FOM Guide to Disease Management

FAQ on
Myeloproliferative Neoplasms
Focus on Polycythemia Vera

25 YEARS
The Max Foundation
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Friends of Max

FAQ

on

Myeloproliferative Neoplasms

Focus on Polycythemia Vera

Editor's Note

Dear Readers,

This is the first in the series of information booklets on the various blood disorders that come under the classification of MPN.

Polycythaemia Vera (PV) is one such condition, a rare blood cancer that affects the bone marrow causing the blood to thicken. It is sometimes called “Erythrocytosis” which involves having a higher-than-normal concentration of red blood cells (erythrocytes) in our blood.

We commonly come across and hear about low haemoglobin (also called anaemia) in our country. In fact, we always equate a high haemoglobin count with "good and prosperous health". However, excess of anything is bad. Similarly, if the haemoglobin is high, especially in people who are old and have concomitant diabetes or high blood pressure, it can be detrimental and lead to ill-health. Herein, we have compiled a booklet of common questions which come to mind when we hear the term "Polycythaemia vera" or "high haemoglobin" from our doctor.

I am grateful to Friends of Max, for going out of their way in formulating this booklet. This will not only increase awareness of this illness but also resolve many queries of our patients.

- *Dr Sumeet Mirgh,*
  Assistant Professor,
  Adult Hematolymphoid and BMT,
  Tata Memorial Centre ACTREC.

FAQ

What is polycythemia vera?

Bone marrow is the factory in our body which makes red blood (called hemoglobin), white blood (called white blood cells), and platelets. Red blood cells carry oxygen, white ones fight infections, and platelets clot your blood to stop bleeding. Polycythemia Vera (PV) is a condition in which your bone marrow makes too many blood cells, especially red blood cells. When you have too many red blood cells, your blood thickens. This increases your risk for several serious health problems. If polycythemia is not treated, it can be life threatening. Right care can give you a long life.

Is polycythemia vera a cancer?

Yes, it is a blood cancer.

What is the cause for polycythemia vera?

Stem cells are the parent cells in our bone marrow which make red blood cells, white blood cells and platelets. Polycythemia is caused by a change in the DNA in these stem cells. This change happens because of mutation of a protein called Janus Kinase 2 (JAK-2) which causes it to not work the way it should. The change occurs more often in men than women, and mostly in patients over 60 years old.

Can polycythemia vera be genetic or hereditary?

Rarely, it runs in families. So there may be genetic factors.

What are the symptoms of polycythemia vera?

Some people do not notice many symptoms, even after diagnosis.

When there are noticeable symptoms, they may include:

- Headache
- Dizziness
- Trouble breathing when lying down
- Shortness of breath
- Dark spots in vision, which come and go
- Itchiness, especially after a bath or shower
- Red skin, especially on the face (looks like a sunburn)
- Numbness, tingling, burning, or weakness in the hands, feet, arms, or legs
- A feeling of fullness in the upper left side of belly

Most of these symptoms are caused by poor circulation, thickening and clotting of the blood.

What if it goes untreated? / What are its complications?

If polycythemia goes untreated it can cause your blood to thicken. This can put you at risk for a number of problems, including:

- Blood clots, which can develop in important organs of body – brain (stroke), heart (a heart attack), or a blocked artery in your lungs (pulmonary embolism) or in your leg muscles (deep vein thrombosis)
- Painful joint swellings (gout)
- Rarely, there can be a complication called myelofibrosis. This is reflected when your hemoglobin and other cell counts remain persistently low, sometimes needing blood transfusions, along with fullness in left side of

your tummy. This happens over a period of time when your bone marrow gets filled with scar tissue. Similarly, another rare but serious complication called leukemia happens when cancer cells are present in blood. It requires treatment with special chemotherapy medicines.

How is it diagnosed?

Because polycythemia is uncommon and develops slowly, you may have it for several years before it is diagnosed. It usually shows up during a blood test. If the blood test shows signs of polycythemia, your doctor may confirm the diagnosis with other tests, such as a bone marrow test or few special tests called EPO level and JAK2 PCR. EPO (short form of erythropoietin) is a hormone in your blood which tells your bone marrow to make blood cells. People who have PV have very low amounts of it. JAK2 PCR study is done on a blood sample, and results are usually available within 10-14 days.

JAK-2 PCR test report is positive in approximately 95% of patients with PV.

What is bone marrow test?

As per your doctor's decision, you may also need a bone marrow biopsy. The results can show your doctor if your bone marrow makes too many blood cells. For this test, your doctor will take samples of your marrow, usually from the back of your hip bone. You lie down on a table and get a shot that will numb the area. Then, your doctor uses a needle to take out a small amount of bone marrow. It is an outpatient procedure, which means you don't have to stay overnight in a hospital. You can get it done in a clinic, a hospital, or your doctor's office.

How is it treated?

Polycythemia cannot be cured. Right treatment can help you manage this disease for many years. The goal of treatment is to make your blood thin. This will help prevent symptoms and complications. If you don't have many symptoms, you might not need treatment for PV right away. Your doctor might prefer to keep a close watch on you.

Treatments include:

- **Drawing blood** (also called phlebotomy). This is often the first treatment. Your doctor or a blood bank personnel takes blood from your vein so you have fewer blood cells. It's just like donating blood. After it's done, your blood will be thinner, and it will flow more easily. You will usually feel better, too. Some symptoms will ease, like headaches or dizziness. Your doctor will decide how often you need phlebotomy. Some people with polycythemia do not need any other treatment for many years.
- **Medication to thin the blood**, such as aspirin. This keeps platelets from sticking together. That makes you less likely to get blood clots, which in turn makes heart attacks or strokes less likely.
- **Medication to reduce the number of red blood cells** you produce. Commonly used is a capsule called Hydroxyurea.
- Rarely, **if Hydroxyurea is not working well**, your doctor may give you another tablet called **Ruxolitinib** to **control number of cells**.

What precautions should I take?

Polycythemia affects your circulation. The most important things you can do, then, are to try to improve your circulation and protect your skin.

Follow these recommendations:

- **Light exercise** like walking to improve your circulation and decrease the risk of blood clots.
- **Quit smoking.** Tobacco narrows your blood vessels and increases the risk of clotting.
- **Care for your skin.** Poor circulation can make sores heal slowly. Keep your skin moist with lotion, and try not to scratch.
- **Bathe or shower** in cool water if warm water makes you itch
- **Avoid extreme temperatures.** Poor circulation can make you more vulnerable to extreme temperatures.

Are there any emergency complications which I should be aware of?

Visit your nearest hospital immediately if you notice any of the following -

- Sudden numbness or weakness of your face, arm, or leg
- Sudden difficulty speaking
- Sudden chest pain
- Sudden dizziness or loss of balance
- Sudden severe headache

These could be symptoms of a clot in the vital organs of your body, which need immediate attention.

Remember – It is common to worry when you find out that you have cancer. Remember that everyone is different and that all cancers are not the same. With the support of your doctor, family, friends, and other people who have polycythemia vera, you will be in the best position to manage it.

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Notes

Notes

Together we share & learn

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