Anomalous Origin of the Left Circumflex Coronary Artery
Recognition, Angiographic Demonstration and Clinical Significance

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SUMMARY
Anomalous origin of the circumflex coronary artery from the proximal right coronary artery or right sinus of Valsalva was recognized in 20 of 2996 patients undergoing selective coronary arteriography (0.67%). The relative frequency of this anomaly demands a high level of anticipation during the performance of selective coronary arteriography to assure that an adequate study has been obtained. Failure to recognize and properly demonstrate the anomaly can be hazardous to patient management.

Two angiographic signs have proved reliable in recognizing the anomalous artery before its selective demonstration. These signs are a profile view of the artery behind the aortic root during left ventriculography (the "aortic root sign") and recognition of absent arterial inflow to a significant area of the posterior lateral left ventricle during selective injections of the main left coronary artery (the "sign of non-perfused myocardium"). These angiographic signs are described and the clinical implications of proper demonstration of the anomalous circumflex coronary artery are discussed.

Additional Indexing Words:
Coronary arteriography
Aortocoronary bypass graft

Case Histories
The total experience is summarized in table 1. Those patients in whom the anomaly assumed clinical importance are presented in more detail.

Patient 1. W. B., a 55-year-old male, experienced an acute myocardial infarction five years prior to evaluation. Coronary arteriography, performed because of the recurrence of angina pectoris, demonstrated an apical left ventricular aneurysm, 50% obstruction of the main left coronary artery, 90% obstruction of the anterior descending coronary artery and 100% obstruction of the right coronary artery. The left circumflex coronary artery was not identified and was assumed to be 100% obstructed. Following aneurysmectomy and saphenous vein bypass grafts to the anterior descending and right coronary arteries, he remained asymptomatic for three years. Repeat coronary arteriography performed because of recurrent angina pectoris demonstrated both vein grafts to be functioning properly. A circumflex coronary artery of relatively large distribution was found to arise anomalously from the right sinus of Valsalva and was 50% proximally obstructed. In retrospect the first study demonstrated absence of arterial distribution to the posterior lateral myocardium and the anomalous circumflex could be identified behind the aortic root in the left ventriculogram.

Patient 2. B. B., a 39-year-old female, was evaluated for progressively symptomatic rheumatic aortic and mitral valvular disease. Coronary arteriography performed at the time of right and left heart catheterization demonstrated anomalous origin of the circumflex coronary artery as a branch of the proximal right coronary artery. Subsequently mitral valvulotomy and aortic valve replacement were successfully accomplished.

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Received May 2, 1974; revision accepted for publication May 31, 1974.
Table 1
Summary of 20 Patients with Anomalous Circumflex Coronary Artery

<table>
<thead>
<tr>
<th>Patient</th>
<th>Mode of origin</th>
<th>Relative area of distribution</th>
<th>Pathology circumflex</th>
<th>Other pathology</th>
<th>Surgery</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. W.B.</td>
<td>55 yr M</td>
<td>Separate</td>
<td>Large</td>
<td>75%*</td>
<td>50% MLCA, 90% LAD, 100% RCA</td>
</tr>
<tr>
<td>2. E.M.</td>
<td>52 yr M</td>
<td>Common</td>
<td>Small</td>
<td></td>
<td>100% LAD</td>
</tr>
<tr>
<td>3. B.B.</td>
<td>39 yr F</td>
<td>Common</td>
<td>Large</td>
<td></td>
<td>MS, MI, AS, AI</td>
</tr>
<tr>
<td>4. J.B.</td>
<td>44 yr M</td>
<td>Common</td>
<td>Small</td>
<td>90%</td>
<td>75% MLCA, 90% LAD, 90% RCA</td>
</tr>
<tr>
<td>5. H.A.</td>
<td>61 yr M</td>
<td>Common</td>
<td>Small</td>
<td></td>
<td></td>
</tr>
<tr>
<td>6. R.B.</td>
<td>40 yr F</td>
<td>Branch, RCA</td>
<td>Large</td>
<td>LAD originated from pulmonary artery</td>
<td></td>
</tr>
<tr>
<td>7. W.J.</td>
<td>66 yr M</td>
<td>Branch, RCA</td>
<td>Small</td>
<td>75%</td>
<td>100% LAD, 100% RCA</td>
</tr>
<tr>
<td>8. E.M.</td>
<td>58 yr M</td>
<td>Branch, RCA</td>
<td>Large</td>
<td></td>
<td>AS</td>
</tr>
<tr>
<td>9. B.J.</td>
<td>71 yr F</td>
<td>Branch, RCA</td>
<td>Small</td>
<td>90%</td>
<td>100% LAD, 90% RCA</td>
</tr>
<tr>
<td>10. R.T.</td>
<td>67 yr M</td>
<td>Separate</td>
<td>Large</td>
<td>90% LAD</td>
<td>CAB × 2: LAD, CX</td>
</tr>
<tr>
<td>11. W.J.</td>
<td>48 yr M</td>
<td>Common</td>
<td>Large</td>
<td>50%</td>
<td>90% LAD</td>
</tr>
<tr>
<td>12. H.W.</td>
<td>39 yr M</td>
<td>Separate</td>
<td>Large</td>
<td></td>
<td></td>
</tr>
<tr>
<td>13. W.K.</td>
<td>32 yr M</td>
<td>Branch, RCA</td>
<td>Small</td>
<td></td>
<td></td>
</tr>
<tr>
<td>14. H.H.</td>
<td>57 yr M</td>
<td>Common</td>
<td>Large</td>
<td>50%</td>
<td></td>
</tr>
<tr>
<td>15. W.J.</td>
<td>63 yr M</td>
<td>Separate</td>
<td>Large</td>
<td>90%</td>
<td>90% LAD, 90% RCA</td>
</tr>
<tr>
<td>16. V.H.</td>
<td>48 yr M</td>
<td>Branch, RCA</td>
<td>Small</td>
<td>50%</td>
<td>90% LAD, 90% RCA</td>
</tr>
<tr>
<td>17. M.B.</td>
<td>43 yr M</td>
<td>Common</td>
<td>Large</td>
<td>100%</td>
<td>50% MLCA, 90% LAD</td>
</tr>
<tr>
<td>18. R.B.</td>
<td>58 yr M</td>
<td>Branch, RCA</td>
<td>Large</td>
<td>50%</td>
<td>90% LAD, 100% RCA</td>
</tr>
<tr>
<td>19. O.H.</td>
<td>53 yr F</td>
<td>Branch, RCA</td>
<td>Small</td>
<td></td>
<td></td>
</tr>
<tr>
<td>20. L.S.</td>
<td>62 yr M</td>
<td>Separate</td>
<td>Large</td>
<td>75%</td>
<td>100% RCA, 50% LAD</td>
</tr>
</tbody>
</table>

*Percentages = maximal arterial lumen diameter narrowing.

Abbreviations: MLCA = main left coronary artery; LAD = left anterior descending coronary artery; RCA = right coronary artery; CX = circumflex coronary artery; MS = mitral stenosis; MI = mitral insufficiency; AS = aortic stenosis; AI = aortic insufficiency; CAB = coronary artery bypass.

Patient 4. J. B., a 44-year-old male, experienced an acute myocardial infarction nine years prior to evaluation. Coronary arteriography performed because of the recurrence of angina pectoris demonstrated severe obstructive lesions of the main left anterior descending, and right coronary arteries. Saphenous vein bypass grafts to the left anterior descending and right coronary arteries resulted in temporary relief of symptoms. Nine months later repeat coronary arteriography, performed because of recurrent symptoms, demonstrated both bypass grafts to be functioning properly. A circumflex coronary artery of relatively small distribution was identified arising from the right sinus of Valsalva and was 90% proximally obstructed. In retrospect, the first study demonstrated absence of arterial distribution to the posterior lateral myocardium and the anomalous circumflex could be identified behind the aortic root in the left ventriculogram.

Patient 6. R. B., a 40-year-old female, complained of progressive chest pain and orthopnea. Coronary arteriography demonstrated anomalous origin of the left anterior descending coronary artery from the pulmonary artery. The circumflex coronary artery arose as a branch of the proximal right coronary artery and had probably contributed to her relatively benign past clinical course by supplying part of the left ventricle with well oxygenated blood. Surgical correction of the anomalous anterior descending artery in this patient has been reported.1

Patient 10. R. T., a 67-year-old physician, had experienced the recent onset of angina pectoris. Coronary arteriography demonstrated 90% proximal obstruction of the left anterior descending coronary artery. In reviewing his cineangiograms it was recognized that the circumflex coronary artery was not demonstrated. Repeat coronary arteriography the following day demonstrated an anomalous but normal circumflex coronary artery arising independently from the right sinus of Valsalva.

Patient 17. M. B., a 43-year-old male, was evaluated for chest pain. Coronary arteriography demonstrated 100% proximal obstruction of the left anterior descending coronary artery. An anomalous circumflex coronary artery was faintly opacified by spillback from injections into the right coronary artery. This was not recognized prior to surgery. He underwent a saphenous vein bypass graft to the anterior descending coronary artery. Two years later repeat coronary arteriography was performed because of recurrent chest pain. The bypass graft was patent, but the circumflex coronary artery was noted to fill retrograde through collaterals from the right coronary artery and was 100% obstructed near its origin.

Patient 20. L. S., a 62-year-old male, was evaluated for progressive angina pectoris. The anterior descending, circumflex and right coronary arteries arose from the right sinus of Valsalva by three separate ostia and were all significantly obstructed. Independent visualization of the three arteries is illustrated in figure 1. He subsequently underwent bypass grafts to all three vessels.

Circulation, Volume 50, October 1974

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Antopol and Kugel first described anomalous origin of the circumflex artery from the proximal right coronary artery or right sinus of Valsalva in 1933. It is difficult to ascertain the true incidence of the anomaly by reviewing current literature. Alexander and Griffith report eight such anomalies among 18,950 consecutive autopsies and Ogden reports 14 examples among a total of 224 coronary artery anomalies identified in an unspecified patient population. Our experience suggests that the anomaly is much more common than these data indicate and is apparently more in accord with the experience of Gensini and Kelly, who include an entry for its identification in their computerized data forms, and Effler, who suggests it be called a normal variant rather than an anomaly.

Relatively little information is available regarding the precise anatomy of the anomaly as defined by direct observation. The following impressions of proximal anatomy are based upon angiography and are basically related to the ability or inability to independently demonstrate the anomalous circumflex and right coronary arteries. The anomalous artery may arise as a proximal branch of the right coronary artery, with the right coronary artery from a common ostium, or from a separate orifice. In figure 2A it is possible to selectively opacify either vessel without the faintest demonstration of the other. In 2B injections of the right coronary artery consistently opacify the anomalous circumflex which arises as a branch (fig. 3). In figure 2C it is possible to direct the catheter tip selectively into either vessel although some degree of opacification of the nonselected artery may occur by spillback of contrast material. In our experience these three possibilities occur with approximately equal frequency. Thus in eight patients (40%) the anomalous circumflex arose as a discrete proximal branch and was consistently opacified during injections of the right coronary artery. In seven patients (35%) spillback of contrast material during injections of the right coronary artery afforded evidence of the anomaly. In five patients (25%) it was possible to selectively opacify the right coronary artery without the faintest evidence of the anomalous circumflex. The anomalous circumflex was selectively catheterized in eight patients (40%).

In these 20 examples the continued course of the anomalous circumflex artery is stereotyped as...
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Figure 3

Patient 19 O. H. Unretouched 35 mm cine frame in a left anterior oblique projection illustrating origin of the anomalous circumflex artery as a discrete branch of the proximal right coronary artery.

previously described. After taking origin by one of the above described possibilities the artery passes behind the aortic root and enters the left atrioventricular groove to distribute as though it had originated as a proximal branch of the left coronary artery (figs. 2 and 4).

The size and quantitative distribution of the anomalous circumflex artery are as variable as that of a normally originating vessel. Thus, when the right coronary artery supplies most of the inferior left ventricular myocardium the terminal circumflex is small and when the diagonal arteries from the main left or proximal anterior descending artery are well developed a typical large obtuse marginal branch is not found. Typical variations in distribution of the anomalous vessel are illustrated in figure 5.

In our experience origin from the right sinus of Valsalva or right coronary artery does not predispose the circumflex coronary artery to a higher incidence of obstructive disease. The presence of obstructive disease, however, especially in a vessel of large distribution, makes it mandatory that the anomaly be recognized and angiographically demonstrated. Since this is the most common deviation from usual coronary artery anatomy encountered in adults, a high

Figure 4

Artist’s three dimensional concept of the origin, course and distribution of an anomalous circumflex coronary artery as seen in a right anterior oblique projection.
level of anticipation should be maintained during the performance of coronary arteriography. In our experience two angiographic signs allow recognition of the anomaly before its direct visualization. These signs can be referred to as the "aortic root sign" and the "sign of nonperfused myocardium."

Aortic root sign: A consistent finding during left ventriculography in a right anterior oblique projection is a profile view of the anomalous circumflex as it courses posteriorly behind the right sinus of Valsalva. This sign is illustrated in the artists' concept (fig. 4) and in a typical cine frame (fig. 6). The single exception to this observation was a patient in whom the artery was completely occluded at its origin and the distal distribution was recognized by retrograde opacification.

Sign of nonperfused myocardium: During selective opacification of the left coronary artery an avascular area in the posterior lateral left ventricular myocardium suggests anomalous origin of the circumflex artery. Subselective injection of the anterior descending artery or separate origin of the circumflex from the left sinus of Valsalva are easily ruled out by left cusp injections. The size of the avascular area is directly proportional to the quantitative distribution of the anomalous artery and may be very obvious or rather subtle. It is unsound to hypothesize hypoperfusion of a significant area of left ventricular myocardium due to congenital absence of arterial inflow. Should this possibility be suspected, a more reasonable alternative is to assume that the study is incomplete. The concept of nonperfused myocardium is illustrated in figure 7.

In searching for the anomalous artery by the transfemoral approach the preformed catheter designed for opacification of aortocoronary vein grafts (Cordis) has been found most useful. This catheter is more versatile than the preformed right coronary catheter which often tends to select only the right coronary artery. After locating the ostium of the right coronary artery a circumflex coronary artery arising independently or from a common origin can be selectively catheterized by directing the catheter tip more posteriorly and at times more inferiorly.

The clinical significance of the anomaly is obvious in patients undergoing angiographic studies for possible obstructive coronary artery disease. In this series the anomalous artery was first recognized in three patients during repeat coronary arteriograms. In all three patients the vessel was significantly obstructed and probably had contributed to continuing clinical problems. One patient underwent repeat angiography to specifically visualize the anomalous artery when it was recognized that a significant area of posterior lateral myocardium had no visible arterial supply on the first study. In one patient aged 40 years the anomaly coexisted with origin of the anterior descending artery from the pulmonary artery and had probably contributed to her relatively benign clinical course. In two patients anticipating possible aortic and mitral valve replacement, recognition of the anomaly had potential importance related to coronary artery perfusion during surgery and to the possibility of compression by the prosthetic valve fixation rings as reported by Roberts and Morrow.9 In two patients, aortocoronary bypass grafts were placed distal to significant proximal obstructions in the anomalous circumflex arteries. The technical aspects of surgery in these two patients were no different from patients.

Figure 7
Retouched 35 mm cine frames illustrating the concept of nonperfused posterior lateral myocardium. The dotted line depicts the posterior lateral cardiac border. Frames (A) left anterior oblique and (B) right anterior oblique from a patient with normal anatomy illustrate typical anterior descending (AD) and circumflex (CX) distributions. Note that the CX approximates the posterior lateral border in both projections. Patient 12 H. W. in (C) left anterior oblique and (D) right anterior oblique projections. A large area of apparently nonperfused myocardium is noted between the anterior descending (AD) and diagonal arteries (D) and the posterior lateral cardiac border. Frames (E) right anterior oblique and (F) left anterior oblique from the same patient illustrate approximation of the posterior lateral border in both views by an anomalous circumflex coronary artery arising independently from the right sinus of Valsalva.
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with normal origin of the circumflex coronary artery.

From this experience it is concluded that anomalous origin of the circumflex coronary artery from the proximal right coronary artery or right sinus of Valsalva is a relatively common anatomic variation. Consistent demonstration of the anomaly is possible if it is anticipated and the above described indirect angiographic signs are recognized during the performance of coronary arteriography. Adequate visualization of the anomaly is essential for proper patient management.

Acknowledgment

The authors wish to acknowledge Drs. Edward Anderson, Loyda Tacogue and Peter Kaplan, each of whom performed one or more of the coronary arteriograms referred to in this report.

References

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Circulation. 1974;50:768-773
doi: 10.1161/01.CIR.50.4.768

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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