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Patent Ductus Venosus in an early term infant with mild respiratory distress – Case report

Maria-Andreea Racean[1], Laura Mihaela Suciu[1,2], Cristina Blesneac[3,4], Marian Pop[4,5], Cristina Oana Marginean[3,6]

[1]Department of Neonatology, Mures Clinical County Hospital, Targu-Mures, Romania
[2]Department of Neonatology, "George Emil Palade" University of Medicine, Pharmacy, Sciencies and Technology, Targu-Mures, Romania
[3]Department of Pediatrics, Emergency Institute for Cardiovascular Diseases and Heart Transplant, Targu-Mures, Romania
[4]"George Emil Palade" University of Medicine, Pharmacy, Sciences and Technology, Targu-Mures, Romania
[5]Department of Radiology, Emergency Institute for Cardiovascular Diseases and Heart Transplant, Targu-Mures, Romania
[6]Department of Pediatrics, "George Emil Palade" University of Medicine, Pharmacy, Sciences and Technology, Targu-Mures, Romania

ABSTRACT

Patent ductus venosus is an extremely rare form of congenital portosystemic shunt that results in the diversion of portal blood into the systemic circulation, decreasing hepatic blood flow and increasing blood volume and toxic substances in the systemic circulation. We present a clinical case of a male infant born at 37 5/7 weeks gestational age with patent ductus venosus. Because very few cases of patent ductus venosus have been reported, the prudent approach it would be to perform serial Doppler studies and laboratory tests to follow the course of the ductus venosus until closure and to intervene if the infant becomes unstable and symptomatic. The medical team decided that the conservative management in this case was a better choice, since the infant remained stable with good growth.

Keywords: patent ductus venosus, early term infant, respiratory distress

INTRODUCTION

Ductus venosus (DV) is the continuation of the umbilical vein, a connection between the portal vein and inferior vena cava that closes functionally in the first minutes after birth and is completely obliterated in 15 to 20 days, at which point ligamentum venosum is formed. However, because of a poorly developed intrahepatic portal system, DV may not close after birth, leading to patent DV (PDV). The DV is normally open for a much longer time than the ductus arteriosus (DA) [1,2,3]. If the DV fail to close, clinical signs may include galactosemia, hypoxemia, encephalopathy with hyperammonemia and hepatic dysfunction with elevated liver enzymes, disturbances of coagulation function and hyperbilirubinemia [4,5].

PDV is a very rare type of congenital portosystemic shunt that results in the diversion of portal blood into the systemic circulation, decreasing hepatic blood flow, increasing blood volume, and toxic substances in the systemic circulation. PDV is associated with the potential for life-threatening complications, with early detection and proper management leading to a good prognosis and the prevention of serious complications [6,7]. PDV is now accurately diagnosed by the use of imaging studies, including abdominal ultrasound, computed tomography (CT) and magnetic resonance imaging [8]. However, the DV has a variable sonographic appearance depending on its stage of involution [9].

The closure of a PDV is now feasible in a minimally invasive way due to the advances in interven-

Corresponding author:
Cristina Oana Marginean
E-mail: marginean.oana@gmail.com

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tional radiological techniques [10]. Treatment options for cases detected with PDV and persistent clinical manifestations include surgical ligation and banding, liver transplantation and transcatheter detachable coil closure [11].

The aim of this case report was to highlight that a rare cause of cholestasis can be represented by the PDV and that is why we need to investigate the presence of a PDV when we confront with a cholestasis without an evident cause.

Written informed consent was obtained from the patient's mother, the minor's legal guardian, for the publication of this case presentation.

CASE PRESENTATION

We present a male infant born at 37 5/7 weeks gestational age to a 23-year-old gesta IV para III healthy mother. This spontaneously conceived pregnancy was unremarkable except the early term labor. The birth took place at a peripheral hospital and the newborn was transferred to our institution at the age 5 hours for a mild respiratory distress syndrome. The amniotic membranes ruptured 10 minutes before birth and the amniotic fluid was clear. The infant's birth weight was 2650 grams (14nd centile), length 50 cm (64nd centile) and cranial perimeter 33 cm (28nd centile). His Apgar scores were 7, 8 and 8 at 1, 5 and 10 minutes respectively. His arterial cord blood measurement was pH 7.17, pCO2: 52 mmHg, lactate: 5.35 mmol/L, BE: -9.2 mmol/L. The maternal ABO blood type was BIII, Rh positive.

CLINICAL COURSE IN THE NEONATAL PERIOD

On clinical examination, the patient was alert, appeared warm and comfortable. The temperature was 36.9°C, the heart rate 148 beats per minute, the blood pressure 79/45 mm Hg, the respiratory rate 69 breaths per minute, with visible subcostal indrawing and nasal flaring, and the oxygen saturation 91% while he was breathing ambient air. The anterior and posterior fontanelles were flat and soft. The lungs were clear on auscultation. The abdomen was soft without distention, tenderness, or palpable masses. No sign of portal hypertension or ascites were evident, no hepatomegaly was detected, and the liver had a smooth surface and sharp edge. The male genitalia appeared normal. There were no dysmorphic features and neurologic examination revealed normal primitives reflexes.

The blood glucose level was 69 mg per deciliter while the patient was receiving dextrose at a glucose infusion rate of 2.5 mg per kilogram of body weight per minute. The initial total serum bilirubin level was 7 mg per deciliter (110 μmol per liter) and the initial direct bilirubin level was 0.58 mg per deciliter-both levels increased during hospitalization. The level of liver enzymes was normal from admission to discharge. The initial level of lactate dehydrogenase was 825 units/liter and the creatin kinase was 789 units/liter.

A sepsis evaluation was performed, ampicillin and gentamicin were started and a viral work up for dilated cardiomyopathy was sent (cytomegalovirus, adenovirus) and were negative. The infant's cultures were negative at 48 hours; therefore, antibiotics were discontinued, his respiratory distress gradually resolved in the next 10 hours and the initial continuous positive airway pressure was suppressed.

RADIOLOGY FINDINGS

A chest x-ray revealed that the lungs were clear, with prominent vascularity in the upper zone. The heart size was abnormal and had a globular (rounded) appearance; the cardiothoracic ratio was greater than 50 percent; the right heart border was doubled; and the costophrenic angles were well-defined, indicating that there is no pleural effusion. These are illustrated by Figure 1.

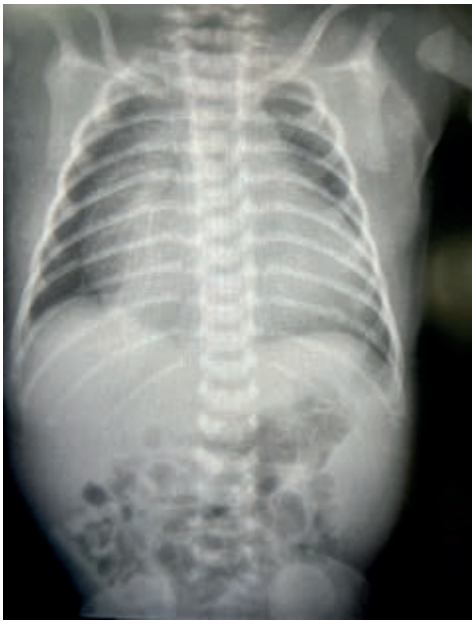

FIGURE 1. Chest radiograph shows diaphragms domed, 9 rib expansion, lungs normal, right liver, left stomach, mosaic bowel gas, cardiomegaly

ULTRASOUND FINDINGS

Cranial ultrasound was notable for mild ventriculomegaly. This is revealed in Figure 2. *Abdominal ultrasound* was completed at 24 hours of age and revealed a communication channel between inferior vena cava and the left portal branch, and the Color Doppler examination confirmed the vascularity of

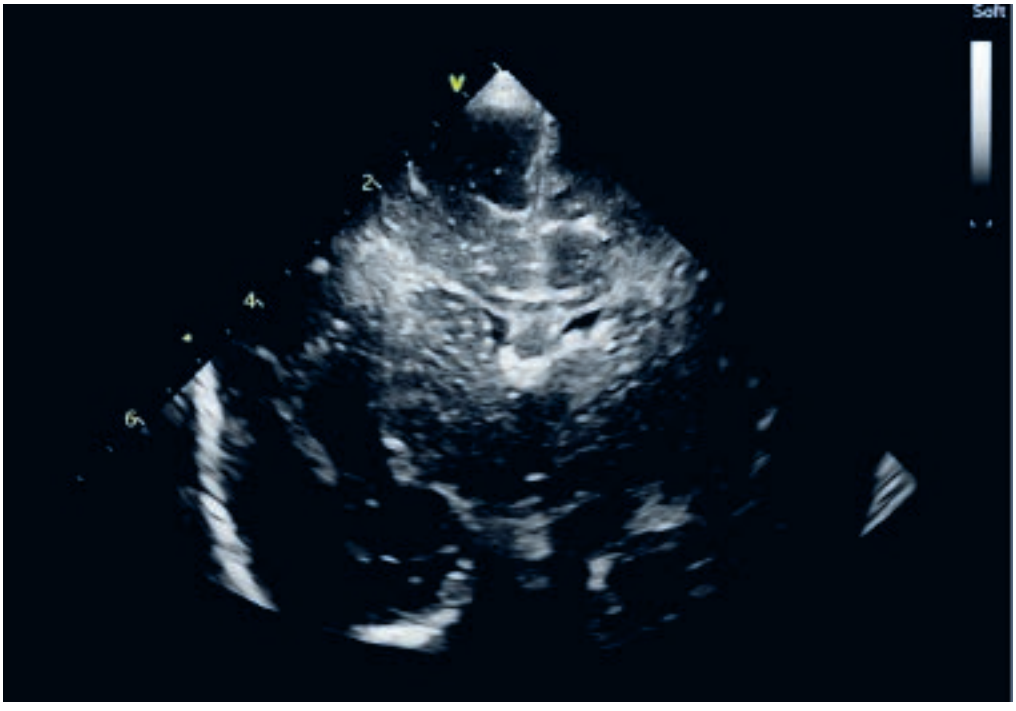

FIGURE 2. Coronal plane, cranial ultrasonography, at the anterior horns of the lateral ventricle and Sylvian fissures

the lesion. In the longitudinal axis the vascular S -shape tubular structure had a maximum diameter of 4.2 mm. Pulsed wave Doppler has shown a similar pattern to that demonstrated in the inferior vena cava. This is illustrated by Figure 3. The portal vein was visualized in the longitudinal axis from the splenomesenteric junction of the liver hilum and the diameter was 3.6 mm. Craniocaudal dimension of the liver on the midclavicular line was 6.2 centimeter. The diameter of the common bile duct was 1.2 mm. Therefore, the cardiology team was consulted for an echocardiographic examination.

Initial *echocardiogram* performed at 36 hours age was notable for elevated right ventricular pressure by tricuspid jet (TR Vmax 3.03 m/sec, TR max PG 36.77 mmHg), high velocity left to right patent DA (PDA), bidirectional shunt across the foramen ovale, right atrium (RA) dilatation, right ventricle (RV) dilatation, RV hypertrophy, mild RV disfunction (TAPSE =8 mm) and 3 Ch-RV fractional area change (FAC=33%), RV diastolic disfunction (Tricuspid E/A= 0.49), left atrial (LA) compression, normal LV systolic function (ejection fraction (EF) by Simpson 55%) with marker of LV diastolic disfunction (Mitral E/A=0.8), IVRT=50 msec.

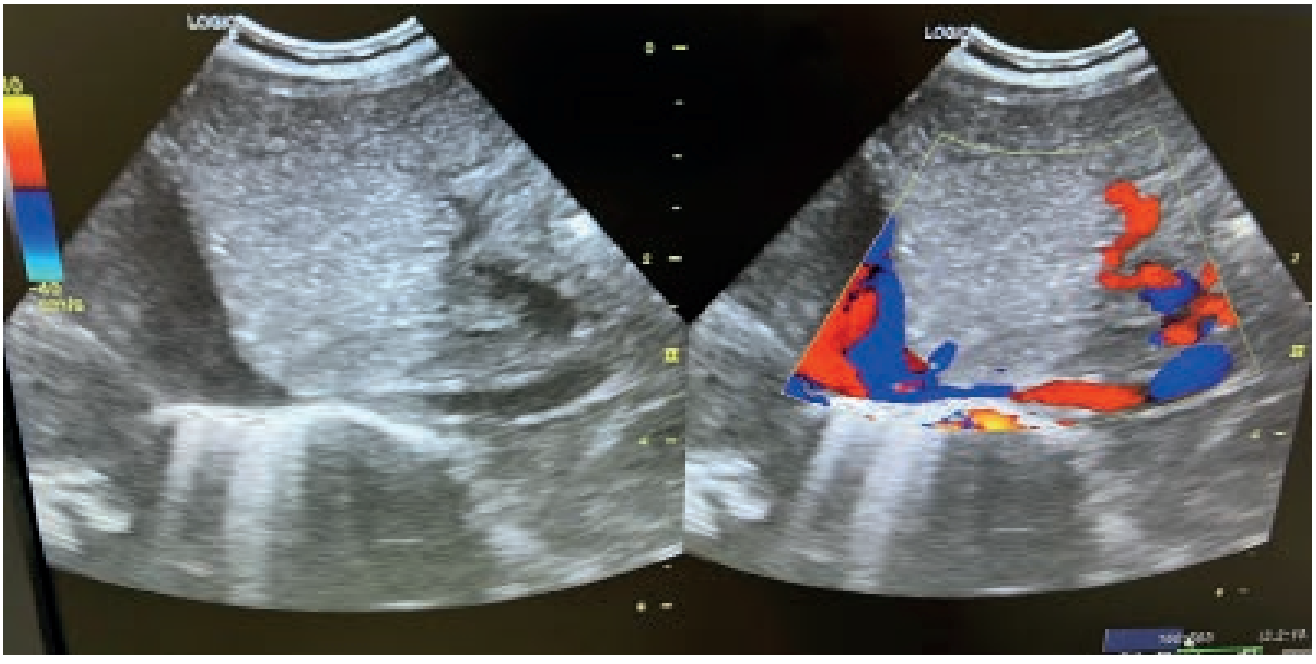

FIGURE 3. A subcostal parasagittal view of the liver and inferior vena cava

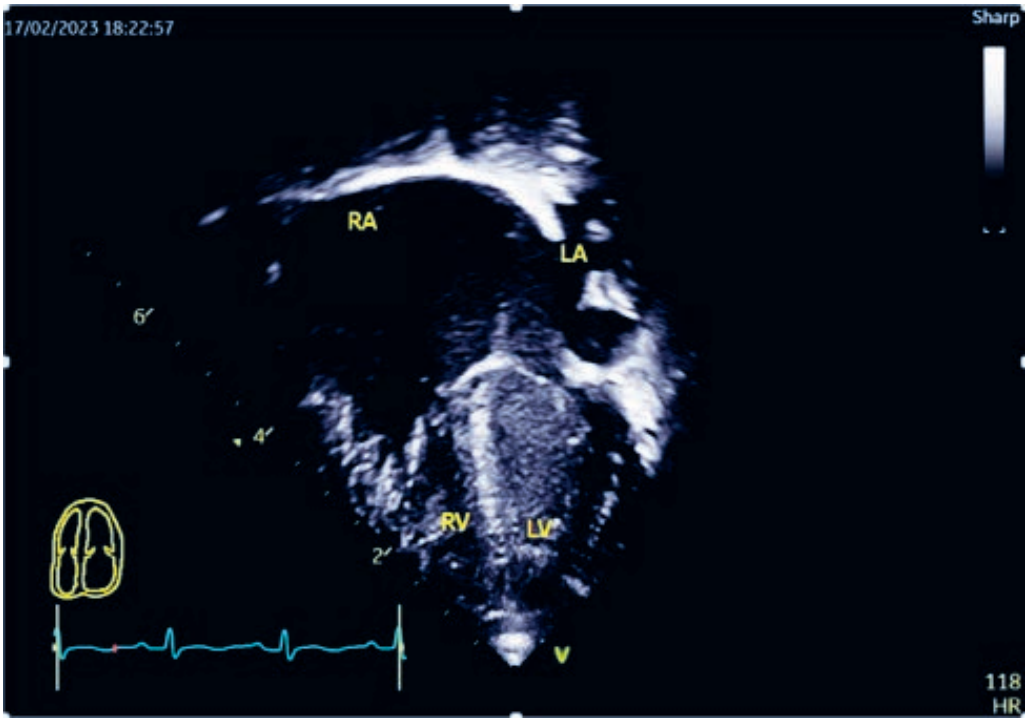

FIGURE 4. Apical 4 chamber view, showing the four chamber and right atrium enlargement

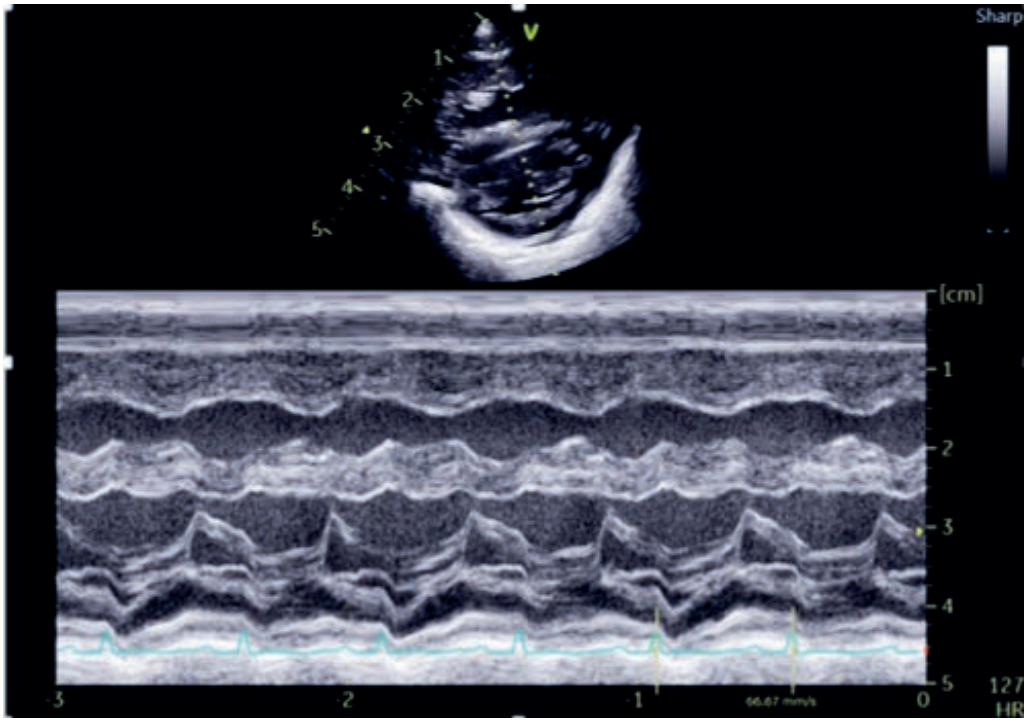

FIGURE 5. Evaluation of shortening fraction using M mode method from short axis view of the left ventricle

Interventricular sept was flat in systole and diastole. A small interventricular septal apical defect was detected with left to right shunt. Echocardiographic images are shown by Figures 4, 5 and 6.

CT angiography (CTA) performed in the thirteenth day of life (DOL) with 3ml of contrast medium (Iomeprol 400) revealed that the chest, cardiac cavities, and large intrathoracic vessels appeared normal. A more pronounced RA was observed, possibly as a result of the saline flush. The liver has a smooth contour and a homogeneous structure without any dilatation of the intrahepatic bile ducts. The presence of a structure with opacification is confirmed in the venous phase: the Arantius ductus venosus, showing a caliber of 6.8 mm, is located between the upper slope of the portal vein immediately before its bifurcation and the inferior retrohepatic vena cava, at approximately 3 cm inferior to the diaphragmatic domes. No other lesions are detected at the level of the abdominal parenchymal organs.

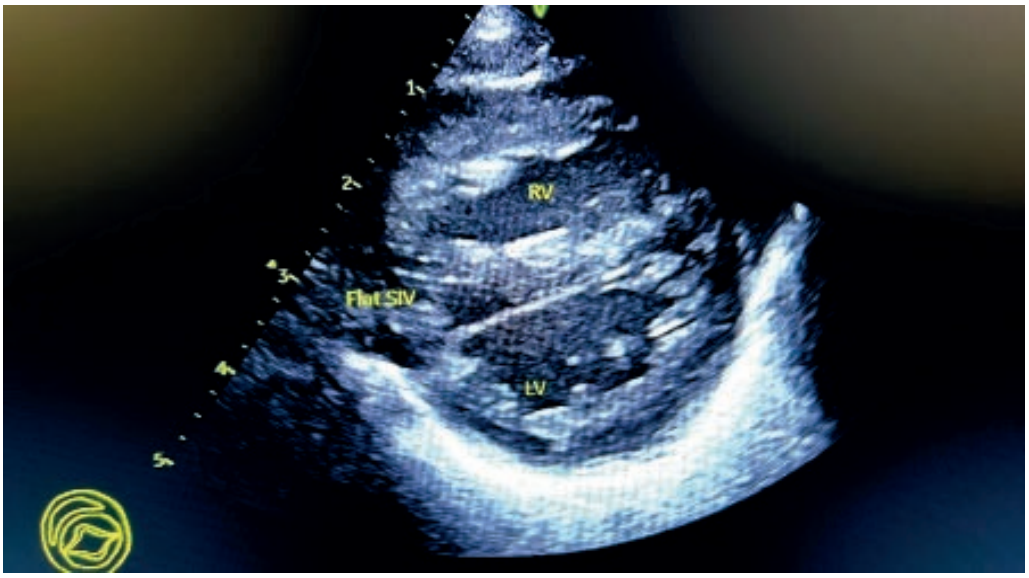

FIGURE 6. Demonstrating septal morphology depending on degree of pulmonary hypertension on the short axis view

Without the collection of pleuro-pericardic or abdominal-pelvic free fluid. Distended intestinal loops with air content. Conclusions: PDV. Cardiothoracic, and abdominal appearance is within normal limits.

DISCUSSIONS

PDV is an uncommon vascular malformation, classified as a type of intrahepatic shunt [4]. Like in our case, the male infant aged 69 days presented by Kamali et al, was afebrile, with no tachycardia or tachypnea present, but with a cholestatic jaundice [2]. In the second DOL of the infant, our patient was referred to a pediatric cardiologist due to the abnormal radiologic aspect of the heart and the cardiothoracic ratio greater than 50 percent. Similar to our case, Sharma et al stated that the infant's DV was primarily identified with ultrasound with color Doppler and it was confirmed by the CTA [10].

The main complication of the portosystemic shunts in the neonatal period is cholestasis [12]. In our infant's case, there was no increase in the hepatic enzymes levels and we believe that the mechanism of cholestasis was due to the decreased intrahepatic flow. However, the partial diversion of the portal blood to the systemic circulation associated with increased ammonia and blood coagulation disturbances in the neonatal period was not the case in our infant. In contrast to our case, Fugelseth et al presented the association between persistent pulmonary hypertension and PDV [3]. Contrary to our case, Poeppelman et al described the case of a male infant with PDV and a complex congenital heart disease, that were associated with hepatic dysfunction, coagulation disturbance and hyperammonemia. After surgical intervention these conditions resolved in that case. [4].

The management of portosystemic shunts in the pediatric population is controversial. In cases of PDV, surgical treatment is recommended if the infant's growth is stunted or if the levels of the hepatic enzyme are rising [11]. Franchi-Abella et al recommend avoiding early closure in the neonatal period due to difficulties related to the complex nature of the lesions and possible spontaneous resolution within 2-5 months after birth. The 5 cases of neonates described by Franchi-Abella et al with intrahepatic shunts that presented with cholestasis, all of them resolved spontaneously [12].

The medical team decided that conservative management in our case was a better choice since the infant remained stable and showed good growth. The infant was discharged from the maternity unit at the seventeenth day of life because he was stable, with an upward growth curve, no signs of encephalopathy, a normal level of liver enzymes and bilirubin and no evidence of portal hypertension. The case will be followed by the pediatric cardiologist at regular intervals until the closure of the PDV occurs.

CONCLUSIONS

In PDV cases, prompt evaluation is required and includes primarily ultrasonography and laboratory tests. Then PDV should be monitored by assessing in infant's growth, performing repeated serology tests and being aware for any signs of portal hypertension such as liver enlargement. Because very few cases of PDV have been reported, the prudent approach it would be to perform serial Doppler studies and laboratory tests to follow the course of the DV until closure and to intervene if the infant becomes unstable and symptomatic.

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