葡萄糖六磷酸去氢酶缺乏症
（又名「蚕豆病」）
知多點

電話熱線：2361 9979
（24 小時；提供葡萄糖六磷酸
去氫酶缺乏症資訊的電話聲帶）
網址：http://www.cgs.dh.gov.hk

葡萄糖六磷酸去氫酶缺乏症是一種保護紅血球的酵素，幫助紅血球維持正常的功能。
造成此症的原因是由於這種酵素的份量低於正常水平，繼而引起所謂的「溶血性貧血」。

溶血性貧血是一種血液疾病，當沒有足夠的葡萄糖六磷酸去氫酶保護紅血球時，大量紅血球會被破壞，溶血性貧血便會發生。而紅血球分解時會導致黃疸、對新生兒來說，這是一個嚴重的情況。若得不到及時的治療，過量的膽紅素（一種可在膽囊內找到的黃色色素）會對腦部造成永久的破壞。引致幼翼、大腦麻痺、聽覺受損，甚至死亡。

但只要此症患者避免某些藥物（中藥及西藥）、食物、化學品及避免感染，會引起溶血性貧血的疾病，他們一般都能健康地生活。
你應該採取以下措施:

- 如你是葡萄糖六磷酸去氫酵素缺乏症患者
  - 一旦發現以上任何症狀，應立即求診。
  - 呼吸困難時，應通知醫生，必要時，應立即送院。
  - 請隨時攜帶懷孕卡。

葡萄糖六磷酸去氫酵素缺乏症有甚麼症狀？

- 病發時，患者皮膚和眼白會泛黃，患者也有可能排出深色尿液。初生兒會顯得顏色偏白，沒有精力和過度眼周，小童或成人可能覺得疲憊，呼吸和心跳變得急促。
- 如發生以上症狀，應立即求診接受治療。

你應該避免:

- *避免使用中藥，例如:
  - 黃酮
  - 膚根
  - 金銀花

*此清單未完全涵蓋所有藥物，請向醫生查詢詳情。

如有任何疑問，請向醫生或藥劑師查詢。
Knowing more about G6PD deficiency (also called favism)

Hotline: 2361 9979
(24 hours; prerecorded tape information about G6PD deficiency provided)
Website: http://www.cgs.dh.gov.hk

This leaflet is designed for families who have children diagnosed with G6PD deficiency, which is a genetic condition. There is no cure for this condition, but the children can lead as normal a life as possible if certain precautions are taken. This leaflet provides the essential information and advice concerning G6PD deficiency. The information and advice provided here is not exhaustive, so it is important that families contact their doctors or the hotline as well as access the website to find out more.

What is G6PD deficiency?

G6PD (Glucose-6-Phosphate Dehydrogenase) deficiency is a genetic condition, also commonly known as favism (after fava beans). The gene causing G6PD deficiency is located on an X chromosome, so it is an X-linked recessive genetic condition. XY are chromosomes that determine the sex of an individual. A girl (XX) will get each X chromosome from her mother and father respectively. A boy (XY) will receive an X chromosome from his mother and a Y chromosome from his father.

A boy will have G6PD deficiency if one X chromosome is defective, while a girl will have the condition only when both X chromosomes are defective.

The figure below shows that a male sibling has a 50% chance to be affected, while a female sibling has a 50% chance to be a carrier (i.e. unaffected but can pass on the condition to their children).

X-linked Recessive Inheritance

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<td>Normal</td>
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It is important to know that while the parents may not be affected by the condition, as in all genetic conditions, the defective gene may be passed to the child from one or both parents.

What are the causes and consequences of G6PD?

G6PD is an enzyme that protects red blood cells and helps them to function normally. G6PD deficiency is therefore the result of having less than usual amount of G6PD in the red blood cells, leading to what is technically called haemolytic anaemia.

Hemolytic anaemia is a blood disorder in which excessive red blood cells are destroyed because there is not enough G6PD enzyme to protect the red blood cells. The breakdown of the red blood cells leads to jaundice and can be a serious problem as far as newborn babies are concerned. If left untreated, excess bilirubin (a yellowish pigment found in bile) can cause permanent brain damage, leading to neurological complications, which includes mental retardation, cerebral palsy and hearing deficit, or even death.

People with G6PD deficiency enjoy good health as long as they avoid certain medications (Chinese and Western), food and chemical substances, and are not exposed to severe infections which may lead to acute hemolytic anaemia.
What are the symptoms of G6PD deficiency?

If affected or at risk of G6PD deficiency, the skin and the white part of the eyes will turn yellow. The affected individuals may also pass dark coloured urine. Newborn babies may look pale and appear lacking energy and excessively sleepy. Older children or adults may complain of tiredness, breathlessness and have rapid heartbeats.

If you observe any of the above symptoms, please seek medical advice without delay.

What you should do

- If you are affected or at risk of G6PD deficiency
  o Consult a doctor without delay when you observe any of the above symptoms.
  o Inform the doctor about the G6PD diagnosis during consultation.
  o Always carry the green card.

- If you are pregnant, you must inform your doctor about any family history of G6PD deficiency.
- If as a mother you are breastfeeding your G6PD deficient baby, you should avoid eating fava or broad beans yourself and must consult a doctor before taking any Chinese or Western medicine.

What you should avoid

G6PD deficient individuals will enjoy normal growth and health if the following precautions are taken.

- Avoid eating fava or broad beans and their products.
- Avoid close contact with mothballs (Naphthalene) and Naphthalene-containing products.
- Avoid taking any drugs unless prescribed by a doctor.

*Avoid Chinese herbal medicines such as
  o Rhizoma Coptidis
    (Huang Lian)
  o Flos Chimonanthi Praecocis
    (Leh Mei Hua)
  o Flos Lonicerae
    (Kam Ngan Fa)
  o Calculus Bovis (Niu Huang)
  o Margaritas (e.g. Counter drugs such as Bo Ying Compound which contains Margaritas, etc.)
  o *Avoid Western medicines such as
    o Certain Antipyretics
    o Antibiotics (e.g. Nitrofurantoin, Nalidixic acid, Sulfamethoxazole)
    o Antimalarials (e.g. Primaquine)
    o Antispasmodics (e.g. Phenzopyridine)

*This list is not exhaustive. Please consult a doctor.

If you have any further questions or concerns, please contact us or consult your doctor or pharmacist.