

Polycythaemia vera

Blood cells are created inside bone marrow, and the number of each type of blood cell is carefully regulated to maintain the correct balance. Polycythaemia vera is characterised by the production of too many red blood cells. It is caused by abnormal function of the bone marrow, but the reasons for this remain unknown. Polycythaemia vera is a rare condition that tends to be slightly more common amongst men and people of Jewish ancestry. Estimates suggest that around five people in every million are affected. Typically, the onset of disease is sometime after the age of 50 years, with a slow but persistent progression. In some cases, the person may go on to develop acute myeloid leukaemia. Other risks of the condition include stroke and heart attack, because the blood is thicker than normal and prone to forming clots. There is no cure but treatment can control symptoms. In many cases, a person diagnosed with polycythaemia vera can expect to live for more than 15 years. Other names for this condition include primary polycythaemia and Vaquez' disease.

Symptoms

The symptoms of polycythaemia vera include:

- Headache
- Dizzy spells
- Itching skin, especially after bathing
- Flushing of the skin, especially the face
- Breathlessness
- Phlebitis (inflammation of the vein)
- Vision problems
- Skin rashes
- Blue tinge to the skin (cyanosis)
- Fatigue
- Gout
- Kidney stones
- Enlarged spleen (splenomegaly).

The cause is unknown

Red blood cells carry oxygen, white blood cells are part of the immune system and help fight infection, while platelets help to form clots after an injury. These cells are created from stem cells within the bone marrow. The numbers and ratio of these blood cells are carefully regulated to ensure the proper balance is maintained. However, the stem cells of a person with polycythaemia vera are abnormal and produce more blood cells than is necessary. Most often, the bone marrow makes too many red blood cells. The reasons for this are unknown. Many of the extra blood cells are also abnormal. Since polycythaemia vera is more common among people with Jewish ancestry, a genetic link could be involved.

Further changes in the bone marrow

Over time, the furiously hard-working bone marrow may stop working altogether and the stem cells are replaced by non-functioning scar tissue. This condition is known as myelofibrosis. On the other hand, around one in 10 people with polycythaemia vera develop acute myeloid leukaemia, which is the most common type of leukaemia affecting adults (particularly older people).

Possible complications

Some of the other complications of polycythaemia vera include:

- Thrombosis - the most common cause of death in people with polycythaemia vera.
- Stroke.
- Heart attack.
- Peptic ulcer.
- Gastric bleeding.
- Heart failure.

Diagnosis methods

Polycythaemia vera is diagnosed using a number of tests including:

- Physical examination
- Medical history
- Blood tests, including complete blood count (CBC)
- Vitamin B12 blood test
- Bone marrow biopsy.

Treatment options

Without treatment, around half of all people with symptomatic polycythaemia vera will die in less than two years. There is no cure, but treatment can extend the person's life span by thinning the blood and reducing the risk of blood clots and other complications. Options include:

- Weekly removal of one unit of blood until red blood cells make up less than 50 per cent of the overall blood volume. This procedure is called either a phlebotomy or venesection.
- The continued removal of blood units, as required.
- Drug therapy to suppress the activity of the bone marrow, such as frequent injections of interferon or tablets called hydroxyurea.
- Drugs to reduce the risk of blood clots, such as aspirin or warfarin.

Secondary polycythaemia

Secondary polycythaemia refers to elevated numbers of red blood cells not caused by bone marrow abnormalities. Usually, anything that reduces the amount of oxygen available to the body prompts the increased production of red blood cells. Some of the factors that may cause secondary polycythaemia include:

- Cigarette smoking
- Lung disease
- Heart disease
- High altitudes
- Certain tumours.

Where to get help

- Your doctor.

Things to remember

- Polycythaemia vera is a type of blood disorder characterised by the production of too many blood cells, typically red blood cells.
- It is caused by abnormal functioning of the bone marrow, but the reason for this remains unknown.
- Polycythaemia vera is a rare condition that tends to be slightly more common amongst men and people of Jewish ancestry.
- There is no cure.
- Treatment options include drug therapy and the regular removal of blood units.

This page has been produced in consultation with, and approved by:

Content on this website is provided for education and information purposes only. Information about a therapy, service, product or treatment does not imply endorsement and is not intended to replace advice from your doctor or other registered health professional. Content has been prepared for Victorian residents and wider Australian audiences, and was accurate at the time of publication. Readers should note that, over time, currency and completeness of the information may change. All users are urged to always seek advice from a registered health care professional for diagnosis and answers to their medical questions.

For the latest updates and more information, visit www.betterhealth.vic.gov.au

Copyright © 1999/2012 State of Victoria. Reproduced from the Better Health Channel (www.betterhealth.vic.gov.au) at no cost with permission of the Victorian Minister for Health. Unauthorised reproduction and other uses comprised in the copyright are prohibited without permission.