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Commentary: Diffuse supravalvar aortic stenosis: Is the enemy of good always better?

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CENTRAL MESSAGE

Surgical decision-making in diffuse supravalvar aortic stenosis is discussed.

Supravalvar aortic stenosis (SVAS) is a rare lesion associated with elastin arteriopathies that may either be familial, as in the case of Williams-Beuren syndrome, or sporadic. The classic presentation is a discrete, hourglass-appearing narrowing at the sinotubular junction. A less common, diffuse variant is observed in 16% to 30% of larger series and can have varying involvement of the ascending aorta, the aortic arch, and, in rare cases, the descending aorta.^{1,2} Occasionally, discrete arch vessel stenoses may be present. In this issue of the *Journal*, Katahira and colleagues³ report the surgical management of an adult with Williams-Beuren syndrome presenting with symptomatic severe aortic valve stenosis who had previously undergone surgical repair for localized SVAS in childhood. Preoperative workup identified diffuse SVAS involving the entire ascending aorta, with a separate proximal stenosis of the brachiocephalic artery.

Although combined aortic valve replacement and ascending aorta and hemiarch replacement is certainly not a novel surgical approach, the late presentation of progressive diffuse ascending aortic stenosis following childhood repair of discrete SVAS is highly unusual. More likely, this patient had diffuse supravalvar aortic stenosis, which manifested most severely at the sinotubular junction. Unfortunately, no information is provided in the case report that

comments on the degree of diffuse aortic involvement at the initial operation.

Importantly, this clinical vignette raises the following question for the surgeon: When should diffuse narrowing of the ascending aorta and/or aortic arch be addressed at the time of primary surgery for sinotubular SVAS? The obvious answer is: it depends. The degree of diffuse narrowing and extent of aortic pathology will alter the risk/benefit calculus in surgical decision-making. Hickey and colleagues⁴ found all surgical reinterventions in their series were for recurrent gradients in the distal ascending aorta or beyond. However, it should be noted that several studies have demonstrated increased operative mortality risk with extensive aortic repair.^{1,5} The most comprehensive multicenter study of SVAS included 76 patients with diffuse-type SVAS and found that the univariate risk factors for mortality were diffuse type and age <12 months.⁶ Nonetheless, most surgeons would agree that the hemodynamic burden of moderate aortic obstruction on the left ventricle should be relieved by patch augmentation or graft replacement. Aggressively addressing significant diffuse supravalvar stenosis at the initial surgical repair will result in a sustained improvement in hemodynamics and reduce the need for future reoperation.

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