

Glucose-6-phosphate dehydrogenase deficiency (favism or G6PD deficiency)



Information for parents

This leaflet aims to provide you with information if you or your child have been diagnosed with G6PD deficiency. If you have any other questions or concerns, please do not hesitate to speak to the team caring for you.

Confirming your identity

Before you have a treatment or procedure, our staff will ask you your **name** and **date of birth** and check your **ID band**. If you don't have an ID band we will also ask you to confirm your address.

If we don't ask these questions, then please ask us to check.

Ensuring your safety is our primary concern.

What is G6PD deficiency?

G6PD is a protein found in red cells in the blood. Red cells carry oxygen and each one lives for about three months before being destroyed. During their life, red cells are exposed to many different damaging substances, which can cause them to be destroyed.

G6PD helps protect the red cells against the damaging effects of these substances, and stops them being destroyed early. If someone has G6PD deficiency, their red cells contain reduced amounts of G6PD protein, and if they are exposed to certain chemicals, such as some foods and drugs, the red cells are destroyed too soon causing anaemia and other problems.

How do you get G6PD deficiency?

G6PD deficiency is inherited, which means that you are born with the condition and it will stay with you all through your life. It is linked to the X chromosome, which means that it is much more common in males than females, although females can be affected. If you have G6PD deficiency, it is possible that you will pass it on to any children of your own, and there are often other family members who have this condition.

Do many people have it?

G6PD deficiency is very common in some parts of the world, especially in countries bordering the Mediterranean Sea and parts of Africa and Asia where more than 10% of people may be affected. It is fairly rare in people from Northern European countries, but does still occur.



What are the symptoms of G6PD deficiency?

Usually people feel completely well with G6PD deficiency unless they are exposed to a drug or chemical that can cause an acute (sudden) attack of red cell breakdown leading to anaemia. Many people have the condition and do not know that they have it.

Babies with G6PD deficiency are sometimes more severely jaundiced (yellowing of the whites of the eyes or skin) than other babies, and can require treatment to reduce the jaundice. This treatment usually involves using ultra-violet light and sometimes blood transfusions.

Children and adults usually develop symptoms only when they are exposed to a substance known to damage red cells and this causes sudden onset anaemia.

Symptoms include:

- looking pale, jaundice
- black or red urine
- back pain
- fever
- generally feeling very ill

These typically occur within 12 hours of exposure to the substance, although this can vary.

What is the treatment for G6PD deficiency?

Most of the time you will not need any treatment, but you should avoid substances known to cause problems. If symptoms of an acute attack develop, it is important to see your GP or go to the A&E Department of your local hospital.

Sometimes the anaemia can become so severe that a blood transfusion is necessary.



Intravenous fluids are often useful and it is obviously important to stop taking any drug that might have caused the problem.

Although attacks can be severe, in most cases people recover quickly without any long-term complications.

What substances should I avoid if I have G6PD deficiency?

1. Fava beans/broad beans

(also called bell beans, English dwarf beans, fever beans, haba beans, hole beans, pigeon beans, tick beans)



This is the only type of food that commonly causes attacks of red cell destruction in G6PD deficiency. Broad beans/fava beans are sometimes eaten in salads and other meals and can cause a severe episode of anaemia, jaundice and black urine, as described earlier.

The effects are worse if large quantities are eaten or if the beans are raw, but even small amounts can cause problems.

Runner beans and baked beans do not cause problems and can be safely eaten.

2. Mothballs

Mothballs contain naphthalene and are used to stop clothes being damaged by moths. They look like sweets and so are sometimes mistakenly eaten by children. This can cause acute anaemia if the child has G6PD deficiency.



3. Drugs

There is a long list of drugs that have been known to cause problems in people with G6PD deficiency. The important drugs to avoid are listed below. Most of the drugs are not used very often, and there is nearly always a safe alternative that will not cause problems.

It is most important to let any doctor, dentist or pharmacist know that you or your child have G6PD deficiency before they either prescribe or give you any drugs.

You can also contact MedicAlert for information on a range of special jewellery that allows you to include a list of all your child's allergies as well as emergency contact details. Find more information at www.medicalert.org.uk

Drugs to avoid in G6PD deficiency

Antibiotic	Anti-malarials	Others
Dapsone	Maloprim	Chinese herbal medicines
Nitrofurantoin	Primaquine	Rasburicase
Sulphonamides (including Co-trimoxazole, Septrin)	Pentaquine	Flutamide
Quinolones (Ciprofloxacin)		
Nalidixic Acid		Methylene blue

Other factors that may cause symptoms

If your child gets a high fever and infection these can also cause them to become unwell.

Occasionally severe episodes occur with no obvious cause, although this is fairly unusual.



Summary

If either you or your child has just been diagnosed with G6PD deficiency, we will normally see you in the Outpatient Department once or twice to confirm the diagnosis and make sure that you or your child is well. It will not normally be necessary to see you regularly in clinic, although it is important that you remember that you have G6PD deficiency and tell anyone prescribing medication that you have it.

If I have any other questions, who should I contact?

You can contact us through the Paediatric Haematology Office on 020 3299 3773.

Sharing your information

We have teamed up with Guy's and St Thomas' Hospitals in a partnership known as King's Health Partners Academic Health Sciences Centre. We are working together to give our patients the best possible care, so you might find we invite you for appointments at Guy's or St Thomas'. To make sure everyone you meet always has the most up-to-date information about your health, we may share information about you between the hospitals.

Care provided by students

We provide clinical training where our students get practical experience by treating patients. Please tell your doctor or nurse if you do not want students to be involved in your care. Your treatment will not be affected by your decision.



PALS

The Patient Advice and Liaison Service (PALS) is a service that offers support, information and assistance to patients, relatives and visitors. They can also provide help and advice if you have a concern or complaint that staff have not been able to resolve for you. They can also pass on praise or thanks to our teams.

PALS at King's College Hospital, Denmark Hill, London SE5 9RS

Tel: **020 3299 3601**

Email: **kch-tr.palsdh@nhs.net**

PALS at Princess Royal University Hospital, Farnborough Common, Orpington, Kent BR6 8ND

Tel: **01689 863252**

Email: **kch-tr.palspruh@nhs.net**

If you would like the information in this leaflet in a different language or format, please contact our Communications and Interpreting telephone line on 020 3299 4826 or email kch-tr.accessibility@nhs.net



