Coronary Artery Fistula with Heart Failure in Early Infancy

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Abstract

Heart failure in early infancy is commonly caused by lesions leading to pulmonary over circulation secondary to left-to-right shunt. This case report describes an unusual cause of significant left-to-right shunt in a 2 months old infant presenting with congestive heart failure, which was diagnosed with transthoracic echocardiography. In this infant, transthoracic echocardiography with Doppler color flow mapping allowed direct visualization of a large right coronary artery to right ventricular fistula that was surgically corrected successfully.

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Received: 03 Jun 2010
Accepted: 27 Jun 2010

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doi:10.5001/omj.2010.73

Introduction

Heart failure in early infancy is commonly caused by left-to-right heart shunts. Such lesions include ventricular septal defects, atrioventriculoseptal defects, complex congenital heart disease with unrestricted pulmonary blood flow, etc. This case report describes an unusual cause of significant left-to-right shunt in an infant presenting with congestive heart failure, which was diagnosed with transthoracic echocardiography.

Case report

A 2 month old full-term female infant, born without complications presented to Royal hospital with congestive heart failure precipitated by chest infection. She was tachypneic with intercostals and subcostal recession. She was pink, had full volume bounding pulses and femoral pulse was easily felt. Her precordium was hyperdynamic and there was grade 3/6 continuous murmur in the left lower sternal border. Her liver was 4.5 cm below the costal margin. An electrocardiogram was normal for her age, and chest radiograph showed cardiomegaly and plethoric lungs. Transthoracic echocardiography showed situs solitus with mildly dilated right ventricle (RV) and normal left ventricle.

There was a 9 mm secundum atrial septal defect with left to right shunt, normal pulmonary venous drainage into the left atrium, intact interventricular septum, non-dilated coronary sinus with no persistent ductus arteriosus (PDA) and normal valves. There was gross dilatation (7 mm) of the right coronary artery (RCA) throughout the whole course along the RV free wall up to apex where it was seen draining into the RV with a 4 mm single exit orifice. (Figs. 1 & 2A)

Color Doppler revealed turbulent blood flow starting from RCA, traversing lateral aspect of RV and finally drained into the RV cavity with Doppler interrogation demonstrating a continuous flow signal, (Fig. 2B). There was no localized aneurysmal formation noted. The calculated pulmonary to systemic blood flow (Qp/Qs) ratio was 2.1:1 and the pulmonary artery systolic pressure was 55 mmHg.

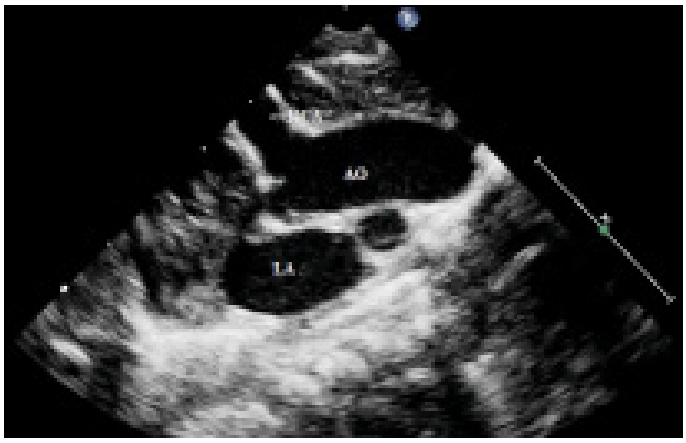
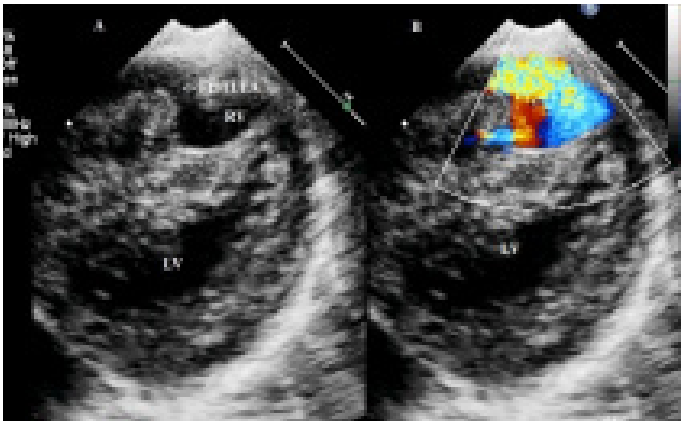


Figure 1: Transthoracic echocardiography showing grossly dilated right coronary artery in an infant with coronary-cameral fistula.
LA=Left atrium; AO=Aorta; RCA=Right coronary artery.

The infant was treated with diuretics and antibiotics, but needed ventilator management. She underwent cardiac catheterization to delineate further the fistula and for possible intervention. Coronary angiogram performed prior to surgery showed large RCA to RV fistula with two large openings into RV each measuring 6 mm in diameter. (Fig. 3)

Since there were two large exit points into the RV, she needed two 10 or 12 mm Amplatzer vascular plugs or two 8/6 Amplatzer PDA device occluders. Also, due to the small weight of the baby (3 kg), a large sheath would be needed for percutaneous procedure with related complications and hence she was advised surgery. She underwent urgent surgical ligation of the fistula along with atrial septal defect closure with uncomplicated recovery.



Figures 2A & 2B: (A) Transthoracic echocardiography showing connection of right coronary fistula into right ventricle. (B) Color Doppler shows turbulent blood flow communicating between the right coronary artery and right ventricle.

LV=Left ventricle. RV=Right ventricle.

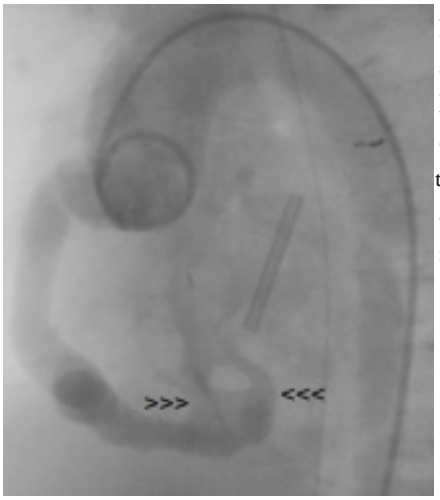


Figure 3: Ascending aortogram showing a large dilated right coronary artery with two fistulous exit openings (arrow heads) into the right ventricle.

Discussion

Coronary artery fistulas (CAF) are uncommon anomalies in which a communication exists between a coronary artery and a cardiac chamber (coronary-cameral fistula) or a great vessel (coronary arterio-venous fistula), bypassing the myocardial capillary network. They are mostly congenital but acquired forms can also occur due to trauma or iatrogenic cause (after cardiac surgery or transcatheter interventions).

Congenital CAF arise due to incomplete closure of embryonic intertrabecular spaces and coronary sinusoids. The functional disturbance caused by CAF is due to myocardial stealing secondary to a diastolic pressure gradient and runoff from high-pressure coronary artery to a low-pressure cardiac chamber with decrease in intracoronary diastolic perfusion pressure.

The coronary artery compensates by enlargement of the donor artery and may become aneurysmal with risk of thromboembolism and rarely, endocarditis and rupture.[1]

The hemodynamic consequences of CAF depend on their origin, size of the communication, the resistance of the recipient chamber, and the potential for development of myocardial ischemia and significant left-to-right shunt. The reported incidence of CAF is about 0.1-0.2% among all cardiac catheterizations.[2] Approximately 50% of pediatric coronary anomalies are CAF. Concomitant congenital anomalies occur in 40% of patients. The origin of the fistulae is variable; right coronary artery (50%), left coronary artery (42%), both coronary arteries (5%). More than 90% of CAF drain into right heart; right ventricle (41%), right atrium (26%), pulmonary artery (17%) and the rest in coronary sinus, left atrium, left ventricle or superior vena cava.[1]

CAF are usually asymptomatic, especially in those less than 20 years of age and who have small CAF.[3,4] They are accidentally discovered during echocardiography or coronary angiography. It tends to manifest in infants less than 2 years of age with heart failure; in young adults with angina, dyspnea on exertion, myocardial ischemia/infarction; and in adults more than 40 years of age with heart failure, atherosclerosis and arrhythmias.[4] The presence of a continuous murmur in the lower sternal border is highly suggestive of a CAF. Differential diagnosis include PDA, ruptured sinus of Valsalva aneurysm, aortopulmonary window, supracristal ventricular septal defect with aortic regurgitation, internal mammary artery to pulmonary artery fistula, and pulmonary or systemic arteriovenous fistula.[1,5]

Traditionally, coronary angiography is the gold standard for imaging the coronary tree and also CAF. Noninvasive imaging with echocardiography, computed tomography, and magnetic resonance imaging (MRI) may facilitate the diagnosis of CAF.[6] Transthoracic echocardiography with color Doppler is non-invasive, easily available, and fairly accurate in diagnosing many forms of CAF. Associated cardiac lesions and ventricular function can be evaluated, but the major limitation is that branching coronary vessels from the fistula are not recognizable. Limitation of transthoracic echocardiography was noted in our patient; namely only one exit orifice of the CAF was seen, and the other orifice was diagnosed by aortography. MRI is a good alternative for imaging proximal coronary anomalies, but not for distal course of CAF.

Multi-detector row computed tomography cardiac imaging has provided excellent distal coronary artery and side branch imaging with better temporal and spatial resolution than MRI and is considered by many as diagnostic modality of choice in imaging of coronary anomalies. If a decision is taken to perform percutaneous

embolization of the fistula, then cardiac catheterization is indicated.[5]

Small fistulas which are haemodynamically and clinically silent usually do not require any treatment. Patients with large fistula, symptoms of heart failure or myocardial ischemia and pediatric patients with high Qp/Qs should be treated early.

Treatment options include surgical repair (patch/suture closure or ligation),[7,8] or transcatheter embolization.[7,9,10] Mavroudis et al. in their series reported 100% success rate and survival with surgery and they recommend elective coil occlusion in patients who fulfill the following criteria; absence of multiple fistulas, a single narrow drainage site, absence of large branch vessels, and safe accessibility to the coronary artery supplying the fistula.[7] Different materials are used in the embolization of CAF; releasable balloons, microcoils/hydrocoils, micro particles and Amplatzer occluder or plug. In asymptomatic cases, regular follow up is advised because there is a chance of spontaneous closure of small clinically silent fistulas.[3,4]

Conclusion

CAF are a rare cause of heart failure in early infancy. Detection of a continuous murmur in a site atypical for PDA and no clear-cut explanation during routine echocardiographic examination should raise the suspicion of CAF.

Acknowledgements

The authors reported no conflict of interest and no funding was received on this work.

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