Role of persistent arterial duct for treatment of neonatal Scimitar syndrome with suprasystemic pulmonary hypertension

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Introduction:
Right ventricular (RV) failure is one of the causes for a poor outcome of neonatal Scimitar syndrome with suprasystemic pulmonary hypertension (ssPH). Here we report on our strategy to use the persistent arterial duct (PDA) for RV decompression.

Case reports:
Two preterm neonates (Pt. 1 31, Pt. 2 35 weeks of gestation) were diagnosed of scimitar syndrome with complete anomalous right pulmonary venous drainage to inferior vena cava, persistent primitive hepatic venous plexus (PPHVP), right pulmonary artery hypoplasia, anomalous systemic arterial supply of parts of the right lung, secundum atrial septal defect (ASD) and PDA. In both patients, anomalous systemic arteries were occluded by transcatheter embolization. In the presence of ssPH despite optimal PH-specific medical therapy, interventional balloon angioplasty of subtotally closed PDA was performed for acute RV decompression. After surgical repair with a pericardial intraatrial baffle and subtotal ASD closure, systolic pulmonary artery pressure declined to half systemic allowing for PDA closure in both patients. In Pt. 1, transcatheter occlusion of a hepatic vein of the PPHVP draining into the Scimitar vein was also performed. At age of 6 months there is no echocardiographic evidence for PH in Pt. 1. In Pt. 2, two restrictive muscular ventricular septal defects leading to increased pulmonary flow were interventionally closed. However, development and recurrence of left pulmonary and Scimitar vein stenosis despite multiple transcatheter interventions led to reappearance of systemic PH in Pt. 2. This patient is in a stable condition at age of 16 months.

Conclusion:
In neonatal Scimitar syndrome with suprasystemic PH, balloon angioplasty of subtotally closed PDA may prevent early RV failure and thus improve the poor outcome of this entity.