

Cardiac Transplantation and Consecutive Minimally Invasive Pectus Excavatum Repair



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Pectus excavatum is a common chest wall deformity with inward deviation of sternum and accompanying ribs. The depression can cause symptomatic cardiac compression, although the cardiopulmonary impact remains controversial. We present 2 cases of cardiac transplantation followed by modified minimally invasive pectus excavatum repair due to the hemodynamic consequences of the pectus deformity.

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Pectus excavatum (PE) is the most common congenital chest wall abnormality.¹ Multiple reports have demonstrated improvement in cardiac function after PE repair.^{2,3} Despite this, there remains a failure to recognize the importance of compression on cardiac function.⁴ We present 2 cases of cardiac transplantation followed by modified minimally invasive pectus repair (MIRPE) due to the hemodynamic consequences of the pectus deformity compressing the transplanted heart.

Case Reports

Patient 1

A 24-year old woman underwent initial cardiac transplantation at 10 years of age for familial hypertrophic cardiomyopathy. The patient developed severe PE during adolescence, and which remained unrepaired (Figure 1A). Fourteen years posttransplant, she developed allograft failure with diastolic dysfunction secondary to antibody-mediated rejection (transplant coronary vasculopathy). She had symptoms of exertional dyspnea and exercise tolerance that were reported to have been present since her teens but worsened significantly over the past year. She was hospitalized for episodes of exertional

syncope and listed for transplantation. A retransplant with bicaval anastomosis was performed; bypass time 232 minutes, graft ischemic time 141 minutes. As her failed graft was not dilated, the anatomic space was extremely limited due to PE deformity. After weaning bypass, multiple attempts over several hours were made to close the sternum. Removal of sternal retractor was not tolerated despite maximal pressor support. The cardiac compression from the PE caused hemodynamic instability with systolic hypotension as low as 30 to 35 mm Hg. The decision was made to repair the PE by modified MIRPE using 2 11.5-inch bars (Figure 1B). The chest was subsequently closed without hemodynamic issues after bars were placed to elevate the sternum. At 19-month follow-up, the patient has no pectus-related issues or sternal instability.

Patient 2

A 34-year old woman presented with nonischemic cardiomyopathy and PE with recurrent episodes of ventricular tachycardia/fibrillation and subsequent cardiovascular collapse requiring emergent extracorporeal membrane oxygenation. The failing native heart had shifted and dilated into the left thorax, as seen on imaging (Figure 2A). She underwent transplant with bicaval anastomosis; bypass time 141 minutes, graft ischemic time 206 minutes. She was successfully weaned off bypass, however the sternum was unable to be closed because of hemodynamic instability from pectus compression on the heart graft in addition to bleeding and coagulopathy. The patient was transferred to intensive care with open sternum for 36 hours. After stabilization and coagulopathy correction, she was returned to the operating room for simultaneous sternal closure and PE repair by modified MIRPE with 2 14-inch bars (Figure 2B). Breast implants were removed simultaneously. At 6-month follow-up, the patient has no pectus-related issues or sternal instability.

Comment

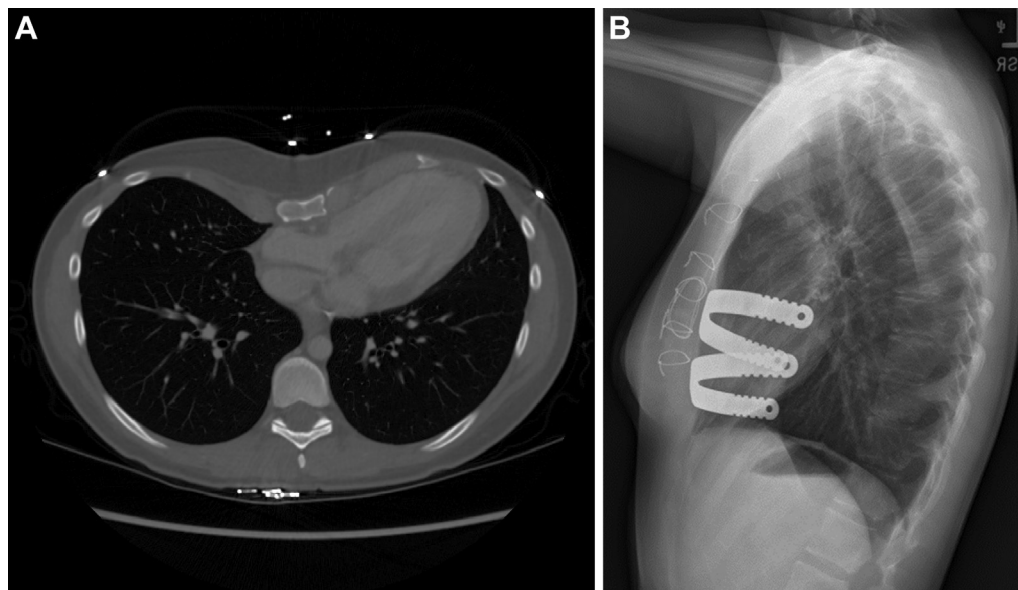
Cardiac compression by PE can cause significant cardiopulmonary impairment, as demonstrated in these cases. Cardiac strain, right heart chamber size, and cardiac output have been shown to be significantly improved with PE repair in native hearts.^{2,3,5} Cardiac graft function was significantly compromised by the PE deformity in these transplant patients and repair was necessary for adequate cardiac function. Neither patient had been followed at our institution prior to presenting with fulminant heart failure. Both had been seen by multiple cardiologists who did not feel the pectus was greater than a cosmetic concern and repair was not recommended. The first patient underwent a transplant

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Figure 1. (A) Computerized tomography imaging: 24-year old woman who presented for retransplantation (past medical history: familial restrictive hypertrophic cardiomyopathy status post orthotopic heart transplantation at 10 years of age) and severe pectus excavatum deformity, Haller index 3.27. (B) Lateral chest roentgenogram posttransplant and repair with 2 bars.



at age 10 years and the pectus developed subsequently in her teen years. She had symptoms including exercise dyspnea and intolerance that progressed over the years but were never attributed to her PE by providers. Pre-operative exertional syncope was a symptom that led to hospitalization and subsequent retransplantation. It is possible that the PE was exacerbating symptoms of the failing graft. It is unknown whether earlier repair of these patients' PE before fulminant heart failure occurred would have delayed failure and decompensation.

Given the degree of cardiac improvement that can be seen with repair, we recommend simultaneous repair of the PE to maximize graft function in patients with significant defects. The right heart is often the more vulnerable of the 2 chambers in the posttransplant setting and PE defects that may cause compression of the graft are planned for repair at the time of transplant. At

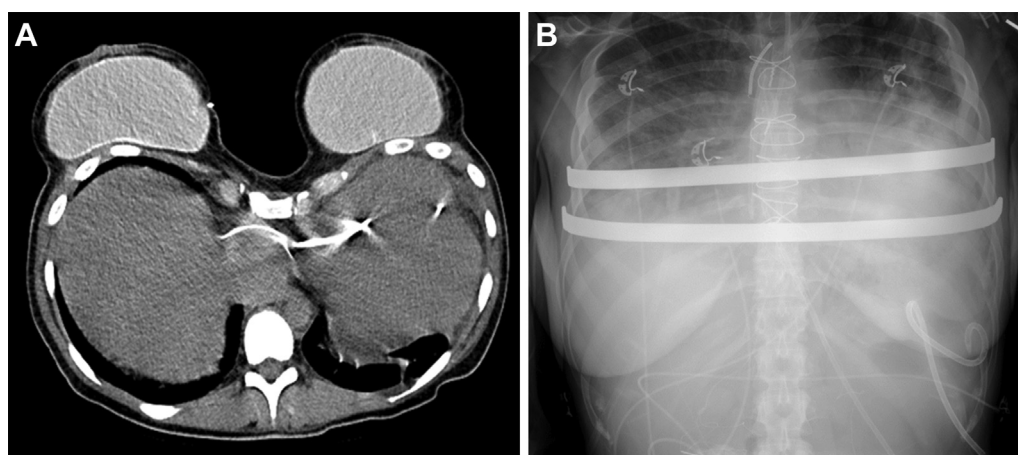
this time, it is unclear whether the risks outweigh the benefits for PE repair in patients undergoing ventricular assist device implantation. Coagulopathy, infectious concerns, and other issues may prevent safe PE repair at the time of transplant and should be included in risk assessment.

For the pectus repair, a modified MIRPE procedure was performed. The patient was positioned supine with arms tucked at the sides for transplant and a midline sternotomy was performed. Once the cardiac transplant was complete and the decision made to close the chest, a modified MIRPE was performed. Our MIRPE procedure has been previously described.^{6,7}

For the modified MIRPE after cardiac surgery, we adhere to 3 principals:

- 1) Construction of a barrier between the heart and intrathoracic Nuss bars

Figure 2. (A) Computerized tomography imaging: 34-year old woman who presented for cardiac transplantation (past medical history: non-ischemic cardiomyopathy) and severe pectus excavatum deformity, Haller index 5.8. (B) Chest roentgenogram posttransplant and pectus repair with 2 bars.



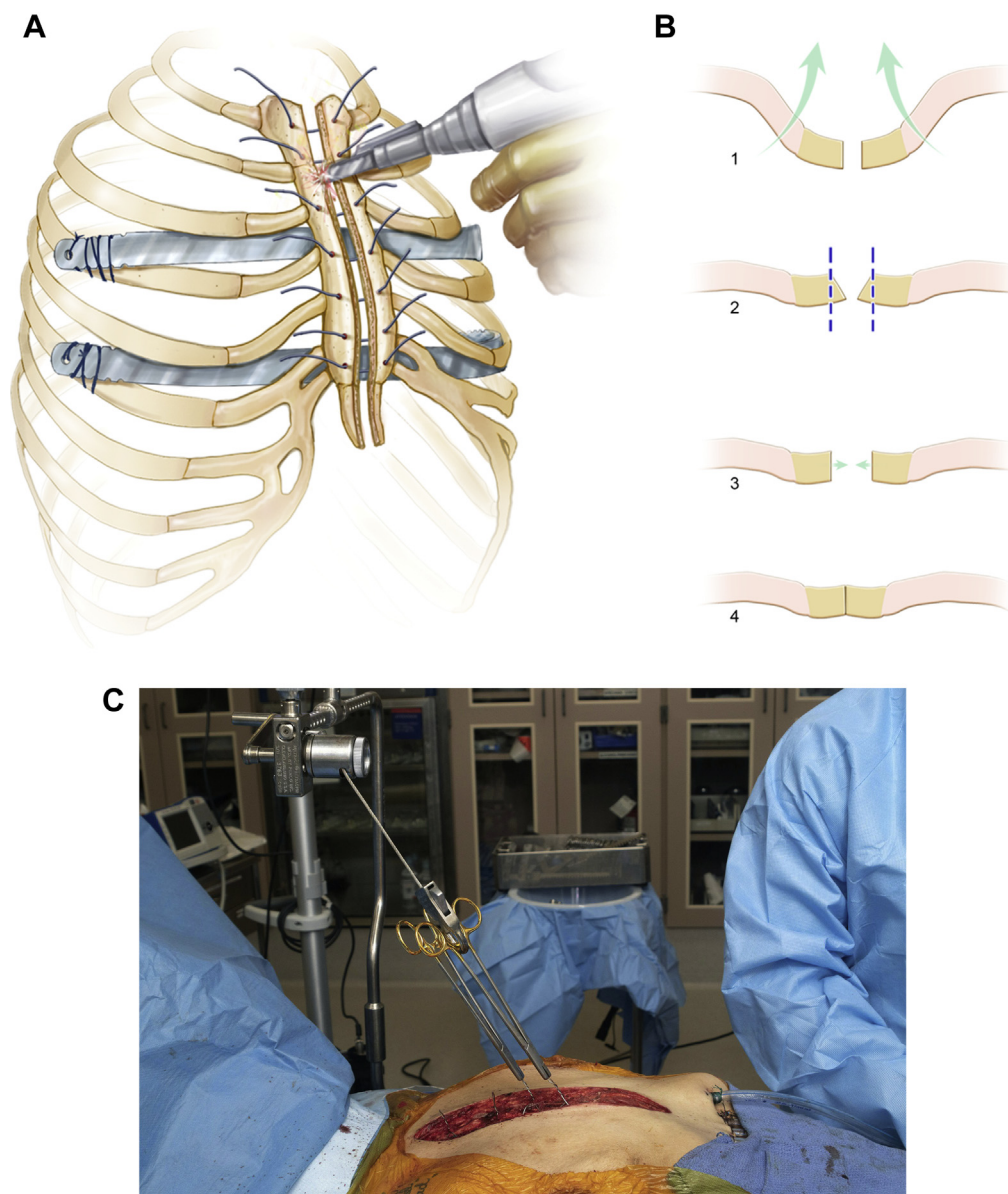


Figure 3. (A) Drawing represents the issue of approximating the cut sternal edges after elevation of the pectus excavatum deformity. (B) Recutting the angled edges allows close approximation after elevation. (C) Intra-operative photograph of the RulTract retractor (RulTract, Inc, Cleveland, OH) connected to cut sternal wires to elevate chest for Nuss bar insertion. (Image used with permission of Mayo Foundation for Medical Education and Research. All rights reserved.)

- 2) Positioning of a minimum of 2 support bars under the sternum
- 3) Close approximation of cut sternal edges by recutting the angle of sternal edges in the elevated position

In order to decrease the risk of adhesions between the heart and pectus bars, as well as facilitate safe bar removal in the future, a barrier was constructed prior to sternal closure. The open edge of the pericardium was sutured to a GORE PRECLUDE pericardial membrane (W.L. Gore & Assoc, Newark, NJ).

Stainless steel wires were then placed for planned approximation of the sternal edges. In most cases, the cut edges of the sternum will be at an angle that prevents close approximation when elevated anterior. This requires recutting the edges with an oscillating bone saw or

osteotome to allow for flush approximation (Figure 3A). Using a bone saw, osteotomies were performed to the edges of the sternum, so that flush approximation occurred in the raised position (Figure 3B).

In case 1, the patient's hemodynamic instability necessitated that the chest be elevated before we could approximate the sternum. In this case, the Nuss bars were placed first and then the sternum closed over. In the second case, the sternotomy was closed and the bars subsequently placed thoracoscopically.⁶ The cut ends of the sternal wires were attached to the RulTract retractor (RulTract, Cleveland, OH) by a needle driver to elevate upward and facilitate repair (Figure 3C).

It is our belief that a modified Ravitch for repair would be prohibitive in these patients because of the immunosuppression required post cardiac transplant

and impairment to healing. The repair of the chest wall deformity can be safely performed utilizing a modified MIRPE. Bars will be planned for removal at 3 years. It is unknown whether immunosuppression would necessitate additional time for support bars to remain in place to minimize regression risk after removal.

In conclusion, simultaneous heart transplant and repair of significant pectus excavatum is feasible and should be considered in patients with significant deformities that may cause compression of the heart graft.

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