Primary Myelofibrosis (PMF)

What this leaflet is about: a scarred bone marrow, the symptoms it causes, and the treatments available.

> Quick emergency advice
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> Sudden breathlessness, chest pain or coughing blood: seek help the same day.
> Fever of 38 degrees C or above, or new and heavy night sweats, needs reporting the same day - infection is more likely and harder to spot.

What is primary myelofibrosis?

In primary myelofibrosis (PMF) the bone marrow gradually becomes replaced by tough, scar-like fibrous tissue. Because the marrow can no longer make enough blood cells, the counts fall - usually the red cells first, causing anaemia and tiredness.

Some people start with high blood counts instead. That earlier phase is called prefibrotic PMF and is why blood tests and a bone marrow biopsy are used to decide when treatment should start.

PMF is a type of myeloproliferative neoplasm (MPN). It is usually driven by a change in JAK2, CALR or MPL.

How will I feel?

The most troublesome symptom is a feeling of fullness or pain in the upper left of the abdomen. This is an enlarged spleen, which has taken over some of the work of making blood cells. It makes you feel full after only a few mouthfuls of food.

- Marked tiredness, usually from anaemia.
- A bloated, uncomfortable or painful abdomen, often on the left side.
- Losing weight without trying.
- Night sweats that soak your clothes.
- Fever, without an obvious cause.
- Intense itching, often worse after a bath.

How is PMF assessed?

Risk scores are used, because they estimate how long the disease is likely to cause problems and whether a transplant should be considered. The scores in use are DIPSS, DIPSS-plus, MIPSS70+ and rIPSS. They combine your symptoms, blood counts, whether you need transfusions and the gene changes in your blood cells.

A bone marrow biopsy, blood tests and a JAK2 / CALR / MPL test are usually needed.

How is PMF treated?

- JAK inhibitors - ruxolitinib, fedratinib, pacritinib or momelotinib. These are the main treatment. They commonly reduce the size of the spleen, improve symptoms and reduce tiredness.
- Interferon - given by injection; often used in younger people, and the only drug group that can alter the disease itself in some people.
- Supportive care - blood transfusions, iron and vitamin supplements, and treatment of infections as they occur.
- Allogeneic stem cell transplant - the only treatment that can cure PMF. It is a major procedure, so it is usually considered for younger or higher-risk patients, or when the disease is progressing.

Your specialist team will weigh the risk scores, your age and general health, and your own wishes, before recommending a plan. You do not have to decide alone, and you can ask for more time.

Living with an enlarged spleen

- Eat small, frequent meals rather than three large ones.
- Avoid bending sharply or straining, and ask for help lifting.
- Tell us straight away if you develop a persistent dull ache or a sudden increase in size of the abdomen.
- Report any fever or night sweats - these can signal infection.

Will it change?

PMF varies a great deal. Some people live for many years with few symptoms on treatment; others progress more quickly. A minority develop acute myeloid leukaemia. Regular blood tests pick up change early, so please keep your appointments even when you feel well.

Where to read more

The US National Cancer Institute and the American Cancer Society publish public patient information on myelofibrosis. Ask us for the printed links.

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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