

Supracardiac total anomalous pulmonary venous connection with right ascending vertical vein

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Case report

A 20-day-old neonate was referred to our tertiary care institute for evaluation of increased precordial activity and feeding difficulty. The birth weight was 2.6 kilograms. On examination the child had tachycardia, increased precordial activity, loud pulmonary component of second heart sound and ejection systolic murmur in the pulmonary area. The transthoracic echocardiogram revealed common chamber (confluence) posterior to left atrium and ostium secundum atrial septal defect with right-to-left shunt and right ventricular dysfunction (fig. 1; video 1*). All four pulmonary veins were connected to a common chamber and draining via right sided ascending vertical vein to right superior vena cava (SVC), one centimetre above the SVC-right atrium (SVC-RA) junction (fig. 2, 3, 4 and 5; video 2, 3 and 4*). There was obstruction across the pathway with severe pulmonary arterial hypertension and no other associated anomalies. The pathway of drainage was confirmed by cardiac computed tomography (fig. 5). It was further confirmed upon surgery. The child underwent surgical correction and was doing well postoperatively.

Total anomalous pulmonary venous connection (TAPVC) is classified as supracardiac, cardiac, infracardiac, or mixed forms [1]. Supracardiac TAPVC [2] occurs due to developmental arrest or atresia of the common pulmonary vein. The common pulmonary vein connects the embryonic lung buds with the left atrium. Collateral channels for pulmonary venous drainage develop via the fetal cardinal venous system. Usually, only one of these channels persists to become the vertical vein that connects the pulmonary venous confluence to the SVC or innominate vein. If left cardinal venous system persists, left vertical vein (LVV) will be present which is usually the case. It usually connects to innominate vein. Rarely the right cardinal venous system persists which results in right vertical vein (RVV). RVV usually has an oblique course and drains to SVC or SVC-RA junction [3]. Anomalous pulmonary venous connections have

wide variability and separate vertical veins can also be seen in heterotaxy syndromes [4]. TAPVC without any associated anomalies but with a right ascending vertical vein is a rare association.

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Figure 1

Transthoracic subcostal four chamber view showing dilated right atrium, right ventricle, ostium secundum atrial septal defect with confluence posterior to left atrium.

RA = right atrium; ASD = atrial septal defect.

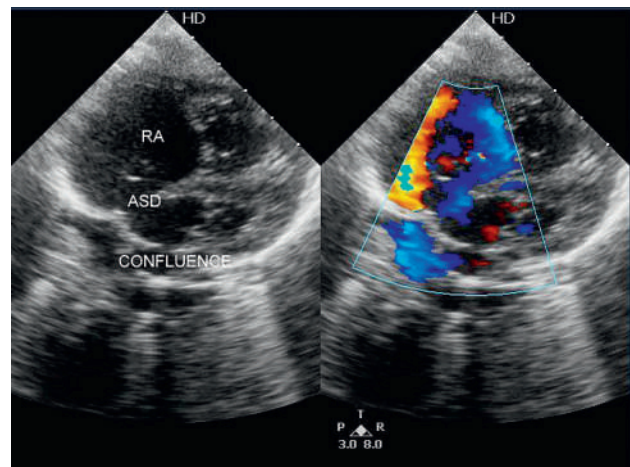
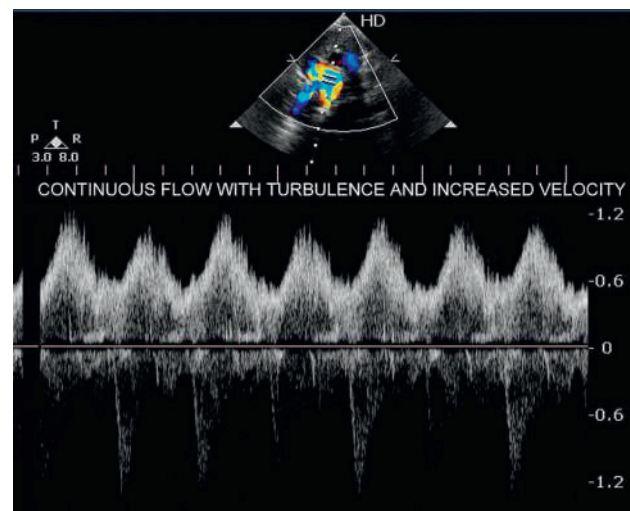


Figure 2

Transthoracic suprasternal long axis view with pulse wave Doppler at the junction of right ascending vertical vein (RVV) with superior vena cava (SVC), showing continuous flow with increased velocity.

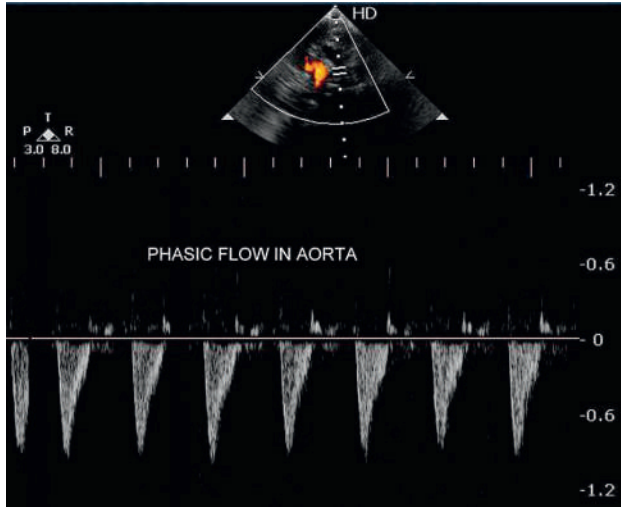


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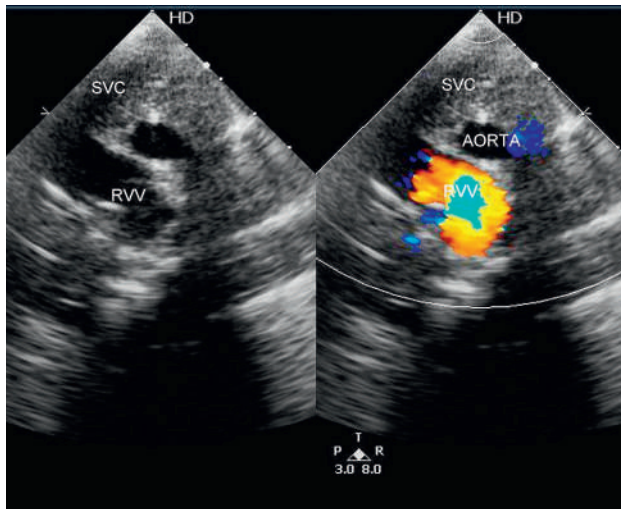
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Figure 3

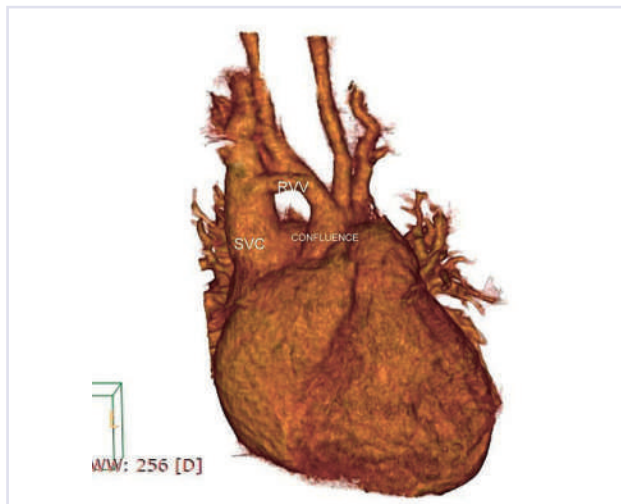
Transthoracic suprasternal long axis view with pulse wave Doppler kept to left of right vertical vein (RVV) showing phasic and pulsatile flow of aorta.

**Figure 4**

Transthoracic modified suprasternal long axis view showing the drainage of right vertical vein (RVV) to right superior vena cava (SVC).

**Figure 5**

Cardiac CT with 3D reconstruction showing right vertical vein (RVV) joining right superior vena cava (SVC) with an almost 90 degree turn.



Acknowledgement

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Video files

You can find the videos on <http://www.cardiovascmed.ch/for-readers/multimedia>.

Video 1: Transthoracic subcostal four chamber view showing dilated right atrium, right ventricle, ostium secundum atrial septal defect with confluence posterior to left atrium.

Video 2: Transthoracic high parasternal view showing superior vena cava (SVC), aorta and pulmonary artery (from right to left) and continuous turbulence in lower portion of SVC.

Video 3: Transthoracic modified suprasternal long axis view showing the drainage of right vertical vein (RVV) to right superior vena cava (SVC).

Video 4: Transthoracic modified suprasternal long axis view showing the drainage of right vertical vein (RVV) to right superior vena cava (SVC) and a point source of turbulence, the probable site of obstruction.

References

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