What is glucose-6-phosphate dehydrogenase (G6PD) deficiency?

North Central London Haemoglobinopathy Network jointly with Whittington Health, Royal Free London, and Luton and Dunstable NHS Foundation Trust

Joint Red Cell Unit
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What is G6PD?
G6PD is an abbreviation for glucose-6-phosphate dehydrogenase. It is an enzyme (chemical) that exists in many cells in the body, protecting them from damage.

What does G6PD have to do with the blood?
G6PD plays an active role in red blood cells that carry oxygen to all parts of the body. It protects these cells against some harmful compounds that may collect when you have a fever or infection, or when you take certain medicines or eat certain foods. Its protective effect is especially important in red cells as these cells don’t have a nucleus, also called the brain of the cell.

How can G6PD deficiency affect me?
If you are deficient in the enzyme, your red blood cells may not be protected from damage when you have a fever, catch an infection, eat certain foods or take some medicines. They break down in the circulation, decreasing the number of whole red cells and causing a ‘haemolytic crisis’. This causes some of the haemoglobin (the part of the cell carrying oxygen around the body) from the broken red cells to be released into the blood.

The haemoglobin is then processed by the liver, giving the eyes and skin a yellow tinge known as ‘jaundice’. Some of the haemoglobin also passes through the kidneys and into the urine, making the urine coca-cola coloured. This is known as haemoglobinuria.

As there are now fewer whole or complete red cells in the circulation, you may become anaemic. You may look pale and if anaemia is severe, you may be breathless, tired and lifeless. This process stops on its own, and when it is over, your bone marrow will make new cells to compensate.
However, while it is happening and for a few days or weeks afterwards (depending on the severity), you will need folic acid and iron to help the bone marrow to make new cells. If the haemoglobin levels go very low or if you become unwell, you may need a blood transfusion.

Newborn babies with G6PD deficiency sometimes have haemolytic crises at birth or soon after, even without having an infection or taking medication. This is probably due to the stress of childbirth. Because of the danger of jaundice in the newborn, these babies may require special treatment for their jaundice. For example, it may be a treatment with special lights (called phototherapy), or occasionally a blood transfusion.

**What causes G6PD deficiency?**

G6PD deficiency is inherited, which means that it is passed on at birth from one or both parents. Most commonly it is passed from the mother. It is found in both males and females but usually affects males more severely. Often when a mother passes it on to her son, she may never have had any symptoms.

G6PD deficiency is also found more commonly in people of Mediterranean, Middle Eastern, Asian and African descent. The severity of deficiency varies among different populations, and people of African descent are generally less affected than other groups.

**Is there any way to prevent G6PD deficiency?**

Since it is inherited, people are born with the deficiency and there is no known way to prevent it.

Fortunately, most people who have it don’t ever have a haemolytic crisis. For many of those who do, it is mild. The most important thing to do is to avoid the triggers for the crisis (please see the next page).
How can I find out if I am G6PD deficient?
The only way to find out is to have a special blood test.

What can trigger a haemolytic crisis in people with G6PD?
1. Infection.

2. Medication. There is a list of medications that can trigger a haemolytic crisis. This can be found in the Pharmacy Formularies, which all doctors, nurses and pharmacists can refer to when you are given a new medication. There will be similar formularies in other countries. Before starting a new medication, it is important that you tell the health professional treating you that you are G6PD deficient. They can then check whether it is safe for you to take it. You should not take non-licensed medicines, such as Chinese medicines, as they have not been tested sufficiently for their safety.

Vaccinations, paracetamol and ibuprofen are all fine.

3. Chemicals, for example:
   • aniline dyes used in dying fabric and leather
   • naphthalene (mothballs).

4. Foods, for example fava (broad) beans. Eating fava beans may be especially harmful to people with G6PD deficiency.

Where can I get more information?

G6PD Deficiency Association
Website: www.g6pd.org

University College London Hospitals NHS Foundation Trust cannot accept responsibility for information provided by external organisations.
Contact details

Haematology advice line (office hours, adults and children):
020 3447 7359

Adult haematology advice line (out of hours):
07852 220 900

Paediatric helpline (out of hours):
• nurse in charge 07961 081 645
• ward T11 south 020 3456 7890 ext. 71103 or 71143

Apheresis:
020 3447 1803

Address: The Joint Red Cell Unit
Department of Haematology
3rd Floor West, 250 Euston Rd
London NW1 2PG

Website: www.uclh.nhs.uk/JRCU

Hospital for Tropical Diseases Travel Clinic

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Appointments for the travel clinic are available on:
Wednesdays 1pm – 5pm
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Please call 020 3447 5999 to book.
Haematology consultants:
Professor John Porter
Dr Sara Trompeter
Dr Perla Eleftheriou
Dr Emma Drasar
Dr Bernard Davis
Dr Farrukh Shah

Specialist nurses:
Bernadette Hylton (adults)
Nancy Huntley (apheresis)
Nina Gorman (children)