Thoracoabdominal aorta replacement for a patient with Marfan syndrome with poor left ventricular function

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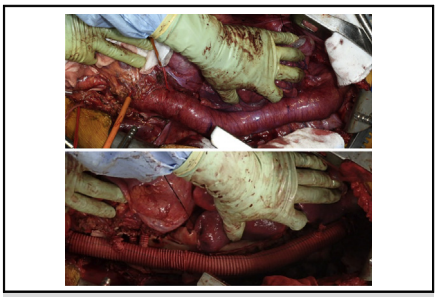


Intraoperative pictures before (*above*) and after (*below*) thoracoabdominal aorta replacement.

CENTRAL MESSAGE

Although extensive aortic surgery for a patient with poor left ventricular function remains challenging, it is feasible if carefully managed, especially for selected cases.

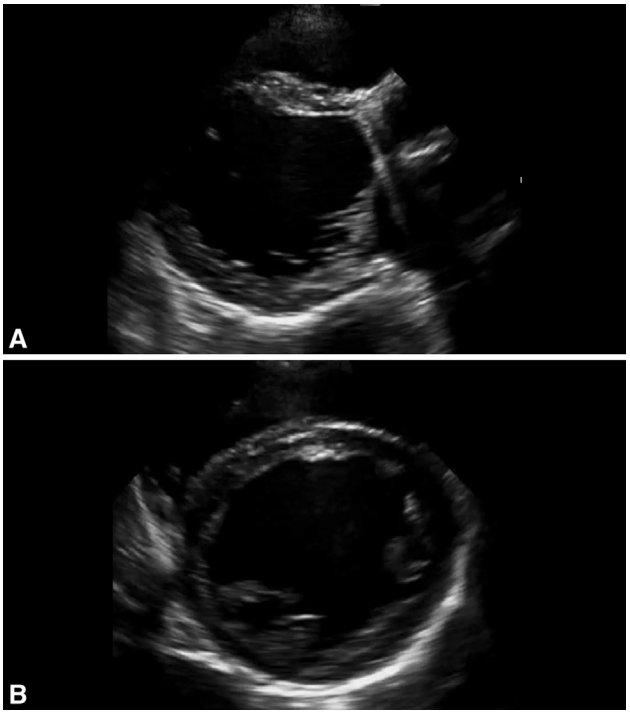
See Commentaries on pages 49 and 51.

Video clip is available online.

Few surgeons advocate extensive aortic surgery with deep hypothermic circulatory arrest (DHCA) for patients with poor left ventricular (LV) function.[1] Here, we describe a successful thoracoabdominal aorta replacement with DHCA, for a patient with significantly reduced LV ejection fraction (LVEF <10%). The patient provided informed consent for the publication.

A 16-year-old male patient with history of Marfan syndrome and left pneumothorax underwent a mechanical Bentall procedure for acute aortic dissection. Postoperative echocardiography demonstrated reduced LV function (LVEF 49%) and computed tomography showed patent false lumen. One year later, he developed dyspnea, LVEF was down to 18%, and the diagnosis of dilated cardiomyopathy was made. Four months later, LVEF became <10% (Video 1). Notably, the diameter of the distal arch increased to 50 mm, and that of the supraceliac aorta increased to 45 mm (Figure 1, *A*, and Video 2). Furthermore, he had a recurrent left pneumothorax with a bulla. Although he was seemingly a transplant candidate, the regulation required us to first repair these aortic lesions to enable listing for transplantation. Additionally, even insurance reimbursement for continuous-flow LV assist device (cf-LVAD) is contingent upon listing for transplant. Therefore, to enable transplant and following long-term survival, he was transferred to us for extensive aortic surgery with significant LV dysfunction.

First, our cardiologist optimized the volume status before the procedure and a right heart catheter study was conducted (Appendix 1). The aorta was exposed via a Stony incision (Figure 2, *top*), and bypass was established by iliac artery perfusion and right atrium (via femoral vein)/pulmonary artery drainage. We placed a LV apical vent and started cooling. At 18°C, we administered potassium chloride into the pump reservoir and put him on DHCA (Appendix 2). During DHCA, the distal arch was opened, an occlusion balloon was placed in the ascending aorta, and crystalloid cardioplegia was administered. Next, proximal anastomosis was performed under retrograde cerebral circulation (Appendix 2), we started side-branch perfusion and rewarming, and iliac perfusion was discontinued. While rewarming, intercostal artery reconstruction was performed, followed by visceral vessel reconstruction (Appendix 3). Distal anastomosis was made at the terminal aorta (Figure 2, *below*). Weaning was smooth under intra-aortic balloon pump (IABP) support. Finally, thoracic surgeons



VIDEO 1. Echocardiography before thoracoabdominal aorta replacement showing extremely poor left ventricular function (a = long axis, b = short axis). Video available at: https://www.jtcvs.org/article/S2666-2507(21)00091-2/fulltext.

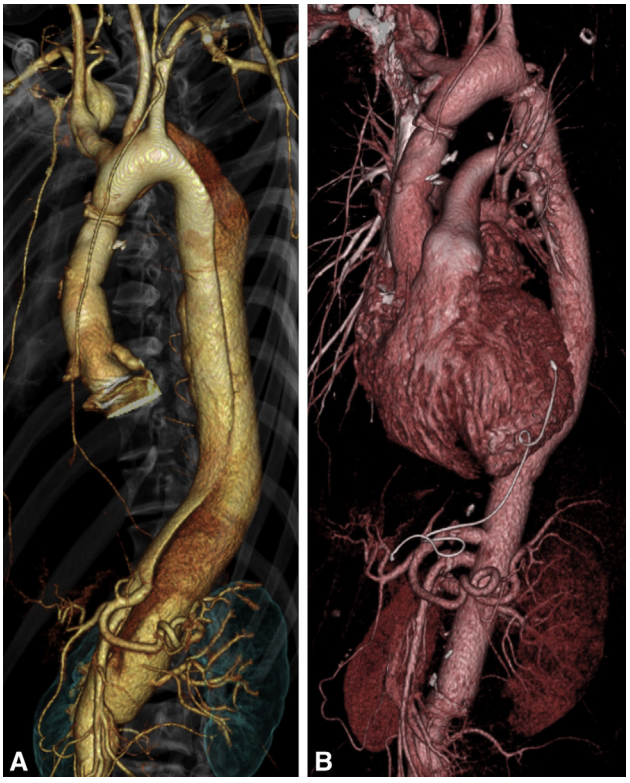
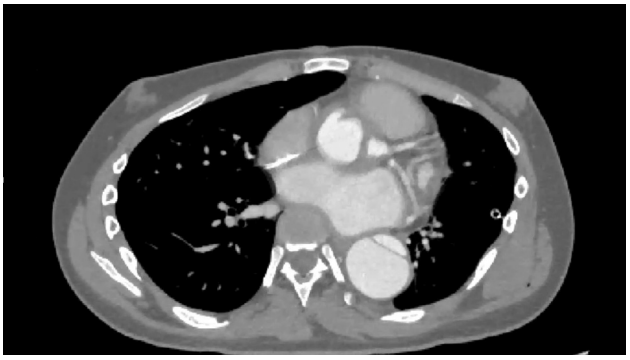


FIGURE 1. Three-dimensional computed tomography. A, Before thoracoabdominal aorta replacement showing Crawford extent II chronic aortic dissection. B, After graft replacement.

conducted a bullectomy. On postoperative day (POD) 2, IABP was removed after extubation. Landiolol was started intraoperatively to control heart rate, which was eventually switched to carvedilol (Appendix 4). On POD 11, echocardiography showed poor LV function as baseline. On POD 14, 3-dimensional computed tomography showed no abnormality (Figure 1, *B*). On POD 30, he was discharged. Three months later, he was cleared for transplant and we implanted a Heartware HVAD (Medtronic, Minneapolis, Minn) with aortic valve replacement (bioprosthetic). He is currently going to college and waiting for transplantation.

Thoracoabdominal aorta replacement with DHCA is an extensive procedure. Even in a center of excellence, its 30-day mortality is 7% to 8% with a 5-year survival of 55%.[2] Particularly, this procedure is challenging for patients with poor LV function.[1] Although a study has shown that decreased LV function is not an independent risk, the authors acknowledge that patients with very low LVEF (<30%) represented only a small minority (0.2%) in their cohort.[3] Most would agree that extremely poor cardiac function is a potential risk for procedures of any kind.

In this context, the situation in Japan is unique. In Western countries, where cf-LVADs are available as destination therapy, conducting isolated aortic procedures without LVAD for patients with significantly reduced LVEF (<10%) is getting rare. In contrast, in Japan, only bridge-to-transplant indication is allowed, implying that listing before cf-LVAD is mandatory. Additionally, the number of donors is limited in Japan, forcing us to create a high threshold for candidacy. Therefore, to enable his candidacy for transplantation, we had to repair our patient's aortic disease, although he could have undergone cf-LVAD in other countries.[4] Another option was implanting an extracorporeal LVAD before the aortic procedure.



VIDEO 2. Computed tomography scan before thoracoabdominal aorta replacement showing chronic aortic dissection. Video available at: https://www.jtcvs.org/article/S2666-2507(21)00091-2/fulltext.

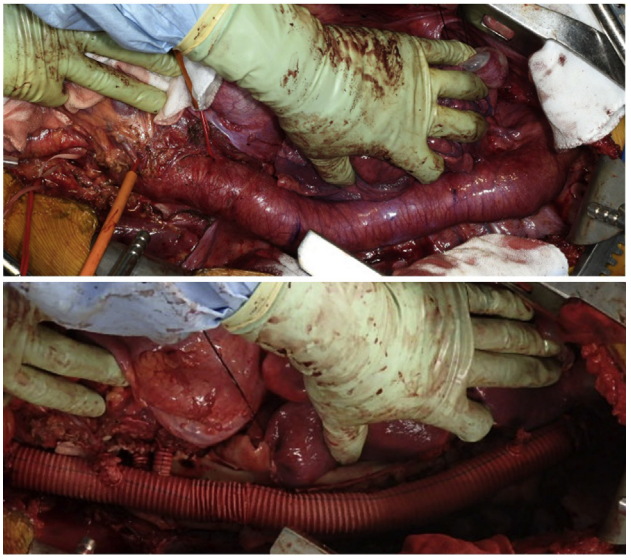


FIGURE 2. Intraoperative pictures before (*above*) and after (*below*) thoracoabdominal aorta replacement.

However, this option might have required some modifications during the procedure to prevent LVAD thrombosis.[5] So, simply performing aortic surgery could be safer.

Well-planned surgical techniques and intensive management are essential for treatment success. In this case, our cardiologists optimized the volume status to avoid decompensation. During the procedure, we paid attention to myocardial protection, cooling, and LV venting. IABP was placed prophylactically before weaning. Intraoperatively, anesthesiologists prepared landiolol to prevent heart failure due to tachycardia. Careful monitoring with daily echocardiography was another key feature. Not only the surgical techniques, but also the thorough perioperative care yielded our successful outcome.

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APPENDIX 1: RIGHT HEART CATHETER INFORMATION BEFORE THE AORTIC PROCEDURE

- Right atrial pressure: 2 mm Hg
- Wedge pressure: 12 mm Hg
- Mean pulmonary artery pressure: 20 mm Hg
- Cardiac index: 1.9 L/min/m^2 (without inotrope)

APPENDIX 2: PUMP TIME INFORMATION AND HOW RETROGRADE CEREBRAL CIRCULATION WAS PERFORMED

- Circulatory arrest time: 20 minutes
- Retrograde cerebral circulation time: 18 minutes
- Total bypass time: 342 minutes
- How retrograde cerebral circulation was performed:

We do not use a superior vena cava cannula for retrograde cerebral circulation when we do deep hypothermic circulatory arrest (DHCA) cases through left thoracotomy. Under DHCA, we clamp the descending aorta and maintain lower body perfusion from femoral arterial cannula. Generally, oxygen saturation of venous-return blood in the right atrium is quite high (mostly 100%). Then, we put a patient in the Trendelenburg position and keep central venous pressure at 20 to 25 mm Hg. By these maneuvers, oxygenated venous blood retrogradely circulates the brain, and de-oxygenated blood eventually comes out from the aortic arch vessels while under DHCA.

APPENDIX 3: INTERCOSTAL ARTERY AND VISCERAL VESSEL RECONSTRUCTIONS

- Intercostal arteries: Th 7, Th 9, and Th 11 in this order.
- Visceral vessels: Right renal, superior mesenteric, celiac, and left renal arteries in this order.

APPENDIX 4: INOTROPIC SUPPORT INFORMATION

The patient returned to the unit under 5 μg/kg/min dobutamine, 0.3 μg/kg/min milrinone, and 0.03 μg/kg/min norepinephrine. Vasopressor was titrated so that we can keep his mean arterial blood pressure between 60 and 80 mm Hg. Milrinone was discontinued on postoperative day 2, and dobutamine was discontinued on postoperative day 6.