Generalized Coronary Arterio-systemic
(Left Ventricular) Fistula
Case Report and Review of Literature

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SUMMARY

A coronary artery-to-left ventricular fistula is a rare finding; to
the best of our knowledge, a total of only 35 cases have been re-
ported. Only 5 cases of a generalized arterio-systemic fistula with
three vessel involvement have been reported in the literature. We
describe another case involving all major coronary arteries. A re-
view of the literature is presented and the data of the reported cases
are analyzed.

A 55 year old woman was examined because of recurrent chest
pain which had persisted for 2 years. On physical examination,
the only abnormal finding was a fourth heart sound. Exertional
chest pain, a positive exercise stress test, and the results of a lactate
extraction study suggested severe myocardial ischemia. Thallium
myocardial scintigraphy showed no evidence of a perfusion defect.
Cardiac catheterization revealed an irregular left ventricular endo-
cardial pattern (Thebesian veins). Selective coronary angiography
showed communicating fistulae of all three major coronary arteries
with the left ventricular cavity. We assume that this vascular
anomaly causes a coronary steal phenomenon and subsequent myo-
cardial ischemia.

Additional Indexing Words:
Coronary artery-to-left ventricular fistula Myocardial ischemia
Clinical assessment

CONGENITAL anomalies of the coronary arteries are a well-known clini-
cal finding.1-3 Coronary arterial fistulae are no longer considered a
rare anomaly.4-8 The majority of these anomalies are arteriovenous con-

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nections between a coronary artery and the right atrium or ventricle; in most cases only one coronary artery is involved. Since generalized coronary artery fistulae with multiple arterio-systemic connections between each of the coronary arteries and the left ventricle are very rare, the clinical picture is not clear.

This report describes a case with generalized coronary artery-to-left ventricular fistulae and a complete review of the literature.

CASE REPORT

A 55 year old woman was admitted to the hospital for evaluation of recurrent chest pain which had persisted for 2 years. She had been admitted twice to coronary care units because of acute severe chest pain lasting longer than 30 min and STT-changes in the electrocardiogram. An acute transmural infarction could be ruled out each time. The clinical diagnosis was non-transmural infarction (non-Q-infarction) because of typical chest pain lasting more than 30 min and persisting STT-changes lasting more than 48 hours.

Chest pain with radiation to the left arm occurred during exercise and was relieved by rest and/or nitroglycerin. There was no history of dyspnea on exertion, paroxysmal nocturnal dyspnea, ankle edema, or high blood pressure.

Fig. 1. Echocardiogram shows mild fluttering of the anterior mitral leaflet during diastole (arrows). This interesting finding was reproduced in different echocardiographic examinations.
pressure. The past medical and family histories were negative except for exertional angina.

Physical examination showed: height 160 cm, weight 59 Kg, arterial blood pressure of 95/45 mmHg and a regular heart rate of 68/min. Palpation of the heart revealed no abnormal heaves or thrills; the point of maximum impulse was in the fifth left intercostal space in the midclavicular line. The first and second heart sounds were normal. There was a fourth heart sound; no murmurs were audible. The central venous pressure and the a- and v-wave in jugular vein inspection was normal. Hepato-jugular reflux was negative, the liver was not enlarged, the spleen was not palpable. There was no ankle edema and pulmonary rales were not audible. All arterial pulses were without pathologic findings upon palpation and auscultation. Multiple blood tests including lipid profile, urea, uric acid, complete blood count and serial enzyme determinations were within normal limits. A glucose tolerance test was negative.

The chest X-ray showed a normal heart silhouette. There was no evidence of ventricular or atrial enlargement or pulmonary arterial hypertension and pulmonary venous congestion. Systolic time intervals and all mechanocardiographic tracings revealed no pathologic findings. The M-mode and bi-dimensional echocardiograms showed normal cardiac chambers, valve patterns and wall thickness, except for a mild fluttering of the anterior mitral leaflet (Fig. 1). The electrocardiograms at rest and during exercise testing are shown in Fig. 2. Pulmonary artery pressures at rest were systolic 21 mmHg, diastolic 10 mmHg, with a mean of 14 mmHg. After an exercise stress test (3 min at 25 watts, bicycle ergometer in supine position) pressures were: systolic 48 mmHg, diastolic 24 mmHg, with a mean of 32 mmHg. Pressure elevation occurred at maximal ST-segment depression (0.3 mV in all leads) and typical chest pain. Thallium 201-myocardial scintigraphy with a maximal load exercise test showed no signs of a myocardial perfusion defect. The coronary sinus lactate measurements during atrial pacing are given in Fig. 3. Typical chest pain occurred during multiple exercise tests and atrial pacing and was reproducible. Holter monitoring for 48 hours revealed no significant supraventricular or ventricular arrhythmias. Cardiac catheterization showed normal pulmonary and right ventricular pressures at rest. Left ventricular pressure was 90/0–13 mmHg, aortic pressure 90/45 mmHg. The cardiac index also was normal. The aortic root injection showed a competent and normal aortic valve. The left ventricular angiogram revealed normal end-diastolic chamber dimensions and contractile patterns; the ejection fraction was 65%. During isometric and maximal systolic contraction, an irregular left ventricular endocardial pattern could be
Fig. 2. Electrocardiogram at rest and during exercise. Note STT-changes at rest and increasing ST-segment depression during exercise. HR = heart rate; BP = blood pressure; W = watt.

Fig. 3. Coronary sinus lactate content shows typical pattern during atrial pacing. Note the relationship between angina and increasing coronary sinus plasma lactate content. AP = angina pectoris; HR = heart rate; SO$_2$ = oxygen saturation.
seen (Fig. 4a); the contrast medium seemed to enter into subendocardial channels/vessels ("Thebesian veins"). Selective coronary angiography (Fig. 4b–c) showed rather dilated coronary arteries without any calcifications or

Fig. 4a. Left ventricular angiogram in the right anterior oblique projection during early systole shows diffuse sinusoidal channels entering the left ventricular myocardium.

Fig. 4b. Coronary angiogram of the left coronary artery. Note the opacification of the left ventricular cavity through a plexus of small vessels.

Fig. 4c. Coronary angiogram of the right coronary artery. Opacification of the left ventricle occurred also through a plexus of small vessels.
narrowing, even the small vessels appeared normal. The contrast material flowed into the left ventricular cavity: a cloudy opacification of the left ventricle could be seen after each injection into the left and right coronary artery. This opacification appeared from a maze of fine vessels. Similarly, opacification of the left anterior descending, the circumflex and the right coronary arteries showed a maze or plexus of small, fine vessels surrounding these arteries, with contrast material flowing into the left ventricular cavity, making it, too, appear cloudy and opacified. The latter occurred only during diastole, with injection of dye into the coronary arteries. The patient was asymptomatic under treatment with isosorbide-dinitrate (20 mg every 4 hours) and 200 mg metoprolol daily during the stay in hospital. During the follow-up period (14 months) there was no exertional angina; electrocardiographic changes (ST-T-changes) were inconstantly present.

**DISCUSSION**

A coronary artery fistula is relatively uncommon. In one series of 55,856 coronary angiographic examinations in adults, there were only 24 cases with coronary arteriovenous fistulae. Coronary arterial fistulae usually arise from a single artery, most frequently the right coronary artery, and usually communicate with the right atrium or ventricle or with the pulmonary trunk. Coronary artery-to-left ventricular fistulae are even less common. Only 35 cases of coronary artery-to-left ventricular fistulae have been reported. A review of 172 reported cases revealed only 4 in which the fistula involved both the right and left coronary arteries. Of the 25 cases of coronary artery-to-left ventricular fistula with one-vessel involvement which have been reported, 13 cases involved the right coronary artery, 10 the left anterior descending and 1 the circumflex artery. Four cases had fistulae involving two major coronary arteries—left anterior descending and circumflex combined in 3 cases, and right coronary artery and left anterior descending together in 1 case. Fistulae simultaneously involving all three major coronary arteries have been documented only 5 times previously.

The first case with multiple arterio-systemic fistulae was described by Reddy et al. In the case described by Kinard and co-workers, communications from all major coronary arteries occurred by way of the Thebesian vein system. Partial atresia of the coronary sinus and hypoplasia of some of the cardiac veins were present. A case with generalized arterio-systemic fistula and involvement of both ventricles was detected at postmortem by Rose. Case 2 in the communication by Cha et al also seems to
Table I. Coronary Artery—Left Ventricular Fistula Reported in Literature

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Ref.</th>
<th>Author</th>
<th>Origin of fistula</th>
<th>Site of drainage</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td>27</td>
<td>Lovitt et al, 1954</td>
<td>LAD</td>
<td>LV</td>
</tr>
<tr>
<td>2.</td>
<td>33</td>
<td>Abbott et al, 1961</td>
<td>RCA</td>
<td>LV</td>
</tr>
<tr>
<td>3.</td>
<td>28</td>
<td>Tanabe et al, 1967</td>
<td>RCA</td>
<td>LV</td>
</tr>
<tr>
<td>4.</td>
<td>4</td>
<td>McNamara, 1969</td>
<td>RCA</td>
<td>LV</td>
</tr>
<tr>
<td>5.</td>
<td>4</td>
<td>McNamara, 1969</td>
<td>LCF</td>
<td>LV</td>
</tr>
<tr>
<td>6.</td>
<td>29</td>
<td>Eguchi et al, 1970</td>
<td>RCA</td>
<td>LV</td>
</tr>
<tr>
<td>7.</td>
<td>30</td>
<td>DeNef et al, 1971</td>
<td>LCA</td>
<td>LV, RV</td>
</tr>
<tr>
<td>8.</td>
<td>26</td>
<td>Galioto et al, 1971</td>
<td>RCA</td>
<td>LV</td>
</tr>
<tr>
<td>9.</td>
<td>31</td>
<td>Okuda et al, 1973</td>
<td>RCA</td>
<td>LV</td>
</tr>
<tr>
<td>10.</td>
<td>32</td>
<td>Vlodaver et al, 1975</td>
<td>LAD</td>
<td>LV</td>
</tr>
<tr>
<td>11.</td>
<td>34</td>
<td>Sastri et al, 1975</td>
<td>LAD</td>
<td>LV</td>
</tr>
<tr>
<td>12.</td>
<td>35</td>
<td>Muraki et al, 1976</td>
<td>RCA</td>
<td>LV</td>
</tr>
<tr>
<td>13.</td>
<td>36</td>
<td>Midell et al, 1977</td>
<td>LAD</td>
<td>LV</td>
</tr>
<tr>
<td>14.</td>
<td>37</td>
<td>Arani et al, 1978</td>
<td>LAD</td>
<td>LV</td>
</tr>
<tr>
<td>15.</td>
<td>38</td>
<td>Kiso et al, 1978</td>
<td>RCA</td>
<td>LV</td>
</tr>
<tr>
<td>16.</td>
<td>25</td>
<td>Dobell et al, 1981</td>
<td>RCA</td>
<td>LV</td>
</tr>
<tr>
<td>17.</td>
<td>25</td>
<td>Dobell et al, 1981</td>
<td>RCA</td>
<td>LV</td>
</tr>
<tr>
<td>18.</td>
<td>24</td>
<td>Chia et al, 1981</td>
<td>RCA</td>
<td>LV</td>
</tr>
<tr>
<td>19.</td>
<td>24</td>
<td>Chia et al, 1981</td>
<td>RCA</td>
<td>LV</td>
</tr>
<tr>
<td>20.</td>
<td>22</td>
<td>Ahmed et al, 1982</td>
<td>LCA</td>
<td>LV</td>
</tr>
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<td>21.</td>
<td>22</td>
<td>Ahmed et al, 1982</td>
<td>LCA</td>
<td>LV</td>
</tr>
<tr>
<td>22.</td>
<td>23</td>
<td>Cheng, 1982</td>
<td>LAD</td>
<td>LV</td>
</tr>
<tr>
<td>23.</td>
<td>45</td>
<td>Brooks et al, 1983</td>
<td>LCA</td>
<td>LV</td>
</tr>
<tr>
<td>24.</td>
<td>46</td>
<td>Santomi et al, 1983</td>
<td>RCA</td>
<td>LV</td>
</tr>
<tr>
<td>25.</td>
<td>47</td>
<td>Nakashima et al, 1983</td>
<td>unknown</td>
<td>LV</td>
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<tr>
<td>26.</td>
<td>20</td>
<td>Rose, 1978</td>
<td>LAD, LCF</td>
<td>LV</td>
</tr>
<tr>
<td>27.</td>
<td>20</td>
<td>Rose, 1978</td>
<td>LAD, RCA</td>
<td>LV</td>
</tr>
<tr>
<td>28.</td>
<td>21</td>
<td>Cha et al, 1978</td>
<td>LAD, LCF</td>
<td>LV</td>
</tr>
<tr>
<td>29.</td>
<td>24</td>
<td>Chia et al, 1981</td>
<td>LAD, LCF</td>
<td>LV</td>
</tr>
<tr>
<td>30.</td>
<td>19</td>
<td>Reddy et al, 1974</td>
<td>LAD, LCF, RCA</td>
<td>LV</td>
</tr>
<tr>
<td>31.</td>
<td>39</td>
<td>Kinard, 1975</td>
<td>LAD, LCF, RCA</td>
<td>LV</td>
</tr>
<tr>
<td>32.</td>
<td>20</td>
<td>Rose, 1978</td>
<td>LAD, LCF, RCA</td>
<td>LV, RV</td>
</tr>
<tr>
<td>33.</td>
<td>21</td>
<td>Cha et al, 1978</td>
<td>LAD, LCF, RCA</td>
<td>LV</td>
</tr>
<tr>
<td>34.</td>
<td>24</td>
<td>Chia et al, 1981</td>
<td>LAD, LCF, RCA</td>
<td>LV</td>
</tr>
<tr>
<td>35.</td>
<td></td>
<td>Present case</td>
<td>LAD, LCF, RCA</td>
<td>LV</td>
</tr>
</tbody>
</table>

Ref. = reference; LAD = left anterior descending artery; LCA = left coronary artery; LCF = left circumflex artery; LV = left ventricle; RCA = right coronary artery; RV = right ventricle.

have involved all three major coronary arteries.21) Chia et al24) reported a case without a prominent Thebesian vein system or concomitant anomalies of the venous system of the heart. No further case has been reported to the
Table II. Generalized Coronary Arterio-systemic Fistulae Reported in Literature,
Overview of the Clinical and Electrocardiographic Findings

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Ref.</th>
<th>Author</th>
<th>Chest pain, duration</th>
<th>Physical findings</th>
<th>X-ray</th>
<th>Electrocardiographic findings</th>
<th>Exercise stress test</th>
<th>Other</th>
</tr>
</thead>
<tbody>
<tr>
<td>27</td>
<td>19</td>
<td>Reddy et al, 1974</td>
<td>10 years</td>
<td>0</td>
<td>norm.</td>
<td>non-spec. STT-changes</td>
<td>not done</td>
<td></td>
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<tr>
<td>28</td>
<td>39</td>
<td>Kinard, 1975</td>
<td>apical systolic murmur</td>
<td>LVE</td>
<td>norm.</td>
<td>neg.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>29</td>
<td>20</td>
<td>Rose, 1978</td>
<td>0</td>
<td>post-mortem</td>
<td>(sudden death)</td>
<td>bicuspid aortic valve</td>
<td></td>
<td></td>
</tr>
<tr>
<td>30</td>
<td>21</td>
<td>Cha et al, 1978</td>
<td>0</td>
<td>S₄</td>
<td>norm.</td>
<td>T-wave changes inferolat.</td>
<td>neg.</td>
<td>CPK-elevation</td>
</tr>
<tr>
<td>31</td>
<td>24</td>
<td>Cha et al, 1981</td>
<td>3 years</td>
<td>0</td>
<td>norm.</td>
<td>ST-depression T-inversion II, aV₂, V₄, V₆</td>
<td>pos.</td>
<td></td>
</tr>
</tbody>
</table>

Ref. = reference; norm. = within normal limits; pos. = positive; neg. = negative; LVE = left ventricular enlargement.

best of our knowledge in which all three major coronary arteries were involved in a complex coronary artery-to-left ventricular fistula system (Table I). The clinical picture and the significance of this condition are not clear. The clinical, physical and electrocardiographic findings of the 5 cases with generalized arterio-systemic fistulae are summarized in Table II. There were no pathological physical findings in the cases involving all three coronary arteries, including ours, but a fourth heart sound was audible in some cases. The pathophysiologic effects of arterio-systemic fistulae are related to the amount of blood draining into the left ventricle. Fistulous bypass flow could produce a coronary steal phenomenon that might induce exertional chest pain, and significant ischemia indicated by STT-segment changes. Fistulae would be diastolic overload of the left ventricle. Kiso et al reported a case with right coronary artery-to-left ventricular fistula with a blood flow through the fistula of about 20% of cardiac output. In our case it seems that there was no severe diastolic volume overload on the left ventricle.

With a large amount of coronary blood draining into the left ventricular cavity, the hemodynamic effects must be analogous to the hemodynamics of
aortic regurgitation.\textsuperscript{19,22} Our patient showed a pulse wave pressure of 40-45 mmHg with an aortic pressure of 95/45 mmHg. There was also diastolic flutter of the anterior mitral leaflet in the M-mode echocardiogram. Diastolic mitral valve flutter is present in aortic regurgitation, in some congenital defects\textsuperscript{48} without aortic regurgitation, and other types of heart disease. In our case there was no aortic regurgitation or a congenital heart disease. This finding may be due to blood flow into the left ventricle during diastole.

Most patients in the literature, like our patient, had chest pain and electrocardiographic changes compatible with ischemia.\textsuperscript{19,24} Our patient had a history of exertional chest pain and severe STT-segment changes in the resting electrocardiogram. The exercise test was strongly positive with reproducible anginal chest pain and downward sloping ST-segment depressions, plus a positive nitrate test. During atrial pacing, lactate values in the coronary sinus increased, especially with the onset of chest pain at a frequency of 150/min. These findings clearly indicate diffuse myocardial ischemia.

Perfusion defects in stress thallium studies are reported by Cheng\textsuperscript{23} and Ahmed\textsuperscript{22}; these findings most probably were due to involvement of only one coronary artery. In our case, all major coronary arteries were involved in a complex arterio-systemic fistula system. No perfusion defect could be detected in thallium-201 myocardial scintigraphy at rest or after maximum exercise stress testing. Balanced and diffuse ischemia may still result in a uniform perfusion scintigram. Exertional chest pain, diffuse STT-segment changes in the electrocardiogram at rest, a positive exercise stress test with significant ST-T-segment depression in all leads, and diminished lactate extraction in the coronary blood in the absence of a perfusion defect in the thallium-201 study, indicate diffuse and generalized myocardial ischemia in our case.

The presence of sinusoidal channels during isovolumetric contraction in the left ventricular angiogram in our patient indicates the existence of a Thebesian vein system\textsuperscript{4,19,21,40,42,43} in the left ventricular myocardium. The existence of such sinusoidal channels in the human heart has been questioned. The Thebesian vein system was thought to be a major venous channel for the right heart only.\textsuperscript{42} Only 2% of the coronary blood flows into the canine left atrium through such a Thebesian vein system.\textsuperscript{43} Some investigators considered these channels as the nutrient supply of the myocardium.\textsuperscript{44} Cha and co-workers\textsuperscript{21} pointed out the absence of any pathophysiologic significance of this "angiographic curiosity". These investigators regarded the fistula-channels as an unusual, prominent Thebesian system which seems to be the embryological etiology of this condition. Although their patients did not suffer from exertional myocardial ischemia, they considered
the well-developed sinusoidal channels (with a possible large amount of fistula flow) as a probable cause of coronary steal, and subsequent coronary ischemia. In conclusion, our findings suggest that this vascular anomaly of arterio-systemic fistula is not just an "angiographic curiosity without any pathophysiologic significance ", as assumed by Cha. Both data in the literature and our findings indicate an association with severe exercise-induced myocardial ischemia, leading to therapeutical consequences of clinical interest. Classic anti-anginal treatment may be effective.

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