Pediatr Cardiol 26:328–330, 2005
DOI: 10.1007/s00246-004-0656-y

Pediatric Cardiology

Surgical Outcome of Coronary Artery Fistulas Repair in Children

M. Malekahmadi,[1] **A. Shahmohammadi**[2]

[1]Pediatic Department, Hajar Hospital, Shahrekord University of Medical Sciences, Shahrekord, Iran
[2]Pediatric Cardiology Department, Iran University of Medical Sciences, Tehran, Iran

Abstract. Coronary artery fistula (CAF) is a rare congenital anomaly that can be complicated by endocarditis, myocardial infarction, or coronary aneurysms. This article reviews the clinical characteristics and surgical outcome of 10 patients with CAF in Rajae Heart Center. From 1990 to 2000, 10 patients (age, 6 months to 15 years; mean age, 8.5 years) were diagnosed with CAF by echocardiography and cardiac catheterization. Six were female and 4 were male. All patients with isolated CAF (9) were asymptomatic. One patient with associated anomaly (mitral valve prolapse with severe mitral regurgitation) had dyspnea on exertion and palpitation. 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 flow ranged between 1 and 1.6. All 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 (range, 1–6 years; mean, 4.2 years) showed no evidence of recurrent or residual CAF. Surgical management of CAF is a safe and effective treatment resulting in 100% survival and clousure rate.

Key words: Coronary artery fistula — Surgical ligation

Symptomatic coronary artery fistulas (CAFs) have been associated with substantial morbidity and mortality at all ages [4, 6–8, 11, 12]. 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 [6]. Incidental detection of CAFs in asymptomatic patients has been noted on coronary angiography, but their clinical significance is unclear [9]. Although some authors recommend elective closure of CAFs regardless of symptoms [6], others advocate conservative management [3]. This study was performed to determine the clinical characteristics and surgical outcome of children with CAFs.

Correspondence to: M. Malekahmadi, *email:* mr_malekahmadi@yahoo.com

Patients and Methods

Patients

The chart databases of the Department of Cardiology, Shaheed Rajae Hospital, were reviewed for all patients with the diagnosis of CAF noted on echocardiography or at catheterization from 1990 to 2001. The study included patients who had isolated CAFs without any important other cardiac anomaly. Patients with CAFs 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 CAFs were detected following intracardiac surgery were also excluded. Ten patients met the inclusion criteria. Patients' charts were reviewed for demographic, symptoms, clinical findings, indication for echocardiography, associated diagnosis, and radiologic and electrocardiograph (ECG) findings at presentation. At follow up, evidence of myocardial ischemia, congestive heart failure, and arrythmias was sought clinically and with electrocardiography. When performed, surgical or cardiac catheterization reports were reviewed. Surgical repair was approached via a median sternotomy and cardiopulmonary bypass (CPB).

Echocardiography

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

Table 1. Clinical, and paraclinical findings and outcome data of 10 patients with CAF

Patient No.	Gender	Age (years)	Reason for echo	Cardiomegally on CXR	ECG	Associated cardiac anomaly	Fistula origin	Exit site	Q_p/Q_s	Follow-up (years)
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	M	12	SMM	0	NL	Q	RCA	RV	1.4	5.0
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	F	15	CMM, DOE	0	LVH	MR, MVP	LCA	PA	1.1	5.5

BVH, biventricular hypertrophy; CMM, continuos murmur; DOE, dyspnea on exertion;.F, female; LAD, left axis deviation; LCA, left coronary artery; LCX, left circumflex; LVH, left ventricular hypertrophy; M, male; MR, mitral regurgitation; MVP, mitral valve prolapse; NL, normal; PA, pulmonary artery; RA, right atrium; RCA, right coronary artery; RV, right ventricle; RVH, right ventricular hypertrophy; SMM, systolic murmur.

left ventricular end diastolic pressures, pulmonary artery (PA) pressure, pulmonary-to-systemic flow ratio, and assessment of left ventricular (LV) function were recorded.

Results

Ten patients had an incidental finding of a small CAF detected by echo and angiography. The salient demographic, clinical, echocardiographic, and outcome data are summerized in Table 1. 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 were noted in 1 patient. This patient, in addition to CAF, had mitral valve prolapse (MVP) and severe mitral regurgitation (MR). A systolic murmur was audible in 6 patients and was assessed as innocent in all. A continuous murmur, which was interpreted as a small patent ductus arteriosus, was audible in 4 patients. No patient had symptoms suggestive of angina or congestive heart failure. Associated cardiac abnormality was present in 1 patient (Table 1). Cardiomegaly was present radiographically in 4 patients. At presentation, 2 patients had electorcardiographic criteria for biventricular hypertrophy—1 patient for left ventricular hypertrophy and 1 patient for right ventricular hypertrophy. No patient had abnormal Q wave or ST segment or T wave changes suggestive of ischemia on EC.

The origin of the CAF was clearly defined by color Doppler and angiography in 10 patients. The origin of the fistula was from the right coronary artery system in 5 patients, from the left coronary artery in 3 patients, and from the left circumflex in 2 patient. Coronary artery dimentions, were normal in all patients. The fistula drained into the right ventricle in 7 patients, right atrium in 2 patients, and pulmonary artery in 1 patient. Cardiac catheterization was performed in all patients. The pulmonary-to-systemic flow ratio ranged from 1 to 1.6. All patients had surgical ligation of CAF by cardiopulmonary bypass and midsternotomy approach. All patients had follow-up from 1 to 6 years (mean, 4.2 years). None had evidence of recurrent or residual fistula.

Discussion

CAF is a rare anomaly and occurs in 0.2–0.4% of cases are congenital heart disease. There are new techniques for closing the CAF, such as transcatheter embolization techniques using coils, a detachable balloon, polyvinil foam, and a double-umberella device. The transcatheter approach is a fairly complicated intervention and requires an experienced operator and interventional specialist with expertise in both coronary arteriography and embolization techniques. Embolization often requires complicated catheter manipulation as well as selection of various catheters and wires. In the literature, there are only case reports and reports of small series [1, 5]. Although there was some experience with patent ductus arteriosus closure by coil and Amplatzer in our center, there was not any experience with CAF clousure using these devices, therefore, surgical treatment was performed in all of our patients.

Of the 10 patients who had undergone surgery, 6 were female and 4 were male. In a study from Thailand, a female preponderance had also been observed [14].

In this study, the most common indication for ECG, as in the Sherwood et al [10] study, was heart murmur. Most patients in our study, as in that of

Sherwood et al [10] and Wang et al [15], were asymptomatic and most included the isolated form of CAF, as in the study of Wang et al [15] and that of Sunder et al [13]. Symptomatic forms were seen more in patients older than 20 years [1, 10, 15].

Electrocardiography revealed that the most common changes were ventricular hypertrophy, as previously reported [14]. The most common finding in chest radiography was cardiomegaly, as reported by Thongtang [14].

In this study, the origin of the CAF was the right coronary artery in 50% of patients and the left coronary artery in 50% of patients, consistent with other studies [2, 14, 16], and the exit site in decreasing order of frequency was the right ventricle, right atrium, and pulmonary artery, as stated in other reports [2, 13, 15]. However, in some studies the common exit site was the pulmonary artery [10, 14]. The range of Q_p/Q_s in this report was 1–1.6, and that in Thohgtang et al.'s [14] report was 1.2–1.6.

All patients in this study were repaired surgically using CPB. There was no mortality, and no residual shunt was found at the time of discharge from the hospital.

No abnormal clinical findings, were encounter in outpatients during follow-up, which ranged from 1 to 6 years (mean, 4.2 years). This was consistent with other reports [14, 15].

Conclusion

Early and proper surgical management of CAF is safe and effective, resulting in 100% survival and closure rates.

References

1. Balanescu S, Sangiorgi G, Castelvecchio S, Medda M, Inglese L (2001) Coronary atery fistulas: clinical conequences and methods of closure. A literature review. *Ital Heart J 2*:669–676
2. Barbosa MM, Katina T, Oliveria HG, Neuenschwander FE, Oliveria EC (1999) Doppler echocardiographic features of coronary artery fistula: report of 8 cases. *J Am Soc Echocardiogr 12*:149–154
3. Hobbs RF, Millit HD, Raghavan PV, Moodie DS, Sheldon WC (1982) Coronary artery fistulae: a ten-year review. *Cleveland Clin Q 49*:191–197
4. Kovalchin JP, Grifka RG, McQuinn TC, Pignatelli RH, Geva T (1995) Right coronary artery to right atrium fistula mimicking a right atrial mass. *Am Heart J 130*:907–909
5. Latson LA, Forbes TJ, Cheatham JP (1992) Trancatheter coil embolization of a fistula from the posterior descending coronary artery to the right ventricle in a two-year-old child. *Am Heart J 124*:1624–1626
6. Liberthson PR, Sagar K, Berkoben JP, Weintraub RM, Levine FH (1979) Congenital coronary arteriovenous fistula. Report of 13 patients, review of literature and delineation of management. *Circulation 59*:849–854
7. Ludomirsky A, O'Laughlin MP, Reul GJ, Mullins CE (1990) Congenital aneurysm of the right coronary artery with fistulous connection to the right atrial atrium. *Am Heart J 119*:672–675
8. Rein AJJT, Yatsiv I, Simcha A (1988) An unusual presentation of right coronary atery fistula. *Br Heart J 59*:598–600
9. Sapin P, Frantz E, Jain A, Nichols TC, Dehmer GJ (1990) Coronary artery fistula: an abnormality affecting all age groups. *Medicine 69*:101–113
10. Sherwood MC, Rockenmacher S, Colan SD, Geva T (1999) Prognostic significance of clinically silent coronary artery fistulas. *Am J Cardiol 83*:407–411
11. Stansel HC Jr, Fenn JE (1994) Coronary arteriovenous fistula between the left coronary artery and persistent left superior vena cava complicated by bacterial endocarditis. *Ann Surg 160*:292–296
12. Starc TJ, Bowman FO, Hordof AJ (1986) Congestive heart failure in a newborn secondary to coronary artery–left ventricular fistula. *AM J Cardiol 58*:366–367
13. Sunder KR, Balakrishnan KG, Tharakan JA, et al. (1997) Coronary artery fistula in children and adults: a review of 25 cases with long-term observations. *Int J Cardiol 58*:47–53
14. Thongtang V, Panchavinnin P, Chaithiraphan S, Sabasakul Y (1996) Congenital coronary artery fistula: a report of 24 patients. *J Med Assoc Thai 79*:630–634
15. Wang S, Wu Q, Hu S, et al. (2001) Surgical treatment of 52 patients with congenital coronary artery fistulas. *Chin Med J (Engl) 114*:752–755
16. Wong KT, Menahem S (2000) Coronary arterial fistulas in childhood. *Cardiol Young 10*:15–20