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CLINICAL VIGNETTE

Prenatal diagnosis of isolated total anomalous pulmonary venous connection (TAPVC) to coronary sinus

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Total anomalous pulmonary venous connection (TAPVC) is the fifth most common cyanotic heart disease, affecting 5.9–7.1 per 100,000 live births [1, 2].

The main feature is that all four pulmonary veins (PV) are not connected to the left atrium (LA) but to the systemic veins, coronary sinus (CS), or right atrium (RA). Four TAPVC types are distinguished based on the PV connect sites: supracardiac, cardiac, infracardiac, and a mixed type. TAPVC can occur as part of complex cardiac malformations, especially in heterotaxy syndrome, or in isolation. Prenatal assessment of the fetal venous system and left venoatrial junction is part of the four-chamber-view evaluation, as stipulated in national and international fetal cardiac screening guidelines [3, 4], however assessment is challenging, and reports indicate that only 1.9% of TAPVC cases are diagnosed prenatally [5].

Prenatal diagnosis of TAPVC is crucial, as it explains postnatal cyanotic heart defect symptoms and enables prompt, sometime lifesaving, treatment planning. The presence of an obstruction at any level of the venous route is the most critical factor affecting outcome and management of the

defect, making the defect one of the few true neonatal cardiac emergencies. Prognosis is very good if detected prenatally and corrected during the neonatal period [2].

The study aim was to identify characteristic sonographic features of TAPVC where there was PV drainage into the CS.

A 35-year-old primigravida at 26 weeks of pregnancy was referred to our department because PV identification by imaging was impossible. Examination revealed normal visceral situs, normal heart size, drainage of the systemic veins into the RA, and concordant atrioventricular connection. The space between the spine and the LA drew our attention as the four PV drained through a collector to the enlarged CS and then to the RA (Fig. 1A, Suppl. video). Imaging also identified the enlarged width of the CS (Fig.1B). In colour and PW Doppler modes, the blood flow spectrum was normal (Fig. 1C). The outflow tracts and upper mediastinum were normal. During the third trimester, the RA and RV became enlarged, and the LA and LV volumes were smaller than normal, however the LV depth was normal and formed the apex of the heart. Trivial tricuspid regurgitation was also detected. Coronary sinus width was 4.9 mm. With TAPVC suspected, examination by a pediatric cardiologist confirmed the diagnosis. The neonate was born at 39 weeks of gestation with vitals of body weight 3300 g, Apgar score 10, venous blood pH = 7.321, and $satO_2$ = 90%.

Postnatal echocardiography and cardiac angiography confirmed that all PV formed a collector behind the LA, and the collector drained into the CS with its kinking (Fig. 1D, Fig. S1). At the level of the CS kinking, a flow gradient of max 16 and mean 12 mm Hg was revealed. The right atrium and ventricle were significantly enlarged (Fig. 1E, Fig. S2), with severe hypertrophy of the right ventricle free wall but normal contractility. Trivial tricuspid valve insufficiency with a retrograde systolic maximal gradient of 34–43 mm Hg, normal right ventricle outflow tract, and widened right and left pulmonary arteries (RPA = 5.2 mm, LPA = 4.7 mm) were observed. Pulmonary blood flow with hypertension symptoms was diagnosed. Atrial septal defect type II of 6 mm with right to left shunt was detected. Left ventricle output was 2.2 L/min/m^2, with normal ejection 75% and shortening fraction 40%. The mitral valve was competent, and the aortic valve was tricomissural and trileaflet, and functioning normally. The coronary arteries originated normally. Imaging identified a normal left aortic arch with normal wide isthmus.

Total correction was done on the 10th day of life: CS drainage was converted from the right to the left atrium. The post-surgery course was not complicated. Follow-up examination proved non-restrictive blood flow through the established coronary sinus to the LA connection. The ventricular systolic function and heart output were normal. The neonate was discharged on the 15th day of life in good general condition, with arterial SatO2 92-95%. Out-patient follow up indicated the child was healthy and developing well.

In conclusion, though difficult, TAPVC can be diagnosed using antenatal ultrasound. In our case, 2D and colour Doppler were chosen for diagnosis. In-utero diagnosis enabled a multidisciplinary approach and optimal prenatal and postnatal patient counselling and treatment.

Article information and declarations

Ethics statement

The institutional review board waived the requirement for a separate ethical approval for this clinical vignette, since the sonographic evaluations were performed as integral parts of routine clinical care, for which informed consent had been previously given by the patient. Data were anonymized.

Author contributions

Anna Wojtowicz — conecpt,analysis and interpretation of data, article draft, literature review, corresponding author.

Beata Zaluska-Pitak — collecting data, article draft, literature review.

Mgadalena Juszcak — literature review.

Hubert Huras — literature review.

Sebastian Goreczny — verification, literature review.

Acknowledgments

None.

Conflict of interest

The authors declare no conflict of interest.

Supplementary material

Supplementary video and Figures S1–S2.

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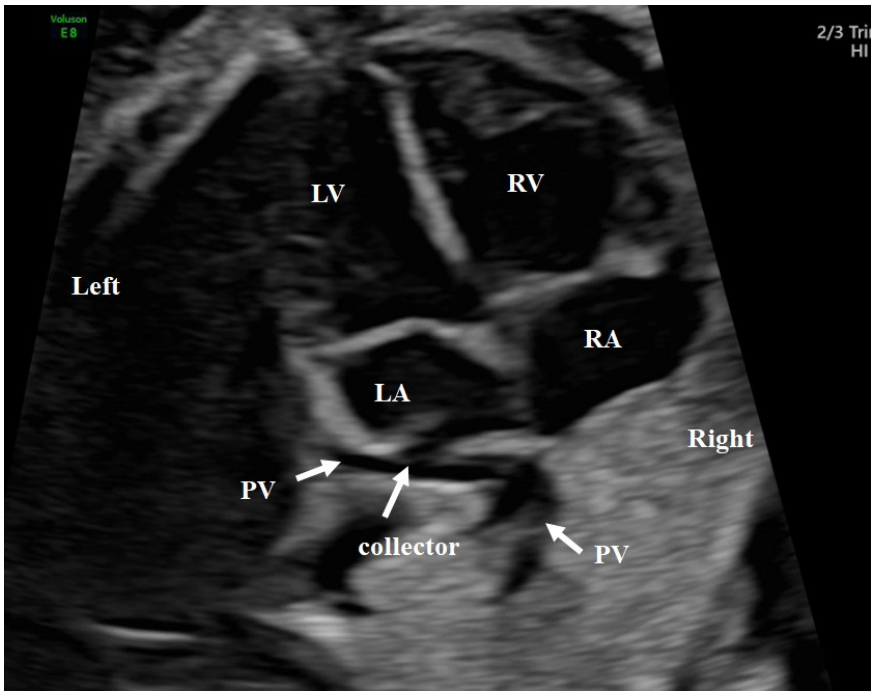
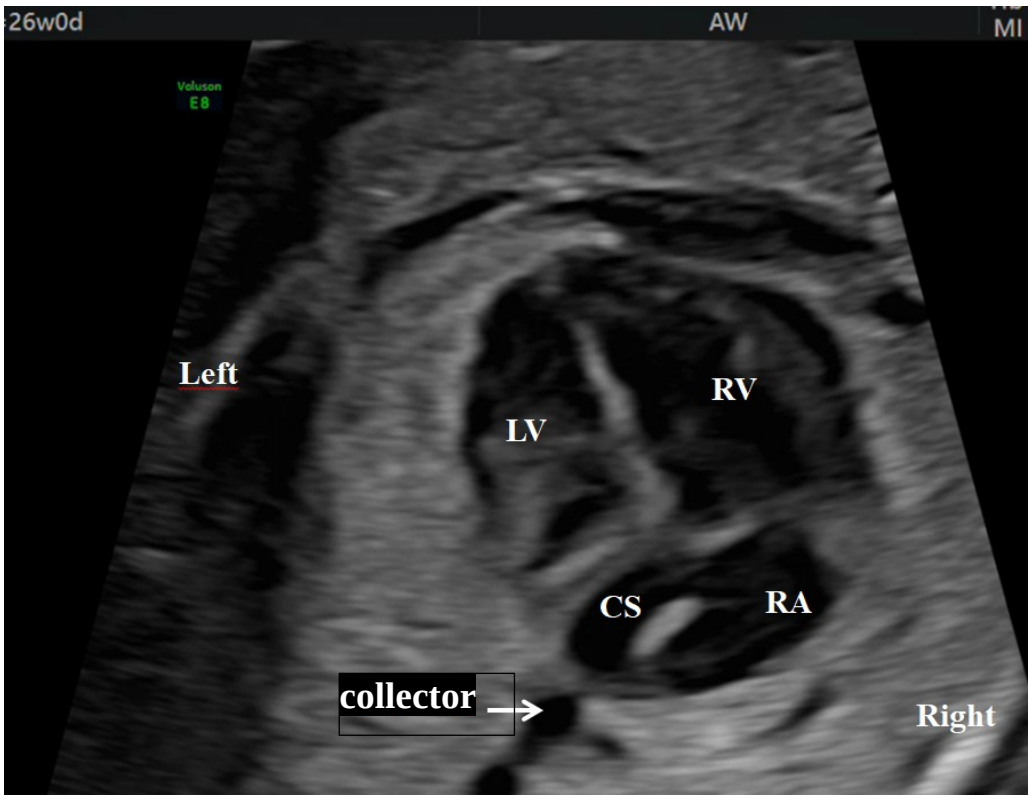
Figure 1. A. Four chamber view at 26 weeks of gestation; the pulmonary veins join a collector visible behind the posterior wall of the left atrium; **B.** Enlarged coronary sinus at 26 weeks of gestation; **C.** 26 weeks of gestation; colour Doppler mode; unrestricted blood flow through the collector; **D.** Postnatal suprasternal view: right and left pulmonary veins draining to pulmonary vein collector. See arrows; **E.** Postnatal modified 4-chamber view: enlarged right atrium (RA) and right ventricle (RV), small left atrium (LA), compressed left ventricle (LV), CS indicates the point where pulmonary collector connects to coronary sinus, the red arrow shows atrial septal defect position (ASD); Ao — aorta; CS — coronary sinus; PV — pulmonary vein

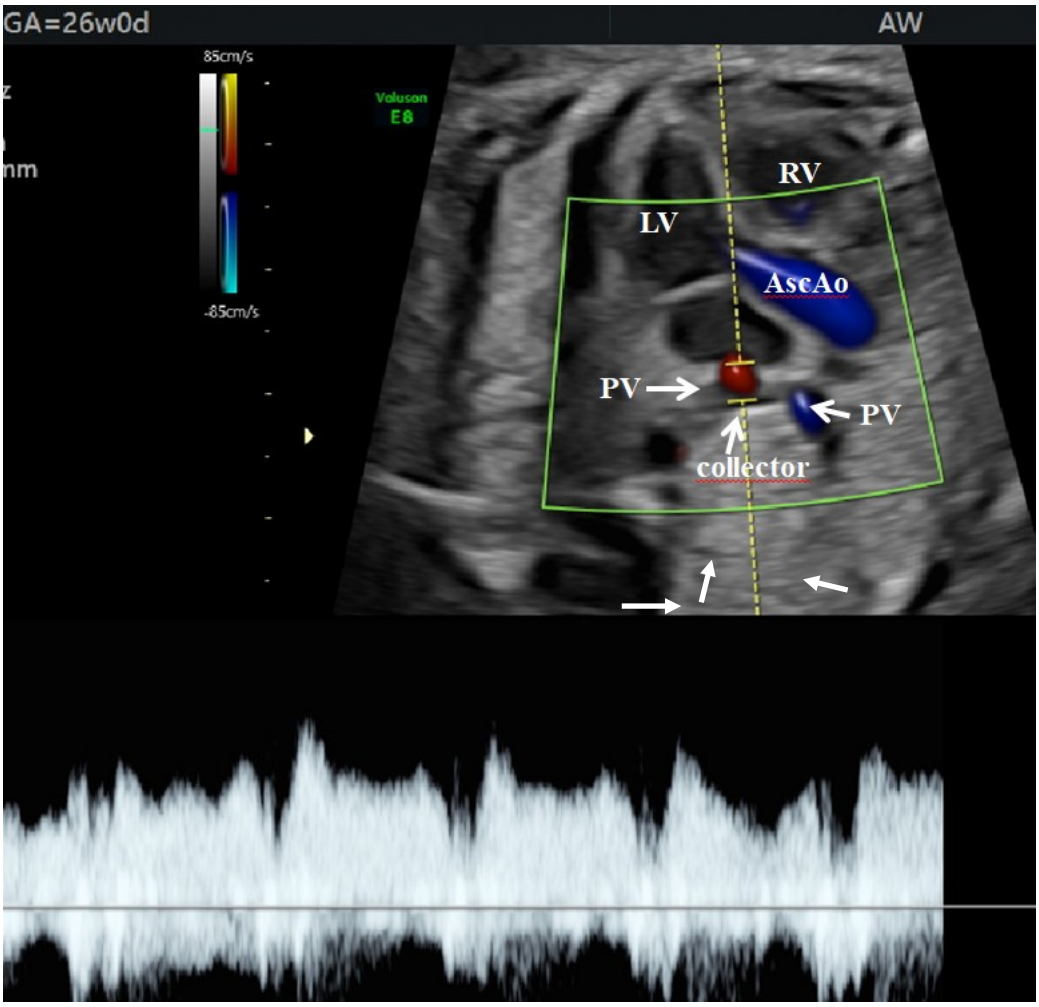
A



B


26w0d
AW
MI
Voluson
E8
Left
RV
LV
CS
RA
collector
Right

C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AW
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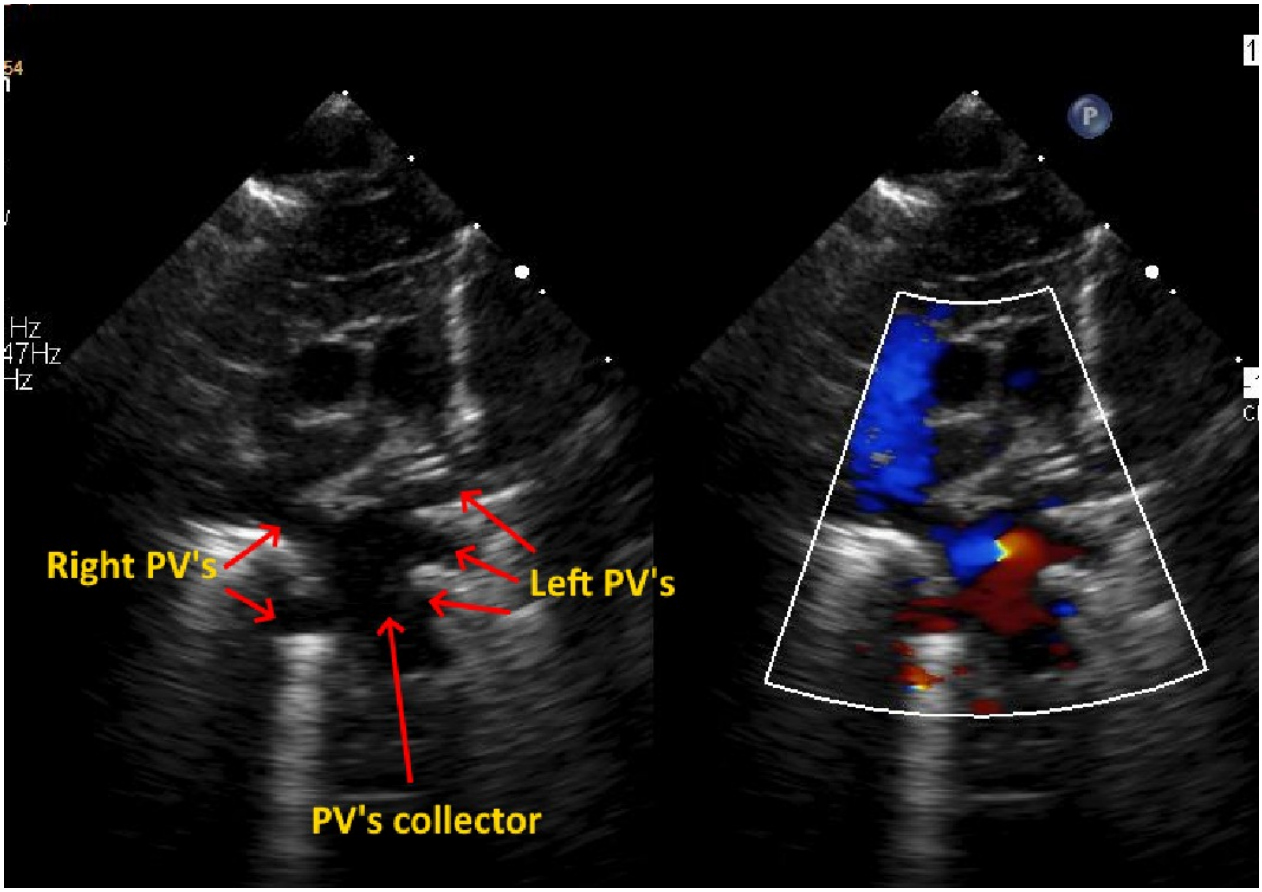
Voluson
E8
RV
LV
AscAo
PV
PV
collector

D



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