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Coronary Cameral Fistula in a Nigerian Infant: An Uncommon Cause of Continuous Murmur

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Case Report

ABSTRACT

A coronary artery fistula involves a sizeable communication between one of the coronary arteries and a cardiac chamber (coronary cameral fistula) or a vein (coronary arteriovenous fistula). Coronary fistulae account for 0.2 to 0.4% of the congenital cardiac abnormalities, and about 50% of the Pediatric coronary vasculature abnormalities. Sixty percent of these fistulae arise from the right coronary artery and terminate mostly (90%) in the right side of the heart. The most frequent sites of termination are the right ventricle, right atrium, coronary sinus, and the pulmonary vasculature. Most children with coronary artery fistulae (CAF) are asymptomatic, and continuous murmur may be audible on routine examinations. The majority of these patients present in adulthood and are usually asymptomatic, often being detected accidentally.

The authors are unaware of any case previously reported in a Nigerian child and hence report the case of a 3 month old boy for its rarity and interest. The girl was referred for evaluation of a cardiac murmur associated with recurrent respiratory tract infection necessitating admissions since birth. Clinical, radiological and electrographic findings were suggestive of a patent ductus arteriosus. A doppler echocardiogram done revealed a coronary cameral fistula from the Left Coronary Artery draining into the Left Atrium.
INTRODUCTION

A coronary artery fistula involves a sizeable communication between one of the coronary arteries and a cardiac chamber (coronary cameral fistula) or a vein (coronary arteriovenous fistula) \[^1\]. It accounts for 0.2 to 0.4% of the congenital cardiac abnormalities, and 50% of the Paediatric coronary vasculature abnormalities. Coronary camera fistula is a type of coronary fistulae \[^1\]. Coronary artery fistulas are thought to arise as a persistence of sinusoidal connections between the lumens of the primitive tubular heart that supply myocardial blood flow in the early embryologic period \[^1\]. Their morphogenesis is thought to begin early with the development of embryological myocardial lacunae when myocardial blood supply is derived from its own lumen. Subsequently the coronary arteries communicate with these intramyocardial trabecular spaces \[^1\]. These sinusoids may involute forming the microcirculation. If the pulmonary valve fuses before the sinusoids contract, RV pressure increases from isovolumic contraction and flow into the sinus is maintained \[^1\]. It also occurs in 45-50% of patients with pulmonary atresia, who have an intact ventricular septum \[^1\].

CASE PRESENTATION

A 3-month old girl referred to Lagos State University teaching Hospital, Ikeja for cardiac evaluation for a murmur. She has been admitted since birth on account of recurrent respiratory tract infection and failure to gain weight. Birth weight was 3.5kg.

Examination revealed a small for age, afebrile infant who weighed 3.65kg. She was tachypneic, tachycardic and had tender hepatomegaly. Her first heart sound was normal but P2 was loud with a Grade 3/6 continuous murmur loudest at the left lower sternal edge.

A clinical diagnosis of patent ductus arteriosus with pulmonary artery hypertension and congestive cardiac failure was done. Chest radiograph and electrocardiograph supported the diagnosis of a patent ductus arteriosus.

A Doppler Echocardiography done revealed a coronary cameral fistula from the left coronary artery draining into the right atrium. Echocardiography diagnosis was confirmed at India where she had transcatheter closure of the fistula and was placed on aspirin and clopidogrel and has been stable on follow-up.
DISCUSSION

Coronary Cameral fistula is a rare cardiac anomaly resulting from aberration in connection between a coronary artery and the cardiac chamber [2]. It is a form of coronary artery fistula which comprises of coronary cameral fistula and coronary artery or arteriovenous fistula [1]. Coronary artery fistula was first described by Krause in 1865 and it accounts for 0.2% to 0.4% of all congenital heart disease [3]. Diagnosis of coronary artery fistulas was done in 0.06% of children during echocardiography [4]. A diagnosis of coronary cameral fistula was done in 0.08% to 0.3% of persons that had diagnostic angiography. The reported incidence of coronary fistulas is possibly underestimated, since in 75% of cases it is asymptomatic [5]. It has no sex or race predilection.
Coronary arteriovenous fistulas are mostly congenital, but cases of traumas, infection, iatrogenic causes have been documented following cardiac surgeries. In most cases, congenital coronary arteriovenous fistulas are asymptomatic with incidental finding of continuous murmur that is associated with a diastolic accentuation. The continuous murmur in a coronary fistula is loudest at the lower sternal edge compared to a patent ductus arteriosus murmur that is loudest at pulmonary region. Confirmation of a coronary fistula is made by coronary angiography or transthoracic echocardiography and inappropriate evaluation could lead to surgery for the wrong diagnosis of a patent ductus arteriosus.

The hemodynamic manifestation depends on the site of origin, the termination of the fistula, as well as size, length and tortuosity of the fistula. In most cases, the origin of the fistula is from the right coronary artery, which is seen in 50% of the cases. In 33% to 42% cases such as was seen in the present case presented, the origin is the left coronary artery. In about 5% of cases, it originates from both the left and right coronary artery.

The termination of coronary artery fistula is mostly to the right heart chambers. Coronary artery fistula terminates in 40% and 26% of cases in the right ventricle and right atrium respectively. In 17% of cases, coronary fistula terminates in the pulmonary artery while in minority of cases; it terminates in the superior vena cava or coronary sinuses and rarely terminates in the left ventricle.

Symptomatic coronary fistula presents usually within the first 2 years of life with congestive cardiac failure, failure to thrive, recurrent respiratory tract infection or arrhythmia. In older patients, symptoms may include dyspnea on exertion, angina, fatigue and palpitations. Only 19% of children with coronary artery fistula are symptomatic. The physiologic manifestations of the fistulas depend on the termination site. A fistula that empties into the left atrium like in the present case results in volume overload that is similar to mitral regurgitation. The drainage to the right atrium has a physiology that is similar to atrial septal defect with increase pulmonary oxygenated blood flow.

In 40% of congenital coronary fistula, there is co-existence of other cardiac anomaly like tetralogy of Fallot, patent ductus arteriosus, hypoplastic left heart syndrome, atrial septal defect, ventricular septal defects and pulmonary atresia with intact ventricle. In the case presented, no other cardiac anomaly was found.

The use of coronary angiography which is the gold standard for making diagnosis is limited by its invasiveness and requirement of extensive skilled operator. Transthoracic echocardiography with colour Doppler which was used in the present case is a useful diagnostic tool that identifies coronary artery fistulas.

Surgical interventions which could be primary or transcatheter closure is required in symptomatic patients with coronary fistula. The first successful surgical closure of a coronary fistula was performed in 1947 by Bjork and Crafoord in a patient with a preoperative diagnosis of patent ductus arteriosus. Transcatheter closure of coronary fistula was first performed by Reidy and co-workers in 1983 and it is a first line surgical intervention in some cases. Spontaneous closure of small asymptomatic fistulas has been reported and surgical intervention is generally not recommended for small size fistulas.
Prevention of thrombosis especially in fistulas with sluggish blood flow with low dose aspirin and oral anticoagulant was beneficial and was prescribed in the present case.

Failure of closure of medium or large fistula such as seen in the current case reported can lead to long term complications. Likely complications include infective endocarditis, arrhythmia, myocardial infarction, aneurysm formation, rupture and death. Progressive dilation and coronary artery steal phenomenon which occurs due to preferential shunting of blood through the fistula rather than myocardial capillaries with clinically manifest as angina \[1\]. Spontaneous closure has been rarely reported. The mortality rate related to surgical repair of coronary artery fistula typically ranges from 0-4%. Variations that may increase surgical risk include the presence of giant aneurysms and a right coronary artery–to–left ventricle fistula \[1\]. Complications of surgery include myocardial ischemia and/or infarction (reported in 3% of patients \[1\]).

Coronary artery fistula can recur (4% of patients) hence here is need for long term follow-up and cardiac evaluation in patients that had transcatheter closure because of the risk of recanalization of the fistula. There is no evidence of recanalization in the present case.

**CONCLUSION**

Coronary camera fistula is rare. It is a cause of continuous murmur and a close differential to Patent Ductus Arteriosus. Echocardiography with a colour Doppler facility and expertise is important in its diagnosis.

**REFERENCES**

5. Latson LA. *Coronary Artery Fistulas: How to manage them. Catheterization and cardiovascular Interventions. 2007; 70:110-116*