Congenital Coronary Arteriovenous Fistula
Report of 13 Patients, Review of the Literature and Delineation of Management

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RONALD M. WEINTRAUB, M.D., AND FREDERICK H. LEVINE, M.D.

SUMMARY  Thirteen new patients and 174 patients previously reported with coronary arteriovenous fistula (CAVF) were reviewed to delineate the course and management of CAVF and to clarify the role of surgical ligation in the young asymptomatic patient. Patients were grouped according to age: 99 patients (four new and 95 reported) were < 20 years old and 88 (nine new and 79 reported) were ≥ 20 years old. Of those under 20 years of age, 19% had preoperative symptoms or CAVF-related complications, including congestive heart failure (CHF) in 6%, subacute bacterial endocarditis in 3% and death in one patient. Seventy-six patients younger than 20 years old had CAVF ligation with only one significant complication. In contrast, 63% of the older group and all of our nine older patients had preoperative symptoms or complications, including CHF in 19%, SBE in 4%, myocardial infarction (MI) in 9%, death in 14% and fistula rupture in one patient. Of the 43 ligated older patients, 23% had surgical complications, including MI in three and death in three. Mean pulmonic-to-systemic flow in the entire group was 1.6:1 and did not differ significantly in those with or without symptoms or complications. One of our patients and one previously reported had spontaneous CAVF closure.

In summary, early elective ligation of CAVF is indicated in all patients because of the high incidence of late symptoms and complications and the increased morbidity and mortality associated with ligation in older patients.

SINCE ITS INITIAL DESCRIPTION in 1865,1 more than 170 patients with congenital coronary arteriovenous fistula (CAVF) have been reported.1-66 The anatomic variants of CAVF and their clinical, diagnostic, hemodynamic and angiographic features have been well-described.16, 26, 30, 56-65 However, the management of patients with CAVF remains controversial,5, 13, 17, 20-23, 36, 40, 48, 49, 50 particularly operative intervention in the asymptomatic patient.

In this study we review our experience with 13 patients who had CAVF, discuss their presentation, clinical course, management, complications and follow-up, review the reported experience with this entity, and clarify guidelines for management of these patients.

Methods
The records of 13 patients who had CAVF diagnosed by cardiac catheterization between 1965-1977 were studied. We reviewed symptoms or complications, clinical, electrocardiographic, radiologic and catheterization findings, follow-up and management. In addition, the previously reported patients who had adequate descriptive data, including symptoms, complications and follow-up course, were tabulated.2-68 The t test was used to analyze statistical significance.

Results
Table 1 gives the age at which CAVF was confirmed angiographically, sex, preoperative symptoms, complications attributed to the fistula, electrocardiographic and chest x-ray findings, fistula course, pulmonic-to-systemic flow ratio (Qp/Qs), associated cardiac abnormalities, surgical complications in those
who had ligation, and follow-up status of these patients. Patients were divided into two groups, those younger than 20 years of age, and those 20 years or older.

All patients younger than age 20 years were asymptomatic, and none of those 20 years and older had symptoms before age 30 years, although all were symptomatic thereafter. Symptoms included dyspnea on exertion in seven patients and angina pectoris in five. No patient under age 20 years had fistula-related complications, and five older patients (ages 39–67 years) had complications, including clinical congestive heart failure (CHF) in four and electrocardiographic findings of myocardial ischemia—classical ST-T wave changes associated with angina—in three.

In all patients heart murmurs were either continuous or had components in both systole and diastole and were heard best at the mid-sternal region over the second to fourth right or left intercostal spaces. All patients had normal systemic arterial pulse pressures. Nine patients, ages 1–41 years, and one patient, age 63 years, had normal ECGs. Two patients, ages 45 and 67 years, both with CHF and fistulae entering the coronary sinus—had atrial fibrillation. Six patients had normal heart size on chest x-ray and seven had roentgenographic evidence of cardiomegaly (cardiothoracic ratio > 0.5). The CAVF drained into the coronary sinus or right atrium in eight patients, the main pulmonary artery in three, and the right ventricle in two. No patient had a fistula entering the left heart.

Three younger patients (ages 1, 14 and 18 years) and three older patients (ages 31, 41 and 57 years) had no detectable Qp/Qs by oximetry, and in the remaining patients Qp/Qs ranged from 1.4:1–2.3:1. Five patients had associated cardiac abnormalities. Two patients had mitral insufficiency, which in one was hemodynamically insignificant and related to prior subacute bacterial endocarditis, and in the other was severe and rheumatic in etiology. In the latter patient mitral insufficiency dominated the clinical presentation and contributed to death 1 month after combined CAVF ligation and mitral valve replacement. Two patients had significant left anterior descending coronary artery stenosis which in one was managed with bypass grafting at the time of CAVF ligation. One patient had a large patent ductus arteriosus which contributed to clinical congestive heart failure and was ligated 12 years before CAVF ligation. Two patients had systemic hypertension which in one was severe.

One patient previously reported by Shubrooks et al. had spontaneous near closure of his CAVF, documented on repeat cardiac catheterization 2 years after the initial diagnosis. Six patients underwent CAVF ligation (one with a coronary artery bypass graft and one with mitral valve replacement). There were no intraoperative deaths, no patient had an intraor perioperative myocardial infarction, and there was one late postoperative death related to associated

<table>
<thead>
<tr>
<th>Age at dx (yrs)</th>
<th>Sex</th>
<th>Preoperative symptoms</th>
<th>Preoperative fistula-related complications</th>
<th>ECG</th>
<th>Chest x-ray</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>M</td>
<td>—</td>
<td>—</td>
<td>nl</td>
<td>nl</td>
</tr>
<tr>
<td>14</td>
<td>M</td>
<td>—</td>
<td>—</td>
<td>nl</td>
<td>nl</td>
</tr>
<tr>
<td>18</td>
<td>F</td>
<td>—</td>
<td>—</td>
<td>nl</td>
<td>Prom MPA</td>
</tr>
<tr>
<td>19</td>
<td>F</td>
<td>—</td>
<td>—</td>
<td>nl</td>
<td>Prom MPA</td>
</tr>
<tr>
<td>30</td>
<td>M</td>
<td>Angina</td>
<td>—</td>
<td>nl</td>
<td>Cardiomeg</td>
</tr>
<tr>
<td>31</td>
<td>F</td>
<td>DOE</td>
<td>—</td>
<td>nl</td>
<td>Cardiomeg</td>
</tr>
<tr>
<td>39</td>
<td>F</td>
<td>Angina</td>
<td>Ischemia</td>
<td>nl</td>
<td>Cardiomeg</td>
</tr>
<tr>
<td>41</td>
<td>F</td>
<td>DOE</td>
<td>—</td>
<td>nl</td>
<td>Cardiomeg</td>
</tr>
<tr>
<td>45</td>
<td>M</td>
<td>DOE</td>
<td>CHF</td>
<td>LVH, A fib</td>
<td>Cardiomeg</td>
</tr>
<tr>
<td>57</td>
<td>M</td>
<td>DOE, angina</td>
<td>CHF</td>
<td>LAD</td>
<td>Cardiomeg</td>
</tr>
<tr>
<td>57</td>
<td>F</td>
<td>DOE, angina</td>
<td>CHF, ischemia</td>
<td>LVH, RAE, LAE</td>
<td>Cardiomeg</td>
</tr>
<tr>
<td>63</td>
<td>F</td>
<td>DOE</td>
<td>—</td>
<td>nl</td>
<td>Cardiomeg</td>
</tr>
<tr>
<td>67</td>
<td>F</td>
<td>DOE, angina</td>
<td>CHF, ischemia</td>
<td>A fib</td>
<td>Cardiomeg</td>
</tr>
</tbody>
</table>

*Also had left anterior descending coronary artery bypass grafting at the time of CAVF ligation.
†Also had mitral valve replacement at the time of CAVF ligation.

Abbreviations: A fib = atrial fibrillation; ASHD = atherosclerotic coronary heart disease; asymt = asymptomatic; cardiomeg = cardiomegaly; CHF = congestive heart failure; CS = coronary sinus; DOE = dyspnea on exertion; dx = diagnosis; ECG = electrocardiogram; F = female; hypert = hypertension; LAD = left anterior descending artery; LAE = left atrial enlargement; LCX = left circumflex coronary artery; LM = left main coronary artery; LVH = left ventricular hypertrophy; M = male; MPA = main pulmonary artery; MR-rheum = mitral regurgitation—rheumatic; MR-SBE = mitral regurgitation—subacute bacterial endocarditis; NA = not available; nl = normal; PDA = patent ductus arteriosus; prom = prominent; Qp/Qs = pulmonary-to-systemic flow ratio; RA = right atrium; RAE = right atrial enlargement; RCA = right coronary artery; RV = right ventricle; spont = spontaneous.
mitral insufficiency. Three of the five symptomatic patients have been asymptomatic after CAVF ligation, and one patient (age 39 years) has had persistent angina after CAVF ligation and coronary artery bypass graft, but no longer has a positive treadmill test. Three patients consented to undergo elective CAVF ligation, one could not be contacted for follow-up, and two patients refused surgery.

Table 2 summarizes the findings in 174 patients reported in the literature\(^1\)\(^\text{-}^{46}\) who had descriptive data concerning age, preoperative symptoms, fistula-related complications, and surgical results, if any. Data concerning the Qp/Qs which were available in 73 patients is also included. Findings in all 174 patients are shown, as are those in the 95 patients younger than 20 years of age, and 79 patients 20 years or older. Among

### Table 1. (Continued)

<table>
<thead>
<tr>
<th>Fistula course</th>
<th>Qp/Qs</th>
<th>Other cardiac lesions</th>
<th>Fistula ligation</th>
<th>Surgical complications</th>
<th>Follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>RCA→RV, LCX→RV</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>2</td>
</tr>
<tr>
<td>LM→RA</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>1</td>
</tr>
<tr>
<td>RCA→MPA</td>
<td>1.6/1</td>
<td>—</td>
<td>+</td>
<td>—</td>
<td>1.5</td>
</tr>
<tr>
<td>RCA→RA</td>
<td>2.3/1</td>
<td>MR-SBE</td>
<td>+</td>
<td>—</td>
<td>2</td>
</tr>
<tr>
<td>LAD→MPA</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>NA</td>
</tr>
<tr>
<td>LCX→CS→RA</td>
<td>2.1/1</td>
<td>ASHD</td>
<td>+*</td>
<td>—</td>
<td>2.5</td>
</tr>
<tr>
<td>LAD→MPA, RCA→MPA</td>
<td>—</td>
<td>PDA, hypert</td>
<td>+</td>
<td>1.0</td>
<td>Asympt</td>
</tr>
<tr>
<td>RCA→CS→RA</td>
<td>1.5/1</td>
<td>MR-rheum</td>
<td>+†</td>
<td>0.1</td>
<td>Death</td>
</tr>
<tr>
<td>RCA→RV</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>10</td>
</tr>
<tr>
<td>RCA→CS→RA</td>
<td>1.4/1</td>
<td>ASHD, hypert</td>
<td>—</td>
<td>4.0</td>
<td>DOE, angina</td>
</tr>
<tr>
<td>LCX→CS→RA</td>
<td>2.2/1</td>
<td>—</td>
<td>—</td>
<td>3.0</td>
<td>DOE</td>
</tr>
<tr>
<td>LM→CS→RA</td>
<td>2.3/1</td>
<td>—</td>
<td>+</td>
<td>0.5</td>
<td>Asympt</td>
</tr>
</tbody>
</table>

Table 2. Presentation, Preoperative Complications, Management and Postoperative Complications in 174 Patients\(^1\)\(^\text{-}^{46}\) with Coronary Arteriovenous Fistula Reported in the Literature

<table>
<thead>
<tr>
<th>Age at dx (yrs)</th>
<th>Preoperative symptoms</th>
<th>Preoperative fistula-related complications</th>
<th>Preoperative symptoms and/or complications</th>
<th>Mean Qp/Qs</th>
<th>Spontaneous closure</th>
<th>Surgical ligation</th>
<th>Surgical complications</th>
</tr>
</thead>
<tbody>
<tr>
<td>All ages (173 pts)</td>
<td>52 pts (30%)</td>
<td>37 pts (21%)</td>
<td>67 pts (39%)</td>
<td>1.60/1</td>
<td>1%</td>
<td>119 (69%)</td>
<td>15 pts (13%)</td>
</tr>
<tr>
<td>Range 1–85</td>
<td>DOE 18%</td>
<td>CHF 12%</td>
<td>MI 4%</td>
<td>73 pts</td>
<td>MI 3%</td>
<td>Ischemia 3%</td>
<td>Recurrence 4%</td>
</tr>
<tr>
<td>Mean 24</td>
<td>Fatigue 6%</td>
<td>MI 4%</td>
<td>Angina 12%</td>
<td>2%</td>
<td>76 (80%)</td>
<td>MI 0%</td>
<td>Ischemia 1%</td>
</tr>
<tr>
<td>Mean 24</td>
<td>Angina 12%</td>
<td>SBE 3%</td>
<td>Death 6%</td>
<td>43 pts</td>
<td>MI 0%</td>
<td>Ischemia 5%</td>
<td>Death 1%</td>
</tr>
<tr>
<td>&lt; 20 years (95 pts)</td>
<td>9 pts (9%)</td>
<td>10 pts (11%)</td>
<td>18 pts (19%)</td>
<td>1.60/1</td>
<td>2%</td>
<td>76 (80%)</td>
<td>5 pts (7%)</td>
</tr>
<tr>
<td>Range &lt; 19</td>
<td>DOE 4%</td>
<td>CHF 6%</td>
<td>MI 0%</td>
<td>43 pts</td>
<td>MI 0%</td>
<td>Ischemia 1%</td>
<td>Recurrence 4%</td>
</tr>
<tr>
<td>Mean 8.3</td>
<td>Fatigue 4%</td>
<td>MI 0%</td>
<td>Angina 3%</td>
<td>2%</td>
<td>76 (80%)</td>
<td>MI 0%</td>
<td>Death 1%</td>
</tr>
<tr>
<td>Mean 8.3</td>
<td>Angina 3%</td>
<td>SBE 3%</td>
<td>Rupture 0%</td>
<td>43 pts</td>
<td>MI 0%</td>
<td>Ischemia 5%</td>
<td>Death 1%</td>
</tr>
<tr>
<td>≥ 20 years (79 pts)</td>
<td>43 pts (55%)</td>
<td>28 pts (35%)</td>
<td>49 pts (63%)</td>
<td>1.61/1</td>
<td>0%</td>
<td>43 (55%)</td>
<td>10 pts (23%)</td>
</tr>
<tr>
<td>Range 20–85</td>
<td>DOE 35%</td>
<td>CHF 19%</td>
<td>MI 9%</td>
<td>30 pts</td>
<td>MI 7%</td>
<td>Ischemia 7%</td>
<td>Recurrence 5%</td>
</tr>
<tr>
<td>Mean 43.3</td>
<td>Fatigue 8%</td>
<td>MI 9%</td>
<td>Angina 22%</td>
<td>1%</td>
<td>MI 7%</td>
<td>Ischemia 7%</td>
<td>Death 7%</td>
</tr>
<tr>
<td>Mean 43.3</td>
<td>Angina 22%</td>
<td>SBE 4%</td>
<td>Death 14%</td>
<td>2%</td>
<td>MI 7%</td>
<td>Ischemia 7%</td>
<td>Death 7%</td>
</tr>
</tbody>
</table>

Abbreviations: MI = myocardial infarction; SBE = subacute bacterial endocarditis; for other abbreviations see table 1.
patients under age 20 years, 91% were asymptomatic, 11% had fistula-related complications and 19% had symptoms or complications. The mean Qp/Qs of patients under age 20 years was 1.6:1. The mean shunt size did not differ significantly between those with symptoms or complications and those without — 1.9:1 vs 1.5:1 (p = 0.075). Only two patients reported previously had documented spontaneous CAVF closure. Of the 76 patients younger than age 20 years who underwent CAVF ligation, five had perioperative complications which were described as recurrent CAVF murmurs in four, and one patient reported early in the surgical experience with this entity died.

Among CAVF patients 20 years of age or older, 55% were symptomatic, 35% had related complications, including 14% who died, and 63% had symptoms or complications. The mean Qp/Qs in reported patients in the older group was identical to that in the younger group — 1.6:1. Among older patients the mean shunt size was not significantly larger in those with symptoms or complications than among those without — 1.6:1 vs 1.4:1 (p = 0.067). There was, however, a significant difference in the Qp/Qs between those older patients with only complications and those with neither symptoms nor complications, 1.7:1 vs 1.4:1 (p < 0.05). In the 43 older patients who underwent CAVF ligation, significant clinical complications related to surgery occurred in 23%, including MI in 7% and death in 7%.

Discussion

Despite many reports, management of CAVF patients has still not been clearly defined. Patients who have CAVF-related symptoms or complications, and those who have large shunts or significant associated cardiac lesions, are generally felt to warrant CAVF ligation. However, most patients with CAVF are asymptomatic and have a relatively small Qp/Qs and their management with elective CAVF ligation is still controversial.

Optimal management of CAVF patients is difficult to assess because most reports of CAVF have been limited by small numbers of patients. Previous investigators have discussed the incidence of symptoms and complications and the results of surgical ligation in patients with CAVF but have not, however, separated these patients by age. We found in our patients, and in our review of the literature, that separating the older and younger patients clarifies both clinical course and management.

As shown in tables 1 and 2, young patients with CAVF are generally asymptomatic and rarely have related complications. For this reason and because of the potential for surgical complications and the rare occurrence of spontaneous CAVF closure, the necessity for elective CAVF ligation has been questioned. Consideration of patients' ages, however, discloses the high incidence of symptoms and complications in older patients, which is otherwise not evident because it is minimized by the large number of asymptomatic, uncomplicated younger patients. Therefore, we believe that elective CAVF ligation in childhood constitutes optimal management. The concept of nonsurgical management of these patients has been so entrenched that one-half of our own study patients (most not under our direct care) have not yet had CAVF ligation.

In general, Qp/Qs in CAVF patients is small regardless of age, and in many, a shunt is not detectable. While mean shunt size was larger in both younger and older patients with symptoms or complications compared with those without, these differences were not statistically significant. It is still likely, however, that shunt size contributes to clinical difficulties with advancing age since there was a significant difference in mean Qp/Qs in older reported patients who had complications alone compared with those who had neither symptoms nor complications, 1.7:1 vs 1.4:1 (p < 0.05). However, for any given patient this small difference in shunt size is not necessarily significant.

Patients with CAVF should have elective ligation, as should patients with asymptomatic patent ductus arteriosus, who, much like the young CAVF patient, rarely have symptoms or complications when young, although incidence of both increase with age. As in other patients with left-to-right shunts, it is likely that the appearance of symptoms and complications with advancing age in CAVF patients reflects the burden of chronic cardiac volume overload, particularly when associated with acquired cardiac and non-cardiac problems that also occur more frequently with advancing age.

The frequent occurrence of angina pectoris in our patients (38%) reflects our relatively older patient population, in whom atherosclerotic coronary artery disease (ASHD) is common. Significant coronary lesions were documented in two of our patients by angiography. In CAVF patients who have associated ASHD, even relatively small shunt flow may exacerbate angina pectoris, and CAVF ligation alone resulted in the disappearance of angina in two of our patients (table 1). There have also been reports suggesting that a "coronary steal" secondary to the CAVF alone causes angina and ischemia in the absence of associated coronary artery disease. Despite the common occurrence of angina in CAVF patients, MI is rare in our experience and in the literature, although it is not an infrequent perioperative complication in older patients undergoing ligation (7%).

Bacterial endocarditis is a rare complication in patients with CAVF but because of its drastic consequences, both pre- and postoperative antibiotic prophylaxis seems advisable. Progressive aneurysmal dilatation of CAVF has been reported; however, documentation of this requires long-term serial angiographic studies. Fistula dissection and rupture are rare in CAVF, and we found only a single reported patient with this complication. We also found only a single report of moderate pulmonary hypertension in our review of the literature, and in no patient was there documented severe pulmonary obstructive disease in the absence of associated cardiac lesions.

Surgical ligation of CAVF can generally be ac-
concluded without cardiopulmonary bypass; however, patients in whom CAVF closure is performed from within the heart, or in whom exposure is difficult, need extracorporeal support. Careful delineation and preservation of myocardial perfusion is essential in preventing intraoperative myocardial ischemia and infarction. There have been excellent reviews of the technical aspects of CAVF ligation. In our study and in the literature, significant surgical complications in the young CAVF patient were rare (7%), and we could find only a single surgical death, which was reported in the early years of the surgical experience with CAVF. In contrast, CAVF ligation in older patients had a higher incidence of complications (23%) and significant clinical complications, including myocardial infarction (7%) and death (7%).

In summary, elective CAVF ligation in childhood constitutes optimal management, even in the asymptomatic patient. Although older patients with CAVF may remain asymptomatic, not develop complications, or have spontaneous CAVF closure, it is important to remember that most unoperated patients develop both symptoms and fistula-related complications with increasing age, and incur increased morbidity and mortality when ligation is performed later in life.

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References

15. Verani MS, Lauer RM: Echocardiographic findings in right coronary artery-right ventricular fistula. Am J Cardiol 35: 444, 1975
43. Valdivia E, Rowe GG, Angevine DM: Large congenital aneurysm of the right coronary artery. AMA Arch Pathol 63: 168, 1957
60. Lowenstein I: Eien seltene miss bildung der coronargefasse. Frankfort Ztsch Path 43: 63, 1932
68. Ogden JA: Congenital variations of coronary arteries. Thesis, Yale University School of Medicine, 1968

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