Acute Promyelocytic Leukaemia (APL)

A Guide for Patients

Leukaemia Care
YOUR Blood Cancer Charity

Introduction

Being diagnosed with acute promyelocytic leukaemia (APL) can be a shock, particularly when you may never have heard of it. If you have questions about APL – 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. You'll also find useful advice about how to get the best from your haematologist, plus practical advice on how to help important people in your life understand such a rare condition.

For more personalised information, talk to your haematologist, clinical nurse specialist or hospital pharmacist.

This booklet was originally compiled by Ken Campbell, MSC (Clinical Oncology) and peer reviewed by Dr George Cherian and Professor David Grimwade, who provided additional support. The rewrite was put together by Lisa Lovelidge and updated by our Patient Information Writer, Isabelle Leach. It has been peer reviewed by Nigel Russell, Professor of Haematology at University of Nottingham, and James Allan, Professor of Cancer Genetics at Newcastle University. We are also grateful to Thea Wilson for her contributions as a patient reviewer.

Throughout this booklet, you will see a number of quotations. These are the real experiences of blood cancer patients and may help you to understand your disease and situation a bit better.

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

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In this booklet

About Leukaemia Care

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

Our services

Helpline

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

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

Nurse service

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

Patient Information Booklets

We have a number of patient information booklets like this available to anyone who has been affected by a blood cancer. A full list of titles – both disease specific and general information titles – can be found on our website at **www.leukaemiacare.org.uk/support-and-information/help-and-resources/information-booklets/**

Support Groups

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

Buddy Support

We offer one-to-one phone support with volunteers who have had blood cancer themselves or been affected by it in some

way. You can speak to someone who knows what you are going through. For more information on how to get a buddy call **08088 010 444** or email **support@leukaemiacare.org.uk**

Online Forum

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

Patient and carer conferences

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

Website

You can access up-to-date information on our website, **www.leukaemiacare.org.uk**.

Campaigning and Advocacy

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

Patient magazine

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

What is APL?

Acute promyelocytic leukaemia (APL) is a rare sub-type of acute myeloid leukaemia (AML) in which there is an increased production of immature, abnormal white blood cells called promyelocytes in the bone marrow. The term acute indicates that the leukaemia develops rapidly rather than being a description of its seriousness.

To understand APL, it is helpful to understand how blood cells are normally produced. Blood cells are produced in the bone marrow, which is spongy tissue found inside bones. Each day, the bone marrow produces more than a trillion new blood cells to replace those that are worn out. Blood stem cells divide to produce either mature blood cells or more stem cells. Only about one in 5000 of the cells in the bone marrow is a stem cell. A blood stem cell, also called a haematopoietic stem cell, may become a myeloid stem cell or a lymphoid stem cell.

A myeloid stem cell becomes one of three types of mature blood cell:

1. Red blood cells that carry oxygen and other substances to all tissues of the body.
2. Platelets that form blood clots to stop bleeding.
3. White blood cells that fight infection and disease; the shortest-lived white cells are called neutrophils.

A lymphoid stem cell becomes a lymphoblast cell and then one of three types of lymphocytes (white blood cells):

1. B-lymphocytes that make antibodies to help fight infection.
2. T-lymphocytes that help B-lymphocytes make the antibodies that help fight infection.
3. Natural killer cells that attack cancer cells and viruses.

In APL, the myeloid cells that are destined to mature into white blood cells (through a process called differentiation) fail to do so and remain as immature promyelocyte cells.

These promyelocyte cells accumulate in the bone marrow leading to a shortage of normal white and red blood cells and

platelets which cause the symptoms of APL.

How common is APL?

APL is considered a rare disease as it affects about two people in every million per year. It represents approximately 10% of all cases of AML. APL affects men and women equally. There are about 160 new cases diagnosed in the UK each year.

APL can be diagnosed at any age. The median age at diagnosis is 51 years.

What causes APL?

The exact cause of APL is not known. However, APL is commonly associated with the PML-RARA gene which is sometimes called the hallmark of APL. The PML-RARA gene is created during a person's lifetime and is not passed on to children. It is the result of a translocation between chromosome 15 and chromosome 17. 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. In the case of APL, the PML gene on chromosome 15 and the RARA gene on chromosome 17 swap and fuse to become the PML-RARA gene.

A small number of APL patients (less than 10%) do not have the PML-RARA gene but have other rarer gene fusions. APL can also develop after receiving certain types of chemotherapy or radiotherapy given for another cancer. This type of APL is called treatment-related APL. The risk of developing APL is highest around three years after the treatment for the prior cancer.

Symptoms and diagnosis of APL

Symptoms of APL

The most common symptoms of APL are similar to those seen in other acute leukaemias and are caused by the bone marrow failing to produce enough normal blood cells. Most patients will experience shortness of breath and tiredness with slight exercise, which is caused by a shortage of red blood cells (anaemia). Anaemia means the body cannot supply enough oxygen to muscles and other tissues. Infections are common and often persistent because of a deficiency in neutrophils, a white blood cell that normally helps to fight infection.

Bruising and bleeding are often seen, and can range from slight bruises in the skin to serious internal bleeding. This is caused partly by a shortage of platelets, but also by a condition called disseminated intravascular coagulation (DIC). Platelets help the body to form a clot at the site of bleeding, so when there are too few, this may cause bruising or slight bleeding.

DIC may happen in other forms of leukaemia but is rare. It affects about four out of five patients with APL. DIC happens when the body forms clots and breaks down clots inside the blood vessels. This uses up the clotting factors in the blood and this can lead to severe, even life-threatening, bleeding. Fortunately, modern treatments are very effective in blocking DIC.

To summarise, common symptoms and their causes are:

- **Anaemia** – breathlessness, easily tired
- **Low white blood cells** – frequent and persistent infections
- **Low platelets** – bruising and/or bleeding
- **DIC** – bruising/bleeding which may be very severe

Diagnosis of APL

If APL is suspected, you'll have a set of tests to confirm the diagnosis. If you're diagnosed with APL, you will have further tests to determine the right treatment for your cancer. It's important that you know and

understand your diagnosis, so you can ask questions and be fully informed of what to expect.

Full blood count

A blood sample is examined using an automatic cell-counting machine and by examining a stained film under a microscope. The cell-counter will usually indicate that there are large numbers of abnormal white cells in the blood. The appearance of the stained blood cells is usually very typical of APL.

Bone marrow examination

If the results of a blood sample show that you may have APL, or another form of leukaemia, a bone marrow sample will be taken, usually from the hip bone. This is done under a local anaesthetic and does not take very long. The bone marrow sample is important to confirm the diagnosis and also for comparison with later samples to show how APL is responding to treatment.

Additional investigations

There is a test that looks for an abnormality called PML-RARA. This is an abnormal fusion gene – PML and RARA are two genes which are normally found on different chromosomes. In APL, the two chromosomes swap over part of their DNA, which joins the PML and RARA genes together. This test is important because the main drugs used to treat APL work directly on the PML-RARA gene. In the very rare cases of APL without the gene, other treatments can be used.

There are also a number of other special investigations that can be done to confirm the diagnosis of APL and to help in planning treatment. Unlike most forms of leukaemia, treatment of APL often starts before all the tests are completed.

These tests may be repeated from time to time during your treatment. This is to find out how the APL is responding to treatment.

"Lots of things were thrown at me in a short amount of time but the sentence I clung on to was 'we are waiting confirmation, but the leukaemia we think you have carries a high cure rate. We should know by the morning.' I remember thinking, please, please let it be that one."

Treating APL

APL is treated in a very different way from other forms of AML; if a patient with APL is given standard treatment, there is a risk of serious problems with their clotting system. Fortunately, it is usually very easy to tell the difference between APL and other types of AML.

APL usually responds very well to treatment and patients with this form of leukaemia have a good chance of being cured.

APL can affect people of any age, but this booklet is about APL in adults. If you are a parent of a child with APL, you should ask their specialist about the differences in treatment and outlook for children.

If you have any concerns, contact your haematologist.

Treatment options

Treatment of patients who are suspected of having APL should be treated immediately, even before the diagnosis is made, because they can quickly develop potentially life-threatening bleeding or blood clotting symptoms. A firm diagnosis of APL using genetic testing can be performed later, and treatment can be discontinued if APL is not confirmed.

Patients with APL are generally subdivided into the following two groups according to their white blood count as treatment recommendations can differ for each group:

1. **Low- to intermediate-risk:** patients with a white blood cell count of 10,000 cells per microlitre of blood or less.
2. **High-risk:** patients with a white blood cell count of more than 10,000 cells per microlitre of blood.

First-line treatment

First-line treatment for APL includes all-trans retinoic acid (ATRA), which is an active by-product of vitamin A. ATRA blocks the effect of the PML-RARA gene that prevents the promyelocyte cells maturing into normal white blood cells (differentiation). ATRA is not a chemotherapeutic drug and is called a differentiating

agent. It is given in combination with another drug in patients with APL to prevent any drug resistance.

ATRA can sometimes be given with chemotherapy drugs called anthracyclines. Anthracyclines, such as daunorubicin and idarubicin, interfere with the DNA and reproduction of white blood cells, including the leukaemia cells. ATRA is given as a capsule, while anthracyclines are given intravenously.

In 2018, NICE approved a drug called arsenic trioxide (ATO) for the first-line treatment of APL in previously untreated patients, with low- to intermediate-risk disease and patients whose APL has returned (relapsed) or did not respond to chemotherapy (refractory). ATO is also a differentiating agent and acts in a similar way to ATRA.

Because differentiating agents have less side effects to chemotherapy drugs, especially anthracyclines, the combination of ATRA and ATO alone is a preferred first-line therapy, particularly as studies found it to be at least as effective as the combination of ATRA and anthracyclines, if not more so, with a reduced risk of disease relapse.

Induction treatment

To achieve remission (induction therapy), the 2019 guidelines from the European Leukaemia Network (European LeukemiaNet) recommend the following regimens:

Low-to-intermediate risk patients: ATRA and ATO

High-risk patients: Both of the following regimens achieve similar results; however, ATO is not approved for high-risk patients by NICE as yet.

- ATRA and ATO plus a cytoreductive chemotherapy such as cytarabine. Cytoreductive means that the chemotherapy reduces the number of cells.
- ATRA plus anthracyclines. The most frequently used regimen being called AIDA.

The treatment for APL that has developed as a consequence of

Treating APL (cont.)

prior chemotherapy is normally similar to APL associated with the PML-RARA gene, although your doctor may choose to use a different drug in this situation.

In addition to induction treatment, patients with APL require supportive care in the form of blood product transfusions to maintain the platelet count and the blood clotting indicators as normal as possible and to prevent the risk of bleeding. Blood chemical levels (particularly potassium and magnesium which are important for electrical conduction in the heart) will be monitored closely. Sometimes it is necessary to also give potassium and/or magnesium supplements.

Consolidation treatment

To consolidate remission in patients who have not received chemotherapy-based treatment, four courses of ATO and seven courses of ATRA are recommended. This can usually be given as an outpatient.

For patients who received ATRA and chemotherapy regimens, two to three courses of anthracycline-based chemotherapy should be given for consolidation therapy. This is usually given as an inpatient.

Minimal residual disease and molecular monitoring

After consolidation treatment has been completed, a bone marrow molecular assessment is recommended to confirm complete response (CR) with negative minimal residual disease (MRD):

- **Complete response (CR), also known as complete remission** – This is based on morphological remission, meaning that a patient has a normal blood count, a normal development of blood cells in the bone marrow and their blast count is at less than 5%.
- **Minimal residual disease (MRD)** - The presence of small, but measurable, numbers of leukaemia cells in the blood of a patient who does not have any clinical presentation of a blood cancer (i.e. the patient is in remission). If there are

no detectable leukaemia cells found, then a patient is said to be MRD-negative.

This is then to be followed by routine monitoring for certain patients, depending on their CR and MRD results as well as their risk classification:

- Routine monitoring may not be necessary for low-risk patients with CR with negative MRD after consolidation.
- MRD monitoring every three months for up to three years after consolidation is recommended for high-risk patients.

Maintenance treatment

For low- to intermediate-risk patients, maintenance treatment after consolidation with ATO and ATRA is not recommended, but for high-risk patients on ATRA and chemotherapy who are showing clinical benefit, maintenance may be initiated with tablets for two years.

Second-line treatment

First-line treatment is generally successful in most patients with APL. However, for patients who haven't gone into first remission or who have relapsed, second-line treatment options are available.

Relapse or being refractory to first-line treatment can occur in any patient with APL, regardless of whether they have been treated with ATRA with ATO or ATRA with chemotherapy. However, these events are uncommon in low- to intermediate-risk patients.

The second-line treatment you have for relapsed or refractory APL will depend mainly on which first-line treatment you were given. If you have had ATRA with ATO as first-line treatment, then you will receive ATRA with chemotherapy, and vice-versa (you will be given ATRA with ATO if you had ATRA with chemotherapy as first-line treatment).

In young, fit patients, an autologous stem cell transplant can be performed. With an autologous stem cell transplant, you are given intensive chemotherapy to destroy all the leukaemia cells. However, as the

Treating APL (cont.)

chemotherapy will also kill your own bone marrow cells, you are given a transplant of your own healthy stem cells which were collected before the intensive chemotherapy. However, in patients who were responding well to ATO and then relapsed, a transplant is not always necessary.

Side effects

Side effects in patients with APL when treated with ATRA and ATO include differentiation syndrome, pseudotumour cerebri, hyperleukocytosis, heart rhythm disturbance (prolongation of QTc interval) and signs of toxicity in the liver.

Side effects common with anthracyclines are cardiomyopathy (disease of the heart muscle) and secondary leukaemia.

Other side effects include:

- Nausea and vomiting
- Alopecia
- Bone marrow suppression (resulting in low levels of white blood cells, red blood cells and platelets)

Differentiation syndrome

Previously known as retinoic acid syndrome (or RA syndrome), differentiation syndrome can affect some patients being treated with ATRA, ATO or both. It occurs when these differentiating agents start allowing the promyelocyte cells to mature. The exact mechanism involved is unknown but it is thought to be an inflammatory response.

Differentiation syndrome is most likely to happen during the first three weeks of treatment and is more common in patients with a high white blood cell count.

Symptoms include:

- Fever
- Cough
- Breathing difficulty
- Fluid in the lungs
- Weight gain
- Fluid in the tissues
- Kidney damage

Differentiation syndrome can usually be treated with steroids. Treatment with differentiating agents may be temporarily reduced or suspended, but this would only happen if the differentiation syndrome is very severe, which is rarely the case. Most current ATRA with ATO regimens include preventative steroid treatment.

Pseudotumour cerebri

Pseudotumour cerebri is seen with both ATRA and ATO, and is more common when these two differentiating agents are given together. Pseudotumour cerebri is a non-harmful increase in pressure in the skull and unrelated to any tumour.

Symptoms include:

- Headaches
- Swelling of the optic disk
- Double vision
- Confusion

If it occurs, pseudotumour cerebri can be treated with steroids and by decreasing the dose of ATRA.

Hyperleukocytosis, leukostasis and disseminated intravascular coagulation

Hyperleukocytosis is said to occur when the white blood cell count is greater than 100,000 per microlitre. This highly increased white blood cell count often leads to complications such as leukostasis and disseminated intravascular coagulation.

Leukostasis occurs when the excessive numbers of white blood cells make the blood viscous and prone to clotting.

Disseminated intravascular coagulation (DIC) is a condition where the small blood clots develop throughout the bloodstream and block blood vessels. Because the increased clotting uses up all the platelet cells and clotting factors to control bleeding, patients are prone to excessive bleeding. This is a very serious complication which should be treated as soon as possible.

In order to prevent DIC from developing, patients with hyperleukocytosis should be

Treating APL (cont.)

treated with anthracycline-based chemotherapy. The use of the chemotherapy drug hydroxycarbamide should be considered for treatment of leukocytosis (a white blood cell count greater than 100,000 per microlitre) in patients treated with combination therapy of ATRA and ATO to keep the white blood cell count low.

Heart rhythm disturbances

Some patients treated with ATRA, ATO or both might show heart rhythm disturbances such as prolongation of the QTc interval on their electrocardiogram (ECG) which may cause heart problems, especially for patients with existing heart disease. Therefore, ECG monitoring before and twice a week throughout treatment is recommended. For patients who show prolongation of the QTc interval on their ECG, treatment should be stopped and restarted when the heart rhythm disturbances subside.

What happens if these drugs do not work for me?

For the majority of patients who do not respond to first-line treatment with ATRA and ATO, or ATRA with ATO and cytarabine in high-risk patients, or patients who relapse at a later point, a good response to second-line treatment is generally achieved with ATRA and anthracyclines, or ATRA with ATO if they received ATRA with anthracyclines as a first-line treatment. While relapse or being refractory to first-line treatment can occur in any patient with APL, this is an uncommon event in low- to intermediate-risk patients.

In the event of requiring second-line treatment, it is worth considering an autologous stem cell transplant in patients who are fit enough to withstand the procedure.

Your consultant will be able to discuss these treatments with you, and any circumstances in which they might become necessary.

Prognosis of APL

For patients who are diagnosed and start treatment promptly, the outcome of APL is very good. Since the introduction of the combination of ATRA and ATO, cure rates for APL of approximately 90% for low- to intermediate-risk and 85% for high-risk patients have been reported.

An important remaining issue is early death which occurs in approximately 10-15% of patients who do not receive immediate treatment. Efforts in this area are focusing on education to improve prompt diagnosis and early management of patients with suspected APL.

Patients who are older, male, have problems with their kidneys (measured by serum creatinine levels) and increased levels of fibrinogen (protein involved in clotting the blood) have a worse prognosis.

Living with APL

Being diagnosed with an aggressive form of leukaemia like APL can be difficult, both practically and emotionally. This chapter will talk about both of these aspects.

Emotional impact of APL

Being told you have cancer can be very upsetting. It can be especially difficult with acute leukaemia as you often get ill suddenly and have to start treatment quickly. There is usually very little time to take in information and start to cope with it.

APL is a rare condition and, because of this, you may need emotional, as well as practical, support. Being diagnosed with a rare disease can impact you emotionally at any point of your journey. It is likely that you will experience a range of complex thoughts and emotions, some of which may feel strange or unfamiliar to you. It is important to know that these feelings are all valid and a normal response to your illness.

Looking after you

Following a diagnosis of APL, you may wish to make changes to your lifestyle. It's important to know your limits and not try to change too much at once. Exactly what you can do will vary and will depend on the treatment you have had, and how fit you were before your leukaemia. Adopting a healthy way of living is about making small, manageable changes to your lifestyle.

You can find more information about living well with leukaemia on our website: **www.leukaemiacare.org.uk/support-and-information/information-about-blood-cancer/living-well-with-leukaemia/**

Diet

Diet plays an important part in coping with cancer and its treatment and recovery. A well-balanced diet can help you feel stronger, have more energy, and recover quicker.

If you're having treatment, you may notice that you lose weight, or your taste or appetite changes. This may be due to the side effects of your treatment

including a sore mouth or nausea and sickness.

Once your treatment has finished though, you should begin to feel better and be able to eat a normal diet. This can take a while after intensive treatment.

Exercise

With some of the side effects you may be experiencing, such as fatigue, the idea of getting out and being active may be the last thing you want to do, but it is important to try and stay as active as possible. This will help to make you feel better and reduce some of the symptoms or side effects you may be experiencing. Speak to your clinical nurse specialist about exercises that may be suitable for you.

Infection

One of the most common problems following a diagnosis of APL is infection. When you have APL, your body is not able to fight infections as well as normal – this is known as immunosuppression. If you have immunosuppression, ordinary infections may occur more often and be more severe or longer lasting. You may also get ill from infections with germs that normally live in your body without causing problems, but which grow more rapidly when your immune system is not working – these are called opportunistic infections.

The neutropenic diet can help protect patients with weakened immune systems.

If you think you may have an infection, you should contact your doctor straightaway. Common symptoms of infection include:

- Fever – a raised temperature (38°C or higher)
- Aching muscles
- Diarrhoea
- Headaches
- Excessive tiredness

The signs and symptoms of infection may be less obvious when you have APL, so if you are in any doubt it is best to contact your doctor and ask for advice.

Living with APL (cont.)

You can help to reduce the risk of infection by taking some simple precautions:

- Wash your hands frequently, especially after using the toilet, and also if you have touched something like a door knob or banister which can be contaminated with lots of germs.
- Try not to spend unnecessary time in crowds; especially if there is an epidemic of flu or another illness.
- You should be very careful to follow food safety advice, such as cleanliness in the kitchen and not keeping food after use-by dates.

Vaccines

Vaccinations may not work as well when you have leukaemia, but it is still recommended that you have your annual flu vaccine. This will still reduce the risk of getting ill and will offer you some protection.

APL patients should avoid having live vaccines which are used for measles, mumps and rubella (MMR) and shingles. If a vaccine is recommended by someone other than your APL specialist, you should check that it is safe first.

Shingles

If you have previously been exposed to chickenpox, you may develop a painful nerve condition called shingles. Even if it was a long time ago, the virus can live dormant for many years and surface when your immune system is supressed. You may be able to receive a vaccine against shingles but, as it is a live vaccine, you should talk to your doctor about this.

Practical support

Work and finances

Being diagnosed with APL means you will need to start treatment straightaway and so you, or someone you know, will need to contact your employer to inform them of your situation. Your condition will mean that you will need to be at hospital frequently at first and you will need to make the appropriate arrangements with your employer with regard to your working arrangements.

You may need to negotiate a reduction in working hours or make an arrangement with your employer for times when you have to go into hospital or for those times when you may not be well enough to go into work.

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

You could provide them with a copy of this booklet or invite them to download it from the Leukaemia Care website at **www.leukaemiacare.org.uk**

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

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

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

Talking about APL

Talking to your haematologist

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

The following gives advice on working well with your haematologist:

- If it's an initial consultation, take along a list of your current medications and doses, and a list of any allergies you may have.
- If you have a complicated medical history, take a list of diagnoses, previous procedures and/or complications.
- Make a list of questions to take to your appointment. This will help the discussion with your haematologist.
- It can be useful to repeat back what you have heard so that you can be sure that you fully understood.
- Note information down to help you remember what was said.
- Be open when you discuss your symptoms and how you are coping. Good patient-doctor communication tends to improve outcomes for patients.

Other tips:

- Bring someone along to your appointment. They can provide support, ask questions and take notes.
- Don't be afraid to ask for a second opinion – most haematologists are happy for you to ask.

You need to tell your haematologist if...

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

Remember, if you choose to start any form of complementary therapy outside of your medical treatment, consult your haematology consultant or clinical nurse specialist prior to taking it. It is important to understand the difference between complementary therapies, used alongside standard treatment, and alternative therapies, used instead of standard treatment. There is no evidence that any form of alternative therapy can treat APL.

For help with talking to your haematologist, you can access more information about APL, including a section on 'Questions to ask your medical team' at **www.leukaemiacare.org.uk/support-and-information/information-about-blood-cancer/blood-cancer-information/leukaemia/acute-promyelocytic-leukaemia** which features a list of questions that you may want to ask.

Talking to other people

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

"I made a conscious decision to be very open about my illness. Telling family was tough. But I encouraged people to ask questions."

It is probably best to focus conversations on the symptoms that you are experiencing, how the condition affects you and how you feel about it. Often people misunderstand and, unfortunately, it will mostly fall to you to educate them as best as you can. Where possible, it's advisable to let people know what you find helpful and unhelpful, in terms of what others say and do. Often people make assumptions

Talking about APL (cont.)

and do what they think helps. For example, saying you look well, recounting stories of others they know with a similar diagnosis, and encouraging you to look ahead and stay positive isn't always what you really want to hear. In many ways, the more you communicate with them the better.

These points may help you:

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

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

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

Glossary

Acute Myeloid Leukaemia (AML)

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

Anaemia

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

Autologous Stem Cell Transplant

A transplant of stem cells derived from part of the same individual receiving them.

Bone Marrow

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

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.

Chromosome

X-shaped, 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 human cells.

Consolidation

A treatment phase given to remove any leukaemia cells that are still present after induction treatment, as this will help reduce the risk of the leukaemia returning.

Fatigue

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

Genes

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

Induction

The treatment phase intended to kill the majority of the leukaemia cells in the blood and bone marrow, and to restore normal blood cell production.

Lymphoid Cell

A cell originating in the bone

Glossary (cont.)

marrow which will eventually become lymphocytes and antibodies.

Maintenance

An ongoing treatment phase intended to prevent any leukaemia cells from recurring. Lower doses of therapy are given often for years.

Neutropenia

Low numbers of neutrophils, making you more vulnerable to infections.

Neutrophils

White blood cells involved in fighting inflammation and infection, specifically bacterial infections.

Platelets

Small blood cells that help the body form clots to stop bleeding.

Red Blood Cells

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

Refractory Condition

A condition for which treatment does not result in a remission.

Relapse Condition

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

Remission

Remission is said to have occurred when the blood cell counts have returned to normal and there are less than 5% of abnormal, immature leukaemia cells still present in the bone marrow. Complete remission is said to have occurred when there are no leukaemia cells anywhere else in the body.

Stem Cells

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

White Blood Cells (or Leukocytes)

The main role of white blood cells is creating an immune response against both infectious disease and foreign invaders. There are several different types of white blood cells, and each has a different role.

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

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

Leukaemia Care
YOUR Blood Cancer Charity