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Portal vein anatomy and pulmonary complications of polysplenia syndrome: difference between patients with biventricular and univentricular heart disease

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Objectives: Polysplenia syndrome is associated with a variety of cardiac anomalies and often accompanied by abdominal and pulmonary vessel abnormalities including portosystemic shunt (PSS), portal vein hypoplasia, pulmonary arteriovenous fistula (PAVF) and pulmonary arterial hypertension (PAH). PSS and PAVF may be congenital or acquired, which makes pathologic condition more complicated. The purpose of this study was to explore the relationship of the type of heart disease and PSS, portal vein hypoplasia, PAVF, and PAH in polysplenia syndrome.
Methods: We conducted a retrospective observational study including 14 consecutive heterotaxia patients with interruption of IVC and azygos or hemiazygos vein connection admitted to our institution during 2002 and 2014. Abdominal and pulmonary vessels were evaluated using ultrasonography, 320-raw multidetector computed tomography and angiography.
Results: Seven patients (age 1-19 years) had biventricular (BV) heart disease and 7 patients (age 0.3-14 years) had univentricular (UV) heart disease. Both BV and UV patients were often complicated by anomalous renal and splenic venous return. In the UV group, PSS was found in 4 patients, PAVFs in 3 patients and PAH in one patient. Two patients had both PSS and PAVFs. The other two patients with PSS did not have PAVF. One patient with PAVF was not accompanied with PSS but demonstrated abnormal pulmonary distribution of hepatic venous flow after Fontan operation with extracardiac total cavopulmonary connenction. Hypoplastic intrahepatic portal vein was found in an infant with the most severe form of diffuse bilateral PAVFs associated with large PSS and PAH prior to any surgical intervention. Ligation of PPS in this patient resulted in incomplete resolution of PAVFs without improvement in PAH. Neither PSS, portal vein hypoplasia, PAVF, nor PAH were found in the BV group.
Conclusions: PAVFs were found in both congenital and postoperative secondary forms. PSS, together with abnormal pulmonary distribution of hepatic venous flow in cavopulmonary circuit in UV heart disease, may contribute to the development of PAVF. A combination of hypoplastic portal vein and PSS may lead to congenital PAVF. Detailed evaluation of the abdominal venous anatomy is crucial in patients with polysplenia syndrome.