

G6P Dehydrogenase Deficiency:

- Also known as FAVISM

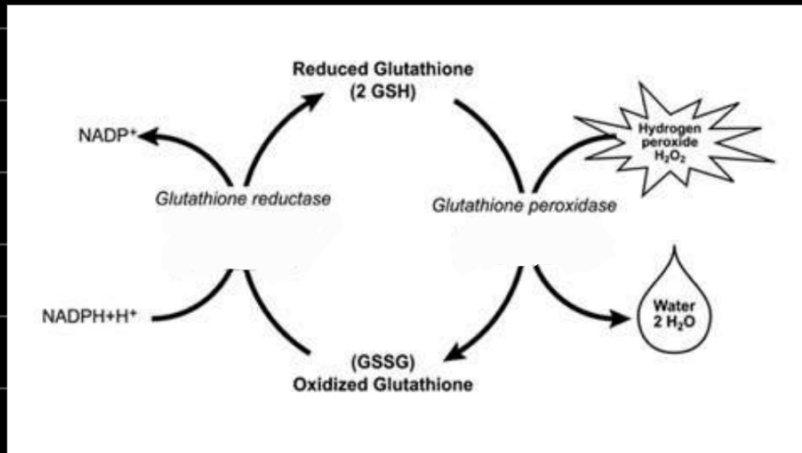
- it is X - Linked recessive genetic condition

- Inborn Error

- Congenital

- Inherited Disorder

- results in reduced intracellular NADPH level which participates in the glutathione cycle to protect cells against hydrogen peroxide.



- NADPH in RBCs is important to keep a high ratio of the reduced glutathione which is vital to protect cells from damaging effect of ROS (detoxification process)
- G6PD enzyme prevents oxidative damage
- G6PD deficiency is characterized by hemolytic anemia, especially in state of oxidative stress such as exposure to infection, some medications and certain foods (e.g. broad or fava beans).
- Oxidative stress is due to imbalance between the generation of ROS or free radicals (e.g. H₂O₂, .OH,...) and the removal by specific cellular enzymes (antioxidants) like glutathione peroxidase (enzyme abundant in cells).
- Oxidative stress depletes the reduced form of glutathione (GSH) and G6P dehydrogenase deficiency disorder can not supply enough NADPH to regenerate GSH from the oxidized one (GSSG) .
- Damaged RBCs are recycled to the spleen. The hemoglobin is metabolized to bilirubin causing jaundice in high concentration .

THE END

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