

# Prenatal diagnosis of total anomalous pulmonary venous connection to the portal vein associated with right atrial isomerism

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**KEYWORDS:** portal vein; prenatal diagnosis; right atrial isomerism; total anomalous pulmonary venous connection

## ABSTRACT

*We report the prenatal diagnosis of total anomalous pulmonary venous connection to the portal system in a 20-week fetus with right atrial isomerism. The apex of the fetal heart pointed to the left, the fetal stomach was on the right, there was a common atrioventricular valve, the left ventricle was small and the abdominal aorta and inferior vena cava were on the left side; all these features were suggestive of right atrial isomerism. An anomalous vein was connected to the portal vein which ascended above the diaphragm and ended in a confluence of pulmonary veins, posterior to the common atrium. Color Doppler imaging helped confirm the diagnosis of total anomalous pulmonary venous connection. The prenatal findings were confirmed on autopsy. Copyright © 2003 ISUOG. Published by John Wiley & Sons, Ltd.*

## INTRODUCTION

Total anomalous pulmonary venous connection (TAPVC) defines an anomaly in which the pulmonary veins fail to connect to the left atrium and drain, either directly or via the systemic veins, into the right atrium. The condition is frequently seen as part of the complex cardiac anomaly of heterotaxy syndrome, especially as part of right atrial isomerism syndrome. Only rarely is it seen as an isolated condition. In different series the incidence varies from 0.4 to 2% of all cardiovascular anomalies.

We report the fetal echocardiographic features including color and pulsed Doppler findings in a case of TAPVC to the portal vein, seen in association with right atrial isomerism.

## CASE REPORT

A 31-year-old woman, gravida 5 para 2, was referred for fetal echocardiography at 20 weeks' gestation, because

of a fetal cardiac malformation in a previous pregnancy. Her first two pregnancies resulted in miscarriages around 8 weeks' amenorrhea. In her third pregnancy the newborn was found to have complete atrioventricular canal defect and subsequently died. Her fourth pregnancy resulted in the delivery of a normal male who was 2 years old at the time of writing. The family had no history of consanguinity.

Ultrasound examination was performed at 20 weeks' gestation using an ATL HDI 5000 (Advanced Technology Laboratories, Bothell, WA, USA) ultrasound machine. Biometry was consistent with gestational age. The apex of the fetal heart pointed to the left (Figure 1) and the fetal stomach was seen on the right side, opposite the apex of the heart (Figure 2). Fetal echocardiography demonstrated a common atrioventricular valve (Figure 1) and the left ventricle was found to be slightly reduced in size. The abdominal aorta and inferior vena cava were seen on the left side. All these features were suggestive of right atrial isomerism. There was an anomalous vein connected to the portal vein (Figure 3), which ascended above the diaphragm (Figure 4) and ended in a confluence of the pulmonary veins, posterior to the common atrium (Figure 5). The pulmonary veins were not connected to the atrium. The features suggested TAPVC to the portal vein. Color Doppler imaging showed flow in the pulmonary veins, confluent vein and anomalous vein, away from the heart and towards the portal vein. On pulsed Doppler examination, the anomalous vein showed continuous, mildly pulsatile flow with a peak velocity of 15 cm/s (Figure 6).

The parents chose to terminate the pregnancy. Autopsy revealed the absence of the spleen, the stomach on the right side, the cardiac apex towards the left side and the common atrioventricular valve, confirming right atrial isomerism. There was a large anomalous vein on the



**Figure 1** Ultrasound image (four-chamber view) of fetal heart showing common atrioventricular valve and apex of the heart pointing to the left. The fetus is lying longitudinally, in cephalic presentation, and the spine is posterior on the right side.

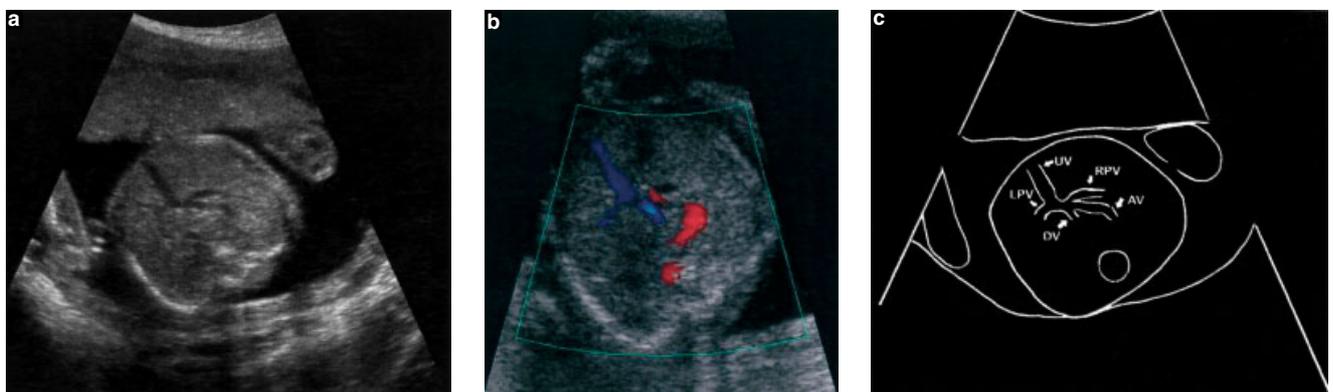
anterior surface of the stomach (Figure 7a). This vein was connected below to the portal vein and extended into the thorax through the esophageal hiatus (Figure 7b). The pulmonary veins were seen to drain into this anomalous vein (Figure 7c). A needle was inserted into the cephalic end of this anomalous vein and contrast was injected; the X-ray taken confirmed the connections of the anomalous vein (Figure 8).

## DISCUSSION

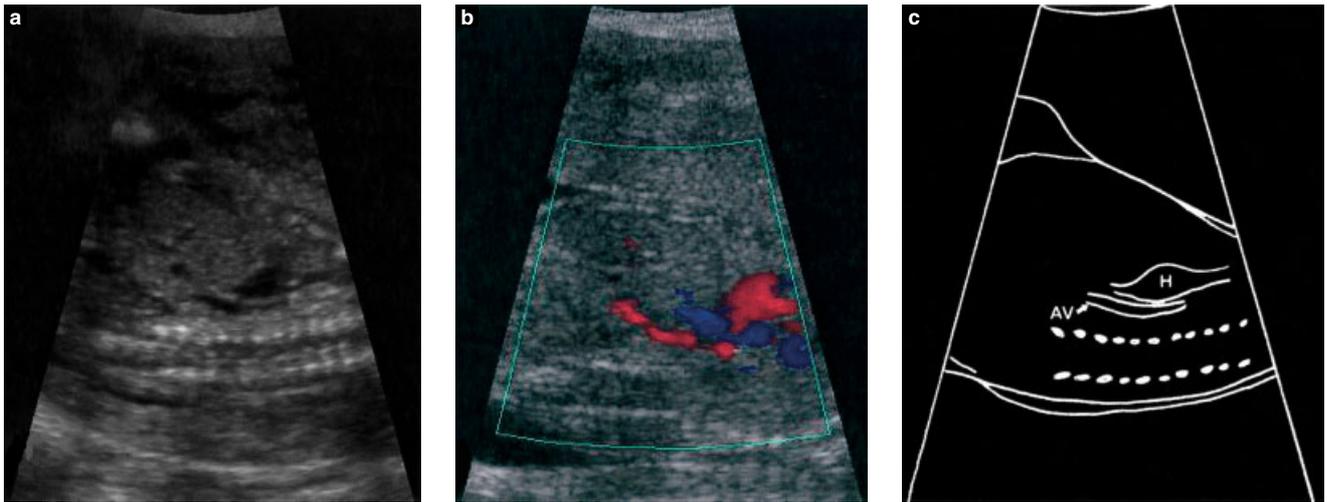
In TAPVC the pulmonary veins fail to connect to the left atrium and they drain by an anomalous vein into the right atrium or systemic veins. The anomalous vein can join a supracardiac site (47%), a cardiac site (31%), an infracardiac site (13%) or multiple sites (9%)<sup>1</sup>. The infradiaphragmatic connection can be to the inferior vena cava, hepatic vein, portal vein, ductus



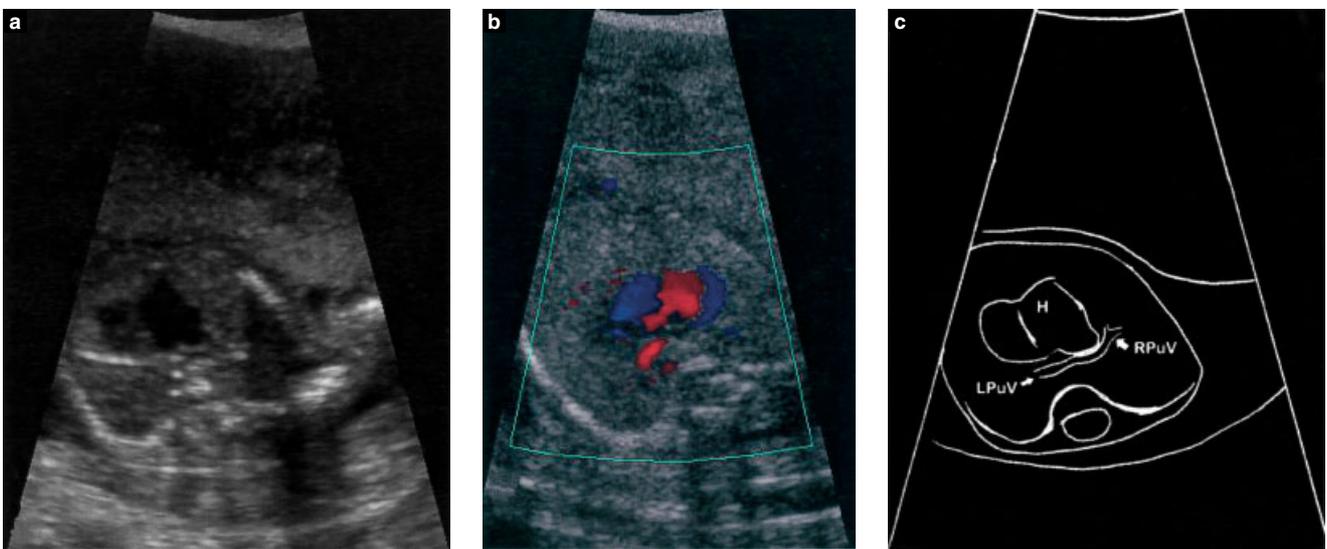
**Figure 2** Ultrasound image (transverse view) through the upper abdomen showing fetal stomach on the right side, opposite to the apex of the heart.



**Figure 3** Grayscale (a) and color Doppler (b) ultrasound images through the upper abdomen showing the confluence of umbilical vein (UV), left and right branches of the portal vein (LPV and RPV), ductus venosus (DV) and the anomalous vein (AV), as indicated in the line diagram (c).



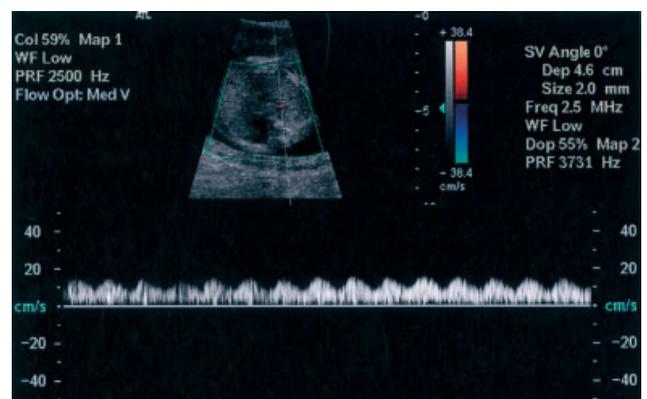
**Figure 4** Grayscale (a) and color Doppler (b) ultrasound images of the vertical portion of the anomalous vein (AV) seen posterior to the heart (H), as indicated in the line diagram (c).



**Figure 5** Grayscale (a) and color Doppler (b) ultrasound images of a transverse section of the chest showing the pulmonary veins (RPuV and LPuV) and their confluence posterior to the heart (H), as indicated in the line diagram (c).

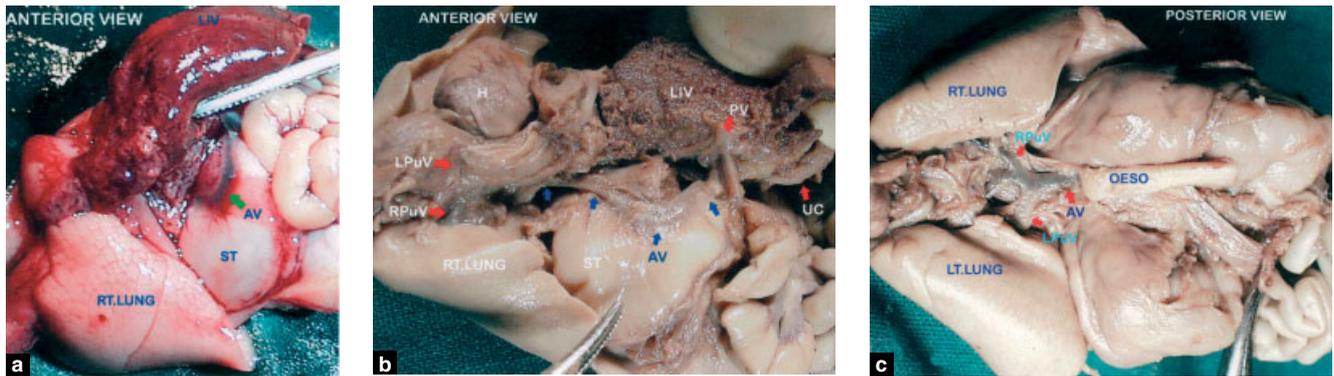
venous, splenic vein or gastric vein. The TAPVC is frequently seen as part of the complex cardiac heterotaxy syndrome, especially as part of right atrial isomerism syndrome. Prenatal diagnosis of TAPVC is beneficial, because affected neonates can deteriorate very rapidly after delivery and surgical treatment can be planned.

Visualization of pulmonary veins has become part of complete fetal echocardiographic examination. Pulmonary veins can be identified in almost all fetuses at 20–21 weeks of gestation<sup>2</sup> and normal velocities relative to gestational age have been determined<sup>3</sup>. The normal pulmonary venous blood flow velocity waveform consists of biphasic forward flow components, suggesting that pulmonary venous flow is influenced by dynamic changes in left atrial pressure. Initial reports on the prenatal diagnosis of TAPVC were based on the appearance on grayscale imaging<sup>4,5</sup>. However, the positive identification of a normal pulmonary venous connection to the

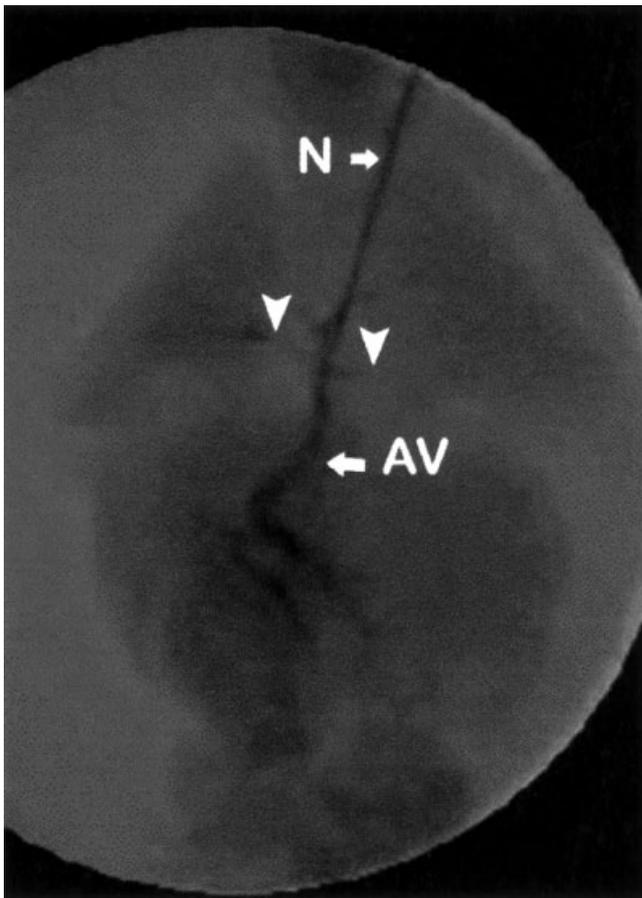


**Figure 6** Pulsed Doppler ultrasound study of the anomalous vein.

left atrium is difficult. The diagnosis has subsequently been based on pulsed Doppler examination of pulmonary vein blood flow<sup>6,7</sup>. In TAPVC, there is a low-velocity,



**Figure 7** Autopsy photographs showing the confluence of pulmonary veins and anomalous vein: anterior views before fixation (a) and after fixation and dissection (b) and posterior view (c). AV, anomalous pulmonary vein; ST, stomach; LIV, liver; RT., right; LT., left; RpuV and LpuV, right and left pulmonary veins; PV, portal vein; UC, umbilical cord; H, heart; OESO, esophagus.



**Figure 8** X-ray image after injection of contrast by a needle (N) into the upper part of the anomalous vein showing the pulmonary veins (arrowheads) and anomalous vein (AV).

mildly pulsatile flow pattern in the pulmonary veins. This represents the lack of the normal influence of the left atrial hemodynamics and is due instead to a distant or dampened connection to a systemic vein. The abnormal pulmonary venous tracing is suggestive of TAPVC and can lead to closer examination and ultimate diagnosis of TAPVC, with the site of connection. The portal vein is a rare site of connection in TAPVC. There is only one previous report of the prospective prenatal diagnosis of TAPVC to the portal vein made at 25 weeks, in which the

authors described the two-dimensional appearance and pulsed Doppler findings<sup>8</sup>.

In our case there were features suggestive of right atrial isomerism. An anomalous vein was seen to be connected to the portal vein. This anomalous vein was traced going through the diaphragm and connecting to a confluence of pulmonary veins. The pulmonary veins were not connected to the left atrium. Color Doppler imaging showed flow towards the portal vein in the anomalous vein. Pulsed Doppler examination of the anomalous vein showed the typical low-velocity, continuous, mildly pulsatile flow characteristic of TAPVC, as described by others. These features were clearly seen at 20 weeks' gestation, the earliest so far reported, and the findings were confirmed at autopsy. The improved resolution of grayscale imaging has facilitated the early diagnosis of this rare condition.

In conclusion, this is a case of a rare form of the infradiaphragmatic type of TAPVC draining into the portal vein. This anomaly was seen as part of right atrial isomerism syndrome.

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