

Prenatal Diagnosis of Coronary Artery Fistula: A Case Report and Review of Literature

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Abstract

We report an antenatally diagnosed large distal coronary artery fistula (CAF) arising from an aneurysmal dilation right coronary artery (RCA) and draining in to the right ventricle (RV) just below the septal leaflet of tricuspid valve posteriorly. A postnatal echocardiogram confirmed the diagnosis. On the second day of life, a percutaneous partial closure of the fistula was performed by placing a Flipper coil (Cook Medical, Bloomington, IN) in the RCA just proximal to the drainage site in the cardiac catheterization laboratory. Follow-up echocardiogram on the day following the procedure showed improved forward flow in the descending aorta with decreased RV size. Our case report suggests that antenatal diagnosis of a CAF may aid in early intervention. Partial closure of the fistula in the cardiac catheterization laboratory is safe and effective.

Keywords

- ▶ prenatal diagnosis
- ▶ coronary artery fistula
- ▶ heart failure

Coronary artery fistula (CAF) is a connection between a coronary artery on one side and a cardiac chamber or great vessel on the other side that bypasses the myocardial capillary bed. CAF was first described by Krause¹ in 1865 and accounts for 0.2 to 0.4% of all congenital heart disease and 14% of congenital anomalies of the coronary arteries. It can occur in isolation or in association with other congenital heart disease.² CAF arises from the right coronary artery (RCA), left anterior descending coronary artery, and the circumflex coronary artery in 52, 30, and 18% of cases, respectively. Over 90% of CAF drain to the right side of the heart, most commonly to the right ventricle (RV) in approximately 40% of the cases, followed by the right atrium, coronary sinus, and pulmonary trunk. Multiple CAF to the left ventricle have also been reported.

In the last two decades, prenatal ultrasound screening has been widely used, and fetal echocardiographic detection of cardiac lesions has become routine. As a result, nine case reports of prenatally diagnosed isolated CAF have been published since 1996.^{2–10} In this report, we describe a case of a prenatally diagnosed RCA fistula to RV with some novel

aspects, followed by a literature review of the echocardiographic findings, management and outcomes of prenatally diagnosed isolated CAF.

Case Report

A 25-year-old multiparous woman with a singleton fetus diagnosed to have a trisomy 21 on amniocentesis was referred to us for a fetal echocardiogram at 31 weeks gestation. The initial obstetric ultrasound suggested a possible diagnosis of complete atrioventricular canal defect and significant flow reversal in the ascending aorta. There was no history of diabetes, hypertension, or drug use in mother. Fetal echocardiogram revealed normal four chamber anatomy with normal relationship of great arteries. All the four valves were morphologically normal with normal Doppler flow pattern. There was a large distal CAF arising from an aneurysmal dilated RCA (▶ **Fig. 1A, B**) draining in to the RV just below the septal leaflet of tricuspid valve posteriorly (▶ **Fig. 1C**). There was abnormal retrograde diastolic flow across the aortic isthmus and ascending aorta (▶ **Fig. 1D**) with normal antegrade flow across

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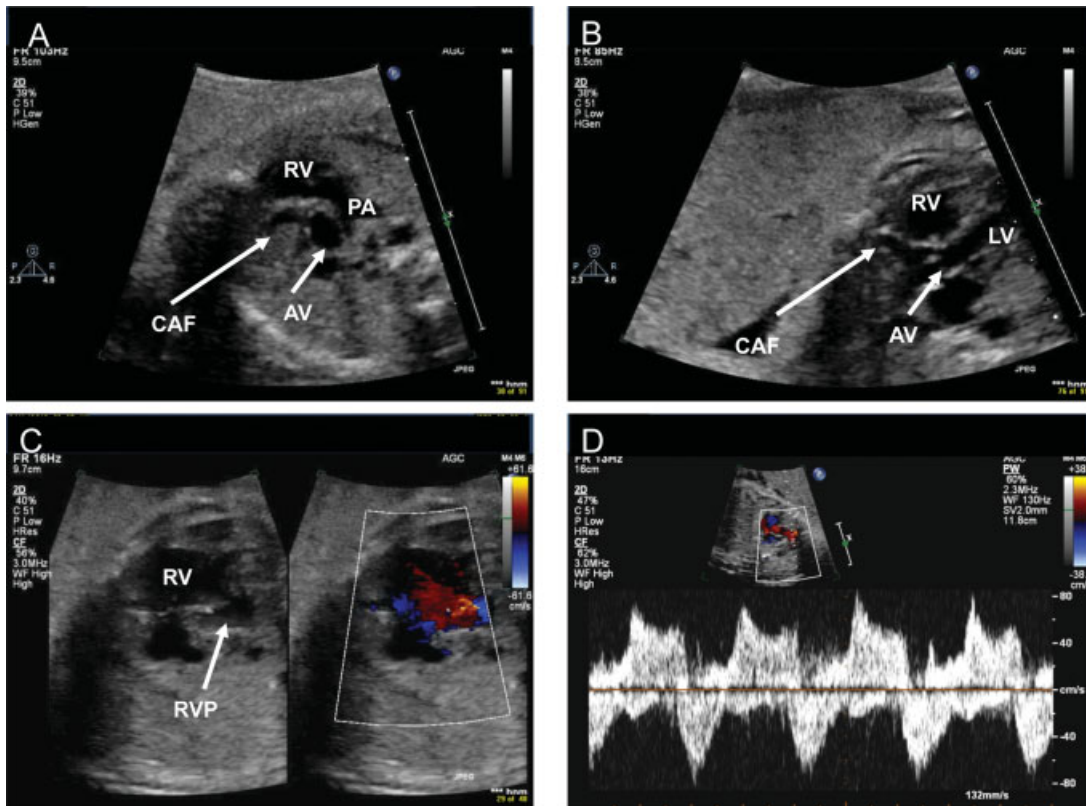


Fig. 1 Fetal images: (A and B) CAF in short axis and apical five chamber view. (C) RVP close to the septal side of tricuspid valve where the CAF was draining. (D) The diastolic flow reversal in the descending aorta. CAF, coronary artery fistula; LV, left ventricle; PA, pulmonary arteries; RV, right ventricle; RVP, right ventricle pouch.

the aortic valve and ascending aorta in systole and no aortic valve insufficiency. The aortic isthmus appeared borderline hypoplastic with a z-score of -1.8 . There was mild cardiomegaly, mild right atrial, and RV dilation (RV major diameter z-score of $+2$) and mild tricuspid valve regurgitation. Biventricular systolic function was normal and there was no evidence of pericardial effusion. The umbilical artery (UA) Doppler pattern was normal with a normal pulsatility index (PI) of 1.1 . The middle cerebral artery (MCA) Doppler pattern was normal with a low normal PI of 1.4 . The ratio of MCA PI to

UA PI was normal at 1.4 . Umbilical venous Doppler patterns were normal. On follow-up echocardiogram, there was further increase in the size and tortuosity of the RCA and CAF. There was a significant diastolic run-off without ventricular dyskinesia. A male fetus was delivered at 37 weeks gestation via cesarean section for the spontaneous onset of labor and breech presentation, with a birth weight of 2.8 kg. On physical examination, he had facial features typical of Down syndrome. His vitals were stable with a heart rate of 156 bpm, respiratory rate of 36 bpm, oxygen saturations of 99% on room

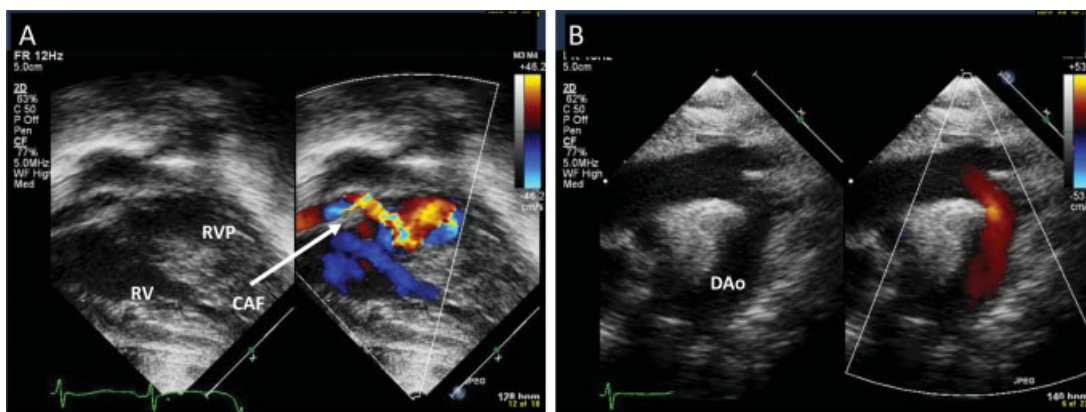


Fig. 2 Neonatal images: (A) Apical four chamber view of fetal echocardiogram showing large CAF arising from the right coronary artery and draining in to RVP. (B) The diastolic flow reversal in the DAo. CAF, coronary artery fistula; Dao, descending aorta; RV, right ventricle; RVP, right ventricle pouch.

air, and upper and lower limb blood pressures of 65/35 and 68/38, respectively. A postnatal echocardiogram confirmed an aneurysmally dilated RCA with a CAF draining into the RV posterior wall close to the tricuspid valve (►Fig. 2A). The RV was mildly dilated and hypertrophied. There was mild tricuspid valve regurgitation. There was significant flow reversal during diastole in the proximal descending aorta with no evidence of coarctation of aorta (►Fig. 2B). There was a small patent ductus arteriosus with left to right shunt. The rest of cardiac anatomy was normal as was biventricular systolic function. Electrocardiogram performed on day 1 of life showed no significant ST–T wave changes or chamber hypertrophy. However, there were frequent premature ventricular complexes.

On the second day of life, a cardiac catheterization procedure was performed which confirmed presence of a distal RCA fistula to RV with RCA diameter of 5 mm. A Flipper coil (5 mm × 4 loops) was placed in the RCA just proximal to the drainage site. Angiography of RCA showed small residual flow, for which no further intervention was attempted in view of concerns of proximal occlusion of the coronary artery. The neonate tolerated the procedure well without complications. The patient was started on intravenous heparin drip for 24 hours and then transitioned to antiplatelet treatment with aspirin and Plavix (Bristol-Myers Squibb, New York, NY). Follow-up echocardiogram on the day following the procedure showed improved forward flow in the descending aorta with decreased RV size. At latest follow-up at 6 months of age, the infant remained asymptomatic and the echocardiogram showed small residual flow across the distal end of fistula with normal biventricular size and function.

Literature Review

Embryologically, the coronary vessels are formed as a result of fusion of intramyocardial vascular islands and microvessels from the aortic root. CAFs may occur in isolation or in association with outflow tract obstructive lesions such as pulmonary valve atresia with intact ventricular septum. In these conditions, chronic fetal hypoxemia and/or elevations in afterload are thought to stimulate development of collateral myocardial vessels that evolve into fistulae.

The clinical presentation of congenital CAF is variable and depends upon the size and site of drainage. Small fistulas can be asymptomatic and may present as a murmur or may be diagnosed incidentally during an echocardiogram. CAFs that drain to the right side of the heart are associated with more symptoms than similar sized CAFs draining to the left. Larger fistulas may manifest as congestive heart failure in early infancy and secondary pulmonary hypertension. With the drop in pulmonary vascular resistance in the weeks after birth, CAFs may result in left to right shunt from the high systemic vascular pressure circuit to the lower pressure pulmonary vessels. The majority of patients presenting in the second and third decade do so with exertional dyspnea or angina due to coronary artery steal. Other reported complications in association with CAF include bacterial endocarditis, arrhythmias due to enlarged right atrium or secondary to

Table 1 Showing the demographics and outcome of the prenatally diagnosed coronary artery fistula in last couple of decades

Study	Origin	Drain	GA	Clinical presentation	Fistula size	Doppler velocity	Coronary size	Qp/Qs	PAP	Intervention age mode	Outcome
Sharland et al, 1996	RCA	RV	20 wk	CHF	3 mm	1.8 m/s	4 mm	N/A	85/47	5 mo	Ventricular fibrillation death
Cotton, 2000	LAD	RV	30 wk	Asymptomatic	N/A	N/A	N/A	N/A	N/A	None	Spontaneous closure
Khan et al, 2003	RCA	RA	22 wk	CHF	4 mm	N/A	N/A	3:1	N/A	3 Wk	Fistula closure
Mielke et al, 2002	LCX	RA	23 wk	Asymptomatic	N/A	2.3 m/s	2 mm	N/A	N/A	9 d	Fistula closure
Hung et al, 2006	RCA	RV	20 wk	CHF	3.8 mm	N/A	N/A	N/A	N/A	12 d	Coil snared Amplatzer device
Karagöz et al, 2008	RCA	RV	33 wk	CHF	6 mm	N/A	N/A	3.2:1	N/A	1 d	Fistula closure
Daniel et al, 2010	RCA	RV	23 wk	CHF	6 mm	N/A	N/A	N/A	N/A	4 d	Fistula closure
Hayashi et al, 2012	RCA	RV	33 wk	CHF	4 mm	3 m/s	5 mm	1.56:1	MAP 50	4 mo	Fistula closure
Zhao et al, 2012	RCA	RV	37 wk	CHF	5 mm	2.6 m/s	9 mm	N/A	MAP 96	6 mo	Fistula closure

Abbreviations: CHF, congestive heart failure; GA, gestational age; LAD, left anterior descending; LCX, left circumflex; PAP, pulmonary artery pressure; MAP, mean artery pressure; N/A, not applicable; RCA, right coronary artery; RV, right ventricle.

steal, thromboembolic events, and premature arteriosclerotic changes within the fistulae.¹⁰ In the last few decades, nine cases of prenatally diagnosed isolated CAF have been reported (► **Table 1**). Seven arose from the RCA and manifested as congestive heart failure whereas the two left-sided CAF were asymptomatic. The earliest reported diagnosis of fetal CAF was at 20 weeks gestation and the latest was at 37 weeks. Fistula size varied between 3 mm and 6 mm with a reported increase in Qp:Qs up to 3.2:1. In the absence of aortic valve regurgitation, significant diastolic run-off from the transverse and ascending aorta on fetal echocardiogram should raise the suspicion of CAF. The association between Down syndrome and ventriculocoronary fistula has been previously reported in a few cases.

The coronary artery from which the CAF arises and the CAF may develop tortuosity and dilation progressively over time because of the excessive flow, increasing the risk of thromboembolism, and coronary artery steal in adults. Therefore, most cardiologists would consider early intervention at the time of diagnosis even in an asymptomatic CAF to prevent complications. Antenatal diagnosis of CAF offers the possibility of early closure to potentially obviate complications. The median age of intervention in the nine antenatally diagnosed CAF cases was 16.5 days (range, 1 day–6 months). Intervention in our case was one of the earliest reported at 2 days of age. The various therapeutic options are surgical ligation or closure by a device or coil in the cardiac catheterization laboratory. Surgical options include either closing the fistula internally from the receiving chamber side or even ligating the aneurysmal coronary artery. Mortality from surgical closure ranges from 0 to 4%. There are reports of myocardial infarction and recurrence of CAF with surgical treatment. Recent reports suggest that transcatheter closure of CAF may have efficacy and safety comparable to surgical treatment. In the reviewed literature, surgery was the modality of management in three patients, percutaneous catheter closure was used in five patients and in one patient, spontaneous closure occurred. Percutaneous closure methods included coil placement in three patients, Amplatzer duct occluder (AGA Medical under St. Jude medical, Plymouth, MN) in one patient and Amplatzer vascular plug (AGA Medical under St. Jude medical, Plymouth, MN) in another. Our case was unique in that a single Flipper coil successfully achieved partial closure of the CAF. As thrombosis and unintended occlusion of coronary artery branches proximal to the closure of a large distal CAF are concerns, partial closure may be a reasonable approach to mitigate the hemodynamic consequences, while minimizing risks.¹¹

In conclusion, our case and the reviewed literature highlight the importance of antenatal detection of CAF to plan

early transcatheter or surgical intervention. Early intervention of CAF appears efficacious and safe and is associated with good intermediate term outcomes.

Notes

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None.

Conflict of Interest

None.

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