Early prenatal diagnosis of right ventricular myocardial sinusoidal-coronary artery connections in pulmonary atresia with intact septum

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A 27-year-old nulliparous woman was referred at 13 weeks' gestation for suspected congenital heart defect (CHD). Two-dimensional ultrasound imaging of the four-chamber view by transvaginal scan revealed a small right ventricle (RV). No antegrade flow could be detected across the pulmonary valve (Fig. 1a).

The patient was asked to return 5 weeks later to review the fetal heart. At 18 week' gestation fetal echocardiogram confirmed hypoplastic RV (length RV/left ventricle 0.4/1 mm). Color Doppler showed no tricuspid regurgitation and turbulent flow at the apex of the RV. From this site a vessel with turbulent flow continued along the RV external wall (Fig. 1b) toward the ascending aorta. This finding, suspicious of a ventriculocoronary fistula between the RV and the right coronary artery, was proved to show typical bidirectional flow (V_{peak} systole 100 cm/s and V_{peak} diastole 80 cm/s) by pulsed Doppler. No antegrade flow could be detected across the pulmonary valve. This, together with retrograde flow from the ductus arteriosus into the pulmonary trunk, lead to the diagnosis of pulmonary atresia with intact ventricular septum (PAIVS) and coronary sinusoids.

Proper counseling was carried out on the clinical and prognostic impact of sinusoids in PAIVS, including the possible correction to univentricular heart physiology. The option of termination of the pregnancy was mentioned. The mother decided to continue the pregnancy.

Proper counseling was carried out relatively on the pathophysiology of the congenital heart disease and on the clinical and prognostic impact of sinusoids. The option of termination of the pregnancy was discussed but the couple declined.

At 21 weeks, the use of the 4D STIC (spatiotemporal image correlation) acquisition in combination with HD-Flow (Voluson E10, GE) allowed us to obtain a better visualization of the aforementioned vessel, connecting the aorta with the RV (Fig. 1c and d; Video clip 1 as Supplementary material, http://links.lww.com/JCM/A260). Delivery was planned at 39 weeks of gestation in a third-level center with pediatric heart surgery where postnatally, the diagnosis of PAIVS and ventriculocoronary fistula was confirmed (Video clip 2 as Supplementary material, http://links.lww.com/JCM/A261).

The PAIVS is a rare CHD accounting for less than 1% of total heart defects with a prevalence of 0.0083 per 1000 live births.[1]

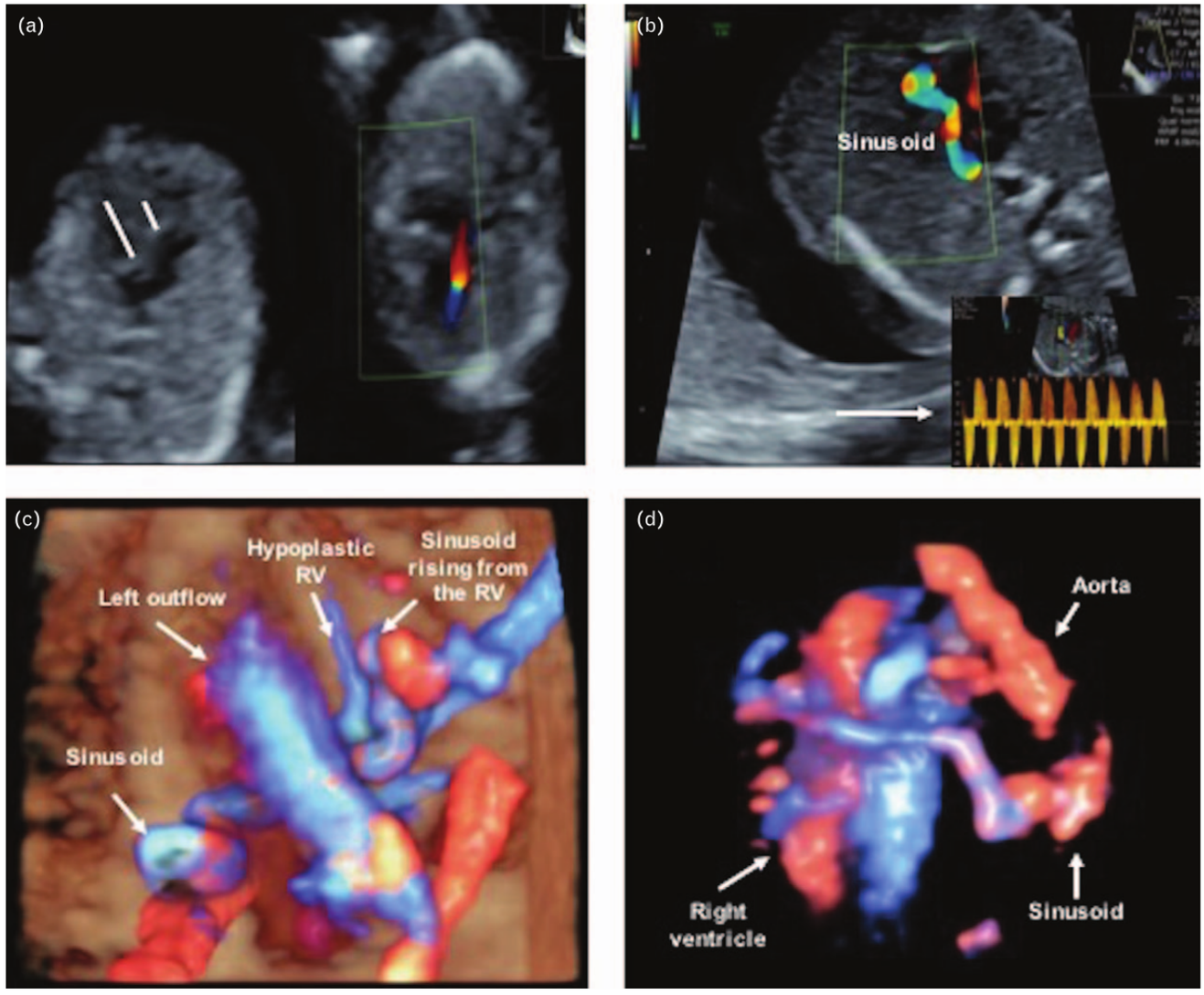
It is characterized by variable degrees of RV hypoplasia with the possibility of persistence of primitive ventriculocoronary connection (VCC), potentially detectable prenatally.[2]

The RV is hypertensive due to lack of blood egress. Consequently, the RV develops abnormal connections with the epicardial coronary arteries, which contribute to decompress the ventricle. These abnormal connections lead to progressive stenosis of the coronary arteries related to high-velocity blood flowing through abnormal connections. Due to progressive stenosis of the coronary arteries over time, the perfusion of some parts of the myocardium depends on the RV, the so-called RV dependent coronary circulation (RVDCC).

The early diagnosis of VCC in PAIVS provides valuable information for postnatal management. In fact, the surgical management of a newborn with PAIVS radically changes if a VCC is present[3]: single ventricular repair (bidirectional Glenn followed by Fontan) should be a consideration in patients with severe RV hypoplasia and patients with RVDCC.

In our case, the baby underwent univentricular palliation.

Fig. 1



The picture is showing the evolution of the case from the first to the second trimester. (a) At 13 weeks, size discrepancy between the left ventricle (4.51 mm) and the right ventricle (1.42 mm); at color Doppler, absent flow through the tricuspid valve and absent filling of the right ventricle were seen; (b) at 18 weeks, a large vessel (maximum diameter 1.4 mm) surrounding the ventricle was seen compatible with a ventriculocoronary fistula in pulmonary atresia with intact ventricular septum; bidirectional flow was seen at pulsed Doppler analysis (white arrow); (c and d) at 21 weeks, the use of 4D spatiotemporal image correlation acquisition in combination with HD-Flow Render mode allowed us to depict the course of the vessel originating posteriorly from the right ventricle, surrounding the left ventricle and terminating in the aorta.

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Conflicts of interest

None declared.

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