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Severe pectus excavatum with tracheal compression presenting with chronic cough

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ABSTRACT

While pectus excavatum is the most common congenital chest wall deformity in children, indications for surgical repair are still debated among experts. Some centers require demonstration of physiologic criteria prior to repair, even in the face of significant anatomic compression. We present a case of a 16-year-old male with severe pectus excavatum who presented with a 3-year history of a chronic barking cough and exercise-induced shortness of breath. Radiographic imaging demonstrated compression of the trachea and bilateral mainstem bronchi secondary to severe pectus excavatum deformity. Despite his severe pectus deformity, preoperative physiologic testing demonstrated normal to near-normal cardiopulmonary function. Minimally invasive repair of the chest wall defect (modified Nuss procedure) provided rapid alleviation of symptoms, and a significant improvement in quality of life.

1. Introduction

Pectus excavatum (PE) is a posterior intrusion of the anterior chest wall occurring in 1:300–1000 live births, making it the most common congenital chest wall deformity in children [1]. The majority of PE cases are mild and asymptomatic thus never require medical attention. A small subset of patients, however, will experience increases in the severity of the depression with rapid adolescent growth, and a significant proportion of these will experience symptoms severe enough to warrant corrective surgery [1–3]. Recent adult data suggest that if surgical repair of the deformity is not performed, a portion of these symptomatic patients will experience a progressive worsening of symptoms and cardiopulmonary function with age [1,4,5]. We present a case of severe pectus excavatum causing tracheal and mainstem bronchi compression in a 16-year-old male presenting with three years of chronic cough and exercise-induced shortness of breath. Anatomic compression was demonstrated, but cardiopulmonary testing was within normal limits.

2. Case report

A 16-year-old male with no significant past medical history presented to the clinic with a three year history of a seal-like barking cough and exercise-induced shortness of breath. He also noted a lack of endurance compared to his peers and occasional chest pain with exercise. He denied a history of wheezing or frequent respiratory infections. Examination revealed a very severe pectus excavatum extending from the superior border of the sternum down to the costal margins. The defect was mildly asymmetric. Computed tomography (CT) and magnetic resonance (MR) imaging of the chest demonstrated marked depression of the anterior chest wall with subsequent severe compression of the trachea as well as mediastinal compression of the left and right mainstem bronchi (Fig. 1). The inferior vena cava was mildly compressed and displaced to the left of midline. There was flattening of the anterior wall of the right atrium and free wall of the right ventricle as well as significant displacement of the heart and mediastinal structures to the left of midline. The internal anteroposterior (AP) diameter (the distance from the anterior most aspect of the vertebral body to the deepest point of the pectus deformity), was determined to be just 7.1 mm. Given his internal transverse thoracic diameter of 249 mm, his calculated Haller index was 35.1. While a Haller index of 2.5 is considered normal, per current guidelines, an anatomic Haller index ≥ 3.2 is considered a candidate for corrective surgery by many centers, while other require abnormal physiologic testing. In the setting of

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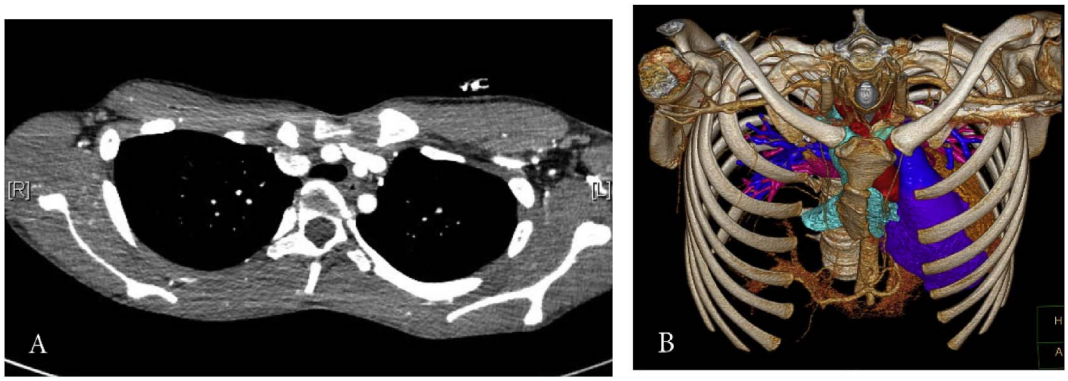

Fig. 1. A/B: Computer Tomography with IV contrast (A) as well as MRI with 3-dimensional reconstruction (B) demonstrating marked depression of the anterior chest well with compression of the trachea, right and left mainstem bronchi, and mild compression of the inferior vena cava. A mediastinal shift to the left of midline is also evident.

tracheobronchial compression as well as diminished anterior chest volume, surgical correction was recommended. In order to perform a Nuss procedure, intraoperative sternal elevation would be required prior to attempting passage of the bars over the heart.

The preoperative echocardiogram demonstrated slight right ventricular anterior free wall deviation due to the sternal defect but systolic function was within normal limits. Cardiopulmonary exercise testing was also conducted preoperatively. Spirometry demonstrated "low normal" pulmonary function; however, the patient did have difficulty providing good effort. Metabolic assessment demonstrated good aerobic capacity with a VO2max of 93% predicted, VO2 at anaerobic threshold of 93% predicted, and a RER of 1.17. No wheezing on auscultation or evidence of bronchoconstriction was appreciated throughout the duration of the test.

Anesthesia was initiated with spontaneous breathing through an LMA due to concerns of expiratory failure upon relaxation secondary to airway compromise. The plan was to emergently start the forced sternal elevation procedure if the anterior mediastinal compression caused sudden cardiovascular collapse. This technique was previously utilized and published for restoration of ventilation after catastrophic anterior mediastinal compression related to tumor undergoing biopsy [6].

With no adverse consequences with neuromuscular relaxation, a double-lumen endotracheal tube was placed. The patient was able to ventilate after positive pressure breaths, but with relatively less CO2 return per breath than had been previously. Bronchoscopy was then performed by the anesthesiologist and confirmed severe compression of the trachea and bilateral mainstem bronchi, but adequate ventilation was maintained.

Bilateral chest wall incisions were made and carried down through the soft tissue. Submuscular pockets were then created in the anterior and lateral chest wall to allow for placement of the bars. To elevate the sternum, a small incision was made on the lateral aspect of the sternum to allow a Lewin spinal clamp to be placed into the sides of the sternum. The clamp was then attached to a Rultract®, which was cranked until the sternum was appropriately elevated, allowing for safe dissection of the plane between the pericardial sac and the sternum. The attachments to the aorta and the thymus were left intact to allow continued elevation of the mediastinal structures off the trachea.

The first 12.5-inch bar was then guided into position thoracoscopically, which corrected the defect significantly. However, a residual inferior sternal depression was still appreciated. A second 12-inch bar was passed inferiorly which provided an excellent correction of the residual defect. A very minimal overcorrection was noted in the inferior aspect. The lateral portions of the bars were then inspected for fit and determined to be appropriate. Two #5 FiberWire™ were placed around each bar and rib to secure the bars in place. Additionally, two additional figure-of-eight sutures were placed on the more superior bar to prevent any additional movement and prevent later stripping. Bilateral intercostal nerve blocks were performed using 0.25% Marcaine with epinephrine. The Lewin clamp was removed, and the incisions were closed in layers and covered with Dermabond®. The patient tolerated the procedure well and was extubated in the operating suite. He was later discharged from the hospital without complication. During his post-operative follow-up, his pectus deformity was completely corrected and he was recovered well (Fig. 2). His 3-year barking cough resolved after surgical repair.

3. Discussion

Pectus excavatum is a relatively common condition with an incidence between 1:300 and 1:1000 live births, and accounts for greater than 90% of all congenital chest wall deformities [1,7,8]. Increasing depression severity is associated with the emergence or worsening of symptoms [1–3]. As mediastinal structures are compressed and/or displaced, symptoms often worsen. Additionally, worsening of symptoms with age may be due to loss of chest wall elasticity and flexibility [1,3]. Such a loss in compliance can lead to cardiac symptoms with only moderate exertion [3,9]. However, requiring evidence of cardiopulmonary impairment as an indication for surgical repair excludes a significant number of symptomatic patients who would benefit from correction of their PE. Several studies have shown that symptomatic patients with or without identifiable physiologic evidence of cardiopulmonary impairment reported significant improvements in their symptoms and an increase in perceived exercise capacity following repair, as well as a ½ standard deviation improvement in cardiovascular function [1,10–12]. Given the compression of the right heart seen on MR and CT, it would be logical to expect the physiologic impairment to be related to right heart dysfunction, rather that the more readily obtainable Cardiac Output or Oxygen-Pulse. Demonstration of anatomic right heart compression in the setting of subjective symptoms supports the argument that surgical repair of PE is indicated in symptomatic patients regardless of physiologic testing results [1].

In this case, a 16-year-old male with symptomatic severe pectus excavatum presented with a three year history of a chronic barking cough and exercise-induced shortness of breath. He also endorsed a lack of endurance compared to his peers as well as occasional exercise-induced chest pain. Radiographic imaging demonstrated evidence of significant mediastinal compression and displacement with a pectus index of 35.1. Despite his severe pectus deformity, preoperative echocardiogram and cardiopulmonary exercise testing was within normal limits with the exception of low-normal pulmonary function. The decision to undergo minimally invasive repair of the pectus excavatum via the Nuss procedure was made based on symptomatology and evidence of tracheal compression. In this case, corrective surgery dramatically improved symptomatology and quality of life despite normal or near-normal cardiopulmonary function [1,3,7–10].

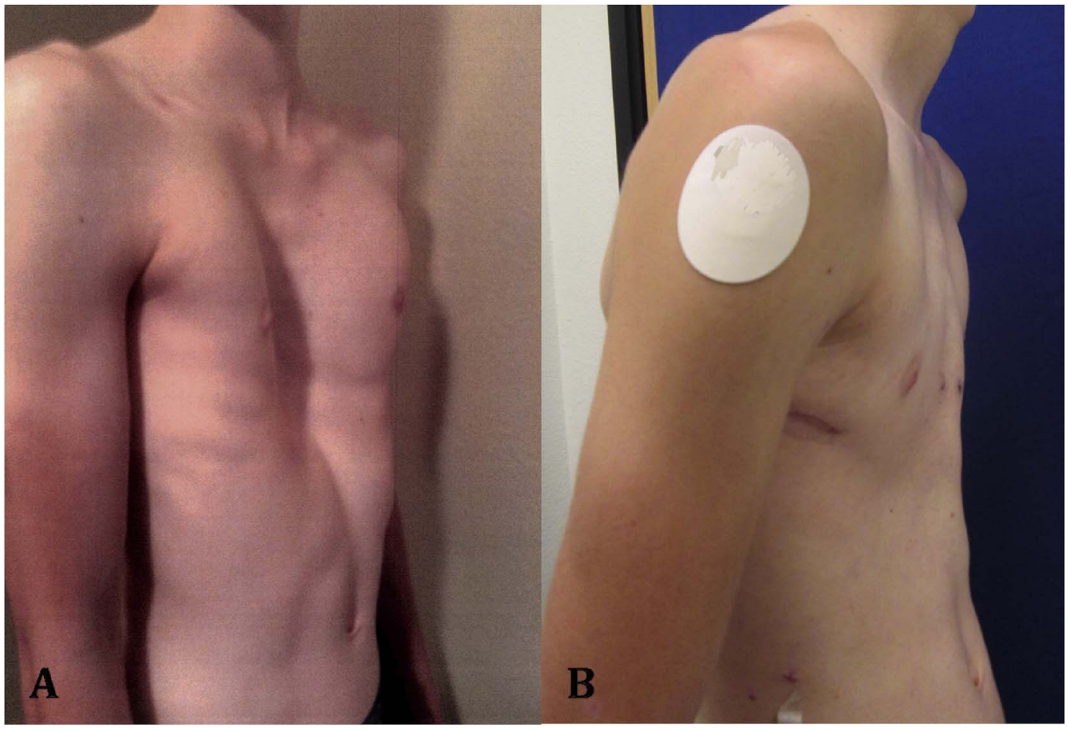

Fig. 2. A/B: Preoperative (A) and postoperative (B) photograph illustrating the surgical correction of the severe pectus excavatum deformity.

4. Conclusion

The indications for corrective surgery in patients with pectus excavatum have long been debated. The current case presents evidence suggesting patients with anatomic compression of mediastinal structures benefit from surgical repair, despite normal cardiopulmonary evaluation. In this case, a patient with symptomatic severe pectus excavatum and a three-year history of chronic cough showed resolution of symptoms after minimally invasive repair of his pectus excavatum. In patients with symptoms reasonably associated with PE, anatomic compression alone in the absence of abnormal cardiopulmonary function testing should be taken into consideration when determining indications for surgical repair.

Patient consent

Consent to publish the case report was not obtained. This report does not contain any personal information that could lead to the identification of the patient.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Conflict of interest

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Appendix A. Supplementary data

Supplementary data related to this article can be found at http://dx.doi.org/10.1016/j.epsc.2018.03.004.

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