Coronary Artery-Pulmonary Artery Fistula in Pulmonary Atresia With Ventricular Septal Defect; Report of 2 Surgical Cases

RUNNING HEAD: Coronary Artery-Pulmonary Artery Fistula

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Abstract

Pulmonary atresia with ventricular septal defect (PA-VSD) is a congenital complex cardiac anomaly. The blood supply to the lungs can be from patent ductus arteriosus (PDA), major aortopulmonary collateral artery (MAPCA) or in very rare cases from coronary artery-pulmonary artery fistula (CAPAF). We had coronary artery-pulmonary artery fistula (CAPAF) in 2 patients which had surgical intervention. In our first patient which was operated 10 years ago, the main pulmonary artery had a source from LMCA. In second patient, the right pulmonary artery derived from LMCA and advanced to the right lung from the posterior of the aorta and the left pulmonary artery was coming out of the PDA.

Keywords Congenital, Coronary artery-pulmonary artery fistula, CAPAF, PA-VSD, Pulmonary atresia, Ventricular septal defect

Introduction

Pulmonary atresia with ventricular septal defect (PA-VSD) is a congenital complex cardiac anomaly. Pulmonary blood supply is usually provided by PDA and MAPCAs in patients with PA-VSD. Rarely, the source of pulmonary blood flow is from coronary arteries. These are called coronary artery-pulmonary artery fistula (CAPAF) [1]. We had CAPAF in 2 patients with PA-VSD. First patient which was operated 10 years ago, the main pulmonary artery had a fistula from the LMCA. In second patient, the right pulmonary artery had a fistula from the LMCA and advanced to the right lung from the posterior of the aorta. The left pulmonary artery was coming out of the PDA. A limited number of case reports were found in the literature.

Case Reports

Patient 1:

A 4-year-old boy with a diagnosis of PA-VSD was admitted to our hospital. Cardiac examination revealed normal S1, S2 and a continuing 3/6 murmur in the left midsternal border. Transthoracic echocardiography (TTE) showed large VSD and pulmonary atresia. It was observed that LMCA was fistulated into main pulmonary artery (Figure 1). Diagnostic catheter angiography was done and the findings in TTE were confirmed. MAPCAs that had a blood supply from the descending aorta to both lungs were identified.

The patient was taken into operation and CPB (cardiopulmonary bypass) was initiated after aortic and bicaval cannulation. Cardiac arrest was done after both left and right pulmonary artery was suspended. Aortotomy was done. Left coronary ostium was found to be highly dilated. It was seen that LMCA came out from the posterior of aorta and bifurcating to Left Anterior Descending (LAD) and Circumflex arteries. After pulmonary artery was opened longitudinally, a fistula from LMCA to pulmonary artery was seen (Figure 2). The fistula between LMCA and main pulmonary artery was transected and the roof over LMCA was closed with a pericardial patch. VSD was closed transatrially and transventricularly with polytetrafluoroethylene (Goretex) patch. 4-5 mm fenestration was opened in the middle of the patch. The right ventricular outflow tract and transected pulmonary artery were reconstructed with a 19 no Labcor stentless valved pulmonary conduit (Labcor, Belo Horizonte, Brazil).

The patient who had pulmonary hemorrhage during intensive care follow-ups was referred to cardiac catheterization. Cardiac catheterization was done for occlusion of MAPCAs after the operation. MAPCAs were closed with coils. Saturation decreased from 97% to 88%. After MAPCAs occlusion, pulmonary artery pressure decreased from 56 mmHg to 51 mmHg. In the postoperative follow-up, the patient was not weaned from mechanical ventilation at the 42th day tracheostomy was done. At the postoperative 50th day, pulmonary hemorrhage was seen again. After serious anti-pulmonary arterial hypertensive medical treatments, pulmonary artery pressure regressed and the pulmonary hemorrhage disappeared. The patient weaned from tracheostomy and was discharged at the third postoperative month. In the postoperative TTE, a two-way shunt was observed in the fenestrated interventricular patch.

The patient was admitted to hospital 8 years later with complaints of ascites and symptoms of NYHA class III. After the TTE, VSD closure was planned due to one-way shunt from left to right causing high pulmonary artery pressure. It was closed with 16 mm muscular VSD occluder. In control TTE, measured right ventricular pressure 50 mmHg, improve right ventricular systolic functions and mild-to-moderate conduit stenosis was seen. The patient was discharged from the hospital. We have been following the patient for 10 years and is still alive with NYHA class I.

Patient 2:

A 40-day-old child with 3350 gr PA-VSD was admitted to hospital. The baby had a tachycardia, difficulty in breastfeeding with a saturation of 90%. Cardiac examination revealed normal S1, S2 and a continuing 3/6 murmur in the left midsternal border.

In the transthoracic echocardiography (TTE), a wide VSD in the subaortic region and ascending aorta coming from the heart as a single root. It was 60% dextroposed. Cardiac catheterization was performed for advanced examination. The right pulmonary artery had originated from LMCA and continues to the right through posterior of ascending aorta. The left pulmonary artery originated from PDA. Dilated LAD was seen at the continuation of LMCA (Figure 3). MAPCA was not detected in this patient.

PDA was kept open with PgE1 infusion until the patient was taken into operation. In this case, CAPAF causing high blood flow to the pulmonary vascular bed and dilated LAD was seen after the origin of the right pulmonary artery from LMCA (Figure 4). The right pulmonary artery was suspended, CPB was initiated after aortic and bicaval cannulation. The right pulmonary artery was divided from LMCA. The stump in LMCA was closed with primary suturing. The left pulmonary artery was transected from PDA and PDA was closed. Right and left pulmonary artery anastomosed posteriorly and augmented with fresh pericardium anteriorly. Modified Blalock-Taussing (m-BT) shunt with 4 mm PTFE graft was done between the right subclavian artery and right pulmonary artery. The patient, who was admitted to intensive care for 2 days and was discharged on the postoperative 9th day.

Discussion

Coronary artery to pulmonary artery fistula is a rare condition in PA-VSD patients. The prevalence of CAPAF has been reported as 7-11% in retrospective case studies in the literature [2]. Collison et al. reported four cases of CAPAF (8%) among 50 PA-VSD patients [3]. In another review, Amin et al. 9 (10%) CAPAF patients were detected in 87 PA-VSD patients [4]. Similar rates were found in other studies. In our study, among 78 PA-VSD patients, 2 (2,5%) patients had CAPAF.

In the literature, CAPAF originates more from the left coronary artery than RCA. It was seen that the most common CAPAF originating from left coronary system was those originating from circumflex coronary artery [2]. In the study of Yadav et al. with 2 patients, both patients had main pulmonary artery originating from LMCA [1]. In our study, CAPAF originated from LMCA in 2 patients.

Another important detail in this pathology is the situation of MAPCAs. MAPCAs were present in 90% of previously reported cases. The remaining 10% did not have MAPCA and fistulas were found to be the sole blood source of the pulmonary circulation [2]. There is a risk of coronary steal in PA-VSD patients with CAPAF but such situation was not reported in the literature [2]. In our patients, we can speculate that increased blood flow through the fistulas to the lungs after we have seen the enlarged pulmonary arteries. But we had not seen any ECG changes in these patients.

Different surgical approaches were applied to these patients according to the experience of the surgeons and pulmonary artery development. In Amin et al. review, all cases had additional MAPCAs, seven patients from the left and two from the right coronary system. One-stage unifocalization was applied to all patients [4]. Collison et al. performed two-stage repair on all four patients. The first stage was unifocalization followed by complete repair [3]. In our first patient we have MAPCAs. In those years, we had less surgical experience and could not do flow study during surgery. As time passed, our surgical strategies have changed. About 6 years ago, we initiated a unifocalization program. Our current approach which is the single-stage unifocalization with intraoperative flow study. That is the reason why we left the MAPCAs in first case during surgery. MAPCAs were closed with a coil after the operation. Our second patient did not have MAPCAs, unifocalization of right and left pulmonary artery and m-BT shunt were done. VSD closure was left for the next operation. Both of our patients are still alive.

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Compliance with Ethical Standards

Conflict of Interest None of the authors have any conficts of interest to declare.

Consent to participate Written informed consent was obtained from the parents.

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