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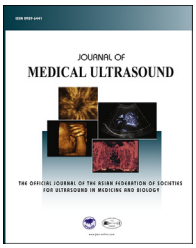

LETTER TO THE EDITOR

Prenatal Diagnosis of Persistent Left Superior Vena Cava is Associated with Coarctation of the Aorta — A Case Report

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Abstract A singleton pregnant woman was found to have persistent left superior vena cava (PLSVC) of the fetus at 22 weeks by ultrasound. Follow-up scans revealed PLSVC, dilated coronary sinus, dominant right heart, some pericardial effusion, and hypertrophy of the right ventricular wall. The woman had an abdominal delivery at 34 weeks due to rupture of membranes. The baby was found to have coarctation of the aorta postnatally and had aortic reconstruction at 31 days of age. A prenatal ultrasound finding of PLSVC might be associated with coarctation of the aorta and it warrants specialist follow-ups and complete workup of echocardiography prenatally and postnatally.

The pregnant woman was referred to our clinic at 22 weeks because of an increased NTD (Neural Tube Defects) risk at 1/113 by the quadruple screen test. The karyotype of the fetus was 46, XX. This was her third pregnancy. Her previous two pregnancies were generally uneventful and both were delivered by Cesarean section. A detailed fetal anatomical ultrasound screening revealed normal fetal anatomy except for a persistent left superior vena cava (PLSVC) and a dilated coronary sinus (Fig. 1a & 1b). A follow-up scan was performed at 26 weeks, and, in addition to the PLSVC and the dilated coronary sinus, the fetus was found to have a dominant right heart, with some pericardial effusion and hypertrophy of the right ventricular wall (Fig. 2a & 2b).

Conflict of interest: The authors have no conflicts of interest relevant to this article.

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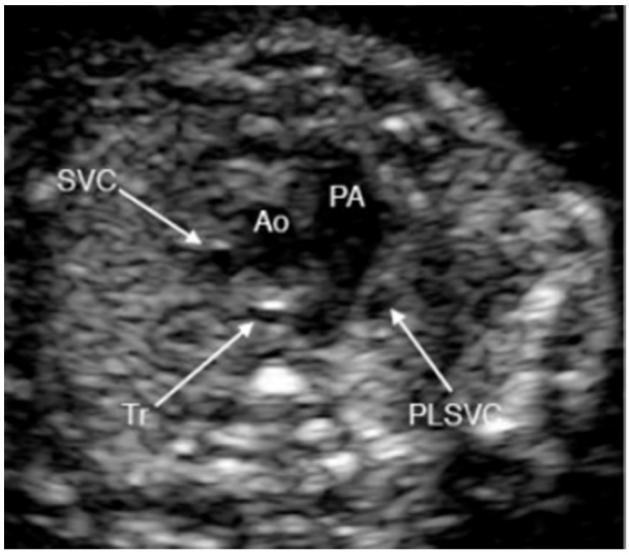

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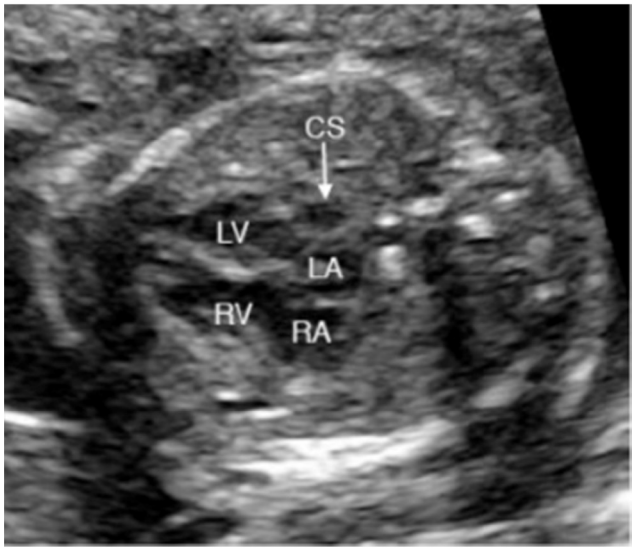

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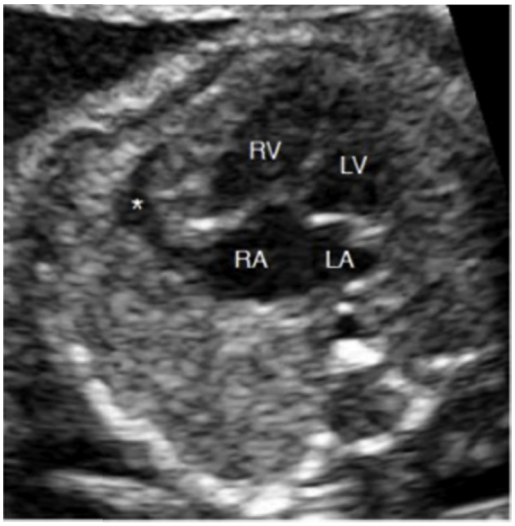
Figure 1 22 weeks: (a) Three-vessel-trachea view. The PLSVC is identified as an additional vessel on the left side of the pulmonary artery. (b) Four-chamber view. The dilated coronary sinus bulges into the left atrium. PA: pulmonary artery, Ao: aorta, SVC: superior vena cava, Tr: trachea, PLSVC: persistent left superior vena cava, LA: left atrium, LV: left ventricle, RA: right atrium, RV: right ventricle, CS: coronary sinus.

An advanced fetal echocardiography was performed by our pediatric cardiologist at 27 weeks, and the findings included PLSVC, a dilated coronary sinus, a patent and unrestricted foramen ovale, pericardial effusion, and a relatively dominant right heart and a small left heart, with pulmonary artery diameter 5.6 mm (Z score: −0.1), aortic



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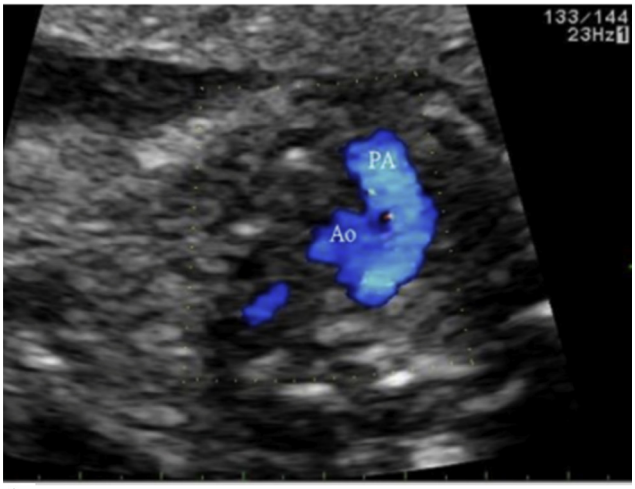

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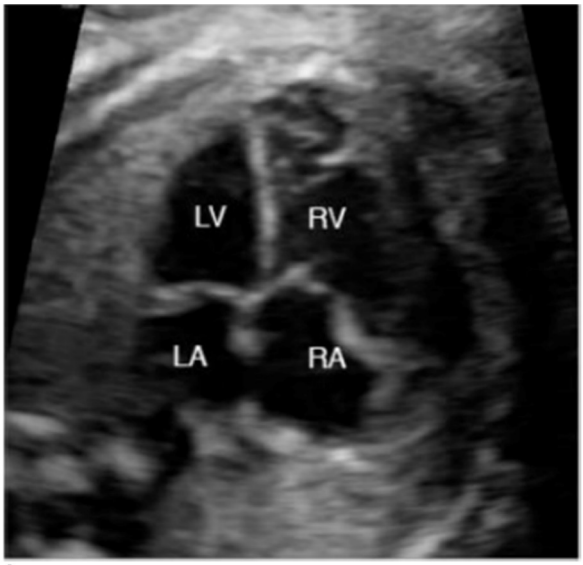
Figure 2 26 weeks: (a) Four-chamber view showing the hypertrophy of the right ventricular wall and mild pericardial effusion (*). (b) Three-vessel-trachea view.



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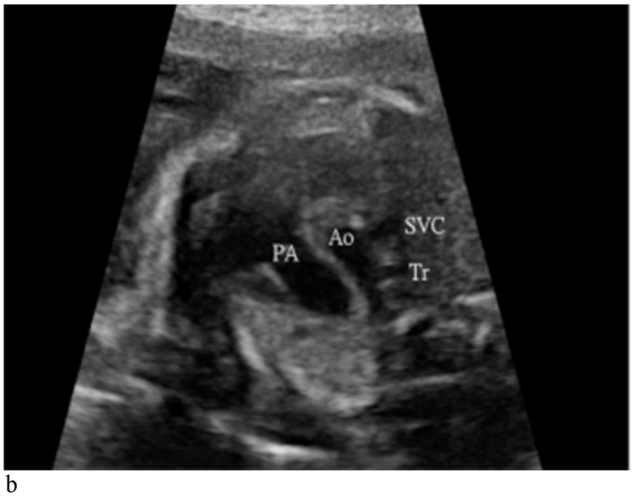

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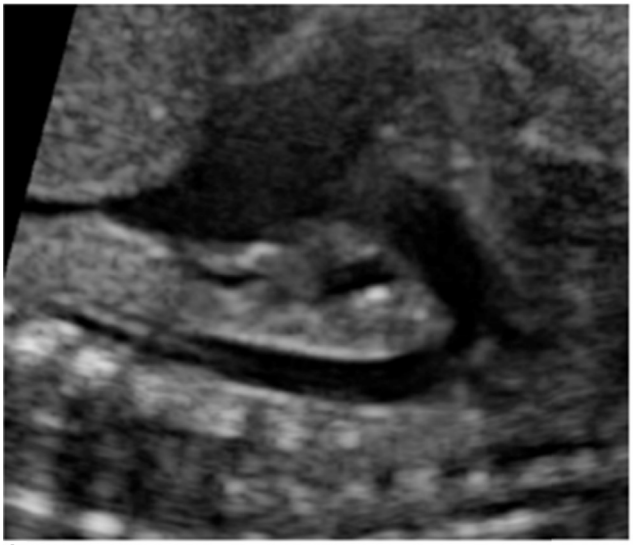

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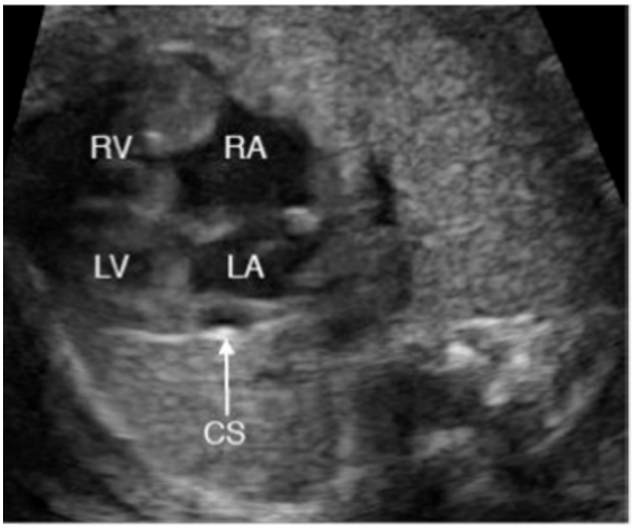
Figure 3 27 weeks: (a) Four-chamber view showing a relatively large right heart as compared to the left heart. (b) Three-vessel-trachea view showing a decreased aortic diameter as compared to the pulmonary artery. (c) Longitudinal view of the aortic arch.



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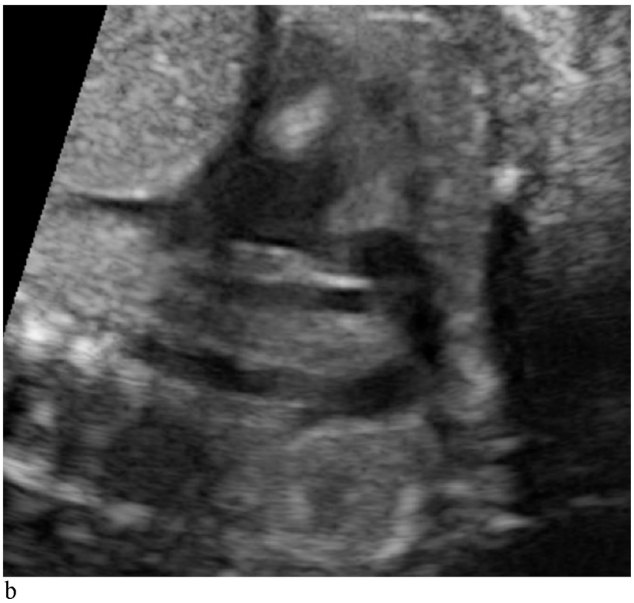

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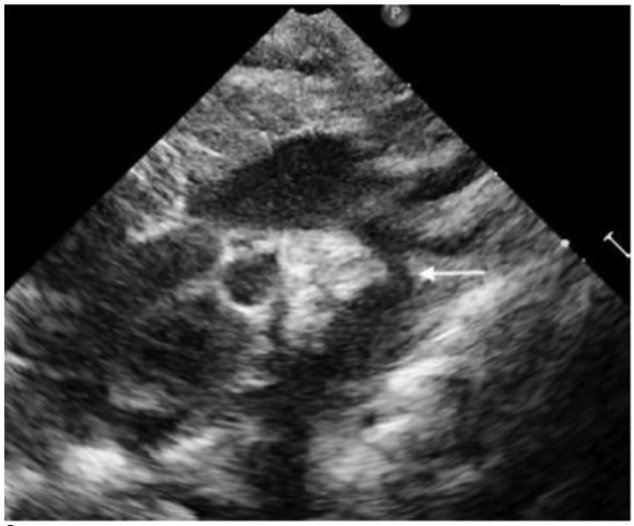
Figure 4 32 weeks: (a) Four-chamber view showing the dilated coronary sinus. (b) Longitudinal view of the aortic arch.

diameter 3.8 mm (Z score: −1.9), mitral valve annulus 5.7 mm (Z score: −2.7), tricuspid valve annulus 6.7 mm (Z score: −1.8), and isthmus width 3.3 mm (Z score: 0.06) (Fig. 3a, 3b & 3c). A follow-up fetal echocardiography was performed at 32 weeks, and the findings were the same, with the right heart becoming relatively more dominant than the left heart, and pulmonary artery diameter 6.5 mm (Z score: −0.59), aortic diameter 5.0 mm (Z score: −1.46), and isthmus width 4.1 mm (Z score: 0.23) (Fig. 4a & 4b). As coarctation of the aorta (CoA) was suspected, the pregnant woman was advised to have echocardiography for the baby soon after delivery (Fig. 5).

She had rupture of membranes at 34 weeks and had an abdominal delivery in a medical center. A postnatal echocardiography on day four revealed a bidirectional PDA, two lower muscular VSD's (1.9 mm, 1.5 mm), PFO 3.8 mm, bicuspid aortic valve with trivial aortic regurgitation, and an increased flow velocity in the descending aorta (PG 15 mmHg), suggesting mild CoA.



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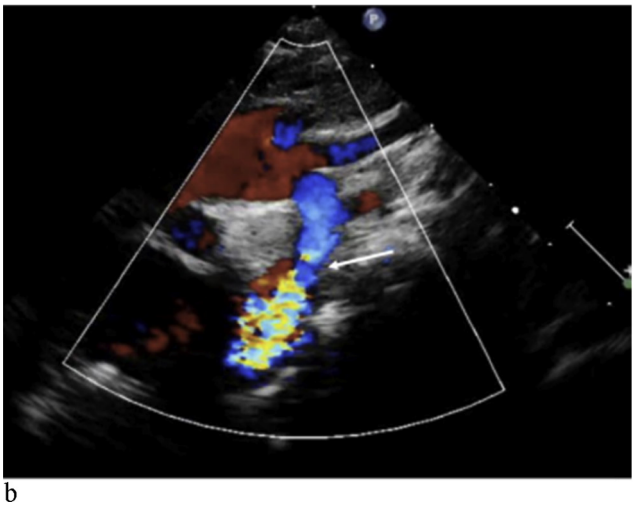

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Figure 5 Neonatal echocardiography on day 4 after birth (a) Suprasternal notch view showing the narrowing aortic isthmus (arrow). (b) Suprasternal notch view with color Doppler showing turbulent flow at the site of the coarctation of the aorta (arrow).

The baby had aortic reconstruction at 31 days of age, and had an uneventful recovery (Fig. 6). At the time of writing, the baby is 11 months of age and is generally healthy.

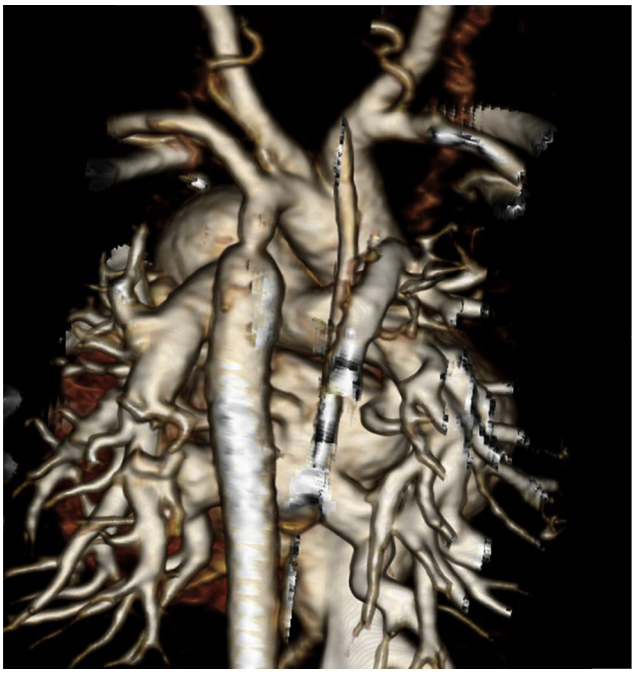

Figure 6 3D CT reconstruction on day 29 after birth.

Prenatal ultrasound finding of PLSVC should prompt a detailed fetal anatomical screening, and further genetic workup should be considered based whether there are associated abnormal findings. Isolated PLSVC is reported to be associated with CoA in 21% of cases [1] and as the prenatal diagnosis of CoA remains a challenge [2], serial follow-up scans during pregnancy and after delivery are suggested.

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