Atypical Chronic Myeloid Leukaemia (aCML)

A Guide for Patients

Leukaemia Care
YOUR Blood Cancer Charity

Introduction

Being diagnosed with Atypical Chronic Myeloid Leukaemia (aCML) can be a shock, particularly when you may never have heard of it. If you have questions about aCML – 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 Mary Frances McMullin and Dragana Milojkovic. We are also grateful to Julie McNeill for their contribution as a patient reviewer.

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

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In this booklet

About Leukaemia Care

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

Our services

Helpline

Our helpline is available 8:30am – 5:30pm 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 aCML?

Atypical chronic myeloid leukaemia (aCML) is a blood cancer characterised by an increase in the reproduction rate of bone marrow (myeloid) cells, most of which are abnormal. In addition, the patient does not have the BCR-ABL1 gene which is present in the 'Philadelphia chromosome'.

aCML is associated with a poor prognosis and, at present, there is no established standard of care. aCML has many clinical symptoms and laboratory results which would suggest the diagnosis of chronic myeloid leukaemia (CML); however, the lack of the BCR-ABL1 gene remains important for its diagnosis and distinction from CML.

In aCML, bone marrow cells start reproducing excessively leading to large numbers of abnormal, immature cells, called blasts. These cells are immature versions of the white blood cells which start life in the bone marrow.

There are different types of white blood cells which include granulocytes, monocytes, macrophages, and dendritic cells. Any of these white blood cell types can reproduce excessively in aCML, but it is generally the granulocytes that do so. Granulocytes consist of three types of cell (neutrophils, eosinophils and basophils), all of which have small granules within the body of their cell, hence their name granulocytes.

In aCML, the bone marrow cells which reproduce uncontrollably are the immature neutrophil cells. Neutrophils are the most common white blood cells accounting for 40 to 80% of all white blood cells. The role of the neutrophil is to protect the body against bacterial infections and inflammation. In addition to having greatly increased levels of immature neutrophil blasts, aCML has myelodysplastic features. Myelodysplastic comes from the Greek words *myelo*, meaning bone marrow, and *dysplasia*, meaning abnormal growth.

Who is affected by aCML?

The incidence of aCML is unknown; however, the estimated incidence of aCML is very small,

representing 0.1 - 0.2% of new cases of leukaemia.

aCML is seen more commonly in patients over 70 years of age, although a few cases in children and teenagers have also been reported. aCML seems to occur equally in males and females. Given the rare incidence of aCML, there is not enough data to determine if there is any racial or geographical variation.

Approximately 40% of patients with aCML will progress into acute myeloid leukaemia, in which case their overall survival is between 12 and 29 months.

What causes aCML?

The exact cause of aCML is unknown. Chromosome abnormalities are found in 20% to 88% of patients with aCML. The chromosome abnormalities which have been commonly seen in patients are trisomy 8 (an extra chromosome 8) and del (20q) (deletion of the q arm of chromosome 20), although other chromosome abnormalities have been described.

Characteristically, patients with aCML do not have the BCR-ABL1 mutation (Philadelphia chromosome) or any of the following chromosome rearrangements:

- PDGFRA (Platelet-Derived Growth Factor Receptor Alpha)
- PDGFRB (Platelet-Derived Growth Factor Receptor Beta)
- FGFR1 (Fibroblast Growth Factor Receptor 1)
- PCM1-JAK2 (PeriCentriolar Material 1-Janus Kinase 2)

A chromosomal rearrangement is a mutation that involves a change in the structure of the chromosome such as a deletion, duplication, inversion, or translocation. 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. Not having these gene mutations is more helpful for the diagnosis of a patient with aCML as they are more characteristic of other leukaemias.

A recent study of the gene

What is aCML? (cont.)

mutations in 35 aCML patients, showed that the most common gene mutations in aCML patients were:

- ASXL1 (Additional SeX combs-Like 1) seen in 60% of patients
- TET2 (Ten-Eleven Translocation-2) seen in 43% of patients
- SRSF2 (Serine/arginine-Rich Splicing Factor 2) seen in 34% of patients
- NRAS (Neuroblastoma RAt Sarcoma) seen in 31% of patients
- SETBP1 (SET binding protein 1) seen in 23% of patients

However, none of these mutations are specific to aCML, and have been described in other leukaemias.

Symptoms of aCML

In aCML, the peripheral blood and bone marrow are always involved, and infiltration of the liver and spleen is also common.

The large numbers of abnormal, immature bone marrow cells which occur with aCML prevent the bone marrow from producing enough healthy blood cells of all types. Symptoms and signs related to the resulting anaemia, thrombocytopenia (low levels of platelets, which are small blood cells that help the body form clots to stop bleeding), or enlargement of the spleen are common and include the following:

- Pale appearance
- Shortness of breath
- Fatigue and weakness
- Infections
- Easy bruising, bleeding or petechiae
- Enlarged spleen and pain/ feeling of fullness below the ribs

Diagnosis of aCML

In the 2016 World Health Organisation Classification of Tumours of Haematopoietic and Lymphoid Tissues classification, aCML is defined as showing all of the following:

- Persistently increased levels of white blood cells (>13x10^9/l; normal range of 4 to 11x10^9/l)
- Immature neutrophil blasts making up >10% of the white blood cells circulating in the blood
- Presence of multiplying abnormal granulocytes, which should be <20% of the blasts present in the blood and bone marrow

There are no specific chromosome abnormalities which have been identified in aCML, making its diagnosis quite difficult. The absence of both the BCR-ABL1 mutation and the chromosome rearrangements of RDGRRA, PDGFRB or FGFR1, or PCM1-JAK2 are a minimum requirement for considering a diagnosis of aCML.

Since there are no gene mutations which are specific to aCML, the diagnosis is strictly based on examinations of the blood and marrow samples.

While the appearance of aCML and CML are very similar, CML can be discounted by the absence of the BCR-ABL1 mutation as it is its diagnostic hallmark. Additionally, examinations of the blood and marrow samples will show the characteristic dysplastic granulocyte cells in aCML.

Diagnostic evaluations

The following tests are used to reach a diagnosis of aCML. As previously mentioned, the examinations of the blood and marrow samples are crucial for making the diagnosis and chromosome analysis enables different types of leukaemia to be excluded.

Complete blood count

A complete blood count with a differential white cell count (to determine which white blood cells are increased) and a blood smear (to evaluate the types of white cells involved) are among the first tests to be done.

Chromosome analysis

As chromosome abnormalities are seen in as many as 80% of patients with aCML, it is important to establish the absence of the BCR-ABL1 mutation, and rearrangements of PDGFRA, PDGFRB or FGFR1 in order to achieve the correct diagnosis.

The most common chromosome abnormalities are trisomy 8 and del(20q), however abnormalities of chromosomes 13, 14, 17, 19 and 12 are also commonly reported. Nevertheless, these chromosome abnormalities are not diagnostic. The common mutations seen with aCML are shown in the section 'What causes aCML'? Unfortunately, none of these gene mutations are specific to aCML.

Of the mutations seen in patients with aCML, the SETBP1 mutation has been described as the nearest thing to a marker for a diagnosis of aCML. The CSF3R (Colony-Stimulating Factor 3 Receptor) mutation, which only occurs in 3% of patients with aCML, should encourage a review of the diagnostic results to make sure there is no alternative diagnosis.

Immunophenotyping

Immunophenotyping is a process which uses antibodies to identify cancer cells based on the types of antigens or markers on the surface of the cells. It can enable the diagnosis of specific types of leukaemia and lymphoma by comparing the cancer cells to normal cells of the immune system.

Unfortunately, in patients with aCML, no specific immunophenotypic characteristics have been reported that could help with diagnosis.

Immunophenotyping does show that cells from biopsies of patients with aCML are positive for the CD14 or CD68R antibodies. This can help identify abnormal monocyte white blood cells, thereby casting doubt on a diagnosis of aCML, which only shows abnormal neutrophil white blood cells.

Prognosis of aCML

Since there have not been any specific chromosome abnormalities identified in patients with aCML, the search for treatments which target abnormalities associated with aCML is more difficult, which impacts on the prognosis of aCML patients.

The prognosis of aCML is also related to its nature as it often runs a more aggressive course. The estimated overall survival for patients with aCML is between 14 and 30 months.

Currently, an allo-SCT is the only potentially curative option in patients with aCML. In 30 - 40% of patients, aCML will evolve into AML, and most of the remaining patients go onto experience bone marrow failure.

Factors predictive for the risk of progression of aCML to AML are enlarged liver or spleen, increased level of monocytes (>3x10^9/l but <8x10^9/l), >5% of blasts in the bone marrow, faulty production of red blood cells, and the need for transfusions.

Factors which are associated with a poorer prognosis for patients with aCML include:

- Increased white blood cell count (>50x10^9/l)
- Increased percentage of immature cancer cells in the blood
- Haemoglobin level <100 g/l (normal range of 135-175 g/l for men and 120-155 g/l for women)
- Age >65 years
- Presence of three or more mutations
- Certain chromosome mutations: ASXL1, TET2, NRAS or SETBP1

Factors which predict a better prognosis for patients with aCML include:

- Certain chromosome mutations: CSF3R - Despite only occurring in 3% of patients with aCML, the CSF3R gene mutation is linked to a better prognosis when it occurs in aCML patients

Treating aCML

Patients with aCML tend to have poor outcomes as there are no specific treatments that have been effective in aCML. Consequently, there are no established treatments or standardised care regimens based on patients' risk factors.

Currently, only an allogeneic stem cell transplantation (allo-SCT) can achieve a cure in patients with aCML.

Allogeneic stem cell transplantation

An allo-SCT is the transplantation of bone marrow stem cells from a matching donor such as a sibling, parent or child. Donors who are not relations may be found through national bone marrow registries.

For patients who are eligible, allo-SCT helps re-establish a healthy bone marrow. Allo-SCT is generally considered for patients under the age of 60 to 65 years, but all patients are considered for this procedure if suitable.

Patients receiving an allo-SCT are given high doses of chemotherapy or radiation to destroy any cancerous cells, and then healthy stem cells from the donor are transplanted into the recipient. Initial tests required for an allo-SCT should be performed when appropriate, irrespective of mutation tests. Patients can be treated with hypomethylating agents (HMAs) or targeted agents (when a relevant mutation has been discovered via mutation testing) to prepare their bone marrow for transplantation.

HMAs are drugs that inhibit DNA methylation, which is known to be essential for cell development and function. Targeted drugs attack the specific abnormalities present in cancer cells that allow them to grow and thrive. Targeted drugs do not simultaneously harm healthy cells the way conventional chemotherapy drugs do.

Since aCML is very rare, especially in patients under the age of 65 years for whom an allo-SCT is more appropriate, experience of the outcomes of allo-SCT in these patients is very limited. Although, it is important to note that aCML patients receiving this treatment are known to relapse very quickly.

Most of the studies of allo-SCTs

are in patients with MDS, which also include a few patients with aCML. However, in an analysis of nine patients with aCML only who received allo-SCTs, all patients achieved complete remission. One patient, whose brother was the donor, relapsed 19 months after the allo-SCT, but following a further allo-SCT with peripheral blood stem cells from his brother, he made a full recovery.

Other treatments available

For patients who are not eligible for an allo-SCT, the treatment strategies which have been successful in patients with other blood conditions, such as MDS, appear to be an option for patients with aCML. These treatments can be adapted to a patient's particular symptoms such as high white blood cell count, enlarged spleen or possible progression to acute myeloid leukaemia.

Ideally, treatment of patients with aCML would result in complete remission, defined as:

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

Hypomethylating agents

Since there are no specific drugs which are effective in the treatment of aCML, the use of hypomethylating agents (HMAs) may provide a possible treatment benefit. Traditionally, this treatment is available for patients with MDS.

Azacitidine (Vidaza®) and decitabine (Dacogen®) are HMAs and act by inhibiting DNA methylation. They have become the standard of care for the treatment of patients with MDS. Azacitidine and decitabine are often used as a bridging therapy for aCML patients who are eligible for allo-SCT, or as stand-alone treatment of patients who do not have the option of an allo-SCT.

Azacitidine has been recommended by the National Institute of Health and Care

Treating aCML (cont.)

Excellence (NICE) as a treatment for adult patients not eligible for allo-SCT and who have MDS, non proliferative CMML and AML, and also for patients with AML with <30% of bone marrow blasts. It is not yet currently recommended for aCML.

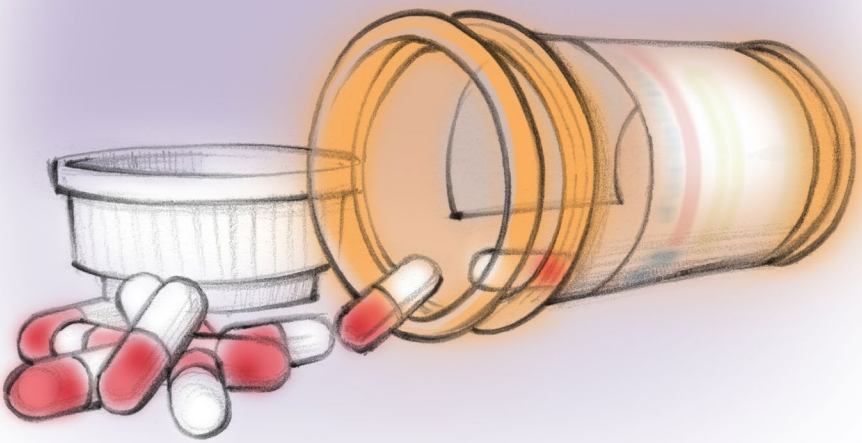
Decitabine has been approved in Europe since 2012 for the treatment of adult patients with acute myeloid leukaemia, who are not candidates for standard induction chemotherapy.

Four recent case reports of patients with aCML have shown some benefit of treatment with decitabine in terms of returning blood counts and bone marrow cell levels to normal.

Additional treatments

Although not yet approved, there are additional treatments which can help in the management of patients with aCML. These include:

- Pegylated interferon alfa: Interferons are naturally-occurring body proteins that send signals to interfere with the ability of viruses to multiply. Pegylation, which is the addition of polyethylene glycol to the interferon alfa, increases the effect of the interferon in the blood.
- Hydroxycarbamide: This anti-cancer chemotherapy drug acts by interrupting the cell cycle and preventing DNA synthesis.
- Erythropoiesis-stimulating agents: These drugs are used to correct anaemia by stimulating the production of red blood cells in the bone marrow.
- JAK inhibitors such as ruxolitinib (Jakavi®) can be effective if the mutation testing has shown the patient has the JAK2 V617F mutation.
- Tyrosine Kinase Inhibitors (TKIs) have been found to treat patients, including those with the ABL-1 EPV6 mutation.



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 the treatment.

Your appointment

Pick a time convenient for you that you know you will be able to attend.

Be sure to let your haematologist know if you notice any new symptoms.

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 be needed?
- What will the tests show?
- How long will it take to get the results back?
- How common is this condition?
- What sort of treatment will be needed?
- How long will the treatment last?
- How will I know if the treatment has worked?
- What will the side effects be?
- Are there any foods or medications that need to be avoided?
- 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; they have seen and heard everything before, so 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 that were performed, and the next steps. Ask also if any intensive treatment or palliative care will be needed.

Your CNS

You will be allocated a specialist nurse for your condition. Keep their contact details as they will be able to support you through the treatment and offer advice.

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 the 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, who can offer additional support.

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 the diagnosis and treatment.

Telling your family

Planning who to tell

Telling your family and friends what is happening 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 the story that you want to tell, the diagnosis, the prognosis, the next treatment steps, and what you expect will happen 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 chance that everything will be okay after treatment."
- "You know I have been feeling unwell for a while. Some tests have been done 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 if they get cancer, that it's their fault, 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.

You may have a positive demeanour, which will obviously be helpful to you during the next steps in the management of the 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 aCML, you may feel alone.

Alternatively, you may feel dealing with your cancer allows you to be around those closest to you. Being around 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 feelings 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 at the cancer diagnosis is natural and normal. You may be angry with yourself, 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, but demanding, goals will help reduce the anger and impatience, 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 energy positively with no negative impact on the body. Talking about feelings and letting them out will also help stop you lashing out at people and keep things calm.

Sadness and depression

You may feel a sense of loss, 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 situation and the illness, treatment and recovery process. However, if this low mood persists for more than several weeks, and you feel hopeless, and lose interest and

Managing your emotions (cont.)

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 the diagnosis and connecting back with your life. Only do as much as you can and try and talk about your thoughts and feelings. This will help lighten your burden and put things into perspective. If you have made any acquaintances or friends in the same position as you, talk to them over coffee as they will understand what you are facing.

Self-confidence

Being forced to adjust from your daily routine during the visits to the hospital for treatment can take its toll. This interruption of your life 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 the diagnosis, and socialising with family, friends, and those in the same position as you. This will help create a supportive atmosphere to get you back to your old self.

Mindfulness and relaxation

Simples practices from mindfulness and relaxation techniques can help you calm the mind, release tension and ease any pain.

- 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 improving 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 aCML. 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 will be identified and addressed, including:

- Dealing with the emotional impact of receiving an aCML 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 an 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 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 a 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 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 the treatment, the physical and physiological side effects of treatments and the psychological impact of aCML 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 pain and symptoms as well as psychological, social and spiritual support for you and your loved ones.

Palliative care aims to reduce the symptoms, control the aCML, extend 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?

Palliative care will be provided by a team of health and social care professionals trained in palliative medicine who will coordinate the 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. The 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 aCML and the impact of the treatments. The therapy may continue because of potential remission and/or useful palliation.

Various pains and other clinical complications can occur such as:

- **Bone pain:** Radiotherapy and/or oral steroids, and sometimes non-steroidal anti inflammatory drugs (NSAIDs), may be used with caution, because they can interfere with the 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.
- **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 the treatment hasn't worked and you are going through palliative care, end of life care may be offered. End of life care begins when it is needed 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 patients enjoy a good quality of life until they die, and to die with dignity. The professionals looking after you will ask 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 this is, 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 the 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 can help to make the decision a little easier.

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

ones, may be something that will be reassuring. External care professionals will be able to visit your home to make sure the 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 symptoms are being controlled and offer a number of services to make the 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 patients' specific needs. Pressures on the NHS mean that your stay will only be as long as strictly required. As soon as the condition requiring hospital admission 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 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 able to help you put everything into place.

Glossary

Antigen

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

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.

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.

Genes

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

Philadelphia chromosome

The Philadelphia chromosome (BCR-ABL) is the most common genetic abnormality associated with adult CML.

Translocation

In genetics, 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.

Trisomy

Chromosomal disorder in which a person has an additional chromosome, resulting in having 47 chromosomes, instead of the normal 46 chromosomes. Down syndrome, where the patients have an extra chromosome 21 is a common example of trisomy.

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

support@leukaemiacare.org.uk

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

Registered charity
259483 and SC039207

Leukaemia Care
YOUR Blood Cancer Charity