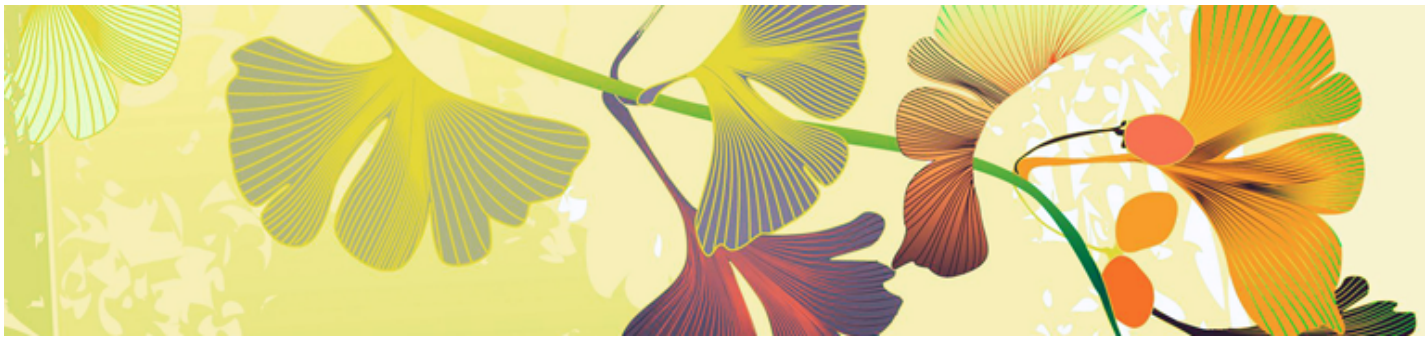

G6PD deficiency (Glucose-6-Phosphate Dehydrogenase deficiency)

G6PD缺乏症

(蠶豆症)(英文)

Definition

Glucose-6-Phosphate Dehydrogenase (G6PD) deficiency is the most common human enzyme deficiency in the world; it affects an estimated 400 million people. G6PD deficiency is also known as "favism," since G6PD deficient individuals are also sometimes allergic to fava beans. G6PD deficiency is an allelic abnormality which is inherited in an X-linked recessive fashion. When someone has G6PD deficiency, complications can arise; hemolytic anemia and prolonged neonatal jaundice are the two major pathologies associated with G6PD deficiency. Both of these conditions are directly related to the inability of specific cell types to regenerate reduced nicotinamide adenine dinucleotide phosphate (NADPH); this reaction is normally catalyzed by the G6PD enzyme. A consequence of this is that G6PD deficient individuals are resistant to the malaria causing parasite.

Symptoms and signs

In G6PD deficient individuals, anemia is usually caused by certain oxidative drugs, infections, or fava beans. When any one of these agents, or their metabolites, enters a G6PD deficient red blood cell, hemoglobin becomes denatured, thus destroying its function as the principal oxygen carrying molecule. A child with G6PD deficiency who is exposed to a medication or infection that triggers the destruction of RBCs may have no symptoms at all. In more serious cases, a child may exhibit symptoms of anemia (also known as a hemolytic crisis), including: paleness (in

darker-skinned children paleness is sometimes best seen in the mouth, especially on the lips or tongue), extreme tiredness, rapid heartbeat, rapid breathing or shortness of breath, jaundice, or yellowing of the skin and eyes, particularly in newborns, an enlarged spleen, dark, and tea-colored urine. In addition to being susceptible to hemolytic anemia, G6PD deficient individuals are also predisposed to prolonged neonatal jaundice. This can be a potentially serious problem as it can cause severe neurological complications and even death.

Prevention and Precaution

The most important measure is prevention - avoidance of the drugs and foods that cause hemolysis, including :

1. NSAIDS (Aspirin, Tylenol, Ibuprophen)

2. Quinolones

3. Drugs metabolized through the liver or known attributions cause blood or liver related problems or hemolysis

4. Sulfa drugs

5. Petrochemically derived substances (This is a long list and gets longer every year. Many artificial foods, dyes and vitamins are included in this list .)

6. Fava Beans and other legumes, products containing sulfites, products containing blue food coloring, and tonic water(contains quinine).

7. Products containing Naphtha, moth balls, and sulfites.

8. Crystal violet: When in doubt, the safest course of action is to avoid the substance. If you react to something not on this list, avoid that substance in the future. Watch carefully for hemolysis signs at all times.

若有任何疑問，請不吝與我們聯絡
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