TWO PEDIATRIC CASES OF THERAPEUTIC EXCHANGE TO TREAT ELEVATED PLASMA-FREE HEMOGLOBIN

Tabish Nabil[1], Stephens S, [2] Annen Kyle[1,2]

Department of Pathology, [1] University of Colorado Anschutz Medical Campus & [2] Childrens Hospital of Colorado, Aurora, Colorado, USA

PURPOSE

When RBCs break down in the bloodstream, hemoglobin (hgb) is released into the plasma. Haptoglobin is the primary protein that binds to hgb, followed by albumin. The reticuloendothelial system rapidly clears hgb from the plasma. In cases of massive intravascular hemolysis, hgb clearance mechanisms may be overwhelmed, accumulating excess plasma free-hgb, which may lead to acute kidney injury and acute tubular necrosis. We present two pediatric cases where Therapeutic Plasma Exchange (TPE) decreased plasma free-hgb.

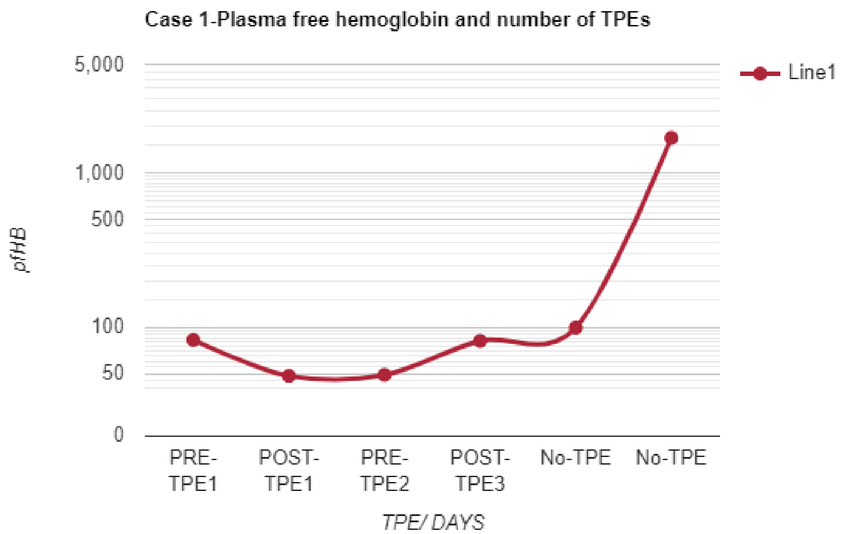
METHODS: CASE 1

Case 1: A 19-day-old male infant born at full term with a genetic deletion of 2q24.3-2q32.1, which includes coarctation of the aorta and bicuspid aortic valve. The patient also had Methicillin Susceptible Staphylococcus Aureus (MSSA) bacteremia and E. Coli UTI, which led to multi-organ failure and Disseminated Intravascular Coagulation (ISTH DIC score 5). Over five days, the patient's platelet count dropped precipitously from 182K to 32K along with multiorgan dysfunction, raising concern for thrombocytopenia-associated multi-organ failure (TAMOF). The patient was started on extracorporeal membrane oxygenation (ECMO) without improvement. The clinical team requested TPE for elevated plasma-free hemoglobin and severe thrombocytopenia. Laboratory tests revealed intravascular hemolysis with elevated Total Bilirubin (29.8 mg/dL) and LDH (6363 U/L) with Serum Creatinine at 1.02 mg/dL. TPE was performed once daily for three days. The plasma hemoglobin (normally 0-3 mg/dL) was 82.6 (pre-TPE-1) & 48.3 (post-TPE-1) at TPE-2 and 49.1 (pre-TPE-2) & 81.8 (post-TPE-3) at the end of the cycle. After completing these procedures, the plasma hemoglobin exponentially elevated to 99.2 and reached a zenith of 1672.5 in the next 2 days: on this day, the parents opted to withdraw care, and the infant passed away.

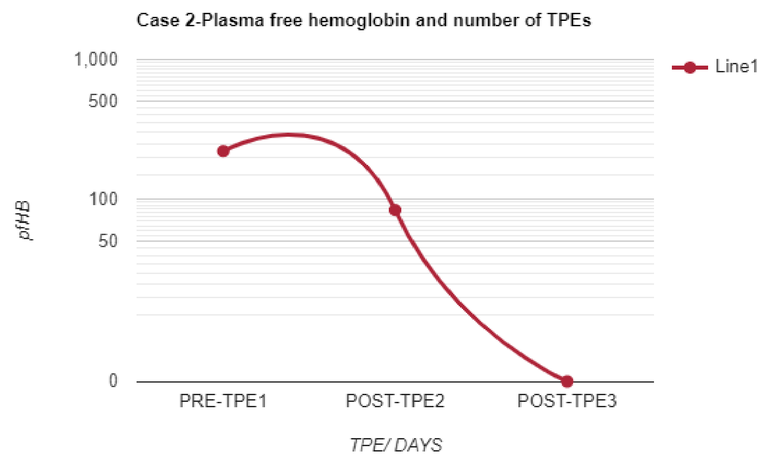
METHODS: CASE 2

Case 2: A 2-day-old female presented with left ventricular failure of uncertain etiology and was put on venoarterial ECMO. She required fresh frozen plasma (FFP), pRBCs, and platelets due to her initial coagulopathy and bleeding. Blood type was O Rh positive, DAT was negative, and antibody screen was negative. Her blood smear showed a few schistocytes with morphologically normal RBCs. She also developed an intraventricular hemorrhage on head ultrasound. She had significant plasma-free hemoglobin levels and elevated LDH without signs of mechanical hemolysis. As her plasma-free hemoglobin was persistently high, the clinical team requested one 2-volume plasma exchange emergently in tandem with the ECMO circuit. PreTPE, the plasma-free hgb was 221 mg/dL; the value was 83.6 on day two after TPE and 0.0 on day three after TPE. Serum creatinine remained within normal limits. The patient made a recovery and is achieving her age-appropriate milestone

GRAPHIC REPRESENTATION



GRAPHIC REPRESENTATION



RESULTS

We demonstrate that TPE reduces plasma-free hemoglobin. Since plasma-free hgb is not dialyzable, TPE may be a viable option for treating elevated plasma-free hgb in critical care.

CONCLUSION

Plasma-free hgb is not categorized in the ASFA guidelines. However, pathophysiology and premise are comparable to Acute Hemolytic Anemia, a Category III indication. Hemoglobinuric acute kidney injury can occur due to several mechanisms, including direct proximal tubular cell toxicity cast formation and other mechanisms and vasoconstriction resulting from plasma free-hgb nitric oxide scavenging. TPE appears effective in reducing plasma-free hemoglobin. However, only one case has previously been reported in the literature. Additional evaluation is needed to determine whether TPE should be recommended as a treatment method.