Clinical and Angiographic Analysis of Congenital Coronary Artery Fistulae in Adulthood

Is There Any New Trend?

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SUMMARY

Fifteen (2.1%) patients were diagnosed as having congenital coronary artery fistula (CAF) in a consecutive series of 704 adult patients undergoing selective coronary arteriography; the incidence was astonishingly higher than previous observations. The presentation of clinical symptoms and the electrocardiographic changes at rest and/or after exercise testing probably attributable to the CAF were observed unexpectedly often in spite of the fact that the magnitude of the shunt seemed not to be significant. With respect to the anatomy of the CAF, the incidence of origination from plural coronary arteries and that of fistulation into the left ventricle were also unexpectedly high. These observations presented a striking contrast to those of previous reports, but we were unable to determine the reason. In four cases, the CAF was ligated either electively or concurrently with mitral valve surgery, and the results were satisfactory. Taking these circumstances into consideration, we should not minimize the impact of a CAF with a seemingly small shunt. (Jpn Heart J 1996; 37: 95–104)

Key words: Coronary artery fistula Congenital Electrocardiogram
Coronary angiography Origination Fistulation Symptom Myocardial ischemia

SINCE the first report of a congenital coronary artery fistula (CAF) by Krause in 1865,¹⁾ more than 400 cases have been reported, and the anatomical and clinical features have been thoroughly discussed in many papers.²⁻⁸⁾ We wondered, however, if our recent observations of this lesion might be different from classical ones with respect to clinical symptoms, electrocardiographic changes,

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and the anatomy of the CAF. We conducted this retrospective study to determine whether or not there was a new trend with this cardiac defect and to define its precise management.

MATERIALS AND METHODS

The background of the cases referred for this study was a consecutive series of 704 adult patients having undergone selective coronary arteriography (CAG) at the Junkankika-Hayashi Hospital from January 1989 to June 1994. They consisted of 434 (61.6%) males and 270 (38.4%) females, ranging in age from 20 to 81 (mean 58.9) years. Fifteen (2.1%) were diagnosed as having congenital CAF based primarily upon the classical definition;⁹⁾ we carefully excluded cases of acquired origin where the CAF was found in patients having cardiac tumors, a mural thrombus in the left atrium or an infarcted left ventricle (LV), and those with a history of cardiopulmonary trauma and/or surgery. There were 7 males and 8 females, ranging in age from 44 to 72 years (mean 60.7). The relevant clinical data are presented in Table I. When the CAF was found to communicate with the right side of the heart, the arteriovenous, that is, the left-to-right shunt ratio was measured by conventional oximetry based on Fick's principle. The

Table I. Clinical Data

Case No	Age/Sex (year)	Symptoms	Heart murmur	CTR (%)	Associated cardiac lesions
1	57/F	Chest oppression	(-)	44.6	(-)
2	64/M	Palpitation	(-)	55.8	(-)
3	63/F	Angina	(-)	46.0	(-)
4	65/F	Chest oppression	Continuous 3/6	47.9	Seg 3 (50%), OMI (-)
5	56/F	Angina	Systolic 2/6	59.2	
6	44/F	(-)	(-)	43.7	(-)
7	55/F	Angina	(-)	57.8	(-)
8	72/M	Angina	(-)	49.0	(-)
9	68/M	Angina	(-)	57.9	(-)
10	61/M	Chest oppression	(-)	42.1	(-)
11	66/M	Angina	(-)	50.0	Seg 9 (75%), OMI (-)
12	70/F	Angina	(-)	59.6	Seg 11 (90%), OMI (-)
13	59/F	Heart failure (NYHA IV)	Systolic 3/6 Diastolic 3/6	66.4	MSR; MVA = 1.8 cm ² Grade 2 MR
14	60/F	SOB (NYHA II)	Diastolic rumble	62.3	Re-MS; MVA = 1.4 cm ²
15	51/M	SOB (NYHA II)	Systolic 3/6	60.4	Op. for ASD and MS (MVR)

The intensity of heart murmurs is presented by the Levine grade as in 3/6, 2/6, and 4/6. The percentages in parentheses indicate the degree of the coronary stenosis. ASD = atrial septal defect; CTR = cardiothoracic ratio; F = female; M = male; MR = mitral regurgitation; MSR = mitral stenosis and regurgitation; MVA = mitral valve area; MVR = mitral valve replacement; NYHA = New York Heart Association Classification; OMI = old myocardial infarction; Op. = operation; Re-MS = mitral restenosis; Seg = coronary artery segment⁸⁾; SOB = shortness of breath.

coronary artery segment from which the CAF originated and the cardiovascular site into which the CAF fistulated were carefully determined on the coronary arteriograms. The coronary artery segmentation proposed by the American Heart Association¹⁰⁾ was used in this study since it was very convenient to indicate the precise site of origination, particularly in the same coronary artery system. The numbers of origins and fistulae of the CAF were carefully examined, and were expressed in terms of single and/or a few, and multiple and/or diffuse. In 15 cases, the clinical symptoms, physical examination data, chest x-ray findings and electrocardiograms (ECGs) at rest and after a routine exercise test using the single and/or double Master method were retrospectively examined. Although the treadmill test using the Bruce protocol was also performed to evaluate the need for surgery, not only for the CAF, but also for other associated cardiac lesions, the results were not included in the above findings. An operative consideration was also presented based on the four cases in which the CAF was surgically closed.

RESULTS

Clinical data and findings on chest x-ray and ECG: Ten (cases 1–10) of 15 cases were not associated with other cardiac lesions, 2 (cases 11 and 12) had significant atherosclerotic coronary stenosis, and the remaining 3 cases had valvular heart diseases (Table I). Of the clinical symptoms, angina and/or chest oppression was observed in 10 cases, shortness of breath (New York Heart Association Class II; NYHA) in 2 cases, and congestive heart failure (NYHA Class IV) or palpitation in one case each. The remaining case (case 6) did not present any symptoms but was referred to us because of abnormal ECG changes after an exercise test performed at another hospital. Therefore, 14 (93.3%) of all 15 cases, and 9 (90%) of 10 cases having no other cardiac lesions were symptomatic. Heart murmurs were heard in 5 cases, although the typical continuous murmur was present in only one (case 4) with an intensity of 3/6 Levine grade. Three of the remaining 4 cases were associated with valvular heart diseases producing heart murmurs. By chest x-ray, the cardiothoracic ratio ranged from 66.4 to 42.1% (mean 53.5), and a ratio larger than 60% was observed in only three cases associated with valvular heart diseases, and it was in these cases that the pulmonary vascularity was increased. The major findings on the ECGs at rest and after exercise testing are summarized in Table II. Of the 10 cases having no other cardiac lesions, including case 4 with a 50% stenosis of the right coronary artery (RCA), 9 cases showed a normal sinus rhythm and one case atrial fibrillation. At rest, three cases presented abnormal changes suggestive of myocardial ischemia. After an exercise stress test, 6 cases showed significant new or increased depres-

Table II. Electrocardiographic Findings

Case No	Rhythm	Abnormal findings	
		At rest	After exercise
1	NSR	(-)	ST-dep. in II, III, aVF
2	Af	(-)	(-)
3	NSR	(-)	T-inv. in V ₁₋₅
4	NSR	(-)	(-)
5	NSR	(-)	ST-dep. in II, V ₄₋₅
6	NSR	(-)	ST-dep. in II, III, aVF
7	NSR	QS in III, Inv-T in III, aVF	ST-dep. in I, II
8	NSR	(-)	T-inv. in V ₂₋₃
9	NSR	Q in aVL	No other change
10	NSR	ST-dep. in V ₃₋₆ , Inv-T in V ₁₋₆	No other change
11	NSR	(-)	(-)
12	NSR	ST-dep. in II, III, aVF, V ₆	No other changes
13	Af	(-)	(-)
14	Af	(-)	(-)
15	Af	(-)	(-)

Af = atrial fibrillation; Inv-T = inverted T-wave; NSR = normal sinus rhythm; Q = large Q-wave; q = small q-wave; QS = QS-pattern; ST-dep. = ST-segment depression; T-inv. = T-wave inversion

sion (more than 2 mm) of the ST segment and/or inversion of the T wave. As a result, 8 (80%) of the 10 cases presented ECG changes suggestive of myocardial ischemia either at rest or after an exercise stress test. On the other hand, of the 5 cases associated with other cardiac lesions, 2 cases of coronary stenosis presented a normal sinus rhythm, while 3 cases of valvular heart disease developed atrial fibrillation. In this group, significant ECG changes suggestive of myocardial ischemia were observed only in case 12, and additional changes after exercise testing were not detectable; 3 cases had atrial fibrillation.

Cardiac catheterization data; CAG and oximetry: As shown in Table III, the CAF originated from both coronary arteries, the left coronary artery (LCA), and the RCA in 9 (60.0%), 5 (33.3%), and one (6.7%) case, respectively. The fistulation sites were the LV, the pulmonary artery (PA), and the right ventricle (RV) in 8, 7, and one case, respectively. Concurrent fistulation into two different sites in the same patient was observed in one case (case 6), while fistulation into the same site from plural coronary systems was seen in 9 cases. Accordingly, in all, 24 coronary artery systems participated in developing CAFs and had 25 sites of fistulation. The number of orifices of the fistulae was one and/or a few for 6 sites of fistulation, while it was multiple and/or diffuse for 19. No relationship was observed between the number and the site of fistulation. In 7 cases, the CAF angiographically showed a left-to-right shunt. The shunt ratio was estimated in 3

Table III. Coronary Arteriographic Findings and Shunt Ratio

Case No	CAF		Left-to-right shunt ratio (%)
	Origin/Fistulation/Number		
	RCA	LCA	
1	RV-brs/RV/M	(-)	Undetectable
2	Seg 1/PA/S	Seg 7/PA/M	Undetectable
3	Seg 1/PA/M	Seg 7/PA/M	8.0
4	Seg 1/PA/M	Seg 7/PA/M	11.0
5	(-)	Seg 7, HL/PA/M	Undetectable
6	Seg 1/PA/M	Seg 6/PA/M, Seg 8, 9/LV/S	Undetectable
7	(-)	Seg 12/LV/S	/
8	4-PD, 4-PL/LV/M	Seg 7, 8, 9, 12/LV/D	/
9	4-PD/LV/D	Seg 7, 8, 12, HL/LV/D	/
10	(-)	Seg 7, 9, 11/LV/M	/
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11	(-)	Seg 7/LV/a few	/
12	4-PD/LV/M	Seg 7, 9, HL/LV/M	/
13	Seg 1/PA/S	Seg 9/PA/S	14.4
14	(-)	Seg 11/R-PA/M	/
15	4-PD/LV/D	All brs/LV/D	/

brs = branches; CAF = coronary artery fistula; D = diffuse; HL = high lateral branch; LCA = left coronary artery; LV = left ventricle; M = multiple; PA = pulmonary artery; R- = right; RCA = right coronary artery; RV = right ventricle; S = single; Seg = coronary artery segment⁶; 4-PD = posterior descending artery of segment 4; 4-PL = posterolateral branch of segment 4.

Figure 1. The diffuse drainage pattern of the CAF into the left ventricle (case 9); the left coronary arteriogram in an anteroposterior projection. The left ventricular anteroapical site is diffusely opacified by the contrast material draining through multiple fistulae originating from the left anterior descending coronary artery (LAD), a high lateral branch (HL), and an obtuse marginal artery (OM).

cases to be 8.0, 11.0 and 14.4% in cases 3, 4, and 13, respectively, but it was not detectable by oximetry in the remaining 4 cases. There were two different patterns of fistulation into the LV. First, the inner layer of the LV myocardium was

Figure 2. The single drainage pattern of the CAF into the left ventricle (case 7); the left coronary arteriogram in a right-anterior oblique projection. An aberrant, straight vessel (the longest arrow) is seen to originate from an obtuse marginal artery (OM) and to drain into the left ventricle with a smoking sign (a short, thick arrow).

diffusely stained with contrast material as a sponge and the dye emptied directly into the LV chamber chiefly at systole. No evident fistulous vessel was seen, and the site and area of the myocardium stained were different in each. It was relatively localized in 6 cases, and in two other cases almost the whole inner layer of the LV was blushed, and even a vague left ventriculogram was presented by the contrast material emptying from the CAF (Figure 1; case 9). This pattern was predominant as observed at 10 (83.3%) of 12 sites of LV fistulation in 8 cases. The second pattern was that one or a few distinct vessels were visualized to directly fistulate into the LV chamber, and the contrast material emptied chiefly during diastole. This pattern was observed in 2 cases (cases 7 and 11) (Figure 2, case 7).

Clinical management and operative considerations: The CAF was electively closed in cases 3 and 4 and was closed at the time of mitral valve surgery in cases 13 and 14, using cardiopulmonary bypass. In cases 3 and 4, the symptom of angina and/or chest oppression completely disappeared and tolerance to daily work was much improved after surgery. The T-wave inversion observed after exercise testing before surgery in case 3 and the continuous heart murmur heard preoperatively in case 4 both disappeared after surgery. Such postoperative improvements, however, could not be supported by thallium myocardial scintigraphy since the method had not shown a myocardial perfusion defect before surgery. At operation in cases 3, 4, and 13, most of the roots of the fistulous arteries were successfully snared before injection of cardioplegic solution via an aortic root cannula, and cardioplegia could be induced as usual with the

aid of snares. Whereas in case 14, we attempted to induce cardioplegia by the usual method before the CAF-related aberrant artery could be controlled since the origin was deep at the root of the left circumflex coronary artery. Cardioplegia, however, could not be obtained with the usual amount of cardioplegic solution, and an additional injection was required. In addition, we unexpectedly observed that the left side of the heart was markedly distended because of the massive drainage of cardioplegic solution from the pulmonary circulation; this urgently necessitated left atrial venting. Thus with cardioplegia the origin of the fistulous artery could be successfully ligated. After this, no further distension of the left side of the heart was observed at the time of the repeat infusions of cardioplegic solution for the subsequent mitral valve replacement. The other 11 cases were subjected to medical treatment, including calcium antagonists, nitro compounds, angiotensin-converting enzyme inhibitors, and their combinations, but the results were inconsistent.

DISCUSSION

Yamanaka and Hobbs have reported that the incidence of coronary artery anomalies in 126,595 adult patients undergoing CAG was 1.33%, and that of congenital CAF was 0.18%.⁷⁾ Many previous reports have described a similar incidence.²⁻⁶⁾ Therefore, the incidence of 2.1% observed in our series was astonishingly high. The real reason was not clear to us. Our equipment, technique and selection criteria for CAG were not significantly different from those used in other studies. The different results might have been related to how carefully we searched for this cardiac defect. Our study series was, however, too small to determine the incidence of congenital CAF and to negate previous observations. Further studies in a large series are required.

In cases with CAFs, myocardial ischemia can arise either from a coronary steal phenomenon or oxygen supply-demand imbalance.¹¹⁾ As a result, it will produce clinical symptoms of exertional chest pain, acute myocardial infarction,¹¹⁾ and even sudden cardiac arrest.¹²⁾ On the other hand, it is generally considered that a CAF with a small shunt rarely presents clinical symptoms and that the extent of the shunt is the most important and reliable indicator for surgical correction. In our study, however, it was only one of the 10 cases having no other cardiac lesions that did not present any clinical symptoms. In addition, only 2 of these 10 cases did not present ECG changes suggestive of myocardial ischemia either at rest or after exercise testing, and one of them was associated with atrial fibrillation. In such cases it is usually difficult to correctly interpret the result of the exercise stress test. Although these observations might be due in part to the fact that the patient selection for this CAG study may have tended to favor symptomatic cases and those presenting heart murmurs or some ECG abnormal-

ity, a similar selection bias must have been present in the previous studies as well. Therefore, it is difficult to determine the true incidence of asymptomatic CAF. However, it can be said that not a few of the adult cases may be symptomatic and/or present some ECG abnormality even when the magnitude of the shunt is considered not significant. Furthermore, in case 13 with a small shunt ratio of 14.4%, the mitral valve was found at operation not to be so severely deteriorated as to cause congestive heart failure by itself. The association of mitral valve disease with CAF was thought to be synergistic, that is, the two lesions might adversely influence the cardiac function in a multiplicative way. Therefore, the surgical indication for CAF should be carefully considered particularly when it is associated with other cardiac lesions. This would be the case when CAF was present in a coronary artery system having an atherosclerotic stenosis. This rare but serious situation was observed in case 11; the fistulae originated from coronary segment 7 and a 75% stenosis was present in segment 9. In this case, close observation would be mandatory although ischemic ECG changes were not observed hitherto and anginal pain could be decreased on medical therapy.

There was a problem in determining the magnitude of the shunt through the CAFs. That is, the amount of the shunt could not always be estimated by conventional oxymetry when the CAF communicated with the left side of the heart as seen in 8 cases in our series. Other precise methods could possibly have demonstrated the hemodynamic significance of the CAF. They included 48-hour dynamic ECG monitoring,¹³⁾ myocardial scintigraphy,^{14,15)} a special calculation method of the left-to-left shunt¹⁶⁾ and coronary sinus lactate measurement,¹⁴⁾ but the results were inconclusive. Recently, Houghton et al¹⁷⁾ have clearly documented a case of CAF, in which the basal coronary blood flow determined by the intracoronary Doppler catheter flow velocity measurement was markedly elevated and the coronary reserve was correspondingly markedly depressed in the affected artery, supporting the concept of a coronary steal as the etiology for angina and ischemic ECG changes. It was, however, also a fact that in the same case they failed to demonstrate the left-to-right shunt by conventional oximetry. This is a very important problem, and there is a serious risk to erroneously judge a CAF as not being clinically significant merely because the magnitude of the shunt is considered small according to the results of conventional oximetry and/or angiography. We should make an effort to describe the myocardial ischemia caused by the CAF by carefully evaluating the clinical symptoms and the results of all possible examination methods even when the shunt appears to be small.

Many papers described the predominance of the origination from a single coronary artery and that of fistulation into the right side of the heart.²⁻⁴⁾ In our series, however, such origination was observed in only 4 cases, whereas origination from plural coronary systems was seen in the remaining 11 cases. Further-

more, fistulation into the LV was unexpectedly frequent as observed in 8 cases, making a striking contrast to the previous observation.²⁻⁴⁾ Lefevre et al¹⁸⁾ also have suggested a higher incidence of LV fistulation. In fact, more than 73 cases of LV fistulation have been reviewed in the literature.^{14,18-21)} Radiologists and cardiologists are expected to completely visualize the CAF with a view toward not only documenting the real features of this cardiac defect but also providing cardiac surgeons with detailed information necessary for precise management.

We observed two different patterns of fistulation into the LV as already described. In light of the observation by Wearn et al,²²⁾ we speculated that the multiple, diffuse pattern of fistulation with blushing of the inner layer of myocardium would have a close relationship to persistent arterio-sinusoidal vessels, and the second pattern presenting one or a few but distinct fistulous vessels would be associated with the presence of arterio-luminal vessels in the myocardial wall. The clinical implications of these two patterns, however, are not yet clear.

An interesting observation at operation was the steal phenomenon of cardioplegic solution through the CAF, which resulted not only in the difficult induction of cardioplegia but also in the unfavorable distension of the left heart. This fact should be kept in mind, and some venting methods or a retrograde cardioplegic procedure as an alternative should be considered when it is necessary to infuse cardioplegic solution before the major fistulous arteries are controlled.

In conclusion, the clinical symptoms and the ECG changes suggestive of myocardial ischemia were observed unexpectedly often in adult cases of CAF with a seemingly small shunt, and the incidence of involvement of plural coronary arteries in CAFs and that of drainage into the LV were also unexpectedly high, making a striking contrast to the previous observations. Therefore, we should not minimize the importance of such CAF, particularly when it is associated with other cardiac lesions. Our series was, however, too small to negate the previous observations. Studies in a large series will further elucidate the clinical significance of CAFs with a seemingly small shunt.

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