G6PD Deficiency

Glucose 6 phosphate dehydrogenase deficiency is an X linked recessive disorder characterized by a deficiency of the enzyme glucose 6 phosphate dehydrogenase (G6PD). This enzyme is involved in the pentose phosphate pathway and supplies reducing substances by converting NADP+ to NADPH. This NADPH is used by glutathione reductase to maintain levels of glutathione in cells, which helps protect red blood cells against oxidative damage caused by free radicals and peroxides. Individuals with this disease commonly exhibit hemolytic anemia in states of oxidative stress. Oxidative stress can result from an inflammatory response and from exposure to certain medications and foods that have high oxidizing potential. Common foods and drugs include fava beans, sulfonamides, primaquine, and anti tuberculosis medications. Because there is no glutathione to protect red blood cells from oxidation, hemoglobin within the red blood cells become denatured forming Heinz bodies. Macrophages in the spleen remove Heinz bodies leading to characteristic bite cells. This disease is more prevalent in Black people because an advantage of this disease is that it confers protection against malaria, particularly Plasmodium falciparum. It is thought that cells infected with the parasite are cleared more rapidly by the spleen giving individuals with the G6PD deficiency gene an evolutionary advantage in malarial endemic environments.

PLAY PICMONIC

X-linked Recessive
X-suit with Recessive-chocolate
This disease is inherited in an X-linked recessive pattern. X-linked inheritance means that the gene causing the trait or the disorder is located on the X chromosome, and because it is recessive, boys are more likely to have this disease.

Hemolytic Anemia
Hemolysing-RBCs from Anemone
Lack of NADPH and glutathione caused by G6PD deficiency causes red blood cells to be susceptible to oxidative damage causing hemolysis.

Inflammatory Response
Lighter in Flames
Inflammatory states can increase production of reactive oxygen species, causing hemolytic anemia in patients with G6PD deficiency in times of infection.

Fava Beans
Fava beans
Fava beans contain alkaloids that can induce a state of oxidative stress, causing hemolytic anemia in patients with G6PD deficiency.

Sulfonamides
Sulfur-match-fondue
Sulfonamides are a group of antibiotic drugs that contain the sulfonamide group. Because the sulfonamide group can act as an oxidant, sulfonamides can induce states of oxidative stress, causing hemolytic anemia in patients with G6PD deficiency.

Primaquine
Prom-queen
Primaquine is a drug used in the treatment of malaria that can generate reactive oxygen species within red blood cells, causing hemolytic anemia in patients with G6PD deficiency.

Anti TB Drugs
Ant-tie on TB-TV
Para aminosalicyclic acid is an anti-tuberculosis drug that can induce a state of oxidative stress, causing hemolytic anemia in patients with G6PD deficiency.

Heinz Bodies
Heinz-ketchup-bottle
Heinz bodies are small round inclusions seen in red blood cells formed by denatured hemoglobin, usually through oxidant damage. Heinz bodies can also be seen in chronic liver disease.

Bite Cells
Bite out of a Cell
Heinz bodies are cleared by macrophages in the spleen via removal of the inclusion and the damaged membrane, leading to characteristic bite cells. Bite cells are defined as abnormally shaped red blood cells with semicircular portions removed from the cell margin, as if someone took a bite out of the cell.

Prevalent Among African Americans due to Increased Malarial Resistance

African American swatting Malaria-mullet-mosquito

This disease is more prevalent in African Americans because an advantage of this disease is that it confers protection against malaria, particularly Plasmodium falciparum. It is thought that cells infected with the parasite are cleared more rapidly by the spleen giving individuals with the G6PD deficiency gene an evolutionary advantage in malarial endemic environments. Other susceptible populations include Kurdish Jews, Sardinians, Nigerians, and Thai people.