

Your guide to

Ruxolitinib (Jakavi®)

Information and advice



MPN  voice



Supporting patients and families affected by MPN

Established by a group of MPN patients over 17 years ago, MPN Voice, registered under the auspices of Guy's and St Thomas' Charity, has a website with a lively online community and up to date information on essential thrombocythaemia (ET), polycythaemia vera (PV), and myelofibrosis (MF).

Backed by leading healthcare professionals, MPN Voice also publishes a newsletter, MPN disease leaflets and drug leaflets, runs regular regional patient forums, funds MPN research and offers a buddy scheme.

For more information visit the website **www.mpnvoice.org.uk**

Email **info@mpnvoice.org.uk**

MPN Voice previously known as MPD Voice

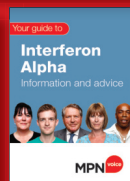


Name: _____

I have the following MPD: _____

Medication: _____

Hospital contact number is: _____



What is this leaflet about?

This leaflet is for people with myeloproliferative neoplasms (MPNs), also known as myeloproliferative disorders, (MPDs).

It offers information about the medicine ruxolitinib or Jakavi®.

In this leaflet you will learn:

- How ruxolitinib works
- The benefits and drawbacks of taking ruxolitinib
- How to take and store ruxolitinib
- Answers to frequently asked questions
- How to feel your best while taking ruxolitinib

The leaflet provides information that can aid your decision about taking ruxolitinib, and is a reference for people already taking this medication. It is important that in addition to reading this leaflet you also read the information provided with your medicine.

What is ruxolitinib?

Ruxolitinib is a medication used to treat myeloproliferative neoplasms (MPNs). It is licensed for the treatment of patients with myelofibrosis (MF) either primary or occurring after polycythaemia vera (PV) or essential thrombocythaemia (ET). Whilst it is also licensed for patients with polycythaemia vera who have not responded or tolerated Hydroxycarbamide it is not however currently readily available under UK prescribing guidelines. It is under investigation in clinical trials for patients with PV.

The drug is known by several names. It goes under the brand name Jakavi/Jakafi® and the molecule is called ruxolitinib.

If you need more information about JAK2 mutation or the types of MPN or myelofibrosis you can learn more on our website at www.mpnvoice.org.uk

Ruxolitinib comes as a tablet and is taken by mouth. Ruxolitinib is currently available in 5mg, 10mg 15mg or 20mg white tablets.

How does ruxolitinib work?

Blood cells originate from stem cells. Stem cells are master cells which divide and mature into the different types of blood cells: red blood cells, white blood cells and platelets. The marrow in our bones acts as a factory for the division and maturation of these stem cells into blood cells. Each new blood cell contains DNA which carries all the instructions the cell needs to grow and function. Once the blood cells have matured they leave the bone marrow and enter our blood stream. In MPNs the processes involved in controlling blood cell production are disordered and for most patients this is thought to be due to over activation of JAK2 a protein found in most cells. For about 75% of patients with MPN this over activation is due to a mutation or change in the genetic sequence of JAK known as JAK2V617F.

Ruxolitinib works by interfering with the function of JAK2 and its relation JAK1. This slows down blood cell production and also the production of signalling chemicals known as cytokines.

When you take ruxolitinib the number of blood cells being made decreases so that your bloodstream will be less crowded, your blood will flow more freely, and you will be less likely to suffer a blood clot or thrombosis. Treatment with ruxolitinib also reduces the size of most patients' spleens towards a more normal size. In addition many of the symptoms, for example sweating, itching, tiredness and pain and fullness after eating which MF and other MPN patients suffer, improve with ruxolitinib therapy.

You will be asked to sign a consent to treatment form before you start taking ruxolitinib.

Are there any drawbacks?

As with all medications you may experience side effects whilst taking ruxolitinib. These are listed in the side effects section below.

Initially you will need blood tests every two to four weeks and then ongoing frequent blood tests and monitoring to ensure that the dose is correct for you.

A small number of people develop a resistance to ruxolitinib, (so that it no longer works) over a period of years, requiring a switch to another treatment.

Non-melanoma skin cancer

Non-melanoma skin cancers (NMSCs), including basal cell, squamous cell, and Merkel cell carcinoma, have been reported in patients treated with ruxolitinib. Most of these patients had histories of extended treatment with chemotherapy medication and prior NMSC or pre-malignant skin lesions. A causal relationship to ruxolitinib has not been established. Periodic skin examinations are recommended for patients who may be at increased risk for skin cancer.

Are there any side effects?

Most people taking this drug tolerate it well and have few side effects. However, it is important that you inform your doctor or nurse if you are experiencing any of the side effects listed below or any develop any new symptoms, even if mild.

Common side effects

Approximately one person in 100 up to one person in five (1–20%) of people taking ruxolitinib will experience some of these common side effects:

- *Reduced red blood cells* If your red blood cells drop too low (anaemia) you may notice that you are becoming breathless and tire easily
- *Reduced platelets* If your platelet count drops too low you may experience nose bleeds, bleeding gums when you clean your teeth, a rash of tiny red spots or increased bruising
- *Reduced white blood cells* If your white blood cells drop too low you may have an increased risk of developing infections. You may experience a high temperature, fever, shivers or chills

If you experience any of the symptoms above you must contact your hospital immediately. Sudden and unexpected changes in blood cell levels may occur, although this is uncommon. Therefore it is important to attend appointments to have your blood counts checked and to discuss any symptoms with your haematologist.

Other common side effects

- painful skin rash with blisters (possible symptoms of shingles (*herpes zoster*))
- high level of cholesterol
- abnormal liver function test results
- dizziness
- headache
- weight gain
- frequently passing wind (*flatulence*)

If you get any side effects, talk to your doctor or pharmacist. This includes any possible side effects not listed here.

Ruxolitinib and infections

Ruxolitinib treatment has been shown to be associated with increased risk of infections varying from simple chest and urine infections to reactivation or occurrence of more serious infections such as shingles, hepatitis, TB and rare infections including a disease called Progressive Multifocal Leucoencephalopathy. It is recommended that baseline blood tests are undertaken to monitor any potential viral infection to prevent reactivation.

Taking ruxolitinib

How to take ruxolitinib

- Take either before or after food, usually twice daily. We recommend that you take the drug at the same time each day
- Wash your hands thoroughly before and after taking the tablets
- Swallow whole with plenty of water
- Do not break or crush the tablets
- It is important to not stop taking your tablets without consulting your haematology team

Dosage

Your doctor, nurse or pharmacist will give dosage instructions. He or she may recommend that you take your medication twice a day. Please be sure to follow the instructions precisely.

Keeping track

It may be helpful to keep a record to remember when to take your tablets and to record any side effects. You may be given a diary or booklet for this purpose.

Storage and disposal of Ruxolitinib

- Store in a dry place at room temperature and definitely less than 30°C
- Ruxolitinib can be dangerous to others. Keep your tablets in a secure location, well out of the reach of children and pets
- Return any unused medication to your local pharmacy or hospital
- Do not dispose of them in the bin or flush them down the toilet

What should I expect?

How fast does it work?

Ruxolitinib may take several weeks to begin having an effect on your cells, but often its effects on spleen and symptoms occur in 2-4 weeks. Your blood cell counts may not stabilise for up to 16 weeks.

How will I feel?

You will hopefully notice a reduction in your MPN-related symptoms. It may help to keep track of these so you can monitor how you are feeling. Most people taking this drug tolerate it well and have relatively few side effects.

Will I need follow up?

You will need more frequent blood tests during the first weeks of treatment to determine how your body is responding to the medication. Once your body has adjusted to the medication you will attend for checks less frequently, perhaps every two to three months. Your kidney and liver function may also be checked with blood tests.

Important tips for taking ruxolitinib

Ruxolitinib works fast to reduce spleen and symptoms but these benefits are rapidly lost when the medication is stopped. This relapse of symptoms and spleen size can occur rapidly and can make patients very ill.

For this reason **it is important not to stop this medication suddenly** and generally your medical team will slowly reduce the dose before stopping this drug.

It is advisable to have an annual cholesterol check with your GP.

If you are admitted to hospital for any reason, emergency or routine it is important that you keep taking ruxolitinib and ask the medical team to liaise with your haematology team.

Keep a regular check on your skin and if any changes are noticed, report these to your haematology team.

Can I take other medicines?

Whenever you take ruxolitinib (or in fact any medication), it is important to inform your medical advisors about all other medications you are taking: this includes medicines prescribed for you as well as any vitamins, herbal supplements or remedies bought in chemists. Always provide the names of these medications and remedies to the hospital doctors, GPs, nurses and pharmacists who are treating you, prescribing additional medications or giving you advice. It can be very helpful to carry a list of the names and dosages of all your medicines to show to your doctor or nurse at appointments.

Some medicines may interact with ruxolitinib because the drug is broken down by an enzyme called CYP3A4. These include:

- Medicines used to treat fungal diseases (such as ketoconazole, itraconazole, posaconazole, fluconazole and voriconazole)
- Medicines used to treat certain types of bacterial infections (antibiotics such as clarithromycin, telithromycin, ciprofloxacin, or erythromycin)
- Medicines to treat HIV infection/AIDS (such as apranavir, atazanavir, indinavir, lopinavir/ritonavir, nelfinavir, ritonavir, saquinavir)
- Medicines to treat hepatitis C (boceprevir, telaprevir)
- Nefazodone, a medicine to treat depression
- Mibefradil or diltiazem, medicines to treat hypertension and chronic angina pectoris
- Cimetidine, a medicine to treat heartburn
- FOOD such as grapefruit and Seville oranges (note these are used in making marmalade)

The following may reduce the effectiveness of ruxolitinib:

- Avasimibe, a medicine to treat heart disease
- Phenytoin, carbamazepine or phenobarbital and other anti-epileptics used to stop seizures or fits
- Rifabutin or rifampicin, medicines used to treat tuberculosis (TB)
- St. John's wort (*Hypericum perforatum*), a herbal product used to treat depression

What if I have other medical conditions?

Ruxolitinib should be used under supervision if you have now or have had any of the following conditions:

- Allergies to any of the ingredients in the medicine (these will be listed on the information leaflet that came with your tablets)
- HIV infection or AIDS
- Previous infections such as TB (tuberculosis) or hepatitis B
- Kidney problems
- If you are planning pregnancy
- If you have radiation therapy planned
- Previous skin cancer or Non-melanoma skin cancer

If you think you may have or have had one of these conditions please discuss this with your doctor.

Frequently asked questions

Can I eat and drink normally?

Yes. We recommend that you eat a normal, healthy diet and drink plenty of water.

Can I drink alcohol?

While it is safe to drink alcohol in moderation whilst taking ruxolitinib, we recommend you do not exceed the recommended weekly limit of 14 units for both men and women. Alcohol can cause dehydration, and it is important to avoid becoming dehydrated if you have an MPN. Please ask your nurse or doctor if you require more information regarding alcohol consumption.

What if I want to have a child?

- We strongly recommend that you use contraception whilst taking ruxolitinib, because this medication can be harmful to a developing foetus
- When planning to conceive or to father a child, you should only stop taking ruxolitinib AFTER discussion with your doctor
- It is imperative to discuss your plans together with your haematologist prior to becoming pregnant or fathering a child. Your doctor can recommend treatment options for you that will not cause harm to your developing baby and will increase your chance of a successful pregnancy
- If you or your partner becomes pregnant while taking this drug please contact your doctor immediately for further advice

Can I breastfeed while taking ruxolitinib?

Breastfeeding while taking ruxolitinib is not recommended.

Who will prescribe ruxolitinib for me?

Your doctor, specially trained nurse or hospital pharmacist will prescribe your medication.

Can I drive?

There is no evidence that taking ruxolitinib would stop you from driving.

Do I need to take any special precautions?

It is important not to stop taking ruxolitinib suddenly as this can cause a withdrawal type syndrome which consists of a return of symptoms and growth of your spleen which may be very sudden and have sometimes made patients very unwell.

Can I have vaccinations such as the flu jab while taking ruxolitinib?

Yes, you can have most vaccinations including the flu vaccine whilst taking ruxolitinib. Some vaccinations are live vaccines e.g shingles vaccine and these should not be taken with ruxolitinib. It is important and we recommend you tell the person giving you the vaccine that you are taking ruxolitinib and we would recommend consulting with your local haematology team.

What to do if...

.....

You have taken too much medicine/someone else has taken your medicine

If you have taken extra tablets or if another person has taken your medication please contact your haematology nurse or doctor as soon as possible.

You were sick shortly after taking your tablets

If this happens just once, take your next dose as usual. If you are sick over a number of days contact your haematology team.

You forget to take a dose

If you have forgotten to take a dose, do not take any extra but take your next dose as normal. If you have forgotten to take a few doses, start taking them again and contact your doctor.

You need to have a medical procedure or operation

You may occasionally be required to adjust or stop your ruxolitinib if you need an operation. It is important that you inform the doctor or dentist planning the procedure or operation that you are taking ruxolitinib and that they coordinate

your treatment with your haematology doctor or nurse. We always recommend that you inform your haematology doctor or nurse if you have any procedures or operations planned. It is very important not to STOP taking ruxolitinib without a discussion with your haematology team.

You feel anxious about taking ruxolitinib

If you have concerns, please discuss this with your doctor or nurse. You can also visit the Living with MPN section of our website www.mpnvoice.org.uk

If you do not want to take this medication

Whether or not to take ruxolitinib is entirely your decision. If after talking things over with your haematologist you still feel uncertain or prefer not to take this medication, you can choose not to take it.

Keep your doctor or nurse in the loop

If you decide not to take ruxolitinib or if you want to stop after you begin treatment, it is important to inform your doctor or nurse of your decision. It is very important that you don't suddenly stop taking your tablets before discussing the best alternatives or other suggestions to safeguard your health with your consultant. Suddenly stopping ruxolitinib can cause a withdrawal type syndrome which consists of a return of symptoms and growth of your spleen which may be very sudden and have sometimes made patients very unwell.

Is there an alternative?

There are other treatment alternatives available and you can discuss additional options with your haematologist. If you wish to read up about them, please visit our website at www.mpnvoice.org.uk and click on "Treatments".

Consent

Your doctor will ask you to sign a consent form when you begin treatment. This confirms that you know why ruxolitinib has been recommended for you, and that you understand the risks and benefits of this treatment.

How to help yourself

If you have an MPN it's important to take good care of yourself. There are many things you can do to feel better.

- Good nutrition is important. Eat a balanced diet including lots of fresh fruit and vegetables, lean protein and whole grains. Drink plenty of water and be careful to prevent dehydration by avoiding excessive alcohol and caffeinated drinks
- Maintain a normal weight and maintain your muscle mass to help keep your cholesterol and blood sugar within normal limits. This is important if you are taking a drug like ruxolitinib which can cause weight gain!
- Exercise is very beneficial for all MPN patients and helps fight fatigue. Be sure to check with your GP and haematologist before launching on any new programme and start slowly and gently if you have not exercised before
- Stop smoking. Ask your GP if you need help

Making the adjustment

It can be disconcerting to start a new medication or find you must increase your dosage. You may feel that your MPN is getting worse or that you are at greater risk of serious medical problems. You may also feel concerned about the long and short term risks of taking this medication. It is normal to feel this way and many people with MPNs have had similar experiences. You may want to discuss your concerns with a family member or friend. MPN Voice also offers a “buddy” programme for interested patients. Please email us at buddies@mpnvoice.org.uk for more information.

It's worth bearing in mind that many people with MPNs have a long life expectancy, and that the treatments are very effective at controlling cell production. You can read more about the psychological aspects of MPN treatment on our website at www.mpnvoice.org.uk under “Living with MPNs”.

Medical exemptions for treatment

MPN patients in England are entitled to an exemption certificate and should obtain a FP92A form from their GP or haematologist. Further details can be found on our website at www.mpnvoice.org.uk in the living-with MPNs, everyday challenges/cost of medication section.

Resources for patients and families

MPN Voice

Registered under the auspices of Guy's and St Thomas' charity, MPN Voice provides patients and families affected by MPNs with a comprehensive range of disease and medication publications, regular newsletters, a buddy scheme and runs regional patient forums as well as funding research into MPNs and drugs trials. The website also offers access to an online community of MPN patients as well as the latest news and reports from leading healthcare professionals.

www.mpnvoice.org.uk

Anthony Nolan

Its vision is to save the lives of everyone who needs a bone marrow or stem cell transplant. Established in 1974, the Anthony Nolan Bone Marrow Register was the founding member of Bone Marrows Worldwide in 1988. Today the register has over 600,000 potential donors on it.

Tel: 0303 303 0303

www.anthonynolan.org

British Heart Foundation

Founded over 50 years ago by a group of medical professionals who wanted to fund extra research into the causes, diagnosis, treatment and prevention of heart and circulatory disease. The website offers health advice to prevent heart disease and practical tips for healthy living.

www.bhf.org.uk

Leukaemia Care

Offers a helpline and website which provides support to anyone affected by a blood cancer.

Tel: 08088 010 444

www.leukaemiacare.org.uk

Blood Cancer UK

Formerly known as Leukaemia and Lymphoma Research, the charity is dedicated to finding the causes of blood cancers including leukaemia, lymphoma and myeloma. It does this through dedicated research focused on finding the causes, improving diagnosis and treatments as well as investment in groundbreaking clinical trials for all blood cancer patients.

Tel: 0808 169 5155
www.bloodcancer.org.uk

Medicines.org

A website run in collaboration with the NHS providing up to date, reliable and understandable information about medicines. A great resource for all medicines.

www.medicines.org.uk

Macmillan Cancer Support

Offers a comprehensive website with cancer support and information about MPNs, chemotherapy drugs, side effects and how best to manage side effects.

Tel: 0808 808 0000
www.macmillan.org.uk

Stroke Association

Offering help and advice for people and families affected by stroke. The website offers tips on recognising the signs of a stroke as well as the preventative lifestyle measures everyone should consider if at high risk of a stroke.

Tel: 0303 3033 100
www.stroke.org.uk

Could you help us?

Your support will enable us to help many more MPN patients and their families



If you have an MPN or know someone with this condition **MPN Voice** aims to offer support and advice.

Visit **www.mpnvoice.org.uk** to find out more about our

- Buddy System
- Medical Alert Cards
- Newsletters
- Treatment and drug leaflets
- Regional patient forums
- Online community of MPN patients
- Funding of some of the latest MPN research
- Backing by leading health professionals

www.mpnvoice.org.uk
email info@mpnvoice.org.uk



MPN Voice is registered under the auspices of Guy's and St Thomas' Charity. Registered Charity No 1160316-15

Order more information

Our **information booklets** can be downloaded from www.mpnvoice.org.uk
Or you can request copies by completing this form. Booklets are free of charge but any donations will be gratefully received, particularly if you are ordering bulk quantities.

We would love to keep you up to date on how we help MPN patients and their families deal with this rare blood disorder. Please tick the boxes below to state your contact preferences

- ☐ I would prefer NOT to be contacted by post
- ☐ I would prefer NOT to be contacted by telephone
- ☐ I am happy to receive email updates

Quantity	Title of booklet
	Anagrelide
	Aspirin
	Busulfan
	Danazol
	Hydroxycarbamide
	Interferon Alpha
	Ruxolitinib
	Newsletter
	A4 MPN Voice Poster
	Travel and Insurance

Please fill in your details below:

Title
First name
Last name
Organisation
Job title
Address
Postcode
Please indicate your age range (tick applicable box) <input type="checkbox"/> 20-30 <input type="checkbox"/> 31-50 <input type="checkbox"/> 51-64 <input type="checkbox"/> 65+
Email
Phone

Please return completed form to
MPN Voice (leaflets)
c/o Department of Haematology, Guy's and St Thomas' Hospital, London SE1 9RT

About this publication and MPN Voice

This document was written by Yvonne Francis, an NHS MPN specialist nurse, with input from consultant haematologists, specialist nurses, psychologists and volunteers of MPN Voice.

You may contact MPN Voice by post:

MPN Voice
c/o Haematology Department
Guy's and St Thomas' Hospital
London
SE1 9RT

or

Email: info@mpnvoice.org.uk

We welcome your feedback. Please write with your suggestions to our postal address or email us at info@mpnvoice.org.uk

You can also contact the Guy's and St Thomas' Charity at info@gsttcharity.org.uk or visit their website for more information at www.gsttcharity.org.uk



MPN Voice is registered under the auspices of Guy's and St Thomas' Charity.
Registered charity No. 1160316-15

This leaflet was first published in November 2013

Revision 3.00 October 2020

For review in two years (October 2022)

© Copyright MPN Voice October 2020

Visit the MPN Voice website to find out more about our

- Buddy system
- Medical Alert Cards
- Newsletters
- Regional patient forums
- Online community of MPN patients
- Funding of some of the latest MPN research

www.mpnvoice.org.uk