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Commentary: You will see them again—sooner or later

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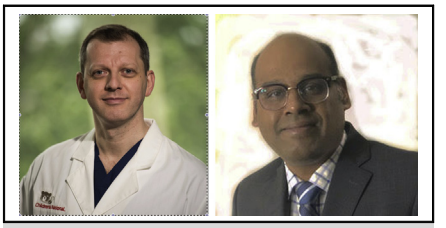

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CENTRAL MESSAGE

Aortic pathology can recur late after initial repair in Williams–Beuren syndrome. Life-long follow-up of these patients is necessary, with a potential for redo surgical repair.

Supravalvar aortic stenosis (SAS) is the most significant cardiovascular manifestation of Williams–Beuren syndrome.[1] Clinically significant SAS has a wide range of phenotypic manifestations.[2] The disease process affecting the aorta is a spectrum ranging from discrete localized thickening of the sinotubular junction to a more extensive involvement of the coronary ostia, ascending aorta, the aortic arch and its branches, the descending aorta, or rarely the renal arteries or intracranial arteries.[1] Discrete and milder forms of SAS can be managed with any of the well-established surgical techniques with good outcomes.[3] Diffuse forms of the disease present a challenging surgical entity. Mild SAS can improve with time or even resolve in a subset of children (about 13%).[4] However, moderate or more severe forms require surgical intervention for hemodynamically significant SAS. An estimated 37.5%, 48%, and 65% of these patients require intervention at 5, 10, and 20 years, respectively.[5]

In this issue of the *Journal*, Katahira and colleagues[6] report the surgical management of complex recurrent SAS with involvement of the ascending aorta and the brachiocephalic artery, with coexisting aortic valve disease. Their patient underwent Doty's procedure (pantaloon patch 2 sinus repair) at the age of 6 years and was regularly followed up. At the age of 41 years, he was diagnosed with severe aortic valve stenosis and recurrent SAS extending into the proximal aortic arch and the origin of the brachiocephalic artery. The patient was successfully treated with a mechanical aortic valve replacement and ascending aortic replacement with hemi-arch augmentation to include enlargement of the stenosed origin of the innominate artery with an excellent outcome.

The nicely presented case highlights some critical aspects of the modified history of SAS. The operative mortality for primary repair of discrete SAS is less than 1%, with 5-year survival approaching 96%.[3] However, the freedom from reintervention for left-sided obstructive lesions is 79% at 5 years and 70% at 10 years. This case report further illustrates that recurrences can occur late after initial intervention and can assume a diffuse form. Therefore, patients with repaired SAS should be on close and probably life-long follow-up to detect recurrences.

The surgical strategy described by the authors appears to be easily reproducible and follows the basic general principles described to protect the end-organs during complex aortic arch surgery. The conduct of cardiopulmonary bypass in these situations is critical to achieve a satisfactory result. Deep hypothermic circulatory arrest, selective cerebral perfusion, and whole-body perfusion are some of the strategies that have been extensively used and studied. Each one of them has their own advantages and disadvantages, and the choice of strategy depends on the preference and expertise of the particular center. The favorable outcome achieved in this patient is a testament not only to the surgical skills but also to the organization and planning of the entire team involved in the operation. We thank the authors for their contribution.

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