Coarctation of the Abdominal Aorta*

Report of a Case and Review of the Literature

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Coarctation usually occurs in the thoracic rather than the abdominal aorta. When the coarctation occurs in the abdominal aorta, it may be congenital or acquired, localized or diffuse, involve the aorta alone, or also narrow the gastrointestinal or renal arteries. In this article, the literature on coarctation of the abdominal aorta is reviewed and a case successfully treated by surgery is reported. The patient had three separate coarctations of the abdominal aorta, stenosis of the origins of both renal arteries, thrombosis of the superior mesenteric artery, and an abdominal aortic aneurysm.

CASE REPORT

The patient, L.M., a 19-year-old white man, was referred for consultation in November, 1961 when a routine pre-employment examination revealed hypertension, an abdominal bruit, and reduced femoral arterial pulsations. Family history was non-contributory. There was no history of rheumatic fever or scarlet fever. He was not limited by symptoms, but on direct questioning admitted weakness and fatigue in the legs with strenuous exercise and nausea after large meals.

The patient was well developed, 69 inches tall, and weighed 140 pounds. The optic fundi were hyperemic and the retinal vessels more tortuous and more prominent than usual. Blood pressure was 210/110 in the right arm, 200/105 in the left arm and 110/90-95 in both legs. Femoral pulses were barely perceptible and no distal pulses were detected. The heart was slightly enlarged with marked left ventricular thrust and palpable presystolic filling impulse at the apex. On auscultation, aortic valve closure was accentuated and a soft protodiastolic filling sound was followed by a loud presystolic gallop. There was a Grade II out of IV ejection systolic murmur heard along the left sternal border and over the carotid and subclavian vessels. In the epigastrium, a palpable systolic thrill and continuous murmur were present. The continuous murmur was also heard in the back at the level of the 10th to 12th thoracic vertebrae. Intercostal vessels were felt at the lower borders of the fifth to tenth ribs and dilated pulsating arteries were palpable at the lower costal margin anteriorly. The legs appeared normally developed with good nutrition of the skin, but the arterial pulses were absent distal to the femoral level as noted. The remainder of the physical examination was negative.

Electrocardiograms showed left ventricular hypertrophy by voltage without T wave inversion in the left precordial leads at rest or after exertion. Chest roentgenograms showed notching of the inferior borders of the fifth to ninth ribs. The ascending aorta was prominent to the right.

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Figure 1: X-ray film of chest showing notching of the fifth to tenth ribs.
but no enlargement of the aortic knuckle or descending thoracic aorta was noted. There was a round area of eggshell calcification situated anteriorly to the second lumbar vertebra; it measured 3 cm. in diameter (Fig. 1). Aortography indicated that the thoracic aorta distal to the left subclavian artery was moderately hypoplastic. Both internal mammary arteries were greatly

**Figure 2:** Antero-posterior and lateral views of abdomen showing calcified aneurysm in front of the second lumbar vertebra.

**Figure 3:** Aortogram showing hypoplastic lower thoracic aorta, enlarged intercostal arteries, and the first coarctation just above the celiac axis.
dilated, as were the sixth to tenth intercostal arteries. The aorta narrowed at a point immediately above the celiac artery and the splenic and hepatic branches of the celiac artery were large. No superior mesenteric artery was visible. A localized area of stenosis at the origin of the right renal artery was apparent, but the left renal artery was of normal calibre. The calcified mass situated immediately distal to the origin of the right renal artery did not fill with contrast media (Fig. 2). The inferior mesenteric artery was huge and blood passed in retrograde fashion through the left and middle colic arteries to the gastro-duodenal artery and remaining colic branches. The abdominal aorta below the inferior mesenteric artery was hypoplastic as were the iliac arteries. A systolic pressure gradient of 110 mm.Hg existed between the thoracic and abdominal aorta and the major change in pressure occurred at the level of the renal arteries (Fig. 3).

No difference between the two kidneys was found on split renal function tests. An I\textsuperscript{131}I-Hippuran scan demonstrated slightly less uptake of radioactivity over the left kidney.

The patient was placed on a 1,000 mg. sodium diet and antihypertensive drugs. During the subsequent six months he received reserpine, chlorothiazide, hydralazine and guanethidine, but the blood pressure was not consistently controlled, rising as high as 260/180 mm.Hg after exertion and falling to 150/100 mm.Hg when standing at rest. With lower blood pressures the patient developed episodes of orthostatic hypotension, particularly in the morning. The physical signs and the electrocardiographic evidence of left ventricular hypertrophy were not altered by this course of treatment and it was felt that in view of the age of the patient, and the difficulty in obtaining effective regulation of blood pressure, surgery should be performed. Operation was performed on June 29, 1962 using an intrapleural, retroperi toneal approach through the left side. The descending thoracic aorta was hypoplastic from the left subclavian artery distally. It measured approximately 1.5 cm. in diameter. Enlarged intercostal vessels were present below the sixth to tenth ribs. The aorta narrowed sharply just above the level of the celiac axis. The celiac artery itself was small but immediately expanded to a large hepatic and splenic artery. The aorta, in the area in which the celiac artery arose, was

**Figure 4:** Drawing of the preoperative anatomy.

**Figure 5:** Drawing of the reconstructive procedure.
somewhat larger than that immediately distal. The superior mesenteric artery was present, but was completely thrombosed. The tissue in this area of the abdominal aorta was exceedingly hard and fibrotic and difficult to dissect. As the aorta in the region of the renal vessels was cleared of this fibrotic tissue, the amplitude of pulsation in the right renal vessel increased notably; when it was completely dissected free of fibrotic tissue, the stenotic area at the origin of the right renal vessel had disappeared. The left renal artery remained small despite removal of the fibrotic tissue, the stenotic area at the origin of the right renal vessel had disappeared. The left renal artery remained small. The right renal artery was an aneurysm arising from the aorta immediately below the renal arteries and constricting the aortic lumen at this level. The inferior mesenteric artery was very large (1 cm.). The aorta continued distally with a diameter of approximately 1 cm. The iliac vessels were very small. Pressures were 205/110 mm.Hg in the thoracic aorta and 100/70 mm.Hg in the aorta at the level of the inferior mesenteric artery. The pressure in the left renal artery was identical to that in the distal portion of the abdominal aorta. The lesion, therefore, consisted of a hypoplastic thoracic aorta from the level of the subclavian artery distally, and a complete, or almost complete, coarctation above the level of the celiac artery with a second complete or almost complete coarctation distal to the celiac artery. The aorta from the second coarctation to the renal arteries was severely hypoplastic, became moderately hypoplastic at the level of the renal arteries, and immediately distal to them was constricted again by the aneurysm. Thus, the abdominal aorta was hypoplastic and contained three separate coarctations. The left renal artery was small. The right renal artery had been narrowed by a periarterial fibrous constricting process. The superior mesenteric artery was occluded. The celiac artery was small at its origin from the aorta but immediately enlarged. The inferior mesenteric artery was huge and was the major blood supply to the gastro-intestinal tract, liver and spleen (Fig. 3).

The aorta between the renal arteries and the inferior mesenteric artery was excised. This area of the aorta included the aneurysm, which was in communication with the aorta through a small orifice and had a smooth endothelial lined wall which was heavily calcified. The resected area of aorta was replaced with a 3/4-inch knitted Teflon graft. A second Teflon graft (diameter 9/16 inch) was then inserted between the left renal artery and the thoraco-abdominal bypass graft. This produced excellent pulsations in the left renal artery. At the completion of the procedure the patient's blood pressure was 130/80 and normal pulsations were present in all palpable abdominal and leg arteries (Fig. 4).

On histologic examination, the wall of the aorta, in the area of the aneurysm, had normal layers but was surrounded by an increased zone of dense fibrous tissue. The wall of the aneurysm itself was composed mainly of fibrous tissue and there were no arterial elastic fibers present. Organized and calcified thrombus lined the interior of the aneurysm. In the sections of the aneurysm wall there was no evidence of a pre-existing inflammatory lesion.

The patient's postoperative course was complicated with many minor crises. On the evening following surgery, a repeat laparotomy was necessary to evaluate a flank hematoma. Continuous oozing from many points was seen, but no large bleeding vessels were found. In the first postoperative week, he had episodes of bleeding from both the gastrointestinal tract and the urinary tract. A recurring left pleural effusion required thoracenteses on three occasions. Persistent fever, anemia and leukocytosis associated with abdominal pain and right lower quadrant tenderness raised diagnostic considerations of appendicitis, retroperitoneal hemorrhage or infection, or arteritis of vessels previously protected from hypotension by the coarctation. It was considered possible that the crampy abdominal pain, nausea and episodic vomiting which followed eating were related to removal of celiac and superior mesenteric ganglia during surgical dissection. At operation, both adrenal veins were ligated, but postoperative evaluation of adrenocortical function revealed no impairment.

The patient returned to normal activity six weeks after surgery. At that time, blood pressure in the arms measured 190/100 mm.Hg and in the legs 170/90 mm.Hg. The femoral pulses were readily palpable and faint pulsations were detected in the dorsalis pedis arteries. Heart size was unchanged and the systolic murmur and atrial gallop rhythm were present.

Evaluation six months and one year after surgery revealed persistent hypertension of mild to moderate degree with values in the upper limbs ranging from 140/70 to 200/110. Blood pressure in the lower limbs was approximately 140/80. The pulsations in the lower extremities and the cardiac findings remained as noted six weeks after operation. The electrocardiogram continued to show evidence of left ventricular hypertrophy. An aortogram showed excellent function of the bypass graft with filling of both renal arteries (Fig. 5).
Eight months after operation, the patient noted enlargement of a pigmented nevus on the right calf. This was widely resected and histologic examination revealed a malignant melanoma without extension to the margin of the specimen. Shortly after, a large, firm node was detected in the groin, which was resected and demonstrated secondary deposits. One year after reconstructive vascular surgery, the patient had evidence of widespread metastases.

**DISCUSSION**

*Nature of the Lesion:*

Whether coarctation of the abdominal aorta is congenital or acquired is controversial. Mayock proposed that it arose from unequal fusion of the two embryonic dorsal aortae (omphalomesenteric arteries) with subsequent obliteration of one of them. Syphilis, rheumatic fever, non-rheumatic inflammation, and neoplasia have been proposed by those who favor an acquired etiology, and the recent reports of Inada suggest a possible association with "pulseless disease." Like "pulseless disease" and renal fibromuscular hyperplasia, this lesion is more common in women, whereas coarctation of the aortic isthmus is more common in men. The dense fibrotic tissue surrounding the area of coarctation is frequently interpreted as evidence of an acquired etiology. Pathologic evidence of neurofibromatosis (Von Recklinghausen's disease) was found both in tissue removed from the site of coarctation and elsewhere in two patients with this disorder. Since there is no conformity to the pathologic picture, it seems reasonable to speculate that cases with long segments of hypoplasia of the abdominal aorta are congenital in origin, whereas cases with more localized lesions may be caused by a non-specific aortitis characterized by the production of dense periaortic fibrous tissue.

Pyoraia, using the classification of D'Abreu, reviewed 27 cases of abdominal coarctation. He noted that 17 were segmentary (63 per cent) and ten involved a considerable length of hypoplastic aorta (37 per cent). Both segmentary and hypoplastic varieties could be suprarenal, at the level of the renal arteries, or infrarenal. In some instances, the hypoplasia began in the lower thoracic aorta or continued distally beyond the iliac vessels; any or all of the branches of the abdominal aorta could be involved by the process. With occlusion or narrowing of the celiac and superior mes...

**Figure 6:** Angiogram six months after surgery showing patent grafts from thoracic to abdominal aorta and to left renal artery.
enteric arteries, as in the present case and that reported by Bayliss,4 the major blood supply to the intestines, liver and spleen was derived from the inferior mesenteric artery. The renal arteries may be normal, hypoplastic, narrowed at the origin, or occluded by thrombosis.

The present example of coarctation of the abdominal aorta appears to represent a combination of two areas of segmental coarctation, one above, and one below the celiac axis. A third area of narrowing was present at the site of the calcified aneurysm. A calcified aneurysm distal to the coarctation was also noted in the case of Bayliss and aneurysms of the right renal artery were reported in the cases of Glenn4 and Senning.9 The mechanism of aneurysm formation distal to coarctation of the aortic isthmus has been studied by Skandalakis,16 and Dunhill,17 and it is reasonable to speculate that any congenital weakness of the aortic wall would be aggravated by the turbulence, cavitation and increased lateral pressure beyond the coarctation. The possibility of superimposed mycotic infection, as noted by Kieffer9 in 19 cases of thoracic coarctation, was favored by the youth of the patient, the localized nature of the aneurysmal bulge, and the calcification in the wall, but there was no history to indicate infection and no pathologic evidence of a specific inflammatory process.

Clinical Features:

The symptoms of coarctation of the abdominal aorta vary with the location and severity of the lesion. When the coarctation is above the renal arteries, the symptoms resemble those of classic coarctation and are related chiefly to hypertension. Leg weakness and intermittent claudication are rare. When coarctation involves the renal vessels, the hypertension may be particularly severe, and heart failure and uremia may occur.18 When the coarctation is infrarenal, hypertension is usually mild, and intermittent claudication of the buttock or calf is usually the presenting symptom.18 The physical signs of hypertension in the upper extremities, and diminished or absent pulsations in the legs permit the diagnosis of coarctation, but the abdominal site of narrowing is indicated by the presence of a thrill and murmur in the epigastrium, and by the prominent collateral arterial circulation over the lower portion of the chest. On radiologic examination, the aortic knob appears normal, and notching is seen on the lower rather than the upper ribs. The precise location of the coarctation and the extent of involvement of other vessels can be established with aortography. Renal function studies may demonstrate impaired function not suspected from the aortograms.

Senning9 proposed that the natural history of this disorder was similar to that of coarctation at the aortic isthmus. He noted that ten of 32 patients with this disorder died before the age of 34. It might be expected that lesions above or involving the renal arteries would constitute a more serious risk.

Surgical Treatment:

Prior to the era of reconstructive vascular surgery, the treatment of this type of coarctation was either impossible or completely unsatisfactory. Several attempts at sympathectomy and nephrectomy are recorded.9 The first case operated upon with reconstructive vascular technique was that of Beattie19 in 1951. Since then, there have been 20 reported cases (Table 1). From perusal of the case reports, several lessons may be learned: (1) exact knowledge of the site and extent of the coarctation must be obtained by angiography prior to surgery; (2) renal function tests may be necessary to demonstrate if renal blood flow or function is impaired; (3) dense fibrous tissue surrounding the aorta, when removed, often leads to marked enlargement of the aorta and its branches; (4) when the renal arteries are not involved, either bypass grafting, patch grafting, or local excision of the coarcted area are equally successful; (5) involvement of the renal arteries makes surgery more difficult and the prognosis less favorable; (6) and extraperitoneal approach through the left side has
## Table 1—Reported Cases of Reconstructive Surgery for Coarctation of the Abdominal Aorta

<table>
<thead>
<tr>
<th>Author</th>
<th>Year Reported</th>
<th>Age</th>
<th>Sex</th>
<th>Location</th>
<th>Type*</th>
<th>Fibrosis</th>
<th>Aneurysm</th>
<th>Repair</th>
<th>Renals Involved</th>
<th>Result</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 Beattie et al.</td>
<td>1951</td>
<td>19</td>
<td>F</td>
<td>Above renal</td>
<td>S</td>
<td>No</td>
<td>No</td>
<td>Bypass graft</td>
<td>No</td>
<td>Good</td>
</tr>
<tr>
<td>2 Albanese et al.</td>
<td>1952</td>
<td>12</td>
<td>F</td>
<td>Below renal</td>
<td>H</td>
<td>No</td>
<td>No</td>
<td>Resection and graft</td>
<td>Yes</td>
<td>Died—Anuria</td>
</tr>
<tr>
<td>3 Glenn et al.</td>
<td>1952</td>
<td>19</td>
<td>F</td>
<td>Above renal</td>
<td>S</td>
<td>Yes—Neurofibromatosis</td>
<td>Yes—Re-Renal</td>
<td>Splenic Art to Thoracic Aorta</td>
<td>No</td>
<td>Good</td>
</tr>
<tr>
<td>4 Guastavino et al.</td>
<td>1956</td>
<td>11</td>
<td>F</td>
<td>Above renal</td>
<td>S</td>
<td>Yes</td>
<td>No</td>
<td>Resection and Anastomosis</td>
<td>No</td>
<td>Good</td>
</tr>
<tr>
<td>5 Gerbaai et al.</td>
<td>1958</td>
<td>11</td>
<td>F</td>
<td>Above renal</td>
<td>S</td>
<td>Yes</td>
<td>No</td>
<td>Bypass graft</td>
<td>No</td>
<td>Good</td>
</tr>
<tr>
<td>6 D'Abreu et al.</td>
<td>1959</td>
<td>40</td>
<td>M</td>
<td>Above renal</td>
<td>H</td>
<td>Yes</td>
<td>No</td>
<td>Bypass graft</td>
<td>No</td>
<td>Good</td>
</tr>
<tr>
<td>7 D'Abreu et al.</td>
<td>1959</td>
<td>58</td>
<td>F</td>
<td>Below renal</td>
<td>S</td>
<td>No</td>
<td>No</td>
<td>Bypass graft</td>
<td>No</td>
<td>Good</td>
</tr>
<tr>
<td>8 Hanson et al.</td>
<td>1959</td>
<td>12</td>
<td>F</td>
<td>Above and at renal</td>
<td>H</td>
<td>Yes</td>
<td>No</td>
<td>Posterior patch graft</td>
<td>Yes</td>
<td>Good</td>
</tr>
<tr>
<td>9 Milloy et al.</td>
<td>1959</td>
<td>18</td>
<td>M</td>
<td>Above, at &amp; below renal</td>
<td>H</td>
<td>No</td>
<td>No</td>
<td>Bypass and subsequent left nephrectomy</td>
<td>Yes</td>
<td>Good</td>
</tr>
<tr>
<td>10 Senning et al.</td>
<td>1960</td>
<td>25</td>
<td>M</td>
<td>Above and at renal</td>
<td>H</td>
<td>Yes—Neurofibromatosis</td>
<td>Yes—Re-Renal</td>
<td>Posterior patch graft</td>
<td>Yes</td>
<td>Good</td>
</tr>
<tr>
<td>11 Senning et al.</td>
<td>1960</td>
<td>25</td>
<td>M</td>
<td>Below renal</td>
<td>H</td>
<td>No</td>
<td>No</td>
<td>Bypass graft</td>
<td>No</td>
<td>Good</td>
</tr>
<tr>
<td>12 Stokes et al.</td>
<td>1960</td>
<td>12</td>
<td>F</td>
<td>Above and below renal</td>
<td>H</td>
<td>No</td>
<td>No</td>
<td>Bypass graft with side arm to renal</td>
<td>Yes</td>
<td>Good</td>
</tr>
<tr>
<td>13 Morris et al.</td>
<td>1960</td>
<td>12</td>
<td>F</td>
<td>Above renal</td>
<td>S</td>
<td>Yes</td>
<td>No</td>
<td>Bypass graft</td>
<td>No</td>
<td>Good</td>
</tr>
<tr>
<td>14 Morris et al.</td>
<td>1960</td>
<td>20</td>
<td>F</td>
<td>Above renal</td>
<td>H</td>
<td>Yes</td>
<td>No</td>
<td>Enderectomy</td>
<td>No</td>
<td>Good</td>
</tr>
<tr>
<td>15 Morris et al.</td>
<td>1960</td>
<td>16</td>
<td>F</td>
<td>Above and below renal</td>
<td>H</td>
<td>No</td>
<td>No</td>
<td>Bypass graft</td>
<td>Yes</td>
<td>Died-Heart Failure</td>
</tr>
<tr>
<td>16 Morris et al.</td>
<td>1960</td>
<td>1</td>
<td>M</td>
<td>Above and at renal</td>
<td>H</td>
<td>No</td>
<td>No</td>
<td>Bypass graft</td>
<td>Yes</td>
<td>Died-Heart Failure</td>
</tr>
<tr>
<td>17 Morris et al.</td>
<td>1960</td>
<td>35</td>
<td>F</td>
<td>Above renal</td>
<td>S</td>
<td>No</td>
<td>No</td>
<td>Bypass graft</td>
<td>No</td>
<td>Good</td>
</tr>
<tr>
<td>18 Indana et al.</td>
<td>1962</td>
<td>43</td>
<td>M</td>
<td>Above renal</td>
<td>H</td>
<td>No</td>
<td>No</td>
<td>Bypass graft</td>
<td>No</td>
<td>Good</td>
</tr>
<tr>
<td>19 Bjork et al.</td>
<td>1964</td>
<td>18</td>
<td>M</td>
<td>Above, at &amp; below renal</td>
<td>H</td>
<td>No</td>
<td>No</td>
<td>Bypass graft with side arm to right renal</td>
<td>Yes</td>
<td>Good</td>
</tr>
<tr>
<td>20 Baird et al.</td>
<td>1964</td>
<td>19</td>
<td>M</td>
<td>Above, at &amp; below renal</td>
<td>H</td>
<td>Yes</td>
<td>Yes (aortic)</td>
<td>Bypass graft with side arm to right renal</td>
<td>Yes</td>
<td>Good</td>
</tr>
</tbody>
</table>

*S=segmental, H=hypoplastic.
many advantages in this type of surgery since it allows the entire abdominal aorta to be visualized and gives adequate exposure for angioplastie procedures on both renal arteries.

Postoperative complications may arise from adrenal insufficiency due to ligation of adrenal veins, mesenteric vasculitis, or gastro-intestinal disturbances due to denervation of the small bowel. Hence, the convalescent period following operation may be prolonged and troublesome.

SUMMARY

Successful surgical correction of abdominal aortic coarctation is reported. The structural abnormalities present included three sites of narrowing of the abdominal aorta, hypoplasia of the thoracic and abdominal aorta, thrombosis of the superior mesenteric artery, narrowing of the right renal artery, and a calcified aneurysm distal to the coarctation. The patient presented with severe hypertension which was not effectively controlled by medical measures. Following reconstructive vascular surgery, the blood pressure in the upper extremities was reduced, and improved arterial pulsations were present in the legs.

The anatomic and clinical features of patients with coarctation of the abdominal aorta are briefly reviewed and problems related to surgical treatment discussed.

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