



**Supportive care**

At all times patients should rely on the advice of their specialist who is the only person with full information about their diagnosis and medical history.

For further advice contact the clinical information team on 020 7269 9060.

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## Introduction

**There has been a steady improvement in survival for patients with leukaemia and related diseases over the last 30 years. Much of this has occurred because of better prevention and management of problems such as infection and bleeding (haemorrhage) and anaemia. Many of these aspects of treatment are similar in patients with different diseases.**

Not all patients will receive all forms of treatment described but most patients are likely to receive at least one of these forms of supportive care.

Although most procedures in supportive care are well standardised, there may be minor differences between hospitals in the way they carry out procedures described in this booklet. In addition to local variations in practice, there may be reasons why a particular patient may require a different or modified form of supportive care. At all times patients should be aware that the members of their healthcare team are the only ones who can assess what is right for them as an individual.

## Blood product support

**All blood cells are made in the bone marrow. Leukaemia and other diseases of the bone marrow typically lead to a reduction in blood cell production. Many patients with leukaemia and related diseases require blood transfusions at some stage during their illness. This is especially true when patients have acute (aggressive) disease or when they require treatment(s) that may affect the ability of the bone marrow to produce blood cells. Patients who receive stem cell transplants will require transfusion support until their transplanted marrow begins producing blood cells.**

Blood supplies in the UK are obtained from unpaid volunteers who donate whole blood at regular intervals. The unit of blood is collected and then usually processed into its separate components — red blood cells, platelets, plasma, blood clotting factors. This is done for two main reasons: patients tend to benefit more when they receive only the blood components that they actually need at the time of their transfusion, and a single unit of blood can benefit several patients when it has been separated into components.

Infection risks are absolutely minimal as all donations are routinely tested for a range of infections that might be spread by transfusion. These include viral infections such as HIV, hepatitis A, hepatitis C and bacterial infections e.g. syphilis. Some common viral infections e.g. cytomegalovirus (CMV) can cause problems for some recipients, who will require specially screened donations. In some situations, for example following stem cell transplantation, the donated blood needs to be irradiated to prevent a condition called transfusion-associated graft vs. host disease (TA-GvHD). Although the risk is lower, certain types of intensive chemotherapy may also be associated with a risk of TA-GvHD; patients in this group will also normally receive irradiated blood products.

In the UK the donated cells are routinely filtered in order to remove white blood cells. This reduces the risk of transmission of a new category of infectious agents, prions. These are thought to be the agents responsible for diseases such as new variant CJD ('mad cow disease'). Transmission of prion diseases by blood transfusion is considered to be extremely rare and only or two instances have ever been reported in the medical literature. As an additional precaution, anyone who has received a blood transfusion in the UK since 1980 is not accepted as a blood donor. Removal of white blood cells from the donated unit also reduces the risk of developing antibodies and this helps to protect against allergic reactions to platelet transfusions.

## Red cell transfusions

Red cells contain haemoglobin which carries oxygen from the lungs, into the blood and through to the tissues. When the number of red cells declines, so does the haemoglobin level in the blood. When the haemoglobin level in the blood falls below normal this is termed anaemia and usually causes fatigue or excessive tiredness. Anaemia can be severe and disabling and may restrict the patient's quality of life to a marked degree. As many as half of all patients with leukaemia or a related condition will be anaemic at the time of diagnosis; many other patients will become anaemic during the course of their illness.

Many patients with leukaemia will require red blood cell transfusions in order to maintain their red cell count and to prevent anaemia. When patients receive large numbers of blood transfusions, there is a possibility that they will start to produce antibodies, which may cause difficulty in finding compatible blood for transfusion. If a patient receives a large number of transfusions (more than 50-100 units) iron within the transfused red cells can accumulate within the body tissues. If iron overload does occur then drugs may be used to help the body excrete the excess iron. To avoid antibody production, and to minimize the risk of iron accumulation, doctors must balance the positive benefit of transfusion against the negative effects. Patients and carers should understand that sometimes doctors decide that it is in the patient's best interests to tolerate a mild degree of anaemia, even if associated with tiredness, in order to minimize the number of transfusions received.

## Platelet transfusions

Patients who are being treated for leukaemia or related conditions are likely to have low numbers of platelets. This is partly a consequence of the disease itself and partly a side effect of treatment. It is standard practice to give platelet transfusions to any patient who has a low level of blood platelets and is consequently at risk of bleeding.

It is not feasible to maintain platelets at near normal levels because transfused platelets have a very short survival in the blood. Frequent platelet transfusions are more likely to cause antibodies to develop, which may make it more difficult to find compatible platelets.

The level at which platelet transfusions are administered will vary according to the clinical circumstances. It is usual to give platelet transfusions every two to three days in an attempt to maintain the platelet count above  $10-20 \times 10^9/l$ . Clinical experience has shown that bleeding problems are very uncommon above this level, even though it is far below normal values. If a patient has to undergo a procedure with a risk of bleeding, for example removal of a tooth, they may receive additional platelet transfusions to achieve a more nearly normal level.

## Erythropoietin for anaemia

**Erythropoietin (EPO) is the chemical signal used by the body to stimulate the bone marrow to produce and release more red cells into the circulation. EPO is made in the kidney and patients with renal disease commonly suffer anaemia due to a lack of EPO. Artificial versions of EPO are now available for treatment of anaemia. EPO treatment has revolutionised the quality of life of patients with kidney failure.**

EPO treatment is also commonly used for treatment or prevention of anaemia in many patients with blood cancers, particularly multiple myeloma, lymphoma and chronic lymphocytic leukaemia. It has been shown to be safe and effective in the majority of patients at improving anaemia due to the disease itself and anaemia arising as a result of treatment.

## Prevention and management of infection

**Patients with leukaemia and related conditions are prone to both bacterial and fungal infections as a result of prolonged low white cell numbers (neutropenia) caused by their illness and its treatment. The ideal solution is to prevent such infections from occurring and there are several measures that can be taken. One of the simplest and most important is hand washing. It is very important that patients should keep their hands as clean as possible and all individuals including staff and visitors who attend to patients must wash their hands prior to touching the patient.**

The British Committee for Standards in Haematology state:

*“Careful attention to hand washing and decontamination before contact with the patient is mandatory for all health care workers and visitors. Patients should be encouraged to challenge staff if hygiene rules are breached.”*

Most patients receiving intensive treatment for leukaemia or a related condition, or receiving stem cell transplantation will be treated in an individual room with en-suite facilities. Flowers or pot plants are not allowed, as these are potential sources of fungi and bacteria. All units treating patients at increased risk of infection will have policies on how such patients should be managed including details of acceptable foods and drinks - it is essential that all food should be free of infection. Most cooked foods are permitted, although it is very important that storage recommendations should be carefully followed. Fruit should be washed and peeled. Salads and uncooked foods are generally best avoided.

Visits should not be allowed by anyone who has evidence of overt infection including viral infections such as colds and flu. Flu is most commonly caught by inhaling airborne virus. It is also possible to transfer the virus by touching the eyes or nose after touching a contaminated surface.

Advice from the Chief Medical Officer to reduce the risk of catching flu (or colds) includes:

- Avoid non essential travel and large crowds of people wherever possible, especially at times of flu outbreaks
- Maintain good hygiene - washing hands frequently in soap and water protects against picking the virus up from surfaces and passing it on
- Frequent cleaning of hard surfaces (e.g. telephone receivers, kitchen surfaces, door handles etc), using a normal cleaning product

## Low Neutrophil Count (Neutropenia)

Neutropenia may increase the risk of bacterial and of fungal infections. Some fungal or bacterial species are very common on healthy people and normally cause no illness (these are known as commensal species). These may cause disease in patients who are neutropenic – this is known as an opportunistic infection.

Patients with neutropenia may not show typical signs and symptoms of an infection. Any patient who develops a high temperature or who becomes generally unwell will be carefully examined and may have special tests to check for infection. When the patient is at home, between treatment courses, they will be advised to contact the hospital if they develop any sign of possible infection – the exact advice may differ between hospitals, but patients will usually be given an advice sheet, indicating the signs and symptoms for which they should be alert.

Patients should call their doctor if they have any of the following:

- Fever above 38°C (100°F) shaking chills or flu-like symptoms
- Sore throat, hoarseness or cough
- A stuffy or runny nose
- Eye or ear drainage
- Burning or pain on urinating

- Burning or pain near the rectum
- Pain, redness or swelling around a surgery or injection site or a wound, pimple or boil
- Sores that heal slowly or fail to heal
- Unusual vaginal discharge or itching

The patient's GP is normally given advice by the specialist on when it is necessary for a patient to be urgently referred to the hospital and when they can safely be treated at home.

## Bacterial infections

If a patient who is neutropenic develops a fever, it is essential that they are immediately given broad-spectrum antibiotic treatment. The initial choice of drug will be made after consultation with local experts in infectious diseases, who will be aware of any local problems of antibiotic resistance. The antibiotic used may be changed once laboratory tests have been carried out to determine which drugs are most effective against the infection. If it is necessary to give intravenous antibiotics, the patient will be admitted to hospital.

Prophylactic treatment is the administration of a drug (in this case an antibiotic) to protect against infection. In some hospitals prophylactic antibiotic treatment is routinely given to all at-risk patients. The potential drawback of this approach is that it may increase the proportion of bacterial infections that are resistant to standard antibiotics.

## Fungal infections

Several of the fungal species, including *Candida* (cause of thrush), may cause disease if the host's immune system is compromised – these are known as opportunistic pathogens. The risk of developing this type of infection is directly related to the severity and duration of neutropenia. Fungal infections often show no specific symptoms or signs and established fungal infections

are very difficult to treat. For this reason it is normal to give anti-fungal treatment early to any patient who has a fever which does not clear up with antibiotic treatment. Anti-fungal drugs are not routinely used for patients with low counts, because they are more toxic than antibiotics and doctors prefer to use them only when the need is clear-cut. Scientific studies are seeking to develop less toxic anti-fungal drugs and also to develop more accurate ways to predict which patients are at greatest risk of developing a fungal infection. This would allow those at most risk to receive prophylactic drug treatment

## **Viral infections**

These may be a particular problem after stem cell transplantation, when a patient's immune system is completely suppressed. One of the most important defences against viral infections lies in minimising exposure; patients who may be at risk should avoid crowds, especially during the 'flu season'. They should tell friends and relatives not to visit if they have symptoms of a cold or flu.

When people have a minor virus infection of the nose or throat it is common for this to lead to a bacterial infection in the same region. This can be very dangerous to immunosuppressed patients as bacterial infection may spread to the lungs (pneumonia) which may be very difficult to treat. It is for this reason that any patients who is known to be immunosuppressed should respond promptly to any signs of a respiratory infection. If such an infection is treated promptly, whilst only affecting the nose and throat, it can usually be effectively treated as an outpatient. If pneumonia develops this may be a life-threatening illness; and even when it is less severe, patients may require inpatient treatment.

One of the commonest viral infections is chickenpox (varicella) with most people in temperate climates having immunity by the age of 14 years. When chickenpox occurs in childhood it is usually quite a mild illness. Many people

are exposed (and acquire immunity) without ever becoming ill. When it occurs in adult life it is almost always more severe, but is rarely dangerous to people with a normal immune system. When people are immunosuppressed, exposure to chickenpox can lead to a serious illness. Relatives and friends must be made aware of this, especially those who have young children. It is important to avoid contact with anyone who has chickenpox or has been in contact with a person with chickenpox. The specialist caring for the patient will be able to advise on when contact is safe. If an at-risk patient comes into contact with chickenpox, their specialist should be contacted immediately, as there are post-exposure treatments which can reduce the risk of serious illness, but these must be applied promptly to be effective.

## **Growth factors to prevent infection**

Body chemicals called growth factors regulate normal blood cell production. An artificial version of a growth factor called G-CSF is now available for treatment of patients. G-CSF stimulates the bone marrow to release neutrophils, which are essential to fight infection. There has been extensive research into the use of growth factors to prevent infection in patients being treated for blood cancers. The conclusion has been that there is no basis for routine use of growth factors after the first stage of treatment (remission induction). It is suggested that they may be of value later in treatment (after consolidation treatment).

Although there are guidelines on the use of growth factors in patients who have fever and are neutropenic (febrile neutropenia), practice still varies between different hospitals. Some units recommend use of growth factors for any patient with febrile neutropenia, while others only use them for patients who are seriously ill. Newer growth factors are longer acting, which may reduce both the cost and the inconvenience of administration of growth factors.

## General precautions against infection

There are certain precautions patients can take to minimize the risk of infection; these are particularly important if a patient is neutropenic, but are advisable at all times.

If you are a patient who is at risk of infection, or you are a carer for someone who is, you should observe the following advice.

### 🔸 Skin care

- Wash hands frequently with soap and water, especially
  - 🔸 Before preparing food or eating
  - 🔸 After going to the bathroom
  - 🔸 Before touching your eyes
  - 🔸 Before touching the inside of your nose and,
  - 🔸 After blowing your nose, coughing or sneezing
- Try to avoid cuts
  - 🔸 Be careful when using knives, nail clippers, scissors or tools
  - 🔸 Use an electric razor rather than a straight, or even safety, razor
- Protect your feet
  - 🔸 Wear slippers or shoes, even indoors. Wear slippers when in hospital
- Protect your hands
  - 🔸 Wear heavy gloves when digging in the garden or working around thorny plants
  - 🔸 Wear oven gloves when handling hot dishes or pans
  - 🔸 Take particular care to avoid burns when cooking or ironing

### 🔸 Other precautions

- Mouth care
  - 🔸 Use a soft toothbrush or oral swab to clean teeth after meals and at bedtime
  - 🔸 Do not use ordinary toothbrushes, dental floss or toothpicks, as these may damage the mouth
  - 🔸 Ask your dentist for advice on how to safely clean your mouth
- Hygiene
  - 🔸 Avoid constipation – use stool softeners and drink plenty of fluids so your stool is soft and moist
  - 🔸 Women should use sanitary towels during their periods, not tampons or douches and should always wipe from front to back after using the bathroom
- Medication
  - 🔸 Ask medical advice before taking aspirin, ibuprofen or any other medicine which might hide signs of infection by lowering a fever. Aspirin and related drugs could also cause bleeding by interfering with platelet function
  - 🔸 Ask your specialist before taking any medication which has not been prescribed
  - 🔸 Paracetamol is a safe all purpose pain killer

### 🔸 Diet

- Patients should avoid
  - 🔸 Soft cheeses
  - 🔸 Raw or undercooked meat, poultry, eggs, fish and shellfish
  - 🔸 Unpasteurised milk and chilled fruit juices
  - 🔸 Raw sprouts, like alfalfa

## Mucositis

**Some chemotherapy drugs can damage the cells that line the digestive tract from the mouth to the anus. This leads to inflammation and soreness that can be very painful. This is called mucositis. When this occurs in the mouth it is called stomatitis.**

- Measures to minimize the risk include:
- Drink lots of fluid. Try diluting fruit juices; the less acidic ones are less likely to sting the mouth
- Use a very soft toothbrush, such as one designed for children, this will make cleaning your teeth much easier
- Ensure you use your mouthwashes as prescribed from the hospital and avoid alcohol-containing mouthwashes. You will probably be given antibacterial and antifungal mouthwashes. If your mouth is painful you may be prescribed one with a local anaesthetic in it; ask your specialist team for advice
- Avoid very hot drinks; often the cooler the drink the more soothing you will find it
- Avoid heavily spiced foods
- Alcohol and smoking will irritate your mouth so try and limit your intake wherever possible
- If your mouth has become very sore you may need to consider a soft diet such as ice cream, rice puddings, mashed potatoes etc
- If you wear dentures leave them out as much as possible (unless your team advises you otherwise) as this will prevent them chafing your gums

- ✦ Hot dogs and luncheon meats unless reheated to steaming hot
- ✦ Refrigerated pates and meat spreads
- ✦ Refrigerated smoked fish and pre-cooked seafood, such as shrimp or crab
- Food preparation
  - ✦ Always clean hands, utensils, food contact surfaces, fruit and vegetables
  - ✦ Cook foods to a high temperature to kill micro-organisms
  - ✦ Separate raw, cooked and ready-to-eat foods when shopping, preparing or storing foods
  - ✦ Chill (refrigerate) perishable food promptly and defrost frozen foods properly
  - ✦ Ask someone to peel fresh fruit for you so that you are not handling it
  - ✦ Salad ingredients must be carefully washed
  - ✦ Meat and poultry should not be rinsed, as this can spread bacteria onto working surfaces

## Management of pain

**Thankfully, pain is not a major problem for most patients with blood cancer. Patients who do experience pain should always report this to their care team so that it can be properly assessed and the precise cause identified and appropriately treated. Pain is commoner in certain conditions, for example multiple myeloma, when pain may arise because of infiltration of the bones by the disease.**

The most important principle in managing pain is to identify the cause and to treat this appropriately. If the pain is due to infiltration by the underlying cancer, such as involvement of the skeleton by myeloma, then the appropriate management would be treatment directed against the cancer such as chemotherapy or localised radiotherapy.

Sometimes pain arises from infection affecting a particular part of the body e.g. skin infection. Occasionally pain arises as a result of bleeding due to low platelets. The cause of the pain must be accurately established so that it can be appropriately treated.

The World Health Organisation (WHO) recommends a three-step analgesia 'ladder' to optimise pain control. Step one of the ladder is the use of nonopioid drugs for mild pain. Patients on step one who do not benefit from nonopioids, or patients with moderate pain, will move to step two which is treatment with mild opioids. Those patients who have severe pain or whose pain fails to respond to step-two strategies will be treated with strong opioids – step three. Adjuvant medications are additional drugs used to treat certain pain types and reduce the dosage of opioids, for example the use of bisphosphonates to control bone disease in multiple myeloma. The WHO strategy recommends that all options at each step should be used before moving to the next step.

Opioid drugs tend to cause stronger side effects and are used when milder drugs are not effective in pain control. The underlying principle is that no patient should experience significant pain problems because of inadequate analgesia.

It is very important that the pain medication should not interfere with platelet transfusion therapy or with other treatments. Aspirin and non-steroidal anti-inflammatory drugs are common painkillers but they should be avoided in patients with blood cancer particularly if they have low blood platelet counts. Long term use of non-steroidal anti-inflammatory drugs, e.g. ibuprofen can also lead to bleeding from the stomach and sometimes can cause kidney damage. Long term use of these drugs should always be discussed with the care team.

Morphine and morphine derivatives are very effective painkillers. It is very uncommon to become addicted to morphine if it is being used specifically to treat pain.

## Tumour lysis syndrome

**Under normal conditions, large numbers of cells die each day, as they have reached the end of their lifespan. When large numbers of cells are killed, for example by cancer treatment, they release their contents into the blood stream. This leads to high blood levels of breakdown products. The body cannot readily deal with such high blood levels.**

The kidneys are particularly vulnerable to damage from tumour lysis syndrome. Drugs such as allopurinol or urate oxidase (Rasburicas™) can protect patients from high uric acid levels. A simple precaution against the other aspects of tumour lysis syndrome is to make sure the patient has plenty of fluids (hydration). Doctors will carefully monitor blood chemistry – a rising potassium level is the biggest risk, if this is severe patients may require dialysis (artificial kidney treatment). A high white count increases the risk of tumour lysis syndrome; the risk of this can be reduced by using a technique called leucophoresis to lower the white count before starting chemotherapy.

## Leucophoresis

**A high white count is a particularly serious event with acute leukaemia. It increases the risk of tumour lysis syndrome and it may lead to blocking of blood vessels in the brain. It is much less likely to be a serious problem in patients with long-standing or chronic leukaemias. Leucophoresis is rarely required unless the white cell count is greatly in excess of  $100 \times 10^9/l$ .**

Leucophoresis is a procedure for physically removing white blood cells from the blood. Blood is passed from the patient into a machine, which separates the blood into red cells, white cells and plasma. The white cells are removed. The red cells, platelets and plasma are returned to the patient. The procedure allows the white cell count to be rapidly reduced. This procedure is usually used in conjunction with chemotherapy and allows the rapid reduction of the thickness (viscosity) of the blood.

In the case of one particular form of acute leukaemia (acute promyelocytic leukaemia) leucophoresis is associated with a risk of severe haemorrhage and is not used for this reason.

## Counselling

It is increasingly common for haematology units to work alongside counsellors. A diagnosis of blood cancer can have very significant implications for individuals and their families. The diagnosis will raise many issues regarding employment, financial considerations, time away from work and time away the family. A serious illness such as a blood cancer can have a significant impact on relationships. It can be very helpful in the circumstances to talk to an individual who is professionally trained as a counsellor. The main role of the counsellor is to be someone with whom the patient and the family can discuss non-medical issues arising from their illness. Counsellors work directly in the patient's interest and work independently of, but alongside, the physicians and nurses. Counsellors can often help to improve communications between the patient and their family and between the patient and the care team. Counsellors can also be particularly helpful in liaising with social services and charitable organisations.

## Summary

One of the most important developments in improving survival from blood cancers has been the ability to prevent and to effectively treat problems such as bleeding and infection. Treatment of these problems is known as supportive care. Supportive care is also of great importance in care of patients who are being treated palliatively rather than with curative intent.

The following patient information booklets are available free of charge from Leukaemia Research. You can download them from our website or request copies by phone or post (see form inside):

Leukaemia and Related Diseases

Acute Promyelocytic Leukaemia

Adult Acute Lymphoblastic Leukaemia (ALL)

Adult Acute Myeloid Leukaemia (AML)

Aplastic Anaemia (AA)

Bone Marrow and Stem Cell Transplantation (BMT)

Childhood Acute Lymphoblastic Leukaemia (ALL)

Childhood Acute Myeloid Leukaemia (AML)

Chronic Lymphocytic Leukaemia (CLL)

Chronic Myeloid Leukaemia (CML)

Hodgkin's Lymphoma (HL)

Multiple Myeloma (MM)

Non-Hodgkin's Lymphoma (NHL)

The Myelodysplastic Syndromes (MDS)

The Myeloproliferative Disorders (MPD)

Clinical Trials

Chemotherapy – what do I need to know?

Donating stem cells – what's involved?

Donor Lymphocyte Infusion (DLI) – what's involved

Supportive care

The Seven Steps – Blood & Bone Marrow Transplantation

Young Adults with a blood cancer – what do I need to know?

Jack's Diary: an illustrated children's book to help young patients understand and deal with blood cancers, treatment and life changes

**Leaflets on a range of associated blood disorders are also available**

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