Aorto-Inferior Vena Caval Fistula of Neoplastic Origin

Hemodynamic and Coronary Blood Flow Studies

By E. Stanley Crawford, M.D., David J. Turell, M.D., and James K. Alexander, M.D.

AORTO-VEHA caval fistula is an uncommon condition. In the cases previously reported it has been due to violent trauma, accidental vascular injury at the time of intervertebral disk surgery, and rupture or erosion of atherosclerotic and syphilitic aortic aneurysms into the vena cava. A survey of the English literature indicates that surgical treatment has been successful in 12 cases. The purpose of this paper is to record an unusual case of aorto-vena caval fistula resulting from neoplasia, with certain physiologic studies secured before and after successful resection therapy. Data relative to hemodynamic changes, coronary blood flow, and myocardial metabolism are presented.

Case Report

A 51-year-old white male springbender was admitted to the Methodist Hospital (case no. 61-3503), Houston, Texas, February 28, 1961. The patient had been well until 1 month prior to admission, when both lower legs began to swell. At the same time he noted enlargement of saphenous vein varicosities bilaterally, which had been present for some years with no symptoms. Initially the swelling was present only during the latter part of the day, but soon it progressed, involving the lower legs and thighs, and was present to some extent upon arising in the morning. The varicosities became larger, and could not be controlled by external elastic support. The patient estimated that he had gained about 30 pounds in weight during the several weeks prior to admission. The patient had noted no other symptoms.

Physical examination revealed a stocky, 207 pound individual in no distress (fig. 1). All peripheral pulses were present, regular at a rate of 100, and forceful with a slight water-hammer quality. Blood pressure taken in the right arm in the supine position was 140/70 mm. Hg. There was a dominant "A" wave in the jugular venous pulse, and venous pressure was estimated clinically to be at the upper limit of normal. There was a soft ejection murmur over the precordium, maximal just inside the apex. The abdomen was covered with enlarged, prominent veins, and an ill-defined, rounded, pulsatile mass could be felt in the lower abdomen just below the umbilicus. Auscultation over the abdomen revealed a machinery murmur, best heard on the right side and in the right flank. Both legs were edematous to the groin and there were many large varicos veins which did not collapse on elevation of the legs. Pulsations could be felt in the veins segments in the thighs, especially near the groin.

Routine studies of blood and urine were normal, as was an electrocardiogram. The serologic test for syphilis was negative. Roentgenographic examination of the chest showed slight enlargement of the cardiac silhouette, particularly in the region of the left ventricle (fig. 2a). Translumbar aortographic examination, performed March 1, 1961, showed an aneurysmal-type shadow overlying the distal aorta with simultaneous opacification of the aorta and inferior vena cava, indicating the presence of an aorto-vena caval fistula (fig. 3a-b).

Physiologic data* secured at the time of right

*The diagnosis was made on clinical grounds by the family physician, George Hart, M.D., Houston, Texas.

†The patient's height was 67.3 inches. Weight fell from 200 pounds immediately preoperatively to 170 pounds postoperatively, due to loss of edema. For this reason, no attempt has been made to express results as units per square meter body surface area. Right heart catheterization was performed in the usual manner and cardiac output estimated by the Fick principle as previously described. The nitrous oxide method was used for determination of coronary blood flow. Plasma volume was measured by the T 1954 dye-dilution method. Blood pH was determined at 37°C, with use of a glass electrode pH meter, and blood gas concentrations were measured with the Van Slyke manometric apparatus. Arterial carbon dioxide tension was calculated from the Henderson-Hasselbalch nomogram. Blood lactate content was measured by the Barker-Summerson method.
heart catheterization, March 2, 1961, are summarized in tables 1-4. As shown in table 1, pulmonary wedge and arterial pressure were at the upper limit of normal or slightly elevated at rest preoperatively, with an appreciable rise in pulmonary artery mean pressure during exercise to nearly double the resting value. Systemic arterial pulse pressure was increased to 88 mm Hg. Intracardiac blood oxygen content determinations yielded no evidence of intracardiac shunt. However, inferior caval blood was 4 volumes percent higher in oxygen content than that in the superior vena cava, a clearly abnormal finding supportive of the presence of a large arteriovenous fistula draining into the inferior cava or

Figure 1
Photograph made with infrared light showing edema of legs and prominence of veins over legs, abdomen, and chest.

Figure 2
Roentgenograms of the chest made before operation (a, upper) and 12 days after operation (b, lower) showing decrease in size of heart after operation.
its tributary veins. With use of the blood oxygen content values shown in table 1, it can be calculated that approximately 75 per cent of the total blood flow through the pulmonary artery was derived from the inferior vena cava, whereas only 50 to 60 per cent are so derived under normal conditions. If it is assumed that the difference between cardiac output determinations before and after operation represents blood flow through the fistula, this flow was 5.7 liters per minute, or 48 per cent of the preoperative cardiac output. As indicated in table 2, resting cardiac output was approximately double the normal value. This was effected by an increase in both cardiac rate and stroke volume. Due to fixed stroke volume, the increase in output with exercise was achieved entirely by a faster heart rate. The systemic arteriovenous oxygen difference at rest was re-
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Table 1

Vascular Pressures and Blood Oxygen Content*

<table>
<thead>
<tr>
<th></th>
<th>Pressures, mm. Hg</th>
<th>O₂ content, Vol. %</th>
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<tbody>
<tr>
<td></td>
<td>RBA</td>
<td>PC</td>
</tr>
<tr>
<td>Pre-op</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Rest</td>
<td>141/59</td>
<td>89</td>
</tr>
<tr>
<td>Exercise</td>
<td>182/83</td>
<td>122</td>
</tr>
<tr>
<td>Post-op</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Rest</td>
<td>135/72</td>
<td>98</td>
</tr>
<tr>
<td>Exercise</td>
<td>167/85</td>
<td>116</td>
</tr>
</tbody>
</table>

*Pressure levels in the right brachial artery (RBA) and right pulmonary artery (RPA) are indicated as systolic/diastolic and mean. The ‘‘pulmonary capillary’’ or wedge (PC) and the right atrial (RA) pressures are mean values. Superior and inferior vena cava are designated SVC and IVC respectively.

Table 2

Hemodynamic Data*

<table>
<thead>
<tr>
<th></th>
<th>Cardiac output (L/min.)</th>
<th>Arteriovenous O₂ diff. (vol. %)</th>
<th>Stroke vol. (ml.)</th>
<th>Heart rate beats/min.</th>
<th>Plasma vol. (L)</th>
<th>Blood vol. (L)</th>
<th>Hematocrit (%)</th>
<th>Peripheral resistance (dynes sec. cm⁻²)</th>
<th>ΔC.O./ΔV.O₂ (ml/min ml.O₂)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pre-op</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>Rest</td>
<td>11.9</td>
<td>2.9</td>
<td>120</td>
<td>99</td>
<td>4.25</td>
<td>7.38</td>
<td>36</td>
<td>565</td>
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<tr>
<td>Exercise</td>
<td>15.7</td>
<td>5.1</td>
<td>112</td>
<td>139</td>
<td>..</td>
<td>..</td>
<td>..</td>
<td>..</td>
<td></td>
</tr>
<tr>
<td>Post-op</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>Rest</td>
<td>6.2</td>
<td>4.5</td>
<td>84</td>
<td>74</td>
<td>3.27</td>
<td>5.27</td>
<td>40</td>
<td>1290</td>
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<tr>
<td>Exercise</td>
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<td>6.3</td>
<td>92</td>
<td>96</td>
<td>..</td>
<td>..</td>
<td>..</td>
<td>950</td>
<td></td>
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</tbody>
</table>

*Preoperative value for peripheral vascular resistance included flow through the fistula. If this is excluded, the calculated value is 1010 dynes sec. cm⁻². Δ C.O./ΔV.O₂ indicates the increase in cardiac output in ml. per 100 ml. increase in O₂ uptake by the body during exercise.

duced. Plasma volume and estimated total circulating blood volume were greatly increased. When calculation of systemic vascular resistance included flow through the fistula, a value below normal was obtained. However, if flow through the fistula was excluded, calculated resistance was within the normal range. With utilization of a figure of approximately 600 ml. increase in cardiac output per 100 ml. increase in body oxygen uptake as the lower limit of normal during steady state exercise, it may be concluded that the patient's response to exercise was within normal limits as regards cardiac output. There was moderate hyperventilation both at rest and during exercise as shown in table 3, with an accompanying respiratory alkalosis. The oxygen uptake of the body at rest was elevated, giving a calculated basal metabolic rate of +31 per cent, even when the preoperative weight was used. Of particular note were the data relative to coronary blood flow and myocardial metabolism at rest shown in table 4. Both myocardial blood flow and oxygen uptake were elevated to levels approximately 150 per cent of the predicted normal, while the coronary arteriovenous oxygen difference remained normal. Calculated coronary vascular resistance was appreciably reduced. The increase in calculated left ventricular work to approximately double the predicted normal, was accompanied by a grossly augmented myocardial lactate uptake.

The patient was given 1.5 mg. of digoxin in three divided doses during the 24 hours of March 2, 1961, and 0.25 mg. every day thereafter while in the hospital. Operation was performed on March 3, 1961, when the patient weighed 200 pounds. The distal abdominal aorta, vena cava, common iliac arteries and veins, and the external iliac arteries and veins were exposed through a long midline abdominal incision. A pulsating tumor mass (fig. 3b) was found overlying the distal abdominal aorta and vena cava. The mass intimately involved the distal aorta and proximal common iliac arteries, overlying the vena cava and right common iliac vein as well. A thrill could be felt over the vena cava and the common iliac veins, which ceased with pressure on the tumor mass. Arterial clamps were placed across the aorta above, and the common iliac vessels below the tumor. The thrill again ceased. The tumor was easily dissected from the vena cava and right common iliac veins. A segment of vena caval wall was removed in the region of the fistula providing a good margin of resection. The defect in the vena cava was closed longitudinally by simple continuous over-and-over suture. The tumor mass was then removed by resecting the distal

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aorta, proximal common iliac arteries and adjacent tissues en masse. Arterial continuity was restored by inserting a bifurcation knitted Dacron graft (fig. 3c). By means of an electromagnetic flow meter with probes in situ before and at the end of operation, it was found that blood flow in the abdominal aorta proximal to the region of the fistula was one third that before resection. Blood flow in the right external iliac artery distal to the region of the fistula was five times that measured before resection. Pathologic examination of the tumor later demonstrated an unclassified malignant mesenchymoma involving the aorta and vena cava.

Postoperative convalescence was uncomplicated. Fluid intake was limited to 1,500 ml. of 5 per cent dextrose in water for the first 4 days. During this period there was an extraordinary diuresis, and body weight fell to 180 pounds. Oral feedings were started 5 days after operation and diuresis proceeded for several days thereafter. Serum electrolyte determinations remained normal during this time. A translumbar aortogram (fig. 3d), performed March 15, 1961, showed normal vessels. Chest roentgenogram the same date revealed an appreciable decrease in the size of the cardiac silhouette (fig. 2b). At discharge, 15 days after operation, the patient weighed 169 pounds, having achieved a total weight loss of 38 pounds from the day of admission.

The physiologic data obtained 2 weeks after closure of the fistula are tabulated in tables 1 to 4. As noted in table 1, systemic arterial pulse pressure fell to normal, largely as a result of the rise in diastolic pressure. Pulmonary wedge and arterial pressures were normal at rest, and the latter remained so during moderate exercise. Inferior vena caval blood contained only 1.5 volumes per cent more oxygen than that in the superior cava, constituting a normal relationship. The hemodynamic data, shown in table 2, were quite normal at rest. Heart rate had fallen to 74 beats per minute, stroke volume to 84 ml., cardiac output to 6.2 liters per minute, and plasma volume to 3.27 liters. Systemic arteriovenous oxygen difference and calculated vascular resistance had risen to normal values of 4.5 volumes per cent and 1,200 dynes sec. cm.\(^{-2}\), respectively. A normal response to exercise in terms of cardiac output was again achieved. As shown in table 3, pulmonary ventilation, body oxygen uptake, and arterial blood pH had fallen to normal at rest. Arterial blood carbon dioxide tension was normal, and the calculated basal metabolic rate was +13 per cent. As indicated in table 4, coronary blood flow, myocardial oxygen and lactate uptake, and calculated left ventricular work were reduced to normal values. Coronary arteriovenous oxygen difference remained unchanged, and calculated coronary vascular resistance rose to normal.

The patient has now been followed for 1 year since discharge from the hospital. There has been no recurrence of leg swelling or other signs previously associated with the fistula. He has, however, developed back pain with radiation into the left leg during the past 3 months. The pain may represent recurrent tumor, although there are no other signs suggesting this.

Discussion

Certain clinical findings in this case are characteristic of aorto-inferior caval fistula. These are rapidly progressive swelling of the lower extremities, distended veins over the legs and abdomen, and machinery-like murmur over the abdomen and flank, in the presence of cardiac enlargement and a high output state. These signs assume significance in view of the incidence of such causative conditions as abdominal trauma and atherosclerotic aortic aneurysm, now remediable by surgical procedures commonly employed in the treatment of aortic disease. Neoplasia would appear to be a very rare cause of this condition. However, in the light of the experience reported here, it should be suspected in each case. Two differentiating factors in the diagnosis are the absence of a history of trauma, and the absence of abnormal aortic calcification suggesting aneurysm in roentgenograms of the abdomen. Curability of malignant tumor by resectional therapy is doubtful, since fistulous formation in general represents an advanced manifestation of the disease process. For this reason and because of the recent onset of appropriately located nerve root pain, permanent cure of the tumor in this patient is unlikely. Despite these limitations of therapy, the hemodynamic palliation, permitting a return to normal activity, seems worth while.

Unfortunately, it has been impossible to classify the tumor precisely in this case. It is undoubtedly of mesenchymal origin arising from periaortic tissue. Renal arteriovenous fistula is not uncommon with certain tumors of the kidney. However, the kidneys and other abdominal organs were normal in this case. There were no testicular abnormalities, and pregnancy tests were negative, excluding functioning metastatic testicular tumor.

Hemodynamic findings in the presence and absence of the disease, and its operative cor-
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Respiratory Data

<table>
<thead>
<tr>
<th></th>
<th>Ventilation (L./min.)</th>
<th>O2 uptake (ml./min.)</th>
<th>Respiratory exchange ratio (R.Q.)</th>
<th>Tidal vol. (ml. BTPS)</th>
<th>Arterial blood CO2 tension (mm. Hg)</th>
<th>Arterial blood pH</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pre-op Rest</td>
<td>12.42</td>
<td>345</td>
<td>.76</td>
<td>16</td>
<td>776</td>
<td>25</td>
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<tr>
<td>Pre-op Exercise</td>
<td>29.20</td>
<td>803</td>
<td>.88</td>
<td>23</td>
<td>1269</td>
<td>26</td>
</tr>
<tr>
<td>Post-op Rest</td>
<td>9.27</td>
<td>278</td>
<td>.77</td>
<td>15</td>
<td>617</td>
<td>37</td>
</tr>
<tr>
<td>Post-op Exercise</td>
<td>19.02</td>
<td>558</td>
<td>.90</td>
<td>16</td>
<td>1187</td>
<td>34</td>
</tr>
</tbody>
</table>

The pronounced increase in pulmonary ventilation found in our patient may be related partially to the elevated oxygen consumption of the body and possibly to the presence of cardiac failure. The tendency to hyperventilation and respiratory alkalosis he demonstrated is quite characteristic of congestive failure of any etiology, though other mechanisms may have been operative. The increase in body oxygen consumption, based on the postoperative reappraisal, was approximately 70 ml. per minute. Even if a heart weight of as much as 500 Gm. is assumed, only 20 ml. per minute or 40 per cent of the total output with exercise was within the normal range, the rise in pulmonary artery pressure and the fall (or absence of increase) in stroke volume with exercise represent abnormal responses. We consider the development of pulmonary hypertension as indicating an elevated diastolic filling pressure in the left ventricle with exercise. This, taken with the fixed or reduced stroke volume, has been interpreted as evidence for myocardial failure of at least modest degree. Pulmonary hypertension at rest with arteriovenous fistula, and failure to increase cardiac output with exercise has been previously reported. This was interpreted by the authors as indicating frank cardiac failure. It is interesting that the abnormal response observed in our patient occurred when he was fully digitalized. Presently available evidence indicates little or no beneficial effect following digitalis administration in the congestive state accompanying arteriovenous fistula. In a study of 12 dogs with chronic aorto-inferior caval fistulas and hemodynamic evidence of myocardial failure, no improvement could be demonstrated following digitalis administration.
change in oxygen consumption can be accounted for on the basis of increased myocardial oxygen requirement, as shown in table 4. A single instance of increased oxygen consumption with systemic arteriovenous fistula has been previously reported, but was not commented upon by the authors. In our patient, the systemic blood flow, exclusive of flow through the fistula, was normal, or at least the same as the postoperative systemic flow. We have no explanation for the observed increase in body oxygen consumption.

Of particular interest in this study are the findings relative to coronary blood flow and myocardial metabolism. The preoperative and postoperative data make it quite clear that the systemic arteriovenous fistula was accompanied by an increase in coronary blood flow and myocardial oxygen uptake, associated with coronary vasodilatation as indicated by the change in calculated vascular resistance and an unchanged oxygen extraction per unit volume of blood flow. A tendency to increased coronary flow with systemic arteriovenous fistula has been reported previously in two cases, but without observations on the effect of obliterating the fistula. This study demonstrates that these changes do occur, and may revert to normal after elimination of the fistula. It may be concluded further that the changes accompanying systemic arteriovenous fistula represent no exception to the general proposition that coronary flow is adjusted to the energy requirements of the heart.

Summary
A case report of a 51-year-old man is presented, who spontaneously developed an aorto-vena caval fistula in the region of a mesenchymal tumor involving both the abdominal aorta and vena cava. The disease was manifested clinically by rapidly progressive and massive leg swelling, prominent abdominal and leg veins, a machinery murmur over the abdomen and in the flanks, and a pulsatile abdominal mass, with signs of cardiac enlargement and a high output state. The fistula was demonstrated preoperatively by translumbar aortography and closed surgically by resection of the tumor en masse, with a part of the vena caval wall and the distal aorta and proximal common iliac arteries. Vascular continuity was restored by lateral suture of the vena cava and by insertion of a bifurcation graft bridging the defect in the aorta and iliac arteries. The patient has been relieved of all clinical manifestations of the fistula to date, 1 year after operation.

Preoperative hemodynamic studies revealed certain alterations characteristic of systemic arteriovenous fistula, including increased pulse pressure, high cardiac output and stroke volume, reduced systemic vascular resistance and arteriovenous oxygen difference, and increased plasma and circulating blood volumes. Some degree of cardiac failure appeared to be present. Coronary arteriovenous oxygen difference was normal, indicating that the observed increase in coronary blood flow closely paralleled the augmented energy requirement of the heart as reflected by the ventricular oxygen uptake. Reversion to normal of the observed changes in systemic hemodynamics, coronary blood flow, and myocardial metabolism was demonstrated 2 weeks postoperatively.

Table 4

<table>
<thead>
<tr>
<th></th>
<th>Coronary flow (ml./100 Gm./min.)</th>
<th>Coronary A-V O₂ difference (Vol. %)</th>
<th>Myocardial O₂ uptake (ml./100 Gm./min.)</th>
<th>Coronary vascular resistance (mm. Hg./ml./100 Gm./min.)</th>
<th>Myocardial lactate uptake (mg./100 Gm./min.)</th>
<th>Left ventricular work (Kg. meters/min.)</th>
</tr>
</thead>
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<tr>
<td>Pre-op Rest</td>
<td>131</td>
<td>11.8</td>
<td>15.4</td>
<td>.74</td>
<td>0.67</td>
<td>11.5</td>
</tr>
<tr>
<td>Post-op Rest</td>
<td>93</td>
<td>12.3</td>
<td>11.4</td>
<td>.71</td>
<td>1.04</td>
<td>0.8</td>
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</table>

References

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