How Should We Treat Coarctation of the Aorta?

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Given decades of experience with coarctation of the aorta, one might expect consensus regarding therapy and certainly regarding agreement on terminology. The advent of balloon dilatation and the study by Shaddy et al.1 prompt a reassessment of what is known about coarctation and awareness of what is not known.

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Coarctation is a narrowing of the aorta located between the first brachiocephalic vessel and the aortic bifurcation. When isolated coarctation is associated with congestive heart failure, it is considered clinically important, but what is “important” when the child is asymptomatic? Shaddy et al.1 use as an entrance criterion “hemodynamically significant native coarctation of the aorta” without giving parameters for “significant.” Is it an elevated right arm blood pressure at rest? A resting arm-leg gradient of 20 mm Hg, or 30 mm Hg? Is it an exercise-induced gradient of 52 mm Hg without gradient at rest? Not considered in the study by Shaddy et al, exercise effect in coarctation further complicates the issue because of limited data, especially simultaneous arm-to-leg measurements.10,11

Surgical repair has been standard therapy for coarctation since 1945.12 Many now recommend balloon dilatation for previously operated coarctation3,4,13,14; some extend this preference to the unoperated or “native” coarctation.5,15,16 Shaddy and colleagues performed a randomized, prospective study on 36 children with unoperated isolated coarctation (ages 3–10 years), comparing surgery (resection and end-to-end anastomosis in 13 of 16) with balloon dilatation (20 patients). There were no deaths. By the authors’ assessment, complication rates and types were considered similar (to be discussed later). Residual stenosis was present in 25% of patients after balloon dilatation and 6% after surgery.

Shaddy et al found a higher incidence of pressure gradient after balloon dilatation than has been reported by other centers after either surgery10,11,17 or balloon dilatation.5,18,19 Residual obstruction can result from stenosis incompletely resolved by the procedure and/or stenosis not addressed by the procedure, usually proximal hypoplasia of the transverse aortic arch.

The therapeutic effect of balloon dilatation is a transmural tear of the vessel wall through the intima and media, depending—in an unoperated patient—on the adventitia to maintain vascular integrity. In the previously operated patient, scar tissue and adhesions can support the vessel during and after balloon dilatation. Thus, balloon dilatation is always a balance between the desire to dilate widely for optimal effect and the fear that too much dilatation will cause vascular disruption. The safe therapeutic range for balloon size in coarctation is narrow. Shaddy et al chose balloons equal in size to the aorta at the left subclavian artery; this is the standard approach.5,15,18,19 However, larger balloons have been used with safety and success.4,20

A hypoplastic aorta proximal to the left subclavian artery is not amenable to balloon dilatation and may be a difficult technical problem at surgery. However, one might ask whether this always requires treatment. There are both physiological and anatomic data showing that proximal narrowing can widen by growth if adequate relief of obstruction is accomplished downstream. It seems likely that the resultant increase in flow transmits better lateral wall stress, which in turn promotes growth of a structurally normal but hypoplastic transverse aorta.

Most children with coarctation are identified in infancy are in a clinical condition that mandates intervention; the study by Shaddy et al was performed in older children (3–10 years). Recommendations for treatment of older children with this condition cannot be simply extended to neonates and infants. Balloon dilatation may be less effective with the more severe, long-segment narrowing of a critically ill infant. The range of balloon size between adequate effect and aortic rupture is narrower in infants, and the femoral vessels are at greater risk in the smaller subject.

Sometimes, an infant or a young child with isolated coarctation does not manifest congestive heart failure. In this group, Shaddy et al say that “many authors continue to advocate elective surgical treatment of coarctation of the aorta after 2 years of age.” We do not agree with this approach for two reasons. First, in an editorial, Waldhausen et al related two cardiovascular catastrophes that occurred spontaneously in “healthy” children with coarctation who were waiting to reach a target age before surgery. Second, the first year of life is known to be critical in the formation of the cerebral blood vessels. We consider, as a working hypothesis, that right arm—and therefore intracranial—hypertension in an infant with unoperated coarctation may induce abnormal development of the cerebrovasculature, resulting in the long known association of berry aneurysm and premature cerebrovascular disease in
adults for coarctation. Therefore, we recommend therapy for coarctation (beyond the first month of life) for a resting upper compartment systolic pressure of >140 mm Hg even without other signs or symptoms.

Complications of balloon dilatation include aortic rupture with demise, ileofemoral occlusion, need for transfusion after balloon dilatation, and aneurysm of the aorta. Surgical risks include mortality, complications of thoracotomy, hemorrhage, aortic aneurysm, and paraplegia.25 One patient in the Shaddy et al surgical group experienced postoperative paraparesis. Spinal cord injury is likely to be a summation effect—the combination of degree of ischemia (e.g., volume of collateral flow, patient temperature) and duration of ischemia (e.g., time of cross-clamping). In surgery, ischemic time (aortic cross-clamping) is measured in minutes; with balloon dilatation, the inflation time is measured in seconds. In addition, balloon dilatation should not interfere with collateral flow. Therefore, balloon dilatation is attractive in the previously operated patient as well as the older unoperated patient with a moderate coarctation who may not have developed collateral vessels. Concerns with regard to paraplegia probably do not apply to the neonate or infant.

To most physicians, “aneurysm” means a weakness in a wall, but this is not correct use. According to Dorland’s Illustrated Medical Dictionary, “aneurysm” comes from the Greek aneurysma, which means widening and is defined as “a sac formed by the dilatation of the wall of an artery, a vein or the heart.” It is ironic that the word that describes the therapeutic effect of balloon dilatation is considered a major complication of that very procedure. There is neither an accepted definition nor understanding of the clinical importance of aneurysm after balloon dilatation. There are no reports of spontaneous rupture late after balloon dilatation, and prior balloon dilatation has not been considered27 a contraindication to subsequent surgical repair of coarctation. In one series,28 aneurysmal dilatation in the area of dilatation was considered an indication for surgical resection, but this view is not generally accepted. During pregnancy, there is increased risk of aortic rupture29,30 in unrelieved coarctation; whether this applies to balloon dilated obstruction—reduced or residual—is unknown but should encourage special vigilance in women of child-bearing age. We believe that the area of intervention should be periodically imaged, but without evidence for clinical importance of “overwidening” of the aorta caused by balloon dilatation, this should not detract from its therapeutic attractiveness.

The inclusion of financial data in clinical studies helps develop awareness of the fiscal implications of medical care. However, in our view, such information should not be used by health plans or government agencies to dictate medical care. The bottom line in medical decisions must remain the best interests of the patient.

What can we conclude about treatment for coarctation? First, one must first obtain a sense of perspective. Fifty years ago, there was no definitive therapy. Twenty years ago, Kilman et al31 hailed our reduction in mortality from 65% to 35%. In 1993, we discuss how to relieve the obstruction with no mortality and without opening the chest! Second, balloon dilatation can be done safely, often with good initial hemodynamic effect, on older children with unoperated coarctation (as well as those with previous surgery). Third, residual arm-leg gradient at rest seems more common after balloon dilatation than after surgery. Fourth, balloon dilatation is preferable when risk for paraplegia is considered high. Fifth, the optimal approach remains undetermined, especially in neonates and small children. At present, an eclectic attitude seems best.

Like the last editorial on this subject from our institution,14 this one ends with the recommendation that the American Academy of Pediatrics develop an advocacy position stating that balloon dilatation is an acceptable form of therapy for both previously operated and unoperated coarctation. Although many of these authors included (e.g., these authors) are not convinced that balloon dilatation is the best therapy for coarctation, especially in neonates and infants, it is clearly an acceptable mode of treatment in older children.

References

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