Polycythaemia Vera (PV)

What this leaflet is about: too many red blood cells, why clotting is the main risk, and how it is treated.

> Quick emergency advice
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> Blood clots in a vein of the leg, the lungs or the brain can be life-threatening and need emergency treatment.
> Sudden breathlessness, chest pain, one-sided weakness or numbness, or sudden blurred vision: seek help the same day.

What is polycythaemia vera?

Polycythaemia vera (PV) is a condition in which the bone marrow makes too many red blood cells. The blood becomes thicker than normal. It is a type of myeloproliferative neoplasm (MPN), which means the cell that makes blood cells grows without proper control.
Almost all cases are driven by a change in a gene called JAK2. This is found on a simple blood test. PV usually develops slowly, often over years, and many people live normal, active lives with it.

How will I feel?

Many people with PV have no symptoms at all. It is often found accidentally on a routine blood test. When symptoms do appear, they are usually:

- A ruddy or flushed face.
- Headaches, dizziness and blurred vision.
- Itching, often worst after a hot bath or after drinking alcohol.
- Tiredness, and sometimes unexplained weight loss or night sweats.
- Gout - a very painful, hot, swollen big toe joint.

Why is clotting the main risk?

Thick blood clots more easily. Clots can form in the deep veins of the leg, travel to the lungs, or form in the brain. This is why low-dose aspirin is often started even when you feel completely well - it is to protect the blood vessels, not to treat pain.

> Seek help the same day for sudden breathlessness, chest pain, a new painful swollen calf, sudden weakness or numbness on one side, or sudden blurred vision.

How is PV treated?

Treatment aims to keep the blood thin enough to flow freely, and to lower the risk of a clot. You will have regular blood tests to guide this.

- Phlebotomy - draining a unit of blood every few weeks to bring the haematocrit down into the target range. This is the mainstay of treatment.
- Low-dose aspirin - usually started once the blood count is under control.
- Interferon (for example Besremi) - given by injection. This is the one treatment that can sometimes reduce the JAK2 change itself, so it is often considered in younger people.
- Hydroxyurea - a tablet when phlebotomy alone is not enough.
- JAK inhibitors such as ruxolitinib - used if PV has developed into myelofibrosis.

Some people also take a medicine to lower uric acid if gout has occurred. Tell your doctor about any new medicine, including over-the-counter ones and herbal remedies, because several interact with these drugs.

Will it change?

PV is a long-term condition that needs monitoring rather than a cure. A minority of people develop myelofibrosis, where the marrow becomes scarred, or acute myeloid leukaemia. Regular blood tests are what detect this early - please keep your appointments even when you feel completely well.

Look after yourself

- Stay well hydrated - aim for about two litres of fluid a day unless you have been told otherwise.
- Keep moving. Regular walking is as useful as more demanding exercise.
- Stop smoking, and keep alcohol to a minimum - both add to clotting risk.
- Wear well-fitting shoes and report any foot ulcer or infection quickly.
- Tell any dentist or surgeon that you take aspirin or a blood thinner.

Where to read more

American Cancer Society and the US National Cancer Institute have public patient information on polycythaemia vera. Ask us for the printed links, or search for the condition on cancer.gov.

When to contact us

If you are worried about yourself, or about the person you are caring for, please get in touch. We would always much rather hear from you early than late. If you have read something in this leaflet that worries you, that is a good reason to telephone us, even if you feel well.

Visit us
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If you cannot reach us, or you are very unwell

Please go to the emergency department of KPJ Ampang Puteri Specialist Hospital, or call the emergency ambulance service. Do not wait for a routine appointment, and do not wait for a reply to an email.

Important medical disclaimer

This leaflet is written for patients and their families and carers. It is general patient education only. It is not a substitute for medical advice, and it is not a substitute for the diagnosis and treatment that your own haematologist, nurse, pharmacist or doctor will give you.

Nothing in this leaflet should be used to change, stop or start a medicine, a test or a treatment. If anything you read here is different from what your healthcare team has told you, follow your healthcare team and ask us about the difference.

Information about medicines names, strengths, test turnaround times and supportive care is current at the time this leaflet was prepared. It may be updated from time to time, and the version printed here may therefore not be the newest one. Please ask reception for the current copy.

If you think that you or someone you care for is seriously unwell, do not wait for an appointment and do not wait for this leaflet to be read. Contact the clinic, or go to the emergency department of KPJ Ampang Puteri Specialist Hospital, straight away.

We Listen. We Care. Evidence-based Medicine.

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