Objectives: Congenital portosystemic venous shunt (CPSVS) is a rare disease with a variety of connections. Some patients of CPSVS presented with pulmonary arterial hypertension (PAH) which is a life-threatening disorder. The clinical perspective of PAH secondary to CPSVS is unknown. The purpose of this research is to bring out the clinical, laboratory and physiological characteristics of this disorder.

Methods: Nineteen patients with CPSVS were studied about clinical, laboratory and our institution.

Results: The median age at the diagnosis of CPSVS is 3 years. Congenital hypoplasia or absence of portal vein was detected in 11 patients. Eight of the 19 patients were identified to have mild to severe PAH (mean pulmonary artery pressure: $50 \pm 17$ mmHg, pulmonary artery resistances: $15.1 \pm 12.9$ U). The serum total bile acid (TBA) and aspartate aminotransferase (AST) levels of the patients with PAH were significantly higher than those of the patients without PAH (TBA: with PAH $150 \pm 62$, without PAH $64 \pm 45$ µ mol/L, $p=0.0099$ AST: with PAH $52 \pm 19$, without PAH $35 \pm 19$, $p=0.0468$). There was no difference in the blood ammonia levels. As treatment for PAH, bosentan or sildenafil: prostacyclin (n=2) and prostacyclin (n=4) were administered. Catheter embolization of the shunt was conducted in 2 patients with PAH. Three of 8 patients with PAH died because of right heart failure. In only one patient with PAH, pulmonary artery pressure was decreased to almost normal range by catheter embolization.

Conclusions: The CPSVS patients with PAH had high levels of TBA. This suggested that the amounts of shunt flow might be related to the onset of PAH. Appropriate catheter intervention may be an effective treatment for the CPSVS patients with PAH whose portal vein pressure was not elevated by balloon test occlusion of the shunt vessels.