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PLASMA EXCHANGE TREATMENT FOR CARDIOGENIC SHOCK IN REFRACTORY KAWASAKI DISEASE: CASE SERIES.

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Introduction: Kawasaki disease (KD), known to complicate coronary artery lesions (CALS) in children, is widely treated by immunoglobulin (IVIG). However, some develop severe hypotensive shock despite this therapy. Plasma exchange (PEX) has been applied for IVIG-resistant KD.

Objectives: NA

Methods: A report of case series.

Results: Case 1. A 2-year-old boy, who underwent IVIG treatment of total 3 g/kg, developed cardiogenic shock on day 7. Echocardiogram (UCG) revealed an enlarged right ventricle compressing the left ventricle, and severe tricuspid regurgitation (TR). Elevated troponin-T levels, reciprocal ST change in II, III, aVF on EKG were suggestive of right ventricular infarction. He was transferred to the pediatric intensive care unit (PICU) under inotropic support, where he was intubated and mechanical ventilatory support was commenced. CXR showed cardiomegaly. EKG demonstrated ventricular premature contractions and atrioventricular block. The UCG showed no CALs, similar right-sided heart findings and fair left ventricular function. Cardiac angiography demonstrated no coronary abnormalities, nor signs of infarction. Cardiogenic shock persisted despite additional administration of IVIG of 2 g/kg. Infusions of milrinone and carperitide, and inhaled nitric oxide all failed to improve cardiac function, with a left ventricular ejection fraction (LVEF) of 40%. PEX (1.5 fold plasma volume) was implemented. LVEF improved to 50% with decreased arrhythmias after the initial PEX. LVEF further improved to 60% with normal cardiac rhythms after completing 4 sessions of PEX. He was extubated on day 12, and was discharged on day 25 without CALs during the course. Case 2. A 4-year-old boy, who underwent IVIG treatment of a total of 4 g/kg, developed cardiogenic shock on day 8. He was admitted to the PICU under inotropic support. CXR revealed cardiomegaly, pulmonary congestion, and pleural effusion. UCG showed no CALs, LVEF of 40%, mild pericardial effusion, mild TR, mild MR, and mild pulmonary regurgitation. He was intubated on day 9, and PEX (1.5 fold plasma volume) was implemented. Hypotension and tachycardia improved immediately, and a total of 3 sessions of PEX was completed. Inotropes were discontinued, and LVEF improved to 63%. He was extubated on day 11, and was discharged on day 19 without CALs during the course.

Conclusion: The series of our patients indicate PEX may be effective to recover from cardiogenic shock with IVIG-resistant KD. Precise mechanism of PEX on improving cardiac function remains unknown. We are conducting cytokine and receptor assays in KD patients,
which may provide further understanding of its disease process and effects of therapeutic modalities. Further study in a larger group is warranted.

References: NA