Tikrit University

College of Nursing

Basic Nursing Sciences

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Child Health Nursing

G6PD deficiency

by:

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Glucose-6-phosphate dehydrogenase (G6PD) deficiency

Hemolysis (destruction of RBC) due to exposure to oxidative stress (triggers).

Because of glucose-6-phosphate dehydrogenase defect

G6PD deficiency is an inherited condition or **X linked recessive disorder**.

The gene is on the X chromosome.

Causes of G6PD deficiency / triggers

1. Bacterial and viral infections
2. Some painkillers and fever-lowering drugs
3. Some antibiotics (most often those with "sulf" in their names)
4. Some antimalarial drugs (most often those with "quine" in their names)
5. After eating fava beans or inhaling pollen from fava plants (favism).

Signs & Symptoms of G6PD Deficiency

Most people with G6PD deficiency don't have any symptoms.

Others might have symptoms of hemolytic anemia if many RBCs are destroyed.

These can include:

1. Pallor is best seen in the mouth, especially on the lips or tongue
2. extreme tiredness or dizziness, Weakness, Confusion
3. Fever
4. Fast breathing or shortness of breath
5. jaundice (the skin and eyes look yellow)
6. Dark-colored urine (tea)
7. Trouble with physical activity

8. Enlarged spleen and liver
9. Increased heart rate

Diagnosis

1. Diagnosis is based on symptoms and supported by blood tests
2. Complete blood count and reticulocyte count;
3. in active G6PD deficiency, Heinz bodies can be seen in RBC/blood film;
4. Liver enzymes (to exclude other causes of jaundice);
5. Lactate dehydrogenase (elevated in hemolysis and a marker of hemolytic severity)
6. Haptoglobin (decreased in hemolysis);
7. A negative "direct antiglobulin test" (Coombs' test) – this should be negative, as hemolysis in G6PD is not immune-mediated.
8. When there are sufficient grounds to suspect G6PD, a direct test for G6PD is the "Beutler fluorescent spot test" is a rapid and inexpensive test that visually identifies NADPH produced by G6PD under ultraviolet light.
9. When a macrophage in the spleen identifies a RBC with a Heinz body, it removes the precipitate and a small piece of the membrane, leading to characteristic "bite cells". However, if a large number of Heinz bodies are produced, as in the case of G6PD deficiency, some Heinz bodies will nonetheless be visible when viewing RBCs that have been stained with crystal violet

Prevention:

1. **Avoid some food**
2. **Some drugs (methiprim, nitrofurantoin, …..)**
3. **Antipyretic drugs (paracetamol, brufen)**

4. **Vaccination against some common pathogens (e.g. hepatitis A and hepatitis B) may prevent infection-induced attacks.**

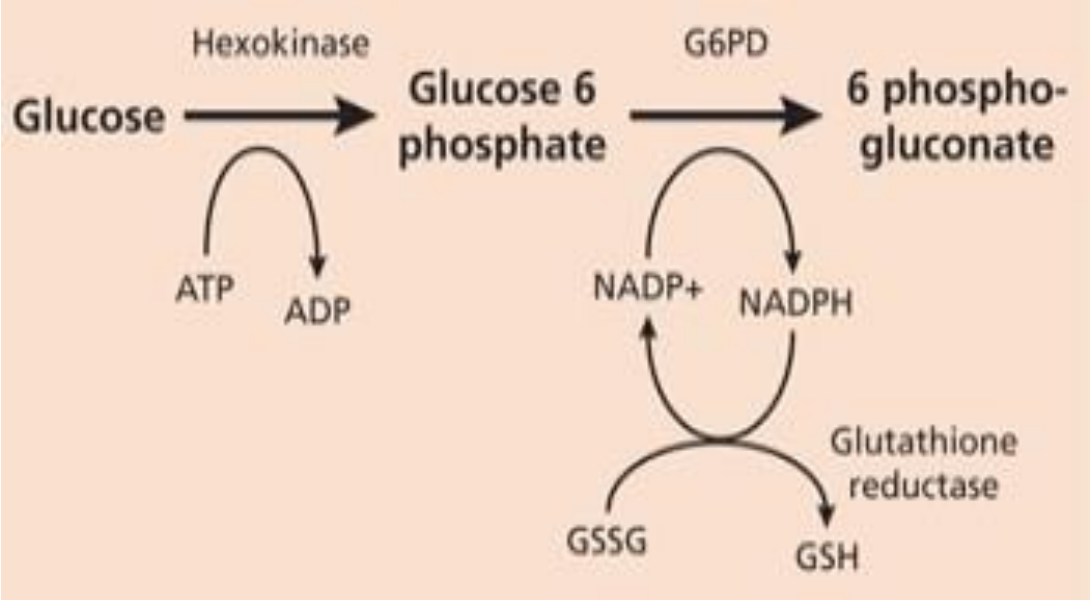
Treatment and Prevention

In acute phase of hemolysis, blood transfusions, or even dialysis in acute kidney failure.

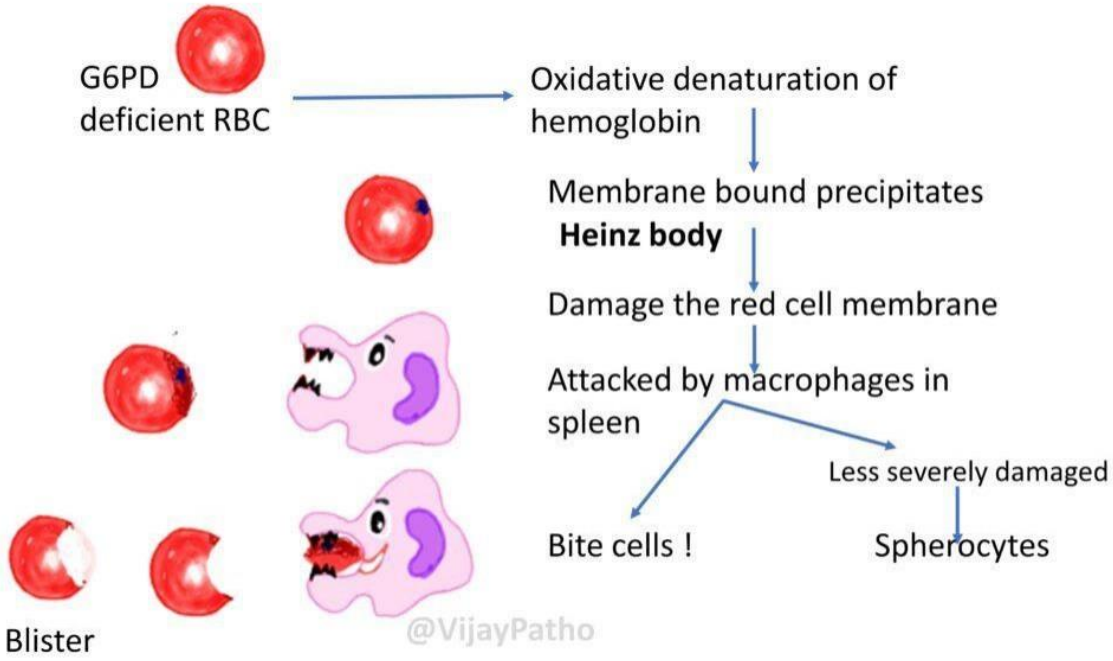
Packed RBC (10 ml/kg)

- Maintaining hydration and urine alkalization protects the kidneys against damage from precipitated free hemoglobin.

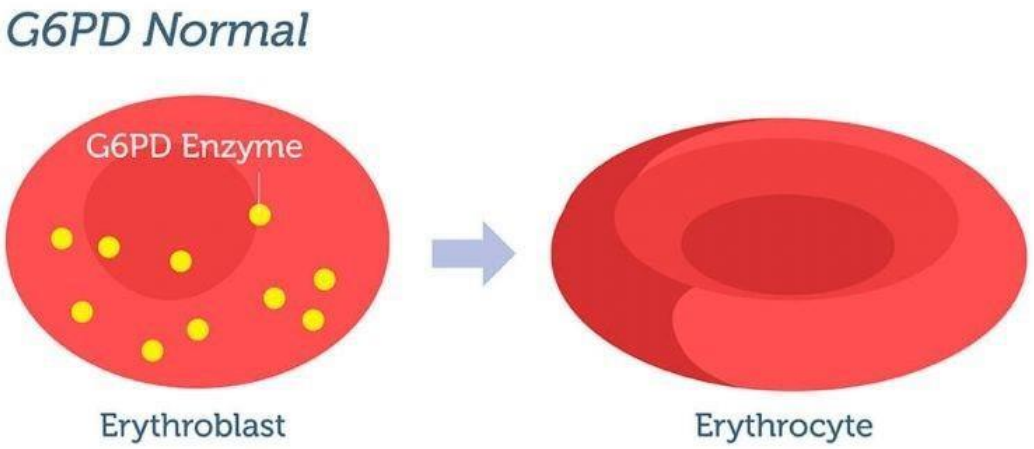
- Some patients may benefit from removal of the spleen (splenectomy) as this is an important site of red cell destruction.

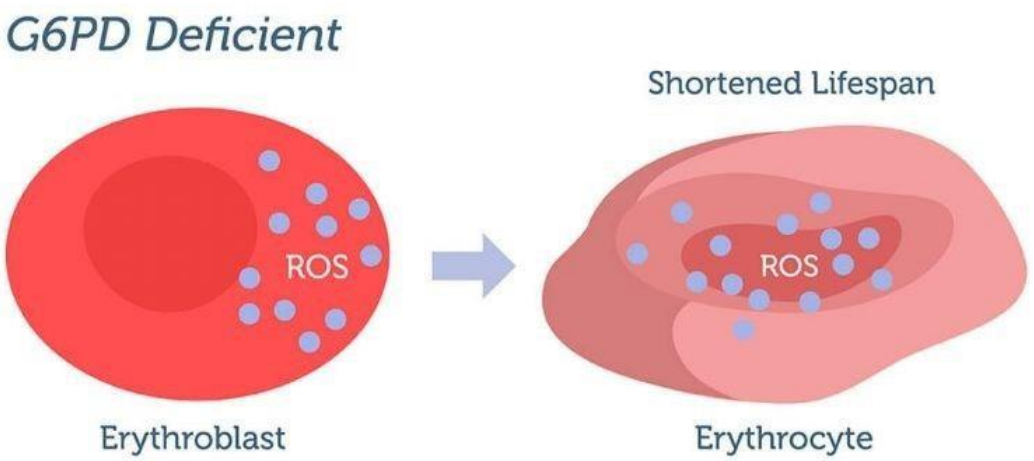


Pathology of G6PD deficiency



Pathology of G6PD deficiency





- **A genetic disorder that results in a deficiency of an enzyme called glucose -6-phosphate dehydrogenase (G6PD) in the blood, which is necessary for the healthy and proper functioning of red blood cells.**