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# G6PD LEVEL DEFICIENCY IN MALRIA

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**Abstract:**

*Glucose-6-phosphate dehydrogenase (G6PD) deficiency is the most common hereditary enzyme disorder and morethan 200 million people have a deficiency of this enzyme. G6PD deficiency is an X-linked enzyme defect, and one of its main signs is the presence of hemolytic anemia. It is a worldwide important cause of neonatal jaundice and causes life threatening hemolytic crisis in childhood. At later ages, certain drugs such as anti-malarial drugs and fava beans cause hemolysis among G6PD deficiency patients. The frequency and severity is influenced by genetic and cultural factors. It is common in Mediterranean, African and some East Asian populations but rare in Bangladeshi peoples. Genetic counseling may be of benefit for patients and their families. Other treatment is symptomatic and supportive.*

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**INTRODUCTION:**

Hemolytic anemia in certain susceptible individuals after ingestion of anti-malarial drugs was first reported in 1926. In the 1950s the cause of the hemolysis was considered to be inside the red cells. It has been proved that the cause of hemolysis is due to the decreased level of glucose-6 phosphatase dehydrogenase (G-6PD)) in the red cells (Banner RL, 1996).

Now the days, G-6PD deficiency is the most common hereditary enzyme deficiency, causing hemolysis. Glucose-6-phosphate dehydrogenase (G6PD) deficiency affects over 200 million people around the world. G6PD lack is a hereditary disorder that regularly influences males. It happens when the body does not have enough of a protein called glucose-6-phosphate dehydrogenase (G6PD). G6PD enables red platelets to work. It additionally shields them from substances in the blood that could hurt them. In individuals with G6PD lack, either the red platelets do not make enough G6PD. Without enough G6PD to secure them, the red platelets break separated. This is called hemolysis. At the point when numerous red platelets are decimated, an individual can create hemolytic paleness. This can cause tiredness, tipsiness, and different side effects.

Red platelets that don't have enough G6PD are sensitive to certain meds, sustenance's, and infection. At the point when these things trigger a fast loss of red platelets over a brief span, it's known as a hemolytic emergency. In these cases, the side effects stop when the reason is no more. In uncommon cases, G6PD lack prompts perpetual pallor paying little respect to introduction to triggers (Elena, 2018)

Signs and Symptoms of G6PD Deficiency are, most individuals with G6PD insufficiency dont have any indications. Others may have side effects of hemolytic weakness if numerous RBCs are obliterated whiteness (in darker-cleaned kids, pallor is at times best found in the mouth, particularly on the lips or tongue) ,outrageous tiredness or wooziness, rapid heartbeat, shortness of breath, jaundice a developed spleen. Mellow indications as a rule need not bother with restorative treatment. As the body makes new red platelets, the sickliness will improve. In the event that indications are progressively serious, a youngster may require care in a hospital (Gaston, 2016).

Some causes G6PD deficiency are Glucose-6-phosphate dehydrogenase inadequacy results from transformations in the G6PD quality. This quality gives directions to making a protein called glucose-6-phosphate dehydrogenase. This catalyst is engaged with the ordinary handling of starches. It likewise shields red platelets from the impacts of conceivably destructive particles called receptive oxygen species, which are side-effects of ordinary cell capacities. Synthetic responses including glucose-6-phosphate dehydrogenase produce exacerbates that keep receptive oxygen species from structure up to lethal dimensions inside red platelets.

In the event that changes in the G6PD quality lessen the measure of glucose-6-phosphate dehydrogenase or adjust its structure, this catalyst can never again assume its defensive job. Accordingly, responsive oxygen species can collect and harm red platelets. Factors, for example, diseases, certain medications, or ingesting fava beans can build the dimensions of receptive oxygen species, making red platelets be annihilated quicker than the body can supplant them. A decrease in the quantity of red platelets causes the signs and side effects of hemolytic sickliness (National Library of Medication, 2017)

G6PD is an enzyme involved in the pentose monophosphate pathway. G6PD deficiency leads to free radical–mediated oxidative damage to red blood cells, which in turn causes hemolysis. This condition is inherited in an X-linked recessive pattern. The gene associated with this condition is located on the X chromosome, which is one of the two sex chromosomes. In males (who have only one X chromosome), one altered copy of the gene in each cell is sufficient to cause the condition. In females (who have two X chromosomes), a mutation would have to occur in both copies of the gene to cause the disorder. Because it is unlikely that females will have two altered copies of this gene, males are affected by X-linked recessive disorders much more frequently than females. A characteristic of X-linked inheritance is that fathers cannot pass X-linked traits to their sons (National Library of Medication, 2017)

The diagnosis depends upon demonstrating decreased activity of the G6PD enzyme through either a quantitative assay or a screening test such as fluorescent spot test Molecular genetic testing can detect mutations in the specific gene known to cause G6PD, but is available only as diagnostic service at specialized laboratories. If doctors suspect a person is G6PD-deficient, they will conduct a variety of blood tests to confirm a diagnosis and rule out other conditions that cause similar conditions. A diagnosis is based upon the identification of characteristic physical findings and symptoms, a thorough clinical evaluation, a detailed patient history, and/or

specialized tests.(National Organization of Rare Disease, 2017)

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**Case Report:**

A Three years old boy was admitted at department of pediatrics of Private Hospital of Lahore because of fever, cough, and sputum for three days and gross hematuria for one day. He is the second brother of two siblings. Prior to this admission, he was admitted due to hemolytic anemia with acute tonsillitis one and half years back. On admission, he looked actually ill, the conjunctivae were pale, the sclera was icteric and throat was infected. The lung sound was coarse and the liver and spleen were not palpable. The laboratory findings are as follows Hb 7.8 gm/dl, Hct 27.3%, reticulocyte count 3.6%, haptoglobin was under 37 mg/dl and there was a picture of hemolytic anemia. The chest X-ray showed pneumonic consolidation on lower lobe and the peripheral blood smear showed microcytic hypochromic anemia, nucleated RBCs, polychromatic, spherocytosis and anisocytosis. On urinalysis, RBC was (++) and protein was (+++). The cold agglutinin test and Combs' test were negative. On the G-6 PD Assay (Dye reduction test) by SIGMA kit, normal control was 30 minutes. However the patient time was prolonged to 6 hours. On the quantitative test for G-6-PD, normal value was 4.60 - 13.50 U/ g Hb but the patient's value was 2.11 U/ g Hb. to maintain the pathway to generate a chemical called glutathione, which in particular form is an antioxidant. The antioxidant is necessary to protect the cell's hemoglobin and its cell wall (red cell membrane). If the level of antioxidant is too low, then the cell's hemoglobin will not bind oxygen (its main purpose); the cell wall will break allowing the cell contents, including the modified hemoglobin, to spill out7.The severity of symptoms associated with G6PDD varies greatly from case to case, depending upon the form of the disorder that is present, and some people have no symptoms at all. When symptoms are present, they may include fatigue, pale color, and shortness of breath, rapid heartbeat, jaundice or yellow skin color, dark urine and enlarged spleen. In the relatively rare, severe, potentially life-threatening cases, symptoms include, in addition to those listed above, others such as: blood in the urine (hemoglobinuria), shock, kidney (renal) failure and congestive heart failure in which the heart is unable to pump blood effectively throughout the body. Most affected individuals, when exposed to fava beans, will experience severe episodes of hemolytic anemia due to such exposure. Chromosomes, which are present in the nucleus of human cells, carry the genetic characteristics of each individual.

**DISCUSSION:**

The diagnosis depends upon demonstrating decreased activity of the G6PD enzyme through either a quantitative assay or a screening test such as fluorescent spot test Molecular genetic testing can detect mutations in the specific gene known to cause G6PD, but is available only as diagnostic service at specialized laboratories. If doctors suspect a person is G6PD-deficient, they will conduct a variety of blood tests to confirm a diagnosis and rule out other conditions that cause similar conditions. A diagnosis is based upon the identification of characteristic physical findings and symptoms, a thorough clinical evaluation, a detailed patient history, and/or specialized tests (National Organization of Rare Disease, 2017)

Most affected people don't require treatment. G6PD insufficiency is regularly best overseen by preventative measures. People ought to be screened for the G6PD deformity before being treated with specific antibiotics, for example, certain anti-microbial, antimalarial and different drugs known to trigger hemolysis in G6PD-lacking people. In people who are G6PD-inadequate, hemolytic frailty from fava beans or from realized medications ought not happen on the grounds that presentation can be maintained a strategic distance from. On the off chance that a scene of hemolytic iron deficiency is because of the utilization of a specific medicine, the causative medication ought to be stopped under a doctor's supervision. On the off chance that such a scene is because of a fundamental contamination, fitting advances ought to be taken to treat the disease being referred to. A few grown-ups may require momentary treatment with liquids to anticipate hemodynamic stun (in which there is lacking supply of blood to the organs) or, in serious situations where the rate of hemolysis is fast, even blood transfusions.

Blood transfusions are bound to be shown in kids than grown-ups, and in kids with fauvism can demonstrate life-sparing. Neonatal jaundice is treated by setting the newborn child under exceptional lights (bili lights) that mitigate the jaundice. In progressively serious cases, a trade transfusion might be essential. This strategy includes evacuating an influenced newborn child's blood and supplanting it with crisp benefactor blood or plasma. Hereditary advising might be of advantage for patients and their families (National Organization of Rare Disease, 2017)

The fundamental treatment for G6PD inadequacy is shirking of oxidative stressors. Infrequently, sickliness might be extreme enough to warrant a blood transfusion. Splenectomy for the most part isn't prescribed. Folic corrosive and iron possibly are valuable in hemolysis, in spite of the fact that G6PD inadequacy more often than not is asymptomatic and the related hemolysis normally is brief. Cell reinforcements, for example, nutrient E and selenium have no demonstrated advantage for the treatment of G6PD insufficiency. Research is being done to distinguish drugs that may repress oxidative-actuated hemolysis of G6PD-inadequate red blood cells (JENNIFER, 2005).

## CONCLUSION:

G6PD deficiency causes problems primarily when the deficiency is complicated by the treatment of malaria. In rare cases, G6PD deficiency leads to chronic anemia regardless of exposure to triggers. Most people with G6PD deficiency don't have any symptoms. Others might have symptoms of hemolytic anemia if many RBCs are destroyed paleness (in darker-skinned kids, paleness is sometimes best seen in the mouth, especially on the lips or tongue) extreme tiredness or dizziness fast heartbeat fast breathing or shortness of breath jaundice (the skin and eyes look yellow) an enlarged spleen dark, tea-colored pee Mild symptoms usually don't need medical treatment. A reduction in the number of red blood cells causes the signs and symptoms of hemolytic anemia. G6PD is an enzyme involved in the pentose monophosphate pathway. G6PD deficiency leads to free radical–mediated oxidative damage to red blood cells, which in turn causes hemolysis. Some causes of G6PD Deficiency are Glucose-6-phosphate dehydrogenase deficiency results from mutations in the G6PD gene. Some adults may need short-term treatment with fluids to prevent hemodynamic shock (in which there is inadequate supply of blood to the organs) or, in severe cases where the rate of hemolysis is very rapid, even blood transfusions. It also protects red blood cells from the effects of potentially harmful molecules called reactive oxygen species.

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