Circular Shunt Physiology: Recognition and Treatment

Introduction: The term circular shunt was first used in 1962 by Shone et al. (1) to describe the pathophysiology resulting from coexistent ventricular septal defect, pulmonary valve stenosis (PS), tricuspid insufficiency (TR), and patent foramen ovale (PFO). In this instance shunted blood returns to the chamber from which it was originated through intracardiac communications without transversing a capillary bed (fig. A). This is manifested by marked cyanosis, tachypnea, ductal dependant pulmonary circulation and diminished cardiac output. Neonates with pulmonary stenosis/ atresia, intact ventricular septum (IVS), persistent ductus arteriosus (PDA) and TR are at risk for developing circular shunt physiology after balloon dilation of the pulmonic valve when pulmonary insufficiency ensues (fig. B). Peripheral perfusion is affected dramatically secondary to decreased cardiac output, resulting in metabolic acidemia.

Case Report:
Case 1: Pulmonary atresia and severe TR. 3.0 kg term neonate with prenatal diagnosis of PS and TR was admitted to cardiac intensive care unit (CICU) on PGE1. Post-natal ECHO revealed pulmonary atresia with no antegrade pulmonary blood flow, dysplastic tricuspid valve leaflets with severe TR, large PDA, and PFO with right to left shunting. On day 3 of life, the child was taken to the catheterization laboratory where he underwent radio-frequency perforation of the pulmonary valve plate followed by balloon valvotomy. On return to the CICU, the child developed metabolic acidemia and bradycardia unresponsive to atropine requiring esophageal pacing. The patient had a cardiac arrest from ventricular fibrillation, was defibrillated after chest compressions and inotropic support was begun. ECHO showed severe pulmonary insufficiency and severe TR, PFO with right to left shunt and large PDA with left to right shunt. The patient was taken emergently to the OR for PDA ligation with prompt improvement in hemodynamics.

Case 2: Critical PS with severe TR. 2.4 kg term neonate with prenatal diagnosis of PS and TR was admitted to CICU on PGE1. Post-natal ECHO confirmed diagnosis of critical PS and demonstrated mild Ebstein’s anomaly of the tricuspid valve with severe insufficiency, 4 mm PDA and 4mm PFO, both with bi-directional shunting. At 24 hrs of life, the child underwent cardiac catheterization and balloon dilation of the pulmonary valve under general anesthesia. Immediately post dilation, oxygen saturation increased and aortic diastolic pressure dropped by 30%. Balloon occlusion of the PDA resulted in a dramatic increase in descending aortic pressure (25mmHg) but a decrease in arterial saturation to 52% on 100% FiO2. PGE1 was discontinued and the patient was extubated on completion of the procedure. Over the first 12 hrs after valvotomy, the child developed a circular shunt with metabolic acidemia, requiring sedation, neuromuscular paralysis and reintubation. Spontaneous ductal closure over the subsequent 48 hrs resulted in improvement in hemodynamics.

Discussion: In neonates with critical PS, IVS, percutaneous balloon valvuloplasty may be the initial treatment of choice (2). However, pulmonary insufficiency is common after this intervention. In neonates with significant TR, the creation of pulmonary insufficiency can result in circular shunt physiology. Both our patients in whom this physiology developed had severe TR prenatally with a structurally abnormal tricuspid valve. Awareness of the potential for this pathophysiology to occur, recognition of its existence and a plan, including the capability for prompt surgical ligation of the PDA and creation of a systemic to pulmonary shunt if cyanosis is present, is warranted.

References: