leukaemia & blood cancer NEW ZEALAND

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APLASTIC ANAEMIA

An information sheet for patients, families and whānau

WHAT IS APLASTIC ANAEMIA?

Aplastic anaemia is a rare blood condition that occurs when your body stops producing enough new blood cells. The word 'aplastic' means the body's inability to create new cells. This means that tissues in the body cannot grow or replicate. Anaemia is when the number of red blood cells (more specifically, the haemoglobin found in red blood cells that carry oxygen) is lower than normal. There are different types of anaemia, aplastic anaemia is one of them.

Blood cells are made in the bone marrow, which is the spongy material inside your bones. In your bone marrow there are cells called blood stem cells. Blood stem cells create the new blood cells in your body. As they mature they become functioning red blood cells, white blood cells or platelets. These three different types of blood cells each have important roles:

- Red blood cells transport oxygen from the lungs to all the cells in the body.
- White blood cells fight infection, bugs and germs.
- Platelets are responsible for stopping bleeding and bruising.

In aplastic anaemia, the bone marrow fails to produce enough red blood cells, white blood cells and platelets.

Approximately 10 people are diagnosed with aplastic anaemia in New Zealand each year. Aplastic anaemia is more commonly diagnosed in people between ages 10 and 20 years old, and over 40 years. There is some specific information at the end of this fact sheet if you are a young person with aplastic anaemia or a parent/guardian of a child with aplastic anaemia.

The exact cause of aplastic anaemia is unknown. We do know that it is not contagious and cannot be passed from person to person. Some risk factors for aplastic anaemia have been identified, this is explained in the next section.

What is acquired vs hereditary aplastic anaemia?

Acquired aplastic anaemia

Acquired aplastic anaemia is more common in children and adolescents, however it can begin anytime in life. Most cases are considered idiopathic, which means they have no known identifiable cause.

Some cases of acquired aplastic anaemia may be caused by chemotherapy and radiation from prior treatments for other diseases. It has also been linked with certain drugs, pregnancy and viral infections (for example, hepatitis B, Epstein-Barr and HIV).

Hereditary aplastic anaemia

Hereditary aplastic anaemia can be passed down through the genes from parent to child or can be caused by a new gene mutation present from birth. It is usually diagnosed in childhood and is less common than acquired aplastic anaemia.

Some inherited conditions (called bone marrow failure syndromes) increase the likelihood that a person will develop the hereditary form of aplastic anaemia. These types of aplastic anaemia can only be diagnosed with specialised testing. These very rare disorders include:

- Congenital amegakaryocytic thrombocytopenia
- Dyskeratosis congenita or other telomere disorders
- Fanconi anaemia
- Shwachman-Diamond syndrome.

What are the symptoms of aplastic anaemia?

The symptoms of aplastic anaemia are a result of low amounts of normal blood cells in the body. These may vary from person to person and will depend on what type of blood cells are low. These symptoms are not specific to aplastic anaemia but occur in any condition where there is a significant deficiency of all blood cells.

Common symptoms from a low red blood cell count:

- Chest pain
- Dizziness
- Fatigue
- Headaches
- Irregular heartbeat
- Lack of energy
- Pale skin
- Shortness of breath while exercising.

Common signs of infection caused from a low white blood cell count:

- Coughing up yellow or green phlegm
- Fatigue or extreme tiredness
- Fever (high temperature of 38°C degrees Celsius or above)
- Shivering or rigors.

Common types of infection in people with aplastic anaemia may include:

- Cellulitis (a skin infection causing redness, swelling and tenderness that can spread)
- Chest infections (including pneumonia)
- Shingles (a rash with tiny blisters, usually localised to one side and area of the body)
- Urinary tract infections (UTI or bladder infection)
- Viral colds and influenza (head cold or flu).

Common symptoms from a low platelet count:

- Bruising easily
- Frequent or severe nose bleeds or bleeding gums
- Minor cuts or injuries that take a long time to stop bleeding
- Red or purple pinhead-sized spots on your skin called petechiae (pe-tee-kee-i)
- Unusually heavy periods in women.

How is aplastic anaemia diagnosed?

Aplastic anaemia is diagnosed by examining samples of your blood and bone marrow. You will need to have a blood test and a bone marrow biopsy.

Blood test

A blood test, called a full blood count (FBC) or complete blood count (CBC), is where a sample of blood is taken from a vein and sent to a laboratory for investigation. The doctor will be able to see how many red blood cells, white blood cells and platelets you have and if these levels are within normal range or are low.

Bone marrow biopsy

To confirm a diagnosis, you will need to undergo a bone marrow biopsy. This involves a doctor taking a sample of bone marrow, usually from the back of the iliac crest (hip bone), and sending it to a laboratory for examination under a microscope. This is needed to rule out other blood conditions or blood cancers. In aplastic anaemia, the bone marrow contains fewer blood cells than normal. See the LBC website for more information on having a bone marrow biopsy.

Other tests

You may need additional tests, usually a combination of blood tests and imaging tests (x-rays, scans and ECG), to look for a possible underlying cause of the aplastic anaemia and assess your general health. These tests are important because they provide a baseline set of results regarding your disease and general health. They may also be used to help select the best treatment for you.

How is aplastic anaemia managed?

The management and treatment of aplastic anaemia may vary from person to person.

Aplastic anaemia can be divided into different groups (classifications) depending on how severely the blood counts are affected. The group you are in will determine the type of treatment that you need. The different groups are called 'non-severe', 'severe' and 'very severe'.

If you have very severe aplastic anaemia then you will need to start treatment. Your blood counts will

be extremely low, which can be life-threatening if not treated. You may have to go into hospital until your condition improves.

If you have non-severe aplastic anaemia you may not initially need treatment. You will be monitored by your doctor and have regular blood tests to see if there is any change in your symptoms and/or blood counts. This is called active monitoring or 'watch and wait'. See the LBC website for a fact sheet on active monitoring.

Management for aplastic anaemia may include:

- Blood transfusions
- Treatment and prevention of infections
- Stem cell transplant
- Immunosuppression drugs
- Other drugs.

Blood transfusions

Blood transfusions are an intravenous (IV) infusion of healthy blood cells donated by another person. Blood transfusions do not cure aplastic anaemia but they do relieve symptoms by providing blood cells that your bone marrow isn't producing. Blood transfusions are usually given in an outpatient department/day ward in the hospital. Transfusions are very safe, although you will need to be closely monitored throughout the transfusion in case of any reaction. If your doctor recommends that you have a blood transfusion they will give you more information about the procedure, any risks involved and give you the opportunity to ask any questions.

The two main types of blood transfusions are:

- Red blood cell transfusion – usually administered over 2-4 hours.
- Platelet transfusion – usually administered over 30 minutes.

Treatment and prevention of infections

Most people with aplastic anaemia are at an increased risk of infection because of their low blood counts.

It is important to let your doctor know immediately if you have any signs of infection as they will want to start you on antibiotics. Infection that is left untreated can quickly get worse and you may deteriorate rapidly. Make sure you know who to call overnight or during the weekend if you were to develop an infection.

You may need to take medications/drugs to help prevent infections. These are called prophylactic antibiotics. Your doctor will let you know if you need to start taking any drugs and how long you need to take them for.

Stem cell transplant

A stem cell transplant (also called a bone marrow transplant) is currently the only curative treatment for aplastic anaemia. A stem cell transplant is a high-risk treatment option and a lot of things are taken into consideration before it is offered by your haematologist.

The aim of a stem cell transplant is to rebuild your bone marrow with healthy stem cells, usually from a donor (called an allogeneic stem cell transplant). The donor may be your sibling or an unknown volunteer who has been genetically matched to you. These donated stem cells replace your immune system with the immune system of the donor.

You will be given high doses of radiotherapy and/or chemotherapy to destroy your stem cells. You then receive donor blood stem cells through an intravenous (IV) drip to replace the ones that were destroyed. You are required to stay in hospital during the whole procedure and recovery.

See the LBC website for more information on stem cell transplants, including picture books for children.

Immunosuppression drugs

Immunosuppression drugs affect the function of the immune system. The aim of this treatment is to reduce the damage being done to the blood stem cells in the bone marrow and reduce or eliminate the need for blood transfusions. This type of treatment can weaken your general immune system and make you more susceptible to infections.

The most common immunosuppression drugs used to treat aplastic anaemia are ATG (anti thymocyte globulin) and cyclosporin.

ATG is a drug made of antibodies (derived from horse or rabbit) which suppress the immune system. It is given as an IV (intravenous) infusion directly into your bloodstream. You will need to be in hospital while you are having this treatment so you can be monitored closely for any reactions. It usually takes about three to six months before ATG starts to work and slowly improves your blood counts.

Cyclosporin is a tablet that also suppresses the immune system and is taken for at least six months (often longer) after the ATG finishes. Most people taking cyclosporin do not experience many side effects. You will need to have regular blood tests to make sure that you are on the correct dose and that the drug is not causing any damage to your liver or kidneys.

Other treatments

Aplastic anaemia caused by radiation and chemotherapy treatments for other conditions usually improves once you complete those treatments.

Pregnant women with aplastic anaemia are treated with blood transfusions. For many women, pregnancy-related aplastic anaemia improves once the pregnancy ends. If that doesn't happen, then treatment is still necessary.

Self-management

If you have aplastic anaemia it is very important to take care of your general health and make good lifestyle choices. These include:

- Eating well
- Keeping active
- Getting enough sleep
- Reducing stress.

It is also important that you:

- **Rest when you need to.** Anaemia can cause fatigue and shortness of breath, even with mild exertion. Take a break and rest when you need to.
- **Avoid contact sports.** Because of the risk of bleeding associated with a low platelet count, avoid activities that may result in a cut or fall.
- **Protect yourself from germs.** You can reduce your risk of infection with frequent handwashing and by avoiding sick people. If you develop a fever or other indicators of an infection, see your doctor immediately.

Coping and support

Tips to help you and your family/whānau cope better with your diagnosis of aplastic anaemia include:

- **Research your disease.** The more you know, the better prepared you'll be to make treatment decisions.
- **Ask questions.** Be sure to ask your health care team about anything related to your disease or treatment that you don't understand. It may help to record or write down what they tell you.
- **Seek support.** Ask family/whānau and friends for emotional support. Consider joining a haematology support group, it may be helpful to talk to others coping with the disease.

It is important to let your health care team know how you are doing, especially if you are not coping well. They can support you in a variety of ways.

Leukaemia & Blood Cancer New Zealand (LBC) facilitates support groups around New Zealand. Contact an LBC Support Services Coordinator to find out more information about support groups and any other help you may need.

Adolescents and young adults (AYA) and aplastic anaemia

If you are a young person with aplastic anaemia your diagnosis may have brought on a variety of emotions and feelings. This is normal. There may be some specific challenges you are faced with and need support to help manage. These challenges may include:

- Change to education (school or tertiary study)
- Financial stress with change in your ability to work
- Change in appearance or altered self-esteem
- Sexual development and fertility
- Psychological changes which may include anxiety and/or depression
- Feeling isolated from friends, family/whānau or your community.

It is important that you feel supported and have someone to talk to if you have any concerns or need help. Your health care team is available if you have any questions or need help. For example:

- Social worker – can help you manage the practical impact such as advice about managing at home, employment or school.
- Psychologist – specialises in helping you manage the emotional challenges such as stress, anxiety and depression.
- Clinical nurse specialist (CNS) – works closely with you and the rest of your health care team to coordinate treatment and help you manage symptoms of aplastic anaemia and side effects of treatment.

You can contact an LBC Support Services Coordinator for support and information. It is also important to have a good network of friends and family/whānau to help support you. It is ok to ask for help.

Looking after your child with aplastic anaemia

Parents cope with a child's diagnosis of aplastic anaemia in different ways and there is no right or wrong reaction. Hearing that your child has aplastic anaemia is extremely distressing and can trigger a range of emotional responses. It is not uncommon to feel upset, angry, helpless and/or confused.

It is not easy to tell a child about a diagnosis of aplastic anaemia. The amount of information will vary with the child's age and level of intellectual and emotional development. Your child's health care team and LBC Support Services Coordinator can help you navigate what and how to include your child in education around their diagnosis, treatment and recovery.

Older children are generally capable of understanding their illness and how this might impact their life. They are understandably very concerned about any potential changes to their appearance, social interactions with friends, and not being able to attend school and hobbies. Encourage them to talk about how they are feeling and give them the opportunity to express their concerns, and to provide them with accurate and relevant information.

Many parents find that their child's behaviour regresses while they are sick or in hospital. This is normal. It is important to re-establish rules and boundaries when possible to help them cope better and feel more secure and relaxed.

Interacting with other children (including siblings) is an essential part of a child's development. Between treatments, or time spent in hospital, encourage them to attend some activities with friends and peers. While they are in hospital or unable to socialise with others, encourage them to video conference or phone siblings or friends.

Important information available online

For more information and to download other fact sheets, see our website **www.leukaemia.org.nz**

QUESTIONS & NOTES

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If you would like to get in touch:
Call: 0800 15 10 15
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Mail: PO Box 99182, Newmarket, Auckland 1149
Or visit one of our Support Services offices in Auckland, Hamilton, Wellington, Christchurch or Dunedin