A 15-year-old boy with a history of d-transposition of the great arteries, status postarterial switch procedure with LeCompte maneuver, and severe neoaoetric regurgitation underwent neoaoetric valve replacement with an Edwards Magna 23-mm pericardial bioprosthesis, along with reconstruction of the ascending aorta with a bovine graft. At routine follow-up 8 months after surgery, he was found to have a harsh systolic ejection murmur over his midprecordium, with a prominent palpable bruit over both carotid arteries. Echocardiography demonstrated a Doppler mean gradient >50 mm Hg through the ascending aorta, but 2-dimensional imaging was unable to define the anatomic lesion because of poor acoustic windows. Left ventricular systolic function was mildly decreased. Subsequent cardiac magnetic resonance imaging documented a long stenosis of the ascending aorta with a minimum diameter of 11 mm.

Given the patient history of multiple and recent cardiac operations, both surgical and transcatheter management options were discussed in a multidisciplinary setting. After presenting these options to the patient and his family, they elected to proceed with endovascular stent placement.

At the time of catheterization, the peak systolic gradient from the left ventricle to the distal ascending aortic arch was 50 mm Hg. Three-dimensional rotational angiography (3DRA) using a DynaCT system (Siemens, Munich, Germany) demonstrated aortic root dilatation with severe narrowing of the midascending aorta to 9 mm (Figure 1 and Movie I in the online-only Data Supplement). Three-dimensional surface renderings of the ascending aorta allowed for sizing of the stent, identification of anatomic landmarks (aortic valve, orifices of the coronary artery, and right brachiocephalic vessel), and establishment of optimal x-ray projection angles (32° left anterior oblique/15° cranial and 97° left anterior oblique/19° cranial) for stent positioning and placement. A 2612 Intrastent LD Max (ev3-Endovascular, Plymouth, MN) open-cell stent on a 16-mm × 4-cm NuMed balloon-in-balloon system (NuMed, Hopkinton, NY) was positioned across the stenosis using 3DRA overlay guidance. Rapid right ventricular pacing at 180 beats per minute was performed during stent placement. The superior edge of the stent was flared using a 22-mm × 4-cm Tyshak II balloon (NuMed). After stent placement, the gradient between the aortic root and distal ascending aorta was <5 mm Hg. Postimplant 3DRA (Figure 2 and Movie II in the online-only Data Supplement) demonstrated an improved caliber of the ascending aorta to 15.5 mm and no evidence of aneurysm, dissection, contrast extravasation, or disruption of surrounding structures. The patient was discharged from the hospital the following day after an uneventful overnight hospital stay.

Contrast parameters for 3DRA injection were based off of a patient weight of 75 kg and height of 172 cm. Both pre-intervention and postintervention 3DRA series were obtained using a contrast ratio of 50:50 Omnipaque:saline dilution. A total dilute contrast volume of 75 mL was injected via a 5F pigtail catheter positioned in the left ventricle at 15 mL/s with a 1-second x-ray delay and 5-second x-ray rotation duration. Rapid right ventricular pacing at 180 beats per minute was performed during rotational angiography.

Radiation dose metrics for the procedure include a procedure cumulative air kinetic energy released per unit mass of 977 mGy. 3DRA accounted for 151 and 143 mGy for the first and second series, respectively, resulting in ≈30% of total radiation dose for the case.

Discussion
To date, endovascular stent placement has been described in 3 patients with nonsyndromic congenital supravalvar aortic stenosis (svAS). Two patients underwent successful stent placement for diffuse svAS. The third case was ultimately successful but demonstrated an acute complication during the procedure. Stent placement was performed using simultaneous left ventricle and ascending aorta angiography for stent guidance with pharmacological (adenosine) induction of transient cardiac arrest to ensure proper stent placement. Despite these precautions, the stent trapped the leaflets of the aortic valve. A novel technique of temporarily pulling back the proximal stent without dislodging it was used and successful but was a heroic maneuver for a potentially catastrophic
complication.2 Additional attempts at endovascular stenting have not been reported.

To our knowledge, this is the first successful treatment of acquired svAS using endovascular stent placement. Multiple characteristics made this patient an optimal candidate for the consideration of endovascular stent placement. First, with a bioprosthetic valve, the coronary ostia and valve cusps were reasonably protected between the struts of the valve, and the radiopaque structure of the valve apparatus provided an easily identifiable landmark for stent positioning. Second, being acquired svAS, the stenotic region was located more superiorly than is typical of congenital svAS. Congenital svAS is more likely to occur at the sinotubular junction, much closer to and sometimes directly involving the coronary ostia. Third, the patient will eventually require surgical intervention for replacement of his bioprosthetic valve and aortic root. Because of the technical complexity of such a surgical procedure in the context of anteriorly translocated pulmonary arteries (LeCompte maneuver) and to avoid another sternotomy, a transcatheter approach was relatively appealing. These considerations made endovascular stenting an attractive alternative to surgical repair in the case of this patient.

This case suggests that endovascular stenting may have a role in acquired svAS in carefully selected cases. The use of rotational angiography in this case allowed for optimal stent selection and precise placement. Immediately on completion of the angiogram, a 3-dimensional reconstruction is rendered, allowing for selection of stent length and width. Imaging planes can be selected on the 3-dimensional workstation to ensure a nonforeshortened length of the stenotic region with visibility of branch arteries and other important nearby structures. Once the desired working angle is noted, the C-arm can be adjusted accordingly. Overlay of the previously acquired 3DRA reconstruction during live, real-time fluoroscopy can provide valuable guidance for stent position, thus avoiding disruption of the bioprosthetic valve, coronary orifices, and brachiocephalic arteries.

Disclosures

None.

References


Figure 1. Baseline 3-dimensional rotational angiogram using a DynaCT system at a projection angle of 32° left anterior oblique and 15° cranial demonstrates severe stenosis of the midascending aorta and dilation of the aortic root.

Figure 2. Three-dimensional rotational angiogram after placement of a 2612 Intrastent LD Max (ev3-Endovascular) open-cell stent demonstrates improvement of the acquired supravalvar aortic stenosis to a diameter of 15.5 mm.
Three-Dimensional Rotational Angiography-Guided Stent Placement for Treatment of Acquired Supravalvar Aortic Stenosis
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