumin has many roles which include preventing fluid from leaking out of the blood vessels, nourishing tissues, and transporting hormones, vitamins, drugs and calcium throughout the body. Levels of blood albumin can be decreased because of a liver disorder or kidney disease.

Alkaline phosphatase

An enzyme found in various tissues throughout the body, with the highest concentrations being found in liver or bone. Raised levels of alkaline phosphatase in the blood are most commonly caused by liver disease or bone disorders.

Amino acids

Organic molecules which are the building blocks for making proteins.

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.

Antibody

A large Y-shaped protein produced by B-cell lymphocytes in response to a specific antigen, such as a bacteria, virus, or a foreign substance in the blood. The antibodies neutralise the bacteria and viruses.

Antigen

A toxin or other foreign substance which induces an immune response in the body, especially the production of antibodies.

β2 microglobulin (beta 2 microglobulin)

A protein found on the surface of most cells in the body and which is shed into the blood, particularly by B lymphocytes and tumour cells. It is used to assess the severity of certain cancers, including multiple myeloma and some lymphomas.

Bilirubin

The breakdown product of red blood cells.

Bone marrow failure

The term used when the bone marrow is unable to keep up with the body's need for white and red blood cells and platelets.

Central nervous system

A part of the nervous system which includes the brain and spinal cord.

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.

ClinicalTrials.gov

ClinicalTrials.gov is a database of trials and includes details of approximately 276,190 research studies in 205 countries.

Complete remission

Complete remission is said to have occurred when the following conditions have been met:

- Blood cell counts return to normal
- Less than 5% of blasts (abnormal, immature, early cells) are still present in the bone marrow
- There are no blast/cancer cells anywhere else in the body

Cytogenetic

Relating to the study of inheritance in connection with the structure and function of chromosomes.

Electrolytes

Salts and minerals in the blood that help conduct electrical impulses in the body. They include sodium, potassium, chloride and bicarbonate among others.

Electrophoresis

A diagnostic tool to visualise fragments of protein molecules.

Fatigue

Tiredness and weakness rendering the patient unable to work or perform usual activities.

Glossary (cont.)

Genes

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

Hypercalcemia

A condition in which calcium levels in the blood are increased. This can result in weakening of the bones, formation of kidney stones, and abnormalities of heart or brain function.

Immunoglobulins

Immunoglobulins, also known as antibodies, are proteins produced by the plasma cells that act by recognising and binding to particular antigens, such as bacteria or viruses, thereby helping in their destruction.

Lactate dehydrogenase

An enzyme that is required during the process of turning glucose (sugar) into energy for body cells. Lactate dehydrogenase is released during tissue damage, and it is therefore a useful marker for common injuries and disease such as heart failure or muscle injury.

Lymph nodes

Components of the lymphatic system (part of the body's immune system) that contain the lymphocytes which produce antibodies and macrophages to digest dead cells. Lymph nodes are swollen with cell fragments in the event of infection or cancer. They are located mainly 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. B-cells produce antibodies that seek out invading organisms. T-cells destroy the organisms that have been labelled by the B-cells, as well as internal cells that have become cancerous. NK-cells attack cancer cells and viruses.

Multiple myeloma

A common blood cancer arising from plasma cells. It accounts for 15% of blood cancers, and 2% of all cancers.

Neoplasm

A medical term for cancer, meaning literally a new and abnormal growth of tissue anywhere in the body.

Plasma cell

A type of white blood cell that produces antibodies and is derived from a B-cell lymphocyte. It is an ovoid (egg-shaped) cell with an off-centre nucleus.

Platelets

One of the types of blood cells that help to stop bleeding.

Prognosis

An indication of how well a patient is expected to respond to treatment based on their individual characteristics at the time of diagnosis or other timepoints during the condition.

Prospective study

A study 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 attempt to provide an answer.

Radiation treatment

Cancer treatment that uses high doses of radiation to kill cancer cells and shrink tumours.

Refractory multiple myeloma

Multiple myeloma in which treatment does not result in a remission, or that gets worse within six months of the last treatment. However, the multiple myeloma may be stable.

Relapsed multiple myeloma

A relapse occurs when a patient initially responds to treatment for multiple myeloma but, after six months or more, response stops. This is also sometimes called a recurrence.

Retrospective study

Study which analyses data from completed studies where the results are already known. Retrospective analysis looks back at existing data that has already been recorded to determine, for example, the characteristics of a disease, the efficacy or safety of a treatment, or factors which

Glossary (cont.)

might have led to a disease. Retrospective analyses are the opposite of prospective studies.

Translocation

In genetics, a translocation is the transfer of one part of a chromosome to another part of the same or a different chromosome, resulting in rearrangement of the genes.

Uric acid

A product of the metabolic breakdown of purine nucleotides which are the chemical building blocks of DNA. Uric acid is a normal component of urine.

Tell us what you think!

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

Suitable for Android, iPhone 7 and above.

Useful contacts and further support

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

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

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

Leukaemia Care

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

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

Bloodwise

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

020 7504 2200
www.bloodwise.org.uk

Cancer Research UK

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

0808 800 4040
www.cancerresearchuk.org

Macmillan

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

0808 808 0000
www.macmillan.org.uk

Maggie's Centres

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

0300 123 1801
www.maggiescentres.org

Citizens Advice Bureau (CAB)

Offers advice on benefits and financial assistance.

08444 111 444
www.adviceguide.org.uk

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

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

Want to talk?

Helpline: **08088 010 444**
(free from landlines and all major mobile networks)

Office Line: **01905 755977**

www.leukaemiacare.org.uk

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Leukaemia Care
YOUR Blood Cancer Charity