

G6PD Deficiency

A plain-English guide for you and your family

WHAT IS HAPPENING

Normally, red blood cells carry oxygen all over your body and stay healthy for about 120 days. To stay strong, these cells need a special enzyme called G6PD that acts like a shield against harmful chemicals. When someone is born with G6PD deficiency, their red blood cells cannot make enough of this shield. Certain triggers—like fava beans, some antibiotics, or even a bad cold—can cause the red blood cells to suddenly break open. This leads to anemia (low red blood cell count), yellowing of the skin, and dark tea-colored urine. Most of the time, people feel fine and only have problems when they encounter these triggers.

What you might feel

- Dark tea-colored or cola-colored urine
- Sudden fatigue and weakness
- Pale skin and mucous membranes
- Yellowing of eyes and skin (jaundice)
- Rapid heartbeat (tachycardia)
- Shortness of breath with activity
- Back pain or abdominal pain
- Fever during destroying red blood cells (hemolytic) episode

Call the doctor right away if...

- Dark tea-colored urine with decreased urine output suggests acute kidney injury from hemoglobin precipitation requiring immediate IV hydration
- Chest pain, severe shortness of breath, confusion, or altered mental status indicates life-threatening anemia needing urgent transfusion
- Hemoglobin below 7 g/dL with ongoing red blood cell destruction (hemolysis) requires hospital admission and transfusion consideration

- Recent exposure to triggering medications (sulfa drugs, antimalarials, aspirin, nitrofurantoin) or fava beans with symptoms starting within 24-72 hours

What the care team will do

- Screen for recent triggers (infection, fava beans, medications) that may have caused red blood cell destruction (hemolysis) (red blood cell breakdown).
- Monitor for signs of acute destroying red blood cells (hemolytic) crisis: dark tea-colored urine, yellowing of the skin and eyes (jaundice) (yellow skin), fatigue, rapid heart rate, and shortness of breath.
- Check baseline and serial complete blood count (CBC) focusing on hemoglobin, hematocrit, and reticulocyte count (young red blood cells).
- Assess for signs of severe anemia: pale skin and mucous membranes, dizziness, chest pain, confusion, and decreased urine output.
- Educate the patient and family about avoiding known oxidative triggers including certain medications, foods, and infections.
- Ensure adequate hydration with IV fluids during destroying red blood cells (hemolytic) crisis to protect kidney function.
- Review the medication list and alert providers to stop any oxidizing drugs immediately.
- Prepare for possible blood transfusion if hemoglobin falls below 7 g/dL or patient shows signs of cardiovascular compromise.

Medicines you may be given — and why

Normal saline (0.9% sodium chloride IV fluid)

This sterile salt water matches the salt concentration in blood. It expands blood volume and increases fluid flowing through kidneys. Higher kidney flow dilutes and flushes out hemoglobin before it can clump and block kidney tubules.

Packed red blood cells (PRBC transfusion)

These are concentrated red blood cells from donated blood with most plasma removed. Each unit contains normal G6PD enzyme and hemoglobin. Transfused cells immediately carry oxygen to tissues and organs that are suffering from low oxygen delivery during severe anemia.

Folic acid (folate)

Folic acid is a vitamin that bone marrow needs to build DNA in new red blood cells. During red blood cell destruction (hemolysis) the marrow works overtime making replacement cells and

uses up folate stores quickly. Supplementing folate ensures the marrow has enough raw material to maintain high red blood cell production rates.

Supplemental oxygen therapy

Increasing oxygen concentration in inhaled air raises oxygen levels dissolved in blood plasma (liquid part). This improves oxygen delivery to tissues even when red blood cell count is low. Higher oxygen partial pressure helps remaining healthy red blood cells load more oxygen molecules onto their hemoglobin.

Acetaminophen (for fever or pain management)

Acetaminophen reduces fever by acting on the brain's temperature-control center and relieves pain by blocking pain signals in the brain and spinal cord. It does not cause oxidative stress like aspirin or NSAIDs. Safe pain and fever control is important because fever increases metabolic demands and many pain medications trigger red blood cell destruction (hemolysis) in G6PD deficiency.

Infection treatment (pathogen-specific antimicrobials when indicated)

Infections generate oxidative stress that triggers red blood cell destruction (hemolysis) in G6PD-deficient patients. Treating the infection reduces inflammatory cytokines (chemical signals) and oxidative free radicals. Lower oxidative stress allows remaining red blood cells to survive and stops ongoing red blood cell destruction. Must choose infection treatments that do not themselves trigger red blood cell destruction.

Where this information comes from

- American Society of Hematology (ASH) guidelines on diagnosis and management of glucose-6-phosphate dehydrogenase deficiency, 2020
- Centers for Disease Control and Prevention (CDC) recommendations for G6PD testing and prevention of drug-induced hemolysis, 2021
- National Institutes of Health (NIH) Genetic and Rare Diseases Information Center G6PD deficiency clinical overview, 2022
- American Academy of Pediatrics (AAP) guidelines on screening and management of G6PD deficiency in newborns and children, 2019

Last reviewed: 2026-08-29