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Descending aortic replacement after Nuss for pectus excavatum in a Marfan patient—Case report

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ABSTRACT

INTRODUCTION: The Nuss procedure for pectus excavatum (PE) repair has been successfully performed in Marfan syndrome (MFS) patients however there is concern for future risk of aortic dilation/rupture and need for emergent access with support bars in place.
CASE PRESENTATION: We present a 45 year-old male with MFS that required descending aortic replacement shortly after modified Nuss repair.
DISCUSSION: The majority of MFS patients have severe PE and repair with the Nuss procedure is not uncommon. The risk for life threatening aortic dilation, dissection, or rupture in such patients is a concern when utilizing this technique. Our work has been reported in line with the CARE criteria.
CONCLUSION: Nuss repair should be considered in MFS patients with technique modifications and careful consideration of future risk of aortic dilation and rupture.

1. Introduction

A large majority of Marfan syndrome (MFS) patients have chest wall deformities with pectus excavatum (PE) being most common [1]. Minimally invasive repair for pectus excavatum (MIRPE, "Nuss") is commonly performed with placement of support bars posterior to the sternum [2]. These bars are left in place for several years. The risk of life threatening aortic dilation, dissection, or rupture in MFS patients is a concern when utilizing MIRPE. A case is presented of a MFS patient after Nuss repair with 2 substernal bars that subsequently required repair of a Type B dissection involving the proximal descending aorta.

2. Presenting concerns

A 45-year-old Asian, married, non-smoking man was referred to Stanford University Medical Center for proximal descending thoracic aortic dilation and consideration of descending thoracic aortic graft replacement on November of 2014. The patient had MFS and history of aortic composite valve graft (St. Jude Medical, St. Paul MN) and mitral valve repair 16 years ago. He was additionally diagnosed in June 2013 with a type B aortic dissection which was being managed medically. The patient had undergone a Nuss repair for his severe PE deformity on March 2014. At the time of Nuss repair, investigations included electrocardiogram (EKG), echocardiogram (ECHO), Computed tomography (CT) scan and a cardiac stress test. Patient's CT scan revealed severe PE deformity (Haller index: 18), with only a 2.3 cm distance between sternum and the anterior aspect of mid thoracic vertebral bodies, intact ascending aortic root replacement, and unchanged chronic Stanford type B aortic dissection from proximal descending thoracic aorta to distal iliac arteries (4.3 cm dilation unchanged at level of left pulmonary artery) (Fig. 1A–D). Cardiopulmonary evaluation revealed significantly reduced functional capacity and $V0_2$ maximum.

On follow up after PE repair, it has been found on CT that his proximal descending thoracic aorta was dilating.

3. Clinical findings

In June 2014, the patient presented to the Emergency Department complaining of sudden onset of shortness of breath and dizziness, and some vague chest pain. The pain was described as mostly left-sided radiating to the left shoulder, positional, and concerning with the history of pain that he experienced with his type B dissection in 2013. A CT scan was done in the emergency room which showed that the proximal extent of the type B dissection had expanded to a diameter of 51 × 45 mm. There was no para-aortic edema or pleural effusion. The descending aorta had a persistent

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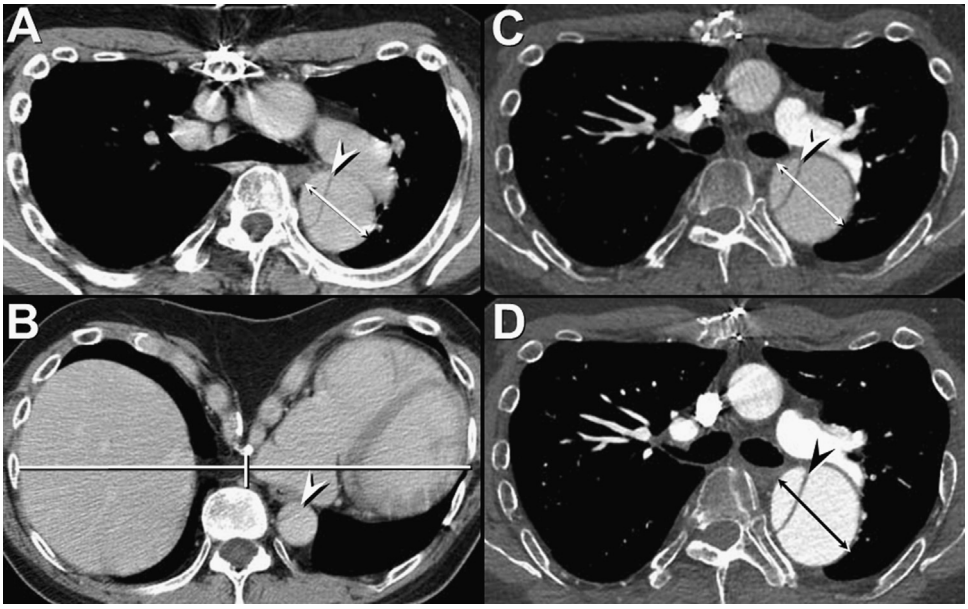

Fig. 1. Axial CTA (computerized tomographic angiography); level of left pulmonary artery shows (A) Stanford type B thoracic dissection (arrowhead) with 4.3 cm dilation of descending aorta (double arrowhead line). Severe pectus deformity (B) (Haller index = 18.15; cardiac compression index = 4.41; correction index = 84.69%). CTA 10 months later (C) shows progressive dilation to 4.7 cm proximal descending aorta (double arrowhead line). CTA at 13 months (D) shows continued dilation (double arrowhead line), now 5 cm.

dissection with a false lumen that extended from the isthmus down into the right iliac and the proximal left iliac. The celiac and right renal came off the false lumen anteriorly. The rest of the aorta was not particularly aneurysmal but the false channel was patent. In a previous CT done in August 2013 showed that the proximal descending aorta just past the isthmus measured 44 × 43 mm. He was admitted for observation and treatment.

The patient denied any recent fever, cold, nausea or vomiting, abdominal pain, constipation, or any urinary symptoms. In addition, he denied any recent heavy lifting or any strenuous activity that preceded these symptoms. He had no recent weight gain, lower leg swelling, paroxysmal nocturnal dyspnea, orthopnea. He stated that he had never had chest pain like this before, and its failure to resolve spontaneously warranted his presentation to the Emergency Department.

On examination his blood pressure was 114/65, a heart rate of 65 beats per minute with irregular rate and rhythm. His examination was remarkable only for mechanical S1 and soft S2 and a 3/6 systolic murmur.

His past medical history was remarkable for lens subluxation in 1985. Patient had history of Cosgrove annuloplasty mitral ring repair with patent foramen ovale closure at the time of root replacement in 1998. Patient had sleep apnea not on continuous positive airway pressure (CPAP).

His family history was negative for pectus excavatum. There is a question of MFS in his children. The patient's father died at age 37 of a suspected aortic rupture.

4. Diagnostic focus and assessment

During his hospital course, echocardiogram demonstrated an ejection fraction of 65%, status post 25-mm St. Jude aortic valve prosthesis conduit and ascending aortic arch graft, aortic valve prosthesis systolic mean Doppler gradient of 9 mmHg, no aortic regurgitation, normal inferior vena cava size, no pericardial effusion.

The patient was commenced on metoprolol 25 twice a day to optimize his blood pressure and heart rate control. His heart rate throughout his hospital admission was between 60 and 80 beats per minute, with a target of 60–70 beats per minute. His systolic blood pressure ranged from 100 to 120 systolic.

Table 1
Timeline.

July 1998	Aortic valve replacement with a Medtronic mechanical and ascending root replacement
June 2013	Acute Type B aortic dissection medically monitored (stable at 4.3 cm)
August 2013.	Hospitalized for high blood pressure; patient was medically stabilized and released
March 2014	Redo Sternotomy with MIRPE with 2 Nuss bars
June 2014	Hospitalized for acute onset dyspnea; patient was medically stabilized and released
November 2014	Replacement of descending thoracic aortic graft

After consultation with cardiothoracic and vascular surgery, along with the cardiology service, decision was made to continue medical management and the patient was discharged on Coumadin with close monitoring.

Follow up CT imaging showed unchanged type B aortic dissection and unchanged dilatation of descending thoracic aorta however the patient was experiencing significant psychologic stress with the uncertainty associated with possible rupture and elected to have the proximal descending thoracic aortic false aneurysm replaced.

The time course for the patient's illness is shown in Table 1. Our work has been reported in line with the CARE criteria [3].

5. Therapeutic focus and assessment

5.1. PE repair

Due to severity of defect and expected cardiac adhesions, a redo-sternotomy was performed with an oscillating hand saw. Adhesions of the right heart were dissected free from the sternum. The sternum was then reapproximated after 30° wedge osteotomy angles were made with the saw blade along sternal edges (Fig. 2A) and

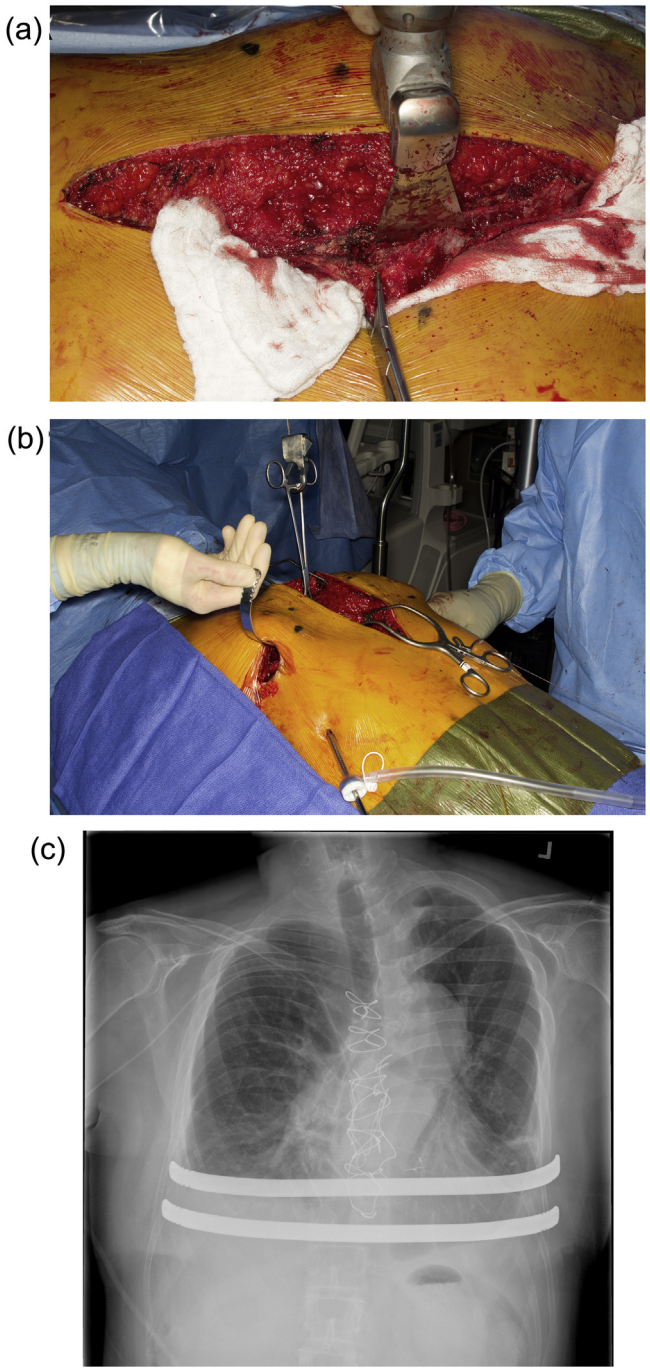

Fig. 2. (a) Oscillating saw is used to contour the angle of cut sternal edges to allow approximation when defect elevated to corrected position, (b) RulTract™ retractor is used for sternal elevation sternum and thoracoscopic bar placement. Posteroanterior chest roentgenogram (c) with 2 support bars without stabilizers.

closed with woven figure-of-eight wires. The RulTract Retractor™ (RulTract Inc., Cleveland, OH) was attached with a perforating bone clamp around sternum to elevate the defect (Fig. 2B) [4]. Thoracoscopic placement of two Nuss bars was performed at 7th and 8th intercostal spaces (Fig. 2C) and secured bilaterally at multiple sites with FiberWire® (Arthrex, Inc, Naples, FL) [5–7]. Postoperatively the patient did well, however, on multiple follow-up CT scans, the false lumen of the proximal descending thoracic aorta continued to expand (Fig. 1C and D). After several months follow up on maximal beta blocker therapy, aortic replacement was performed.

5.2. Descending aortic repair

The patient was positioned left lateral semi-decubitus (shoulders at 60° and hips at 15–20°) to allow access to the left thorax and right femoral vessels (Fig. 3a). A posterolateral thoracotomy was performed and the 4th intercostal space was entered without exposure of the pectus bars. A retractor could not, however, adequately open the intercostal space due to bar fixation of the lower intercostal spaces. The 4th and 5th ribs were shingled posteriorly which provided access to the arch and descending aorta without needing to adjust or remove the bars.

After cannulation of the right femoral vessels, the arch was clamped and partial cardiopulmonary bypass (CPB) commenced. The proximal descending aorta was clamped, the aortic false lumen opened, and the large primary intimal tear (Fig. 3b) identified just beyond the left subclavian artery take off. The dissection flap was excised, and a full-thickness aortic cuff dissected free at the level of left subclavian artery. The proximal graft anastomosis (20 mm woven double velour, Atrium Maquet, Hudson, NH) was constructed (Fig. 3c) and the proximal clamp then moved to the graft for left subclavian artery reperfusion. The distal aortic clamp was removed and the distal dissected aortic false lumen opened. The remainder of the dissection flap was excised to 5 cm beyond where the elephant trunk "waist" anastomosis would be constructed. Beyond the end of the flap rectangular excision, 3 cm of the flap was filleted open. A previously-prepared elephant trunk graft (22 mm woven double velour) was inserted into the common chamber of distal aorta and distal anastomosis completed. The elephant trunk was then extracted, and graft-to-graft anastomosis performed (Fig. 3d). Air was evacuated from the graft, clamps removed, and bypass weaned. The thoracotomy was closed in the standard fashion. No manipulation of bars or sutures securing them was required.

Postoperative imaging revealed bars and chest wall reconstruction were undisturbed.

6. Follow-up and outcomes

At 12 months follow up, CT scan with contrast performed demonstrated stable aneurysm size and anatomy and PE repair remains intact with planned bar removal 3 years post MIRPE.

7. Discussion

More than 2/3 of patients with MFS have pectus deformities [1]. Repair with MIRPE is frequently performed with 3 years recommended for the support bars to remain in place [2]. The true risk of emergent thoracic procedures following PE repair is unknown. Prior surgical interventions and sternotomy also complicate repair and increase risks. In this patient, the severity of the defect and mediastinal adhesions led us to perform redo median sternotomy prior to repair as it was felt that safe crossing of the mediastinum by thoracoscopy was not possible.

One significant concern is impediment to thoracic access, should it be necessary, with retrosternal bars. Access through a median sternotomy requires either removal or cutting of a stainless steel bar. Due to the integrity of bars, removal is recommended and should require less than 10–20 min for most patients [8]. Replacement of bars at procedure completion should be considered when possible to prevent PE recurrence for MFS patients less than 3 years from MIRPE [2]. Access to the descending aorta through left lateral thoracotomy can also be limited when longer bars and a left-sided stabilizer is present. In this patient, shorter bars with bilateral suture fixation facilitated access through the left intercostal space. Unbending of bars for lateral access is an option

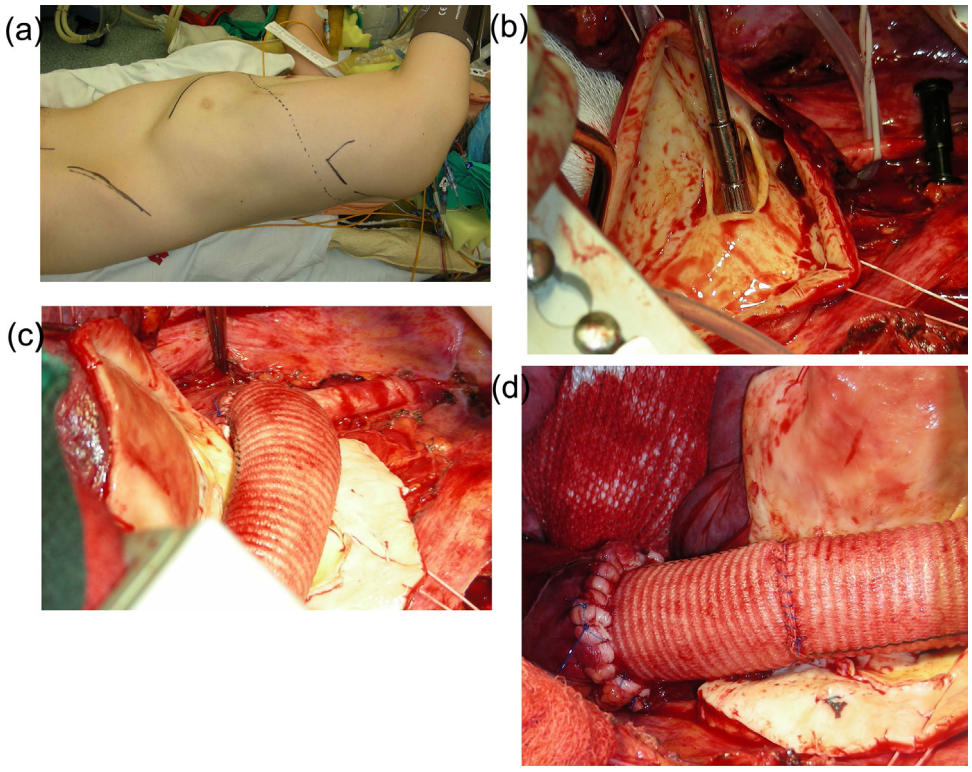

Fig. 3. (a) Patient positioning for posterolateral thoracotomy, (b) large primary intimal tear in proximal descending thoracic aorta, (c) completed proximal anastomosis, (d) distal elephant trunk graft completed.

when necessary. The Nuss set contains a tool that easily unbends and re-bends the lateral bar ends (Biomet Microfixation, Pectus Removal Benders, Jacksonville, FL). With suture fixation at multiple sites bilateral, removal of suture on the left for unbending will not significantly destabilize the bars after re-contouring. Reapproximation of the bars and stable course can be expected despite removal of FiberWire and manipulation of the bar on the left.

Repair of Marfan patients should be considered following multidisciplinary planning with cardiology and pediatric/cardiothoracic surgery. Consideration of the risk for aortic dilation and rupture may include the use of shorter bars and bilateral suture fixation should access to the thorax be needed during the period bars are retained.

8. Patient consent

The patient provided informed consent for publication of this case report.

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