

# Prenatal diagnosis of obstructed supracardiac total anomalous pulmonary venous connection at 23 weeks with successful immediate postnatal surgical correction

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## ABSTRACT

**Diagnosis of isolated total anomalous pulmonary venous connections (TAPVCs) is relatively rare in fetal life, especially in early gestation. We report a case of a fetus diagnosed with the supracardiac type of TAPVC at 23 weeks gestation, with evidence of obstruction to connection of the common vertical vein to the superior vena cava. The neonate had a critical presentation at birth and underwent an emergency surgical repair immediately after birth with excellent outcome on short term follow-up with the resolution of pulmonary artery hypertension.**

**Keywords:** Antenatal diagnosis, fetal echocardiography, pulmonary artery hypertension, supracardiac total anomalous pulmonary venous connection, total anomalous pulmonary venous connection

## INTRODUCTION

To diagnose isolated total anomalous pulmonary venous connection (TAPVC) is relatively rare in fetal life, especially in the early gestation. We report a case of a fetus diagnosed with the supracardiac type of TAPVC at 23 weeks gestation, with evidence of obstruction to connection of the common vertical vein to the SVC. The neonate had a critical presentation at birth and underwent an emergency surgical repair immediately after birth with excellent outcomes on short term follow-up with resolution of pulmonary artery hypertension.

## CASE REPORT

A 32-year-old primigravida was referred for fetal echocardiography at 23 weeks of gestation in view of right heart enlargement and inability to demonstrate drainage of pulmonary veins to the left atrium. Fetal

echocardiography performed at our center showed significant right-left chamber asymmetry with dilatation of the right-sided chambers in the four-chamber view [Figure 1a and Video 1]. The left atrium was small with a smooth posterior wall, and the pulmonary veins were not seen connecting to the left atrium [Figure 1b]. The outflow tracts were normally related and unobstructed and there were no abnormal systemic venous connections. The area behind the heart showed a common chamber measuring 1.5 mm in diameter between the LA and the descending aorta [Figure 1c]. The pulmonary veins were demonstrated as joining this common chamber [Figure 1d]. The superior vena cava (SVC) appeared dilated, and a channel with turbulent high velocity was seen draining into the SVC in the three-vessel trachea view [Figure 1e and Video 2]. Doppler evaluation of this channel showed continuous flow into the SVC, suggesting obstruction at the site

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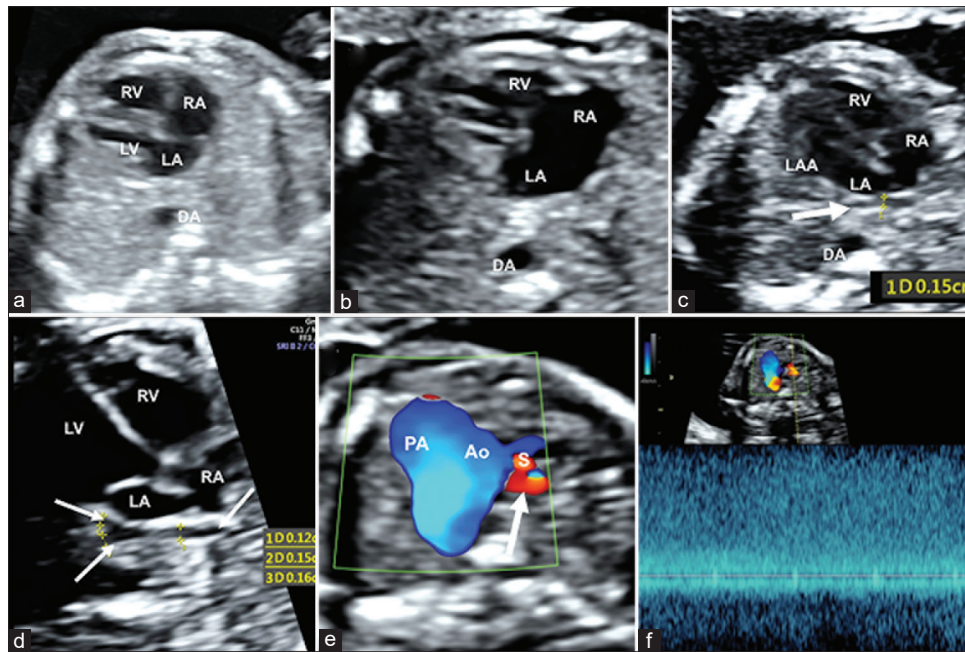
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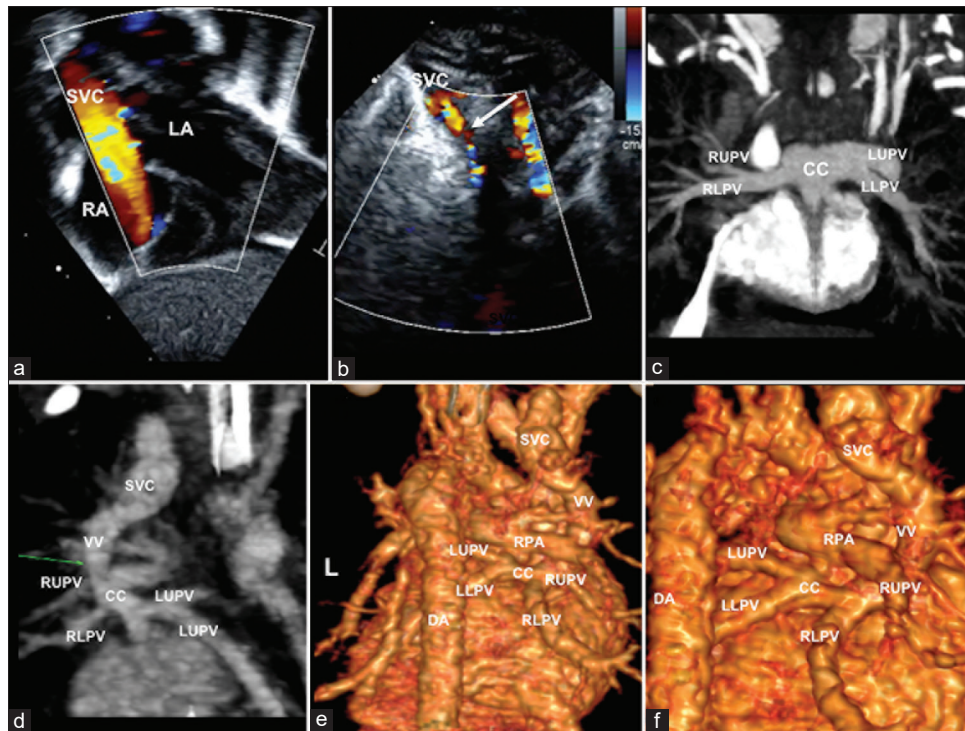
**Figure 1:** Fetal echocardiography imaging of supracardiac TAPVC to the SVC with obstruction. This sequence of images depict the fetal echocardiographic diagnosis of obstructed type of supracardiac type of TAPVC in a 23 week fetus. (a) The 4-chamber view with dilatation of RA and RV. (b) A relatively smooth wall of the LA with absence of the pulmonary vein connections. (c) The presence of an abnormal channel (arrow) in the area behind the LA. (d) The drainage of the individual pulmonary veins (arrows) into this channel. (e) A 3-vessel view with color Doppler showing the common channel draining into the SVC (s) with flow aliasing suggesting turbulence and obstruction. (f) Pulse wave Doppler of the vertical vein drainage site into SVC showing continuous flow suggestive of obstruction. RA: Right atrium, RV: Right ventricle, LA: Left atrium, LV: Left ventricle, LAA: Left atrial appendage, DA: Descending aorta, PA: Pulmonary artery, Ao: Aorta, SVC: Superior vena cava, TAPVC: Total anomalous pulmonary venous connection

of joining of the channel into the SVC [Figure 1f]. A diagnosis of the obstructed type of supracardiac TAPVC was made. The findings were re-confirmed in a repeat scan at 34 weeks, and the parents were counseled about the expected postnatal course and the need for urgent surgical intervention after birth. Following prenatal counseling, the family opted for postnatal cardiac care and surgical correction. Hence, the delivery was planned in a tertiary pediatric cardiac surgical facility for expediting postnatal cardiac care and avoiding the risks of neonatal transport.

At 39 weeks' gestation, the mother was admitted in our hospital with a history of premature rupture of membranes. In view of significant cephalopelvic disproportion, an emergency cesarean section was performed, and a male baby weighing 2.4 Kg was delivered. The Apgar score at 1 and 5 min after birth was 8 and 9, respectively. The baby presented with significant cyanosis, tachypnea with a respiratory rate of 60–70 breaths/min, suprasternal and subcostal retractions. Pulse oximetry in room air showed a saturation of 70% immediately after birth. The baby's respiratory distress worsened rapidly, requiring mechanical ventilation. The chest radiography revealed signs of severe pulmonary venous congestion. Postnatal echocardiography confirmed the prenatal findings. The SVC was markedly dilated [Figure 2a]. The suprasternal

view revealed a vertical vein ascending to the right of the SVC; the vertical vein appeared to have a tortuous course before draining into the SVC [Figure 2b]. The drainage appeared obstructed at the ascending vertical vein on color flow mapping [Video 3]. There was evidence of severe pulmonary arterial hypertension with a moderate-sized ASD. Computed tomography (CT) angiogram was done in order to confirm the pulmonary vein anatomy and delineate the anatomy of the vertical vein more clearly. CT revealed the presence of reasonable sized individual pulmonary veins which joined together from both sides at multiple levels with a small confluent chamber. The vertical vein was narrow and tortuous and it ascended to the right of the midline to join the right-sided SVC [Figure 2c-f]. A tight stenosis was noted at the confluence site measuring approximately 2.2 mm. Three-dimensional rendering showed that the vertical vein was very thin along the entire length, causing obstruction throughout its course.

In view of the unstable hemodynamics and low oxygen saturations despite ventilation, the neonate underwent an emergency open-heart surgery within 2 h after birth. The anatomic findings were confirmed on surgery. A side-to-side anastomosis was performed between the pulmonary venous confluence and left atrium. The vertical vein was identified lateral to the right SVC



**Figure 2: Postnatal imaging (echocardiography and high resolution 256 slice CT with 3D reconstruction) confirming the diagnosis.** This sequence depicts the postnatal echocardiography and high resolution 256 slice CT reconstruction of the anatomy of the lesion. (a) Neonatal echocardiography showing dilated RA and SVC with increased flow in SVC as shown by flow aliasing. (b) Depicts the drainage site of the vertical vein (arrow) into the SVC; the vertical vein is very narrow with turbulent flow. (c) Depicts a coronal view in 256 slice CT showing the confluence of the 4 pulmonary veins forming a CC. (d) A sagittal view in 256 slice CT showing the individual pulmonary veins forming the confluence and ascending up as VV into the SVC. (e and f) Depict the 3D reconstruction of the anatomy of the TAPVC. The individual pulmonary veins are clearly seen joining together forming a very short CC which ascends posterior to the RPA as the obstructed VV into the SVC. RA: Right atrium, LA: Left atrium, SVC: Superior vena cava, LUPV: Left upper pulmonary vein, LLPV: Left lower pulmonary vein, RUPV: Right upper pulmonary vein, RLPV: Right lower pulmonary vein, CC: Common chamber, VV: Vertical vein, DA: Descending aorta, TAPVC: Total anomalous pulmonary venous connection, CT: Computed tomography, 3D: Three dimensional

and looped. ASD was closed using pericardial patch leaving behind a small ASD. The intraoperative course was uneventful, except for systemic PA pressures while coming off bypass. The patient had supra systemic PA pressures in the immediate postoperative period, and hence, the sternum was kept open for 3 days. Poststernal closure, the baby was gradually weaned and extubated on the 11<sup>th</sup> postop day with stable hemodynamics. The baby required high flow nasal canula support for about 6 weeks due to persisting mild respiratory distress. The child was discharged on the 45<sup>th</sup> postoperative day and at the time of discharge, the baby was active, alert and feeding well. Postoperative echocardiogram revealed unobstructed pulmonary vein anastomosis with no residual PAH. At 5 months follow-up, the baby had gained weight (4.8 kg) and there was no residual pulmonary vein obstruction or pulmonary hypertension on follow-up echocardiogram. Cardiac medications were weaned off, and the baby is scheduled for further follow-up for the reassessment of PA pressures at 1 year of age.

## DISCUSSION

TAPVC is rare congenital heart disease, accounting for 1%–3% of all cardiac malformations. There are very few reports describing diagnosis of isolated TAPVC during fetal life.<sup>[1]</sup> There are several reasons of this, including difficulty in imaging pulmonary veins in the prenatal period and the lack of hemodynamic variations due to the presence of TAPVC in the fetus in view of the very small amount of cardiac output coursing through the fetal lungs. Most cases of TAPVC in the fetus were reported in the third trimester, with very few reports of early or mid-trimester diagnosis of this condition as an isolated defect.<sup>[1]</sup> In a recent series of 10 cases of postnatally confirmed isolated TAPVC seen at a single institution, the prenatal diagnosis was made at a mean gestational age of 31 weeks (range 25–37).<sup>[1]</sup> In our case, the diagnosis was suspected very early in pregnancy and confirmed at 22 weeks. Several indirect echocardiographic signs have been proposed as possible clues for suspecting TAPVC in the fetus.<sup>[2–4]</sup> These include

chamber asymmetry with right-sided dominance, small left atrium with a small posterior wall (the bald left atrium) and dilatation of the receiving systemic vein-like SVC or inferior vena cava.

The right heart dilatation is typically late in fetal life due to the fact that the proportion of cardiac output distributed to the lungs, and hence, the pulmonary venous return is typically low in early gestation. Obstruction of the pulmonary venous confluence may be extremely difficult to detect in fetal life owing to the low pulmonary blood flow in the fetus. However, the presence of in-utero pulmonary venous obstruction can have a significant impact on the fetal lungs due to in-utero pulmonary venous hypertension and could become a risk factor for persistent pulmonary hypertension in the postnatal period even after surgical correction.<sup>[5]</sup> When there is a severe obstruction to the pulmonary venous drainage, reactive pulmonary arteriolar constriction occurs and this coupled with decreased PO<sub>2</sub> of blood perfusing the lungs affects pulmonary vascular smooth muscle development. In a study of infants with obstructed TAPVC, the pulmonary vascular smooth muscle was markedly increased in pulmonary arterioles and more peripheral vessels also showed increased smooth muscle development.<sup>[5]</sup> The degree of obstruction of the pulmonary venous channel (the right-sided vertical vein in our case) connecting the pulmonary veins to the systemic veins is one of the most important determinants of the hemodynamic effects and clinical manifestations of the anomaly. In our case, the entire length of the vertical vein was narrow, causing severe pulmonary venous congestion. The cardiac output was maintained entirely by the foramen ovale despite the presence of severe pulmonary venous congestion and pulmonary edema.

This case report illustrates the importance of prenatal diagnosis and planned peri-partum care in a tertiary pediatric cardiac facility in neonates with critical, life-threatening forms of CHD.<sup>[6]</sup> Multiple studies from high-income countries have shown the positive impact of prenatal diagnosis on improving the peri-operative outcomes in neonates with critical CHD.<sup>[7-11]</sup> Neonatal transport is very challenging in situations like our case where the neonate became symptomatic at birth and rapidly deteriorated, needing mechanical ventilation. The infant with TAPVC with obstruction to venous return requires urgent surgery. In the current era, the surgical outcomes for obstructed TAPVC in the neonate have significantly improved with a survival rate of >95%.<sup>[12]</sup> Re-stenosis of the anastomotic site between the pulmonary veins and the left atrium has been reported in around 15%–20% of infants following surgery, but with recent improvements in technique, the incidence of this complication has been reduced considerably.<sup>[12]</sup> In our case, the prolonged postoperative course was explained by the persistence of pulmonary hypertension due to the effects of in-utero pulmonary venous hypertension, pulmonary arteriolar hyperplasia, and vasoconstriction. These

gradually improved following the relief of the obstruction to the pulmonary venous flow and improved oxygenation due to prompt and early surgical intervention.

## CONCLUSIONS

Prenatal diagnosis of obstructed forms of TAPVCs is feasible in early fetal life, and this can have a significant impact on surgical outcomes after neonatal repair.

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

### Conflicts of interest

There are no conflicts of interest.

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