Pectus excavatum (PE) is a common disorder of the chest wall that affects 1 in 500 of the general population. Affected individuals have cosmetic and, less commonly, cardiopulmonary consequences that typically present during the adolescent years. The latter is appreciated only in those with severe phenotypes, especially if associated with a “straight back” that further limits the anterior-posterior thoracic dimensions. Although the cardiopulmonary complications of PE have been well described, we describe a unique presentation of a patient who had experienced years of intractable syncope before receiving corrective surgery for severe PE. The role of cardiovascular magnetic resonance imaging (CMR) in her diagnosis and subsequent treatment is also highlighted.

A 22-year-old woman presented with a long-standing history of recurrent syncope, beginning in early adolescence. These episodes typically occurred after prolonged periods of standing or walking, beginning with a prodrome of diaphoresis and blurred vision and culminating in loss of consciousness. She described self-taught maneuvers that reduced these symptoms, including twisting her torso to the side and performing rapid, shallow breathing. In spite of these maneuvers, she continued to be limited by frequent syncope and ultimately stopped attending school at 16 years of age.

On examination, she had a marked pectus deformity with a straight back appearance. Her heart sounds were normal, with a Grade II out of VI systolic ejection murmur at the left sternal border. There was no jugular venous distention and no peripheral edema. Following identification of the chest wall deformity further questioning revealed that both her brother and paternal uncle had severe PE, suggesting an autosomal dominant inheritance pattern.

On initial investigation, her ECG showed an incomplete right bundle-branch block. A transthoracic echocardiogram was performed but was of poor quality owing to the patient’s severe PE. Pulmonary function testing showed a restrictive pattern with a vital capacity of 2.50 L (66% of predicted) and total lung capacity of 3.32 L (65% of predicted). The patient was unable to undergo cardiopulmonary exercise testing because of the rapid onset of light-headedness.

Tilt table testing was performed with elevation of the bed to 80°. After 2 minutes, the patient insisted on twisting her torso dramatically to the side and in doing so maintained consciousness for 15 minutes. When instructed to straighten her posture, she lost consciousness at 2 minutes and 30 seconds, with a documented drop in both blood pressure and pulse wave amplitude.

Cardiovascular magnetic resonance imaging was performed using a 1.5 Tesla MRI (magnetic resonance imaging)
After routine breath-held cine imaging in serial short-axis and long-axis orientations, free-breathing real-time cine imaging was performed. This was done in axial planes to identify the location of maximal PE deformity. This demonstrated marked expiratory compression of the inferior vena cava (IVC) with near obliteration of the intrathoracic space between the sternum and thoracic vertebrae (Figure 1A and 1B). Standard, retrospectively gated cine imaging was then performed in axial orientations at the level of maximal compression seen on real-time imaging. This was done at both end inspiration and end expiration (Figure 1C and 1D). The cross-sectional area and anterior-posterior diameter of the IVC decreased from 6 cm² and 21 mm during inspiration to 1.2 cm² and 3 mm during expiration, respectively. Finally, phase contrast flow imaging was performed in identical slice orientations for the determination of flow velocities at maximal inspiration and expiration while avoiding Valsalva maneuver (Figure 1E and 1F). Figure 2 demonstrates a significantly increased mean IVC flow velocity at end expiration with marked blunting of the normally biphasic flow pattern, suggesting obstruction to caval flow.

An expiratory-phase chest computed tomography scan was also performed and demonstrated an anterior-posterior intrathoracic distance of 1.2 cm (Figure 3). The pectus severity index on this study was calculated at 18.8 (severe ≥3.5).

Because of the severity of her PE and persistent symptoms, the patient underwent a minimally invasive surgical repair of her pectus deformity by use of the Nuss procedure, a pectus bar being inserted via bilateral video-assisted thoracoscopy. She tolerated this procedure well with excellent postsurgical cosmetic results. Figure 4 shows a chest radiograph that demonstrates placement of the pectus bar at the inferior sternum. A predischarge chest computed tomography scan (Figure 3) subsequently revealed no IVC compression during maximal expiration and marked improvement in the minimal expiratory thoracic depth from 1.2 cm to 5.7 cm. Four months after surgery, the patient reported no further syncopal episodes, being able to stand and walk for extended periods of time without difficulty. She has returned to school and is reporting a significant improvement in exercise tolerance and overall activity level.

Discussion

Severe PE can lead to significant morbidity through direct limitations on pulmonary and cardiac performance as well as through associated disorders, such as scoliosis, mitral valve prolapse, and musculoskeletal chest pain. A reduction in upright cardiac output in comparison with age-matched control individuals has been well documented in affected persons and is believed to be due to the restrictive effects of the sternum on right ventricular filling. However, syncope is an exceptionally rare complication in PE, with only 1 prior case reported in the literature, to our knowledge.
Crossland et al\(^5\) reported on a 7-year-old girl with anterior Raphe syndrome who had undergone sternal repair of a midline defect at 3 years of age. In this case, a chest computed tomographic scan revealed compression of the right ventricle. This was thought to be the cause of her syncope resulting from reduction in cardiac output. No physiological testing or dynamic imaging was performed in this case. The child did not have syncope after reconstructive surgery.

The present case demonstrates conclusive evidence of expiratory sternal compression of the IVC with associated flow restriction by dynamic CMR. Position dependency of syncope was confirmed during tilt-table testing, suggesting that this patient’s learned “twisting” maneuvers likely shifted the sternum away from the IVC to maintain flow and avoid syncope. Similarly, the rapid shallow breathing technique adopted by the patient likely avoided syncope through avoidance of significant expiratory compression.

The role of current CMR imaging techniques in patients with PE has not been previously described, to our knowledge. Significant incremental data were provided by the use of dynamic CMR imaging in this case, especially given that echocardiographic imaging was characteristically poor. The physiological consequences of this patient’s sternal compression were well characterized by both real-time cine CMR imaging and MRI flow assessment.

In conclusion, this case highlights an exceptionally rare complication of severe PE and the role of CMR in assisting in its diagnosis and successful management. In select cases of PE, cardiovascular MRI, inclusive of real-time cine imaging and phase contrast flow imaging, should be considered to assist in therapeutic decision making.

**Disclosures**

None.

**References**

Pectus Excavatum With Compression of the Inferior Vena Cava: A Rare Cause of Recurrent Syncope
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