



Glucose-6-Phosphate Dehydrogenase Deficiency in the Setting of Cardiac Surgery

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Background

- Glucose-6-Phosphate Dehydrogenase (G6PD) deficiency is an X-linked genetic disorder that afflicts >500 million people
- G6PD is associated with **hemolysis and premature cardiovascular disease** due to the **inability to mitigate excess oxidative stress**
- Few case reports discuss cardiac surgery in this population and orthotopic heart transplant has not been described
- Cardiac surgery and **CPB** are **underappreciated sources of oxidative stress**

Case Description

- A 51 YM presented with angina, EKG demonstrated STEMI, and angiography revealed an occlusive pLAD thrombus with embolization to the LCx. During thrombectomy and balloon angioplasty he suffered cardiac arrest.
- After ROSC, a percutaneous left ventricular assist device was placed. **His course was complicated by hemolysis raising suspicion for G6PD deficiency** (Figure 1A) and he was transferred to our institution for ECMO and heart transplant evaluation. He was cannulated on ECMO and listed UNOS status 2E.
- He received an **orthotopic heart transplant** 22 days after presentation.
- **Intraoperatively, he developed hyperthermia, dark urine, and his hemoglobin decreased, raising concern for ongoing hemolysis.**
- **A partial exchange transfusion with oxygenated blood was used to reduce ongoing hemolysis**
- Quantitative analysis of his G6PD function was performed 3 months later, his mean activity was 18% of normal, confirming the suspected G6PD deficiency diagnosis.

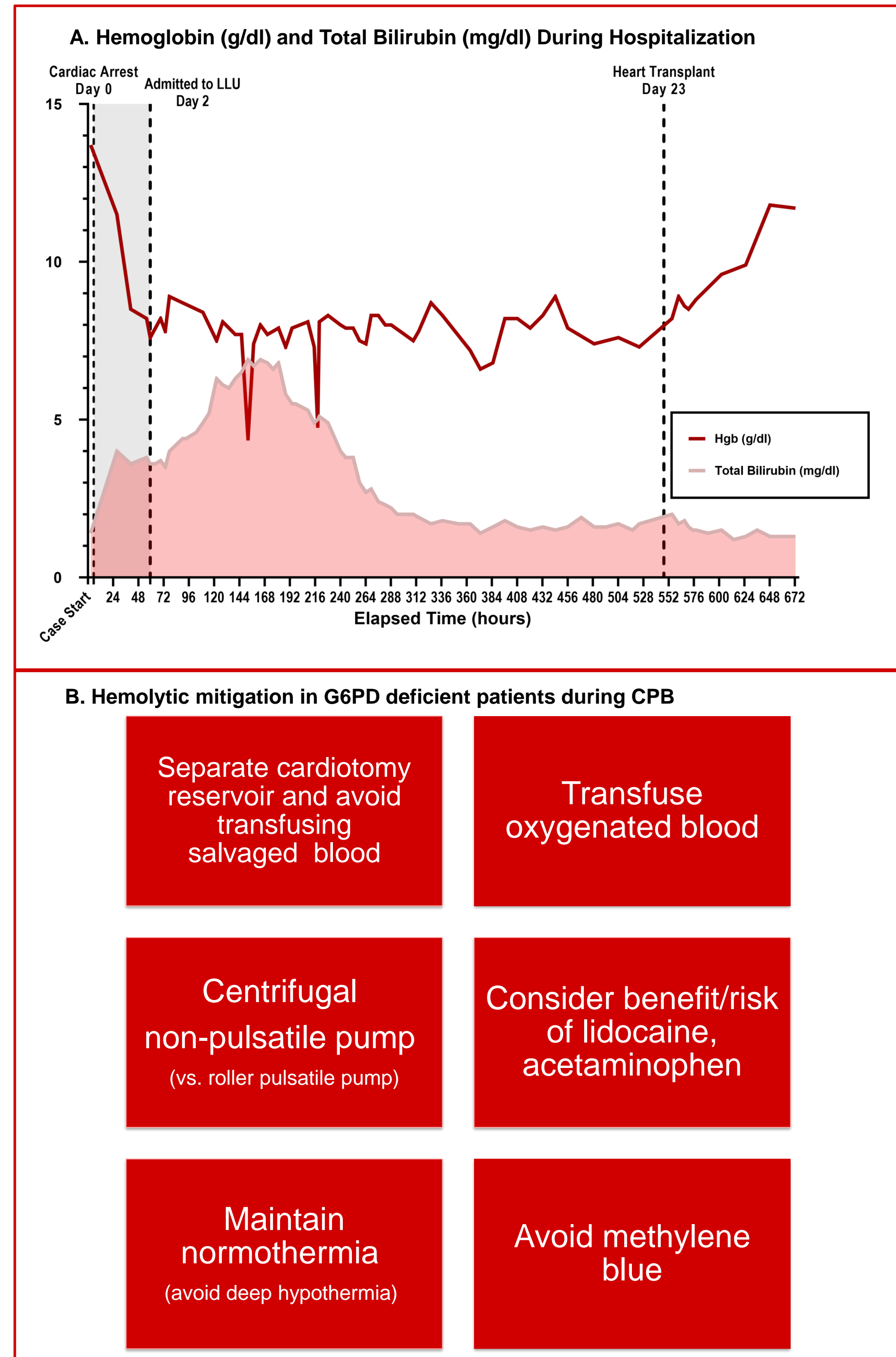


Figure 1. A. Hemoglobin (Hgb, g/dL) and total bilirubin concentration (mg/dL) during hospitalization, admission = October 25, 2023 (Day 0); gray shading indicates length of time not in our facility **B.** Interventions for mitigating hemolysis in G6PD deficient patients during Cardiopulmonary Bypass (CPB)

Discussion

- G6PD deficiency cannot be diagnosed during acute hemolysis and requires a high-index of suspicion.
- Hypothermia, transfusion of deoxygenated or traumatized red blood cells may encourage development of ROS and precipitate hemolysis (Figure 1B).
- CPB is an underrecognized precipitant of oxidative stress and may trigger hemolysis in G6PD deficient patient.

Conclusion

- Intraoperative management should aim to reduce oxidative stress.
- Partial exchange transfusion may be helpful during an acute crisis.

References

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