Anomalous origin of the left main coronary artery from the right pulmonary artery: a case report

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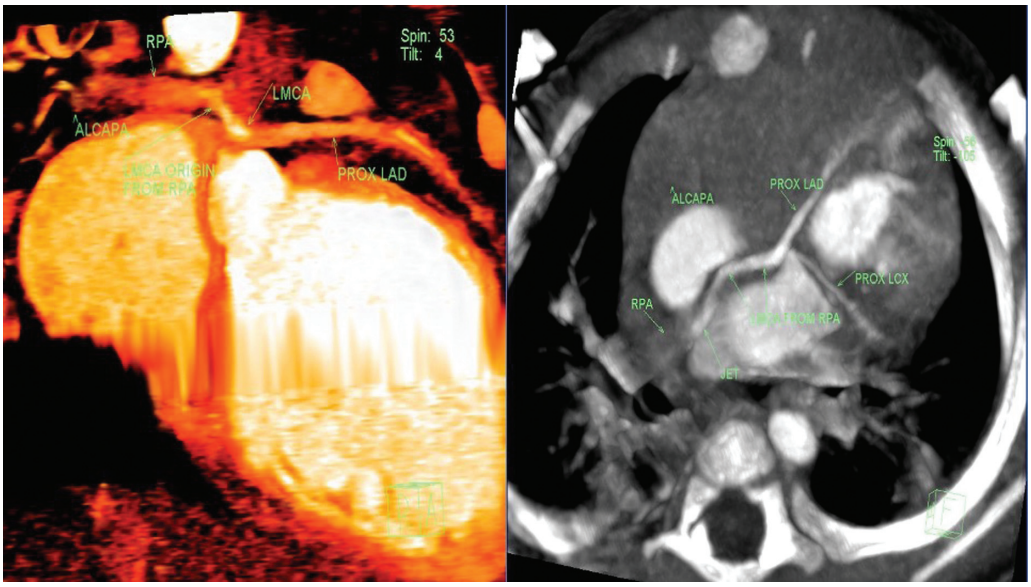
Anomalous origin of the left coronary from the right pulmonary artery (AOLCARPA) is an extremely rare anomaly, which has been reported in only a few patients worldwide [1]

An 8-month-old male patient weighing 5.7 kg came to our hospital with chief complaints of tachypnea while feeding and failure to thrive. ECG of the patient showed sinus tachycardia. The patient had a heart rate of 140/min and saturation of 99% on room air. Chest radiograph showed cardiomegaly. Transthoracic echocardiography showed moderate mitral regurgitation, dilated left atrium and ventricle, severe left ventricular (LV) dysfunction, global LV hypokinesia with an LV ejection fraction of 15–20%. Flow pattern in the left anterior descending artery and the left circumflex artery was reversed and the right coronary artery (RCA) was dilated. The left main coronary artery (LMCA) origin was not clear. As the diagnosis was unclear, computed tomography coronary angiography (CTCA) was done to delineate the coronary anatomy, which showed an anomalous origin of LMCA from the right pulmonary artery (RPA) (Figs. 1 and 2Figs 1, 2). Blood investigations including complete blood count, and liver and kidney function tests were normal. Preoperative medication of the patient included digoxin, spironolactone, furosemide, and milrinone infusion. The patient was posted for corrective repair of anomalous origin of the left coronary artery from the RPA.

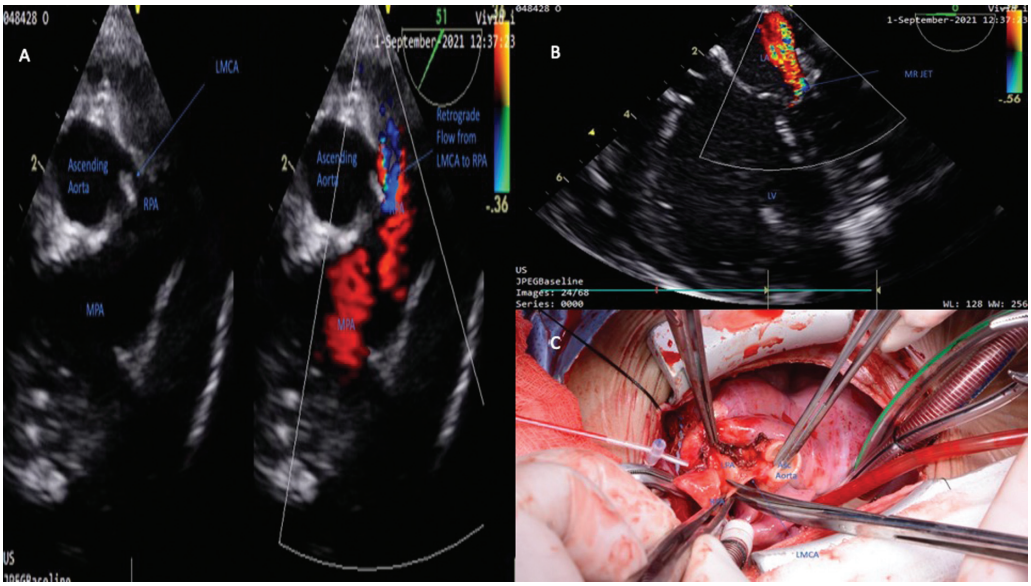
On the day of surgery, the patient was taken to the operating room. Preoperative inotropic support of intravenous infusion of milrinone 0.5 μg/kg/min was continued. After attaching a multiparameter monitor with a five-lead ECG, noninvasive blood pressure monitor, and a pulse oximeter, general anesthesia was induced with intravenous fentanyl 2 μg/kg, midazolam 0.1 mg/kg, vecuronium 0.1 mg/kg, and endotracheal intubation was done. Femoral artery was cannulated for invasive arterial blood pressure monitoring, and central venous catheter was inserted in the right internal jugular vein. Anesthesia was maintained with isoflurane and intermittent boluses of fentanyl and vecuronium. Intraoperative ventilation was instituted with the goals of avoiding hypocarbia, hyperoxia, and alkalosis. An FiO_2 of 0.35 was kept with PaO_2 targets of 80–100 mm Hg to maintain pulmonary vascular resistance End-tidal carbon dioxide ($EtCO_2$) was maintained between 40 and 42 mm Hg. Pre-bypass transesophageal echocardiography (TOE) examination was carried out with a pediatric 9T probe (Vivid I, GE; Vingmed Ultrasound, Horten, Norway). ME ascending aorta short-axis view showed the origin of LMCA from RPA (Fig. 2a). Midesophageal four-

Figure 1



Computed tomography coronary angiography showing left main coronary artery (LMCA) arising from the right pulmonary artery (RPA). PROX LAD, proximal left anterior descending artery.

Figure 2



(a) Midesophageal (ME) ascending aortic short-axis (SAX) view showing retrograde flow from left main coronary artery (LMCA) to right pulmonary artery (RPA). (b) Midesophageal (ME) four-chamber view showing dilated left ventricle (LV) and mitral regurgitation jet. (c) Origin of LMCA from RPA. LPA, left pulmonary artery; Asc Aorta, ascending aorta.

chamber view showed dilated LV and moderate mitral regurgitation (Fig. 2b). Heparin 400 IU/kg was given, and cardiopulmonary bypass was instituted.

Cardiopulmonary bypass was established using aortobicaval cannulation with looping of both pulmonary artery branches. Myocardial preservation was obtained with antegrade cold blood potassium cardioplegia into the aorta and main pulmonary artery. Under low pump flows, LMCA origin was identified originating from the RPA (Fig. 2c). Medial trap door was made in the aorta and LMCA was anastomosed to the aorta (coronary translocation). Posterior annuloplasty of the mitral valve was performed through the trans-septal approach. Weaning from the cardiopulmonary bypass was uneventful with minimal inotropic support (milrinone 0.5 µg/kg/min; Epinephrine 0.04 µg/kg/

min). The chest was kept open with a sterile membrane cover electively and the patient was shifted to the pediatric ICU with stable hemodynamics.

AOLCARPA is an extremely rare congenital coronary anomaly. Only 10–20% of neonates survive till adulthood, and it depends on the adequacy of LV perfusion through collaterals to LCA from RCA and the proportion of coronary steal from the LCA to pulmonary artery [2] Coronary angiogram is usually done to delineate the anatomy and for the diagnosis. In this case, the origin of LMCA could not be visualized on transthoracic echocardiography (TTE) leading to inconclusive diagnosis. CTCA delineated the exact anatomy and showed its origin from RPA. Intraoperative TOE examination also revealed it as a case of AOLCARPA.

Anesthetic management of patients presenting with ALOCARPA includes rapid and smooth intubation, while avoiding swings in blood pressure and heart rate. A decrease in pulmonary vascular resistance is avoided by preventing hyperventilation, hyperoxia and hypocarbia [3] The aim is to maintain a PaO_2 of 80–100 mm Hg and an $EtCO_2$ of 40–45 mm Hg. Both reduction and increase in afterload can hamper RCA perfusion and stroke volume, respectively. An optimum preload is required to ensure adequate cardiac output.

CTCA and TOE examinations are useful modalities for delineating the coronary anatomy and help in planning the perioperative surgical and anesthetic management of the patient.

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Conflicts of interest
There are no conflicts of interest.

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