



Coronary Artery Fistula with Double Outlet Right Ventricle: a Case Report

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Abstract

The majority of coronary artery fistulas (CAFs) are congenital. The anomaly accounts for 0.4% of congenital heart defects and approximately 50% of pediatric coronary vasculature anomalies. Twenty percent of people with congenital CAFs have other concomitant cardiac anomalies, most frequently aortic and pulmonary atresia and patent ductus arteriosus. It is worthy of note that CAF with the tetralogy of Fallot has also been reported. Here we describe a patient with a double outlet right ventricle in association with a coronary artery fistula.

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Introduction

Coronary artery fistula (CAF) is a rare, usually solitary, anomaly that accounts for approximately 0.4% of congenital heart defects. The majority of cases in the pediatric population are congenital in nature, while acquired fistulas are often iatrogenic as a result of coronary insult.¹ There are some reported cases of congenital CAF in association with the tetralogy of Fallot (TOF)²⁻⁵ and some iatrogenic cases detected postoperatively in patients with TOF.

Case report

A 14-year-old girl was referred for cardiac surgery with a diagnosis of TOF. She had had central cyanosis and exertional dyspnea since birth and had not been able to walk since she

was two years old. On examination, she was underdeveloped with central cyanosis, clubbing of fingers, and a squatting position. A right ventricular impulse and systolic thrill were palpable along the left sternal border. The second heart sound was single. Arterial oxygen saturation was about 55% on admission.

Transthoracic echocardiography showed a double outlet right ventricle (about 75% overriding of the aorta toward the right ventricle, lack of fibrous continuity between the posterior aortic valve leaflet and anterior mitral valve leaflet, and side-by-side position of aorta and pulmonary artery); large perimembranous ventricular septal defect; hypoplastic right ventricular outflow tract, pulmonary artery, and pulmonary artery branches; thickened pulmonary valve; small atrial septal defect; severe right ventricular enlargement

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and hypertrophy; and continuous flow and turbulancy in the ascending aorta toward the main pulmonary artery, which could be a fistular connection or aortopulmonary window. There was also a continuous flow and turbulancy between the descending aorta and left pulmonary artery, which could be a small patent ductus arteriosus or a collateral vessel.

Catheterization showed atrial septal defect, large ventricular septal defect, severe subvalvular pulmonary stenosis in oximetric and pressure studies, and simultaneous opacification of both aorta and pulmonary artery in right ventricular injection. In addition, no patent ductus arteriosus was detected in aortography. In selective coronary angiography, a large fistula from the proximal left anterior descending to pulmonary artery was apparent (Figure1).

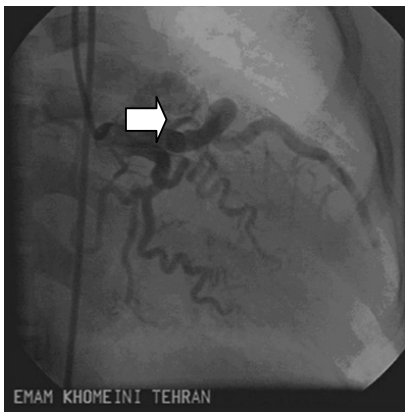


Figure1. Selective coronary angiography demonstrated ectatic coronary arteries and a large fistula from proximal of left anterior descending artery (LAD) to pulmonary artery

Discussion

CAF is a rare, usually solitary, anomaly that accounts for approximately 0.4% of congenital heart defects. The majority of cases in the pediatric population are congenital in nature, possibly arising from the persistence of sinusoidal–coronary arterial connection, while acquired fistulas are often iatrogenic because of coronary insult or the patient’s having undergone endomyocardial biopsies. Congenital fistulas most frequently arise from the right coronary artery system, and the great majority (~90%) exit into the right heart structures, including the vena cava, coronary sinus, or pulmonary arteries.¹ Twenty percent of people with congenital CAF have other concomitant cardiac anomalies, most frequently aortic and pulmonary atresia and patent ductus arteriosus.⁶

In this patient with a diagnosis of a double outlet right ventricle with a subaortic ventricular septal defect and subpulmonary stenosis, the clinical scenario and management algorithm were similar or identical to those of TOF. Coronary artery anomalies are encountered in up to 12% of patients with TOF. The most common variants include the left anterior descending arising from the right coronary artery or

separately from the right sinus of Valsalva. Many of these variants result in a major coronary artery coursing anterior to the pulmonary outflow tract, which in turn complicates the ultimate surgical repair.

The recently acquired ability to repair TOF early in life, regardless of the coronary artery anatomy, has largely been the reason for many academic centers to no longer perform routine coronary artery angiography prior to surgery.¹

Catheterization and coronary angiography were performed in this patient to clarify the suspicious flow and turbulancy in the ascending aorta toward the pulmonary artery and the descending aorta toward the left pulmonary artery. Coronary angiography showed a CAF between the left anterior descending and pulmonary artery, which helped the patient by providing a left to right shunt.

In the existing literature, there are some reported cases of congenital CAFs with TOF,²⁻⁵ but no cases with a double outlet right ventricle have been reported. Therefore, this patient is the first reported case of CAF with a double outlet right ventricle. Surgery was performed with central shunting from the ascending aorta to the pulmonary artery because a corrective surgery was impossible due to the patient’s hypoplastic pulmonary artery branches. The oxygen saturation increased from 55% to 78% after surgery.

Conclusion

This case was interesting because of association of double outlet right ventricle and coronary artery fistula. As in this case we should consider coronary artery fistula especially in patients with tetralogy of fallot or double outlet right ventricle both in echocardiography and catheterization.

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