Perioperative Concerns of Anomalous Origin of the Left Main Coronary Artery From the Pulmonary Artery

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To the Editor,

Anomalous origin of the left main coronary artery from the pulmonary artery (ALCAPA), or Bland–White–Garland syndrome, is a rare type of congenital heart disease. It affects 1 in 300,000 live births.¹ It is one of the most common causes of myocardial ischemia and infarction in children. Most of these patients will not survive for a year.² Diagnosing this abnormality in adulthood is rare. We present a case of successful surgical repair of this coronary anomaly.

A 25-year-old female had no medical history before suddenly lost consciousness. Her family immediately called emergency medical services and initiated cardio-pulmonary-cerebral resuscitation (CPCR). After 10 minutes of CPCR, the emergency medical services arrived and activated an automated external defibrillator. She was sent to the hospital nearby after return of spontaneous circulation. Coronary angiography revealed left coronary artery (LCA) orifice in the pulmonary artery (PA) and congenital anomaly with right coronary artery (RCA) collateral circulation. She was transferred to our hospital for surgery.

In the operating room, standard monitoring showed initial heart rate = 116 bpm, blood pressure = 136/62 mmHg, room air SpO₂ = 98%. After right radial arterial catheterization, general anesthesia was induced with IV midazolam 2.5 mg, fentanyl 50 mcg, propofol 200 mg, and rocuronium 50 mg. She was intubated with #7.0 endotracheal tube and controlled ventilation keeping EtCO₂ at 38–40 mmHg. Sevoflurane and O₂-air mixture were used to keep adequate entropy values of 40–60. Transesophageal echocardiography revealed slightly reduced left ventricular (LV) systolic function and hypokinesis of the LV anterior wall.

After midline sternotomy and pericardiotomy, the diagnosis of ALCAPA was confirmed. The RCA was severely dilated and tortuous. The surgeon ligated the anomalous origin of LCA in combination with saphenous vein grafting between LAD and aorta. Under inotropic support with dopamine (3–5 mcg/kg/min), the cardiopulmonary bypass weaning successfully. The patient had an uncomplicated perioperative and postoperative course.

ALCAPA is the commonest congenital coronary anomaly. According to the collateral circulation development, it can divide into the infantile and the adult types. The adult type has better collaterals from the RCA, which provide adequate LV perfusion.³,⁴ It accounts for 10–15% of patients to survive in later childhood.² With the pulmonary pressure decreasing, it may cause a reversal of flow in the left coronary circulation to pulmonary circulation. This situation was known as the “coronary steal phenomenon” and result in myocardial ischemia, myocardial infarction, LV dysfunction, and mitral regurgitation, leading to sudden cardiac death.

The anesthetic goals are to preserve adequate preload, contractility, and keep myocardial oxygen balance. Because of extensive RCA collateral circulation to LCA territory in adult ALCAPA, once decreased pulmonary vascular resistance (PVR) will amplify RCA-to-PA shunt, low FiO₂, and higher EtCO₂ were set to maintain normal to higher PVR.⁴,⁶

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In this patient, we kept entropy values of 40–60 to assure the depth of anesthesia. Perioperative fentanyl (25–50 mcg) and nicardipine (0.5–1 mg) bolus were used to manage hypertension and tachycardia. The systemic vascular resistance (SVR) is inversely proportional to cardiac output and also affects LV systolic wall stress. Maintain normal to low SVR can provide better RCA perfusion and myocardial oxygen balance. Being familiar with the pathophysiology of ALCAPA, the patient can have better pre-bypass management.

References