Coarctation of the Aorta in an Adult*
Problems of Diagnosis and Management
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CASE REPORT

A 41-year-old man was initially evaluated 6 years previously by his family physician, found to be hypertensive, and managed with pharmacologic agents. Over the next several years, control of his BP became increasingly difficult, requiring multiple agents. He was referred to a cardiologist for further evaluation. Throughout the entire period, he has remained asymptomatic.

The patient’s history revealed that 3 years ago, he underwent echocardiography, which disclosed a strong likelihood of a bicuspid aortic valve that, by Doppler interrogation, was mildly regurgitant. The mitral valve was believed to demonstrate prolapse with mild regurgitation. He was also found to have hyperlipidemia and was treated with simvastatin. Results of a treadmill exercise test at that time were normal. His mother had undergone mitral valve replacement several years previously; otherwise, no other cardiovascular disease was evident in family members. Current medications consisted of hydrochlorothiazide/losartan, 25/100 mg/d; amlodipine, 5 mg/d; and simvastatin, 20 mg/d.

Physical examination disclosed a BP of 160/94 mm Hg and a heart rate of 75 beats/min. Precordial pulsations were normal. Auscultation disclosed a late-peaking systolic murmur heard well at the apex; however, it was also heard over the entire thoracic cage and upper back (Fig 1). No diastolic murmurs were audible. Simultaneous palpation of the radial and femoral pulses disclosed a significant delay of the latter. The systolic pressure in the lower extremities was 130 mm Hg, determined with a Doppler probe over the pedal vessels, yielding an ankle/brachial index of 0.85. The remainder of the examination was normal.

Chest radiography disclosed clear lung fields, with the cardiac silhouette at the upper limits of normal. The aortic arch demonstrated slight tortuosity, but notching of the ribs was absent.

The echocardiogram disclosed mild dilatation of the left ventricle (diastolic diameter, 5.5 cm) and left atrium (4 cm). Interventricular septum was mildly thickened (1.3 cm), and the left ventricle contracted normally. The aortic valve was bicuspid in configuration and displayed mild regurgitation. The ascending aorta was mildly dilated; however, Doppler echocardiographic interrogation of the descending aorta was technically impossible, thus precluding detection of an area of stenosis with increased flow velocity.

Because of the strong suspicion of coarctation of the aorta, cardiac catheterization with aortography was performed. Initial attempts using the right femoral approach were unsuccessful because of marked arterial tortuosity beyond the coarctation; therefore, the right radial artery was used for access. This disclosed coarctation of the aorta located just distal to the left subclavian artery (Fig 2). This was associated with a 35-mm Hg gradient on pullback. Intra-cardiac study disclosed normal systolic function of the left ventricle, normal coronary arteries, and the presence of mild aortic regurgitation.

QUESTIONS FOR CONSULTANTS

1. What treatment would you recommend for this man?
2. What are the pros and cons for surgical correction vs stent implantation to correct this defect?
3. If mechanical intervention corrected the co-

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arced aortic segment, what is the likelihood that his hypertension would be cured or ameliorated?

4. If this patient were to refuse any type of mechanical intervention in favor of attempts at more effective pharmacological control of the BP, what would be the likely long-term risk and outcome of this strategy?

**Comments by Consultants**

*Chetan Varma, MD, and Peter McLaughlin, MD*

**What treatment would you recommend for this man?**

All patients with significant coarctation, including those with proximal systemic hypertension, whether symptomatic or asymptomatic, warrant intervention.¹ A coarctation is considered significant if the gradient is > 20 mm Hg between arm and leg by BP cuff with or without proximal systemic hypertension, or in the presence of upper-extremity hypertension accompanied by echocardiographic or angiographic evidence of aortic obstruction.² This patient has increasingly difficult control of BP requiring multiple drug therapy; therefore, an interventional or surgical approach to therapy is warranted. The options are surgical repair or percutaneous intervention, and we would recommend the latter.

**What are the pros and cons for surgical correction vs stent implantation to correct this defect?**

For aortic coarctation, intervention may be surgical or percutaneous. Surgical repair remains the “gold standard” against which newer therapies must be compared.

Primary operative repair of coarctation of the aorta in adults consists of either end-to-end anastomosis (usually the preferred method for initial repair), interposition graft, patch aortoplasty, or arch augmentation. In this case, because of the length of the narrowing, an end-to-end tubular graft is a safer long-term solution than a patch aortoplasty. The risk of aneurysm in the wall opposite the patch is very high 15 to 20 years after the operation. Aneurysms with end-to-end grafts do occur, but rarely. The mortality associated with surgical repair is approximately 1%. The incidence of complications is approximately 2%. The most significant morbidity from surgical repair of coarctation is spinal cord ischemia that results in postoperative motor impairment. Patch repair has resulted in an appreciable increase in recoarctation and development of false aneurysms compared with end-to-end anastomosis. The rate of recoarctation after a patch repair may be as high as 30% and aneurysm formation up to 35% with long-term follow-up.³ Other surgical risks are recurrent laryngeal nerve palsy, phrenic nerve injury, and...
rebound hypertension in the early postoperative phase. Collateral circulation increases with age. Although collateral vessels provide perfusion to the lower extremities and to the spinal cord during cross clamping and repair, the vessels are fragile and a hazard for blood loss requiring care during the initial incision and mobilization of the aortic segment. The operation is usually followed by an approximate 1-week hospital stay, and postoperative rehabilitation is necessary. The presence of a painful lateral thoracotomy and large scar may be an issue.

Balloon angioplasty with stenting can be as effective as surgical repair in relieving stenoses, with similar mortality risk. Stenting prevents elastic recoil and may also reduce the incidence of aneurysm formation by limiting the amount of dilatation required and thus reducing the degree of traumatic injury to the aortic wall.

In this case, the marked tortuosity may be a technical problem because the operator was unable to pass a wire across the lesion from the femoral route. However, once a wire is across from the radial approach, it is possible to snare this and use it to pull a wire up from below.

Early results after stent implantation for both native and recurrent coarctation are good. In many cases, there is near-complete resolution of the coarctation gradient. A review of nine published series with a combined total of 225 patients showed a reduction of gradient with a mean residual gradient of 5 mm Hg. Complications are unusual, and include stent migration and vascular complications at the site of arterial cannulation. Aneurysm formation is uncommon (<5%), although detailed follow-up is often incomplete in the published series and follow-up length relatively short compared to surgical series. A common procedural hazard is femoral artery injury.

Intervention has a similar mortality risk to surgery and several advantages over surgery: the minimal risk of spinal cord injury, probable next-day discharge, no scar, possibly less aneurysm formation; therefore, we would recommend catheter intervention for this patient. If technically this approach were not possible, then surgery would be indicated.

If mechanical intervention corrected the coarcted aortic segment, what is the likelihood that his hypertension would be cured or ameliorated?

The major risk factor for postoperative systemic hypertension is the duration of preoperative coarctation hypertension. The outcome in terms of BP after stent implantation seems to be comparable to that after surgical repair of coarctation in adults. In our own series of 49 patients in Toronto, mean BP was 127/75 mm Hg at a mean follow-up of 20 months after stenting. Seven of the patients continued to have systolic hypertension, and 14 patients were receiving antihypertensive medication. If this patient’s hypertension fails to resolve with intervention, it is generally responsive to medical therapy.

If this patient were to refuse any type of mechanical intervention in favor of attempts at more effective pharmacologic control of the BP, what would be the likely long-term risk and outcome of this strategy?

A significant coarctation causes a proximal pressure load with consequent left ventricular hypertrophy, and ultimately heart failure. The mean survival of patients with untreated coarctation is 35 years, with 75% mortality by 50 years of age. Death in unrepaired patients is usually due to heart failure (30%), aortic rupture/dissection (20%), infective endarteritis/endocarditis (20%), cerebral hemorrhage (10%), concomitant aortic valve disease, or premature coronary artery disease. Hypertension is not a necessary precondition for rupture because hemorrhage can occur in normotensive patients long after successful coarctation repair.

The most common associated congenital malformation is a bicuspid aortic valve that imposes inherent risks. In this case, neither aortic stenosis nor significant regurgitation is present. The aortic root may harbor cystic medial necrosis, a bicuspid aortopathy, that rarely extends further than the ascending aorta. The clinical course of a bicuspid valve with coarctation of the aorta is the same as that of an isolated congenitally bicuspid aortic valve.

![Figure 2. Angiogram of the thoracic aorta (left lateral view), demonstrating the area of coarctation (arrow), associated with considerable tortuosity of the adjacent arteries.](http://journal.publications.chestnet.org/pdfaccess.ashx?url=/data/journals/chest/21993/ on 04/28/2017)
Editor’s Comment

As noted previously, there is increasing evidence that the preferred therapy for coarctation of the aorta is balloon dilation. This approach seems to possess a high primary success rate with resolution of the obstruction and long-term durability. There is a low incidence (<2%) of aneurysm formation following balloon dilatation. The additional use of stents seems to be gaining popularity in contemporary interventions. Because of the geometry and angulation of the stenotic segment, however, it was believed that this patient would be best managed by surgical repair rather than through a percutaneous approach. He has, therefore, been scheduled for elective repair in the near future.

References

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