Surgical Outcome in Coronary Artery Fistula Repair in Children

Mohammadreza Malek Ahmadi [1*] MD, Akbar Shahmohammadi[2] MD, and A. Hussein Tabatabaei,[3] MD

Abstract

Background- Coronary artery fistula (CAF) is a rare congenital anomaly that can be complicated by endocarditis, myocardial infarction or coronary aneurysms. The purpose of this article is to review the clinical characteristics and surgical outcome in 10 patients with CAF.

Methods- From 1990 to 2000, ten patients (aged 6 months to 15 years with a mean of 8.5 years) were diagnosed with CAF via echocardiography and cardiac catheterization. Six were female and 4 were male. All the patients with isolated CAF (9) were asymptomatic. One patient with associated anomaly (MVP with severe MR) had dyspnea on exertion and palpitation.

Results- Five fistulas originated from the right coronary artery, three from the left and two from the left circumflex. Drainage was to the right ventricle (7), right atrium (2) and pulmonary artery (1). The ratio of pulmonary to systemic flows ranged between 1 to 1.6. All the patients had surgical ligation. In the symptomatic patient, in addition to ligation, mitral valve replacement was performed. There was no operative or late death. Follow-up evaluation ranging from 1 year to 6 years with a mean of 4.2 years showed no evidence of recurrent or residual CAF.

Conclusion- Surgical management of CAF is a safe and effective treatment, resulting in 100% closure rate ***(Iranian Heart Journal 2003; 4 (4):35-38).***

Key words: Coronary artery fistula ■ Surgical ligation ■ Outcome

Symptomatic coronary artery fistulas (CAF) have been associated with substantial morbidity and mortality all ages.[1-6] CAFs that have not been detected or closed in childhood have been reported to become symptomatic in adulthood because of chronic volume load and ischemia.[1] Incidental detection of CAF in asymptomatic patients has been noted on coronary angiography, but its clinical significance is unclear.[7] Although some authors have recommended the elective closure of CAF regardless of symptoms,[1] others have advocated conservative management.[8]

The present study determines the clinical characteristics and surgical outcome in children with CAF.

Patients and Methods

Patients

The chart databases were reviewed for all patients with the diagnosis of CAF noted on echocardiography or at catheterization from 1990 to 2001.

1) Assistant Professor of Pediatric Cardiology, Shahrekord University of Medical Sciences
2) Associate Professor of Pediatrc Cardiology, Shaheed Rajaie Cardiovascular Medical Center
3) Associate Professor of Pediatrc Cardiology , Dr. Shariati Hospital, Tehran University of Medical Sciences.
From the Pediatic Department, Hajar Hospital, Shahrekord University of Medical Sciences, Shahrekord, Iran,
Correspondence to: M. Malek Ahmadi, MD, Pediatic Department, Hajar Hospital, Shahrekord University of Medical Sciences, Shahrekord, Iran
email: Mr_Malekahmadi@Yahoo.Com Tel: 0311-2337247, O913-2088153, Fax: 0381-2243715

The study included patients who had isolated CAF without any other important cardiac anomaly.

Patients with CAF or sinusoids associated with congenital cardiac malformation, such as pulmonary atresia with intact ventricular septum, or mitral stenosis and aortic atresia were excluded. Patients in whom CAF were detected following intracardiac surgery were also excluded. Ten patients met the inclusion criteria. Patients' charts were reviewed for demographics, symptoms, clinical findings, indications for echocardiography, associated diagnosis and radiologic and ECG findings at presentation. At follow-up, evidence of myocardial ischemia, congestive heart failure and arrythmias were sought clinically and by means of electrocardiography. After the procedures had been performed, surgical and cardiac catheterization reports were reviewed.

Echocardiography

Echocardiograms were performed using available cardiac scanners with transducer frequency and focus appropriate for the patient's size. The origin, course and exit site of the fistula were recorded for each patient. Coronary artery diameter was determined and considered normal if the measurements were within 2SD of normal controls.

Cardiac catheterization

Cardiac catheterization data and angiography were reviewed when available. The origin, course and exit site of the fistula, right and left ventricular end diastolic pressures, PA pressure, pulmonary to systemic flow ratio and assessment of LV function were recorded.

Results

Ten patients had an incidental finding of a small coronary artery fistula detected on echo and angiography. The salient demographic, clinical, echocardiographic and outcome data are summarized in Table I. The mean age at diagnosis was 8.5 years (range 6 months to 15 years). The primary indication for echocardiography at presentation was murmur in 9 patients. Dyspnea on exertion and palpitation in addition to murmur was seen in one patient. The latter patient had mitral valve prolapse (MVP) and severe mitral regurgitation (MR) in addition to CAF. A systolic murmur was audible in 6 patients and was assessed as innocent in all. A continous murmur, which was interpreted as a small patent ductus arteriosus, was audible in 4 patients. None of the patients had symptoms suggestive of angina or congestive heart failure. Associated cardiac abnormality was present in one patient (Table I). Cardiomegaly was present radiographically in 4 patients. At presentation, 2 patients had electrocardiographic criteria for biventricular hypertrophy, one patient for LVH and one patient for RVH. None of the patients had abnormal Q wave or ST-segment or T wave changes suggestive of ischemia on electrocardiogram.

The origin of the coronary artery fistula was clearly defined by color Doppler and angiography in 10 patients. The origin of the fistula was from the right coronary artery system in five patients; from the left coronary artery in 3 patients; and from the left circumflex in 2 patients. Coronary artery dimensions were normal in 10 patients. The fistula drained into the right ventricle in 7 patients, right atrium in 2 patients and pulmonary artery in one patient. Cardiac catheterization was performed in 10 patients. The pulmonary to systemic flow ratio ranged from 1 to 1.6. All the patients had surgical ligation of CAF via cardiopulmonary bypass and midsternotomy approach. All the patients

had follow-up from 1 to 6 years (mean 4.2 years). None of the patients had evidence of recurrent or residual fistula.

Discussion

CAF is a rare anomaly and constitutes 0.2% to 0.4% of congenital heart diseases. Of the 10 patients who had undergone surgery, 6 were female and 4 were male. In a study from Thailand, a female preponderance has also been observed.[9]

The age of the patients was between 6 months and 15 years old. In a report from Wong et al., age distribution is from 2 days to 16 years.[10]

In this study, the most common indication for echocardiography, similar to that in Sherwood's study,[11] was heart murmur. Most of the patients of our study, like those in the study of Sherwood[11] and Wang,[12] were asymptomatic. Moreover, isolated forms of CAF, as reported in the study of Wang[12] and Sunder,[13] was the most common form. Symptomatic forms were seen more often in patients older than 20 years.[9,12,13]

In electrocardiography, the most frequent changes were ventricular hypertrophy as noted by other studies,[9] and the most common sign in chest radiography was cardiomegaly as observed by Thongtang.[9]

In the present study, like other studies,[9,10,14] the origin of the fistula was from the RCA in 50% and from the left coronary artery system in 50%. The exit site in decreasing order of frequency was the right ventricle, right atrium and pulmonary artery, respectively, as stated by some reports.[12,13,14] However, in some reports, the pulmonary artery is the most common exit site.[9,11] The range of QP/QS in this report is 1-1.6, whereas in Thongtang's report it is 1.2-1.6.[9]

Surgery with cardiopulmonary bypass was performed on all our patients. The mortality rate was zero, and no residual shunt was found before patients were discharged from the hospital.

No clinical symptoms were found in our patients during a follow-up that ranged from 1 to 6 years (mean 4.2 years), like other reports.[9,12]

Table I: Clinical, paraclinical findings and outcome data in 10 patients with CAF.

Pt. No	Gender	Age (yr)	Reason for echo	Cardiomegaly on CXR	EKG	Associated cardiac anomaly	Fistula origin	Exit site	QP/QS	Follow up (yr)
1	F	9	CMM	0	NL	0	RCA	RV	1.2	3.9
2	F	3.5	CMM	+	RVH	0	LCX	RA	1.3	1.0
3	F	13	SMM	0	NL	0	RCA	RV	1.4	4.2
4	F	5	SMM	+	BVH	0	LCA	RA	1.3	2.4
5	F	0.5	SMM	+	BVH	0	LCX	RV	1.6	7.5
6	F	15	SMM & DOE	0	LVH	MR, MVP	LAD	PA	1.1	5.5
7	M	6	CMM	0	NL	0	LCA	RV	1.0	5.5
8	M	14	SMM	0	NL	0	RCA	RV	1.5	4.0
9	M	7	SMM	+	NL	0	RCA	RV	1.3	6.0
10	M	12	CMM	0	NL	0	RCA	RV	1.4	5.0

BVH: Biventricular hypertrophy, **MR:** mitral regurgitation , **CMM:** Continous murmur , **MVP:** mitral valve prolapse, **DOE:** Dyspnea on exertion , **NL:** normal, **F:** female, **PA:** pulmonary artery, **LCA:** Left coronary artery, **RCA:** Right coronary artery, **LCX:** left circumflex, **RV:** Right ventricle, **LVH:** left ventricular hypertrophy, **RVH:** Right ventricular hypertrophy, **M:** male, **SMM:** systolic murmur

Conclusion

Early and proper surgical management of CAF is a safe and effective treatment, resulting in 100% closure rate.

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