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Commentary: An opportunity to better characterize coronary ostial stenosis in patients with Williams syndrome and other elastin arteriopathies

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A group from Stanford has analyzed outcomes of patients with Williams syndrome and other elastin arteriopathies who underwent relief of coronary ostial stenosis along with relief of supra-aortic and pulmonary stenosis.² The group has also published their results with relief of branch pulmonary artery stenosis in this difficult patient population.³ As we understand more about the pathophysiology of vascular obstruction related to elastin gene deletion at 7q11.23,⁴ the ultimate treatment for these patients would be potentially related to gene therapy that could enhance elastin formation and prevent smooth muscle proliferation. These patients have debilitating vascular obstructions that result in ventricular hypertrophy, dysfunction, and coronary insufficiency. Catheter-based therapies have not proven to be very durable, and hence surgical relief of obstruction has been the gold standard. There are multiple phenotypes with variable areas of obstruction, including the supra-aortic region of the aortic and pulmonary valve, branch pulmonary arteries, aortic arch, and coronary arterial origin. The group at Stanford presents their experience with repair of coronary artery ostial lesions in conjunction with relief of supra-aortic aortic and/or pulmonary stenosis. This group included 16 patients of whom 11 had Williams syndrome and 5 had elastin arteriopathy. Of the 16 patients, 7 underwent repair of bilateral ostial lesions and the remainder had repair of either the right or left coronary ostium. The surgical

Postoperative aortic root angiogram demonstrating widely patent left main coronary artery after patch augmentation, from Anderson et al.¹ Used with permission.

CENTRAL MESSAGE

Preoperative testing can help identify coronary artery lesions in patients with Williams syndrome and other elastin arteriopathies.

technique consists of utilizing a contiguous homograft patch to augment the coronary arterial ostium and also to address the supra-aortic stenosis. This technique and results are similar to those reported by a group from Boston Children's Hospital.^{1,5} The indication for coronary artery intervention in this report are manifold: preoperative evidence of myocardial ischemia, preoperative imaging with evidence of obstruction, intraoperative evidence of ostial obstruction, and finally evidence of coronary artery insufficiency after surgical repair of supra-aortic aortic or pulmonary stenosis. There is a clear opportunity here to help this complex patient population. This opportunity exists in the preoperative testing and imaging period; clear imaging of coronary artery origins and either flow measurements in the coronary artery or intracoronary ultrasound to detect any flow limiting lesions. This preoperative testing can be very helpful in operative planning. If there is evidence of greater than mild flow limitation in the coronary arteries, these patients may benefit from intervention on the coronary artery because of significant ventricular hypertrophy present in the vast majority of these patients. As clinicians, it is imperative that we pay close attention to coronary artery ostia and proximal coronary arteries in these complex patients so that these lesions can be accurately addressed at time of surgery. As the report

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from Stanford indicates, relief of coronary artery ostial lesions is something that can be done with excellent outcomes.

References

1. Anderson ND, Borisuk MJ, Hoganson DM, Rathod RH, Baird CW. Three-patch aortic root reconstruction with extended left main coronary artery patch augmentation in neonates and infants. *Semin Thorac Cardiovasc Surg.* 2019;31:99-101.
2. Mainwaring RD, Collins RT, Patrick WL, Martin E, MacMillen KL, Hanley FL. Surgical repair of coronary artery ostial stenosis in patients with Williams and elastin arteriopathy syndromes. *J Thorac Cardiovasc Surg.* 2021;162:212-9.
3. Martin E, Mainwaring RD, Collins RT II, MacMillen KL, Hanley FL. Surgical repair of peripheral pulmonary artery stenosis in patients without Williams or Alagille syndromes. *Semin Thorac Cardiovasc Surg.* January 17, 2020 [Epub ahead of print].
4. Pober BR, Johnson M, Urban Z. Mechanisms and treatment of cardiovascular disease in Williams-Beuren syndrome. *J Clin Invest.* 2008;118:1606-15.
5. Wu F, Mondal A, del Nido PJ, Gauvreau K, Emani SM, Baird CW, et al. Long-term surgical prognosis of primary supravalvular aortic stenosis repair. *Ann Thorac Surg.* 2019;108:1202-9.