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## 401.BLOOD TRANSFUSION

**Successful Use of Therapeutic Plasma Exchange Following Red Blood Cell Exchange for Severe Acute Chest Syndrome: A Single Institution Experience**

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**Introduction:** Severe acute chest syndrome (ACS) is a potentially fatal complication of sickle cell disease (SCD) that can progress to multiorgan failure. Treatment includes red blood cell (RBC) exchange to improve oxygenation and reduce vaso-occlusion by decreasing the proportion of hemoglobin (Hb) S. Patients with poor response to RBC exchange are at high risk for mortality.

Therapeutic plasma exchange (TPE) may be beneficial in cases of severe organ dysfunction refractory to RBC exchange. TPE is postulated to remove pro-inflammatory cytokines, reduce circulating free hemoglobin, and decrease complement products that contributes to vaso-occlusion and inflammation. The use of TPE for severe ACS and multiorgan failure has been rarely described in adult patients and is even less commonly described for pediatric patients.

**Aim:** To describe use of TPE for the management of severe ACS with multiorgan failure in two pediatric patients and one young-adult patient who did not respond to standard medical management with RBC exchange transfusion.

**Methods:** Case reports

**Case 1:** A six-year-old male with homozygous SCD (HbSS) and moderate persistent asthma was admitted for ACS. His course was complicated by hypoxemia requiring intubation, oliguria, and hypotension. On hospital day 3, he underwent RBC exchange with four units of RBCs. Despite decrease of Hb S to 15%, he remained hypoxic on high ventilatory support and developed acute respiratory distress syndrome. After multidisciplinary discussion, TPE was trialed in hopes of improving oxygenation and reversing multiorgan dysfunction. He underwent 3 TPEs, each with 1.5 total plasma volume (TPV) using 5% albumin (days 4, 5 and 7). After a single session of TPE, his oxygenation saturations improved to the 90s, and his ventilator support was weaned. He was extubated on day 8 and was discharged on day 13 in stable condition.

**Case 2:** A sixteen-year-old male with HbSS, recurrent hyper-hemolytic syndrome, and sickle cell hepatopathy was admitted for medical optimization before scheduled cholecystectomy. His Hb was raised pre-operatively with erythropoietin, in lieu of RBC transfusion, due to history of severe refractory hyper-hemolysis with transfusions. He underwent uncomplicated cholecystectomy on hospital day 2. On day 3, he developed ACS, septic shock, and an exacerbation of known hepatopathy. He was intubated and underwent RBC exchange with 8 units of RBCs. His Hb S decreased to 20% but he had no improvement in clinical status. High-dose steroids and TPE were then trialed. The use of TPE was dual purposed to reduce inflammatory cytokines contributing to systemic dysfunction and to prevent hyper-hemolysis following exchange transfusion. He underwent 3 TPEs each with 1.5 TPV using 5% albumin for three days (days 5, 6, and 7). Throughout his course of TPE, he was extubated (day 6), weaned off pressor support (day 6), and his liver dysfunction began to improve (day 7). He was discharged home on day 16.

**Case 3:** A thirty-five-year-old female with HbSS without chronic complications was transferred from an outside hospital where she presented 3 days prior with ACS and progressive neurologic deterioration with minimal responsiveness. Imaging revealed diffuse pulmonary infiltrates and extensive cerebral micro emboli consistent with fat embolism syndrome. She underwent RBC exchange on the day of arrival to our facility (hospital day 4) with 5 units of RBCs. Her Hb S decreased to 20% but she had no improvement in neurologic function. After multidisciplinary evaluation, TPE was trialed alongside high-dose steroids. She underwent 5 TPEs each with 1.0 TPV using 5% albumin (days 5, 6, 8, 9, and 12). Shortly after initiating TPE, she began recovering

neurologically with eye-opening, response to noxious stimuli, and tracking (day 6). After 3 days of TPE, CRP fell from 21 to 5 and ESR from 49 to 23 (day 8). She followed commands on day 10 and was taken off oxygen on day 15. She was discharged to a rehabilitation facility on day 30 with significant improvements in mental status, cognition, and functional abilities.

Conclusions: TPE for three to five days should be considered in patients with severe ACS and multiorgan failure with high risk of mortality when standard management fails. The early initiation of TPE likely contributed to the successful recoveries of these three cases. Further studies are needed to delineate the role of TPE in the management of acute complications of SCD.

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