World Journal of Vascular Surgery

Case Study
Published: 12 Mar, 2021

Unexplained Syncope: A Case of Pulmonary Arterial Hypertension Associated with Congenital Extrahepatic Portocaval Shunt

Jiang-you Wang* and Han Chen

Department of Cardiology, Wuhan Asia Heart Hospital and Wuhan Asia General Hospital, China

Case Study

An 18-year-old male smoker without significant medical history presented to the emergency department complaining of post-exercising syncope of 2 weeks. Physical examination showed no obvious positive signs, only A2<P2, P2 slightly hyperactive. Emergency electrocardiogram showed no abnormality. The transthoracic echocardiogram demonstrated mild pulmonary valve and tricuspid valve regurgitation, widening of the pulmonary artery and its branches, pulmonary hypertension (mild to moderate, pulmonary systolic blood pressure: 47 mmHg, diastolic blood pressure: 38 mmHg), left ventricular EF56% (Figure 1 and 2). Chest X-ray showed pulmonary hypertension (Figure 3). Laboratory examination showed liver function: serum albumin 38.6 g/l, serum total bilirubin 21.9 umol/l, serum direct bilirubin 6.9 umol/l total bile acid 157.5 umol/L. The rheumatic antibody test, infectious disease screening, thyroid function was not abnormal. Right cardiac catheter showed pulmonary artery pressure 70/25/47 mmHg, pulmonary resistance 6.6 wood, pulmonary circulation resistance index 711 Dyn/s/cm-5/m^2, Pulmonary Arteriolar Wedge Pressure (PCWP) 10 mmHg, cardiac output 6.9 l/min, cardiac index 4.01 l/min/m^2, blood pressure 110/65/80 mmHg. After pulmonary vasodilatation test (vantawil) showed pulmonary artery pressure 62/28/39 mmHg, pulmonary resistance 5.6 wood, pulmonary circulation resistance index 567 Dyn/s/cm-5/m^2, cardiac output 6.01 L/min, cardiac index 3.51 L/min/m^2, blood pressure

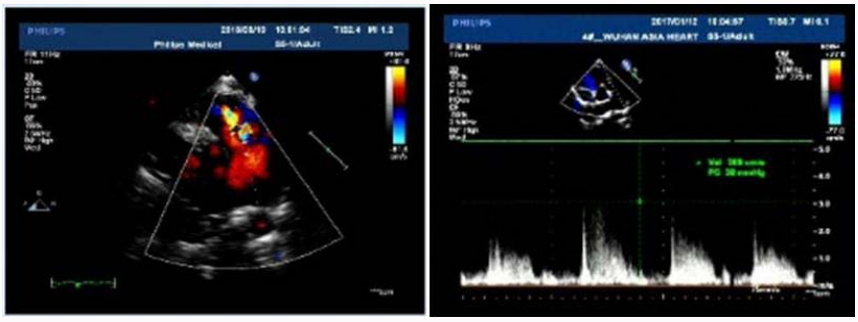

Figure 1: Slight regurgitation signal was observed at the mouth of the diastolic pulmonary valve, with a velocity of 3.1 m/s and a pressure difference of 38 mmHg.

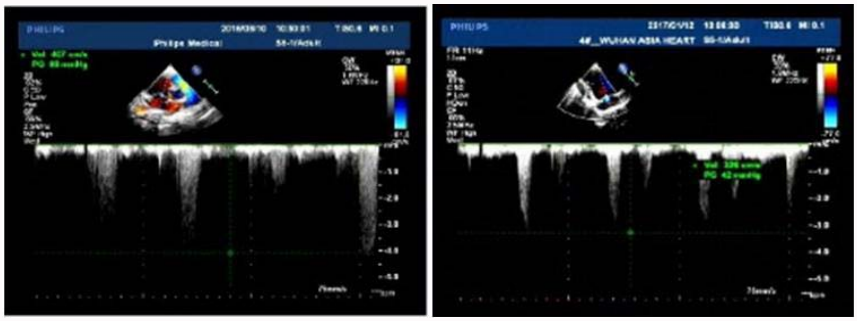

Figure 2: A slight regurgitation signal was observed at tricuspid systolic stage, with a velocity of 3.3 m/s and a pressure difference of 42 mmHg, and a pulmonary artery systolic pressure difference of 47 mmHg was estimated.

OPEN ACCESS

Correspondence:
Jiang-you Wang, Department of Cardiology, Wuhan Asia Heart Hospital and Wuhan Asia General Hospital, Wuhan, 430022, China, Tel: 8615172496706;
E-mail: drwangjiangyou@163.com

Received Date: *06 Feb 2021*
Accepted Date: *08 Mar 2021*
Published Date: *12 Mar 2021*

Citation:
Jiang-you Wang, Chen H. Unexplained Syncope: A Case of Pulmonary Arterial Hypertension Associated with Congenital Extrahepatic Portocaval Shunt. World J Vasc Surg. 2021; 4(1): 1028.

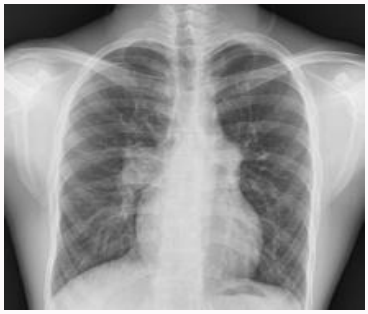

Figure 3: The pulmonary segment is prominent. The right lower pulmonary artery has a dry nodular dilatation with an enlarged right heart.

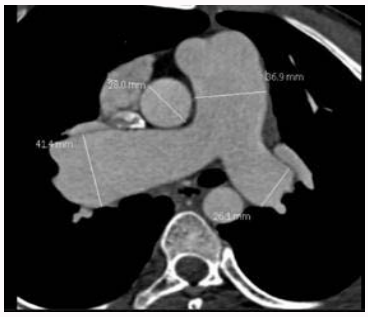

Figure 4: The pulmonary arteries and their branches widened markedly.

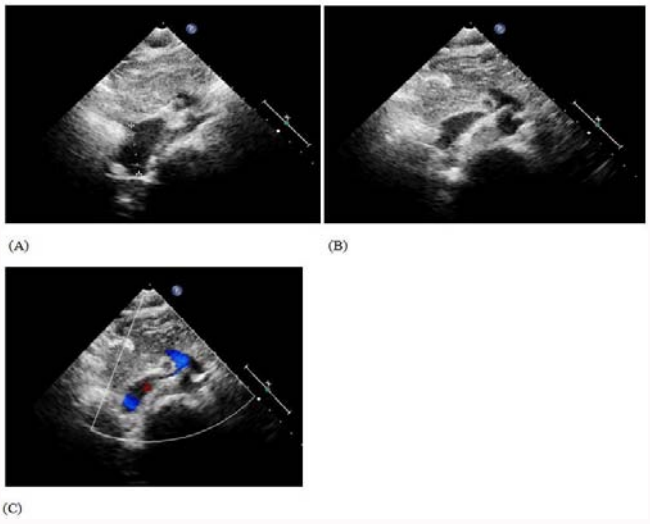

Figure 5: A: The inferior vena cava widened markedly; B: Part of the portal vein drains into the inferior vena cava and part of it runs to the liver; C: Color Doppler imaging showed that part of venous blood flowed into the inferior vena cava, part of it went to the liver, and the blood flow into the liver was retrograde.

120/67/85 mmHg. Pulmonary artery CTA showed the diameter of the main pulmonary trunk was about 36.9 mm, and the diameter of the left and right pulmonary arteries were 23.8 mm and 31.4 mm, respectively (Figure 4). Abdominal ultrasound showed congenital extrahepatic portocaval shunt (Figures 5A-5C). Abdominal enhanced CT showed congenital extrahepatic portocaval shunt (Abernethy type II shunt) (Figures 6A-6D).

Pulmonary CT of the patient further excluded pulmonary embolism, but abdominal ultrasound and enhanced CT scan of the abdomen suggested congenital extrahepatic portocaval shunt, and

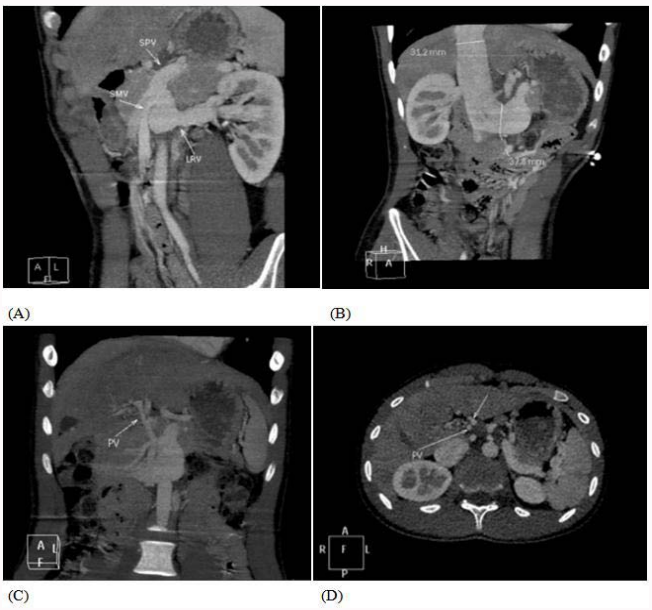

Figure 6: A: The superior enteric vein (SMV) and Splenic Vein (SPV) converge to form the Portal Vein (PV), with most of the portal vein entering the inferior vena cava; B: The inlet diameter is about 37.8. The inferior vena cava widened markedly; C and D: The portal vein gives off another slender branch to the liver. Portal vein is obviously slender.

the correlation between congenital extrahepatic portocaval shunt and pulmonary hypertension should be further considered. Patients who planned to undergo portal shunt occlusion were refused to receive small-dose diuretic, anticoagulant and targeted drug therapy for pulmonary hypertension. The patient's physical strength is better than before, and there is no syncope.

Congenital Extrahepatic Portocaval Shunt (CEPS) is a congenital anomaly observed predominantly in females in which the splanchnic blood bypasses the liver and drains directly into the Inferior Vena Cava (IVC). After the first description of this entity by Abernethy in 1793, fewer than 30 cases of CEPS have been reported [1]. Pulmonary hypertension related to CEPS is an extremely rare condition: to date, only a few cases have been reported in the medical literature [2-3]. CEPS, also known as Abernethy malformations, are classified into 2 types based on the presence or absence of portal blood flow within the hepatic parenchyma. The first type is congenital atresia of the portal vein (end-to-side anastomosis, an Abernethy type 1 shunt), in which the superior mesenteric and splenic veins can join the IVC separately (type 1a) or as a confluence (type 1b). The second type other is hypoplasia of the portal vein with a resultant partial portocaval shunt (side-to-side anastomosis, an Abernethy type 2 shunt). Each type of shunt is associated with unique clinical manifestations: A type 1 shunt, as was present in this patient as a type 1b shunt, is more common in females and is often complicated by congenital cardiac defects, hepatic masses, gastrointestinal and vascular anomalies [4]. However, a type 2 shunt is more common in males and is rarely associated with other malformations.

Pulmonary hypertension is well recognized as a complication of portal hypertension in chronic liver diseases [5-6], but only a few cases of pulmonary hypertension have been reported that are the result of CEPS (type 1b) [2-3]. CEPS, namely portal vein malformation and extrahepatic portal vena cava shunt, the mechanism of pulmonary hypertension has not been fully elucidated. According to current

literature reports, the reason may be that the blood in the portal vein system shunt through the hepatic bypass, and the vasoactive substances enter the pulmonary circulation without liver inactivation, resulting in the imbalance between pulmonary endovascular substances and vasoconstrictor substances, which leads to pulmonary angiectasis or pulmonary vascular lesions, and eventually leads to pulmonary hypertension [7-9]. Clinicians encountered unknown causes of pulmonary hypertension in young patients and should note that congenital portocaval shunt may cause pulmonary hypertension.

References

1. Howard ER, Davenport M. Congenital extrahepatic portocaval shunts: The Abernethy malformation. J Pediatr Surg. 1997;32(3):494-7.
2. Ohno T, Muneuchi J, Ihara K, Yuge T, Kanaya Y, Yamaki S, et al. Pulmonary hypertension in patients with congenital portosystemic venous shunt: A previously unrecognized association. Pediatrics. 2008;121(4):e892-9.
3. Imamura H, Momose T, Kitabayashi H, Takahashi W, Yazaki Y, Takenaka H, et al. Pulmonary hypertension as a result of asymptomatic portosystemic shunt. Jpn Circ J. 2000;64(6):471-3.
4. Morgan G, Superina R. Congenital absence of the portal vein: Two cases and a proposed classification system for portasystemic vascular anomalies. J Pediatr Surg. 1994;29(9):1239-41.
5. Mandell MS, Groves BM. Pulmonary hypertension in chronic liver disease. Clin Chest Med. 1996;17(1):17-33.
6. Umeda N, Kamath PS. Hepatopulmonary syndrome and portopulmonary hypertension. Hepatol Res. 2009;39(10):1020-2.
7. Liehr H, Grün M, Thiel H, Brunswig D, Rasenack U. Endotoxin-induced liver necrosis and intravascular coagulation in rats enhanced by portacaval collateral circulation. Gut. 1975;16(6):429-36.
8. Parratt JR, Sturgess RM. Evidence that prostaglandin release mediates pulmonary vasoconstriction induced by *E. coli* endotoxin. J Physiol. 1975;246(2):79P-80P.
9. Starzl TE, Watanabe K, Porter KA, Putnam CW. Effects of insulin, glucagon, and insuling/glucagon infusions on liver morphology and cell division after complete portacaval shunt in dogs. Lancet. 1976;1(7964):821-5.