Pediatr Cardiol 23:415–419, 2002
DOI: 10.1007/s00246-002-1334-6

Pediatric Cardiology

Coronary Artery Fistulas in Neonates, Infants, and Children: Clinical Findings and Outcome

K.-S. Hsieh, T.-C. Huang, C.-L. Lee
Department of Pediatrics, Veterans General Hospital-Kaohsiung, 386 Ta-Chung 1st Rd, Kaohsiung, Taiwan

Abstract. Ten patients were diagnosed with coronary artery fistula (CAF) between 1991 and 1998 in our department. The ages of patients ranged from 3 days to 12 years. Five patients were male and five patients were female. Nine patients had single CAF and 1 patient had dual CAFs. CAFs of 7 patients originated from the left coronary artery, and CAFs of 3 patients originated from the right coronary artery. CAFs of 7 patients terminated at the right ventricle, CAFs of 2 patients terminated at the right atrium, and the CAF of 1 patient terminated at the pulmonary artery. Four patients were diagnosed with CAF in the neonate period. All presented with congestive heart failure. Medical therapy was successful in treating congestive heart failure in 2 of these patients, but the other 2 needed operations. One patient presented with subacute bacterial endocarditis at 12 years of age requiring surgical intervention. One patient had a large left-to-right shunt that was surgically repaired. One patient with dual CAFs underwent coil embolization and the larger CAF achieved complete embolization, but the smaller CAF failed. Follow-up data revealed that 1 patient who received an operation died of sepsis due to recurrent bronchiolitis 6 months later. Nine patients were asymptomatic. Because complications including endocarditis may be encountered in later life, we suggest early closure with coil embolization.

Key words: Coronary artery fistula — Neonate — Embolization

Congenital coronary artery fistulas (CAFs) are rare. The advent of color Doppler echocardiography has increased the detection of this anomaly [10]. When diagnosed during infancy or childhood, most of them are asymptomatic. Whether to treat or only observe during infancy or childhood is still controversial. Because complications, including heart failure, myocardial ischemia and angina, infective endocarditis, atrial fibrillation, and rupture are found in later life [3, 5], some authors suggest early closure [1, 3]. However, others advocated conservative management based on the experience that the CAF did not have an adverse clinical outcome in childhood and adolescence [11]. In this study we report 10 cases with congenital CAF and discuss their clinical outcomes.

Patients and Methods

The records of all patients with the diagnosis of CAF made by color Doppler echocardiography between 1991 and 1998 in the pediatric department of Kaohsiung Veterans General Hospital were studied. Color Doppler echocardiography was performed by pediatric cardiologists. Diagnosis of CAF was made when the whole length of CAF (origin, exit, and pathway) could be clearly seen by color Doppler echocardiography. By this method, we found 7 patients who had the CAF. On the other hand, if the origin of CAF could not be determined, then the presence of high-velocity flow in the coronary artery was used as an indicator of the coronary artery from which the CAF originated. By this method, we found 2 patients who had the CAF. In rare conditions, when only continuous high-velocity turbulent flow entering a cardiac chamber or vessel was seen by color Doppler echocardiography, the CAF should be highly suspected and catheterization should be done to confirm the presence of the CAF. By this method, we found 1 patient who had the CAF. Patients with CAF associated with pulmonary atresia with intact ventricular septum were excluded (2 patients). There was a total of 10 patients in our study. We reviewed the 10 patients' chart for presenting symptoms, signs, electrocardiogram (ECG), chest x-ray, echocaridography, catheterization findings, surgical reports, follow-up, and management.

Results

In our study period, 5035 new patients had echocardiographic examinations. Ten patients were found to have CAF. The incidence rate is approximately 0.2%. Seven patients underwent cardiac catheterization.

Correspondence to: C.-L. Lee

The clinical, echocardiography, and catheterization findings and outcome of the 10 patients are listed in Table 1. The age of patients ranged from 3 days to 12 years. Five patients are male and 5 patients are female. In all cases the reason for echocardiography was heart murmur. Among the 10 patients, 4 patients less than 1 month old had congestive heart failure, 1 patient had fever, the other 5 patients had no symptoms or signs. Except for cardiomegaly, chest x-ray revealed no other specific findings. Electrocardiography had no abnormal Q waves, ST segment, or T wave changes suggestive of ischemia.

CAFs of 7 patients originated from the left coronary artery, and CAFs of the other 3 patients originated from the right coronary artery. CAFs of 7 patients terminated at the right ventricle, CAFs of 2 patients terminated at the right atrium, and the CAF of 1 patient terminated at the pulmonary artery. The origin of CAF was misdiagnosed echocardiographically in 2 patients. Patient 2 was diagnosed to have CAF originating from the left coronary artery, but catheterization showed CAF originating from the left anterior descending coronary artery. Patient 8 was diagnosed to have one CAF by echocardiography, but catheterization showed two CAFs terminating at the right ventricle via a common tortuous aneurysm (Fig. 1A).

The pulmonary to systemic flow ratio (Q_p/Q_s) values were between 1 and 2.7. Two patients had values greater than 1.5 and were considered to require operation. Associated cardiac defects were found in two patients. One had mild valvular pulmonary stenosis, and the other had apical muscular ventricular septal defect.

Four patients underwent surgical operation. Patient 1 underwent surgery due to recurrent bronchiolitis and congestive heart failure. Although the congestive heart failure improved by medication with small left-to-right shunt ($Q_p/Q_s = 1.2$), he required surgery to close the ventricular septal defect and ligate the fistula. The operation findings revealed that the CAF was 0.3 cm and the ventricular septal defect was 0.4 cm in diameter. Patient 2 underwent surgery due to congestive heart failure and large left-to-right shunt with $Q_p/Q_s = 2.7$. She was operated on at 1 month of age. The operation findings revealed a CAF of about 1.4 cm in diameter. Patient 7's operation was due to large left-to-right shunt with $Q_p/Q_s = 1.7$. He was operated on at 1.5 years of age. The operation findings revealed a CAF of about 0.5 cm in diameter. Patient 10 was found to have heart murmur and fever. Vegetation and CAF were seen by echocardiography; therefore, CAF with endocarditis was diagnosed. Blood culture also revealed *Staphylococcus aureus*. The patient's history revealed that he had received a dental operation and he did not have antibiotics prophylaxis. He had received a magnetic resonance imaging examination and a CAF about 1.0 cm in diameter was found. After 6 weeks of antibiotics treatment, the patient recovered with no cardiac sequale. He went to another hospital for operation. About 6 months later, he returned to our hospital for echocardiography and was determined to be healthy.

Follow-up periods ranged from 2 months to 7 years, with a mean of 2.9 years. Patient 2 had an operation at 1 month of age. Frequent bronchiolitis was noted. She was admitted to our ward several times. Unfortunately, she expired at 7 months of age due to bronchiolitis with sepsis (culture revealed osacillin-resistance *S. aureus*). Patients 3 and 4 had congestive heart failure in the neonate period. After medication, congestive heart failure improved. Now they are in good health with no medication. In patient 8, coil was inserted to occlude the CAFs. The larger CAF achieved complete embolization (Fig. 1B). The smaller CAF failed because the patient's heart rate dropped when the delivery catheter passed through the CAF. Therefore, we stopped the procedure for coil insertion. She still had a small CAF with good health.

Discussion

CAF diagnosed in the neonate period is rare. Our four neonate patients had congestive heart failure. This association had not been documented in the literature. The diagnosis of congestive heart failure is based on the presence of tachypnea with cardiothoracic ratio in chest x-ray more than 0.55 and/or left ventricular shortening fraction in the M-mode echocardiography less than 0.28. After medication, congestive heart failure improved in two patients. However, two patients still had congestive heart failure that needed operation. The operation findings revealed large and moderate CAFs (1.4- and 0.5-cm diameter). The clinical presentations are different from those in Sherwood et al.'s study [11]. Their study included two neonates with CAF. The neonates in their study had cardiomegaly without congestive heart failure.

Seven patients underwent cardiac catheterization. Among them, two patients had large CAF and received operations. One patient had a small CAF with concomitant ventricular septal defect. His clinical presentation had congestive heart failure, so he also received operation. We attempted coil occlusion in another four patients. Patient 8 had two CAFs; the larger CAF was completely occluded with Gianturco coil, but the smaller CAF failed due to complications. The remaining three patients (patients 3, 6, and 9)

Table 1. Clinical Data, Echocardiography, Catheterization Findings, and Outcome of CAF

Patient No.	Gender	Age	Chest x-ray	ECG	Symptom/ sign	Echo			Catheterization				Outcome
						Origin	Exit	Others	Origin	Exit	Others	Q_p/Q_s	
1	Male	3 days	Cardiomegaly	LVH	CHF	RCA	RV	VSD	RCA	RV	VSD	1.2	Operation at 3 months old
2	Female	4 days	Cardiomegaly	LVH	CHF	LCA	RV	Nil	LAD	RV	Nil	2.7	Operation at 1 month old; died of sepsis at 7 months old
3	Female	7 days	Normal	Normal	CHF	LCA	RV	Nil	LCA	RV	Nil	1.3	CHF in neonate period; asymptom in later life
4	Female	17 days	Normal	Normal	CHF	LCA	RA						Asymptom
5	Male	1 month	Normal	Normal	None	LCA	RV						Asymptom
6	Female	5 months	Normal	Normal	None	LCA	RA	Nil	LCA	RA	Nil	1.2	Asymptom
7	Male	1 year	Normal	Normal	None	RCA	RV	PS	RCA	RV	PS	1.7	Operation at 1.5 years old
8	Female	4 years	Normal	Normal	None	LCA	RV	Nil	Dual LCA	RV	Nil	1.3	Larger CAF: coil embolization; Smaller CAF: left
9	Male	5 years	Normal	Normal	None	LCA	PA	Nil	LCA	PA	Nil	1	Asymptom
10	Male	12 years	Normal	Normal	Fever	RCA	RV	Vegetation					Endocarditis; operation at 12 years old

CAF, coronary artery fistula; CHF, congestive heart failure; LVH, left ventricle hypertrophy; LCA, left coronary artery; PA, pulmonary artery; PS, pulmonary stenosis; Q_p/Q_s, pulmonary to systemic flow ratio; RA, right atrium; RCA, right coronary artery; RV, right ventricle.

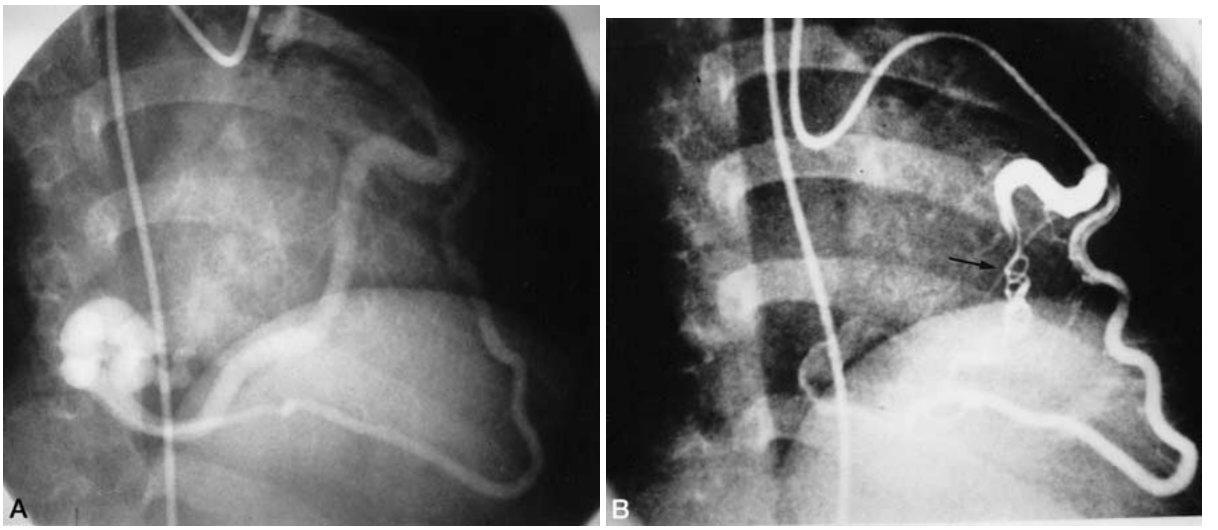

Fig. 1. Anteroposterior projection of selective left coronary arteriogram. (**A**) Dual fistulas originating from the left coronary artery and terminating at the right ventricle via a common tortuous aneurysm. (**B**) Complete coil (*arrow*) embolization of one fistula.

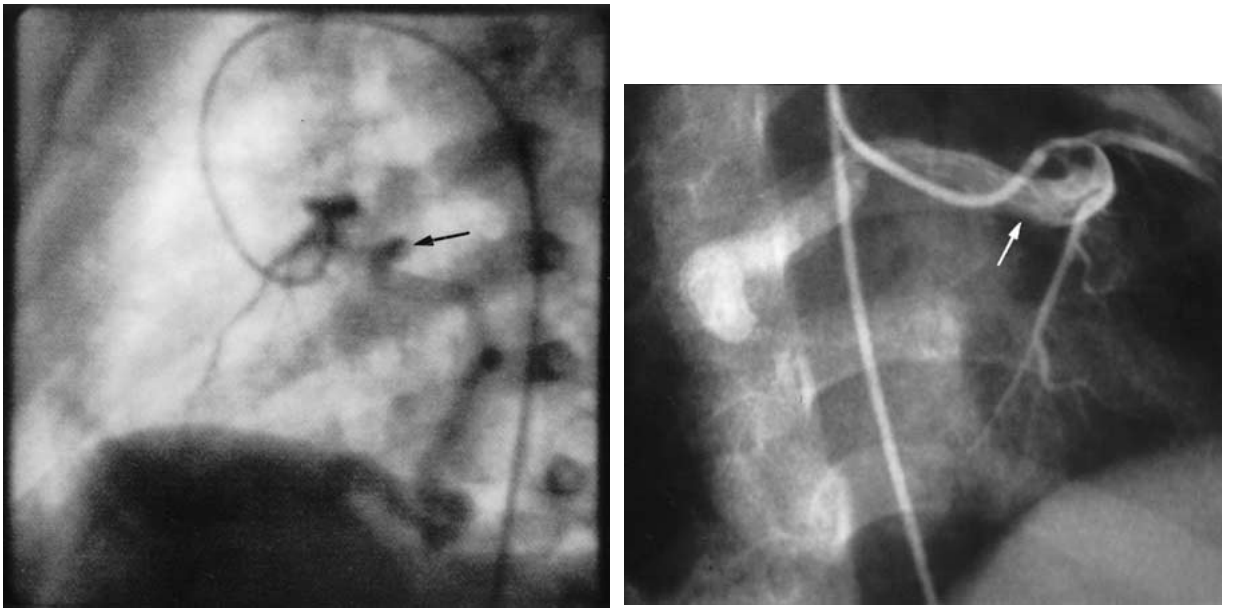

Fig. 2. Lateral projection of selective left coronary arteriogram revealed tortuous fistula (*arrow*) originating from the left coronary artery and terminating at the right ventricle.

Fig. 3. Anteroposterior projection of selective left coronary arteriogram revealed tortuous fistula (*arrow*) originating from the left coronary artery and terminating at the right atrium.

failed in coil occlusion due to the difficulty of the procedure. The fistula pathways of patients 3 and 6 were too tortuous to pass through the catheter (Figs. 2 and 3). The fistula of patient 9 was too small to pass through the catheter (Fig. 4). Although our experience for coil occlusion is limited, we believe transcatheter coil occlusion of the CAF is a reasonable alternative to surgical closure [8, 9]. However, not all patients can undergo coil embolization. In Mavroudis et al.'s [4] study, only 37% of patients could have coil embolization. They believe the factors prohibiting the use of coil embolization are distal fistula, adjacent vessel at risk, need for concomitant distal coronary bypass, large fistula, and young age. However, in our experience, the difficulty of the procedure in cases of small size and tortuous pathway of CAF is

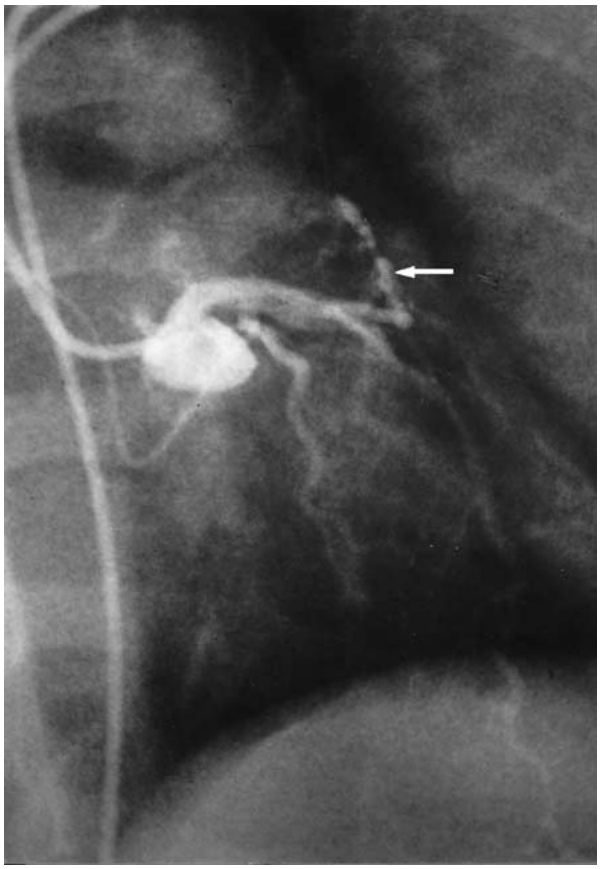

Fig. 4. Anteroposterior projection of selective left coronary arteriogram revealed small fistula (*arrow*) originating from the left coronary artery and terminating at the pulmonary artery.

the major problem. In addition to standard Gianturco coil embolization, detachable balloon [2], polyvinyl alcohol foam [12], double umbrella devices [8], and platinum microcoil [13] are other transcatheter closure methods. Once transcutaneous closure of CAF is selected, a suitable technique for closure of CAF may be chosen.

Whether to treat CAF in childhood or not is controversial. CAF can cause symptoms including cardiac failure, myocardial ischemia and angina, infective endocarditis, atrial fibrillation, and rupture in adult patients [3, 5]. Based on these complications, early closure of CAF in childhood regardless of symptoms is suggested [1, 3]. However, some authors have advocated conservative management [11]. They consider that clinically silent CAF does not have an adverse clinical outcome in childhood and adolescence. Conservative management with continued follow-up is sufficient. However, such reports state that life-long risk of infective endocarditis is not known. In our study, infective endocarditis did happen in one patient. To avoid this complication, even a small CAF should be treated.

Once transcutaneous embolization of CAF is selected, the coronary artery and CAF relationship should be clearly recognized. Left coronary artery fistula combined with right coronary artery atresia has been reported [1, 8]. In this situation, the fistula may serve as a supplying artery to the right ventricle. Moskowitz et al. [7] recommended 5 minutes of ballon occlusion of the fistula prior to embolization to evaluate possible ischemic or conduction changes. After embolization, follow-up of these patients is important because myocardial infarction has been reported in a 14-year-old girl 10 years after surgical closure of CAF [6].

References

1. Farooki ZQ, Nowlen T, Hakimi M, Pinsky WW (1993) Congenital coronary artery fistulae: a review of 18 cases with special emphasis on spontaneous closure. *Pediatr Cardiol 14*:208–213
2. Krabill KA, Hunter DW (1993) Treatment of congenital coronary arterial fistula with a detachable balloon. *Pediatr Cardiol 14*:176–178
3. Liberthson RR, Sagar K, Berkoben JP, Weintraub RM, Levine FH (1979) Congenital coronary arteriovenous fistula: report of 13 patients, review of the literature and delineation of management. *Circulation 59*:849–854
4. Mavroudis C, Backer CL, Rocchini AP, Muster AJ, Gevitz M (1997) Coronary artery fistulas in infants and children: a surgical review and discussion of coil embolization. *Ann Thorac Surg 63*:1235–1242
5. McNamara JJ, Gross RE (1969) Congenital coronary artery fistula. *Surgery 65*:59–69
6. Mesko Zg, Damus PS (1998) Myocardial infarction in a 14-year-old girl, ten years after surgical correction of congenital coronary artery fistula. *Pediatr Cardiol 19*:366–368
7. Moskowitz WB, Newkumet KKM, Albrecht GT, et al (1991) Case of steel versus steal: coil embolization of congenital coronary arteriovenous fistula. *Am Heart J 121*:909–911
8. Perry SB, Rome J, Keane JF, Baim DS, Lock JE (1992) Transcatheter closure of coronary artery fistuals. *J Am Coll Cardiol 20*:205–209
9. Reidy JF, Anjos RT, Qureshi SA, Baker EJ, Tynan MJ (1991) Transcatheter embolization in the treatment of coronary artery fistulas. *J Am Coll Cardiol 18*:187–192
10. Shakudo M, Yoshikawa J, Yoshida K, Yamura Y (1989) Noninvasive diagnosis of coronary artery fistula by Doppler color flow mapping. *J Am Coll Cardiol 13*:1572–1577
11. Sherwood MC, Rockenmacher S, Colan SD, Geva T (1999) Prognostic significance of clinically silent coronary artery fistulas. *Am J Cardiol 83*:407–411
12. Strunk ML, Hieshima GB, Shafton EP (1991) Treatment of congenital coronary arteriovenous malformations with microparticle embolization. *Cathet Cardiovasc Diagn 22*:133–136
13. Wolf DD, Terriere M, Wilde PD, Rediy JF (1994) Embolization of a coronary fistula with a controlled delivery platinum coil in a 2-year-old. *Pediatr Cardiol 15*:308–310