Descending thoracic and abdominal aortic coarctation in the young: Surgical treatment after percutaneous approaches failure

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Descending thoracic and abdominal aortic coarctations are characterized by a segmental narrowing that frequently involves the origin of the visceral and renal arteries. Optimal primary treatment is debated, being reported for both surgical and percutaneous complications. We describe our surgical experience with two youths presenting with failure of distal descending aortic stenting and with abdominal aortic coarctation post-balloon angioplasty, and associated thrombosis of a stented right renal artery and stenosis of the origin of the superior mesenteric artery (SMA). In both cases, a longitudinal aortoplasty was performed with a polytetrafluoroethylene (PTFE) patch, using simple aortic cross-clamping. Renal thrombosis and SMA stenosis were managed with eversion technique. In-hospital course was uneventful. Midterm follow-up showed absence of significant restenosis and better control of hypertension. In order to refrain from operating on these patients as long as possible, and also because of the very high risk of a redo-surgery, we think that an initial balloon angioplasty should be considered. Surgical management can be adopted, even after failure of percutaneous treatments, with satisfactory short- and midterm vessels patency. (J Vasc Surg 2008;47:865-7.)

Descending thoracic and abdominal aortic coarctations are very rare diseases that occur in young patients.[1,2] These forms, also defined as middle aortic syndrome, are related to congenital disorders or to acquired nonspecific aortitis, such as Takayasu syndrome, Williams syndrome, neurofibromatosis, fibromuscular dysplasia, retroperitoneal fibrosis, and mucopolysaccharidosis.[3-5] Aortic specimens have a similar pathology, characterized by chronic inflammation and collagen proliferation that leads to segmental narrowing of the distal descending thoracic or abdominal aorta, and often involves the origin of the visceral and renal arteries.[2-5] Optimal primary treatment is still undefined, with both surgical and percutaneous approaches reporting acute and chronic complications.[1,3,5-8] We describe our surgical experience with two youths after percutaneous approaches failure.

CASE REPORT

Patient 1. A 12-year-old girl was transferred to our hospital for an abdominal aortic coarctation. Initially, she had episodes of cyanosis and the cardiac evaluation showed hypertensive cardiomyopathy. One year prior, the young girl was submitted to a right renal artery stenting and abdominal aortic balloon angioplasty to treat a nonhemodynamically significant gradient. On admission, she was tachycardic with a 2/6 systolic murmur auscultated at the apex, absent femoral pulses, and mild renal insufficiency (creatinine 1.45 mg/dL). The echo Doppler demonstrated a severely dilated cardiomyopathy of the with severe left ventricular hypokinesis (LVDd = 67 mm, E.F.= 15%) and mild to moderate mitral regurgitation. Cardiac catheterisation showed obliteration of the stented right renal artery, severe abdominal coarctation just below the renal arteries, and severe stenosis of the origin of the superior mesenteric artery. The NMR confirmed the vascular lesions, associated hypertrophic Riolan's arc, (Fig 1). She was on four medications to control hypertension and congestive heart failure. Primary aortic stenting was not considered because of the need to perform right renal revascularization. The aorta was approached through a thoraco-phreno-laparotomy (TPL) in the ninth intercostal space with left kidney left in its bed. The right renal artery was exposed for approximately 2 cm from the origin. No adjuncts were utilized for preventing spinal cord ischemia, except passive hypothermia at 32° of esophageal temperature. Renal protection was administered using cold lactates at 4°. Eparin 1 mg/kg was given before simple aortic cross-clamping, positioned at supradiaphragmatic level. Left postero-lateral aortotomy was carried out longitudinally, from the celiac axis to the subrenal abdominal portion. The stent was carefully removed from the right renal artery using the eversion technique and a distal renal arteriotomy on the common trunk was performed to confirm the absence of intimal flaps. Renal arteriotomy was than sutured with interrupted stitches using polypropylene 7/0. Accordingly, eversion technique was adopted also to remove a fibrous plaque at the ostium of the superior mesenteric artery, without necessity of distal arteriotomy. Aortoplasty was accomplished from the celiac axis to the subrenal abdominal aorta using PTFE patch. Aortic cross-clamping time was 42 minutes.

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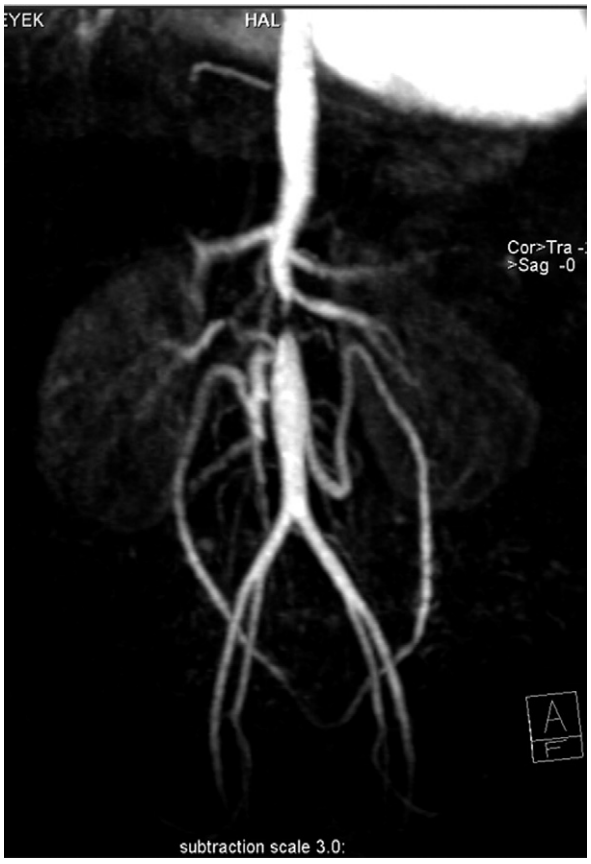

Fig 1. Preoperatory angioNMR showing severe abdominal coarctation just below the renal arteries.

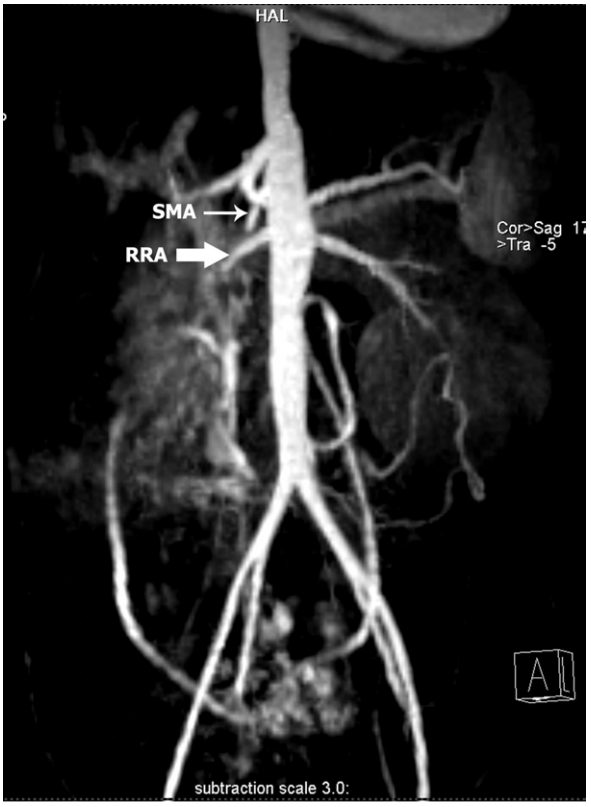

Fig 2. Severe stenosis of the origin of the superior mesenteric artery.

Postoperatory course was uneventful, with restoration of normal plasmatic renal function (creatinine 0.38 mg/dL). A postoperative color-flow Doppler evaluation revealed normal abdominal aortic diameter, patency of the right renal artery and superior mesenteric artery, confirmed by a NMR control (Fig 2). A 2-year follow-up Doppler sonography showed normal patency of the abdominal aorta. The superior mesenteric artery and the right renal artery were also patent with a trivial and a moderate increase in peak systolic and diastolic velocities related to an approximately 30% restenosis, respectively. The young girl has normal renal function and only requires two medications to control her hypertension and congestive heart failure.

Patient 2. A 10-year-old boy with hypertension and the absence of femoral pulses was admitted to our hospital. He had been previously treated, 6 months prior, with a primary stenting for a distal thoracic aortic coarctation. After that, he developed recoil of the aortic stent, and 3 months later, he underwent balloon angioplasty, with a decrease in gradient from 35 mm Hg to 15 mm Hg. A subsequent 30-day follow-up examination detected the absence of femoral pulses. At that time, he was taking three medications to control his hypertension. Preoperative CT scan showed a severe aortic stenosis at the supraceliac level. Surgical treatment was similar to the previous patient, through a TPL in the ninth intercostal space, using simple aortic cross clamping. No adjuncts were utilized for preventing spinal cord ischemia and renal failure, except passive hypothermia. Left postero-lateral aortotomy was carried out longitudinally, from the supradiaphragmatic aorta to the infrarenal abdominal portion. The aortic stent was gently removed and aortoplasty was performed using a PTFE patch. Simple aortic cross clamping was 33 minutes. Postoperatory course was uneventful, with normal plasmatic renal function (creatinine 0.56 mg/dL) and no in-hospital need for anti-hypertensive therapy. A 16-month follow-up Doppler sonography showed normal patency of the supra-infradiaphragmatic aorta. The young boy required only one medication to control hypertension at that time.

DISCUSSION

The results of surgical repair for descending thoracic and abdominal aortic coarctation reported in the literature are limited to a small number of experiences and patients. They range from reports of an absence of surgical mortality with good long-term results[1,5] to significant operative mortality with early postoperative complications and incidence of graft occlusion.[6] Accordingly, the results of using balloon angioplasty, indicated as an alternative treatment to

avoid surgery on the developing aorta, have been infrequently reported.[3,7-9] The existing literature reports encouraging results with no mortality rate, but limitations are described, related to technical failures,[3,7] iatrogenic aortic dissections,[3,8,9] and restenosis.[3] Use of aortic stenting has been reported only to treat complications such as dissections and recurrent stenosis.[3] Although stents have been successfully used to treat restenosis in other locations,[10] its long-term effect on luminal patency in a young population is unknown. During later life, because of the increasing diameter of the aorta over time, stent may result in hemodinamic obstruction, potentially compromising later surgery. This justifies refraining from operating on these patients as long as possible.[11] Nonspecific aortitis, typically expressed in the young age, involve all layers of the aortic wall and frequently the origin of the visceral and renal arteries by a chronic inflammatory process with connective tissue proliferation, cell infiltration, and fibrosis. Studies have also shown that the inflammatory processes are present in a continuous length of the aorta, producing extensive wall changes, even in the angiographically normal areas,[12] which may explain the incidence of restenosis after surgical and/or the percutaneous approach. Surgical strategies include aortoplasty and bypass grafting from the ascending or supraceliac aorta to the infrarenal aorta.[1,5,6] In patient 1, aortoplasty was planned for two reasons: to manage the segmental extensions of the coarctation and to perform a longitudinal aortotomy to safely remove, using the eversion technique, the thrombosed stent from the right renal artery and treat the stenotic ostium of the superior mesenteric artery in the same manner. The satisfactory surgical result in patient 1 influenced our decision in the second less complex patient. Patient 1 had preoperative renal insufficiency, and, accordingly, cold lactates at 4° were administered for preventing renal failure. In patient 2, we decided not to use adjuncts for preventing spinal cord ischemia, except passive hypothermia at 32° of esophageal temperature, because of the luxurious collateral circulation.

Our experience, although limited, shows that in pediatric patients, surgery for coarctation of the distal thoracic and abdominal aorta is feasible and indicated when an elevated risk of stent recoiling/thrombosis exists, or if aortic rupture occurs following further dilatation.[1,5] Although the incidence of endovascular failure is high in these patients, we think that an initial balloon angioplasty should be considered in order to refrain from operating as long as possible, and also, because of the very high risk of a redo-surgery. Surgical management can be adopted, even after failure of percutaneous treatments, with satisfactory short and midterm vessels patency and control of blood pressure, cardiac and renal function.

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