LEBEN MIT
POLYCYTHAEMIA VERA

POLYCYTHAEMIA VERA HAS MANY FACES

Patient information

English

Dear Patient,

A diagnosis of polycythaemia vera brings with it uncertainty and, above all, many questions. What does this rare disease mean? How does it develop? What are the symptoms and what are the possible risks? And how can the disease be treated?

This brochure is designed to provide you with scientifically sound answers to these important questions. Once the risks of PV and the treatment options are clear in your mind, you will definitely feel less threatened by the disease because then you will be aware of the positive prospects offered by modern medical treatment.

You will also know then that you can help to keep the disease well under control and that you can lead a normal life. Being aware of any changes in how you are feeling and discussing new symptoms with your doctor are the very things that will help to keep the PV in check.

Another tip: use the card at the end of the brochure to get more free information on PV.

We wish you an interesting read and all the best!

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Physical stamina

Quality of life

The disease

What is PV?

Polycythaemia vera: a rare bone marrow disease

Have you or has someone close to you been diagnosed with the disease polycythaemia vera (PV)? Then you may already know that PV is classified as a chronic blood cancer in which the formation of new blood cells in the bone marrow is interrupted.

Polycythaemia vera belongs to the group of “rare diseases” (RD)[1] and can occur at any time of life. Every year, up to 1,500 people in Germany develop polycythaemia vera.[2] It usually affects older people (median age at diagnosis 65 years), but can also occur earlier.[1] Important to know: Nowadays, PV can be successfully treated using various means and with medication.

What happens during polycythaemia vera?

This disease of the blood-forming system means that too many blood cells are formed in your bone marrow. All types of blood cells may be affected: red and white blood cells and platelets. This overproduction affects the red blood cells in particular, resulting in the blood becoming thicker.[1]

You can read more about the occasionally serious consequences on page 10.

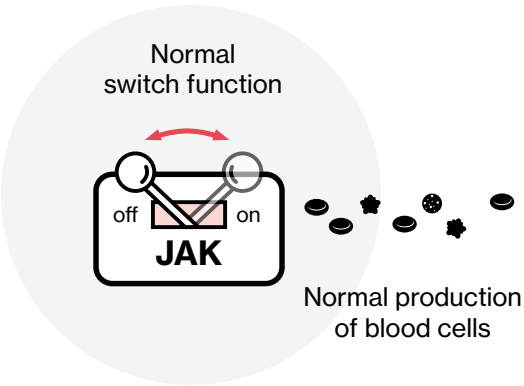
Enzymes switch to overproduction: the Janus kinases (JAK)

You are probably wondering why production of the blood-forming cells gets out of control at all. Today we know that almost all patients with polycythaemia vera have a genetic change in their blood-forming cells: the mutation of the enzyme Janus kinase 2 (JAK2).[1] This protein controls biochemical processes in the body.

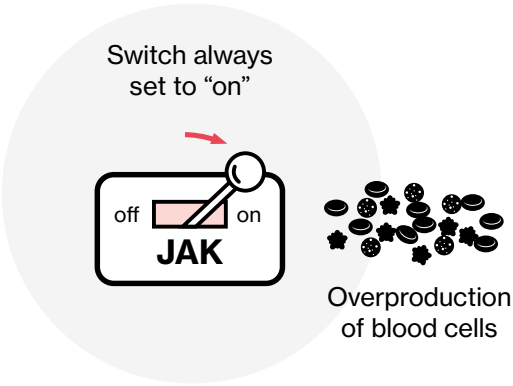
The enzyme JAK2 plays an important role in the blood-forming cells: normally, it switches the multiplication of blood-forming cells on or off. However, because of the JAK2 enzyme mutation, this faulty switch is permanently set to “on”. The result is uncontrolled overproduction of blood cells – especially red blood cells, but the number of white cells and blood platelets can also be increased.[1]

How the JAK1/2 switch works

Healthy person/Normal JAK function



PV patient/JAK overactivation



Why is polycythaemia vera dangerous?

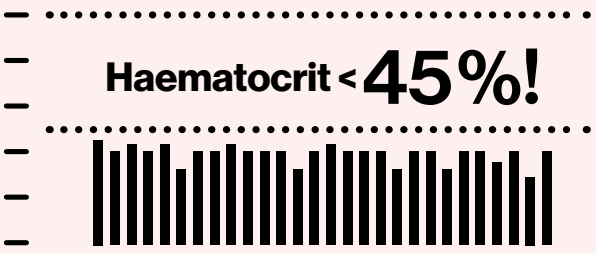
Thicker blood, higher risk: heart attack and stroke

What are the risks when you have PV? What makes polycythaemia vera so dangerous is that, if left untreated, it can cause a fatal heart attack, stroke, thrombosis[1] or a pulmonary embolism. The reason for this is that your blood becomes thicker and more viscous because it contains more and more red blood cells and thus solid components. Haematocrit is the name for the part of the blood that is made up of these solid components.

The haematocrit in your blood should not exceed 45 %[1], because above this the risk of blood clots forming is greatly increased. For this reason, your doctor will check your haematocrit levels at regular intervals and possibly introduce measures to reduce them.[1]

Briefly

Your haematocrit levels must not exceed 45 %

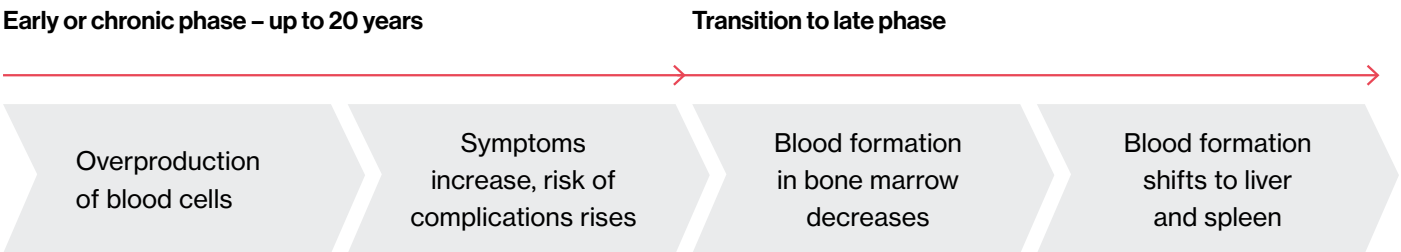


How does PV progress?

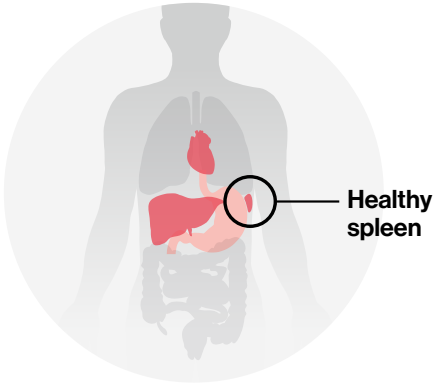
You cannot predict exactly how polycythaemia vera will progress in your case. Progress varies from person to person. However, there is a general trend of progression in the development of polycythaemia vera – in two phases.[1] At an early stage, many patients feel almost free of symptoms. Symptoms such as extreme tiredness (fatigue) occur only gradually. Since the overproduction of blood cells can increase without treatment, the risk of thrombosis[1,4], heart attack, and stroke also grows.

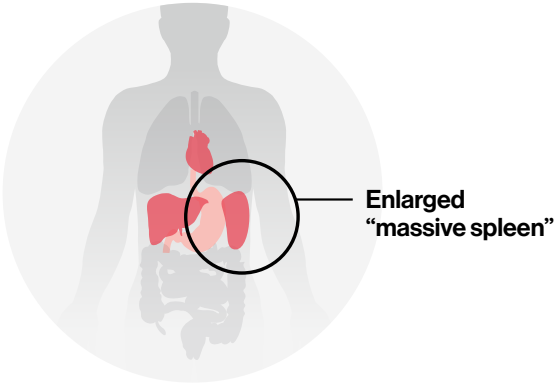
Later in this phase, so-called **cytokines** may increasingly trigger symptoms. Cytokines are messenger substances that are formed as an immune response where inflammation is present. They release a concentration of additional immune cells (leukocytes) at the site of the inflammation and "activate" them. Following their activation, these immune cells also form cytokines in order to intensify the immune response. This "snowball effect" can then trigger the symptoms.

The most common symptoms are intense itching and severe fatigue with loss of physical stamina (details on page 21). The good news is that if polycythaemia vera is diagnosed early, your doctor can successfully treat both blood cell overproduction and the symptoms and stabilise the disease. With the right treatment, it is now possible to achieve almost normal life expectancy.



The late phase – fewer blood cells and spleen enlargement





The risk of complications such as a heart attack or stroke increases further. You may also experience new or increased symptoms caused by cytokines such as extreme fatigue, itching, concentration disorders and pain.

During the late phase, blood cell overproduction becomes underproduction[1]: the bone marrow develops fibrous tissue[3,4] and can form fewer and fewer new blood cells. As blood cells are also part of the immune system (white blood cells) and blood clotting (platelets), you become more susceptible to infection and tend to bleed more easily. If the number of red blood cells also falls further, you may become anaemic. As a result, your physical stamina levels may continue to decline.

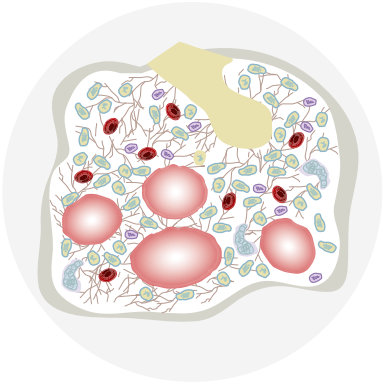
In this phase, the liver and spleen take over blood formation through the colonisation of stem cells from the bone marrow. The spleen, in particular, may increase in size as a result (so-called splenomegaly)[1,5], which can lead to pain in the upper abdomen or an early feeling of fullness at mealtimes.

Late and rare: myelofibrosis and acute myeloid leukaemia

In rare cases, polycythaemia vera may transition to acute myeloid leukaemia (AML)[1,4] or myelofibrosis (MF).[6] If your doctor finds indications of such a transition during regular follow-ups, a new bone marrow examination is useful.

Myelofibrosis: the bone marrow is fibrous

Myelofibrosis (MF) develops when the bone marrow is replaced by fibrous and connective tissue. The special feature of MF is that blood cell production decreases in the bone marrow. As the result of blood formation shifting to other organs (liver or spleen), these organs can become enlarged.



Acute myeloid leukaemia: too many immature blood stem cells

Acute myeloid leukaemia (AML) is a malignant disease. It is characterised by too many immature blood stem cells in the blood and bone marrow. This hinders the normal development of white blood cells and consequently the fight against infection by the immune system.

What are the signs of PV?

Polycythaemia vera – a second opinion is often required

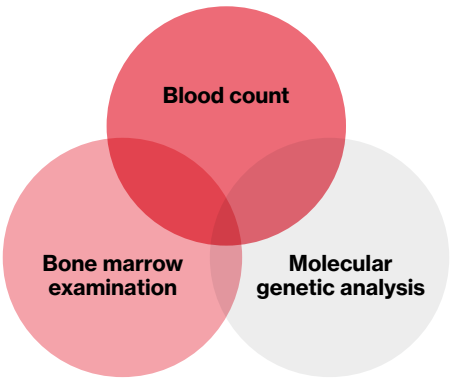
In your case, polycythaemia vera may only have been diagnosed at the second or third doctor's visit.

Many symptoms of polycythaemia vera cannot be properly interpreted at first, as they also occur in other, much more common diseases – these symptoms range from itching and night sweats to headache, dizziness, vision disorders, nose bleeds and/or ringing in the ears.[3]

- Some affected persons come to the doctor with symptoms of circulatory disorders in the hands and feet, with skin redness or blue coloration of the lips[1,3]
- In other patients, polycythaemia vera is determined by chance because the blood values show irregularities
- Thrombosis or a sudden heart attack can also subsequently lead to the diagnosis of polycythaemia vera
- The symptoms of itching and chronic fatigue are often not initially recognised as symptoms of PV at all

How is it possible to get a reliable diagnosis of polycythaemia vera?

As the physical symptoms of polycythaemia vera are often not conclusive, the doctor needs to follow up possible signs using a very targeted approach. If you are suspected to have PV, your doctor or the haematologist (specialist for blood diseases) will perform the following tests: blood count, molecular genetic analysis and bone marrow biopsy.[3]



Blood count: normal blood levels[3]	
Number of red blood cells	4.3–5.6/pl (men) • 4.0–5.4/pl (women)
Number of white blood cells	3.8–10.5/nl
Number of platelets	140–345/nl
Haemoglobin	13–17 g/dl (men) • 12–16 g/dl (women)
Haematocrit	42–50 % (men) • 38–44 % (women)
Erythropoietin	10–25 U/l
Bone marrow examination	
Number/appearance of blood-forming cells	
Molecular genetic analysis	
Gene mutations (especially JAK2 mutation)	

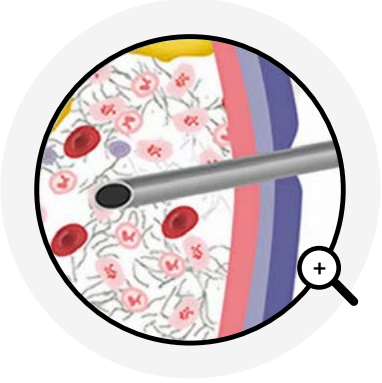
pl = picolitre = 1 trillionth of a litre; **nl** = nanolitre = 1 billionth of a litre; **U/l** = units per litre

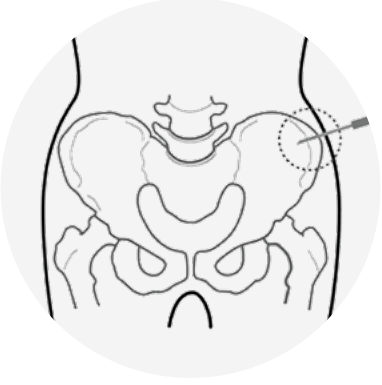
Why do you need a blood count?

The blood count shows your doctor any irregularities, such as increased haematocrit and haemoglobin levels. This can be an initial indication of polycythaemia vera. If blood platelet and white blood cell levels are also elevated, this may reinforce the suspicion. A further molecular genetic analysis will show whether you have a genetic defect in the JAK2 gene (the JAK2 gene contains the blueprint for the JAK2 enzyme – page 9). Elevated blood levels and a JAK2 gene defect already mean that your doctor will be able to diagnose PV with a relative degree of certainty.

The bone marrow biopsy

Bone marrow is usually taken from the iliac crest. This is done to determine cell density and cell types.[5]





Why is a bone marrow biopsy so important?

In order to be absolutely sure whether it is really a case of polycythaemia vera, an examination of your bone marrow is usually also necessary.[5] Your bone marrow sample shows under the microscope whether and how the blood-forming cells of the bone marrow are changed. In this way, your haematologist can diagnose or rule out polycythaemia vera in most cases.[5] During the bone marrow examination, a doctor takes a small amount of bone marrow directly from the pelvic bone for laboratory analysis.

Is polycythaemia vera hereditary?

You have probably already asked yourself this question. The answer is a clear no. Based on current knowledge, the JAK2 mutation of polycythaemia vera is not hereditary. The defect in the JAK2 gene only develops in the bone marrow over the course of the person's life.[1] Since neither egg cells nor sperm cells are affected by this, people with polycythaemia vera cannot pass on the genetic defect to their children.

Symptoms

Common symptoms

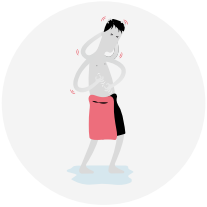
Symptoms that are often overlooked

Perhaps you have experienced the physical symptoms of polycythaemia vera yourself – symptoms that it causes, but which you have not associated with the disease at all. Many patients only experience these symptoms, such as fatigue, itching, or difficulty concentrating, after years have passed, although some may have them at an earlier stage. If they are not recognised as being disease-related, they cannot be specifically treated either.

These debilitating symptoms are caused primarily by so-called cytokines – messenger substances of the immune system that are released in the bone marrow when there is an inflammation present. The symptoms can severely impair your quality of life because they have a negative impact on your physical stamina levels and, consequently, your ability to work to a greater or lesser extent.

Below is an overview of the most common symptoms.[5] Pay particular attention to:

Severe tiredness (fatigue)



Itching and burning of the skin (often increased by contact with water)

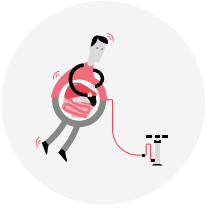
Feeling full quickly during mealtimes

Difficulty concentrating

Fever

Inactivity

Night sweats



Abdominal discomfort

Bone pain

Unintended weight loss

What you can do

If you suffer from polycythaemia vera, it is important that you document all symptoms and tell your doctor about them – even if you get new symptoms. This is the only way your doctor can provide you with the best treatment for your needs! You can record your symptoms using the MPN10 Symptom Assessment Form[7] developed by doctors. You can bring the form with you when you visit your doctor so you can discuss the symptoms.

Skin complaints

When you have polycythaemia vera you also need to pay special attention to your skin because this is where you may experience problems caused both by the disease as well as the treatment (more on treatment objectives and options from page 32 onwards).

Disease-related problems
These problems are caused by the disease itself. This includes the itching (pruritus) already mentioned, especially after bathing/showering, or even painful red areas on your skin.

Treatment-related problems
Treatment-related means that the problems are linked to the use of hydroxyurea, often used to treat PV. Noticeable signs of this are dry skin and so-called actinic keratosis – palpable skin roughness that is a precursor of white skin cancer. The occurrence of leg ulcers (ulcerations; see page 23) may be the result of either the treatment or the disease.

Watch out for: skin alarm signals

You should always keep a regular eye on your skin. Particularly, however, if you are being treated with hydroxyurea for polycythaemia vera.

This is because you may also have reactions to the treatment as well as disease-related side effects such as itching. Some common problems with hydroxyurea are listed here:

Dry skin
The skin is often raw, dull, scaly or chapped, with possible redness and cracks

Inflammation of blood vessels
Usually at first pinhead-sized signs of bleeding on the legs; these can extend over a larger area and look like a bruise

Ulceration
A painful, inflammatory reddened hole in the skin, sharply punched out, it can be due to illness or treatment

Actinic keratosis
Light damage, for example, on your forehead, cheeks or the backs of your hands in the form of palpable raw, scaly, reddish, raised areas on the skin, which can lead to white skin cancer

What you can do

Check your skin regularly and tell your doctor if you see anything unusual.

Circulation problems & thrombosis

As polycythaemia vera is a blood formation disorder, and too many blood cells impede blood flow, the disease can also cause circulation problems in the hands and feet, blue discoloration of the lips and areas of red skin on the face. Headaches, dizziness, impaired vision, nosebleeds and/or ringing in the ears are some of the possible consequences. It is essential that you tell your doctor if you notice any of these symptoms, despite the treatment.

Risk of thrombosis: if thick blood forms clots

Too many red blood cells thicken your blood and increase your risk of suffering thrombosis (blood clots) in the larger blood vessels.
If a clot **blocks an important artery**, this can trigger a **heart attack** or **stroke.**

If a clot **blocks a vein**, deep vein thrombosis may develop that can even lead to a life-threatening pulmonary embolism.

- **Deep vein thrombosis (DVT):** a blood clot in a deep vein, mainly in the legs. Pain, swelling, redness and enlarged surface veins may all be signs of DVT.

- **Pulmonary embolism:** this can occur if the clot caused by DVT dissolves, circulates in the bloodstream and clogs one of the pulmonary arteries. Pulmonary embolism is a serious life-threatening complication.

Actively prevent thrombosis – some tips[5]

- Eat a healthy, balanced diet including plenty of fruit and vegetables, and drink a sufficient amount, at least two litres, of water, tea or other unsweetened drinks every day
- Give up smoking
- Take regular exercise – physical activity can have a positive impact on your symptoms and reduce the risk of thrombosis
- Avoid sitting for long periods, for example, in the car, on long-haul flights or at your desk
- A good way to encourage circulation in your legs is to stand up regularly and walk around a bit

Other complications

Complication spleen enlargement

Another complication is also associated with the blood formation problem in the bone marrow: spleen enlargement. As the bone marrow is increasingly replaced by connective tissue and becomes more and more fibrous at a later stage in the disease, it loses its ability to form new blood cells. The blood formation process moves to the spleen and liver. This leads to spleen enlargement and associated pain in the upper abdomen.

What you can do

Your doctor should perform an abdominal ultrasound scan once a year to determine the exact size of your spleen. The enlargement can also be felt for manually. It is important to treat spleen enlargement as soon as possible!

MPN10 Symptom Assessment Form[7]

Symptoms: signs that the treatment has succeeded

Diseases from the group of myeloproliferative neoplasms (MPN) such as polycythaemia vera (PV) and myelofibrosis (MF) are often accompanied by debilitating symptoms. They can significantly restrict quality of life.

Disease-related symptoms include fatigue, fever, inactivity, itching, night sweats, or a feeling of fullness. In the medical guidelines, these symptoms are significant for achieving treatment objectives or as a prognosis factor. In other words, laboratory results alone do not determine the appropriate form of treatment for you, or the success of the treatment. Your quality of life is also a consideration. Therefore, you should contact your doctor if you notice any of the symptoms becoming worse.

MPN10 Symptom Assessment Form – Documenting how you feel

The MPN10 Symptom Assessment Form helps you to systematically recognise, record, and assess the symptoms of your disease and document them for your doctor.

It was specially developed by experts for myeloproliferative neoplasms (MPN) and is designed to show you and your doctor how effective your current treatment is and how much (or how little) your symptoms are affecting your daily routine.

For this purpose, experts have documented the ten most common symptoms of MPN diseases. The assessment takes place using a scale from 0 to 10, where 0 stands for “not present” and 10 for “worst extent imaginable”. This allows you to assess the severity of your symptoms very precisely and to monitor development by comparing the total weekly scores.

MPN10 SYMPTOM ERFASSUNG

Name:

Datum:

Der MPN10 Symptomerfassungsbogen hilft Ihnen, die Symptome Ihrer Erkrankung zu erkennen und aktiv zu verfolgen.

1. Bitte beurteilen Sie Ihre Erschöpfung (Mattheit, Müdigkeit) **während der letzten 24 Stunden.**
   Kreuzen Sie dazu die Zahl an, die das schlimmste Ausmaß Ihrer Erschöpfung am besten beschreibt.

SYMPTOME	keine Erschöpfung										schlimmste vorstellbare Erschöpfung
Erschöpfung (Fatigue)	0	1	2	3	4	5	6	7	8	9	10

2. Kreuzen Sie bitte die Zahl an, die das Ausmaß Ihrer Schwierigkeiten mit jedem der folgenden Symptome **in der letzten Woche** beschreibt.

	nicht vorhanden										schlimmste erdenkliche Form
Schnelles Völlegefühl beim Essen	0	1	2	3	4	5	6	7	8	9	10
Bauchbeschwerden	0	1	2	3	4	5	6	7	8	9	10
Inaktivität	0	1	2	3	4	5	6	7	8	9	10
Konzentrationsschwierigkeiten*	0	1	2	3	4	5	6	7	8	9	10
Nachtschweiß	0	1	2	3	4	5	6	7	8	9	10
Juckreiz**	0	1	2	3	4	5	6	7	8	9	10
Knochenschmerzen	0	1	2	3	4	5	6	7	8	9	10
Fieber (> 37,8 °C)	0	1	2	3	4	5	6	7	8	9	10
Unbeabsichtigter Gewichtsverlust#	0	1	2	3	4	5	6	7	8	9	10

Berechnen Sie Ihren MPN10 Symptomwert für ein Gesamtbild
Ihrer MPN Symptomlast, indem Sie die angekreuzten Zahlen aufaddieren: **Total** _____

Adaptiert von Emanuel RM et al. Myeloproliferative Neoplasm (MPN) Symptom Assessment Form Total Symptom Score:
Prospective International Assessment of an Abbreviated Symptom Burden Scoring System Among Patients With MPNs. J Clin Oncol. 2012; 30 (33): 4098–4103.
* im Vergleich zur Zeit vor meiner Diagnose; ** besonders nach Kontakt mit Wasser; # in den letzten 6 Monaten

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Ideally, you should fill in the sheet regularly or as agreed with your treating doctor, but before, you visit the doctor at the least. Enter the date and your name.

First, tick the severity of your exhaustion (fatigue) over the past 24 hours.

Tick the severity of your other symptoms over the past week.

Calculate the total symptom score by adding the individual scores together.

Therapy

The objectives of your polycythaemia vera treatment

A treatment that accompanies your life

Polycythaemia vera is a chronic disease. Therefore, standard treatment consists of procedures doctors can use to successfully manage your disease and alleviate symptoms in the long term, enabling you to lead as normal a life as possible.[1] Maintaining your quality of life and physical stamina plays a very important role in this regard.

Avoid complications, alleviate symptoms

The treatment of polycythaemia vera has three important objectives as its focus[1]:

The first and primary objective is to reduce the long-term overproduction of your blood cells (haematocrit level <45%) to improve your blood flow. The aim of this is to prevent complications such as thrombosis, heart attack, or stroke.

The control of clinical symptoms and thus the maintenance of quality of life according to the medical guidelines is another important objective. Untreated symptoms such as being plagued by itching and persistent fatigue can sometimes drain a lot of your strength, reduce your physical stamina levels, and have such an impact on your everyday life that you are unable to work. This is why recognising and alleviating individual symptoms are of significant importance for treatment.

The third objective contributes to the stabilisation of your disease for as long as possible. Therapeutic measures tailored to the progress of your disease, taken at an early stage, are designed to prevent the later transition to other diseases such as myelofibrosis (MF) and acute myeloid leukaemia (AML).

What you can do

Talk to your doctor about what you are expecting personally from the treatment. In your case, better quality of life may play an important role if you are battling symptoms such as fatigue or are plagued by itching on a daily basis. Your doctor can adapt the current treatment if it does not alleviate these disease-related symptoms.

What treatment options are open to you?

First off: there is no universal treatment for polycythaemia vera. Depending on how far your disease has advanced, various methods and medications can be used to

- reduce the number of blood cells and improve the flow properties of the blood,
- slow down blood clotting to avoid blood clots,
- relieve symptoms such as itching and fatigue.

Dilute the blood – bloodletting and ASA

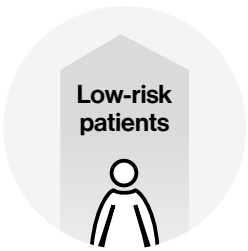
Bloodletting is the first remedy in your treatment regimen – an age-old treatment proven to reduce your haematocrit levels quickly and easily. Bloodletting involves your doctor taking a sample of between 300 and 500 ml of your blood.[1,3] The goal is to keep your haematocrit levels, in other words, the solid components in your blood (red blood cells), below 45 %. This will allow the blood to become thinner and reduce your risk of blood clot formation and complications such as thrombosis, heart attack, or stroke. Your doctor may also prescribe a drug treatment with acetylsalicylic acid (ASA).[5] ASA has the property of “diluting” the blood and thus preventing the clumping of platelets. In this case, your doctor will decide whether you will be able to tolerate ASA.

Mild chemotherapy – reduce the risk of complications

You may be at particular risk of developing complications if you are over 60 years old, for example, or have a history of vascular occlusion. Your doctor may then also prescribe cytoreductive treatment with hydroxyurea or interferon in addition to bloodletting. Cytoreductive treatment is also used to reduce the number of blood cells you have.

Reasons for cytoreductive treatment[1]

- Age ≥60 years old
- Already had vascular occlusions in the past



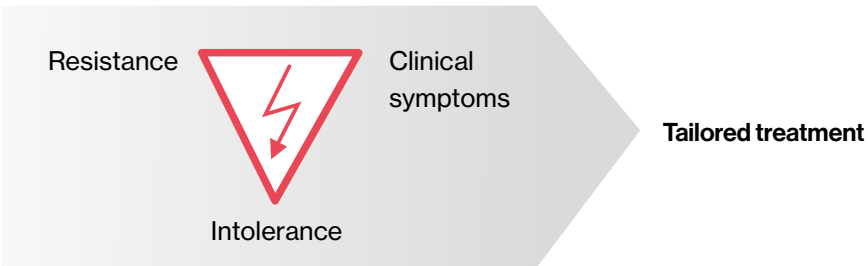
- Poor tolerance or limited feasibility of phlebotomies
- Inadequate haematocrit control (6 or more phlebotomies per year required within 2 years)
- Increase in spleen size
- Progressive blood cell formation
- Inadequate control of disease symptoms
- Insufficiently controllable risks of the cardiovascular system

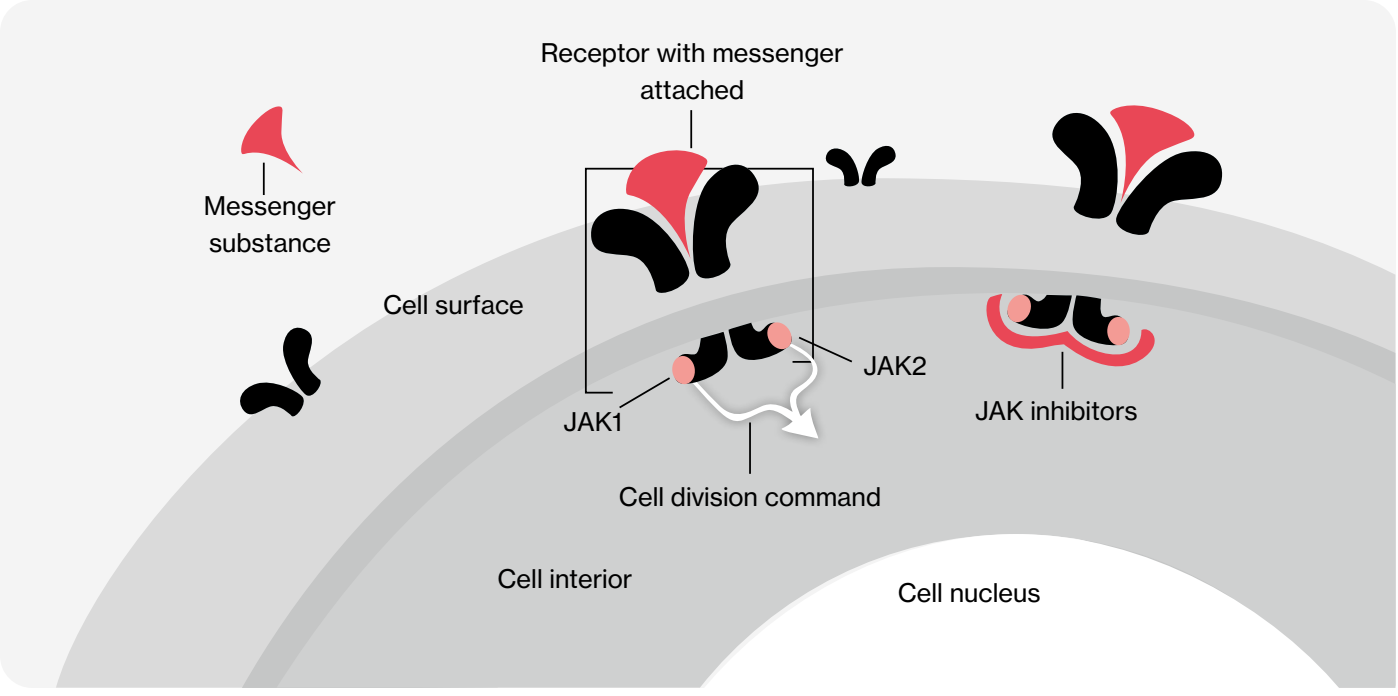
If the initial steps don’t work for you ...

It is certainly possible that the steps just mentioned won’t work for you. There may be various reasons for this, for example, if the medication doesn’t work or you don’t respond adequately to it, or you experience side effects. If this is the case, your doctor will adapt your treatment and introduce different medication – for example, a medicine such as a so-called JAK inhibitor.

Janus kinase inhibitors: directly halt the increase in the number of blood cells

JAK inhibitors are a type of medication that directly inhibits signals in cells. They are called targeted treatment because they work at the location where the disease develops – in the cell. Depending on the disease, they block very specific enzymes that have an important signal function in the cell (see also page 9).





JAK inhibitors used in polycythaemia vera block the activity of the enzymes JAK1 and JAK2.

If JAK2 in particular is halted, the number of new blood cells is also reduced. JAK inhibitors can thus help to achieve the important treatment goal “haematocrit <45 %”.[1,5] In addition, they can alleviate symptoms such as itching and fatigue. JAK inhibitors belong to a group of medications that are generally known as tyrosine kinase inhibitors (TKI).[1,6]

When do changes need to be made to your treatment?

The triggers for adjustments: insufficient effect, intolerance and symptoms

If mild chemotherapy is used to treat you, the medication may need to be adjusted in certain situations. The reasons for this are:

- The medication is not effective or is not effective enough. Treatment with chemotherapeutic agents is designed to reduce the high cell counts in the blood of polycythaemia vera patients. However, it can happen that the treatment is not effective or is not effective enough. This is known as therapy resistance.

- You are not able to tolerate the medication, and experience severe side effects. This is known as intolerance. The medication needs to be discontinued as a result.

- New symptoms occur or existing symptoms worsen. Regardless of whether or not you are being treated with mild chemotherapeutic agents, you may still experience debilitating symptoms such as itching or severe fatigue. Even though the chemotherapeutic agents reduce the cell count in the blood, they do not reduce the debilitating symptoms associated with polycythaemia vera.

In all three cases, there is a need for change. One possible option is targeted therapy with a so-called tyrosine kinase inhibitor (TKI).

Be aware of any side effects or problems tolerating your treatment, as well as possible symptoms, and tell your doctor about them. Your doctor is relying on the information you provide in this respect! In particular, use the MPN10 Symptom Assessment Form to document your symptoms.

You need to pay special attention to your skin, as treatment-related skin problems can also occur that may be caused by polycythaemia vera or treatment with hydroxyurea.

What you can do

Be especially aware of any side effects or problems tolerating your treatment, as well as possible symptoms, such as extreme fatigue, and tell your doctor about them. Your doctor is relying on the information you provide in this respect! In particular, use the MPN10 Symptom Assessment Form to record symptoms.


MPN10 SYMPTOM ERFASSUNG

Name:

Datum:

Der MPN10 Symptomerfassungsbogen hilft Ihnen, die Symptome Ihrer Erkrankung zu erkennen und aktiv zu verfolgen.

1. Bitte beurteilen Sie Ihre Erschöpfung (Müdigkeit, Müdigkeit) **während der letzten 24 Stunden.**
Kreuzen Sie dazu die Zahl an, die das schlimmste Ausmaß Ihrer Erschöpfung am besten beschreibt.

SYMPTOME	keine Erschöpfung									schlimmste vorstellbare Erschöpfung
Erschöpfung (Fatigue)	0	1	2	3	4	5	6	7	8	9	10

2. Kreuzen Sie bitte die Zahl an, die das Ausmaß Ihrer Schwierigkeiten mit jedem der folgenden Symptome **in der letzten Woche** beschreibt.

	nicht vorhanden									schlimmste vorstellbare Form
Schwäche/ Mangel an Energie/ Antriebslosigkeit	0	1	2	3	4	5	6	7	8	9	10
Beschwerden beim Duschen/Baden	0	1	2	3	4	5	6	7	8	9	10
Hautjucken	0	1	2	3	4	5	6	7	8	9	10
Konzentrationsschwierigkeiten*	0	1	2	3	4	5	6	7	8	9	10
Nachtschweiß	0	1	2	3	4	5	6	7	8	9	10
Juckreiz**	0	1	2	3	4	5	6	7	8	9	10
Knochenschmerzen	0	1	2	3	4	5	6	7	8	9	10
Fieber (> 37,8 °C)	0	1	2	3	4	5	6	7	8	9	10
Unbeabsichtigter Gewichtsverlust*	0	1	2	3	4	5	6	7	8	9	10

Berechnen Sie Ihren MPN10 Symptomwert für ein Gesamtbild
Ihrer MPN-Symptomlast, indem Sie die angekreuzten Zahlen aufaddieren: **Total**

Physical stamina

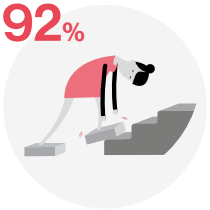
Maintain your physical stamina levels

If your daily life becomes a burden

As a chronic disease, polycythaemia vera brings changes into your life. These can vary in intensity. For example, a number of patients feel quite well for a long time. However, in the course of the disease, the problems can increase to a significant extent in some people. Most patients, for example, experience several symptoms at the same time. The cause is mainly so-called cytokines, messenger substances in the immune system, which are excessively released.

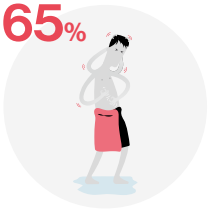
The most common problems[8,10]

92%



Severe tiredness (fatigue)

65%

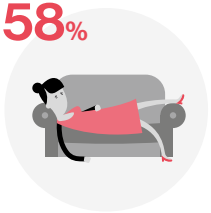


Itching and burning of the skin (often increased by contact with water)

61%

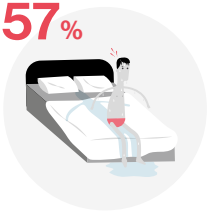
Difficulty concentrating

58%



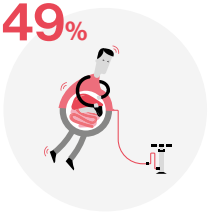
Inactivity

57%



Night sweats

49%



Abdominal discomfort

Consequences that many patients feel

You also actively play important roles in your life – in your partnership, with relatives and friends. However, if the strength to do this is missing, important contacts may dwindle, or hobbies have to be abandoned. Simple everyday activities become challenges as your physical stamina levels gradually decline. Your mobility is restricted, so that travelling and even a car journey become problematic. Surveys among patients with polycythaemia vera show the effects.[9,10]

Limitations in family and social life

63% Slightly restricted

11% Significantly restricted

Impact on everyday activities

48% Slightly restricted

10% Very significantly restricted

Work restrictions **33%**

Illness-related reduction in working hours **15%**

Tackle symptoms – initiate the correct treatment

Bloodletting and, if also necessary, mild chemotherapy with hydroxyurea are successful measures for controlling your blood count. However, they will not help to alleviate symptoms you may have such as fatigue or itching. So-called targeted therapy may help to alleviate your symptoms. However, your doctor first needs to accurately record the symptoms. Therefore, you need to be aware of and document your symptoms.

Quality of life

How to get your quality of life back

What you can do

Keeping a diary helps you both to collect positive experiences and insights as well as to record any changes in your health. This information is also important for your doctor in order to assess the status of your disease.

Stay alert for any changes

How would you rate your overall condition? Your physical well-being? How about your independence in your everyday life? And do you feel up to fulfilling your social engagements? It is precisely these personal assessments that determine your quality of life; your quality of life is nothing more than your personal “feel-good balance sheet”.
Polycythaemia vera has an impact on many aspects of your well-being. Therefore, it is important that you keep an eye on any disease-related changes in your everyday life – but also your positive opportunities. Very consciously note the following:

- whether and how your condition has changed
- what contributes to your well-being and what does not
- whether you notice side effects or symptoms

What exercise can do for you

Physical activity has many positive effects on your overall condition and is recommended by doctors even during rehabilitation after severe diseases. As a polycythaemia vera patient, there are many areas in which you will benefit from physical activity:

- Exercise improves blood flow and reduces the risk of thrombosis
- Exercise has been shown to reduce the risk of heart attack and stroke
- Physical stress promotes blood flow and thus the supply of oxygen to the body
- Moderate training alleviates tiredness and fatigue
- Chronic fatigue in PV can also be positively influenced
- Sport has a mood-enhancing effect, as endorphin secretion is stimulated
- Regular training increases confidence in your own body and promotes self-esteem
- In particular, endurance sports such as hiking and Nordic walking are suitable for improving your physical performance if you suffer from polycythaemia vera

What you can do

Talk to your doctor or a physiotherapist or sports therapist about the ways in which you can undertake regular physical activity, the forms of exertion or types of sport and exercise that are suitable for you, and how often you can be active.

Relaxation helps you take everything in your stride

Chronic diseases are a burden – they bring uncertainty with them that may lead to anxiety and fear. In short: stress. Proven relaxation techniques will help you to manage the consequences of stress actively and more successfully and regain quality of life as a result. With regular relaxation, you can have a tangible impact on:

- tension and cramps
- symptoms of exhaustion
- impaired concentration or memory

Here is a brief overview of the most well-known methods of relaxation for which a relatively wide range of information and courses is also available:

- Autogenic training – promotes physical and mental relaxation with autosuggestive ideas
- Meditation – ensures inner calm and concentration
- Progressive muscle relaxation – the tensing and relaxing of certain muscle groups
- Visualisations – calm yourself with the idea of positive environments and situations
- Yoga – combines physical fitness with meditative elements for more serenity

Healthy nutrition follows simple rules

Especially in chronic diseases, nutritional condition plays an important role: with the right supply of nutrients, you will contribute to your physical and mental function and prevent nutrition-related deficiencies. Conversely, it is a well-known fact that patients whose energy and nutritional requirements are not met often have less stamina and a reduced quality of life. The benefits of a balanced and healthy diet have even been established in cancer treatment. However, you do not need to conduct a specific search for a special diet for your disease – there is actually no such thing!

If you do not have any particular problems eating and drinking, a diet that is also beneficial for healthy people is recommended. This diet follows a few easy-to-understand guidelines and allows a lot of room for personal taste.

What you can do

Learn about the 10 tips on healthy and balanced nutrition from the Deutsche Gesellschaft für Ernährung [German Nutrition Society] and try to implement them: **www.dge.de**
If you have nutritional problems, it is important to talk to your doctor about this.

Helping people understand: four tips on how to involve family and friends

1. No false secrets

Polycythaemia vera is a decisive turning point in your life to date. However, the burdens and the changes that you will need to make in this regard will also be felt by those close to you. It therefore makes sense to inform your family and friends about the disease. This helps prevent misunderstandings or people coming to the wrong conclusions, for example, if you no longer have as much time for others. Give the people you trust a brief description of the disease and, if necessary, tell them about the limitations and problems associated with the disease.

2. Talk about expectations

People from your close circle will want to help you or actively support you. However, advice is not always welcome. Tell them honestly if at any stage you do not want to talk about the disease, or if their help is too much for you. Some people may not know how to deal with you and your disease. An open exchange helps both sides.

3. Accept help

With polycythaemia vera, sometimes you may be less able to cope with your daily routine due to doctors' appointments or physical complaints.

Do not hesitate to accept help, whether it is with household tasks, errands, or if you simply need to talk. It can also be of valuable support if a family member goes with you to the doctor.

4. Partnership and sexuality

In patients with polycythaemia vera, symptoms such as chronic fatigue, itching, pain, fever and night sweats can also limit the need for physical closeness. Tell your partner if you feel physically unwell and as a result, you do not have any need for closeness.

As targeted therapy also works to alleviate symptoms, it can have a positive impact on sexuality. Sexuality is an important component of partnership. If you experience persistent problems, a discussion with the doctor may be useful in order to clarify the exact causes and initiate medical treatment.

Supporting the treatment: three recommendations for your consultation with the doctor

1. Become well informed!

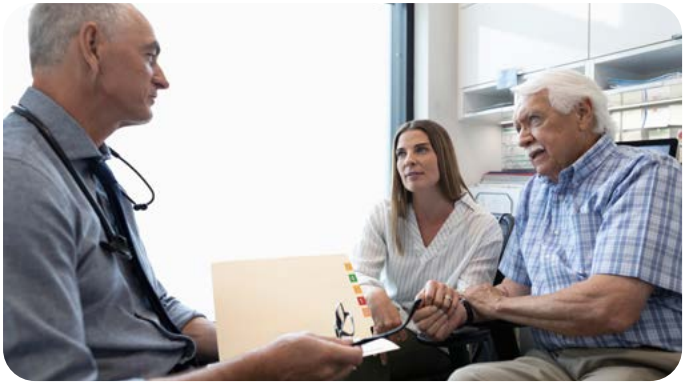
With a chronic disease such as polycythaemia vera, it is particularly important to have a good working relationship with your doctor because the treatment will be with you for life. The more you trust and the more actively you work with your doctor, and the more information you provide about your health, the better placed your doctor will be to help you.

2. Ask specifically

After all, information is important for you, too. A clear explanation about your disease, the treatment options and the objective of the therapy will help you feel you are being well looked after.

Therefore, always feel free to ask if you want to know anything else or there is something you have not understood. Some patients do not do this because they think they have to make do with the information they were given.

The better your doctor understands what wishes or questions you have and what your concerns are, the better they can be addressed! This applies in particular in relation to your experiences during the treatment – in terms of new symptoms, for example: what you tell the doctor can be important in terms of adapting the treatment to your needs.



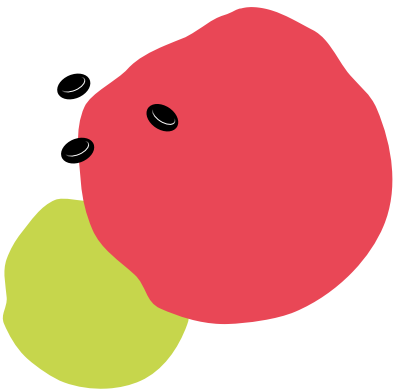
3. Say what is on your mind

Trust grows when you communicate openly with your doctor. The “chemistry” is right if you feel that the doctor understands your concerns. However, if you do not feel that you have a good relationship with the doctor, you need to address the reasons. Tell your doctor if you have doubts, are missing certain information or do not understand a procedure. This is the only way to clarify something.

Perhaps you cannot find common ground on important issues even after you have discussed them. Then you may decide whether you would like to seek a second opinion or look for a doctor with whom the “chemistry” is better.

And finally: a main contact person

Ideally, you should have a constant medical contact person who is familiar with and is up to date with your entire medical history. Your GP or a haematologist specialising in diseases of the blood, who usually also makes the diagnosis in polycythaemia vera cases, can take on this role. You should always tell all the doctors treating you about your disease!



In the workplace: two recommendations for working life

You may be able to continue to pursue your profession. However, you may leave your job or take time out from it and return to your workplace at a later date. Whatever you decide, you will need to tell the people you work with. Below, you will find several tips for communicating with managers and colleagues.

1. What your manager needs to know

You are legally obliged to inform your employer immediately if you are going to be absent from work and to tell your employer how long you are likely to be gone. If you are not fit for work for longer than three days, you need a medical certificate. Depending on the employment contract or the collective pay agreement applicable to you, this can also be requested earlier.

You do not need to provide your employer with the exact diagnosis as it is subject to data privacy regulations. However, in a small company, telling your manager about the disease at an early stage may be the sensible thing to do. This way, he or she can enable you to attend your doctor's appointments and/or adapt your workload and responsibilities to your health situation.

2. The right amount of information for your colleagues

It is, of course, your decision whether or not you discuss your illness with your colleagues. Think about which of your colleagues you would most like to talk to and what you want to say exactly, and you will find it much easier to have a conversation.

Be aware of what support or assistance you need from your colleagues and talk about it openly. In doing so, it is up to you how much information you want to reveal about yourself and how much you want your colleagues to be involved.

It will be time to talk about your illness once visible signs of the disease appear or if side effects occur due to the treatment. A good strategy can be to give your colleagues initially only the information regarding your state of health that is necessary for working together.

Glossary

Links & addresses

Glossary

Acute myeloid leukaemia (AML)
A malignant disease that develops if there are too many immature blood-forming cells in the blood and bone marrow. This influences the development of the white blood cells that fight infection. The “acute” in the name refers to the fact that the disease can progress quickly.

Blood count
A number of values representing different components of the blood. It is possible to specify a value for each blood component, for example for white blood cells, red blood cells or platelets.

Blood clot (or thrombus or thrombosis)
A clot consisting of congealed blood in a blood vessel. If the clot dissolves and moves through the bloodstream, it is called an embolus. If this then closes off a blood vessel completely, an embolism develops.

Blood cells
Red blood cells, white blood cells and platelets.

Bone marrow
The soft, blood-forming tissue that fills the bones and contains immature blood cells, so-called stem cells. These can develop further to become red blood cells, which transport oxygen around the body, white blood cells, which fight infections, or platelets, which assist in blood clotting.

Chronic
A chronic disease is a disease that is long-lasting. The name is derived from the Greek word “chronikos”, which means long-lasting. Myeloproliferative neoplasms, including PV, are regarded as chronic diseases because they progress slowly and many symptoms of myeloproliferative neoplasms are chronic.

Cytoreductive therapy
Treatment with the goal of removing as many of the blood cells that are growing in an uncontrolled way as possible.

Deep vein thrombosis (DVT)
A blood clot in a deep vein in the thigh or leg. The blood clot can break away and move within the bloodstream as an embolus.

Embolism
If a blood clot (thrombus) dissolves in a blood vessel and moves through the bloodstream, it is called an embolus. If this then closes off a blood vessel completely, an embolism develops.

Fatigue (exhaustion)
A feeling of weakness and tiredness that limits your ability to work or do other activities. Exhaustion can be acute and occur suddenly or be chronic and long-lasting.

Haematocrit
The proportion of cellular components in the blood. Red blood cells account for the largest proportion at 96%. Haematocrit values below 45% can reduce the risk of thrombosis and cardiovascular complications.

Heart attack
Death of heart muscle tissue due to lack of blood supply. The lack of blood supply usually results from a complete blockage of one or more coronary vessels that supply the heart with blood.

JAK2
JAK2 is a protein that occurs in all humans and controls blood cell production in bone marrow. It is part of a communication path along which biological messages are transmitted to the cells.

Janus kinase (JAK)
Enzyme in the cell that controls the rate at which cells (such as blood cells) grow and multiply through signal molecules.

Mutation
A change in the genetic material.

Myelofibrosis (MF)
Fibrosis or scarring of the bone marrow, which is accompanied by severe anaemia and often by enlargement of the spleen.

Myeloproliferative neoplasms (MPNs)
A group of blood and bone marrow disorders. Four main types of MPN make up around 95% of all MPNs: myelofibrosis, essential thrombocythaemia, polycythaemia vera, and chronic myeloid leukaemia (CML).

Night sweats
Severe hot flushes that occur at night and lead to severe sweating.

Phlebotomy (or bloodletting)
Procedure for reducing your blood volume.

Platelet (or thrombocyte)
A disc-shaped component of the blood that assists in blood clotting. During normal blood clotting, the platelets clump together (aggregation). Although platelets are often counted as blood cells, they are actually fragments of large bone marrow cells.

Polycythaemia vera (PV)
PV is one of the sub-types of myeloproliferative neoplasms and results from an overproduction of blood cells, particularly red blood cells.

Prognosis
The probable course or probable outcome of a disease.

Pruritus
Itching. Pruritus can occur due to dry or ageing skin, contact reactions of the skin, food allergies, side effects of medications, cancer diseases, kidney or liver diseases, parasites or for unknown reasons.

Pulmonary embolism
Blockage of the pulmonary artery, or a branch of it, leading to the lungs, often triggered by a thrombus.

Spleen
Abdominal organ that acts as a filter for blood and pathogens.

Splenomegaly
Name for an acute or chronic enlargement of the spleen (Greek: Spien).

Symptom
Sign of a disease; often characteristic of a specific disease.

Thrombosis
If a blood clot forms in a blood vessel; this can happen both in an artery and in a vein. The clot itself is called a thrombus (see blood clot).

White blood cells (Leukocytes)
Blood cells that the body forms to fight infection.

Links & addresses

mpn-netzwerk e. V.
c/o Deutsche Leukämie- &
Lymphom-Hilfe e. V.
Haus der Krebsselbsthilfe
Thomas-Mann-Straße 40
53111 Bonn
info@mpn-netzwerk.de
www.mpn-netzwerk.de

Deutsche Krebsgesellschaft e. V.
Straße des 17. Juni 106–108
10623 Berlin
030 32203290
service@krebsgesellschaft.de
www.krebsgesellschaft.de

Deutsche Krebshilfe
Buschstraße 32
53113 Bonn
deutsche@krebshilfe.de
0228 729909 05
www.krebshilfe.de

Deutsche Leukämie- & Lymphom-Hilfe (DLH)
Haus der Krebsselbsthilfe
Thomas-Mann-Straße 40
53111 Bonn
0228 33889200
www.leukaemie-hilfe.de

INKA – Informationsnetz für Krebspatienten und Angehörige
Reuchlinstraße 10–11
10553 Berlin
030 44024079
info@inkanet.de
www.inkanet.de

Krebsinformationsdienst (KID) Deutsches Krebsforschungszentrum
Im Neuenheimer Feld 280
69120 Heidelberg
0800 4203040
krebsinformationsdienst@dkfz.de
www.krebsinformationsdienst.de

Unabhängige Patientenberatung Deutschland (UPD)
Littenstraße 10
10179 Berlin
0180 3 117722
info@patientenberatung.de
www.patientenberatung.de

Fatigue-Informationstelefon (FIT)

06221 2444

Deutsche Fatigue Gesellschaft e. V. (DFaG)

Maria-Hilf-Straße 15
50677 Köln
0221 931159 6
info@deutsche-fatigue-gesellschaft.de
www.deutsche-fatigue-gesellschaft.de

Comprehensive information on polycythaemia vera can be found at:

www.leben-mit-blutkrankheiten.de/pv
www.mpn-netzwerk.de
www.spotlightonmpn.com
www.leukaemie-hilfe.de
www.deutsche-fatigue-gesellschaft.de
www.krebshilfe.de
www.onkopedia.com

Novartis Pharma – Medical Information Service

If you have medical questions about Novartis products or your illness, which is being treated with Novartis products, then you are welcome to contact us, the Novartis Pharma Medical Information Service, at

Telephone: 0911 273 12100*
Fax: 0911 273 12160
infoservice.novartis@novartis.com
www.infoservice.novartis.de

* Mon–Fri from 8:00 a.m. to 6:00 p.m.

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Notes

Life thrives on ideas …
Keep in touch!

Use the accompanying card to request further information on PV. You can also find out what's new in relation to PV on the MPN patient days.

For more information see
www.leben-mit-blutkrankheiten.de/pv

The initiative “Living with PV & MF” offers patients, relatives and interested parties communication and suggestions on central topics of the disease.

By registering, you indicate your interest in receiving further information on the disease polycythaemia vera (PV) or myelofibrosis (MF) and give us your consent to the sending of additional materials.

Data processing as part of the “Living with PV & MF” programme

I agree that Novartis Pharma GmbH, Roonstr. 25, 90429 Nuremberg, collects, processes and uses my above-mentioned data in order to support me within the scope of the above-mentioned programme and provide me with further information on the disease polycythaemia vera (PV) or myelofibrosis (MF) by post or email (depending on my information).

Right of revocation

- I am aware that I will can withdraw consent at any time without giving reasons with future effect by post or email to Novartis Pharma GmbH, Roonstr. 25, 90429 Nuremberg, infoservice.novartis@novartis.com.
- I am aware that my further participation in the programme “Living with PV & MF” is not possible in this case and that my participation automatically ends on receipt of the revocation.

General data protection information for processing

The legal basis for the processing is your consent, Art. 6 para. 1 letter a of the General Data Protection Regulation [GDPR].

Novartis uses both group-internal services and external third parties for processing. For these purposes, personal data may be forwarded to and processed, stored and used by the following bodies:

- companies affiliated with Novartis worldwide, particularly in Switzerland, India and the United States.

If Novartis transmits personal data outside the EU or the EEA, Novartis shall always ensure by means of appropriate contracts that the recipient complies with an appropriate level of data protection in accordance with European data protection law.

You are also entitled to request information (Art. 15 GDPR), correction (Art. 16 GDPR), deletion (Art. 17 GDPR), restriction of processing (Art. 18 GDPR) and data portability (Art. 20 GDPR) under the respective legal requirements. In addition, in accordance with Art. 21 para. 1 GDPR, you have a right to object to the processing, insofar as this is based on Art. 6 para. 1 sentence 2 f GDPR. To exercise your rights, please contact: Infoservice, Novartis Pharma GmbH, Roonstr. 25, 90429 Nuremberg, infoservice.novartis@novartis.com.

You can also contact the Novartis data protection officer for general questions. The current data protection officer can be reached at the following address: Novartis Business Services GmbH, Data Privacy, Industriestr. 25, 83607 Holzkirchen, Germany, datenschutz@novartis.com. Finally, in accordance with Art. 77 GDPR, you also have the right to file a complaint with the competent supervisory authority if you believe that the processing of your personal data is not lawful. The supervisory authority responsible for Novartis is the Bavarian State Office for Data Protection Supervision, Promenade 27 [Schloss], 91522 Ansbach, https://www.lda.bayern.de/de/index.html.

You can find further information on the transfer of data and the storage periods at https://www.novartis.de/sites/www.novartis.de/files/Consolidated_Patients_Final%20Clean_Mar%202018_DE-DE.pdf.

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