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Perioperative & Critical Care: Case Report

Pulmonary Atresia Intact Ventricular Septum With Anomalous Left Coronary Artery From Pulmonary Artery

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A patient with known pulmonary atresia and intact ventricular septum and ductal stent presented with low cardiac output and arrythmia. Intraoperatively, the patient was found to have an anomalous left coronary artery arising from the pulmonary artery. After reimplantation of the left coronary artery to the aortic root and placement of a central shunt, the patient progressed well and was discharged home. A high index of suspicion and clear diagnostic visualization of both right and left coronary artery origins are needed to diagnose this highly fatal yet treatable lesion properly.

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Pulmonary atresia with intact ventricular septum (PA-IVS) is an uncommon congenital heart disease with an incidence of 4 to 8 of 100,000 live births.[1,2] An anomalous origin of a coronary artery from the pulmonary trunk has been mentioned to be associated with this condition.[3] We describe a case of PA-IVS in a patient with an incidental diagnosis of anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) who survived to discharge.

The patient was born at the referring quaternary care hospital at 37 weeks of gestation; the infant weighed 3000 g and had respiratory failure. The patient was given a postnatal diagnosis of PA-IVS with right ventricular (RV)-dependent coronary circulation because there was only a right aorto-coronary connection from the aortic root and evidence of RV sinusoids on an echocardiogram. At 10 days of life, this diagnosis was confirmed by an aortic angiogram at the time of cardiac catheterization, and a patent ductus arteriosus (PDA) stent was placed. A week after stent placement, the patient experienced tachycardia and ST-segment changes with progressively rising troponin levels concerning for myocardial ischemia. An esmolol drip was started, with suboptimal correction of the rhythm and ischemia. The echocardiogram reported to show mildly depressed left ventricular dysfunction. The family was recommended heart transplantation as the only viable option. The parents requested transfer to our facility for a second opinion.

The echocardiogram at our facility reconfirmed membranous PA-IVS with a moderately hypoplastic tricuspid valve annulus with limited inflow, a moderately hypoplastic right ventricle, and a normal right coronary artery origin. No left main coronary artery origin was initially visualized, but a large vessel branching into multiple distal channels was seen adjacent to the aortic root with to-and-fro flow, in the area of the RV outflow tract. This large vessel (later confirmed to be the dilated left main coronary artery) was presumed to be a coronary fistula from the right ventricle communicating to the left and possibly right coronary artery system (Figure 1). Given the unusual appearance of this presumed coronary artery fistula, an urgent cardiac computed tomographic (CT) angiogram was performed. Initial review of CT images showed similar findings of a dilated coronary vessel in close proximity to the aortic root, giving rise to the left anterior descending artery and likely left circumflex artery with additional fistulas communicating to the RV cavity.

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The patient was taken to the operating room to replace the PDA stent with a restricted aorto-pulmonary shunt to limit pulmonary over-circulation, which was suspected to be leading to low cardiac output and cardiac ischemia, and to attempt creating an aortocoronary connection to the left coronary system. Intraoperatively, during removal of the PDA stent at the pulmonary end, the left main coronary artery was found to be anomalously arising from the pulmonary root (Figure 2). An ALCAPA repair in the fashion of a Takeuchi tunnel was performed using autologous pericardium and the opened pulmonary root (Figure 3). A central systemic-to-pulmonary shunt was placed, and successful chest closure was performed 6 days after surgery followed by extubation. On further review of the admission imaging studies, the ALCAPA origin and severely dilated left main coronary artery bifurcating into left anterior descending and circumflex branches were clearly identifiable (Figure 2). In addition, there were several channels connecting to the proximal left main coronary and left anterior descending arteries that were likely RV-coronary sinusoids. The patient was discharged home 8 weeks post-operatively with minimal cardiac medication.

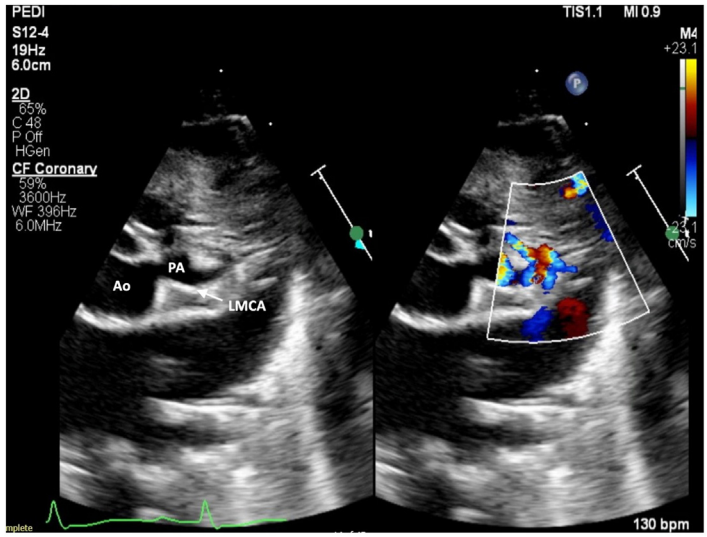

FIGURE 1 Preoperative echocardiographic parasternal short-axis images in 2 dimensions (2D) and color Doppler, showing the left main coronary artery (LMCA) arising from the pulmonary artery (PA) with turbulent to-and-fro flow on color Doppler images. There is dropout artifact between the aorta (Ao) and the pulmonary artery. (bpm, beats per minute.)

COMMENT

ALCAPA is a rare congenital cardiac condition that occurs in approximately 1 of 300,000 live births or 0.5% of children with congenital heart defects.[4-6] It carries a high mortality during infancy without timely intervention.[5,6] These patients often present in cardiogenic shock, heart failure, or arrythmias secondary to myocardial ischemia. Because pulmonary vascular resistance decreases after birth, it causes coronary steal leading to intermittent hypoperfusion of cardiac tissue. Our patient with PA-IVS received multiple detailed cardiac diagnostic tests, including echocardiography, cardiac catheterization angiography, and cardiac CT angiography; despite the multitude of diagnostic imaging at multiple hospitals, the diagnosis of ALCAPA was missed preoperatively. The exceedingly rare association of PA-IVS and ALCAPA, coupled with the complex anatomy of the RV-coronary sinusoids and mistaken presumption of the dilated left main coronary artery as a fistulous vessel or a large sinusoid, all contributed to the missed finding of ALCAPA until its subsequent diagnosis in the operating room. Specifically, the lack of any left coronary artery origin from the aorta was mistakenly assumed to imply that the left coronary supply was exclusively from RV sinusoids, and no RV angiogram was performed to confirm this incorrect assumption. Our case report illustrates how a high index of suspicion and clear diagnostic visualization of both right and left coronary origins are needed to diagnose this highly fatal, yet treatable, lesion properly in patients with or without congenital heart disease. In this particular case, the diagnosis of ALCAPA and its subsequent repair significantly changed the treatment strategy and prognosis of this patient and obviated the need for neonatal cardiac transplantation.

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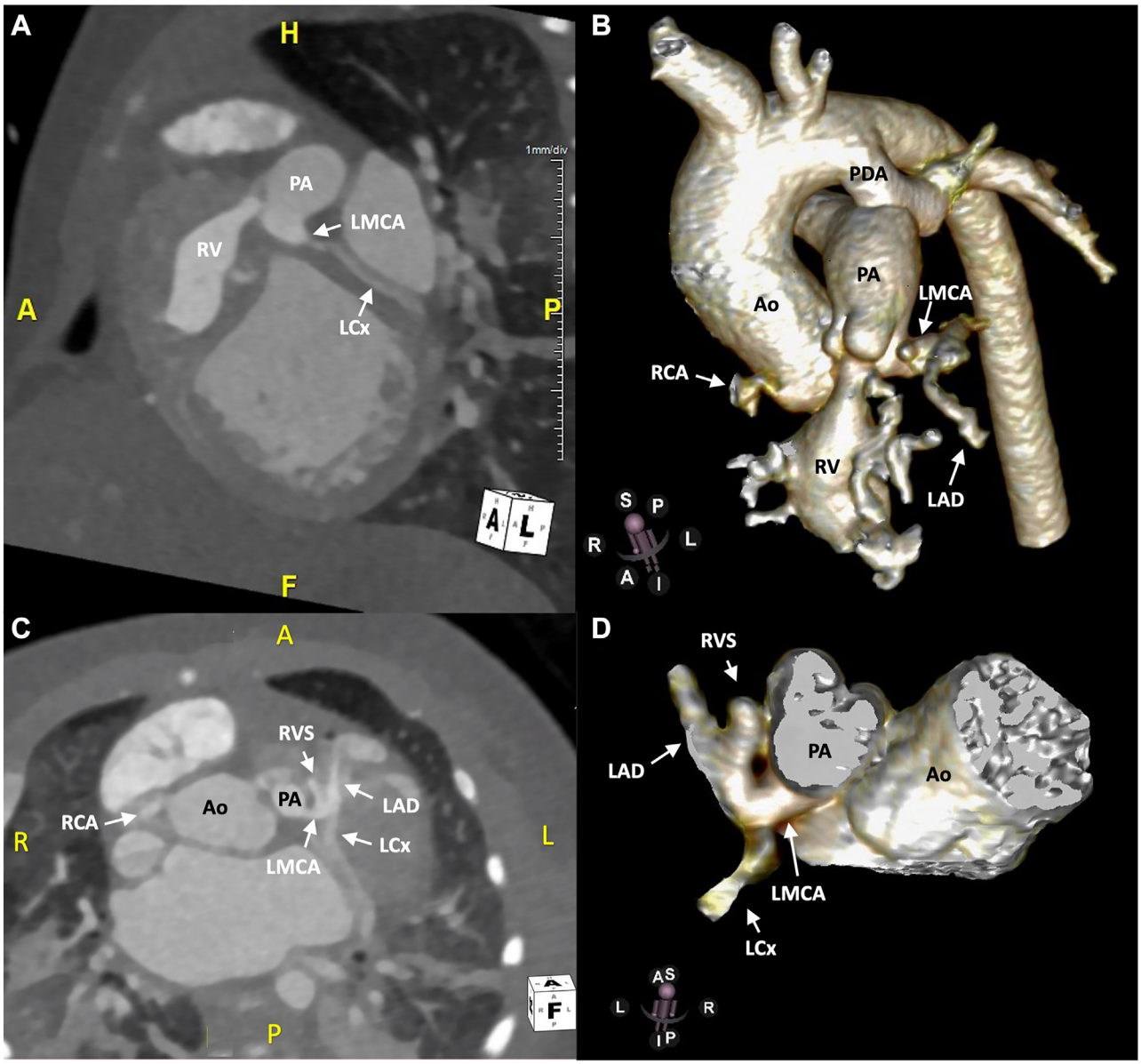

FIGURE 2 (a) Preoperative cardiac computed tomographic angiogram (CTA) with contrast enhancement gated to midsystole in the oblique sagittal view showing the hypoplastic right ventricular outflow tract with membranous pulmonary atresia and a dilated left main coronary artery (LMCA) arising from the pulmonary artery (PA), with the left circumflex (LCx) branch arising from the LMCA. (b) A 3-dimensional volume-rendered reconstruction of the cardiac CTA showing the hypoplastic right ventricle (RV) with coronary sinusoids, the dilated LMCA arising from the PA, the left anterior descending (LAD) branch arising from the LMCA, and the right coronary artery (RCA) arising normally from the aortic root. The arterial ductal stent is also seen extending into the left pulmonary artery. (c) Cardiac CTA oblique axial view showing the LMCA arising from the PA, with the LAD branch arising from the LMCA. There is also a left coronary–RV sinusoid (RVS) connecting to the proximal LMCA. (D) Modified 3-dimensional volume-rendered reconstruction from the cardiac CTA showing the dilated LMCA arising from the PA and not the Ao, and the LAD and LCx branches arising from the LMCA. (A, anterior; F, feet; H, head; I, inferior; L, left; P, posterior; PDA, patent ductus arteriosus; R, right; S, superior.)

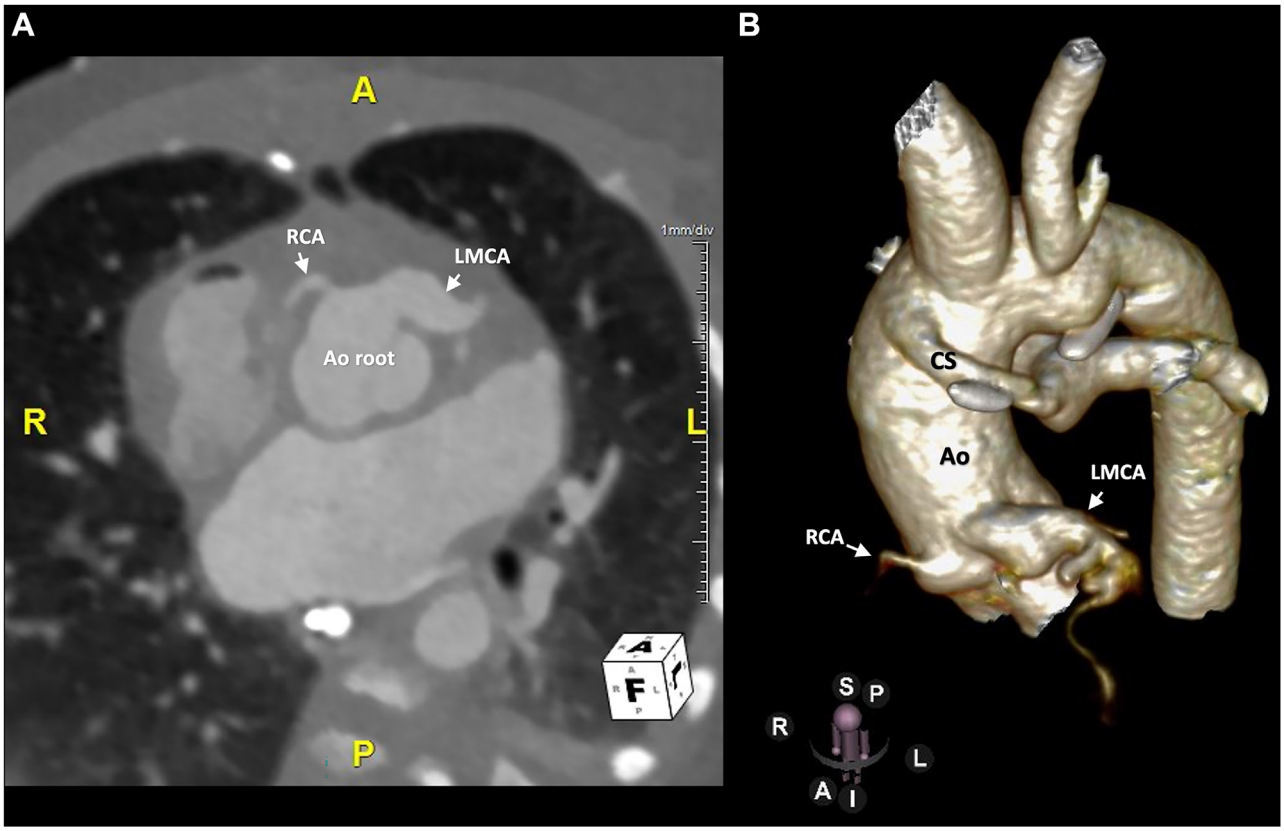

FIGURE 3 (a) Postoperative cardiac computed tomographic angiogram with contrast enhancement gated to midsystole in the oblique axial view showing the aortic root (Ao root) with the right coronary artery (RCA) and the reimplanted, dilated main left coronary artery (LMCA) arising from the right sinus of Valsalva. (b) A 3-dimensional volume-rendered reconstruction of the cardiac computed tomographic angiogram showing the RCA and the reimplanted and dilated LMCA arising from the Ao root, as well as the central shunt (CS) arising from the ascending aorta supplying the branch pulmonary arteries. (A, anterior; F, feet; H, head; I, inferior; L, left; P, posterior; R, right; S, superior.)

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