Management of a patient with Glucose-6-Phosphate Dehydrogenase Deficiency. A Case Study

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INTRODUCTION

- Glucose 6-phospahte dehydrogenase (G6PD) deficiency is an enzymatic disorder of red blood cells in humans.
- G6PD causes a spectrum of diseases including neonatal hyperbilirubinemia, acute hemolysis, neonatal jaundice, and chronic hemolytic anemia (a pathological condition known as methahemoglobinemia).
- About 400 million people are affected by this deficiency worldwide; in the United States of America, black males are commonly affected with a prevalence of around 10% and G6PD is an X-linked inherited disorder which has a higher prevalence in African, Asian, Mediterranean, or Middle Eastern descent.
- In glucose 6-phosphate dehydrogenase (G6PD) patients, a cytoplasmic enzyme prevents oxidative damage to cells by promoting detoxification of free radicals. The most common allergies associated with this condition include sulfa, aspirin and flava beans.
- There is a dearth of information on safe pharmacologic agents used in conscious sedation of a patient with G6PD without causing acute hemolytic events (methahemoglobinemia). Thus, the aim of this case study is to review management protocols for the safe and effective treatment of patients with G6PD.

THE CASE

- **Presenting Patient:** 5 Years, 5 months African American who male presented to the Pediatric Dental Clinic of Howard University College of Dentistry.
- **Chief Complaint:** Mom reported my son had episodes of mild dental-pain especially during eating.
- **Medical History:** Glucose-6-phospahate dehydrogenase deficiency (G6PD) with no history of acute hemolysis. Patient is allergic to sulfa, aspirin and flava beans.
- **Extra oral examination:** No lymphadenopathy, no facial swelling, and no facial asymmetry
- **Intra oral Examination:** Revealed early mixed dentition, poor oral hygiene with high plaque accumulation. Deep cavities in all patient's primary posterior teeth which correlated with radiographical evidence. Patient has a Brodsky of I and Mallampati of class I
- **Behavior:** Frankl 2 which necessitated a pharmacologic behavior management.
- **Treatment** : We had a medical consult with patient's primary care provider and all treatment completed under Conscious sedation with midazolam (1.0 mg/Kg) and hydroxyzine (1.0 mg/Kg).

LITERATURE REVIEW

- This review of literature is an electronic database search done on PubMed to obtain relevant published information on safe pharmacologic agents for use in conscious sedation of a patient with G6PD without causing acute hemolytic events.
- Keyword phrases searched include **Glucose 6 phosphate dehydrogenase, G6PD, Glucose 6 phosphate dehydrogenase and dental management, Glucose 6 phosphate dehydrogenase and Favism, Glucose 6 phosphate dehydrogenase and hemolytic anemia, Glucose 6 phosphate dehydrogenase and conscious oral sedation.**
- Important information was extracted to meet the objectives of the literature, viz. background information on Glucose 6 phosphate dehydrogenase, pathophysiology and clinical manifestation.

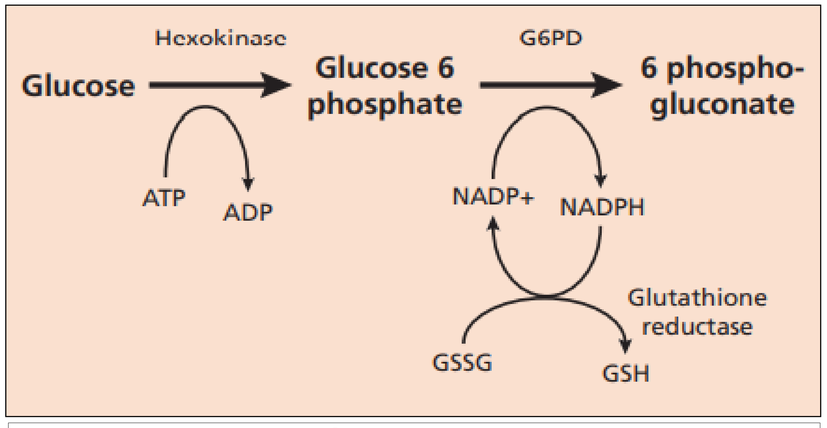

Figure 1. The Pentose Phosphate Pathway.[5] G6PD catalyzes NADP+ to its reduced form, NADPH, in the pentose phosphate pathway. (G6PD = glucose-6-phosphate dehydrogenase; ATP = adenosine triphosphate; ADP = adenosine diphosphate; NADP+ = nicotinamide adenine dinucleotide phosphate [oxidized form]; NADPH = reduced NADP; GSSG = oxidized glutathione; GSH = reduced glutathione

Table 1: Classes of Glucose-6-phosphate dehydrogenase (G6PD) enzyme Variants

Class	Level of deficiency	Enzyme activity	Prevalence
I	Severe	Chronic nonspherocytic hemolytic anemia in the presence of normal erythrocyte function	Uncommon; occurs across populations
II	Severe	Less than 10 percent of normal	Varies: more common in Asian and Mediterranean populations
III	Moderate	10 to 60% of normal	10% of blacks in the United States
IV	Mild to none	60 to 150% of normal	Rare
V	None	Greater than 150% of normal	Rare

DISCUSSION

- There is dearth of information on the drugs used for the safe pharmacologic behavior management of patients with G6PD deficiency since it was discovered by Carson *et al*, in 1956.
- The detailed mechanism of acute hemolytic crisis is not fully known and understood, but it certainly can result from the inability of G6PD- deficient cells to withstand the oxidative damage produced directly or indirectly by an exogenous trigger.
- Through our search of literature, medications such as prilocaine, topical benzocaine, acetaminophen, aspirin, penicillin, and antimicrobial agents such as sulphonamides have all been implicated to cause acute hemolytic crisis or methahemoglobinemia.
- However, midazolam and hydroxyzine which belong to the class of benzodiazepines and antihistamine respectively has no history of precipitating acute hemolytic crisis or (methahemoglobinemia). On using these two medications, we adhered with the dose outlined in the AAPD handbook, as a dose of 1.0 mg/Kg of hydroxyzine and 0.5 mg/Kg of midazolam were used.
- Though our patient cried throughout the procedure and sedation experience was deemed ineffective, the drugs combination proved successful in the treatment of our patient because acute hemolytic crisis did not occur.

CONCLUSION

- Patients with G6PD deficiency who are anxious and have lower Frankl scores, will benefit from pharmacologic behavior management using midazolam and hydroxyzine.
- When treating a G6PD deficiency patient, the best management strategy to prevent acute hemolytic crisis or methahemoglobinemia are to avoid using any pharmacologic drugs that will precipitate acute hemolytic crisis or methahemoglobinemia
- In addition, it is necessary for pediatric dentists to know how to recognize children with suspected G6PD or favism, not only for counseling the parents, but also for taking all precautions regarding dental restorative treatment, particularly whenever pharmacologic behavior management is indicated.
- Furthermore, to help provide care for these patients, medical consult with the primary care provider should be exploited when in doubt.

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