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Case report

Prenatal diagnosis of anomalous drainage of the right superior vena cava to the left atrium

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Abstract

Anomalous drainage of the right superior vena cava to the left atrium, as an isolated abnormality, is a very rare form of congenital heart disease that causes postnatal hypoxemia. There is only one other case reported in the literature of this anomaly diagnosed prenatally. We describe a case of isolated right superior vena cava to the left atrium diagnosed prenatally by our fetal cardiology team after obstetrical referral for a dilated superior vena cava, and we discuss the differential diagnosis of in utero dilation of the superior vena cava.

Key words: right superior vena cava to the left atrium, biatrial drainage of the superior vena cava, systemic venous anomalies, dilated superior vena cava, fetal/prenatal diagnosis.

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Case report

A 29-year-old G7P1504 African-American woman was referred to Vanderbilt University Medical Center's fetal cardiology clinic at 26 1/7 weeks' gestation for a dilated right superior vena cava (RSVC) appreciated on obstetrical ultrasound. Her pregnancy was complicated by a history of stillbirth (20 weeks' gestation) and neonatal demise (24 weeks' gestation), polysubstance abuse, and chronic hypertension.

The initial fetal echocardiogram demonstrated a dilated RSVC (diameter 5.2 mm; inferior vena cava diameter 3.3 mm in comparison) with prominent innominate (3.6 mm) and azygous veins (Figure 1) and with an unusual flow pattern in the rightward aspect of the left atrium (LA) in the four-chamber and long-axis views (Figure 2). Sagittal images suggested anomalous connection of the RSVC to the LA and biatrial drainage of the RSVC, with the RSVC "overriding" the atrial septum via a sinus venous defect and with associated partial anomalous pulmonary venous drainage (Figure 3). No cerebral arteriovenous malformations had been appreciated on obstetrical ultrasound, and middle cerebral artery Doppler patterns (MCA peak systolic velocity: 46.4 cm/sec; multiples of the median [MoM] 1.37) were not consistent with fetal anemia. The fetal heart was otherwise structurally normal with normal atrioventricular and ventriculoarterial connections and with normal connection of the inferior vena cava (IVC) to the right atrium (RA). A follow-up evaluation at 30 1/7 weeks' gestation revealed similar findings.

A male infant was born at 39 1/7 weeks' gestation via emergency Cesarean section for prolonged fetal heart rate deceleration and thick meconium. His APGAR scores were 8 and 9. He was initially admitted to the special care nursery, but oxygen

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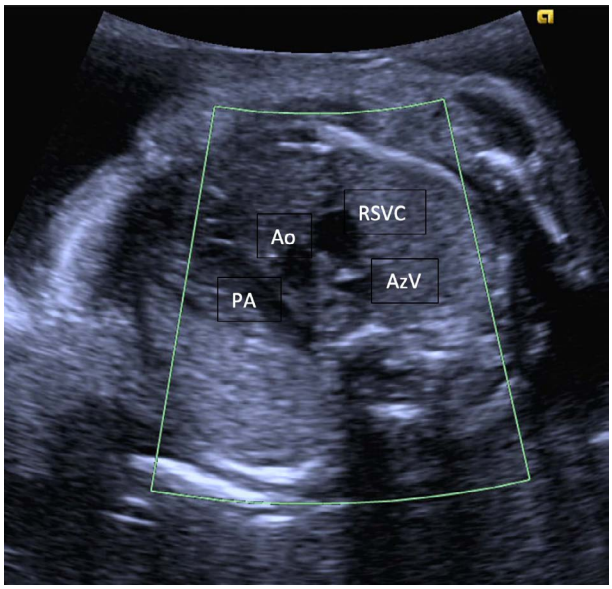

Figure 1. Three-vessel-trachea view demonstrating a dilated right superior vena cava (RSVC) and a dilated azygous vein (AzV)

PA – pulmonary artery, Ao – aorta.

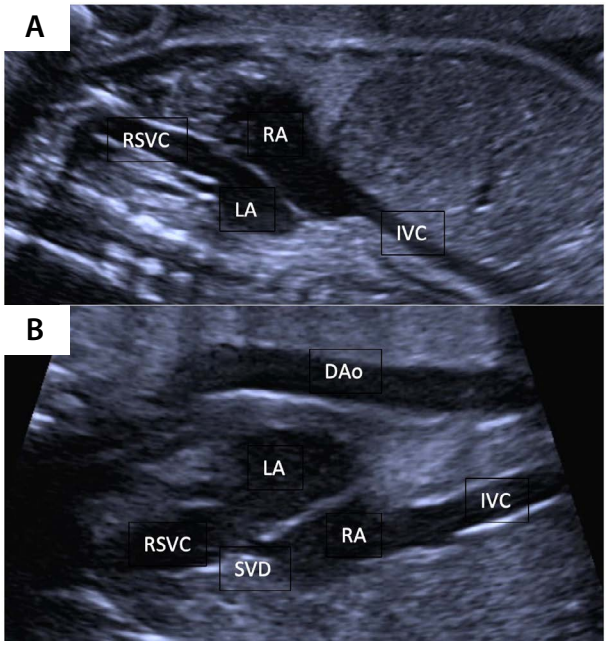

Figure 3. Sagittal images demonstrating (**A**) anomalous insertion of the right superior vena cava (RSVC) into the left atrium (LA) and (**B**) biatrial drainage with the RSVC "overriding" the atrial septum via a sinus venous defect (SVD)

RA – right atrium, IVC – inferior vena cava, Dao – descending aorta

desaturations to 50-60% required positive pressure ventilation and transfer to the neonatal intensive care unit. Because he was weaned off this support in 24 hours, the respiratory complications were attributed to transient tachypnea of the newborn.

Postnatal transthoracic echocardiogram confirmed an RSVC emptying into the LA, with the right upper pulmonary vein entering the LA at the RSVC-LA junction; imaging also identified an unrestrictive atrial communication, variously described as a superior secundum atrial septal defect and as a sinus venous defect (Figure 4). Left to right shunting across

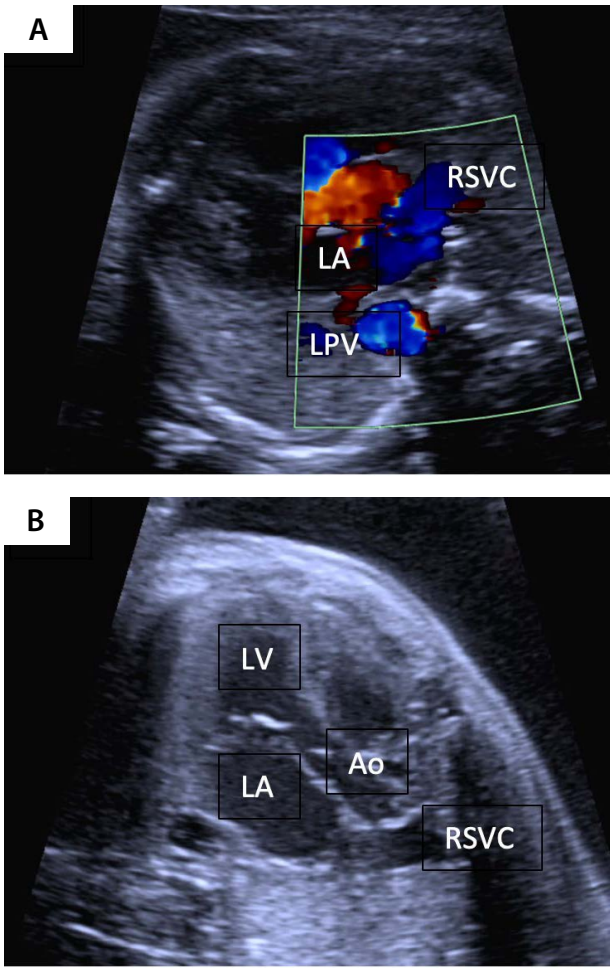

Figure 2. Four-chamber view demonstrating an unusual flow pattern in the rightward aspect of the left atrium (LA) from the right superior vena cava (RSVC) (**A**). Long-axis view demonstrating entrance of a dilated RSVC into the LA (**B**)

LPV – left pulmonary vein, LV – left ventricle, Ao – aorta.

the atrial communication allows partial drainage of the RSVC into the RA. There is no SVC connected to the RA, but the IVC enters the RA normally. The patient's oxygen saturations were in the upper 80s to low 90s at hospital discharge and 92-99% at subsequent follow-up visits in the pediatric cardiology clinic. He has not had any other apparent cardiac symptoms to date.

At 12 months of age, he underwent cross-sectional imaging, which demonstrated a sinus venosus defect with partial anomalous pulmonary venous drainage of the right upper lobe pulmonary veins to the RSVC, which itself inserts anomalously into the LA (Figure 5). Surgical repair is anticipated in the coming months; he is currently doing well at 21 months of age.

Discussion

Anomalous drainage of the RSVC to the LA, as an isolated abnormality (Figure 6), is a very rare form of congenital heart disease (0.5%) that causes hypoxemia [1, 2]. It was first described in 1914 as biatrial drainage of the SVC [1, 3, 4]. There is only one other case reported in the literature of this anomaly diagnosed prenatally [1].

Various theories have been suggested to explain the embryologic origin of this developmental malformation, which is effec-

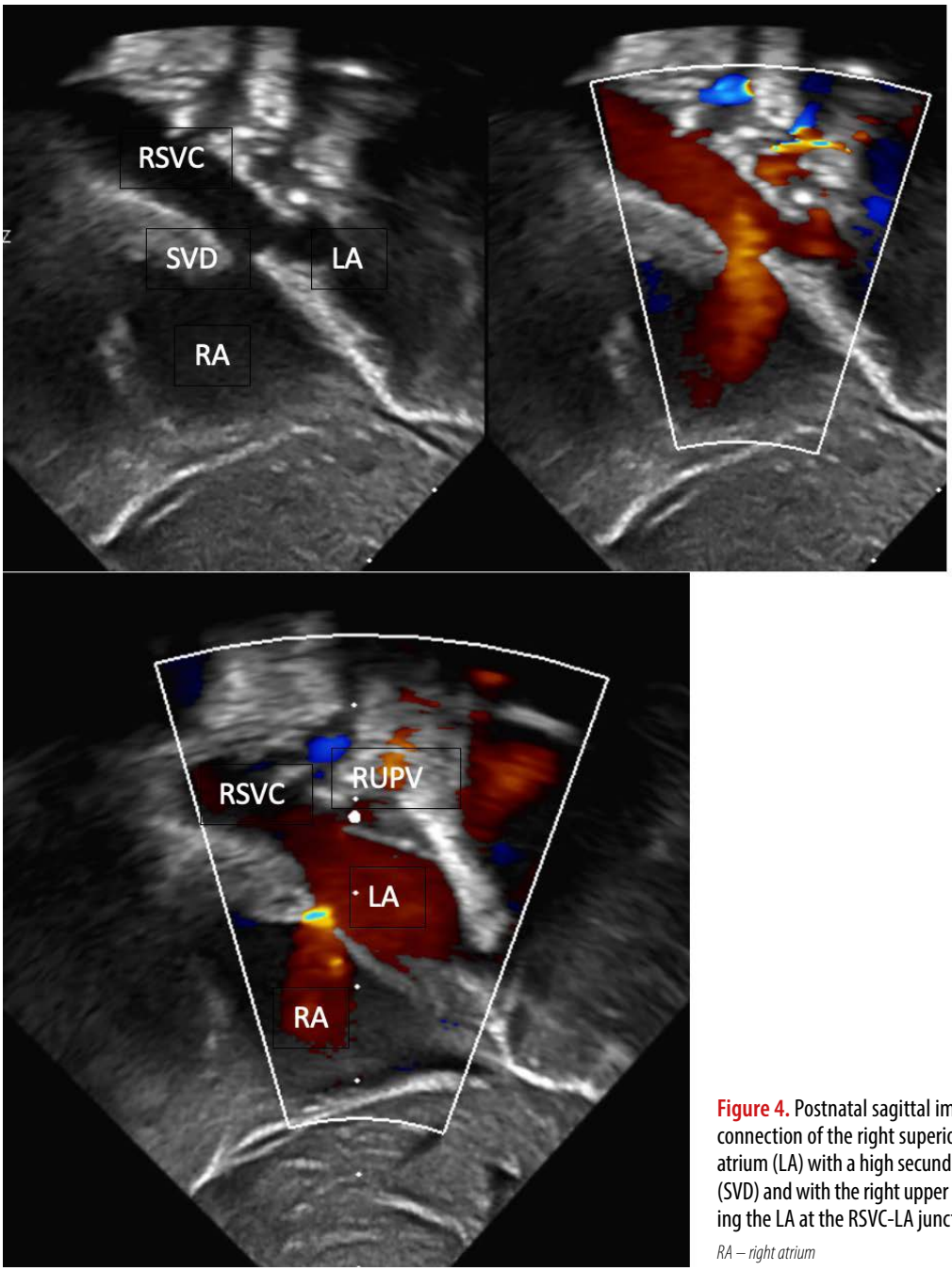

Figure 4. Postnatal sagittal imaging confirming anomalous connection of the right superior vena cava (RSVC) to the left atrium (LA) with a high secundum versus sinus venous defect (SVD) and with the right upper pulmonary vein (RUPV) entering the LA at the RSVC-LA junction

RA – right atrium

tively a type of interatrial communication [5]. The original theory by Kirsch et al. in 1961 encompassed an abnormal position of the right horn of the sinus venosus, involving "relative leftward and cephalic distortion that resulted in placing the aperture of the SVC alone in the LA" [6]. An alternative theory postulated that the cephalic portion of the right valve of the sinus venosus fuses with the atrial septum superior to the coronary sinus inlet, forming a seal that prevents the SVC from draining into the RA [7]. In 2003 the Drs. Van Praagh described this abnormality as a localized cavopulmonary venous defect in the wall between the RSVC and the right upper pulmonary veins; the posterior wall of the RSVC is contiguous with the anterior wall of the right pulmonary veins, creating a venovenous bridge [8]. If the RSVC blood flows preferentially into the LA, its right atrial orifice will become atretic such that the RSVC drains into the LA [8]. Dr. Anderson et al. recently refuted this theory, stating that the SVC and the pulmonary veins never share a common wall; rather, the right pulmonary veins can drain anomalously into the SVC while retaining their connection to the LA [9].

Although usually associated with mild degrees of cyanosis and hypoxemia, anomalous drainage of the RSVC to the LA is not expected to cause problems prenatally or in the neonatal period. It may be associated with anomalous pulmonary venous connections, and careful prenatal and postnatal assessment for associated pulmonary venous abnormalities is warranted [10]. Those without atrial septal defects typically have lower baseline oxygen saturations [10]. There is a low incidence of associated cardiac or extracardiac anomalies [1].

Cross-sectional imaging with CT or cardiac MRI can help confirm a prenatal diagnosis of biatrial or left atrial drainage

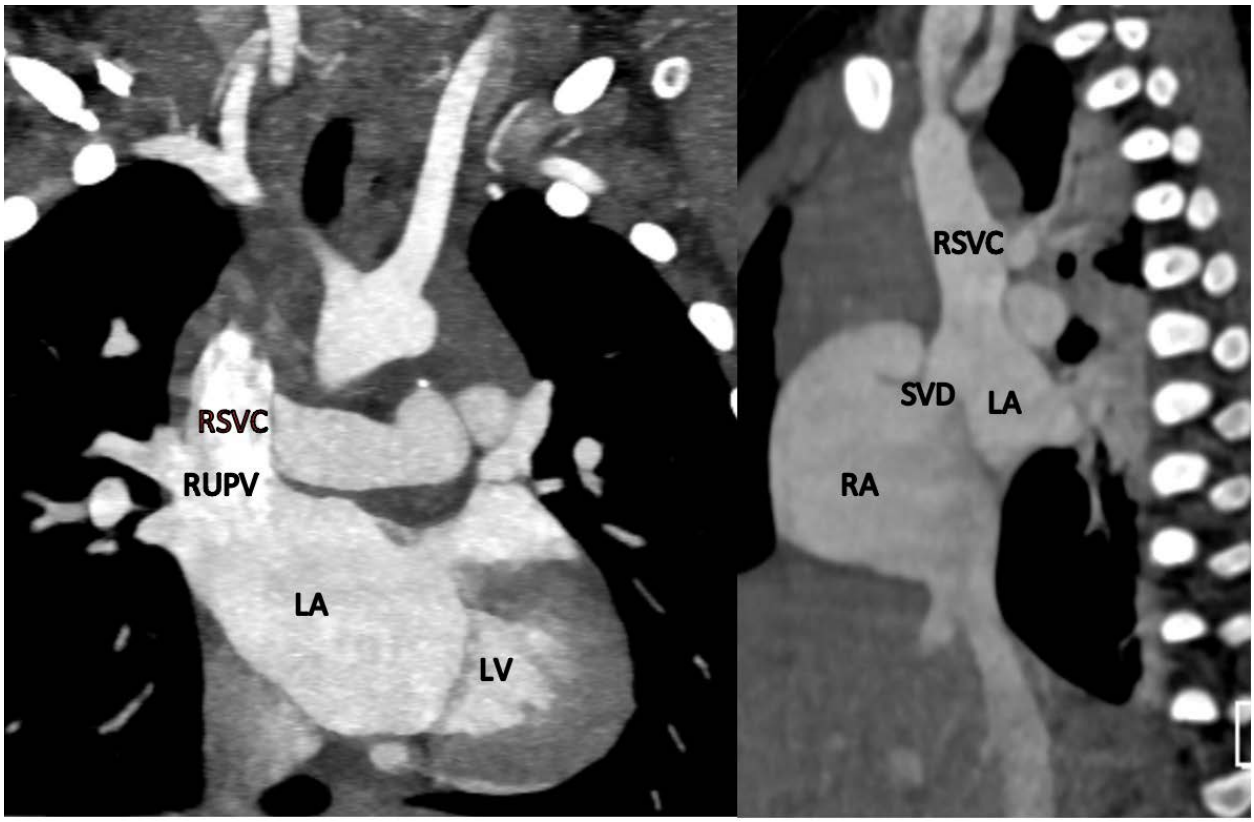

Figure 5. CT scan demonstrating a sinus venosus defect (SVD) with partial anomalous pulmonary venous drainage of the right upper lobe pulmonary veins (RUPV) to the right superior vena cava (RSVC), which itself inserts anomalously into the left atrium (LA)

LV – left ventricle, RA – right atrium.

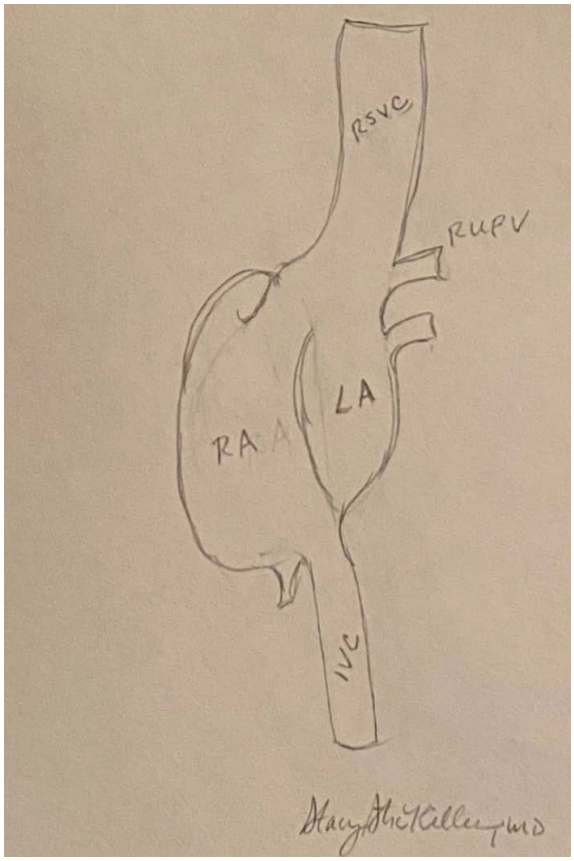

Figure 6. Schematic drawing: right superior vena cava (RSVC) to the left atrium (LA), sagittal image

LV – left ventricle, RA – right atrium, RUPV – right upper pulmonary vein.

of the RSVC and further define the combined anomalous systemic and pulmonary venous drainage [5, 11]. Surgical repair includes trans-section and re-anastomosis of the RSVC to the RA/right atrial appendage to prevent the long-term complications of cyanosis and right-to-left shunting, including brain abscesses, paradoxical emboli, renal and splenic infarcts, and atrial arrhythmias associated with right atrial dilation [8-11]. A Warden procedure transects the SVC superior to the SVC-pulmonary venous junction, when there is associated partial anomalous pulmonary venous drainage, to preserve pulmonary venous return to the LA [11, 12].

Dilation of the RSVC prompted referral to fetal cardiology for our patient and for the only other prenatally-diagnosed case of anomalous drainage of the RSVC to the LA in the literature [1]. The differential diagnosis of *in utero* dilation of the RSVC includes abnormalities that either obstruct drainage into the RA or increase blood flow into the RSVC [1]. These include vein of Galen or other cerebrovascular arteriovenous malformations, fetal anemia, thoracic masses/tumors causing SVC obstruction, supracardiac total or partial anomalous pulmonary venous return, and anomalous drainage of the RSVC into the LA.

In the coronal imaging plane, a vein of Galen malformation appears as a tubular, cystic, anechoic structure ("comet tail") below the corpus callosum and cavum septum pellucidum; it is superior to the thalami and contiguous with the dilated sagittal sinus [13]. Cardiac manifestations of this cerebrovascular arteriovenous malformation range from asymptomatic cardiomegaly

with preserved ventricular function to severe congestive cardiac failure and persistent pulmonary hypertension [13, 14]. Cardiac chamber dilation, especially right heart enlargement, hydrops, flow reversal in the aortic arch from "cerebral steal", and abnormal flow patterns (elevated systolic and diastolic velocities) in the MCA may be apparent prenatally [14]. Dilation of the RSCV out of proportion to the IVC occurs because of increased volume load to the cerebral venous system leading to increased blood flow through the RSVC [14]. Hoda et al. reported 2 patients with both vein of Galen malformations and anomalous drainage of the RSVC to the LA, who were only mildly symptomatic; they concluded that the combination of abnormalities may provide a physiological advantage by ameliorating the left-to-right shunt from the arteriovenous malformation [15].

Fetal anemia may be due to red blood cell alloimmunization, infection (especially with parvovirus B19, CMV, and toxoplasmosis), hemoglobinopathies, fetomaternal hemorrhage, placental/fetal tumors, and complications of monochorionic placentation [16, 17]. In the setting of fetal anemia, more oxygen-rich blood from the umbilical vein is shunted to the brain; this increased flow to the brain increases venous return to the RSVC and dilates this vessel and the right heart. An MCA-PSV > 1.50 MoM correlates with moderate to severe fetal anemia [16, 17].

Thoracic masses, including lymphoma, teratoma, neurogenic tumors, foregut cysts, hemangiomas, lymphatic malformations, chest wall sarcomas and hamartomas, vascular malformations, pleuropulmonary blastoma, and congenital pulmonary airway malformations, can cause RSVC dilation by obstructing flow into the RA [18, 19]. Fetal MRI may help identify these abnormalities prenatally [19].

With supracardiac total or partial anomalous pulmonary venous return (TAPVR/PAPVR), dilation of the RSVC occurs from pulmonary venous drainage through this vessel. While prenatal diagnosis of TAPVR remains challenging, identifying a smooth-walled left atrium, an increased/hypoechoic "space" between the LA and the descending aorta, and a confluence posterior to the LA ("twig sign") can aid in this diagnosis [20-22].

Finally, as demonstrated in our case and in the case reported by Vassallo et al. [1], drainage of the RSVC to the LA can cause RSVC and innominate vein dilation with a relatively normal-sized IVC because of associated partial anomalous venous drainage to the RSVC. Those cases of RSVC to the LA without associated pulmonary venous anomalies may not manifest such enlargement.

Conclusions

Anomalous drainage of the RSVC to the LA, as an isolated abnormality, is a very rare form of congenital heart disease that causes postnatal hypoxemia, which may be mild and not clinically apparent in the newborn period. Most reported cases in the literature have been identified in later childhood or adulthood, but universal pulse oximetry newborn screening may increase postnatal detection. Prenatal diagnosis, though challenging, is possible in the current era, as our case illustrates, and can prepare families and clinicians for postnatal care. *In utero* dilation of the RSVC should prompt careful evaluation for extracardiac, systemic venous, and pulmonary venous anomalies and may facilitate prenatal detection of this rare cardiac anomaly.

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Conflict of interest

The author declares no conflicts of interest.

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Stacy A.S. Killen (ORCID: 0000-0002-6458-3669): research concept and design, collection and/or assembly of data, data analysis and interpretation, writing the article, critical revision of the article, final approval of the article.