Pulmonary arterial hypertension secondary to congenital extrahepatic portosystemic shunt — a serious complication of a rare disease

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Kardiol Pol. 2021;
79 (11): 1288–1289;
DOI: 10.33963/KP.a2021.0096

Received:
July 3, 2021

Revision accepted:
August 30, 2021

Published online:
August 31, 2021

Congenital extrahepatic portosystemic shunt (CEPSS) also known as Abernethy malformation, resulting from the embryonic development of the portal vein, is a rare condition in which splanchnic venous flow bypasses the liver. Volume overload, microthrombosis, and prolonged exposure to vasoactive substances that are normally metabolized by the liver are factors which lead to pulmonary arterial hypertension (PAH) [1, 2].

A four-year-old girl was admitted to the Cardiology Department due to suspicion of pulmonary hypertension. She had been diagnosed prenatally with the absence of venous duct and direct connection of umbilical vein to the right atrium causing fetal heart failure requiring transplacental digoxin treatment.

On admission, she was in World Health Organization Functional Class (WHO-FC) II with the oxygen blood saturation of 97%. Echocardiography revealed elevated right ventricular systolic pressure (RVSP) to 45 mm Hg. The N-terminal pro-brain natriuretic peptide (NT-proBNP) level was low (55 pg/ml, N <190 pg/ml). The typical diagnostic approach to pulmonary hypertension revealed no abnormalities. Despite normal transaminases and coagulation values, specific liver tests were performed based on prenatal history. Hyperammonemia (158 µg/dl, reference range 19–87 µg/dl) and liver ultrasound suggested CEPSS. Abdominal computed tomography (CT) showed a hypoplastic portal vein with hypoplastic branches, a wide connection vessel between the portal vein and inferior vena cava (Abernethy type IIa) (Figure 1A). The patient was accepted for percutaneous embolization of portosystemic shunt. The procedure was done from the internal jugular vein approach. During the balloon occlusion test (temporary shunt closure) (Figure 1B), no changes in the portal vein flow were recorded. On the basis of the diameter of the fully inflated balloon, the size of the implant was selected. Then, using three-dimensional (3D) overlay derived from CT on fluoroscopy (HeartNavigator, Philips Healthcare, Eindhoven, Netherlands) as a roadmap (Figure 1C), Amplatzer Vascular Plug II 16 mm was deployed (Figure 1D). After successful shunt closure, the right cardiac catheterization (Figure 1E) was performed: the mean pulmonary arterial pressure was 37 mm Hg, the mean systemic arterial pressure 53 mm Hg, the capillary wedge pressure 12 mm Hg, pulmonary vascular resistance 4.3 Wood units × m^2, the cardiac index 5.83 l/min/m^2; the acute vasoreactivity test was negative. PAH due to CEPSS was confirmed and bosentan therapy was initiated. During the follow-up, the ammonia level decreased and normalized 3 months after the procedure. Abdominal ultrasound demonstrated no signs of portal hypertension. Unfortunately, after stable 9 months on single PAH therapy, the patient deteriorated (WHO-FC III, NT-proBNP 4804 pg/ml, RVSP 113 mm Hg) (Figure 1F) and required treatment intensification (sildenafil was added). After 24 months of follow-up, the patient is in the intermediate-risk group (WHO-FC II, NTproBNP 283 pg/ml, RVSP 83 mm Hg) on dual PAH therapy currently without the need for treatment intensification.

PAH is a rare, but serious complication of CEPSS leading to RV failure and death [1, 2]. Percutaneous treatment of CEPSS is feasible and allows normalization of the liver

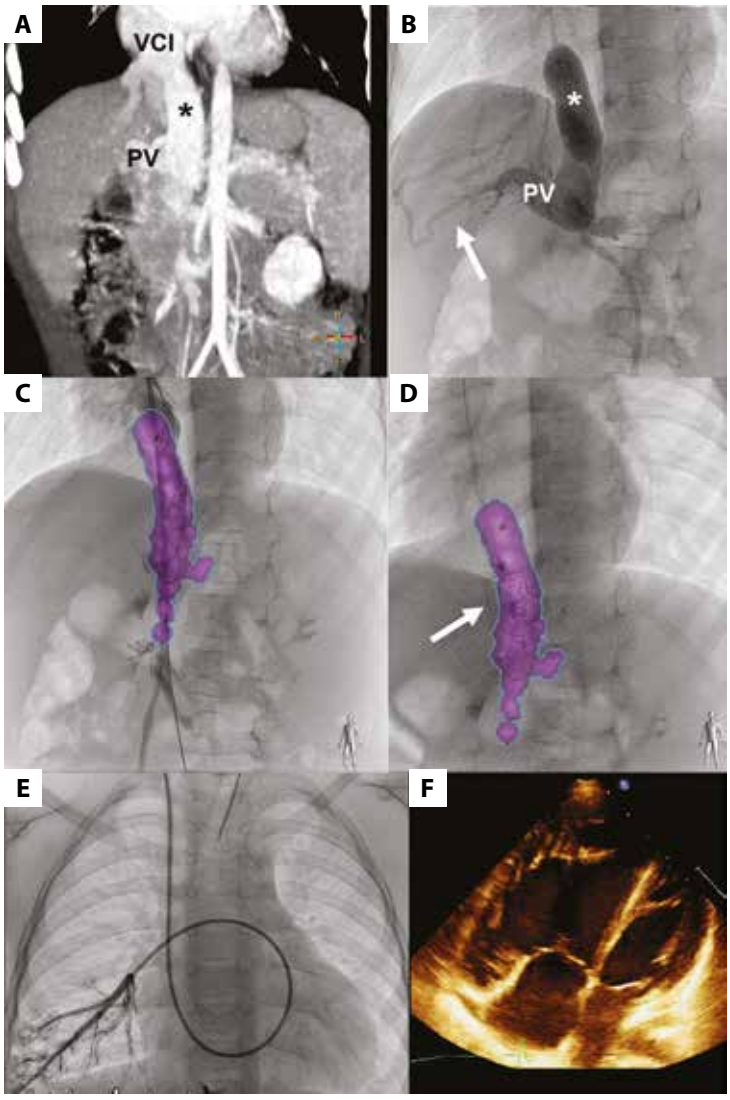

Figure 1. A. Computed tomography of the abdomen shows a large shunt (asterisk) between extrahepatic PV and VCI. **B.** Angiography through an inflated balloon (16 mm) catheter (asterisk) positioned across the shunt shows intrahepatic portal veins (arrow). **C.** Fluoroscopy fusion with CT derived model of the shunt (violet structure) — control angiography through the long sheet to confirm position. **D.** Amplatzer Vascular Plug II (arrow) positioning based on CT derived model overlay. **E.** Peripheral angiography shows decreased arborization of pulmonary arteries. **F.** Echocardiography: the enlarged right ventricle and the right atrium, the left ventricle compressed

Abbreviations: CT, computed tomography; PV, portal vein; VCI, vena cava inferior

function after shunt closure [2, 3]; however, the impact on regression of PAH is not well known. Early diagnosis and shunt closure of CEPSS may prevent the progression or even reverse PAH [4], but in most cases, PAH persisted and required PAH-specific therapies [1–3, 5]. No improvement after early CEPSS closure in this case indicates that volume overload and vasoactive substances may not be the only factors responsible for the development of PAH and the PAH worsening may result from the natural progression of pulmonary vascular disease.

Article information

Conflict of interests: None declared.

How to cite: Migdał A, Koleśnik A, Żuk M, Brzezińska-Rajszys G. Pulmonary arterial hypertension secondary to congenital extrahepatic portosystemic shunt — a serious complication of a rare disease. Kardiol Pol. 2021; 79(11): 1288–1289, doi: 10.33963/KP.a2021.0096.

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