Surgical management of recurrent coarctation from diffuse elastin arteriopathy

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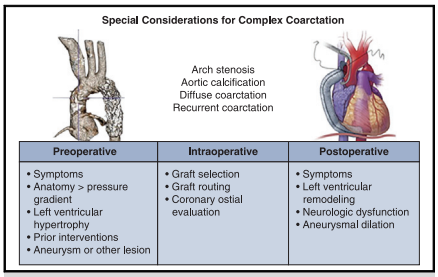


Special considerations for complex coarctation including diffuse or recurrent coarctation

CENTRAL MESSAGE

Ascending-to-descending aortic bypass offers a safe and reliable solution for recurrent complex aortic coarctation. Patch augmentation can be used to address bilateral outflow obstruction.

Video clip is available online.

Elastin arteriopathy (EA) involves decreased elastin in the arterial wall, causing stenosis.[1] Nonsyndromic EA is rare, occurring in only 1 of 20,000 live births,[2] and thus management guidelines are limited.[1,3] Here we describe a patient with congenital EA complicated with aortic, pulmonary artery (PA), and coronary artery stenosis despite multiple surgical repairs and stenting.

The patient provided written consent and permission for this publication; Institutional Review Board approval was not required.

CASE DESCRIPTION

Our patient was an 18-year-old female with diffuse non-Williams EA, with worsening exertional dyspnea and low exercise tolerance on stress testing. She had a history of supravalvular aortic stenosis and coronary artery stenosis, for which she had undergone 3 previous cardiac surgeries, including patch augmentation of the ascending and transverse aortic arch as well as branch pulmonary arteries, coronary artery osteoplasty, and multiple stents placed in the aorta and coronary arteries. At the time of presentation, she had a resting blood pressure of 135/59 mm Hg. Her cardiac stress test demonstrated an exaggerated early blood pressure response to 233/75 mm Hg.

Echocardiography showed supravalvular aortic stenosis at the sinotubular junction (1.5 cm) and moderate supravalvular PA stenosis (peak/mean gradient, 52/30 mm Hg). There was mild right ventricular hypertrophy and moderate left ventricular (LV) hypertrophy with preserved systolic function. The peak/mean gradient across the supravalvular aortic area was 49/15 mm Hg, with mild residual obstruction at the distal aortic arch. Mild aortic regurgitation was observed with a posteriorly directed jet through the left ventricular outflow tract of 2.6 m/s. On computed tomography scan, the descending thoracic aorta measured 11 mm, and 2 collaterals were noted arising from the proximal descending thoracic aorta (Figure 1).

The patient and family desired a long-term surgical solution to her recurrent symptoms. We planned an ascending-to-descending graft that included an ascending aortic plasty to offload the ascending and arch gradient and patch augmentation of the right PA to relieve the outflow obstruction (Video 1).

Resternotomy was performed safely. After aortic and bicaval cannulation, the descending aorta and main PA were

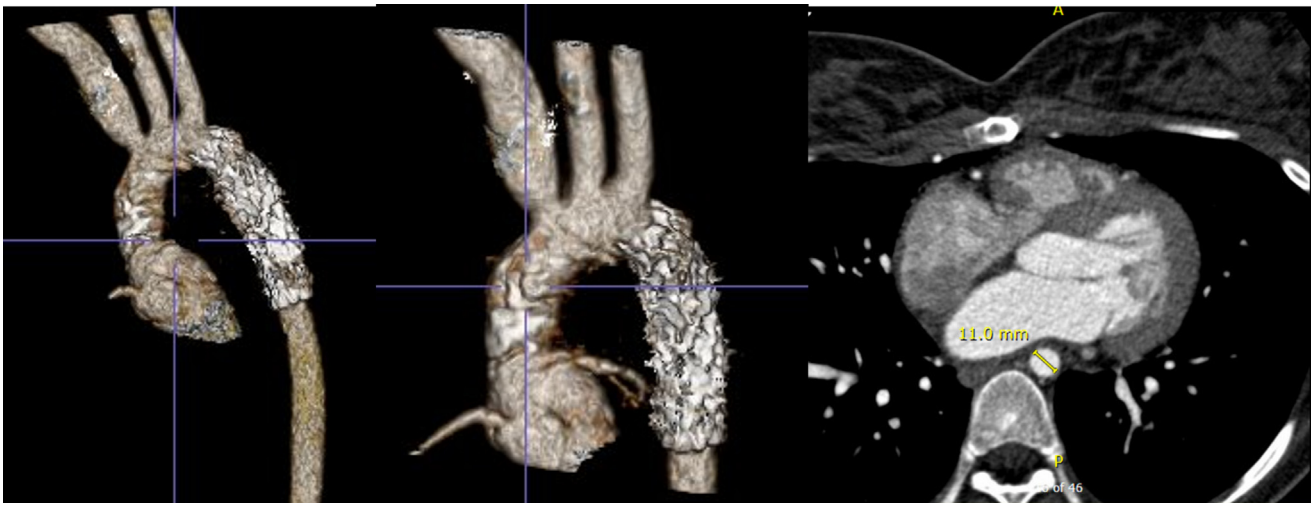


FIGURE 1. Preoperative computed topography scan showing stenosis of the ascending and descending aorta with graft. The descending aorta measured 11.0 mm.

dissected. The posterior pericardium was opened, and the descending thoracic aorta was exposed and dissected for a segment of approximately 8 cm while on bypass (Figure 2, *A*). After application of 2 straight clamps, the distal anastomosis of a 14-mm beveled Dacron tube was performed (Figure 2, *B* and *C*). After testing for leaks, the graft was routed behind the internal vena cava to the right side of the atrium for subsequent insertion into the ascending aorta (Figure 2, *D-F*). An incision was made on the left aspect of the PA and extended into the left PA and main PA sinus (Video 1). A bovine pericardial patch was used to augment the supravalvular region of the main PA (Video 1).

Under cardioplegic arrest, the aortotomy incision was extended into the previously implanted Dacron patch toward the noncoronary sinus. The heel of the ascending graft was anastomosed proximal to the noncoronary sinus and the toe across the supravalvular aortic stenosis, addressing the supra-aortic stenosis. The total pump time was 134 minutes, cross-clamp time was 26 minutes, and descending aorta clamp time was 16 minutes.

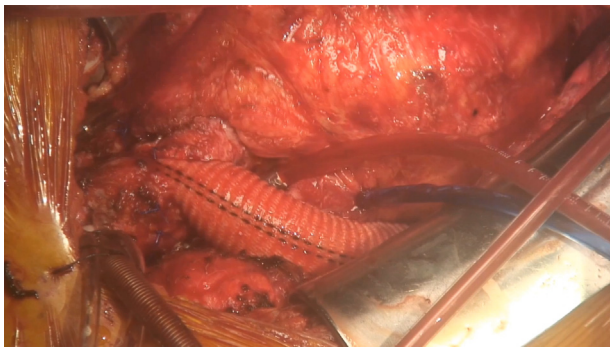
Postoperative echocardiography showed reductions in the peak aortic gradient to 2 mm Hg (0.71 m/s) and in the pulmonary valve gradient to 27.1 mm Hg (2.6 m/s). A computed tomography scan revealed a patent graft with no kinking and no focal PA stenosis (Figure E1). The patient was discharged on postoperative day 8 once her upper and lower extremity gradient resolved.

At a 3-month follow-up, she had returned to normal activity levels, and her blood pressure had normalized without medication (122/77 mm Hg). At a 10-month follow-up, echocardiography showed regression of LV size and wall thickness with good systolic function.

DISCUSSION

"Complex coarctation" refers to recurrent coarctation or cases associated with other cardiac lesions. Repair of complex coarctation is challenging and requires specific considerations. While a hemodynamic gradient (>20 mm Hg) is a standard indicator of coarctation, it can be blunted in patients with diffuse stenosis and proximal flow obstruction. Thus, decision making should incorporate symptoms, anatomic features, and LV hypertrophy. Resolution of LV remodeling, as seen in this case, confirms significant obstruction.

Although transcatheter interventions are typically the first-line approach, patients with EA often experience recurrent coarctation following prior procedures.[4] Data comparing surgical and transcatheter procedures in such patients are limited. The use of semi-rigid stents may be



VIDEO 1. Step by step surgical video demonstrating the technical aspect of the procedure. Video available at: https://www.jtcvs.org/article/S2666-2507(25)00061-6/fulltext.

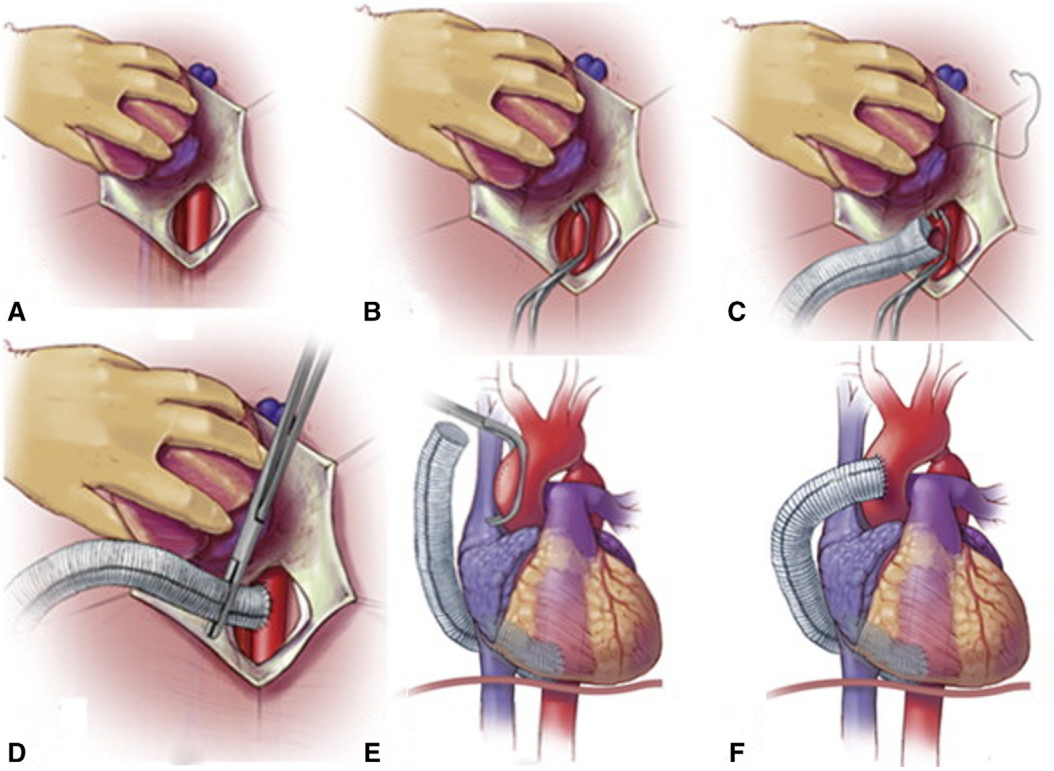


FIGURE 2. Illustrations showing the technique of ascending-descending posterior pericardial aortic bypass. A, The descending thoracic aorta is dissected out and exposed after aortic and bicaval cannulation. B, A distal segment is selected, and a clamp is applied. C, The distal anastomosis of a 14-mm beveled Dacron graft is constructed with continuous 4-0 polypropylene suture. D, Air is removed from the graft. E, The graft is routed behind the inferior vena cava to the right atrium for proximal anastomosis. F, The proximal anastomosis is completed on the lateral aspect of the ascending aorta using a side-biting clamp. Reproduced with permission from Elsevier.[4]

a concern in hypertensive or tissue-disordered aortas, as they may worsen ventricular afterload and vascular properties. In these cases, surgical treatment of recurrent coarctation is safe and associated with a lower rate of reintervention.[5] Ascending-to-descending bypass offers good long-term outcomes in those ineligible for repair.[4,5]

CONCLUSIONS

Managing outflow tract obstruction caused by diffuse arteriopathy is less well-documented and necessitates a tailored approach. Patch augmentation can be an effective method to reduce systemic afterload. For cases of residual aortic coarctation following stenting procedures, an ascending-to-descending bypass offers a safe and reliable solution for recurrent stenosis.

Conflict of Interest Statement

The authors reported no conflicts of interest.

The *Journal* policy requires editors and reviewers to disclose conflicts of interest and to decline handling or reviewing manuscripts for which they may have a conflict of interest. The editors and reviewers of this article have no conflicts of interest.

References

1. Stamm C, Friehs I, Moran AM, et al. Surgery for bilateral outflow tract obstruction in elastin arteriopathy. *J Thorac Cardiovasc Surg*. 2000;120(4):755-763. https://doi.org/10.1067/mtc.2000.107477
2. Merla G, Brunetti-Pierri N, Piccolo P, Micale L, Loviglio MN. Supravalvular aortic stenosis: elastin arteriopathy. *Circ Cardiovasc Genet*. 2012;5(6):692-696. https://doi.org/10.1161/CIRCGENETICS.112.962860
3. Mainwaring RD, Hanley FL. Surgical techniques for repair of peripheral pulmonary artery stenosis. *Semin Thorac Cardiovasc Surg*. 2016;28(2):418-424. https://doi.org/10.1053/j.semtcvs.2016.07.003
4. Said SM, Burkhart HM, Dearani JA, Connolly HM, Schaff HV. Ascending-to-descending aortic bypass: a simple solution to a complex problem. *Ann Thorac Surg*. 2014;97(6):2041-2048. https://doi.org/10.1016/j.athoracsur.2014.02.030
5. Brown JW, Ruzmetov M, Hoyer MH, Rodefeld MD, Turrentine MW. Recurrent coarctation: is surgical repair of recurrent coarctation of the aorta safe and effective? *Ann Thorac Surg*. 2009;88(6):1923-1931. https://doi.org/10.1016/j.athoracsur.2009.07.024

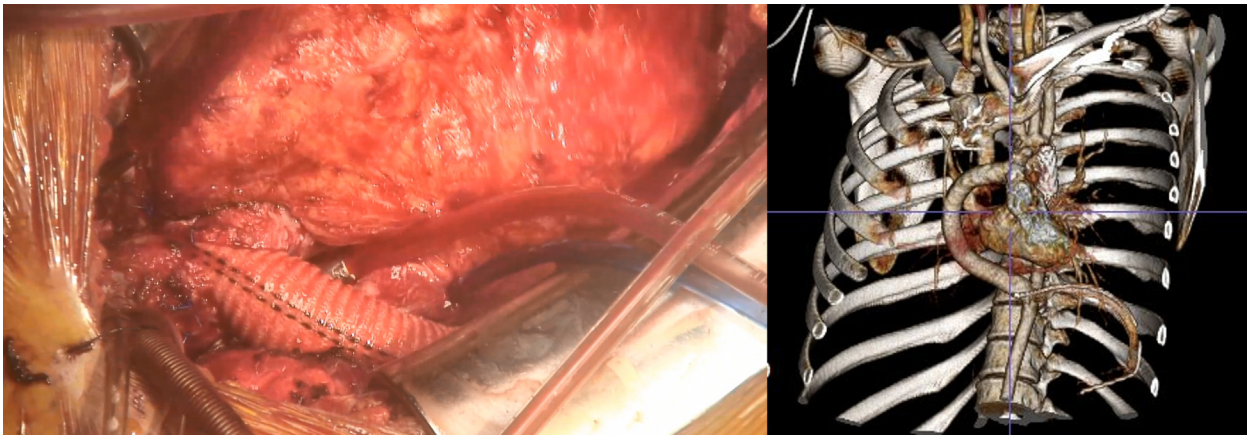


FIGURE E1. Intraoperative image of ascending-descending posterior pericardial aortic bypass and pericardial patch augmentation of the pulmonary artery. Postoperative reconstructed computed tomography scan showing patent bypass graft.