Fusion of the Left Aortic Cusp to the Aortic Wall with Oclusion of the Left Coronary Ostium, and Aortic Stenosis and Insufficiency

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SUMMARY
The case of a 16-year-old girl with a unique anomaly of the coronary arterial tree is presented. This report concerns the first known case of isolated adherence of the left coronary aortic valve cusp to the aortic wall. This produced total obstruction of the left coronary artery ostium, angina pectoris, syncope, aortic stenosis, and aortic insufficiency. Freeing the adherent cusp from the aortic wall restored patency of the left coronary artery, relieved the aortic insufficiency, and resulted in disappearance of the angina pectoris and syncope. This case adds to the growing spectrum of nonatherosclerotic coronary abnormalities which are capable of producing myocardial ischemia.

Additional Indexing Words:
Angina pectoris Myocardial ischemia Syncope

Lesions of the coronary circulation capable of provoking myocardial ischemia assume immense significance in view of their frequent occurrence and high incidence of catastrophic consequences.1-3 Coronary atherosclerosis is the basis of this process in the vast majority of instances.1-3 Congenital and acquired nonatherosclerotic lesions of the coronary arterial tree capable of producing myocardial ischemia are of particular interest because of their rarity, their ability to result in obscure forms of heart disease, their unusual presentations and manifestations, and ultimately because of the ease with which remedial surgery can frequently be performed, provided the nature of these disorders is understood.4,5 This report concerns the first known case of isolated fusion of the left coronary aortic valve cusp to the aortic wall. This produced complete obstruction of the left coronary artery ostium, angina pectoris, syncope, aortic stenosis, and aortic insufficiency. The most gratifying and important feature of this case was its correction by a relatively simple surgical procedure.

Report of Case
C. C. (DUMC H4 4181), a 16-year-old girl, was referred to Duke University Medical Center for evaluation of syncope and chest pain. Since the age of 6 years, she had a total of four episodes of syncope during or immediately following vigorous exertion. She also had experienced infrequent but typical angina pectoris with heavy exercise since that age. No history could be obtained of chest trauma, syphilis, arthritis, rheumatic fever, or endocarditis.

Physical examination revealed a healthy appearing, 16-year-old girl. The pulse rate was
80/min and regular; blood pressure 130/60 mm Hg in both upper extremities; and the respiratory rate was 16/min. There were no abnormal venous waves or peripheral signs of aortic insufficiency outside of a modestly widened pulse pressure. The point of maximal cardiac impulse was forceful, suggestive of left ventricular hypertrophy, and located 1 cm lateral to the midclavicular line in the fifth intercostal space. The heart sounds were normal, and there were no clicks or gallops. A grade III/VI systolic ejection murmur was heard maximally over the second intercostal space to the right of the sternal border, and a grade IV/VI high-pitched, blowing decrescendo diastolic murmur of aortic insufficiency was heard maximally along the left sternal border. Findings on the remainder of the physical examination were normal.

Laboratory data revealed a hemoglobin of 12.8 g/100 ml with normal leucocyte and differential counts. Urinalysis and serum electrolytes including calcium and phosphorus were normal. The VDRL, Reiter protein, latex fixation, and lupus erythematosus studies were negative. The electrocardiogram (fig. 1) revealed normal sinus rhythm. Slight ST-segment elevation over the lateral precordial leads consistent with early repolarization was present. Treadmill exercise produced a rise in heart rate from 80 to 170/min, and at this time the patient experienced typical angina pectoris but did not develop distinct ST-T wave changes of myocardial ischemia. Four-view chest x-rays with barium swallow showed mild dilatation of the ascending portion of the thoracic aorta and slight prominence of the left ventricle. The lungs and bony thorax were normal (fig. 2).

The clinical, electrocardiographic, and radiologic findings suggested that the aortic insufficiency was of relatively minor degree. It was, therefore, difficult to account for the history of angina and syncope, and accordingly cardiac catheterization was undertaken.

**Cardiac Catheterization Findings**

Cardiac catheterization (table 1) revealed normal pressure measurements except for minimal elevation of the left ventricular end-diastolic pressure and a 20 mm Hg systolic gradient across the aortic valve. Cardiac index and systemic arteriovenous oxygen difference were normal. Indicator-dilution curves did not reveal evidence of an intracardiac shunt. An aortic root cineangiogram (fig. 3) demonstrated a dilated ascending aorta with moderately severe aortic regurgitation and delayed filling of the left coronary artery by collateral channels. Late in the injection a...
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Table 1

Summary of Cardiac Catheterization Data and Metabolic Studies

<table>
<thead>
<tr>
<th></th>
<th>Preoperative</th>
<th>Postoperative</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Catheterization studies</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Heart rate (beats/min)</td>
<td>90</td>
<td>80</td>
</tr>
<tr>
<td>Pressure (mm Hg)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mean right atrial</td>
<td>7</td>
<td>3</td>
</tr>
<tr>
<td>Right ventricular</td>
<td>30/6</td>
<td>27/3</td>
</tr>
<tr>
<td>Pulmonary artery</td>
<td>30/12 (16)</td>
<td>27/10 (14)</td>
</tr>
<tr>
<td>Left ventricular</td>
<td>130/12</td>
<td>120/5</td>
</tr>
<tr>
<td>Aortic root</td>
<td>110/70</td>
<td>100/68</td>
</tr>
<tr>
<td>Systemic A-V O$_2$ (vol %)</td>
<td>3.30</td>
<td>3.54</td>
</tr>
<tr>
<td>Cardiac index (L/min/m$^2$)</td>
<td>3.8</td>
<td>3.5</td>
</tr>
</tbody>
</table>

**Preoperative myocardial metabolic studies**

<table>
<thead>
<tr>
<th></th>
<th>Resting</th>
<th>Isoproterenol infusion (4 min 1µg/min)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Heart rate (beats/min)</td>
<td>90</td>
<td>132</td>
</tr>
<tr>
<td>Mean aortic pressure (mm Hg)</td>
<td>82</td>
<td>82</td>
</tr>
<tr>
<td>Tension-time index (mm Hg sec/min)</td>
<td>2107</td>
<td>2681</td>
</tr>
<tr>
<td>Myocardial A-V O$_2$ (vol %)</td>
<td>9.8</td>
<td>8.8</td>
</tr>
<tr>
<td>Myocardial A-V lactate (mm/L)</td>
<td>0.01</td>
<td>0.32</td>
</tr>
<tr>
<td>% Lactate extraction</td>
<td>8</td>
<td>19</td>
</tr>
<tr>
<td>ECG changes</td>
<td>Nil</td>
<td>Nil</td>
</tr>
<tr>
<td>Chest pain</td>
<td>Nil</td>
<td>Nil</td>
</tr>
</tbody>
</table>

membranous structure which ultimately proved to be an adherent left coronary cusp was seen covering the left coronary ostium. Using the Sones and Shirey$^6$ and the Judkins$^7$ technic only two aortic cusps could be selectively opacified. The left coronary cusp and left coronary artery could not be entered. Selective injection into the right coronary artery showed a larger than normal vessel with numerous large collateral channels arising from its proximal and distal portions to anastomose with and fill the entire left coronary system, in a retrograde fashion (fig. 4A and B). After filling the left anterior descending and circumflex coronary arteries, contrast material flowed retrograde through a normal main left coronary artery to opacify a blind sac. This sac represented the malplaced left coronary cusp of the aortic valve which adhered to the aortic wall, thereby totally covering the left coronary ostium. The blind sac remained opacified for several seconds after the coronary vessels had cleared since there was no communication between it and the aortic lumen (fig. 5). In addition this structure distended with each diastole and collapsed with each systole. This suggested that this leaflet of the aortic valve was adherent by its margin only and still flexible. Selective injection into the right coronary cusp and the noncoronary aortic cusp of the aortic valve revealed that each was competent, and contrast material was seen refluxing into the left ventricle through the space left by the adherent left coronary cusp. The cusp margins were moderately thickened. The aortic root was rotated counterclockwise so that the right coronary cusp and ostium were located anteriorly and to the left of the midline, the noncoronary cusp more rightward and anterior than normally, and the adherent left coronary cusp was completely posterior and slightly rightward (fig. 6).

Resting myocardial oxygen extraction (arterial and coronary sinus sampling) was normal, but myocardial lactate extraction was borderline abnormal (extration 8%). Isoproterenol infusion (1µg/min for 4 min) increased heart rate from 90 to 130 beats/min, with a rise in the calculated tension-time index from 2,107 to 2,681 mm Hg sec/min. This did not produce pain or electrocardiographic changes, and the myocardial lactate extraction increased from 8% to 19%, while the oxygen extraction decreased from 9.3 to 8.8 vol % (table 1).

**Surgical Procedures**

Although the magnitude of aortic insufficiency by itself did not appear sufficient to warrant surgical correction, the history of angina pectoris and syncope indicated that establishing communication between the left coronary artery and the aortic lumen might be beneficial. Since the angiographic findings suggested that the left coronary cusp was still intact, pliable, and

Circulation, Volume XLI, May 1970
adherent only by its margin, the operative correction was approached with the hope that freeing the adherent cusp would restore antegrade flow in the left coronary artery and simultaneously correct the aortic regurgitation.

Through a median sternotomy and employing total cardiopulmonary bypass and systemic hypothermia of 32C, the ascending aorta was exposed. Numerous large tortuous collaterals from the proximal main right coronary artery were seen coursing over the anterior right ventricular surface and anastomosing with branches of the left coronary artery. The anterior right ventricular wall in the region of pulmonary outflow tract was impressively thinned and white raising the possibility of a previous myocardial infarction or fibrosis of this region. No further evidence of myocardial infarction such as surface electrocardiographic mapping was sought. No paradoxical pulsations were noted. With a clamp on the distal ascending aorta, an incision was made in the proximal aorta exposing the aortic valve. No evidence of valvular or supravalvular aortic stenosis was found. The counterclockwise rotation of the aortic root was confirmed so that the right coronary cusp was on the left side (fig. 6). The right and noncoronary cusps were easily identified and both were moderately thickened and demonstrated slight nodularity of their free edge. Initially the left coronary cusp and left coronary ostium could not be identified.

After perfusion of the right coronary artery was begun, a membranous structure, located posteriorly and toward the right side of the annulus, was seen to distend. At this point, it was appreciated that this structure was the left coronary cusp with its normally free edge adherent to the aortic wall, thereby creating a blind sac totally covering the left coronary ostium. The cusp margin was incised and completely freed from the aortic wall. Arterialized blood gushed from the left coronary ostium as soon as it was opened. The left coronary cusp was easily freed, and except for mild thickening of its edge, was intact and smoothly

Figure 3
The aortic root cineangiogram reveals dilatation of the ascending aorta and moderately severe aortic insufficiency. The adherent aortic cusp is seen. Ao = aorta; LV = left ventricle; RCA = right coronary artery; LCA = left coronary artery; AAC = adherent aortic cusp.
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Figure 4

A. Selective injection into the right coronary artery reveals that the left coronary artery fills from the right coronary artery by way of a large tortuous conal artery and prominent septal perforating branches. A saclike structure representing the adherent left aortic cusp covers the left coronary artery ostium.

B. Graphic reproduction of figure 4A. LAD = left anterior descending coronary artery; Circ A = circumflex coronary artery; SPA's = septal perforating arteries; AVB = anterior ventricular branch.

lined with a glistening surface. The blood perfusing the right coronary artery and the blood flowing retrogradely from the left coronary artery both had an oxygen saturation of 98%. The aortotomy was closed, and since no gross evidence of aortic insufficiency was found, prosthetic aortic valve replacement was not undertaken. Biopsy of the right atrial appendage was obtained. Immediately following electrical defibrillation of the heart, a left bundle-branch block pattern was noted. This persisted until discharge.

Postoperative convalescence was rapid and complete. Transient atrial flutter occurred on the second day which was easily terminated by cardioversion.

Follow-up Examination

Three months after operation, the patient was readmitted for follow-up study. She was completely free of symptoms. Physical examination revealed a pulse rate of 70/min and blood pressure of 110/70 mm Hg. There was no edema nor abnormal venous waves. The heart was not enlarged. A grade III/VI, harsh systolic ejection murmur was heard best over the aortic area, and there was a faint grade I/VI diastolic blow along the left sternal border audible only during full expiration and in the leaning forward position. The remainder of the physical examination was normal. The electrocardiogram demonstrated normal sinus rhythm. The previously noted left bundle-branch block had disappeared. Chest x-rays revealed the heart and lungs to be normal. Repeat catheterization (table 1) revealed normal pressures throughout with persistence of a mild aortic systolic gradient (20 mm Hg). The aortic

Figure 5

A later stage after the injection than figure 4. The catheter is still in the right coronary artery. Contrast material has left the right coronary artery, while portions of the left coronary artery and the blind sac formed by the adherent aortic cusp remain opacified. AAC = adherent aortic cusp; LCA = left coronary artery.
abnormalities of the semilunar valves or great vessels as found in this patient has not been previously reported. However, adherence of an aortic cusp to the aortic wall with resultant obstruction of the right or left coronary artery has been reported in five patients, but in each instance this was associated with supravalvular aortic stenosis.\textsuperscript{5-12} In three of these patients\textsuperscript{5, 10, 12} the obstruction of the coronary artery was felt to be responsible for, or contribute to, the symptom angina. In two patients, aortic insufficiency was present. However, in none of these patients was the cause for the valvar adherence evident. It is possible that our case may represent a forme fruste of supravalvular aortic stenosis.

It is impossible to conclude with any degree of certainty whether the anomaly in our patient was congenital or acquired. An inflammatory process of the aorta or aortic valve, or both, would seem to be a logical setting for adherence of an aortic cusp. Although a history of rheumatic fever was not elicited and no evidence of rheumatic carditis was found in the right atrial biopsy, a rheumatic etiology cannot be excluded. No clinical evidence of any other type of inflammatory process capable of involving the aorta or valves was found. While the irregular thickening of the valve surfaces may represent old inflammatory

root cineangiogram demonstrated very trivial aortic regurgitation and prompt antegrade opacification of the left coronary arterial system (fig. 7). Selective right coronary cineangiography demonstrated a normal right coronary artery with absence of the previously noted extensive collateral channels. Since antegrade opacification of the left coronary artery was seen on the aortic root cineangiogram, selective catheterization of that vessel was not undertaken.

At present, 15 months after operation, the patient continues to be totally free of symptoms.

**Discussion**

Isolated adherence of a coronary cusp of the aortic valve to the aortic wall without other
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disease, this may also be due to incomplete valvular differentiation.13-15 If this anomaly were indeed the consequence of an antecedent episode of aortitis, it is surprising that such a lesion of the aortic cusp has not been previously described in cases of known inflammatory valvular disease. A recent review of the embryologic development of the semilunar valves and their abnormalities does not offer any evidence which would support the concept of a congenital etiology in this case.16 Despite this, the early age of onset of symptoms suggests a congenital etiology. The rotation of the origins of the coronary orifices also suggests some type of developmental anomaly. Several reports cite incomplete differentiation and dysplasia of the aortic17, 18 or pulmonic valves15 as a cause of stenosis of these valves. A similar mechanism may explain the persistent aortic valvular systolic gradient of 20 mm Hg in this patient.

In this patient, angina pectoris was the major basis for the diagnostic investigation. The presence of this symptom in cases of aortic insufficiency usually implies massive left ventricular volume overloading19, 20 or a special condition such as syphilis20-22 or aortic dissection23 compromising one or both coronary ostial orifices. The amount of aortic insufficiency judged to be present in this case did not appear to be sufficient to produce symptoms of myocardial ischemia. Syphilitic ostial stenosis was excluded on the basis of the young age21 at which symptoms began and the negative blood serologic test for syphilis.24 Aortic dissection likewise seemed remote in a young patient lacking features of those diseases which predispose to dissection.25 Syncope, an unusual symptom in uncomplicated aortic insufficiency, was an additional reason for undertaking diagnostic investigation in this patient.

Although electrocardiographic evidence of myocardial ischemia was lacking, the patient's history was typical for the anginal syndrome. No documentation of the mechanism of the syncopal episodes was available, but a transient ventricular tachyarrhythmia provoked by myocardial ischemia seemed plausible. During resting conditions, myocardial oxygen extraction was normal and lactate extraction was in the low and borderline range.26 Although myocardial oxygen consumption was not measured, it can be assumed that the rise in heart rate, tension-time index, and myocardial inotropy during the isoproterenol infusion was accompanied by an increase in myocardial oxygen consumption.27, 28 The narrowing of the myocardial arteriovenous oxygen difference during isoproterenol infusion suggests that left coronary blood flow increased. With total obstruction of the left coronary artery ostium, this augmented left coronary blood flow had to be derived through collateral channels. Since aortic perfusion pressure was unchanged, the rise in coronary blood flow must have resulted from a drop in coronary vascular bed resistance. Whether this auto-regulation occurred as a result of a drop in resistance at the level of the collateral channels themselves or in the left coronary vascular bed, or both, cannot be determined.

The pattern of coronary arterial collateral filling and flow closely resembles that following ligation of a left coronary artery that arises anomalously from the pulmonary artery. It is also similar to the flow observed in cases of a single coronary artery. When the left coronary cusp was freed, during the operation, fully saturated blood from the perfused right coronary artery escaped from the left coronary ostium into the aortic lumen. Since the left coronary artery was not perfused at this point, the open ostium permitted diversion of myocardial blood supply in a fashion similar to that seen with anomalous origin of the left coronary artery from the pulmonary artery. This, in fact, may have been the reason for the development of the left bundle-branch block pattern.

If the white area on the right ventricular epicardial surface does in fact represent previous infarction or fibrosis, the possibility that the extensive collateral network supplying the left coronary artery may have acted to divert blood away from the region of myocardium normally supplied by the right coronary artery can be raised. Moreover, the fact that
our patient experienced typical angina during the treadmill exercise test without developing ischemic ST-segment and T-wave changes supports the idea that her angina may have reflected right ventricular ischemia. It is interesting to compare this patient’s anomaly to others in which the entire myocardial blood supply is derived from a single coronary artery. Cases of single coronary artery and ligated anomalous left coronary artery arising from the pulmonary artery derive their entire coronary blood flow from one coronary artery. In the absence of additional congenital or acquired cardiac abnormalities, the former anomaly is usually not associated with heart disease,29–31 and ligation of a left coronary artery which arises anomalously from the pulmonary artery usually terminates myocardial ischemia when present preoperatively. A recent report32 cites a single coronary artery as a cause of myocardial infarction and congestive heart failure, while ligation of an anomalous left coronary artery has not been uniformly associated with termination of myocardial ischemia.33, 34 Myocardial ischemia in these anomalies might be due to a mechanism similar to that in our patient.

Although aortic valvuloplasty has been largely abandoned except for children with congenital aortic stenosis,35 our patient’s unique anatomic setup could be corrected by a simple plastic procedure. The result of the surgery was indeed gratifying. Postoperative evaluation demonstrated normal antegrade filling of the left coronary artery and only a faint trace of residual aortic insufficiency. The failure to opacify the collateral vessels postoperatively does not necessarily imply regression of these channels but may merely be due to the fact that the left coronary artery system now has a pressure equal to that in the right coronary artery system.

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Circulation, Volume XLI, May 1970
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Circulation. 1970;41:849-857
doi: 10.1161/01.CIR.41.5.849

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

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