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The pediatric interventionist “lost” in the upper abdomen – diagnosis and management of congenital portosystemic venous shunts, liver hemangioma and portal vein thrombosis in children

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Objective: Congenital and acquired diseases affecting the portal vein system and liver hemangioma may become subject of invasive diagnosis and treatment using modern catheter techniques.

Methods: In an institutional case series all consecutive pediatric patients with different types of portosystemic shunt malformations (PSM), liver hemangioma (LH) and portal vein thrombosis (PVT) were reviewed for diagnostic and therapeutic interventional management.

Results: Twelve children at a median age of 2.4 years (range 0-21) and weight 12.6 kg (2.7-62) with different types of PSM (n=6), infantile LH (n=3), and PVT (n=3) were treated between May 2005 and December 2013. Invasive hemodynamic diagnostics focussed on evaluation of pulmonary arterial hypertension (testing) and angiographic diagnostic investigation, which was performed by selective angiography of mesenteric artery, retrograde wedge angiography by occlusion of fistula, or direct angiography of fistula. Catheter interventional treatment in PSM – if suitable – included stepwise partial (n=2) or complete (n=2) occlusion depending on hypoplasia of intrahepatic portal veins using Amplatzer vascular plug, detachable coils, and diabol-shaped covered stents. For LH interventional closure of feeding hepatic arteries was performed using detachable coils. Due to the comorbidity of LH with Kasabach Meritt syndrome we had one postinterventional death.

Conclusions: Diagnostic catheterization with angiography and hemodynamic evaluation provides important information regarding pulmonary and portal hypertension in order to plan any staged catheter interventional partial or complete closure of portocaval fistula, if suitable. Special attention has to be given to neonates with LH, which are severely ill, and target-oriented and quick interdisciplinary team work is necessary to achieve optimal clinical outcome.