Clinical, Angiographic, and Hemodynamic Findings in Patients with Anomalous Origin of the Coronary Arteries

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SUMMARY Clinical and angiographic features of 31 patients with anomalous coronary artery origin are reviewed. Of 17 aberrant circumflex arteries from the right sinus of Valsalva or artery, each was retro-aortic, six atherosclerotic, and seven irrigated a small area of myocardium. Of seven anomalous right coronary arteries from the left sinus of Valsalva or artery, each was ante-aortic and two atherosclerotic. Aberrant origin of the circumflex or right coronary artery was a benign anomaly.

The proximal course of seven aberrant left coronary arteries from the right sinus of Valsalva or right coronary artery was related to clinical events. The anomaly was best demonstrated in the lateral view. In five cases, coronary blood flow during exercise and myocardial metabolism during pacing were assessed. Two had had a previous infarction with nonatherosclerotic arteries. Exercise coronary blood flow was normal in four and low in one patient. Pacing resulted in lactate production in two with atherosclerotic and one without atherosclerotic arteries. We conclude that anomalous left coronary origin from the right sinus of Valsalva can result in significant myocardial ischemia and infarction.

THE WIDESPREAD APPLICATION of coronary arteriography has resulted in more frequent detection of patients with anomalous coronary artery origin. Clinical interest in these rare anomalies has increased since sudden death has been reported to be a complication of aberrant left coronary origin from the right coronary artery or right sinus of Valsalva when the proximal course of this vessel is between the aorta and pulmonary artery. The predisposing factors which lead to sudden death in some patients with

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this anomaly are unknown. Coronary vascular hemodynamics have not been studied in a systematic manner because the diagnosis of this anomaly is rarely made ante-mortem. We have reviewed the clinical and angiographic features of 31 patients with anomalous coronary artery origin and have assessed the coronary vascular responsiveness to pacing-induced tachycardia and exercise in a subgroup of five patients with aberrant left coronary origin from the right coronary artery or right sinus of Valsalva.

Material and Methods

A review of 3,750 patients having coronary arteriography performed at the Montreal Heart Institute between January 1, 1972, and May 1, 1975, revealed 31 adults with anomalous coronary artery origin. Selective coronary arteriograms were performed by a percutaneous transfemoral ap-
proach which used preformed polyethylene catheters.7 Cine coronary arteriograms were filmed with a 35 mm Mitchell camera at 60 frames/sec utilizing a 6 inch Picker image intensifier. Seventeen of the 31 adults were men and 14 women; their mean age was 50 (range 25–70) years. Clinical characteristics of each patient were recorded at the time of arteriography. Seventeen patients had coronary arteriography done for typical angina pectoris or for a previous history of myocardial infarction at a young age. Two patients were studied for atypical angina pectoris and three for nonspecific chest pains. Eight patients underwent coronary arteriography during catheterization for valvular heart disease and one patient during catheterization for a cardiomyopathy. The coronary arteriograms were reviewed by two experienced angiographers, who reached agreement on the diagnosis of anomalous origin and the course of the coronary arteries.

Protocol

Seven patients had aberrant left coronary origin from the right sinus of Valsalva. Coronary artery distribution, pertinent medical history, and angiographic features are summarized in table 1. Left ventricular wall motion was assessed by visual inspection of the left ventriculogram and ejection fraction was calculated by previously described methods. A previous myocardial infarction was in all cases documented by clinical history, serial electrocardiograms, and sequential changes in cardiac enzymes. Five patients in this subgroup were reevaluated an average of 15 months (range 3–26 months) postarteriography. The project was explained to each of the five patients prior to hospitalization and informed consent obtained prior to the procedure.

Graded Exercise Test

One day prior to catheterization a maximal treadmill test utilizing 12-leads was performed following a Bruce test modified by a 3 minute warm-up at 1.7 mph and 5% grade. Standard criteria were used for stopping the test. An ischemic response was registered when there was flat or downsloping ST-segment depression greater than one mm below the corresponding P-R segment measured 0.08 seconds after the J point.

Catheterization

The patient was brought to the catheterization laboratory the following morning in the fasting state. A 100 cm size 8F Ganz thermodilution catheter was inserted from the right antecubital vein into the main coronary sinus. The external thermistor or distal pacing electrode of the Ganz catheter was positioned just inside the coronary sinus ostium and was maintained in that position throughout the study. Proper placement of the catheter was established by injection of 3 cc of Renografin 76% into the sinus with fluoroscopic identification of the ostium. The stability of the catheter position during the procedure was checked by periodic fluoroscopy. An 18 gauge polyethylene catheter was inserted percutaneously into the right radial artery. Both catheters were connected to Statham P23Db transducers and were adjusted so that zero pressure was at the midchest position. The electrocardiogram was continuously monitored.

Table 1. Clinical and Angiographic Features of Patients with Anomalous Left Coronary Origin from the Right Sinus of Valsalva or Right Coronary Artery

<table>
<thead>
<tr>
<th>Patient/Age/Sex</th>
<th>Description of coronary anatomy</th>
<th>History of MI</th>
<th>Class*</th>
<th>LVEDP* (mm Hg)</th>
<th>EF</th>
<th>LV wall motion</th>
</tr>
</thead>
<tbody>
<tr>
<td>JB: 25/F</td>
<td>LAD anterior to PA, Cx posterior to Ao, separate diagonal branch from left aortic sinus; non-ath. (fig. 5)</td>
<td>antero-septal</td>
<td>asymptomatic</td>
<td>12</td>
<td>0.59</td>
<td>Mild antero-apical hypokinesis</td>
</tr>
<tr>
<td>EB: 43/M</td>
<td>Left main posterior to Ao, non-ath.</td>
<td>anterior</td>
<td>I</td>
<td>12</td>
<td>0.60</td>
<td>normal</td>
</tr>
<tr>
<td>RB: 45/F</td>
<td>Left main posterior to Ao, non-ath. (fig. 2)</td>
<td>none</td>
<td>nontransmural</td>
<td>5</td>
<td>—</td>
<td>normal</td>
</tr>
<tr>
<td>LB†: 49/F</td>
<td>Left main between PA and Ao, non-ath.</td>
<td>none</td>
<td>asymptomatic</td>
<td>12</td>
<td>—</td>
<td>normal</td>
</tr>
<tr>
<td>JL: 39/M</td>
<td>Left main between PA and Ao. Complete occlusion of proximal LAD, middle Cx and middle RCA</td>
<td>inferior</td>
<td>III</td>
<td>10</td>
<td>0.35</td>
<td>akinesia, anterolateral wall; severe hypokinesis inferoapical wall</td>
</tr>
<tr>
<td>ML: 60/M</td>
<td>LAD between PA and Ao, Cx posterior to Ao. 90% St Cx; 75% St distal RCA (fig. 4)</td>
<td>none</td>
<td>III</td>
<td>12</td>
<td>—</td>
<td>normal</td>
</tr>
<tr>
<td>KW: 70/F</td>
<td>Left main between PA and Ao. 90% St RCA; 60% St Cx (fig. 3)</td>
<td>none</td>
<td>III</td>
<td>20</td>
<td>0.54</td>
<td>normal</td>
</tr>
</tbody>
</table>

*Functional Classification Angina of the Canadian Cardiovascular Society.19
†Coxistent mitral stenosis.

Abbreviations: LVEDP = left ventricular end-diastolic pressure; LAD = left anterior descending; MI = myocardial infarction; Cx = circumflex; ECG = electrocardiogram; RCA = right coronary artery; RBBB = right bundle branch block; Ao = aorta; Abn. = abnormality; PA = pulmonary artery; Non-ath. = non-atherosclerotic; St = stenosis; EF = ejection fraction.
The patient was allowed to stabilize and resting measurements were taken. Each set of measurements consisted of simultaneous blood samples from the radial artery and coronary sinus which were analyzed for pH, PO_2, and lactate by previously described techniques and amplified pressures recorded at paper speed of 25 and 100 mm per second on a multichannel Sanborn photographic recorder. The patient was paced from the coronary sinus at 120 and 150 beats/min for 6 min at each level with the above measurements made during the last three minutes at each level. Pacing was terminated and five to ten minutes were allowed to elapse while blood pressure and heart rate returned to prepacing levels. A second set of control measurements was acquired prior to exercise and expired gas was collected for determination of oxygen consumption. Coronary sinus blood flow measurements were made and resting cardiac output determined by injection of 5 mg of indocyanin green into the coronary sinus with collection from the radial artery. The patient performed supine exercise for 7 min utilizing a bicycle ergometer with 60 watts resistance. Pressure measurements, simultaneous blood samples for oximetry, and coronary sinus blood flow measurements were made during the last three minutes of exercise. Expired gas was collected during exercise for determination of oxygen consumption and exercise cardiac output was performed following recording of coronary sinus blood flow.

Calculations
Peak systolic blood pressure was averaged over 10 beats and thermodilution coronary blood flow determinations were calculated by previously described formulae utilizing a Hewlett-Packard 9830A calculator with digitizer. Cardiac output was calculated according to the Williams formula.

Results
The incidence of patients with anomalous coronary artery origin was 31/3750 (0.83%) in our angiographic population. These 31 patients were subdivided into three groups.

I. Anomalous Left Coronary Origin from the Right Sinus of Valsalva or Right Coronary Artery (seven patients)

ANGIOGRAPHIC FEATURES (FIG. 1)
The left main or left anterior descending coronary artery passed posterior to the aortic root in two patients, between the aorta and pulmonary artery in four patients, and anterior to the pulmonary artery in one patient. The distal distribution of the coronary arteries was normal in all cases.

Retro-aortic Course of the Left Main Coronary Artery (two patients). The left main coronary arose from the proximal part of the right coronary artery in both patients. Its proximal course was best seen in the lateral view (fig. 2).

Left Main or Left Anterior Descending Coronary Artery Between the Aorta and Pulmonary Artery (four patients). There were three patients in whom the left main coronary artery coursed between the great vessels. Patient KW typified the angiographic features of this group (fig. 3). In each patient the first septal branch came off the left main trunk before its division into the left anterior descending and circumflex arteries. The fourth patient had a left anterior descending coronary artery which coursed between the great vessels and an aberrant circumflex artery which followed a retroaortic course (fig. 4).

Left Anterior Descending Coronary Artery Anterior to the Pulmonary Artery (one patient). The angiographic features of this anomaly are illustrated in figure 5.

CLINICAL AND HEMODYNAMIC FEATURES
Of seven patients with anomalous left coronary origin (table 1) three had significant arterial stenoses and were evaluated for typical angina pectoris. Four patients had nonatherosclerotic arteries; two were evaluated for a myocardial infarction which occurred at a young age, one for paroxysmal atrial tachycardia and nonspecific chest pains, and one patient during preoperative evaluation for valvular heart surgery. Two of the seven patients were not restudied because 1) KW (fig. 3) had received bypass grafts to the right coronary artery (130 ml/min intraoperative flow; 2 mm diameter) and the left circumflex artery (70 ml/min intraoperative flow; 1.5 mm diameter); and 2) LB had severe mitral stenosis and systemic pulmonary hypertension.

Of the five patients restudied, both with coronary atherosclerosis (JL, ML) experienced angina and produced

![Diagram](http://circ.ahajournals.org/)

**Figure 1** Diagrammatic representation of anomalous left coronary origin from the right sinus of Valsalva or right coronary artery (RCA). A) Retro-aortic course of the left main coronary artery (LMCA). B) LMCA between the aorta (Ao) and pulmonary artery (PA). C) Left anterior descending (LAD) between Ao and PA. Retro-aortic course of the circumflex (Circ). D) LAD anterior to PA. Circ. retro-aortic. Diagonal branch (Diag.) from the left coronary sinus.
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Figure 2  Patient RB; lateral view. The RCA arises normally from the right coronary sinus (RCS). The LMCA is a branch of the proximal RCA and passes posterior to the aortic root in a large concave arc prior to its bifurcation into the LAD and circumflex (Cx) arteries.

Figure 3  A) Patient KW; lateral view. The left main coronary artery (LMCA) arises from the right coronary sinus (RCS) anterior and adjacent to the conus branch (CB). It courses directly laterally and is seen end-on giving off the first septal branch (S1) before it branches into the left anterior descending (LAD) and proximal circumflex (Prox Cx) arteries. B) The left anterior oblique position shows the LMCA between the Ao and PA giving off S1 at its first branch. A 60% stenosis (arrow) is seen at the origin of the Cx beyond the diagonal branch (Diag.). C) Right anterior oblique position; semi-selective injection into the RCA shows the origin of both the RCA and LMCA from the RCS. There is a 90% stenosis in the mid-RCA (arrow).
lactate during pacing at 150 beats/min (fig. 6). During exercise-induced angina, both increased coronary blood flow normally* and myocardial oxygen extraction remained relatively constant (fig. 7). Hemodynamic findings were normal in a third patient (RB) without atherosclerosis. JB, a 25-year-old woman, had an anteroseptal myocardial infarction complicated by right bundle branch block and complete heart block requiring temporary pacing. Coronary arteriography done three months postinfarction is shown in figure 5. Myocardial lactate extraction was normal during pacing (fig. 6) and coronary blood flow increased 76% over resting values during exercise (fig. 7). EB, a 43-year-old man, had a nontransmural anterior myocardial infarction complicated by hypotension and pulmonary edema. His coronary anatomy was similar to that illustrated in figure 2. Myocardial lactate production occurred during pacing (fig. 6) and myocardial oxygen extraction increased 33% accompanied by a decrease of 39% in coronary blood flow during exercise (fig. 7). This patient did not have angina or ischemic

*In six normal patients evaluated in our laboratory utilizing the above protocol thermodilution coronary sinus blood flow increased 105 ± 72% (mean ± 1SD) during supine exercise.

**Figure 4** Patient ML; lateral view. Semi-selective injection in the right coronary sinus (RCS) shows the proximal part of the left anterior descending artery (LAD) coursing directly anterior between the Ao and PA. The circumflex artery (Cx) arises from the RCA and courses retro-aortically. There is a 50% stenosis in the mid-Cx (white arrow).

**Figure 5** A) Patient JB; lateral view. The LAD arises from the proximal part of the RCA and courses superior, anterior and to the right of the pulmonary trunk to arrive in the anterior interventricular sulcus where it gives off septal branches (S). The RCA originates normally in the right coronary sinus (RCS) and gives off the Cx which courses retroaortic. B) The right anterior oblique view shows the LAD coursing superior and anterior to the pulmonary artery.

**Table 2.** Maximal Treadmill Data of Patients with Anomalous Left Coronary Origin from the Right Sinus of Valsalva or Right Coronary Artery

<table>
<thead>
<tr>
<th>Patient</th>
<th>HR (beats/min)</th>
<th>BP (mm Hg)</th>
<th>HR × BP (10^-3)</th>
<th>HR (beats/min)</th>
<th>BP (mm Hg)</th>
<th>HR × BP (10^-3)</th>
<th>Result</th>
<th>Reason for stopping</th>
</tr>
</thead>
<tbody>
<tr>
<td>JB</td>
<td>83</td>
<td>130/75</td>
<td>107.9</td>
<td>180</td>
<td>204/120</td>
<td>367.2</td>
<td>Incomplete RBBB became complete RBBB</td>
<td>fatigue</td>
</tr>
<tr>
<td>EB</td>
<td>68</td>
<td>142/80</td>
<td>96.6</td>
<td>176</td>
<td>212/90</td>
<td>373.2</td>
<td>neg.</td>
<td>fatigue</td>
</tr>
<tr>
<td>RB</td>
<td>131</td>
<td>164/100</td>
<td>214.8</td>
<td>182</td>
<td>174/90</td>
<td>310.7</td>
<td>neg.</td>
<td>fatigue</td>
</tr>
<tr>
<td>JL</td>
<td>87</td>
<td>132/90</td>
<td>114.8</td>
<td>112</td>
<td>170/90</td>
<td>190.4</td>
<td>pos</td>
<td>angina</td>
</tr>
<tr>
<td>ML</td>
<td>79</td>
<td>172/100</td>
<td>135.9</td>
<td>152</td>
<td>192/130</td>
<td>291.8</td>
<td>I, aVL, V₅₋₆ pos</td>
<td>angina</td>
</tr>
</tbody>
</table>

Abbreviations: HR = heart rate; BP = blood pressure; neg = negative; pos = positive; RBBB = right bundle branch block.
electrocardiographic changes recorded during pacing and exercise.

Each of the five patients had a normal increase in cardiac index with exercise (fig. 8). Treadmill exercise tests were negative for ischemia in each of the patients without atherosclerosis (JB, RB, EB) but positive for ischemia in both patients with significant arterial stenoses (JL, ML). There were no arrhythmias recorded during exercise and no complications (table 2).

II. Aberrant Circumflex Origin from the Right Coronary Artery or Right Sinus of Valsalva (17 patients)

ANGIOGRAPHIC FEATURES (FIG. 9A)

The typical angiographic findings of this anomaly are illustrated in figure 10. The aberrant circumflex arose from the right coronary artery or right sinus of Valsalva and coursed retroaortic in all cases. In four cases circumflex arteries had significant atherosclerosis. The distal distribution of the aberrant circumflex perfused a small segment of the heart in seven patients (fig. 10), a large segment in seven patients (fig. 11); and segments intermediate between the two extremes in three patients.

CLINICAL FEATURES

Nine patients had nonatherosclerotic coronary arteries; five were studied during catheterization for valvular heart disease, one for atypical angina pectoris, two for nonspecific chest pains, and one patient for a cardiomyopathy. The remaining eight patients had significant atherosclerosis and typical angina pectoris. Ten of the 17 patients had associated cardiac abnormalities. Three patients had mixed mitral and aortic valve disease, three had aortic valve disease, two had mitral valve disease, one had idiopathic hypertrophic sub-

![Figure 6](http://circ.ahajournals.org/)

Effect of exercise on hemodynamic parameters in patients with anomalous left coronary artery from the right coronary artery or right sinus of Valsalva. Left panel) Each patient increases their pressure-rate product normally with exercise. Central panel) Coronary blood flow increases normally with exercise in four patients including JL and ML (increases of 112% and 239% respectively) who had significant coronary atherosclerosis. EB decreases coronary blood flow 39% with exercise. Right panel) Myocardial oxygen extraction remains relatively constant with exercise in RB, JL, ML. However, EB significantly widens his A-V $O_2$ difference with exercise. JB is not shown.
aortic stenosis, and one patient had a congestive cardiomyopathy.

III. Aberrant Right Coronary Origin from the Left Coronary Sinus (seven patients)

ANGIOGRAPHIC FEATURES (FIGS. 9B, 9C)

In six patients, the abnormal origin of the right coronary was in the left sinus of Valsalva in front of the left coronary artery ostium (fig. 12). In the seventh patient the right coronary took origin from a terminal branch of a large circumflex artery (fig. 13). The aberrant right coronary artery was free of atherosclerosis in five patients.

![Diagram of coronary artery origins](http://circ.ahajournals.org/)

**Figure 9** A) Aberrant circumflex origin from the RCA or right sinus of Valsalva. B) Aberrant RCA origin from the left coronary sinus. C) Aberrant RCA from the distal Circ.

CLINICAL FEATURES

Three patients had nonatherosclerotic coronary arteries; two were studied during catheterization for valvular heart disease and the third for atypical angina pectoris. The remaining four patients had significant arterial stenoses and were studied for typical angina pectoris. Of three patients with associated cardiac abnormalities, there was one ventricular septal defect, one bicuspid aortic valve, and one patient with mixed mitral and aortic valve disease.

Discussion

Pathologic reviews have classified several different anatomical variations that can occur in patients with anomalous aortic origin of the coronary arteries. There are few angiographic descriptions of these anomalies. Most were thought to be benign until sudden death was reported as a complication of aberrant left coronary origin from the right sinus of Valsalva when this vessel coursed between the aorta and pulmonary artery. The mechanism of sudden death is unknown and coronary vascular hemodynamics have not been systematically evaluated in patients with aberrant left coronary origin from the right sinus of Valsalva. We have reviewed the angiographic and clinical features of 31 patients with anomalous aortic coronary artery origin and have assessed coronary vascular hemodynamics in five patients with aberrant left coronary origin from the right sinus of Valsalva.

The total incidence of patients with anomalous aortic coronary artery origin was 0.83% of our angiographic population. The most frequent anomaly was aberrant circumflex origin from the right coronary artery or right sinus of Valsalva, which occurred in 0.45% of cases. Angiographic features characteristic of this anomaly include 1) detection of an avascular area in the segment of heart normally perfused by the circumflex artery after selective injection of the left coronary and 2) direct visualization of the circumflex after injection into the right coronary or selective injection into the ostium of the left circumflex artery in the right sinus of Valsalva. A third angiographic feature described by Page et al. is the "aortic root sign" in which a profile view of the anomalous circumflex is seen as it courses posteriorly behind the right sinus of Valsalva during left ventriculography in the right anterior oblique position. The quantity of left ven-
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Figure 10  A) lateral view. The LAD arises in the left coronary sinus and gives off diagonal arteries (Diag.). There is a relatively avascular area in the segment of ventricle normally perfused by the Cx. B) lateral view. The Cx comes off the RCA to supply the avascular area seen in the lateral view. The distal run-off of this aberrant Cx perfuses a small segment of the left ventricle.

The lateral view. The LA D arises in the left coronary sinus and gives off diagonal arteries (Diag.). There is a relatively avascular area in the segment of ventricle normally perfused by the Cx. The distal run-off of this aberrant Cx perfuses a small segment of the left ventricle.

tricular myocardium perfused by an aberrant circumflex can vary. In 41% of our patients and in 40% of the patients described by Page et al., the aberrant circumflex artery covered a small territory of the left ventricle. Aortocoronary bypass graft distal to a proximal stenosis in a small aberrant circumflex artery would be technically difficult. A large distal distribution of a diseased aberrant circumplex can make it a suitable recipient for a bypass graft, e.g., intraoperative graft flow was 70 ml/min and intraluminal diameter 1.5 mm in one patient.

We found a high incidence (47%) of coexistent valvular lesions associated with aberrant circumflex origin. A slightly lower incidence has been reported by others. The average age of patients with aberrant circumflex origin in our population was 51 years: we routinely perform coronary arteriography on patients older than 40 years undergoing catheterization for valvular heart disease. This could increase the number of patients falling into this group although a congenital association between aberrant circumflex origin and valvular lesions is suspected.

Aberrant right coronary origin from the left sinus of Valsalva is a less frequent coronary anomaly occurring in 0.19% of our angiographic population. During arteriography, this anomaly should be suspected when the right coronary artery is not visualized after contrast injections into the right sinus of Valsalva. The aberrant vessel can be demonstrated by selective injections into the aberrant ostium which invariably is in front of the ostium of the left coronary artery. The distal distribution of the right coronary artery is the same as in the general population. Aortocoronary bypass graft distal to a proximal stenosis in an aberrant right coronary can provide satisfactory flow and one of our patients had a graft distal to a 50% stenosis with an intraoperative flow of 70 ml/min.

Figure 11  Frontal view. Hyperselective injection in the Prox Cx which arises from the right coronary sinus (RCS). A large segment of the left ventricle is perfused by the Dist Cx and its marginal branches (Marg.).

Figure 12  Frontal view. The RCA arises in the left coronary sinus (LCS) in front of the LMCA and passes anterior, superior and to the left of the aorta before occupying the right anterior atrioventricular groove.
We did not find an increased incidence of atherosclerotic plaques resulting from the aberrant origin and course of the circumflex or right coronary arteries, a finding similar to those of others. In addition, the presence of these anomalies in the absence of atherosclerosis did not result in arrhythmias, typical angina pectoris, or myocardial infarction.

Anomalous left coronary origin from the right sinus of Valsalva or right coronary artery was rare, forming 0.19% of our angiographic population. It is important to distinguish the course of the aberrant left coronary artery relative to the great vessels in this group of patients since sudden death has been reported in adolescents and young adults in whom this artery coursed between the aorta and pulmonary artery. We found the lateral position was best followed by the right anterior oblique position for delineating the proximal course of the aberrant opacified left coronary. Those patients with this anomaly who are predisposed to sudden death usually are uncovered in adolescence. Only two of the 16 patients with this anomaly reported in the literature who died suddenly were older than 22 years of age. One of the two patients, a 56-year-old man described by Benson, also had several 90% stenoses in a dominant right coronary artery. Although the author concluded that death was secondary to the aberrant left coronary passing between the aorta and pulmonary artery it is equally possible that death was the result of a lethal arrhythmia caused by ischemia in the territory of the stenotic right coronary artery. We have presented two patients (ML, KW) with this anomaly and atherosclerotic disease who are 60 and 70 years of age. Other investigators have also reported patients with this anomaly in the later decades of life.

Several hypotheses have been postulated to explain the mechanisms leading to sudden death. These include 1) compression of the aberrant artery between the aorta and pulmonary artery, 2) spasm, torsion, or kinking of the aberrant left coronary on itself, 3) an anatomical derange-

ment at the takeoff of the anomalous vessel, and 4) a congenitally small left coronary artery system. It is unlikely that the pulmonary artery with normal pressures could occlude or constrict the aberrant left coronary artery distended with systemic pressure. We have presented a 49-year-old woman with this anomaly (LB) who had systemic pulmonary hypertension. Although her aberrant left coronary coursed between the great vessels she did not have angina or evidence of myocardial ischemia. An intraluminal narrowing of the aberrant left coronary artery at its takeoff has been described in several of the adolescents who have died suddenly. We did not observe this feature in the four adults we presented with this anomaly. Two of these four patients (JL, ML) with class III angina severe atherosclerosis were able to augment coronary blood flow significantly with exercise-induced angina pectoris. The normal increase in coronary blood flow despite angina can be explained by heterogeneity of blood flow distribution with underperfusion in ischemic areas and augmented flow in nonischemic areas dilated by reactive hyperemia. Both patients had distal coronary anatomy unsuitable for bypass grafts. We would conclude that the isolated angiographic demonstration of this anomaly in an adult with mild cardiac symptoms is not an indication for prophylactic aorto-coronary bypass surgery.

Sudden death has not been reported in patients in whom the left coronary artery arises from the right coronary artery or right sinus of Valsalva and courses posterior to the aorta or anterior to the pulmonary artery. We have described a 25-year-old woman (JB) with nonatherosclerotic arteries who had an anteroseptal myocardial infarction complicated by complete heart block and in whom the aberrant left coronary passed anterior to the pulmonary artery. Lactate metabolism during pacing and exercise coronary blood flow were normal. Pachinger and coworkers have described a 43-year-old man with this type of anomaly who had severe angina pectoris in spite of nonatherosclerotic arteries and who had lactate production and a deterioration in left ventricular performance during pacing-induced angina. We have described a 43-year-old man (EB) with nonatherosclerotic arteries who had a myocardial infarction complicated by pulmonary edema in whom the aberrant left coronary coursed posterior to the aorta. Myocardial lactate production occurred during coronary sinus pacing and coronary blood flow diminished during exercise accompanied by a widening of the A-VO₂ difference. The reason why there was a decrease in total coronary flow during exercise and production of lactate during pacing in this patient and not in a second patient (RB) with the same anomaly and nonischemic equivalents is not clear.

We would conclude that certain patients with aberrant left coronary origin from the right coronary artery or right sinus of Valsalva can develop severe myocardial ischemia, infarction, and sudden death. It is difficult to know which patients with this anomaly diagnosed at angiography will have future acute ischemic episode(s). Clinical history, close angiographic study of the origin of the aberrant vessel accompanied by assessment of 1) myocardial lactate metabolism during pacing, 2) coronary sinus blood flow during exercise, and 3) response to maximal exercise testing may be useful in detecting those patients predisposed to future ischemic events.
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Clinical, angiographic, and hemodynamic findings in patients with anomalous origin of the coronary arteries,
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