Heart & Lung 43 (2014) 367–370

Contents lists available at ScienceDirect

Heart & Lung

journal homepage: www.heartandlung.org

Primary pulmonary vein stenosis in a premature infant without bronchopulmonary dysplasia: A case report

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ARTICLE INFO

Article history:
Received 27 January 2014
Received in revised form
13 March 2014
Accepted 18 April 2014

Keywords:
Pulmonary vein
Stenosis
Premature

ABSTRACT

Primary pulmonary vein stenosis (PVS) presenting in childhood is uncommon and is related to premature with bronchopulmonary dysplasia (BPD). Here we present a premature infant with primary PVS and without BPD. In our case, a 19-month-old girl was diagnosed with PVS, atrial septal defect and patent arterial duct by echocardiography and selective pulmonary artery angiography. Interestingly in the first 2 month after birth, there was no clue of PVS by bed echocardiography in the patient. It is important to focus on the pulmonary vein blood velocity in premature infants and very low birth weight infants even without BPD.

Introduction

Pulmonary vein stenosis (PVS) is a rare condition characterized by the narrowing of one or more pulmonary veins at their left atrial opening. Primary PVS is most frequently associated with cardiac malformations such as anomalous pulmonary venous return.[1] It can lead to worsening pulmonary hypertension and cardiac failure in the pediatric population. Neither surgery nor transcatheter interventions have yielded satisfactory long term results.[2] Recent years have seen significantly improved perinatal survival for extremely premature infants, resulting in a number of infants and children suffering with its common pulmonary sequela, bronchopulmonary dysplasia (BPD).[3] Recent case and epidemiologic studies have identified a hitherto-unappreciated association between the PVS and BPD.[4–6] Here we report a premature infant with primary PVS and without BPD at our institution.

Case report

A 19-month-old girl was admitted for heart surgery with a secundum atrial septal defect (ASD) and patent arterial duct (PDA). The patient was the mother's third child from her second pregnancy, the younger of twins and born at 36 weeks gestation. She was delivered by cesarean section and her birth weight was only 1.09 kg. Her Apgar scores were 7 and 8 at 1 and 9 min, and she was given oxygen, was intubated and received parenteral nutrition in the neonatal intensive care unit. She had mild perinatal indirect hyperbilirubinemia managed by phototherapy and, as demonstrated on bed echocardiography, an atrial septal defect, a ventricular septal defect, a patent arterial duct and pulmonary hypertension. Further more, the color Doppler and pulse wave Doppler demonstrated normal pulmonary vein velocity (Fig. 1). Then she was received diuretic hydragogue to decrease the cardiac load. She developed neonatal sepsis 3 weeks later and received injected antibiotics and gamma globulin. After 2 months, her body weight was 1.90 kg and she was discharged from hospital. The patient was not received surfactant and was not required positive pressure ventilation in NICU.

Her vital signs were pulse 118/min, respiratory rate 26/min, weight 6 kg and oxygen saturation 98% in room air. Physical examination revealed a soft upper left parasternal systolic murmur with a prominent pulmonary second heart sound. Electrocardiography showed sinus rhythm. Chest radiography showed a slightly enlarged heart shadow and lung field congestion. There was no evidence of abnormal branch tract or lung development on computed tomography of the chest. Transthoracic echocardiography showed atrial septal defect (secundum type, Ø 10 mm), patent arterial duct (Ø 1.0 mm), abnormal increased right upper pulmonary vein blood velocity (193 cm/s), and pulmonary hypertension as estimated by the tricuspid reflex (ΔP 56 mm Hg) (Fig. 2). Heart catheterization demonstrated a pulmonary capillary wedge pressure of 19/10 mm Hg (mean 11), main pulmonary artery pressure 70/38 mm Hg (mean 54), right ventricular pressure 87/0 mm Hg (mean 3), and left ventricular pressure 126/1 mm Hg (mean 16). Selective pulmonary vein angiography showed stenosis of four pulmonary veins at the venoatrial junction (Fig. 3).

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http://dx.doi.org/10.1016/j.hrtlng.2014.04.018

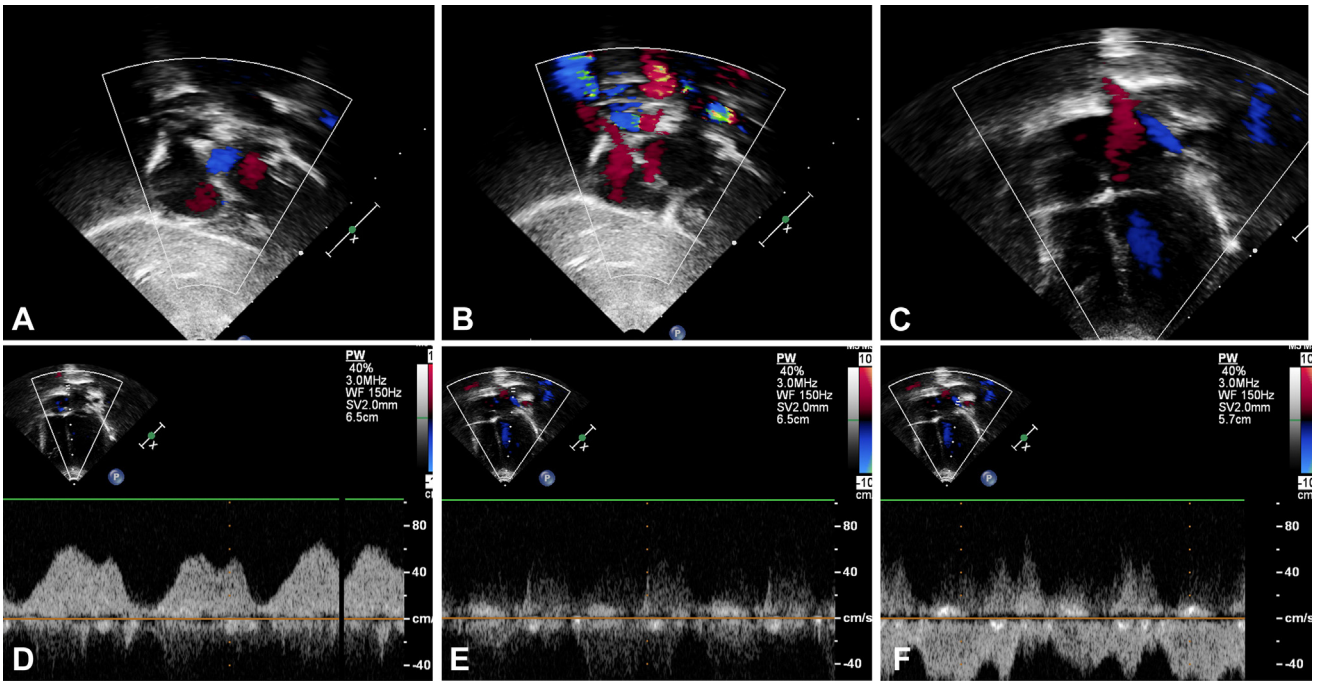

Fig. 1. (A) The two atrium view showed atrial septal defect (right to left shunt) with color Doppler. (B) The two atrium view showed atrial septal defect (left to right shunt) with color Doppler. (C) The four chamber view showed normal blood reflux in right upper pulmonary vein with color Doppler. (D) Right upper pulmonary vein blood flow velocity demonstrated by continuous wave Doppler. (E) Left upper pulmonary vein blood flow velocity demonstrated by continuous wave Doppler. (F) Left down pulmonary vein blood flow velocity demonstrated by continuous wave Doppler.

Discussion

Primary PVS is a rare anomaly in association with anomalous pulmonary venous return. However, there is recent evidence of increased risk of primary PVS in premature infants with BPD. A recent clinicopathologic population study by Drossner and colleagues identified prematurity as being highly associated with PVS (odds ratio: 10.2; 95% confidence interval: 4.7–22.6; $P < 0.001$); of 26 cases of PVS, 11 (42%) had comorbid BPD.[4] The pathogenetic hierarchy of PVS is not clear. Drossner et al have speculated that many of the causal mechanisms of BPD, especially its small vascular component, might contribute to the endovascular proliferative stenosis observed in PVS, while PVS itself may contribute to the syndrome of BPD through its induction of vascular congestion, pulmonary edema, and increased right-sided heart pressures.[4] Sadr et al[7] postulated that abnormal cytokine production, flow abnormalities and post-surgical stress may be triggers of the process. The autopsy of an premature infant with PVS and BPD showed fibrous ridge in pulmonary vein obstructing atrial ostia and microscopically the lungs showed patchy edema, severe capillary congestion,

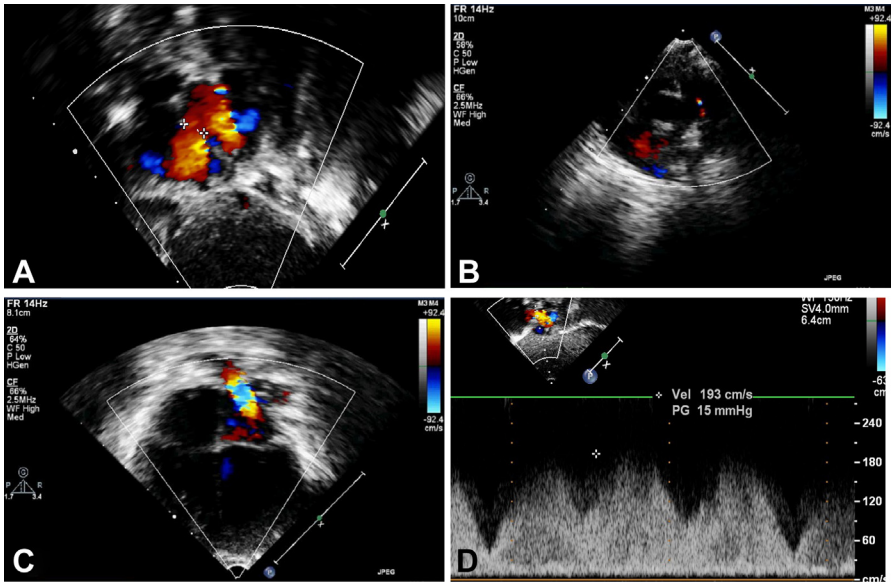

Fig. 2. (A) Shunt from the left atrium to the right atrium, indicating atrial septal defect. (B) A small patent arterial duct shunt on a short axis view. (C) Right upper pulmonary vein flow was brightly colored, indicating a high blood flow velocity. (D) Pulmonary vein blood flow velocity (193 cm/s) demonstrated by continuous wave Doppler.