Case Report

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Pulmonary Atresia with Ventricular Septal Defect: Rare Presentation with Coronary-to-Pulmonary Artery Collaterals from Both Right and Left Coronaries

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Abstract

Pulmonary atresia with ventricular septal defect and coronary-dependent pulmonary circulation arising from both major coronary arteries is rare. Dependence of pulmonary blood flow on the coronaries and the risk of early development of pulmonary vascular obstructive disease warrant early surgical repair in these patients. We report a case of a ten-month-old infant with pulmonary atresia with ventricular septal defect and coronary artery-to-main pulmonary artery connections who was successfully managed with ligation of the coronary fistulas and intracardiac repair.

Keywords

coronary artery anomaly, pulmonary atresia with ventricular septal defect, fistula, collateral blood flow

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Introduction

Pulmonary atresia with ventricular septal defect (PA/VSD) is known for its heterogenous sources of pulmonary blood supply. The ductus arteriosus and aortopulmonary collateral arteries are the usual sources of pulmonary blood flow. Fistulous communication between coronary arteries and pulmonary arteries as the sole source of pulmonary blood flow is rare. We report a very rare case of pulmonary atresia with VSD with coronary-dependent pulmonary circulation arising from both major coronary arteries.

Material and Methods

A ten-month-old infant presented with complaints of difficulty with feeding, tachypnea, and an antecedent history of bronchopneumonia. Examination revealed a hyperdynamic precordium, with harsh continuous murmur and mild cyanosis. Pulse oximetry showed an oxygen saturation of 86%. Echocardiographic evaluation showed dilated origin of both the right and left coronaries. Both the coronary arteries supplied the main pulmonary artery (MPA) through large fistulous communications. There was no evidence of ischemia on the electrocardiogram. Cardiac computed tomography displayed collaterals from both the coronaries to the pulmonary artery and absence of other aortopulmonary collaterals (Figure 1).

During surgery, both right and left coronary ostia had normal origin from the respective aortic sinuses. The right coronary artery gave off a large collateral just after its origin which traversed across the base of the aorta to join the MPA. The left main coronary artery gave off a collateral to the MPA and then went on to divide into left anterior descending artery and left circumflex artery. There was no communication between the MPA and the right ventricle. Intraoperative transesophageal echocardiography confirmed these findings (Figure 2).

The collaterals were dissected out and circled with vessel loops at their respective origins (Figure 3). Aorto-bicaval

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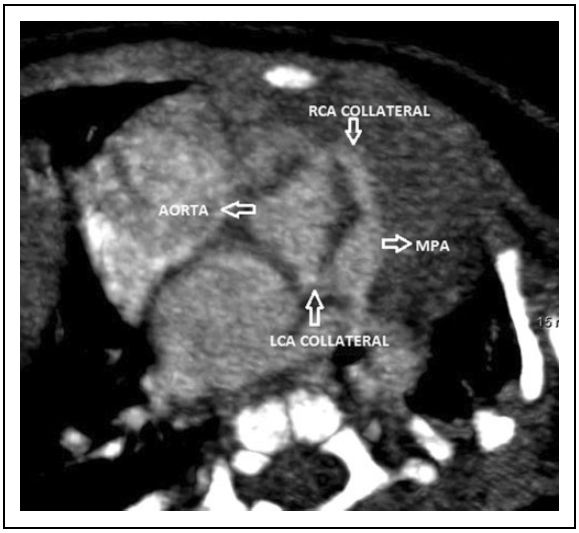

Figure 1. Cardiac computed tomography displaying collaterals from both the left coronary artery (LCA) and right coronary artery (RCA) to the main pulmonary artery (MPA).

bypass was established and cardioplegic arrest achieved after snugging both branch pulmonary arteries to avoid run off. Both coronary collaterals were transfixed and ligated close to the MPA. Intracardiac repair was done with VSD closure and reconstruction of the right ventricular outflow tract with bovine pericardium and a polytetrafluoroethylene monocusp. Post procedure the right ventricle to left ventricle pressure ratio was 0.7. The pulmonary artery pressure was 52 mm Hg and the left atrial pressures 8 mm Hg, suggestive of pulmonary artery hypertension. Postoperative transthoracic echocardiography performed in the intensive care unit showed a peak systolic pulmonary artery pressure of 51 mm Hg and the patient was started on oral sildenafil. The postoperative period was uneventful, and the child made good recovery.

Comment

The incidence of pulmonary atresia with VSD is 2.5% to 3.4% of all congenital heart disease; 0.07/1,000 live births.[1] Rarely the source of pulmonary artery blood flow is a fistula from a coronary artery. Literature search reveals 53 patients with PA/VSD who were reported to have MPA blood flow through coronary-to-pulmonary artery fistulas.[2] The fistulas originated from either the right or the left coronary artery in most patients. In four cases, the patients had single coronary artery anatomy. To date, there are only two previous reports of coronary-to-pulmonary artery fistulas from both coronaries.[2]

Most patients present with features of increased pulmonary blood flow and signs of congestive cardiac failure.[3] Our patient also presented with features of failure. There was however no evidence of ventricular dysfunction on echocardiogram. In the absence of cardiac catheterization, the anatomic status of the distal pulmonary vascular bed was unknown. Coronary ischemia has been rarely documented in these patients as coronary flow is dependent on the state of the distal coronary bed as long as there is no proximal stenosis.[4] The lack of evidence of ischemia on the electrocardiogram in spite of the presence of fistulas from both coronaries supports this theory. In our patient, the pulmonary artery pressure remained high after surgery in the presence of low left atrial pressure. This suggests the early development of pulmonary arterial hypertension in these patients.

In the absence of other major aorto-pulmonary collateral vessels, increased pulmonary blood flow in these patients is generally associated with favorable pulmonary artery anatomy, making them suitable for primary intracardiac repair with or without the use of a right ventricle-to-pulmonary artery conduit. In our patient, posterior continuity was achieved using a small patch of bovine pericardium (Figure 4). Homograft was not used due to lack of availability. Dependence of pulmonary

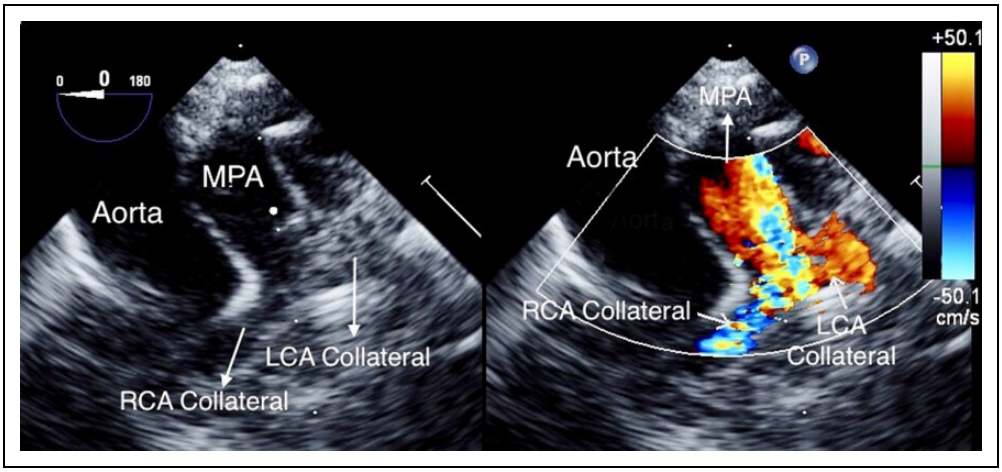

Figure 2. Intraoperative transesophageal echocardiographic image showing the fistulous connections of the right and left coronary arteries to the main pulmonary artery.

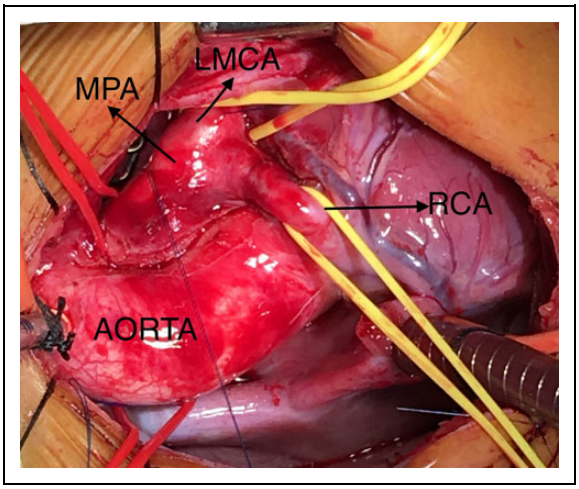

Figure 3. Both right and left fistulas have been dissected out and circled with vessel loops at their respective origins.

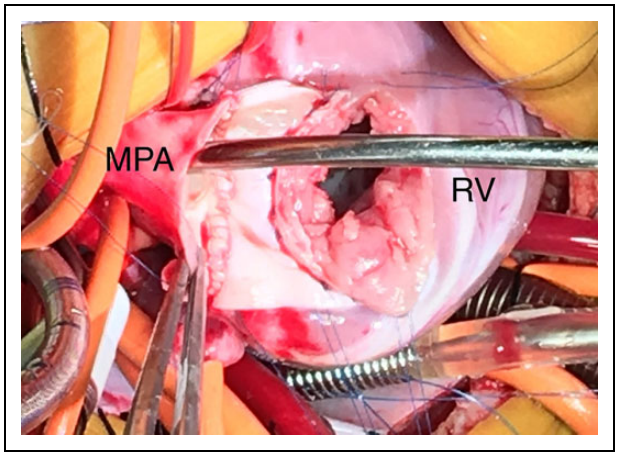

Figure 4. Right ventricular outflow tract reconstruction performed using pericardial patch; fistulous communications from the coronary arteries have been ligated.

blood flow on the coronary arteries and the risk of early development of pulmonary vascular obstructive disease warrant early surgical repair in these patients.[5]

Authors' Note

Permission was granted by the patient's parent to publish the case report. We had full control of the study design, the methods used, outcome parameters, analysis of data, and production of the written report.

Declaration of Conflicting Interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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