

# MDCT evaluation of intramyocardial-sinusoids-coronary artery communications in a neonate with pulmonary atresia and intact ventricular septum

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## Abstract

A patient of tetralogy of Fallot with complete atresia of the pulmonary outflow tract with ventriculocoronary connections is presented. MDCT imaging revealed left coronary sinus, with a large fistula draining into the free wall of hypoplastic right ventricular cavity with tortuous channel arising from right ventricular outflow, and communicating with proximal limb of the fistula forming a complete loop suggesting a right ventricle-to-left coronary sinus sinusoid.

**Key words:** Intramyocardial sinusoids; multidetector computed tomography; pulmonary atresia

## Introduction

The right ventricular coronary connections in a patient with pulmonary atresia with intact ventricular septum is a rare presentation. A patient of Tetralogy of Fallot with complete atresia of the pulmonary outflow tract with ventricular coronary connections is presented. Multidetector computed tomography (MDCT) provides valuable information on the status of the obstructed right ventricle with ventricular coronary connections as well as coronary artery (CA) stenosis or atresia.

## Case History

A full-term 5-day-old boy developed cyanosis and oxygen desaturation at 20 hours. Chest radiography revealed

oligemic lungs, and ECG showed tall, bifid *P* waves with features of right ventricular hypertrophy. Clinical examination showed a stable active baby, saturating above 85%, with a continuous murmur on the upper left sternal border. Echocardiography revealed pulmonary atresia, hypoplastic right ventricle with intact interventricular septum, suspicious left coronary artery fistula (CAF), secundum atrial septal defect (ASD) with right to left shunt and small persistent ductus arteriosus (PDA) filling confluent pulmonary artery branches. He was started on prostaglandin E infusion at 0.05 mcg/kg/min. MDCT examination was carried out for the evaluation

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of proposed coronary fistula and pulmonary vascular anatomy. A prospective gated 64-slice MDCT performed with iodinated contrast revealed the coronary fistula [Figure 1] and origin of confluent pulmonary arteries from patent ductus arteriosus [Figure 2A]. The right pulmonary artery was hypoplastic with a high-grade narrowing at the beginning [Figure 2B]. CT imaging revealed left coronary sinus with a large fistula draining into the free wall of hypoplastic right ventricular cavity [Figures 2C and D]. Another tortuous channel was noted arising from the right ventricular outflow and communicating with proximal limb of the fistula forming a complete loop suggesting a right ventricle to left coronary sinus sinusoid [Figures 2C and D]. Filling of left and right CA was noted to originate from distal and proximal limb of the sinusoid [Figures 3A and B], respectively. Multiple small sinusoids were noted emerging from the right ventricle along [Figures 3C and D] with the creation of unnamed coronary branches.

Cardiac catheterization confirmed the CT findings [Figures 4A and B] with suprasystemic pressure in the right ventricle. No CA branches were noted to originate from aorta. The patient was put on short-term indomethacin therapy, was transported to the cardiac intensive care unit, and was planned for future corrective surgery.

## Discussion

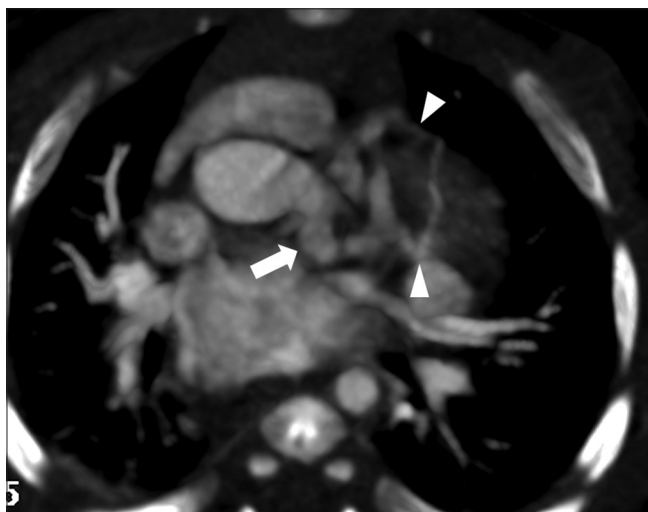
This case can be referred to as right-to-left shunt occurring in a case of pulmonary atresia with intact ventricular septum and subsequent development of intramyocardial-sinusoids-CA communication.<sup>[1]</sup> Significant variation of coronary supply may be noted in similar patients, and the choice of right ventricular decompensation (RVD) depends on proper

CA support to the left ventricle.<sup>[2,3]</sup> In patients with RV to CAFs without coronary stenosis, RVD could result in a right ventricular “steal” phenomenon from the aorta into the RV during diastole. Whereas, in patients with RV to CAFs with coronary stenosis, RVD could lead to a right ventricular steal if the stenosis is distal to the fistulas and steal and/or ischemia if the stenosis is proximal to the fistulas.<sup>[2,3]</sup>

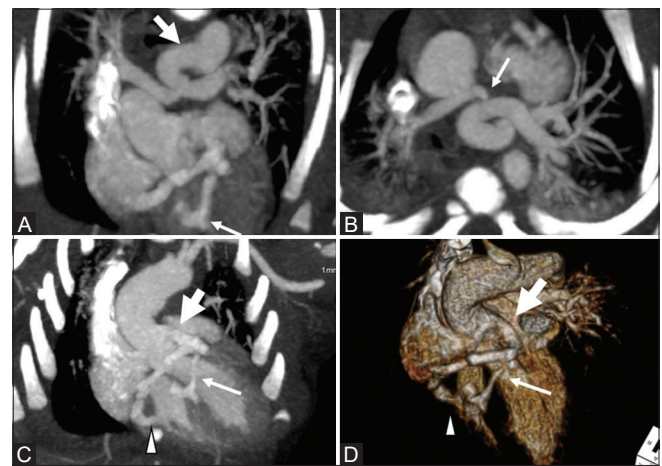
The coronary arteries in these patients may directly originate from intramyocardial sinusoids without any communication with aorta or pulmonary artery.<sup>[2,3]</sup> A thick-walled right ventricular cavity with suprasystemic pressure is known to initiate the formation of intertrabecular spaces and sinusoids of embryonic blood bed, which eventually connects with coronary vascular bed and form anastomosis between it and the ventricular lumen.<sup>[4]</sup> The same pathogenesis may have contributed to the creation of several intramyocardial-sinusoids-CA branches in our case.

A CAF is an abnormal connection that directly connects one or more coronary arteries to a heart chamber or to major thoracic vessels without an interposed capillary bed. CAF that arises from a CA and then terminates into a chamber of the heart is known as coronary cameral fistulas as seen in our case.<sup>[5]</sup> About 60% of CAFs originate from the right CA and drain into the right-sided heart chamber or great vessel as seen in our case. CAF draining into the left heart chambers is very rare.<sup>[5]</sup>

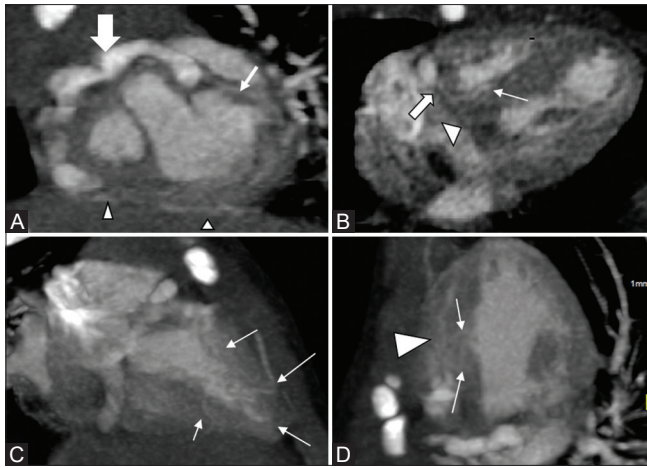
The majority of CAFs are congenital in origin and complications do not occur until after the age of 20 years.<sup>[6,7]</sup>



**Figure 1:** Axial maximum intensity projection (MIP) image of multidetector computed tomography (MDCT) (A) shows communication of the fistula with left coronary sinus (thick arrow) and forming a complete loop with the distal limb of the fistula surrounding the right ventricle (arrowheads)



**Figure 2 (A-D):** Coronal, axial MIP, and volume rendering image of MDCT (A) shows confluent pulmonary artery originating from a large persistent ductus arteriosus (PDA) (thick arrow). Hypoplastic right ventricle with the fistula (arrow). High-grade narrowing noted (B) at the origin of the right pulmonary artery (arrow). 2D CT image (C) and 3D CT image (D) show communication of the fistula with left coronary sinus (thick arrow) and entering free wall of the right ventricle (arrowhead). The distal limb of fistula is seen to originate from the right ventricle outflow (arrow) to form a complete loop. No other coronary ostium was identified from the aortic sinus

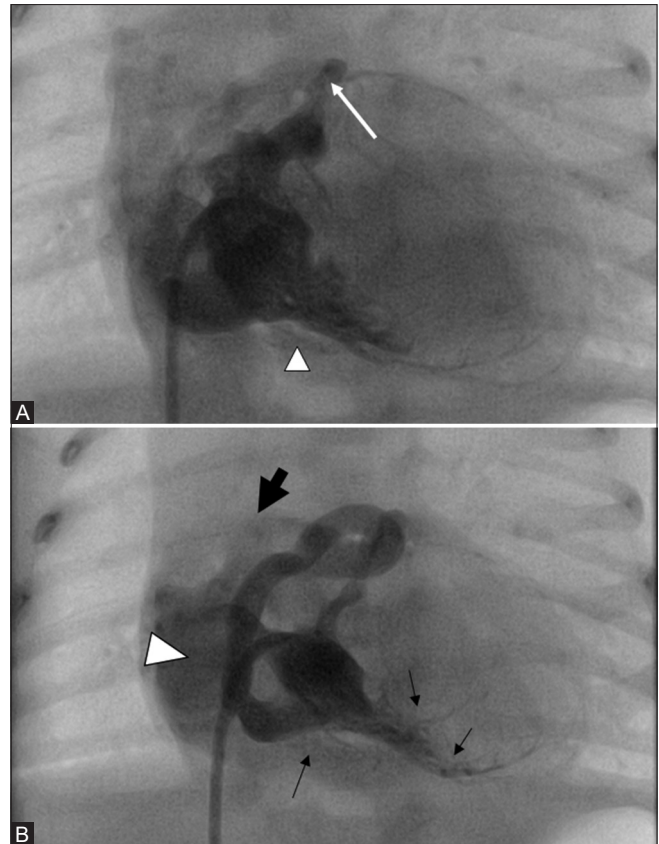


**Figure 3 (A-D):** 2D MDCT image shows (A) origin of left coronary (arrow) and right coronary (arrowheads) artery from the proximal and distal limb of fistula (thick arrow), (B) origin of right coronary artery from distal limb of the fistula (thick arrow) and giving posterior descending artery (arrow) and posterior left ventricular branches (arrowhead), (C) multiple sinusoids are seen to arise from right ventricle coursing to form unnamed coronary arteries (arrows), (D) two of the large sinusoids (arrows) arising from right ventricle (arrowhead) and coursing through the interventricular septum toward left ventricle

Although the majority are asymptomatic, some may present with myocardial ischemia, aortic insufficiency, and sudden death.<sup>[8,9]</sup> Arrhythmias and congestive heart failure can also occur due to left-to-right shunt and volume overload. Around 20% of patients with a CAF may be associated with other congenital heart anomalies such as Tetralogy of Fallot, aortic atresia, pulmonary atresia, atrial and ventricular septal defects, and patent arterial duct.<sup>[5]</sup>

CAF are mostly treated conservatively with serial follow-up.<sup>[5]</sup> In a limited number of cases, these CAF may close spontaneously.<sup>[5,10]</sup> Earlier diagnosis with proper management can prevent cardiac complications. The management of CAF is carried out either with surgical correction or transcatheter occlusion.<sup>[11]</sup> Typically, direct ligation of the CAF at the drainage site is preferred because it should eliminate the possibility of myocardial ischemia.<sup>[10,11]</sup> In cases with compromised blood flow to the myocardium, grafting of the involved distal CA is suggested.

Management of pulmonary atresia with intact ventricular septum is challenging because of the wide anatomic variations. Systemic-to-pulmonary shunt, bidirectional Glenn shunt, closed pulmonary valvotomy with the Blalock–Taussig (BT) shunt, transvalvular right ventricular outflow tract reconstruction with BT shunt and catheter valvotomy are used for the RV decompression at the initial stage,<sup>[12]</sup> after initial intervention, definitive surgeries including biventricular, 1.5 ventricular, or Fontan procedures are performed, respectively, based on RV development and the approaches of the initial intervention.<sup>[12]</sup> Zheng *et al.* reported the survival rates of 93.7 and 88.2% as mid-term



**Figure 4 (A and B):** Right ventricular angiogram shows (A) origin of the left coronary (arrow) and right coronary (arrowhead) artery from proximal and distal limb of the fistula (thick arrow). The proximal (arrowhead) and distal loop of the fistula forms a complete loop and finally enters the left coronary sinus

outcomes in one-stage surgery group and staged surgery group, respectively, without statistical difference, which is comparable with other studies.<sup>[12]</sup> In addition, a relatively low number of deaths were observed among patients with three types of definitive repair, which indicated that selecting an appropriate surgical strategy depending on the degree of RV hypoplasia and the patients' age could be achieved with a low mortality rate.<sup>[12]</sup>

In conclusion, MDCT evaluation of the intramyocardial-sinusoids-CA communications is possible: this approach provides accurate information for planning, navigation, and more future non-invasive assessment of the sinusoids and may reduce the dependency on invasive contrast angiography.

#### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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### Conflicts of interest

There are no conflicts of interest.

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