

Combined Heart Transplantation and Total Replacement of Thoracic Aorta in Marfan's Syndrome With Recurrent Aortic Dissection: A Case Report

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ABSTRACT

It is extremely rare for a patient to need simultaneous heart transplantation (HTx) and replacement of the thoracic aorta. A 23-year-old woman with Marfan's syndrome underwent Bentall's operation and replacement of the ascending aorta (AsA) due to a type A aortic dissection (AD) in August 2001. In March 2005, she began to experience dyspnea on exertion and was found to have a huge pseudoaneurysm at the aortic root, which had caused dehiscence of the aortic conduit. In July 2009, she suffered acute chest pain followed by hypotension and cold sweating. The computed tomography (CT) scan showed a recurrent dissection with a long intimal tear extending from the arch to the mid-portion of the descending thoracic aorta (DTA). Due to technical difficulties in the repair of the aortic root, she was placed on the HTx waiting list. The next day, she received a donor heart and underwent combined HTx and total replacement of the thoracic aorta. Explantation of the heart improved the exposure of the DTA. With the use of a vascular ring connector (Vasoring), the operation was successfully performed without need for a blood transfusion. The patient was still well at 2 years after the operation. Simultaneous replacement of the heart and the whole segment of the thoracic aorta is technically possible in Marfan patients who are complicated with aortic dissection.

TO TREAT aortic dissection (AD) in patients with Marfan's syndrome is technically demanding. Surgery for recurrent dissection in these patients is especially difficult.

CASE REPORT

In August 2001, a 23-year-old woman with Marfan's syndrome suffered acute chest pain for the first time, undergoing an emergency Bentall's operation for type A AD. During the operation, an intimal tear in the aortic root required replacement of a segment of the ascending aorta (ASA). She remained healthy until March 2005 when she began to develop dyspnea on exertion. The echocardiogram showed a huge pseudoaneurysm arising from the aortic root with dehiscence of the aortic conduit from one third of the annulus. However, the patient was not willing to undergo redo surgery due to the lack of symptoms. It was not until July 2009 when she suddenly developed excruciating pain over the anterior chest and back so that she was admitted to the hospital for further treatment. The computed tomography (CT) scan showed a recurrent dissection with a long tear arising from the aortic arch to the mid, descending thoracic aorta (DTA) (Fig 1). After discussion with her family, we proposed 2 choices: (1) if HTx was available in a short time, we would perform combined HTx and total replacement of the thoracic aorta; or (2) if HTx was not available, we would redo Bentall's operation and replace the whole segment of the thoracic

aorta. She was put on the waiting list for HTx. After 12 hours of waiting we were informed that she had a donor heart.

Surgical Procedure

Before performing the median sternotomy, we anastomosed bilateral carotid arteries using 8-mm vascular grafts. Another 8-mm vascular graft was anastomosed to the left common femoral artery. A cannula was inserted into the right common femoral vein. The patient was then put on cardiopulmonary bypass (CPB) through the right carotid artery, left femoral artery, and right femoral vein. During CPB, we performed the median sternotomy and opened the pericardium. After achieving moderate hypothermia, the systemic circulation was stopped while the brain was perfused with oxygen-saturated blood at a rate of 1200 cc/min.

The heart and AsA were excised and an intraluminal graft inserted into the DTA. The intraluminal graft included a gel-sealed vascular graft and a vascular ring connector (VRC; Vasoring, Sunwei Technology Co, Taipei, Taiwan; Fig 2). Using a Coda

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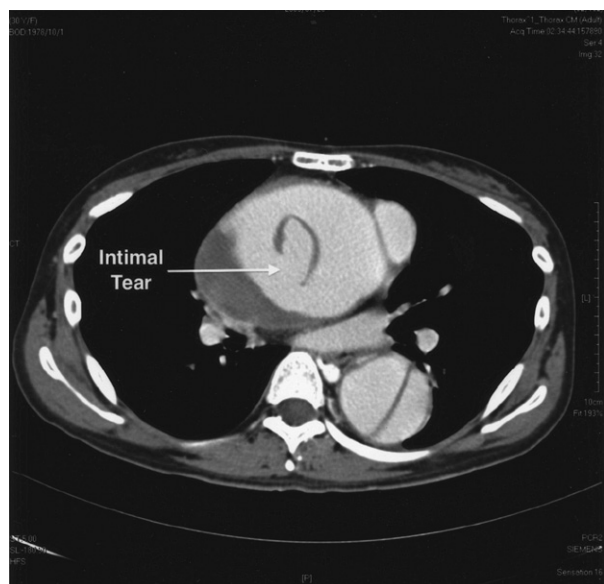


Fig 1. Preoperative CT scan showed aortic dissection with intimal tear at AsA and DTA.

balloon catheter (Cook Medical Inc., Bloomington, Ind, United States) from the femoral artery, the ringed graft was pulled into the DTA through the aortotomy. The Vasoring was further secured with 2 nylon tapes from the outside of the lower DTA. The other end of the intraluminal graft was then anastomosed to the AsA of the donor heart. Finally, the 2 carotid vascular grafts were brought to the mediastinum and anastomosed to the anterior wall of the donor aorta. The procedure was smooth and did not require blood transfusion. The patient recovered but stayed for 2 months in the hospital due to leukocytopenia caused by the immunosuppressive agents. She was well at 2 years after the surgery with an excellent follow-up CT study (Fig 3).

DISCUSSION

Due to technical difficulties, combined HTx and total replacement of the thoracic aorta has not been reported in patients with Marfan's syndrome. It might be considered impossible when the dissection is recurrent and the entry tear involves both the arch and the DTA.

After explantation of the heart, the accessibility to the

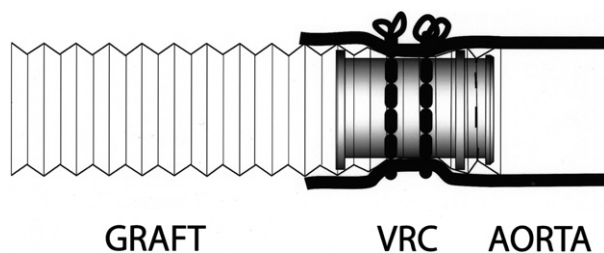


Fig 2. Making intraluminal graft with VRC. The VRC was inserted into the vascular graft and was fixed with stitches. The ringed graft was then inserted into the aorta and was secured with 2 tapes from the outside of the aorta.

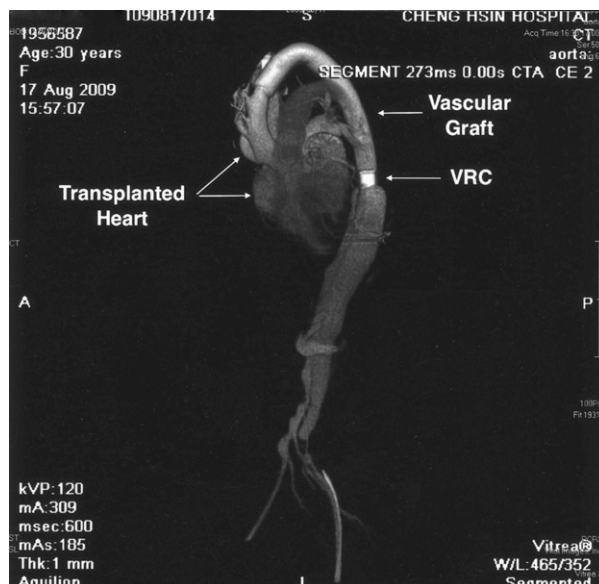


Fig 3. Postoperative CT scan showed total replacement of the thoracic aorta with intraluminal graft.

DTA may improve due to the good exposure. Our experience showed that even though the heart was present, we still could use a balloon catheter to bring the ringed intraluminal graft to the lower DTA to repair the dissection. Good surgical exposure was not necessary due to the anastomosis using an intraluminal graft.

Sutureless intraluminal grafts were introduced to repair AD in the late 1980s and early 1990s. The early results were exciting due to the reduced bleeding compared with conventional suture techniques.¹ Unfortunately, several years after the surgery pseudoaneurysm formation was noted in many patients.^{2,3} We found that the cause of pseudoaneurysm is probably the lack of a furrow on the ring that is crucial for fixation between the aorta and the intraluminal graft, such as cases using the Meadox (Meadox Medical, Oakland, NJ) in prosthesis. To prevent dislodgement, we used a new VRC, i.e., the Vasoring. Our initial results with its use are excellent in surgery for aortic dissection.⁴

In conclusion, simultaneous replacement of the heart and the whole segment of thoracic aorta was technically possible in a Marfan patient complicated with an aortic dissection.

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