Coronary Arteriovenous Fistula

By Israel Steinberg, M.D., Janet S. Baldwin, M.D., and Charles T. Dotter, M.D.

Angioangiography and physical findings established the diagnosis of a coronary arteriovenous aneurysm in a 6-year-old asymptomatic schoolgirl. During the 7 years that have ensued, her heart size, electrocardiogram, and general condition have remained normal. Review of the literature discloses 13 autopsy-proved coronary artery fistulas and 8 cases wherein the diagnosis was established by exploratory thoracotomy or cardiac catheterization. The data indicate a wide variation in the clinical significance of the lesion and lead to the conclusion that management is best established on an individual basis.

Abnormal communications between coronary arteries and the coronary venous system constitute arteriovenous lesions in the strictest sense of the word. To use the term arteriovenous with reference to communications between the coronary arteries and structures such as the right atrium, right ventricle, and the pulmonary artery is open to criticism from a semantic point of view, but appears to have become accepted through common usage. In this report, arteriovenous refers to any fistulous communication through which blood is shunted into vascular channels without first passing through capillaries. Although all coronary fistulas exhibit dilatation usually aneurysmal in degree, the converse is not true (i.e., not all coronary aneurysms are due to fistulas). Coronary arteriovenous fistulas are potentially serious lesions that have been successfully treated surgically; their diagnosis is, therefore, a matter of concern to all physicians. Thirteen of 21 previously reported coronary arteriovenous fistulas were revealed at autopsy and in 8 additional cases the diagnosis was established during life by thoracotomy or cardiac catheterization.\(^1\)-\(^{23}\) In the case to be described, a continuous high-pitched "superficial" murmur, best heard to the left of the lower sternum, suggested the presence of an arteriovenous fistula while angioangiography demonstrated aneurysmal dilatation of a coronary vessel.

Case Report

A 6-year-old schoolgirl was referred for consultation in May 1949 because of a heart murmur first heard a year previously. The medical history indicated that her birth followed a normal pregnancy and that physical development and functional capacity had been normal. Under medical observation she has continued to develop normally to her present age of 13 years. She has always been an energetic child, participating in all ordinary childhood activities save for highly competitive sports.

On physical examination, abnormal findings were limited to the heart and have shown no significant change during the 7-year period of observation. The pulse and apex beat were normal. A thrill was not detected. The intensity of the heart sounds was unremarkable. Normal duplication of the second sound in the pulmonary area was observed with the patient in the supine position. A striking cardiac murmur was present. This murmur was loudest in the left fourth and fifth interspaces at the midclavicular line, less intense at the left sternal border, and faintly audible at the angle of the left scapula. It was not transmitted into the axilla.

A peculiar superficial quality was noted by all observers. The murmur, moderately high pitched and hollow in quality, began late in systole while its greatest intensity occurred during early diastole, throughout which phase it was continuous (fig. 1). The intensity became markedly accentuated during expiration. Blood pressure in the arms was 106/72 mm. Hg; in the legs, 115/80.

Roentgenographic examination of the heart, electrocardiographic studies and functional tests were negative and have remained so.

Prior to angioangiography, the diagnosis of arteriovenous fistula involving diaphragmatic or anterior chest wall vessels was suspected because of the location, late onset, and very superficial quality of the continuous murmur. Angioangi-
2nd Rt. i.s. R
d
2nd Lt. i.s. L
d
4th Lt. i.s. L
d
Apex SI SM SD

Fig. 1. Stethogram showing maximum focus of the continuous murmur at fourth left interspace. Note accentuation throughout mid and late third of diastolic phase. (Tracing courtesy of Dr. J. Scott Butterworth.)

ography (figs. 2, 3), performed January 10, 1952, revealed a tortuous, markedly dilated vessel or group of vessels apparently located in the anterolateral wall of the heart and filling simultaneously with the ascending aorta. Film detail was impaired by patient movement, and it was consequently impossible to delineate with certainty the proximal portion of the involved coronary artery, apparently the right, or the site of the abnormal communication. Considered together, however, the angiographic and physical findings are regarded as diagnostic of a coronary arteriovenous fistula with aneurysmal dilatation of the shunt-bearing vessels. The late onset and maximum diastolic intensity of the murmur are consistent with shunting during the diastolic period of coronary blood flow; the area of maximum intensity and superficial quality of the murmur are consistent with the course of a coronary artery.

During the 7-year period of observation, there has been no disproportionate increase in heart size. Physical development and functional capacity remain excellent. It is planned to continue this patient's medical observation on a routine basis. Should there be subsequent appearance of symptoms or functional disability, surgery will be reconsidered and in this event coronary arteriography and cardiac catheterization can be employed to achieve accurate anatomic and functional diagnoses.

**Review of Literature**

Cases of coronary artery fistula have been included in published reports dealing with other types of coronary lesions, particularly aneurysms. In certain instances the published evidence suggests that abnormal communications were overlooked; in others, mention of arteriovenous fistula appears to have been incidental to the description of coronary aneurysms. This is putting the cart in front of the horse, since arteriovenous fistulas are an important cause of coronary aneurysm. Scott's recent review of coronary artery aneurysms included 5 stated examples of abnormal communications in a total series of 48 aneurysms. A review of the literature on coronary aneurysm indicates that there has been a general failure to recognize the etiologic importance (and sometimes the presence) of arteriovenous fistulas in this condition. Twenty-two cases of coronary arteriovenous fistula, briefly summarized in table 1, are reviewed in this report in order to demonstrate the features of this lesion. Except for case 22, our case, all were collected from previously published case reports. Certain cases purposely were not included in this review.

**Description of Twenty-One Previously Reported Cases of Coronary Arteriovenous Communications**

Case 1. Abbott in 1908 reported the findings in a 60-year-old woman who died accidentally; detailed data were not recorded. The left coronary artery took anomalous origin from the pulmonary artery and there was a "crab apple sized" dilatation of the right coronary artery close to its origin from the aorta, believed to be due to a communication with the anomalous left coronary artery. Abbott stated, "The course of blood was towards the pulmonary artery."

Case 2. Halpert described a case of congenital arteriovenous fistula wherein a branch of the right coronary artery was aneurysmally dilated and communicated with the coronary sinus via an anastomotic loop that exhibited histologic characteristics of both arteries and veins. Aneurysmal, serpentine dilatation involved the structures through which the abnormally shunted blood passed; branches of the right coronary artery distal to the "feeder" branch were not remarkable. Although cardiac symptoms or signs were not known to have been present, the heart weighed 500 Gm. and the left ventricle was hypertrophied.

Case 3. Nagoya and Takahashi described a marked, diffuse, serpentine aneurysmal dilatation of the circumflex branch of the left coronary artery that communicated directly with the coronary sinus (also dilated). Cardiac symptoms had been present for 10 years prior to death from myocardial
Fig. 2. Coronary arteriovenous fistula in an 8-year-old asymptomatic girl. **Top.** Frontal angiocardiogram showing the left atrium, left ventricle, and aorta. Below the left ventricle there is a cluster of large coiled vessels (arrow). **Bottom.** Tracing of top.

Fig. 3. Same case as figure 2. **Top.** Lateral angiocardiogram. Cirsoid, markedly dilated coronary vessels (arrow) fill simultaneously with the aorta and are shown near the apex of the heart. **Bottom.** Tracing of top.

Infarction of their 72-year-old patient, and a murmur had been audible for many years.

**Case 4.** Löwenheim found a right coronary arteriovenous fistula in a 62-year-old man who exhibited cardiac failure and hypertension (260/100 mm Hg) for at least 2 years prior to death. At autopsy, the heart weighed 835 Gm, the left ventricle being markedly hypertrophied.

**Case 5.** Harris reported the case of a 43-year-old man stated to have been free of cardiac symptoms (no clinical data were recorded save for the fact that death was due to cerebral hemorrhage). Autopsy revealed a diffuse, cirsoid aneurysmal dilatation of the right coronary artery that communicated with the right atrium through a 1-mm opening. The heart weighed 380 Gm.
<table>
<thead>
<tr>
<th>Age, race, sex</th>
<th>Reference, author, year (Diagnosis by)</th>
<th>Pathologic data</th>
<th>Clinical data</th>
<th>Reported symptoms of cardiac disease</th>
<th>Remarks</th>
</tr>
</thead>
<tbody>
<tr>
<td>Case 1 60 WF</td>
<td>Abbott¹ 1908 (Autopsy)</td>
<td>Dilated RCA → PA via anom. LCA. Aneurysm size of a &quot;crab apple.&quot; Heart weight not stated</td>
<td>Blood pressure: Not stated</td>
<td>Murmurs: None described</td>
<td>No</td>
</tr>
<tr>
<td>Case 2 54 WM</td>
<td>Halpert² 1930 (Autopsy)</td>
<td>Diffusely dilated RCA → dilated coronary sinus via interven. loop which had charac. of art. and vein. Heart weight = 500 Gm.</td>
<td>Blood pressure: 75/70</td>
<td>Murmurs: Systolic only</td>
<td>No</td>
</tr>
<tr>
<td>Case 3 72 WM</td>
<td>Nagoya³ 1932 (Autopsy)</td>
<td>LCA circumflex → coronary sinus. Diffuse serpentine dil. involved structures. Heart weight = ?</td>
<td>Blood pressure: 280/100</td>
<td>Murmurs: None described</td>
<td>Increasing symptoms of heart failure for at least 2 years.</td>
</tr>
<tr>
<td>Case 4 62 WF</td>
<td>Lowenheim⁴ 1932 (Autopsy)</td>
<td>RCA → R. coronary vein via &quot;narrow&quot; orifice. Walls thicken. Diffuse aneurysmal dilat. involved parts. Cardiac hypertrophy, esp. LV. Heart weight = 885 Gm.</td>
<td>Blood pressure: 150/65</td>
<td>Murmurs: None described</td>
<td>Not recorded</td>
</tr>
<tr>
<td>Case 5 43 WM</td>
<td>Harris⁵ 1937 (Autopsy)</td>
<td>Branch of RCA → RA via 1 mm. orifice; diffuse cirrhotic dil. of coronary sinus and involv. parts RCA. R. heart much dilated. Heart weight = 380 Gm.</td>
<td>Blood pressure: 120/70</td>
<td>Murmurs: None described</td>
<td>No</td>
</tr>
<tr>
<td>Case 6 43 WF</td>
<td>Emminger⁶ 1946 (Autopsy)</td>
<td>Diffusely dil. RCA → Great card. vein thru small orif. Coronary sinus → RA by 2 ostia; origin RCA = 1.4 cm. Heart weight not given</td>
<td>Blood pressure: 150/65</td>
<td>Murmurs: None described</td>
<td>Not recorded</td>
</tr>
<tr>
<td>Case 7 84 WM</td>
<td>Scott⁷ 1948 (Autopsy)</td>
<td>LDCA and anom. branch RCA → PA. 17 aneurysms of shunt-bearing arts. Non-shunt bear. coronaries normal. Largest aneurysm clotted. Heart weight = 780 Gm.</td>
<td>Blood pressure: 120/70</td>
<td>Murmurs: None described</td>
<td>No</td>
</tr>
</tbody>
</table>

* In living patients, age given at time diagnosis was established.
| Case 8 1 NM | Brown⁸ 1949 (Autopsy) | Accessory LCA → RV. Narrowest diameter was 5 mm. Heart weight not stated | 98/65 | Machinery | No | Sudden death attributed to tracheobronchitis in absence of other reason |
| Case 9 76 WF | Baylis⁹ 1952 (Autopsy) | Serpentine, dilated branch of LADCA received small branch of RCA → PA. AV orifice = 1 cm. Heart dil. and hypert. Heart weight = 430 Gm. | 160/70 | Machinery | No evidence of functional disability except term. CHF. Pt. steadfastly denied ever having had dyspnea and orthopnea | Death from terminal heart failure and metastatic carcinoma of breast. Lesion believed due to anom. origin of an accessory coronary artery from the pulmonary artery |
| Case 10 85 WM | Colbeck¹⁰ 1954 (Autopsy) | Fusiform aneurysm of RCA through a diffusely dil. serpentine vessel → RA via 4 small orifices. Card. hyper.; marked pulmonary edema. Heart weight = 554 Gm. | 170/80 | Systolic | Heart failure with dyspnea and edema for 18 months before death | Marked enlargement of LV on x-ray. Death from acute pulmonary edema |
| Case 11 58 WF | Davison¹¹ 1955 (Autopsy) | LCA → coronary sinus. Cardiac hypertrophy. Heart weight = 490 Gm. | 125/55 | Machinery | Heart failure—ascribed directly to shunt | Death due to lesion. Cardiac cath.: PA pressure = 75/25 mm. Hg. Shunt = 3.9 l./min.; 55% of LV output |
| Case 12 55 WM | Knoblich¹² 1956 (Autopsy) | "AV fistula of the heart" with serpentine dil. LCA entering aneurysm sac and thereafter → vein leading to dil. myocard. capillaries. Heart weight = 400 Gm. | 220/110 | "Diastolic" | Heart failure and symptoms of hypertension | Exact route of AV shunt not clear from findings. Presence of significant shunt-flow evidenced by dilated vessels. Death from subarachnoid hemorrhage |
| Case 13 25 NF | Valdivia¹³ 1957 (Autopsy) | Huge arteriovenous aneurysm RCA which → RA. Heart weight = 1800 Gm. (Including aneurysm). | 120/70 | "Double" | Chronic heart failure with palpitation, fatigue and dyspnea | Neither angiocardiology nor exploratory thoracotomy allowed a correct diagnosis during life. Symptoms and death due to arteriovenous fistula |
| Case 14 * 15 WM | Bioreck¹⁴ 1947 (Surgery) | Probable anomalous dilated branch of LCA → PA. Anomalous branch ligated with disap. of thrill | 135/80 | Continuous | Decreased exercise tolerance | Surgery done for suspected PDA. Ligation of feeder artery resulted in disappearance of murmur. No recent follow-up |

* In living patients, age given at time diagnosis was established.
### Table 1.—Case Summaries—Continued

<table>
<thead>
<tr>
<th>Age, sex</th>
<th>Reference, author, year (Diagnosis by)</th>
<th>Pathologic data</th>
<th>Blood pressure</th>
<th>Murmurs</th>
<th>Reported symptoms of cardiac disease</th>
<th>Remarks</th>
</tr>
</thead>
<tbody>
<tr>
<td>Case 15</td>
<td>Paul15 1949 (Surgery)</td>
<td>Dilated, tortuous RCA → coronary vein at its junction with coronary sinus. No attempted ligation</td>
<td>110/75</td>
<td>Continuous</td>
<td>No</td>
<td>Pre-op. diagnosis of arteriovenous fistula; exact location in doubt but coronaries consid. Ligation not attempted. Pt. was asymptomatic (and still had murmur) at age 16</td>
</tr>
<tr>
<td>Case 16</td>
<td>Gross17 1949 (cited by Paul) (Surgery)</td>
<td>Cirsoid, bulging nest of vessels in wall of LV. Probable drainage into low pressure structure on basis of thrill. No attempted ligation</td>
<td>Not stated</td>
<td>Continuous</td>
<td>Not clear. “Know heart disease since infancy.” Had diagnosis of PDA and SBE made pre-op.</td>
<td></td>
</tr>
<tr>
<td>Case 17</td>
<td>Sondergaard19 1955 (Surgery)</td>
<td>LCA → RV. Also PDA. Both ligated with disappearance of murmurs</td>
<td>Not stated</td>
<td>Continuous double</td>
<td>Dyspnea on exertion</td>
<td>Operation for suspected PDA. Had been treated for SBE 6 weeks before operation. Not ligated. Apparently living at age 24</td>
</tr>
<tr>
<td>Case 18</td>
<td>Davis21 1956 (Surg. &amp; Cath.)</td>
<td>LCA → infundibulum of RV. Ligated with disappearance of thrill</td>
<td>110/80</td>
<td>Continuous</td>
<td>No</td>
<td>Surgery for suspected PDA. Ligation resulted in apparent cure. Pre-op. cardiac cath. showed shunt volume of 1.7 L./min., 26% of LV output</td>
</tr>
<tr>
<td>Case 19</td>
<td>Moxen22 1956 (Surgery)</td>
<td>Tortuous, cirsoid dilatation of LADCA which → LA (probably). Ligation with disappearance of murmur</td>
<td>130/60</td>
<td>Continuous also systolic</td>
<td>“Slight dyspnea on extreme exertion”</td>
<td>Normal cardiac catheterization. Diagnosis made at surgery. Ligation resulted in apparent cure</td>
</tr>
<tr>
<td>Case 20</td>
<td>Walther23 1957 (Catheter)</td>
<td>Coronary artery → middle cardiac vein</td>
<td>130/84</td>
<td>Continuous</td>
<td>No</td>
<td>Cardiac catheterization: Fully saturated blood from middle cardiac vein. Shunted blood amounted to 47% of LV output</td>
</tr>
<tr>
<td>Case 21</td>
<td>Walther23 1957 (Surgery)</td>
<td>RCA → PA or pulmonary conus. Ligation not attempted</td>
<td>120/70</td>
<td>Continuous</td>
<td>No</td>
<td>Surgery for suspected PDA. Ligation not attempted. Post-op. Cardiac Cath.: PA pressure = 33/17 mm. Hg. Shunt volume = 2.0 L./min., 29% of LV output</td>
</tr>
<tr>
<td>Case 22</td>
<td>Present case (Angiography)</td>
<td>Coronary arteriovenous fistula</td>
<td>106/72</td>
<td>Continuous</td>
<td>No</td>
<td>No evidence of disability during 7 years of medical observation</td>
</tr>
</tbody>
</table>

*In living patients, age given at time diagnosis was established.
Case 6. Emuninger* described an arteriovenous fistula consisting of a serpentine, diffusely dilated right coronary artery that communicated with the great cardiac vein in a 43-year-old woman. The fistula was thought not to have caused cardiac disability; death was due to uremia complicating glomerulonephritis. Her blood pressure had been 150/65 mm Hg. Detailed clinical data were not reported. In addition to the fistula, autopsy revealed that the markedly dilated coronary sinus opened into the right atrium through 2 fairly small ostia.

Case 7. Scott¹ in 1948 reported the case of an 84-year-old man without cardiac symptoms who died following prostatectomy. The patient's chest x-ray had revealed a right atrial aneurysm. At autopsy, there were found to be 17 aneurysmal dilatations involving 2 vessels, an accessory right coronary artery and a branch of the left anterior descending coronary artery. The largest aneurysm measured 10 by 8 by 8 cm and was almost or completely clotted. A murmur had not been heard, and it is believed possible that the occlusion by laminated thrombus was responsible for this. The 2 aneurysm-bearing coronary arterial branches joined together and thereafter, through a common vessel, affected a small (1 mm) communication with the pulmonary artery. As usual, the non-shunt-bearing coronary arteries were normal. The heart weighed 780 Gm.

Case 8. Brown and Burnett⁴ described the case of a 13-month-old boy whose symptoms suggested patent ductus arteriosus, and who died of a purulent tracheal bronchitis. Physical findings included the demonstration of a pronounced thrill along the left sternal border. In addition, a continuous, machinery-like murmur was loudest in the third and fourth left interspaces. Both thrill and murmur seemed close to the surface. Blood pressure was 98/65 mm Hg. Autopsy revealed the pressure of an abnormal communicating vessel arising adjacent to the normal left coronary artery and passing into a small recess in the right ventricle. The authors regarded it as a large left coronary artery "which had anastomosed early in development with intertrabecular spaces in the myocardium and thus established communication with the chamber of the right ventricle." The right ventricle was stated to be hypertrophied and moderately dilated. Neither the weight of the heart nor the appearance of the left ventricle was included in the description of pathologic findings.

Case 9. Bayliss and Campbell's case⁵ consisted of a diffusely dilated communication between coronary and pulmonary arteries in a 76-year-old woman who was said to have been in good health until 2 years prior to death of terminal heart failure associated with carcinoma of the breast. She had had a loud, continuous murmur for at least 30 years. Blood pressure was 160/70 mm Hg. Autopsy revealed that a large, tortuous, sacular aneurysm arose from the left anterior descending coronary artery and communicated with the right anterior sinus of Valsalva of the pulmonary artery. The heart weighed 430 Gm.; there was moderate enlargement and hypertrophy of both ventricles.

Case 10. Colbeck and Shaw⁶ reported a coronary arteriovenous fistula in an 85-year-old man whose death in acute pulmonary edema occurred in a setting of heart failure of at least 18 months' duration. A harsh, grade IV systolic murmur without a thrill was present and the blood pressure was 170/80 mm Hg. The electrocardiogram showed atrial fibrillation and left bundle-branch block. At autopsy, the heart weighed 554 Gm and left ventricular hypertrophy and dilatation were present. There was also moderate enlargement and hypertrophy of the right atrium and ventricle. The lesion consisted of a large, fusiform aneurysm of the right coronary artery with connection to the right atrium by means of a serpentine anomalous communicating vessel.

Case 11. Davison et al.⁷ reported a case wherein autopsy disclosed an arteriovenous fistula consisting of a diffuse, cirsoid dilatation of the circumflex branch of the left coronary artery which, after a serpentine course, entered an aneurysmally dilated coronary sinus. The patient, a 58-year-old woman, had noted palpitations for 10 years before death. Although at first clinical and electrocardiographic studies were normal, she subsequently developed a systolic murmur, increasing exertional dyspnea, orthopnea, fatigue, and the onset of atrial fibrillation. Ten months before death, a systolic and diastolic machinery-type murmur was heard over the pulmonary area. A systolic thrill was present. Blood pressure was 125/55 mm Hg. Cardiac catheterization disclosed the pulmonary arterial pressure to be 75/25 mm Hg and indicated the presence of a major left-to-right shunt at the atrial level. The total cardiac output was determined and equaled 7.1 L per minute; while the shunt volume was 3.9 L per minute (55 per cent of the total cardiac output). Death was ascribed to congestive heart failure. At autopsy, the heart weighed 490 Gm.; all its chambers showed dilatation and hypertrophy.

Case 12. Knoblich and Rawson⁸ reported the autopsy finding of a long, tortuous, dilated, sclerotic, descending branch of the left coronary artery that communicated with a sacular coronary vein located in the ventricular septum. The patient, a 55-year-old man, had had systemic hypertension for years with blood pressures as high as 220/110 mm Hg. Death was due to rupture of a congenital aneurysm of the basilar artery. A cardiac murmur had been present for at least 37 years; hypertension and cardiac enlargement had been known to
CORONARY ARTERIOVENOUS FISTULA

exist for at least 10 years. A harsh, grade III, diastolic apical murmur was heard repeatedly. Electrocardiographic findings suggested acute posterior-wall infarction with extension into the septum. At autopsy, a coronary arteriovenous fistula located in the interventricular septum was encountered. This was presumed to have caused the electrocardiographic changes. The heart weighed 600 Gm., the left ventricular wall measured 17 to 22 mm. in thickness, that of the right 5 mm.

Case 13. Valdivia et al.\(^13\) recently described the autopsy finding of a large, presumably congenital aneurysm of the terminal end of the right coronary artery communicating with the right atrium. Their patient, a 25-year-old woman, had been known to have a cardiac murmur since childhood. X-ray studies 4 years prior to death demonstrated marked enlargement of the heart shadow. Angiocardiography done at that time did not result in satisfactory opacification. An exploratory operation established the presence of a large vascular tumor or aneurysm that could not be removed. Death occurred 4 years later due to heart failure. The diagnosis had not been established during life, despite angiocardiography and exploratory thoracotomy. At autopsy, a huge arteriovenous aneurysm was discovered. Its external measurements were 17 by 23 by 14 cm. The heart and aneurysmal mass together weighed 1,800 Gm. The aneurysmal cavity emptied into the right atrium through a 2-mm. opening.

In summary, of thirteen autopsied cases of coronary arteriovenous fistula collected from the literature, the structures in communication with coronary arteries were: coronary sinus, 4 cases (cases 2, 3, 5, and 11); coronary veins, 3 cases (cases 4, 6, and 12); pulmonary artery, 2 cases (cases 1 and 7); right atrium, 2 cases (cases 10 and 13); and right ventricle, 1 case (case 8).

CORONARY ARTERIOVENOUS MALFORMATIONS ESTABLISHED DURING LIFE

In 8 previously reported instances, exploratory thoracotomy (7 cases) or cardiac catheterization (1 case) has established with reasonable certainty the diagnosis of coronary arteriovenous fistula. In 4 of these 8 patients, ligation of the communicating vessel was accomplished with apparent success.

Case 14. Biörk and Crafoord\(^14\) in 1947 reported the finding at surgery of a communication between an aberrant branch of the left coronary artery and the main pulmonary artery. Their patient, a 15-year-old boy, had had decreased exercise tolerance for several years and exertional dyspnea since age 14. At age 7, a presystolic murmur was noted over the pulmonary area. It later became accentuated, exhibited systolic and diastolic components, and finally developed into a continuous murmur. The preoperative blood pressure was 135/80 mm. Hg. Radiologic examination demonstrated a rounded left heart border, prominent pulmonary artery segment, and hilar dance. The electrocardiogram was normal. Although the murmur was not regarded as characteristic, it suggested patent ductus arteriosus. At surgery, a thickened, abnormal vessel, probably arising from the left coronary artery, was seen to enter a pea-sized aneurysm-like sac on the pulmonary artery. That the continuous murmur was caused by this vessel was established by its ligation and the subsequent recovery of the patient. Four months later at follow-up examination, there was recorded a moderately harsh systolic but no diastolic murmur.

Case 15. Paul et al.\(^15\) described a coronary arteriovenous communication that was discovered at surgery for a suspected patent ductus arteriosus. In their patient, a 9-year-old boy, blood traversed a diffusely dilated right coronary artery, a large venous plexus, and a coronary vein en route to the coronary sinus. No murmur had been heard at birth, but at the age of 2, in association with a transient hemiplegia of unknown etiology, a loud systolic murmur was first noted. Following scarlet fever at age 5, a loud apical systolic murmur, followed by a third heart sound and diastolic rumble, was described. Subsequently, the diastolic rumble disappeared, while there was a loud, continuous murmur along the lower right sternal border with maximum intensity occurring during systole. Blood pressure was 110/75 mm. Hg. The preoperative differential diagnosis was arteriovenous fistula involving the chest wall, the lung, or the coronary vessels. At operation, the coronary arteriovenous fistula was encountered, but its ligation was not attempted for fear of jeopardizing the myocardial blood supply. The patient continues to be well and is free of symptoms at 23 years of age.\(^16\)

Case 16. Gross,\(^17\) at thoracotomy for suspected patent ductus arteriosus, encountered a probable arteriovenous fistula of the coronary circulation in a 16-year-old boy. Heart murmurs had been present since birth and, preoperatively, a harsh, continuous murmur and thrill were maximum over the third and fourth left intercostal spaces adjacent to the sternum. Six weeks before surgery, the diagnosis of streptococcal endocarditis complicating patent ductus arteriosus had been made and a course of penicillin was given with apparent good results. At surgery, complete identification of the components of the fistulous communication was not possible, although a nest of bulging, thin-walled, vascular structures about 3.5 cm. in diameter and associated with a thrill was encountered.
in the lateral wall of the left ventricle, slightly below the tip of the left atrial appendage. Ligation was not attempted. Gross states that the patient is currently performing hard manual labor without difficulty.\textsuperscript{19}

\textbf{Case 17.} Sondergaard\textsuperscript{20} ligated an aneurysmal connection between the left coronary artery and the right ventricle of an underdeveloped 11-year-old boy. Exertional dyspnea had been present since age 5, at which time a murmur had been noted. Preoperative physical examination revealed a continuous murmur (with thrill) possessing 2 areas of maximal intensity, one in the second and the other in the fourth left interspace. The second pulmonary sound was accentuated. At operation, a patent ductus arteriosus and an aneurysmal fistula were encountered. Both were ligated with the disappearance of all murmurs. Recent information discloses that the patient is now in excellent health.\textsuperscript{20}

\textbf{Case 18.} Davis et al.\textsuperscript{21} reported ligation of an anomalous left coronary artery communicating with the right ventricle in a 19-year-old Negro girl. Although the patient described no cardiac symptoms, a continuous machinery-like murmur was encountered, maximal in the pulmonary area. The electrocardiogram was normal, and there was no radiologic evidence of cardiac enlargement. Blood pressure was 110/80 mm. Hg. Catheterization of the heart disclosed normal right heart pressures, but indicated the entrance of highly oxygenated blood at the level of the outflow tract of the right ventricle. At operation, there was found to be diffuse dilatation of the left coronary artery, a branch of which entered the infundibular area of the right ventricle. Since temporary occlusion of this vessel resulted in disappearance of the thrill, ligation and division were accomplished. A small segment of the vessel was removed for histologic examination and found to be normal. Sixteen months postoperatively, the murmur had not reappeared. Although the electrocardiogram remained normal, postoperative x-ray films suggested slight cardiac enlargement (possibly due to pericardial reaction).

\textbf{Case 19.} Mozen\textsuperscript{22} reported the ligation of a fistulous vessel connecting a cirroid, aneurysmal dilatation of the anterior descending branch of the left coronary artery and the left atrium. The patient, a 10-year-old boy, had had a heart murmur since infancy, although his growth and development had been normal. Preoperatively, a soft systolic thrill was felt over the apex of the heart, and there was a loud continuous to-and-fro murmur with maximum intensity in the pulmonary area. A lower-pitched systolic murmur was also audible at the apex. Blood pressures ranged between 120/80 and 130/60 mm. Hg. Electrocardiographic and radiologic studies as well as cardiac catheterization revealed no abnormality. At operation, the cirroid aneurysmal dilatation of the left descending coronary artery was found to be about 5 to 6 cm. in length and 0.5 to 1.3 cm. in diameter. Many large branches terminated in the left ventricular myocardium. One branch 1.5 cm. long and 0.5 cm. in diameter apparently emptied into the left atrium; ligation of this branch resulted in complete obliteration of the harsh thrill that it caused. It was stated, "this was a fistula which was carrying large amounts of blood from the high pressure aneurysm into the low pressure left atrium." In their report, published a year after surgery, the author stated that "heart sounds have remained normal."

\textbf{Case 20.} Walther et al.\textsuperscript{23} reported the findings in 2 patients where the diagnosis of coronary arteriovenous malformation had been established during life. The first patient was an asymptomatic 8-year-old white girl who had developed normally in every respect and in whom a heart murmur had been heard during infancy. Her blood pressure was 130/84 mm. Hg, and the principal abnormality consisted of a grade V, roaring, continuous murmur (with thrill) maximal in the fourth and fifth left intercostal spaces close to the sternum. Electrocardiogram was unremarkable; slight left atrial enlargement was observed radiologically. At cardiac catheterization, the coronary venous system was entered and fully saturated blood withdrawn from the middle cardiac vein. The shunt-volume was calculated to be 5.2 L. per minute as compared to a cardiac output of 5.9 L. Pulmonary arterial pressure was 30/13 mm. Hg, and the right ventricular pressure was 30/6 mm. Hg.

\textbf{Case 21.} A second patient\textsuperscript{24} reported by Walther et al. was a 14-year-old girl, who was known to have had a heart murmur since infancy, developed normally, and was asymptomatic. Blood pressure was 120/70 mm. Hg. A continuous machinery-type murmur was audible at the left sternal border, maximal in the second interspace. Radiographs were said to reveal slight increase in the transverse diameter of the heart (the published reproduction speaks for a left ventricular enlargement). The electrocardiogram was normal. Thoracotomy for a suspected patent ductus arteriosus failed to confirm the diagnosis. Instead, a thrill was palpated over the moderately enlarged pulmonary artery. The cause of this thrill was traced to an aneurysmally dilated right coronary artery that entered the right ventricular myocardium just proximal to the pulmonary valve. Since the thrill could not be completely abolished by compression of the dilated right coronary artery, ligation was not done. It was concluded that there was a communication between the right coronary artery and the outflow tract of the right ventricle or pulmonary artery. Cardiac catheterization subsequently revealed a high pulmonary
CORONARY ARTERIOVENOUS FISTULA

artery oxygen content. Calculated shunt volume was 2 L per minute, and was an impressive 28 per cent of the total cardiac output.

DISCUSSION
Pathologic and Physiologic Considerations

Coronary arteriovenous communications are rare. The present series of 22 collected cases, probably the largest to date, indicates that in comparison to coronary arteriovenous fistulas primary tumors of the heart are relatively commonplace lesions since they are reported over 10 times as frequently!

It seems reasonable to postulate that congenital coronary arteriovenous fistulas may result from either or both of 2 forms of embryologic maldevelopment. The first consists of an embryonic arrest in the normal differentiation of the coronary vasculature. Probably the most primitive example of this type of developmental defect is the persistence of embryonal vascular sinusoids in the myocardium. Grant30,31 drew attention to the transient presence of blood-filled intertrabecular spaces within the myocardium of the normally developing mammalian heart. These spaces communicate freely with each other, also with the ventricular cavities and with the developing coronary vasculature. Postulating that arrest or faulty differentiation of the normal intertrabecular spaces gives rise to coronary vascular malformations, Wilson and Grant32,33 cited an example of multiform anomalies of the coronary and cardiovascular systems in a 14-month-old patient wherein blood-filled myocardial spaces communicated with coronary vessels. Burchell34 in 1939 described large endothelium-lined sinuses within the wall of the left ventricle of an adult dog stating that the sinuses appeared to have functioned as arteriovenous fistulas. Such sinusoids have been observed repeatedly in human autopsy material33,35-39 usually in association with anomalous origin of the left coronary artery from the pulmonary artery. In certain instances a significant arteriovenous shunt evidently occurred; in others, it apparently did not. The vascular resistance of such lesions determines the extent of abnormal shunting. Because of the difficulty in assessing shunt-function in reported cases of persistent myocardial sinusoids, these lesions have not been included in the present review of coronary arteriovenous communication.

Certain congenital arteriovenous fistulas are believed to reflect a developmental defect occurring at a later stage in the fetal maturation of the coronary system than that resulting in the persistence of primitive myocardial sinusoids. In support of this, is the fact that while the gross lesion of the usual fistula may appear quite complex, many of the changes are secondary to the shunt. Unlike the undifferentiated sinusoids, the vessels of the more mature lesion bear a reasonable resemblance to arteries and veins. To extend this reasoning, the coronary arteriovenous malformation could be regarded as a more mature lesion than the usual peripheral arteriovenous fistula where innumerable functioning or potentially open sites of transcapillary communication exist. This fortunate difference is probably essential to successful surgery in coronary arteriovenous lesions for if, as in peripheral lesions, new openings were to develop following the ligation of evident sites of communication, death might well be hastened rather than postponed.

The second major mechanism responsible for the formation of coronary artery fistulas involves the anomalous origin of a coronary artery. As was exemplified by Abbott's case1 and by other cases not included in this review,40-43 intercoronary anastomoses may allow a reversal in the direction of flow through the anomalous coronary artery. If the coronary resistance at the anastomotic site is sufficiently low, the pressure gradient is such as to favor flow from the normally originating coronary artery through the anomalous branch into the pulmonary artery or the right ventricle.

Despite the frequency with which the term arteriovenous aneurysm is used, confusion appears to exist concerning the relationship between coronary arteriovenous communications and coronary artery aneurysms.
Authors have reviewed the findings in collected cases of coronary aneurysm without mentioning or stressing the important etiologic role of arteriovenous communications. That the association is important is evidenced by analogous extracoronary arteriovenous shunts. Given a sufficient shunt volume acting over a long enough period, there must result dilatation of the shunt-bearing structures. This is true of the vessels involved in peripheral arteriovenous shunts of any cause; it is true of the structures carrying recirculated blood in patients with interatrial septal defects; and it is also true of coronary arteriovenous communications. In several published cases of "arteriosclerotic" or "mycotic-embolic" coronary aneurysms, diffuse dilatation (usually termed a cirrroid or serpentine aneurysm) points directly to a probably undetected but causative fistulous lesion. Particularly convincing in this respect are cases where the dilatation involves a coronary artery down to and including one of its branches but spares other branches and the vessel itself beyond the point where the dilated shunt-bearing branch is given off. Since the relationship was apparently overlooked, previously reported cases cannot serve as basis for quantitating the etiologic relationship. The frequency with which current reports have dealt with aneurysmal dilatation secondary to arteriovenous communications rather than with coronary artery aneurysms offers evidence of the increasing awareness of this previously unrecognized causative relationship. The study of known instances of shunt-caused aneurysms explains why the primary communication escaped autopsy detection in others. In case 7, for example, 17 aneurysms existed, the largest measuring 10 cm. in maximum diameter, while the orifice of communication was but 2 mm. wide. Similarly, case 13 exhibited an aneurysm measuring 23 cm. in diameter, but a communicating orifice of only 2 mm. An argument that such large aneurysms could not result from such tiny communications is not valid; the contrary, of course, is the case.

It is difficult to explain a genuine continuous murmur on the basis of simple aneurysmal dilatation. It would be illogical to assume that arteriosclerosis is the cause of the majority of coronary aneurysms, since, as has been emphasized by Scott, while coronary aneurysm is rarely seen, coronary arteriosclerosis is, comparatively speaking, neither rare nor as likely to give rise to large dilatations. Furthermore, the fact that arteriosclerosis is present in a coronary artery aneurysm is not surprising, since secondary atheromatous changes occur in a wide variety of aneurysms.

The pathophysiology of coronary fistulous communications is essentially that of arteriovenous fistulas elsewhere in the body with the exception that the former may result in myocardial ischemia in addition to the usual increase in cardiac output. Regardless of the site, in any functional fistulous communication, blood flows from a high pressure system to a low pressure system. If there were no pressure gradient, there would be no flow. As a consequence of the increased volume of flow there occurs dilatation of the affected coronary arteries and the vessels or chambers receiving the shunted blood. The clinical consequences are almost entirely determined by the shunt-volume and its duration. The shunt-volume is, in turn, dependent upon coronary vascular resistance. Thus, when persistent embryonal sinusoids are the sole lesion present, physiologic consequences would tend to be slight due to a fairly high coronary resistance.

Given sufficient shunt-volume acting over a long enough period of time, left ventricular hypertrophy and failure must inevitably occur. In coronary, as in other systemic arteriovenous communications, there may occur an increase in blood volume, heart rate, and pulse pressure, as well as other evidences of high-output failure.

Clinical Features

Age, Sex, and Race. The ages of the 22 patients comprising this series ranged from 13 months to 85 years. In view of the small
series and wide age range, the figure for average age would be more misleading than useful. The 8 living patients were relatively young compared to the autopsied patients. It is not surprising that the published case reports of the living patients are of relatively recent vintage in view of the heightened interest and diagnostic abilities resulting from the recent emergence of surgery as a definitive method of treatment for many congenital cardiovascular lesions. Of the entire group of cases, 12 were male, 10 female. Three patients were Negroes.

*Symptoms and Signs.* Cardiovascular symptoms occurred in 11 of the 22 patients, but the lesion was apparently regarded as responsible for symptoms in only 6 of these; 2 autopsied cases (cases 11 and 13) and 4 surgically proved cases (cases 14, 16, 17, and 19). In 3 or 4 cases, the statement that symptoms had not existed apparently was based upon hearsay rather than clinical observation. The autopsy findings cast doubt upon the contention that coronary arteriovenous fistula is usually a benign lesion. Findings such as those in case 13 leave little doubt as to the potentially serious nature of the anomaly. Cardiac symptoms are those of heart failure due to the increased cardiac output occasioned by a left-to-right shunt. Five cases exhibited clear-cut evidence of chronic heart failure; and among the reported symptoms and signs of this were dyspnea, peripheral edema, pulmonary congestion, and fatigue.

*Laboratory Data.* Electrocardiographic studies in our case and in the other 7 patients diagnosed during life were not remarkable. The electrocardiograms of the autopsied patients were frequently abnormal. Case 7 had premature atrial beats and a shifting pacemaker. Case 9 exhibited left axis deviation and left ventricular predominance (moderate hypertension may have been a contributory factor). Case 11 had a normal electrocardiogram 5 years prior to death but exhibited terminal atrial fibrillation and right axis deviation without ventricular predominance. Case 10 showed atrial fibrilla-

tion and left bundle-branch block. In case 12, there existed for at least 3 years, an unchanging elevation of the S-T segments suggestive of septal injury. The patient had severe hypertension for several years. At autopsy a coronary artery aneurysm, located in the septum, was discovered and believed to be responsible for the electrocardiographic changes. The authors, Knoblich and Rawson, suggested that the fixed electrocardiographic abnormality favored some lesion other than myocardial infarction. Generally speaking, the data from the 22 patients comprising this series suggest that electrocardiographic changes, when they exist, are not very helpful in establishing the diagnosis of coronary arteriovenous communications.

Cardiac catheterization was performed in 5 of the patients of this series. In case 19, shunted blood apparently entered the left atrium and shunt volumes could therefore not be calculated (other findings at catheterization were normal in this patient). In the 4 others, cases 11, 18, 20, and 21, hemodynamic data are available and have been summarized by Walther et al. The observed shunts ranged from 1.7 L per minute to a maximum of 5.2 L per minute. In this lesion, the pulmonary flow is equal to the left ventricular output, and the latter represents the sum of the peripheral flow plus the shunt flow. Expressed as percentages of left ventricular output, the shunts were as follows: case 11, 55 per cent; case 18, 26 per cent; case 20, 47 per cent; case 21, 29 per cent. The mean shunt volume of all 4 patients was 39 per cent. Stated differently, about 40 per cent of the work of the heart was wasted. Pressures in the pulmonary artery of case 11 were 75/25 mm. Hg, indicating the development of pulmonary vascular disease. Case 18 revealed normal right heart pressures, while cases 20 and 21 showed slight pulmonary hypertension (probably this was significant since both were children). It is evident that the determination of blood oxygen content in various regions may be highly significant in establishing the diagnosis
of coronary arteriovenous shunting of the blood, particularly where the communication occurs via a coronary vein that is successfully catheterized. Lacking this, the findings could mimic more common cardiac anomalies such as interatrial and interventricular septal defects, patent ductus arteriosus, or aortico-pulmonary septal defects. Similar blood oxygen findings could result from rupture of an aortic sinus into one of the right heart structures.

X-ray examination by conventional technics is of value insofar as there are present grossly recognizable changes in the cardiovascular silhouette. The demonstration of an aneurysmal dilatation, when it exists, might be of aid in distinguishing coronary arteriovenous fistulas from high interventricular septal defects or patent ductus arteriosus. Radiologic appraisal of heart size offers a useful means of gauging the functional significance of the lesion. In 7 of the autopsied cases, roentgenograms were available; 5 were described and said to reveal cardiac enlargement. Colbeck and Shaw identified intracardiac calcification that at autopsy was found to lie within the coronary artery aneurysm (case 10). In case 13, the chest film showed marked enlargement of the cardiac silhouette due to the massive aneurysm that was present. In Scott’s patient (case 7), a large globular mass causing esophageal displacement was demonstrated adjacent to the right heart border. Of the 8 living patients, Biörck and Crafoord’s patient (case 14) revealed rounding of the left heart border as well as enlargement and increased pulsation of the pulmonary artery and hilar branches. The findings reverted to normal following successful surgical interruption of the fistula. Both of Walther’s patients (cases 20 and 21) were reported to have shown slight enlargement of the heart and prominence of pulmonary arteries on radiographic examination. In the remaining 5 cases, including our own, the heart was normal in size and contour and abnormalities of the pulmonary artery or pulmonary vasculature were not recognized. Following surgery, in case 18, there was slight enlargement of the heart 16 months after operation, a finding possibly attributable to postoperative pericardial changes.

Special contrast visualization offers much in the diagnosis of coronary arteriovenous fistulas. Angiocardiography can be expected to reveal moderate- to large-sized coronary artery aneurysms providing technically adequate studies are made. Neither angiocardiography nor thoracotomy resulted in a correct antemortem diagnosis in case 13; the former because of marked cardiac dilatation and consequent excessive dilution of the contrast agent, the latter because the anatomy of the lesion was such as to discourage manipulation for diagnostic or therapeutic reasons. Angiocardiography was performed in our patient and made the correct diagnosis possible. Cirsoidal dilatation of the feeding coronary artery was clearly visualized, although the site of the fistula was not determined. The study suffered from patient movement during the examination, a factor that now can be eliminated by the use of millisecond exposure technics. It is obviously desirable to employ studies in 2 projections (or stereoscopic sequencing) and rapid serial-recording technics.

Coronary arteriography by the controlled approach of occlusion aortography stands to offer the most precise anatomic diagnosis of coronary arteriovenous fistula that can be achieved during life, and should prove superior to surgical exploration in this respect. To date, there has been no opportunity to demonstrate this conviction. Angiocardiography offers a more useful method for study of the heart and great blood vessels as a whole, but is distinctly inferior in the ability to produce satisfactory coronary visualization. Selective technics such as aortography, while providing optimum opacification of particular regions, suffer from their inability to reveal structures that in terms of blood flow are proximal to the site of injection. Inasmuch as angiocardiography is simpler to perform, bears a somewhat lower risk than aortography, and reveals a
CORONARY ARTERIOVENOUS FISTULA

A greater total amount of information about the heart as a whole, it is recommended as the initial procedure of choice if there is any doubt about the exact nature of the lesion in question. In coronary arteriovenous communications, angiocardiography during opacification of the right heart structures should demonstrate a filling or mixing defect at the site of shunts as large as those in the 4 cases studied by cardiac catheterization. It should be emphasized that any compromise in technical quality, particularly the use of detail-destroying slow exposures will seriously limit the approaches just discussed.

Diagnosis. The diagnosis of a coronary arteriovenous fistula depends upon the evaluation of a continuous precordial murmur especially when its location is unusual. Most continuous, machinery-like murmurs are best heard at the pulmonary area and reflect the presence of a patent ductus arteriosus. A somewhat lower location of the site of maximal intensity might indicate an aortico-pulmonary septal defect or a ruptured aortic sinus aneurysm, 2 other lesions capable of causing similar murmurs. In the latter, the abrupt onset of the murmur in association with chest pain, provides a valuable clinical lead to the diagnosis. Unlike coronary arteriovenous fistulas, pulmonary arteriovenous fistulas often are associated with other vascular anomalies and exhibit a strong tendency to occur in families. When a murmur is present, it is also located over the lung fields. Pulmonary arteriovenous fistulas are usually identifiable by conventional chest roentgenography and are obvious by angiocardiography. 47 Arteriovenous fistulas of the chest wall, muscles, and internal mammary vessels 48, 49 also may enter into differential diagnoses.

On the basis of this collected series of cases, it appears that murmurs due to coronary arteriovenous fistulas may be heard at various locations depending upon the site of the fistulous communication. In our case, since the fistula was close to the apex of the heart, the resultant murmur was similarly located, appeared to take origin close to the stethoscope and was intensified during expiration.

If a continuous murmur is judged to represent something other than a patent ductus arteriosus (and location may be the most important means of making the distinction), additional studies such as cardiac catheterization, angiocardiography, and arteriography provide means for making a nonsurgical diagnosis in the living patient.

Prognosis and Treatment. The prognosis in this lesion like other arteriovenous shunts depends upon the volume and duration of the shunt as well as the ability of the heart to handle the increased load. 44 The cases reviewed here demonstrate the wide range of functional disability that can result. Thus, the most useful generality referable to prognosis and therapy would be the suggestion that the findings exhibited by the patient are a better guide than any set policy, conservative or otherwise. It may be worthwhile to summarize the evidence bearing upon prognosis provided by this series.

By repeated use of "remarkably well tolerated" 112, 15 and other similarly optimistic comments, several previous authors have indicated their belief that coronary arteriovenous fistulas do not carry a poor prognosis. It should be noted, however, that with 1 exception, 23 personal observations have been limited to single cases. Furthermore, a critical review of published reports suggests that the optimism of some authors is unsupported by their data.

Knoblich and Rawson 12 reported the autopsy findings in an arteriovenous fistula of the heart that occurred in a 55-year-old business man who died of a subarachnoid hemorrhage. In ascribing the marked cardiac enlargement that was present (600 Gm.) to the patient's hypertension (220/110 mm. Hg), they made no mention of the frequent association of arteriovenous shunts and systemic hypertension. Noting marked serpentine dilatation of the left descending coronary artery and a sacular vein with which it communicated, the authors attributed this to "markedly reduced capillary
beds through which blood entering the terminal portion of the artery had to pass.'

This interpretation and the assumption that the patient's hypertension was independent of his arteriovenous fistula are both inconsistent with the observed hemodynamic behavior of arteriovenous fistulas including those in this series. Reduction in flow through vessels leads to the opposite of dilatation; the reverse occurs in arteriovenous fistulas. When they concluded that "arteriovenous fistulas of the heart do not appreciably shorten the life span," Knoblich and Rawson appear to have disregarded the possibility that their patient who died in 1955 at the age of 55 years would, but for his illness, have enjoyed a life expectancy of 19.3 additional years.\(^{61}\)

The age at death of some of the patients in this series might seem to support the contention that longevity is the rule in coronary arteriovenous fistula. This is simply not true. The mean age of death of 13 autopsied patients was 55 years, while the range was from 1 to 85 years. Four patients died without reaching their forty-fourth year of life. The average figure for life expectancy at the age at which the 13 autopsied patients in this series died is 21.6 years.* This figure could mislead the unwary, since it fails to take into account other causes of death. Inasmuch as it reflects actuarial factors such as the year in which death occurred and the age and sex of the patients, it is more meaningful than a figure expressing the average age at death. This short series demonstrates that longevity is possible with a coronary arteriovenous fistula, but provides little or no useful information about the life expectancy of patients with this lesion.

Convincing evidence exists to show that coronary arteriovenous malformations are indeed potentially serious lesions. Heart weights were stated in 9 of the 13 cases where autopsy was done. Without exception, the figures were above normal. Heart weights ranged from 380 to 1,800 Gm. with a mean of over 700 Gm.

In this series of 13 autopsied patients, death was directly attributed to the shunt only twice (cases 4 and 11). Although the authors did not discuss the cause, it is apparent that the death of case 13, a 25-year-old woman reported by Valdivia et al.\(^{13}\) was a result of her fistulous communication. Review of the autopsied cases indicates that death probably resulted from congestive heart failure in 6 of the reported patients (cases 3, 4, 9, 10, 11, and 13). In a total of 12 of the 22 patients comprising this series, there were definite cardiovascular symptoms. Although myocardial infarction (case 3) and hypertension (cases 4, 10, and 12) may have been independent contributory factors, it is equally possible that both may have resulted from or added to the disability due to the fistulous communication. The tendency for gangrenous changes to occur in extremities affected by peripheral arteriovenous fistulas is well known and suggests that coronary arteriovenous communications may predispose to or aggravate myocardial ischemia and infarction. Similarly, it is apparent that when a sufficient shunt volume exists, there will necessarily be an increase in the systemic, systolic, and pulse pressures. For example, although the lethal heart failure in case 4 probably was due to hypertension (260/100 mm. Hg), it is also probable that the hypertension was due to or aggravated by the fistula. The pulse pressure of 160 mm. Hg offers support to this interpretation as do many reports of reversible but serious hypertension associated with peripheral arteriovenous lesions.

In 4 of the 22 cases comprising this series, cardiac catheterization allowed the calculation of the volume of shunted blood. This ranged from 2.0 to 3.9 L. per minute, with the average shunt of the 4 patients accounting for slightly more than 40 per cent of the average cardiac output. Although shunts of
CORONARY ARTERIOVENOUS FISTULA

this magnitude may be well tolerated in many individuals, they necessarily represent a threat to the patient's well being and longevity.

Certain lesions would be difficult or impossible to correct by presently available surgical methods. On the other hand, in all of the 4 instances wherein an attempt was made to ligate the involved coronary artery or branch, the patients appear to have been cured, as evidenced by the disappearance of the continuous murmurs in all cases, and symptoms in the 2 cases where they existed preoperatively. Ligation was not deemed advisable in 3 of the 7 patients subjected to thoracotomy on the basis of the anatomic conditions encountered. Furthermore, the 3 patients who were explored without ligation appear to have survived none the worse for the procedure. Thus, it appears that surgery may offer much to patients suffering with this lesion. The possibility of successful surgical management will be dependent upon the type of communication present in a given case. Where a single anomalous vessel is present, the surgeon should attempt to demonstrate that occlusion of this channel is safe and eliminates the thrill. Final closure should probably not be carried out until it is demonstrated that clinical or electrocardiographic evidence of impaired myocardial blood supply does not follow temporary occlusion of the shunt-bearing branch. In complex, cirsoid communications surgical obliteration of the anomalous channels might prove to be hazardous by virtue of technical difficulties and because the multiple ligations requisite for closure might produce regional myocardial ischemia. It is in such cases that the lesion is most apt to cause serious functional consequences, however, and again the decision must be based upon the evidence provided by the case in question.

In our case, because of the asymptomatic course and absence of cardiac enlargement, as well as the normal and unchanging electrocardiographic findings during 7 years of observation, surgical exploration has not to date been considered necessary. Insofar as there has been no evidence of progression of the disease, the time gained has already brought the patient into a new era of cardiac surgery—the surgery of the open heart. In this respect, her chances for cure have been materially improved. Since, as in other types of arteriovenous fistulas, bacterial endocarditis represents a potential complication, antibiotic prophylaxis is used for dental extractions and other minor surgical procedures. Medical observation will be continued with emphasis placed upon functional testing. Should there be evidence of progression of the disease, surgery will be seriously considered.

SUMMARY

Twenty-one cases of coronary arteriovenous communications have been collected from the literature and a single case has been added. Coronary arteriovenous communications appear to result from 1 or a combination of 2 types of mechanisms causing defective fetal development.

Type 1. Defective Differentiation of Coronary Capillary Elements. Primitive sinusoidal elements of the coronary system may persist or undergo defective development resulting in communications between the coronary arteries and the coronary veins or the chambers of the heart. If such communications present a sufficiently low resistance to coronary flow, an arteriovenous shunt is the result.

Type 2. Anomalous Coronary Artery Origin with Intercoronary Communication. When, through defective formation of the primitive bulbar septum, a coronary artery takes its origin from the pulmonary artery or the right ventricle, intercoronary anastomoses sometimes permit blood to flow from the high pressure systemic coronary circuit into the low pressure pulmonary circuit. This mechanism, though a common cause of coronary arteriovenous fistulas, is a less frequent manifestation of anomalous coronary artery origin, since inter-coronary vascular resistance usually prevents significant
shunting. It is apparent that defective capillary differentiation and anomalous coronary artery origin are often jointly responsible for coronary arteriovenous fistulas.

Perhaps as many as a third of previously reported coronary artery aneurysms have resulted from congenital coronary artery fistulas, a fact sometimes overlooked.

The diagnosis of coronary arteriovenous fistula can be made with certainty and precision during life. It may be suspected on the basis of a continuous murmur resembling that of patent ductus arteriosus but atypical in location. Catheterization of the heart may be diagnostic and can be expected to reveal the presence and volume of a left-to-right shunt. It also might prove to confuse the diagnosis by suggesting a more common cause of such a shunt. Angiocardiography is capable of diagnostic findings. Coronary arteriography, though yet to be used in coronary arteriovenous fistula, probably offers the best means of obtaining a precise anatomic diagnosis during life.*

The prognosis in this lesion varies widely in keeping with the varying anatomic and functional abnormalities that may exist. In some instances there has been long life with apparent freedom from symptoms; in others, heart failure due to the demand for increased cardiac output or associated hypertensive disease has caused death of patients with this lesion.

Surgical ligation of the artery or arteries carrying blood to the fistula should be attempted if there is evidence of progressive disease. In a given case, surgery may be curative or it may be impossible, depending upon the pathologic anatomy that is present.

ADDENDUM

R. M. Cooley and R. B. Sloan (Radiology of the Heart and Great Blood Vessels, Baltimore, Williams & Wilkins, 1956, p. 226, fig. 141) have also illustrated an asymptomatic child with a coronary arteriovenous fistula. Diagnosis was suspected after clinical studies and confirmed by angiocardiography.

*This has recently been achieved by Dr. B. A. Gasul and associates and briefly described by Dr. E. A. Fell.**

STEINBERG, BALDWIN, AND DOTTER

SUMMARIO IN INTERLINGUA

Vinti-un casos de communication inter arteria e vena coronari esseva colligite ab le litteratura. Un vinti-secunde caso es addite. Communicationes inter arteria e vena coronari pare resultar ab 1 o un combination de ambes de 2 typos de mechanismo effectuante un defective disveloppamento fetal.

Typo 1. Differentiation Defective del Elementos Capillari Coronari.—Primitive elementos sinusoide in le systema coronari pote perverso o sequer un disveloppamento defective que resulta in communicationes inter le arterias coronari e le venas coronari o le cameras del corde. In casos in que tal communicationes presenta un sufficientemente basse resistentia al fluxo de sanguine coronari, le resultato es un derivation arterio-venose.

Typo 2. Origine Anormal del Arteria Coronari con Communication Intercoronari.—Quando, in consequentia del formation defective del primitive septo bulbar, un arteria coronari prende su origine ab le arteria pulmonar o ab le ventriculo dextere, anastomoses intercoronari permette a vices un fluxo de sanguine ab le systemic circuito coronari con su alte pression verso le circuito pulmonar con su basse pression. Iste mechanismo es un causa communs de fistulas inter arteria e vena coronari, sed illo es minus frequentemente un manifestatation del origine anormal del arteria coronari, proque le resistentia vascular intercoronari preveni usualmente grados significante de derivation. Il es apparen-te que le defective differentiation capillari e le origine anormal del arteria coronari es frequentemente coresponsable pro le formation de fistulas inter arteria e vena coronari.

Il pare que multes e possiblemente non minus que un tertiio del previemente reportate aneurysmas de arteria coronari esseva le resultato de congenite fistulas de arteria coronari. Iste facto es non infrequentemente negligite.

Le diagnose de fistula inter arteria e vena coronari pote esser estabite con certitude e con precision durante le vita del paciente. Ilo pote esser suspicione super le base de un murmure continue que resimila le mur-
mure de patente ducto arteriose sed differre ab isto in su location. Catheterisation del corde pote stabilir le diagnose. Il es a expectar que illo revela le presentia e le volumine de un derivation sinistro-dextere. Il es etiam possibile que catheterisation confunde le diagnose per suggerer un causa plus commun del derivation. Le angioangiographia es capace a contribuer datos de valor diagnost. Arteriographia coronari — ben que non ancora usate in casos de fistula inter arteria e vena coronari—representa probablemente le melior metodo pro obtener un precise diagnose anatomic durante le vita del patiente.

Le prognose in casos de iste lesion varia extensemente, de acordo con le varie anormalitates anatomic e functional que pote esser presente. In certe casos, longe periodo de supervventia con apparente absente de symptomas esseva notate. In alteres, insufficientia cardiæ—causate per le requirimento de un augmentate rendimento cardiæ o per le presentia associate de morbo hypertensive—ha causate le morbo de patientes con iste lesion.

Le ligation chirurgic del arteria o del arterias que porta sanguine verso le fistula debe esser essayate in casos de evident morbo progressive. In le caso individual, intervention chirurgic pote esser curative o illo pote esser impossibile, in dependencia del anatomic pathologic que es presente.

REFERENCES

17. Gross, R. E.: Cited by Paul et al.3
18. —: Personal communication.
20. —: Personal communication.
25. Rigdon, R. H., and Vandergriff, H.: Aneu-


STEINBERG, BALDWIN, AND DOTTER


42. BROOKS, H. S.: Two cases of an abnormal coronary artery of the heart arising from the pulmonary artery: With some remarks upon the effect of this anomaly in producing cirrhoid dilatation of the vessels. J. Anat. & Physiol. 26: 26, 1885.


Coronary Arteriovenous Fistula
ISRAEL STEINBERG, JANET S. BALDWIN and CHARLES T. DOTTER

Circulation. 1958;17:372-390
doi: 10.1161/01.CIR.17.3.372
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1958 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/17/3/372

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation is online at:
http://circ.ahajournals.org/subscriptions/