

The clinical characteristics of coronary artery fistula anomalies in children and adults: A 24-year experience

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Abstract

Objectives: The aim of our work is to investigate the clinical characteristics of coronary artery fistula (CAF) anomalies in South Vietnam.

Methods: This is a retrospective analysis of 119 patients with diagnosis of definite CAF between January 1992 and April 2016. The demographic, clinical, echocardiographic, and angiographic characteristics and management of CAF with short-term outcomes are described.

Results: The median age was 15 years (range, 1-79 years), with 49 male (41%) and 70 female (59%). There were 77 symptomatic patients (64.7%) and 91 patients (76.5%) who presented with a murmur. The electrocardiogram was abnormal in 45.4% and cardiac enlargement or increased pulmonary vasculature were seen in 76 patients (63.9%) on chest X-ray. The sensitivity of echocardiography for CAF diagnosis was 79%. The source of the fistula was most often from the RCA (54%), most commonly to right atrium (34.5%) or right ventricle (31.1%). In comparison with surgery, transcatheter closure had a shorter hospital length of stay (5.4 ± 3.8 days vs 12.6 ± 6.5 days, $P = .02$) and better postprocedural left ventricular ejection fraction ($67.9 \pm 8.1\%$ vs $62.9 \pm 6.0\%$, $P = .03$).

Conclusion: The majority of fistula in this study originated from the RCA and terminated in the right atrium or the right ventricle. Transcatheter and surgical closure are both relatively safe and effective, with the potential for shortened length of hospital stay following transcatheter closure.

KEYWORDS

coronary artery fistula, clinical, echocardiographic and angiographic characteristics, surgical closure, transcatheter closure

1 | INTRODUCTION

Coronary artery fistula (CAF) is a rare anomaly of the cardiovascular system, defined as an abnormal connection between a coronary

artery and a cardiac chamber or vessel. CAFs are observed in 0.002% of the general population¹ and account for 0.2%-0.4% of all congenital heart disease cases.^{2,3} The most common symptoms and complications of CAF include dyspnea, myocardial ischemia, arrhythmia, congestive heart failure, pulmonary hypertension, infective endocarditis, and aneurysm rupture.^{4,5} Surgical or transcatheter

Huynh and Truong contributed equally to this manuscript.

intervention is recommended in patients who have symptoms, unexplained ventricular systolic or diastolic dysfunction or enlargement, or a large fistula regardless of symptomatology.⁶ We report the demographic, clinical, echocardiographic, and angiographic characteristics of CAF in patients encountered over a 24-year period and compare the short-term outcomes between surgical and transcatheter closures.

2 | MATERIALS AND METHODS

2.1 | Study design

This study was performed at The Heart Institute and Tam Duc Hospital of Cardiology, which are tertiary care referral hospitals located in Ho Chi Minh City, South Vietnam. The hospital charts of patients admitted with the diagnosis of a coronary artery fistula between January 1992 and April 2016 were retrospectively reviewed. Two-dimensional echocardiography was performed in the parasternal short-axis view of the aorta to visualize the diameter of the left and right coronary arteries. Color Doppler was performed from the parasternal, apical, and subcostal views to identify the high-velocity flow from the coronary artery through its fistulous connection into the cardiac chamber or vessel. A total of 119 consecutive patients with diagnosis of definite CAF were eligible for inclusion. This study was approved by the institutional review boards (IRB) of the two participating hospitals. Informed consent was waived because of the retrospective nature of the study.

2.2 | Statistical analysis

Continuous variables are expressed as mean \pm standard deviation (SD) for normal distributions and median + interquartile range for nonnormal distributions. Categorical variables were represented as frequencies and percentages. For the evaluation of qualitative variables, we used the Chi-Square test. To test for significant differences between continuous variables in two groups, independent sample *t*-tests were performed. Statistical analysis was performed using the SPSS 22 software program (SPSS Inc., Chicago, IL, USA). A *P* value of $<.05$ was considered statistically significant.

3 | RESULT

3.1 | Baseline characteristics

Baseline characteristics and clinical findings on admission for the 119 CAFs cases were shown in Table 1. The median age of patients was 15 years (range, 1-79 years), with 49 male (41%) and 70 female (59%). There were 77 patients (64.7%) who presented with symptoms, and 91 (76.5%) who presented with a murmur, most commonly being a prominent continuous murmur. Interestingly, a murmur was not apparent in up to 24% patients (Table 1). Pulmonary hypertension, severe heart failure, valve regurgitation, and atrial flutter or fibrillation were the most common

TABLE 1 Characteristics of study sample

Clinical characteristics	<i>n</i> = 119
Age	15 (1-79)
Female	70 (58.8)
Symptom	
Asymptomatic	42 (35.3)
Chest pain	22 (18.5)
NYHA I-II	32 (26.9)
NYHA III-IV	13 (10.9)
Others ^a	10 (8.4)
Murmur	
No	28 (23.5)
Continuous murmur	56 (47.1)
Systolic murmur	31 (26.1)
Diastolic murmur	4 (3.4)
ECG	
Sinus rhythm	65 (54.6)
Atrial flutter/fibrillation	13 (10.9)
Other abnormality ^b	41 (34.5)
Chest X-ray	
Normal	43 (36.1)
Abnormal	76 (63.9)
Cardiac enlargement	43 (36.1)
Cardiac enlargement and increased pulmonary vasculature	33 (27.7)

Note: Continuous variables are expressed as median and interquartile range. Categorical variables are presented as *n* (%).

^aPalpitation, presyncope, tachyarrhythmia, infective endocarditis.

^bRight ventricular hypertrophy, left ventricular hypertrophy, right bundle branch block, T wave inversion > 2 leads, ST segment depression > 0.5 mm at least 2 leads, premature atrial or ventricular complex.

complications (Table 2). Most patients with symptoms, complications, or electrocardiographic abnormalities were 20 years of age and older, while a continuous murmur was most frequent in those less than 20 years of age (Table 3).

3.1.1 | Electrocardiogram and chest X-ray

The electrocardiogram was normal in 54.6%. Left or right ventricular hypertrophy, right bundle branch block, T wave inversion, and ST segment depression were present in 34.5% of all cases, while atrial flutter or fibrillation accounted for 10.9% (Table 1). Evidence of cardiac enlargement or increased pulmonary vasculature was seen in 76 patients (63.9%) on chest X-ray (Table 1).

3.1.2 | Imaging

Transthoracic echocardiography was performed in all patients. The sensitivity of echocardiography for CAF diagnosis was up to 79% (Table 4). The mean dimension of the coronary artery origin was

TABLE 2 Complication of coronary artery fistula in the study sample

Complication	n = 119
Severe HF, NYHA III-IV	13 (10.9)
Atrial flutter/fibrillation	13 (10.9)
Significant valvar regurgitation	14 (11.8)
Endocarditis	3 (2.5)
Coronary aneurysm	11 (9.2)
Rupture of coronary aneurysm	1 (0.8)
Thrombus in coronary aneurysm	3 (2.5)
Death	1 (0.8)
Pulmonary hypertension	36 (30.3)

Note: Categorical variables are presented as n (%).

Abbreviations: HF, heart failure; NYHA, New York Heart Association.

11.6 ± 4.9 mm with a left ventricular ejection fraction of 65 ± 7%. Patients above 20 years of age had a large dimension of the coronary artery origin compared to those aged 20 or under, although not statistically significant (12.8 ± 5.3 mm vs 11.00 ± 4.67 mm, *P* = .244). Thirty-one patients (25%) underwent multidetector computed tomography (MDCT) and 57 patients (48%) underwent coronary angiography. The entry and exit points of CAF varied with the right coronary artery (RCA) involved in 64 patients (54%) and the

TABLE 3 Comparison in two age subgroups

	Age < 20	Age > 20	<i>P</i>
	(n = 65)	(n = 54)	
Male	30 (46.2)	19 (35.2)	.31
Symptomatic	25 (38.5)	52 (96.3)	<.001
Murmur	55 (84.6)	35 (64.8)	.08
ECG abnormality	25 (38.5)	29 (53.7)	<.001
CXR abnormality	46 (70.8)	30 (55.6)	.04

Note: Categorical variables are presented as n (%).

TABLE 4 Echocardiographic characteristics in 119 patients with CAF

Sensitivity	94 (79%)
Dimension of the coronary artery origin (mm)	11.6 ± 4.9
Coronary artery/aorta ratio	0.58 ± 0.21
LVEF (%)	65 ± 7
Mean PAP (mm Hg)	33.5 ± 9.1
Proportion of pulmonary hypertension	36 (30.3)
Abnormal wall motion	7 (5.9%)
Cardiac enlargement	89 (74.8%)

Notes: Continuous variables are expressed as mean ± standard deviation. Categorical variables are presented as n (%).

Abbreviations: LVEF, left ventricular ejection fraction; PAP, pulmonary artery pressure.

left coronary artery involved in 48 patients (40%). Seven patients (6%) had involvement of both coronary arteries. The most common fistula position was the right atrium (34.5%) or right ventricle (31.1%), mostly commonly involving a single fistula (Tables 5 and 6, Figure 1). Thirteen patients (10.9%) had two or more fistulas and 9% of patients were associated with other forms of congenital heart disease, including atrial septal defect, ventricular septal defect, or a patent arterial duct (Table 5).

3.1.3 | Treatment

Conservative medical management was elected in 22 patients (18.5%). Eighty-six patients (72.3%) underwent surgical closure and 11 patients (9.2%) had transcatheter closure (Table 7). In comparison with surgery, hospital length of stay was shorter in patients following transcatheter closure. Although postprocedural left ventricular ejection fraction was better in patients following transcatheter closure, both groups had a normal average postprocedural value (Table 8). There was one death in the surgery group due to aneurysmal rupture.

TABLE 5 Characteristics of coronary artery fistula in the study

Characteristics	n = 119
Coronary artery fistula	
Right coronary artery	64 (53.8)
Left coronary artery	48 (40.3)
LM, LAD	37 (31.1)
LCx	11 (9.2)
Both right and left coronary artery	7 (5.9)
Fistula position	
Right atria	41 (34.5)
Right ventricle	37 (31.1)
Left atria	3 (2.5)
Left ventricle	15 (12.6)
Pulmonary trunk	20 (16.8)
Coronary sinus	3 (2.5)
Number of fistula	
1 hole	106 (89.1)
≥2 holes	13 (10.9)
Other heart defects	
Other congenital heart disease ^a	11 (9.2)
Valvular heart disease, coronary stenosis	13 (10.9)
Coronary aneurysm	11 (9.2)
Thrombus in coronary aneurysm	3 (2.5)
Endocarditis	3 (2.5)

Note: Categorical variables are presented as n (%).

Abbreviations: LAD, left anterior descending; LCx, left circumflex; LM, left main.

^aAtrial septal defect, ventricular septal defect and patent ductus arteriosus.

TABLE 6 Positional characteristics of coronary artery fistula in the study

	Right coronary artery (n = 62)	Left coronary artery (n = 50)	Left and right coronary artery (n = 7)
Right atria	22 (35.5)	19 (38)	0 (0)
Right ventricle	30 (48.4)	7 (14)	0 (0)
Left atria	0 (0)	2 (4)	1 (14.3)
Left ventricle	8 (12.9)	7 (14)	0 (0)
Pulmonary trunk	1 (1.6)	13 (26)	6 (85.7)
Coronary sinus	1 (1.6)	2 (4)	0 (0)

Note: Categorical variables are presented as n (%).

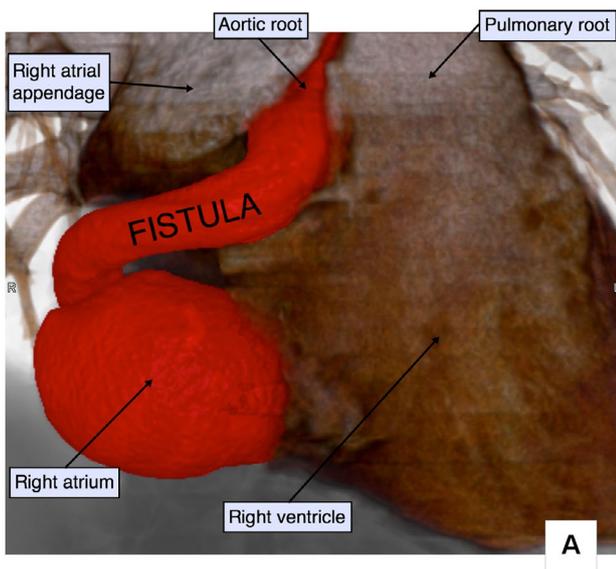


FIGURE 1 This 3D endocast computed tomography reconstruction demonstrates the most common coronary arterial fistula, communicating from the proximal right coronary artery to the right atrium. The aortic root, fistulous connection, and right atrium are highlighted in red. The portion of the right atrium between the right atrial appendage and the distal right atrium opacified from the fistulous connection is poorly visualized due to the differences in opacification

4 | DISCUSSION

Approximately two-thirds of patients presenting with CAF were symptomatic at presentation, although most patients under 20 years of age were generally asymptomatic. Furthermore, dyspnea was more common in the younger cohort, while chest pain presented more commonly in the older cohort. This is consistent with the CAF cohort reported by Liberthson et al.⁷ In their study of 13 new and review of 174 previously reported patients, only 19% of those under 20 years of age had preoperative symptoms, in contrast to 63% of those aged 20 years or over. In the reports by Song et al and Lo et al of pediatric-aged patients, 84% and 74% patients had no symptoms,

TABLE 7 Characteristics of treatment in the study sample

	n = 119
Treatment method	
Surgery	86 (72.3)
Transcatheter closure	11 (9.2)
Conservative therapy	22 (18.5)
Hospital lengths (days)	12 ± 6.5
Residual fistula	8 (6.7)
In-hospital mortality	1 (0.8)

Notes: Continuous variables are expressed as mean ± standard deviation. Categorical variables are presented as n (%)

TABLE 8 Comparison of surgical and transcatheter therapy

	Transcatheter intervention (n = 11)	Surgery (n = 86)	P
Hospital length of stay (days)	5.4 ± 3.8	12.6 ± 6.5	.02
Preoperation LVEF (%)	66.5 ± 5.9	65.5 ± 6.7	.65
Postoperation LVEF (%)	67.9 ± 8.1	62.9 ± 6.0	.03
Postoperation PAP (mm Hg)	26.4 ± 4.2	27.3 ± 5.5	.69
Residual fistula (%)	2 (18.2)	6 (7.0)	.42
Unsuccessful technique (%)	1 (9.1)	1 (1.2)	.49
Complication ^a (%)	1 (9.1)	6 (7.0)	.72
In-hospital mortality (%)	0 (0)	1 (1.2)	.73
Reintervention (%)	1 (9.1)	4 (4.6)	.46

Notes: Continuous variables are expressed as mean ± standard deviation. Categorical variables are presented as n (%).

Abbreviations: LVEF, left ventricular ejection fraction; PAP, pulmonary artery pressure.

^aVentricular arrhythmia, infection, pericardium effusion, coronary artery rupture, myocardial infarction.

respectively.^{8,9} On the other hand, in the reports by Albeyoglu et al and Abdelmoneim et al of 21 adult patients, the majority were symptomatic.^{10,11} This is likely explained by the inclusion of older patients.

Similarly, the majority of patients presented with a murmur on examination (continuous murmur in nearly half), which was more common in the younger cohort. Lo et al and Albeyoglu et al both reported similar proportions of patients presenting with a murmur, 77.8% and 85.7%, respectively.^{8,10} Half of their patients presented with electrocardiographic abnormalities and 64% with cardiac enlargement or increased pulmonary vascular on chest X-ray, similar to prior reports and the present series.⁷ Older patients tend to have a larger dimension of the coronary artery in CAF that may in part be explained by the fact that young patients with a large shunt are more likely to undergo intervention at the time of admission. Patients with a small shunt who are asymptomatic will have conservative medical

management and follow-up. The fistulas may increase in size over time, although this does not occur invariably and the fistula pattern might affect the fistula size since larger shunts occur with CAF to the right side rather than the left side of the heart.^{6,12}

In our study, the source of the fistula was most often from the RCA with the left main coronary artery and its branches being much less involved. The most common fistula pattern was from RCA to right ventricle or right atrium, consistent with prior studies.¹³⁻¹⁵ However, other studies have suggested that the majority of fistulas originate from the left main coronary artery or its branches and terminate in the right atrium, right ventricle or pulmonary trunk.^{10,16} These differences may be related to a smaller sample size or differences between varying ethnic populations.

Our study observed comparable rates of successful fistula closure between transcatheter and surgical procedures, although the residual fistula rate appeared higher (difference not statistically different) in the transcatheter group. Both closure strategies had a low rate of post-procedure complications and mortality. Not surprisingly, transcatheter closure had a shorter hospital length of stay and was associated with better postoperative left ventricular ejection fraction when compared to surgical closure. Transcatheter closure of CAF has become a feasible and effective alternative therapy to surgical closure with good outcome in previously published studies,¹⁷⁻¹⁹ with good long-term results.^{13,15,20} Although we did not assess long-term outcomes, our study supports the relative efficacy and short-term safety of both transcatheter and surgical closure.

4.1 | Limitations

This is a retrospective study of a relatively small sample size, and does not evaluate the natural history or long-term treatment results. However, our aim was to better characterize the phenomenon in the Southeast Asian population. This study is also limited in that the data were collected from patients seen at two tertiary care hospitals in South Vietnam which are large referral centers for congenital heart disease, and may not be representative of other populations. However given the low prevalence of the condition, a focused patient stream was necessary to study a meaningful number of patients.

4.2 | Conclusion

CAF is a relatively rare coronary congenital anomaly, with patients commonly coming to clinical attention following auscultation of a murmur or presenting with symptoms. The majority of fistula in this study originated from the right coronary artery and terminated in the right atrium or the right ventricle. Transcatheter and surgical closures are both relatively safe and effective, with the potential for shortened length of hospital stay following transcatheter closure.

CONFLICT OF INTEREST

The authors declare that they have no conflicts of interest with the contents of this article.

AUTHOR CONTRIBUTIONS

Concept/design: Huynh, Truong

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