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Option C (glucose-6-phosphate dehydrogenase) is correct. G6PD deficiency is an X-linked recessive trait and is the most common inborn error of metabolism. A deficiency of G6PD, the rate-limiting enzyme of the pentose phosphate pathway, impairs the synthesis of the reduced form of nicotinamide adenine dinucleotide phosphate (NADPH). By maintaining glutathione in its reduced state, NADPH plays a key role in the ability of cells to defend against oxidizing agents (e.g., peroxide). Erythrocytes have no alternative routes for NADPH synthesis. **Oxidative stress** caused by some drugs (e.g., sulfonamides) or by infections can lead to hemolysis in patients with G6PD deficiency. Intravascular hemolysis is the primary type of hemolysis, and hemoglobinuria is present.

Option A (branched-chain alpha-keto acid dehydrogenase complex) is incorrect. The branched-chain alpha-keto acid dehydrogenase complex is responsible for metabolizing branched-chain amino acids (leucine, isoleucine and valine). A deficiency in this enzyme causes maple syrup urine disease, which causes sweet-smelling urine at birth and leads to neurological symptoms and death unless treated by diet. In this case, an underlying deficiency of Glucose-6-phosphate dehydrogenase (G6PD) has caused **hemolytic anemia** due to increased **oxidative stress** as a result of a sulfa medication.

Option B (glucose-6-phosphatase) is incorrect. **Glucose-6-phosphatase** deficiency causes von Gierke's disease, which is a type I glycogen storage disease. It is not associated with a **hemolytic anemia**. In this case, an underlying deficiency of Glucose-6-phosphate dehydrogenase (G6PD) has caused **hemolytic anemia** due to increased **oxidative stress** as a result of a sulfa medication.

Option D (homogentisate 1, 2-dioxygenase deficiency) is incorrect. A deficiency in homogentisate 1, 2-dioxygenase deficiency causes alkaptonuria, in which homogentisic acid, a compound produced during the

At the bottom of the interface, it shows "Block Remaining: Untimed" and "Question Time: Untimed". There are also "Lock" and "End Block" buttons.

Expert rationales for every option with every question, some in Step 1 including Dorland's definitions.

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Item 1 of 21 Mark Previous Next Lab Values Notes Calculator Show Answer & Explanation

Show Answer & Explanation

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Expert rationales for every option with every question, some in Step 1 including Dorland's definitions.

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