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Thoracic insufficiency: A novel surgical approach

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1. Introduction

Thoracic insufficiency syndrome is defined as inability of the spine and thorax to maintain normal respiratory function and postnatal lung growth. It includes a heterogeneous group of congenital and developmental chest wall and spine deformities that occur early in life and can lead to progressive restrictive respiratory disease [1,2]. In some instances, the chest wall and spine abnormalities cause significant restriction of the lungs and compreass on the airways and mediastinal structures including the great vessels and esophagus. The clinical manifestation of this compression presents with shortness of breath which may be related to pulmonary and or hemodynamic abnormalities such as hypercapnia, pulmonary hypertension, or cor pulmonale. In severe cases, thoracic insufficiency syndrome may lead to premature death.

Most surgical therapeutic options described for thoracic insufficiency focus on correction of the spine deformity and osteotomies when fused ribs are present [2]. The Ravitch and Nuss procedures are used specifically for pectus excavatum deformity [3]. New spine devices include growth-friendly fusionless titanium spine distraction devices and growth guidance systems, e.g. the Shilla procedure [4]. These growth-friendly devices can be expanded repeatedly to promote vertebral growth and minimize spine curvature over time [2].

The Ravitch procedure was first developed for the surgical treatment of congenital anterior chest wall deformities including pectus excavatum and carinatum [5]. It addresses the underlying chest wall abnormality by removing abnormal cartilages and fixing the sternum in a more normal or anatomic position, supporting the correction with a bar until the cartilages heal. The Nuss procedure is now more commonly used than the Ravitch and is a more minimally invasive approach to an excavatum deformity [6].

Aortopexy is a technique that is most commonly used in the treatment of tracheomalacia [7]. Its efficacy is based on the anatomic proximity of the trachea, aorta and innominate artery such that anterior displacement of the great vessels will also pull the trachea anteriorly and thus open the airway.

For any diagnosis of thoracic insufficiency there are varying underlying structural abnormalities that contribute to the clinical symptoms, and therefore, a single surgical approach to these patients may not be effective in relieving their symptoms. For this patient, there were no options to reconstruct her spine and instead, the Ravitch procedure and aortopexy were used in combination to modify her otherwise normal anterior chest to open her mediastinal structures compressed posteriorly by her spine.

2. Case history

2.1. Preoperative evaluation

The patient is a 17-year-old girl who was referred to Pulmonary Medicine and General Surgery for a severe chest deformity. She had costovertebral dysostosis with significant thoracic lordosis. She had a long history of symptoms upon referral including worsening exercise intolerance over the prior year with shortness of breath and wheezing after walking up a single flight of stairs. She also required BiPAP for respiratory support at night due to pronounced obstructive sleep apnea with recurrent hypopneas and hypoxemic events, but no hypoventilation on polysomnogram. Physical exam was significant for severe thoracic lordosis and a pronounced anterior-posterior diameter, but no pectus deformities. She had asymmetric breath sounds diminished on the right side, forced expiratory wheezing from her left chest, and a 1/6 systolic ejection murmur. Initial evaluation for her shortness of breath included a chest and spine radiograph that showed lordosis, incomplete segmentation of multiple thoracic vertebrae and confirmed no pectus deformities. Her pulmonary function tests were consistent with severe restrictive lung disease with a forced vital capacity (FVC) of 0.86 L (31% of predicted value) and a forced expiratory volume in 1 s (FEV_1) at 0.70 L (27% predicted) with an arm span height. Her echocardiogram showed a bicuspid aortic valve without insufficiency, cardiac compression, and pulmonary hypertension with a right ventricular pressure approximately 40% of systolic pressure. She also had a mildly dilated aortic root.

A CT angiogram of the chest showed severe thoracic lordosis causing compression of her distal trachea by her vertebral bodies (Fig. 1). Cardiac compression, particularly her left atrium and right pulmonary artery was compressed at the level of her spine deformity protruding into her chest. Her right mainstem bronchus and esophagus were also compressed. Blood gas analysis showed no evidence of

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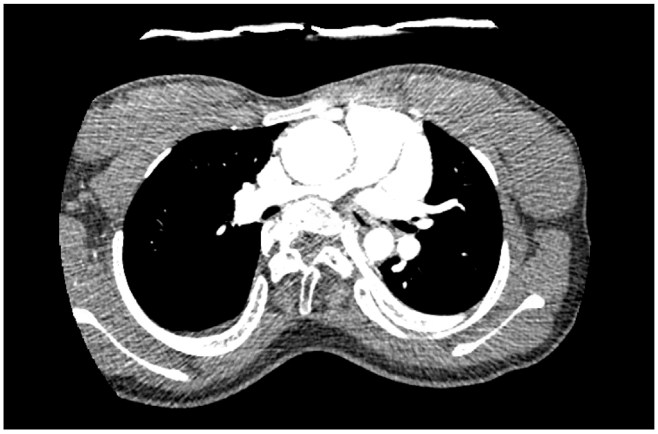

Fig. 1. CT of chest showing cross sectional imaging of the compression of mediastinal structures.

hypercapnia and oxy-hemoglobin saturation was 96% breathing room air.

Following these significant CT findings, a ventilation and perfusion (V/Q) scan were obtained. The nuclear medicine scan showed a drastic V/Q mismatch with lung ventilation of 45% on the left and 55% on the right and perfusion of 80% on the left and only 20% on the right (Fig. 2).

After assessment of these studies and a multidisciplinary discussion between the patient's providers from Cardiology, Pulmonary, Orthopedic Surgery and General Surgery, a surgical approach was deemed appropriate for correction of this complex anatomic thoracic problem with symptoms due to the significant compression of her mediastinal structures. There were multiple surgical options discussed, including anterior vertebral body wedge resection to reduce the mediastinal compression. However, it was determined that the options relating to spinal correction would not result in adequate relief of pulmonary artery compression to relieve her symptoms and improve her cardiopulmonary status.

Instead, a novel hybrid procedure was proposed creating a pectus carinatum deformity with a Ravitch-type procedure, and combining this with an aortopexy to bring the aorta anteriorly, pulling it away from the compressed pulmonary artery and improving right pulmonary artery outflow. In addition, the procedure might help open the compressed distal trachea and right mainstem bronchus.

2.2. Operative technique

Intraoperatively, the procedure was initiated with a sternotomy incision and dissection was carried down to the sternum. The pectoralis muscles were then dissected free from the sternum bilaterally exposing the costal cartilages from the second costal cartilage to the xiphoid process. All of the costal cartilages were excised bilaterally the entire length of the sternum by incising the perichondrium and excising them with a Freer elevator in the usual Ravitch type fashion.

The mid portion of the manubrium was divided using a burr drill bit and osteotome. This was completed through both the anterior and posterior tables. This effectively connected the second intercostal space bilaterally. The area of maximal compression of the mediastinal contents had been identified on CT as 3.5–5.5 cm inferior to the sternal notch. The manubrium was also cut at this point to obtain adequate sternal extraction. The perichondrium was then released bilaterally at ribs 2, 3, and 4, taking care to preserve the intercostal arteries as well as the internal mammary arteries bilaterally. The adhesions posteriorly between the sternum and the heart were also released. The combination

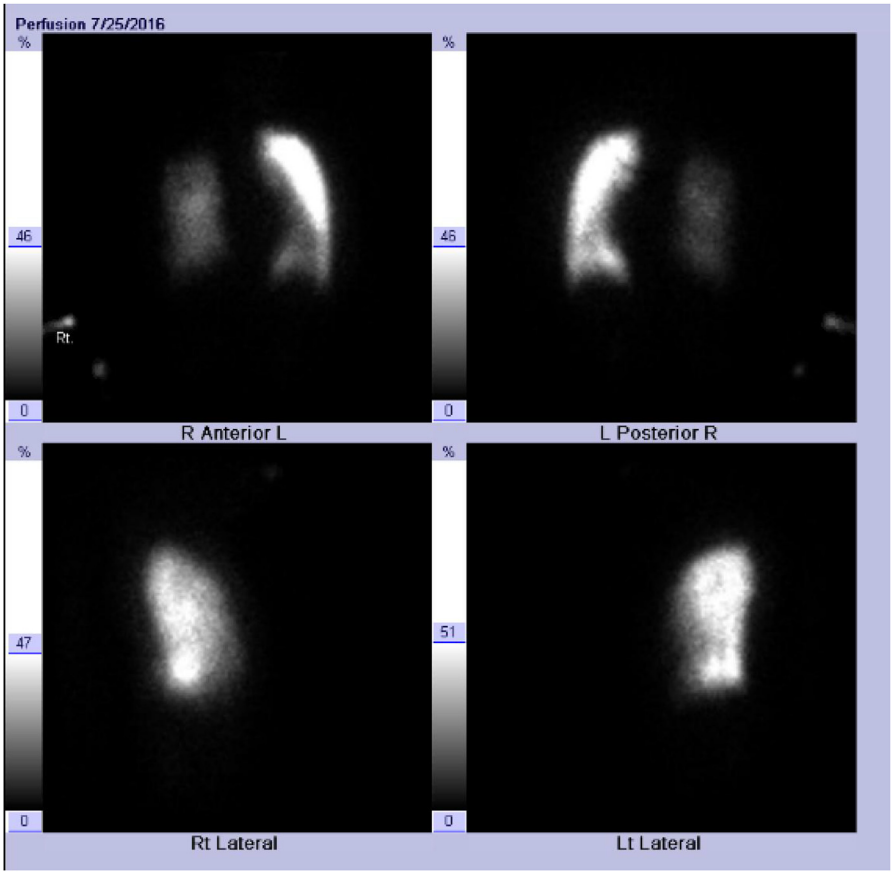

Fig. 2. Preoperative VQ scan showing the VQ mismatch with 80% perfusion on the left and 20% on the right.