

CONGENITAL MINI-FOCUS ISSUE

ADVANCED

CASE REPORT: CLINICAL CASE

Pulmonary Atresia With Ventriculocoronary Arterial Connections and a Large Conoventricular Septal Defect



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ABSTRACT

Ventriculocoronary arterial connections are typically found in patients with pulmonary atresia with an intact ventricular septum. This report describes a case of ventriculocoronary arterial connections in a patient with pulmonary atresia with a ventricular septal defect. Our case supports recent data suggesting a primary coronary artery developmental anomaly in pulmonary atresia. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2019;1:545–8) © 2019 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

HISTORY OF PRESENTATION

A full-term infant, gestational age 39 weeks, with a prenatal diagnosis of tricuspid atresia was born by elective cesarean section to a gravida 3, para 1, abortus 1 mother who had pre-eclampsia. The infant's birth weight was 3.5 kg, her height was 50 cm, her head circumference was 48 cm, and her Apgar scores at 5 and 10 min were 10. No clinical feature suggesting a genetic syndrome was observed. Her vital signs were normal, with a heart rate of 163 beats/min, blood pressure of 75/42 mm Hg (mean 53 mm Hg, and

oxygen saturation on room air of 98%. On physical examination, the baby was acyanotic, without dyspnea. The pulses were normal and equally palpable. Lung fields were clear, and precordial auscultation noted a normal first heart sound and a single second heart sound with a continuous murmur at the upper left sternal border. Chest radiography showed levocardia with a normal heart size and decreased pulmonary vascular markings, with dark lung fields. The electrocardiogram revealed sinus rhythm, a heart rate of 163 beats/min, and right atrial and left ventricular hypertrophy, with a normal QRS complex axis of 80°.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis included cyanotic congenital heart defects with a normal heart size, including tetralogy of Fallot, tetralogy of Fallot with absent pulmonary valve, pulmonary atresia (PA) and ventricular septal defect (VSD), tricuspid atresia, and single ventricle with pulmonary stenosis.

LEARNING OBJECTIVES

- Ventriculocoronary arterial connections are possible in pulmonary atresia with ventricular septal defect.
- Ventriculocoronary arterial connections have specific embryological origins.

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Informed consent was obtained for this case.

Manuscript received April 12, 2019; revised manuscript received July 25, 2019, accepted September 11, 2019.

**ABBREVIATIONS
AND ACRONYMS****PA-IVS** = pulmonary atresia
with intact ventricular septum**PA-VSD** = pulmonary atresia
with ventricular septal defect**VCAC** = ventriculocoronary
arterial connection**VSD** = ventricular septal defect**INVESTIGATIONS**

Echocardiography showed situs solitus, levocardia, and normal systemic and pulmonary venous return. The infant also had atrioventricular concordance and a large ostium secundum atrial septal defect 5.13 mm with bidirectional shunt, as well as a unipartite right ventricle with a hypoplastic tricuspid valve (size, 5.05 mm [Z-score, -4.96])

([Figure 1](#), [Video 1](#)). The left ventricle and mitral valve were normal. Ventriculoarterial concordance, muscular PA, aorta dextroposition, and a large anterosuperiorly malaligned conoventricular septal defect of 4.81 mm, with bidirectional shunting, were also observed. Confluence of the central pulmonary arteries could not be established. The pulmonary artery branches were of normal size (right pulmonary artery, 3.69 mm [Z-score, -1.47]; and left pulmonary artery, 3.83 mm [Z-score, -0.79]), with blood supply from a tortuous and large vertical patent ductus arteriosus. Only 1 coronary ostium was seen in the left coronary sinus, with a branch taking an unusual course anterior and right to the aorta ([Figure 2](#), [Video 2](#)). Color Doppler imaging revealed ventriculocoronary arterial connections (VCACs) with a retrograde flow in systole

and normal antegrade flow in diastole ([Figure 3](#), [Video 3](#)). The patient had a left aortic arch and no aortic coarctation.

MANAGEMENT

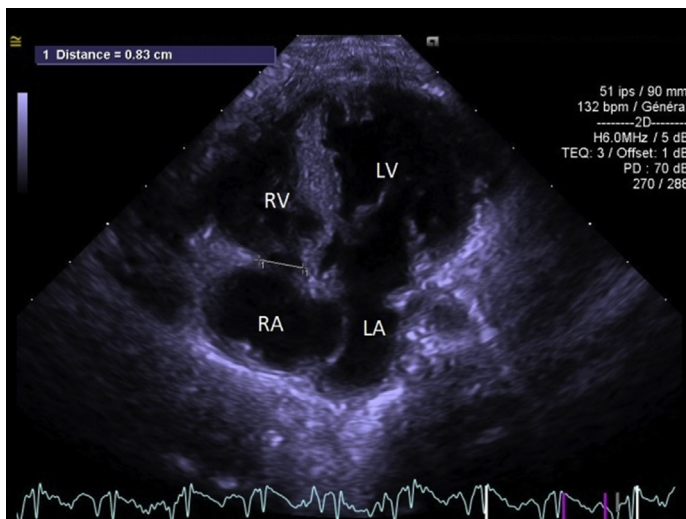
Prostaglandin infusion was administered and was gradually reduced to a minimal dose of $0.01 \mu\text{g}/\text{kg}/\text{min}$. The patent ductus arteriosus remained large, with a saturation $>90\%$, along with clinical and biological signs of heart failure, thus advocating the use of loop diuretic agents. To decide on further management, we completed the morphological evaluation with multislice computed tomography, which confirmed the single coronary artery anatomy with VCACs, as well as confluence of the central pulmonary artery ([Figure 4](#), [Video 4](#)).

DISCUSSION

PA with VSD (PA-VSD) and PA without VSD are 2 cyanotic congenital heart diseases with different origins and pathogenesis. PA with intact ventricular septum (PA-IVS) is characterized by heterogeneous right ventricular development, an imperforate pulmonary valve, and possible extensive VCACs. The pulmonary arteries are usually confluent and supplied by a left-sided arterial duct (1). Conversely, PA-VSD is characterized by an anterosuperiorly malalignment conoventricular septal defect with a spectrum of obstruction at the pulmonary valve level from imperforate pulmonary valve to total absence of the pulmonary trunk. The right ventricle is of normal size, and the central pulmonary arteries are usually hypoplastic or absent, with congenital systemic collateral arteries (2). To the authors' knowledge, VCAC has not been reported in a patient with PA-VSD.

VCACs are connections between the ventricular intertrabecular spaces and the main coronary arteries through thick-walled vascular communications. They are typically found in PA-IVS with gross underdevelopment of the right ventricle and have also been observed in various congenital heart diseases with hypoplastic right and left heart but always in the setting of an intact ventricular septum (3). Previously, it was assumed that atresia of the pulmonary valve was the primary morphogenic event leading to right ventricular hypertension and development of the fistulous communication with the coronary arteries. The development of VCACs has been considered secondary to hemodynamic alterations caused by the combination of tricuspid valve competence and PA.

FIGURE 1 Echocardiographic Apical 4-Chamber View Showing the Hypoplastic Right Ventricle



The right ventricle is unipartite and the tricuspid valve annulus is hypoplastic. See [Video 1](#).
LA = left atrium; LV = left ventricle; RA = right atrium; RV = right ventricle.

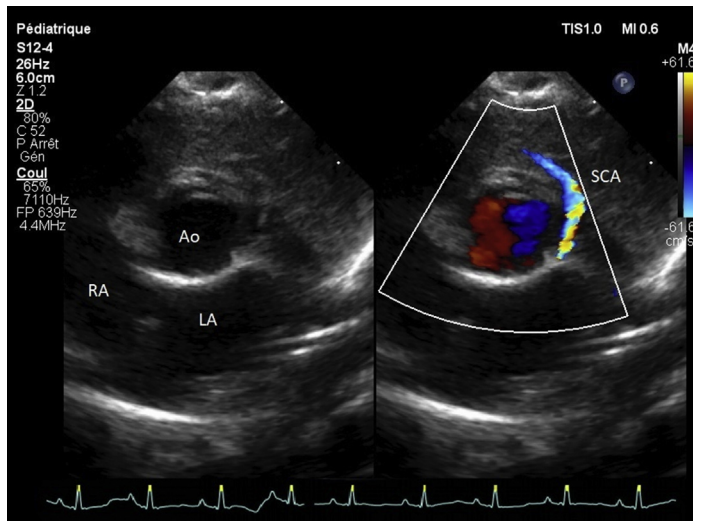
On the basis of increased ventricular pressure, the persistence of embryonic ventriculocoronary communications has been postulated to be the cause of VCACs (4). Blood flow would be preferential through the VCACs and not through the pulmonary valve, thereby promoting development of PA during gestation. This hypothesis may not be true, however, because clinical and pathological data have revealed the existence of VCACs during fetal life before PA was established (5). The coexistence of VCACs and pulmonary stenosis has also been reported (6). Our case of VCACs in a patient with a VSD adds clinical evidence supporting the hypothesis of a primary coronary developmental anomaly that may contribute to the formation of VCACs in some patients with PA-IVS.

Experimental data showed that coronary vascular development derives from the epicardium and is not dependent on outflow tract septation. Inhibiting epicardial formation by mechanical intervention and molecular disruption in avian and mouse models resulted in abnormal coronary arteries with VCACs and no pulmonary stenosis (7). Embryological studies supported the notion of a different developmental insult producing PA-IVS, and investigators proposed a posterior second heart field abnormality as a possible cause of PA-IVS with VCACs, as opposed to a primarily anterior second heart field-directed outflow tract septation abnormality for PA-IVS without VCACs. The same investigators suggested a progression to valvular PA resulting from a VCAC blood steal phenomenon (8).

Recent histopathological studies are still stirring the debate by stating that VCACs are, in fact, the already existing vessels of Wearn. The arterioluminal vessels of Wearn originate from the main trunks of coronary arteries and from the subendocardial arteries, and they deliver blood from the coronary arteries to the atria and ventricles. As they progress distally, these vessels may give off capillary branches and may lose their arterial characteristics, thus becoming a simple endothelial-lined tube. PA-IVS with and without coronary arteriopathy would be manifestations of a continual disease spectrum (9).

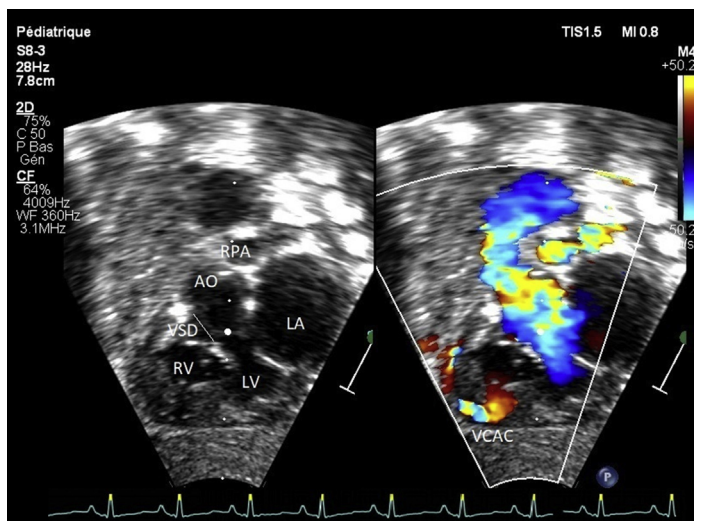
According to these embryological theories, prenatal diagnosis of PA-VSD with a hypoplastic right ventricle should not rule out associated VCACs. Therefore, when severe right ventricular hypoplasia exists and precludes biventricular circulation, physicians can provide specific information on prognosis of a single ventricle with high early mortality associated with VCACs and discuss whether to provide intensive

FIGURE 2 Echocardiographic Left Parasternal Short-Axis View in Doppler Color Mode Showing the Single Coronary Artery

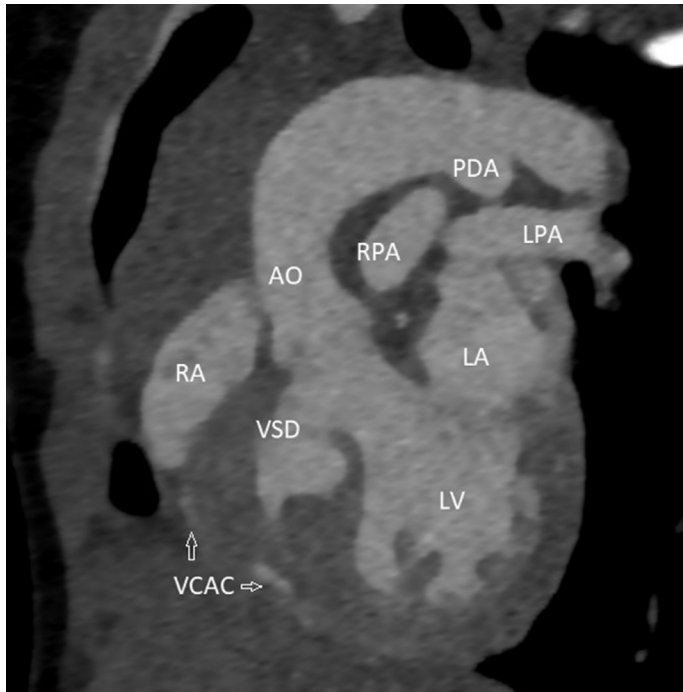


Coronary flow is retrograde in systole. See [Video 2](#). AO = aorta; SCA = single coronary artery; other abbreviations as in [Figure 1](#).

FIGURE 3 Echocardiographic Subcostal Short-Axis View in Doppler Color Mode Showing the VCAC Blood Steal



During the systole, the flow is going from the right ventricle to the ventriculocoronary arterial connections (VCAC). See [Video 3](#). RPA = right pulmonary artery; VSD = ventricular septal defect; other abbreviations as in [Figures 1 and 2](#).

FIGURE 4 Multislice Computed Tomography in a Modified Sagittal View Showing the VCACs

Ventriculocoronary arterial connections (VCAC) arise from the hypoplastic right ventricle associated with a large conoventricular septal defect. See [Video 4](#). LPA = left pulmonary artery; PDA = patent ductus arteriosus; other abbreviations as in [Figures 1 to 3](#).

care, palliative care, or termination of pregnancy (10). In addition, fetal cardiac interventions to relieve pulmonary stenosis with an intact ventricular septum or PA-IVS may improve hemodynamics but may not alter the primary development of VCACs.

FOLLOW-UP

A Blalock-Thomas-Taussig shunt procedure was performed on day 7. At 5 months, her average saturation decreased to <75%, with ductal-associated pulmonary artery coarctation, thus prompting the Glenn procedure with central pulmonary artery repair as a first step in a univentricular palliation program.

CONCLUSIONS

PA-VSD includes a wide spectrum of disease with great morphological heterogeneity. Our case supports recent data suggesting a primary coronary developmental anomaly in PA, rather than the persistence of embryonic VCACs because of elevated right ventricular pressure.

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KEY WORDS congenital heart defect, coronary circulation, coronary vessel anomaly, pulmonary atresia, right ventricle, ventricular septal defect

APPENDIX For supplemental videos, please see the online version of this paper.