

Positional Right Ventricular Obstruction in Pectus Excavatum



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Pectus excavatum is one of the most common congenital chest wall deformities. The degree of sternal depression, which may result in compression of the right heart by the chest wall, is variable. While typically asymptomatic, there are various symptoms that can result from severe pectus excavatum. We report on a patient with severe pectus excavatum leading to dynamic obstruction of the right ventricular outflow tract in the seated position. © 2017 Elsevier Inc. All rights reserved. (Am J Cardiol 2017;119:1288–1289)

Pectus excavatum is one of the most common congenital chest wall deformities. The degree of sternal depression, which may result in compression of the right-sided cardiac by the chest wall, is variable. Although typically asymptomatic, there are various symptoms that can result from severe pectus excavatum.¹ We report on a patient with severe pectus excavatum leading to dynamic obstruction of the right ventricular outflow tract (RVOT).

Case Report

A 19-year-old man with a history of severe pectus excavatum presented with orthostatic dizziness. He was a tall thin man in no acute distress with prominent pectus deformity. When in supine position, his heart rate was 70 beats/min, and his blood pressure was 100/60 mm Hg. When sitting upright, his heart rate increased to 100 beats/min with a blood pressure

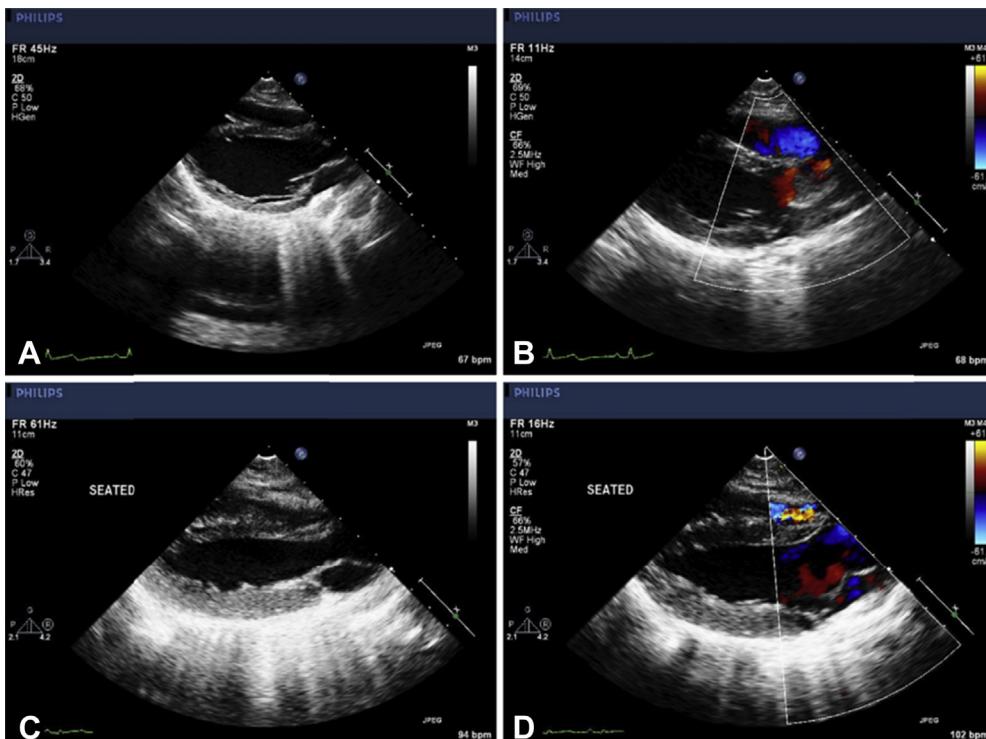


Figure 1. Parasternal long-axis echocardiography images in the left lateral decubitus position (A). The RV appears mildly reduced in size but by color Doppler (B), there is no aliasing of flow (evidence of compression). Additional parasternal long-axis views with the patient seated upright demonstrate near complete collapse of the RV (C) and evidence of turbulent flow (evidence of compression) (D). RV = right ventricle.

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of 98/70 mm Hg. The point of maximal impulse was nondisplaced, and pulses were normal and equal. His electrocardiogram demonstrated sinus rhythm with incomplete right bundle branch block and right axis deviation. A transthoracic echocardiogram performed in the left lateral decubitus position was normal. However, when images were obtained in the seated position, there was significant

compression of the right ventricle by the deformed sternum and a peak RVOT gradient of 16 mm Hg (**Figure 1**).

Symptomatic patients with pectus excavatum often present with signs of exercise intolerance out of proportion to their degree of pulmonary restriction.¹ The location of the deformity is critical, and even small degrees of depression can result in significant right-sided cardiac compression and impaired venous return or right ventricular outflow. Decreased exercise capacity, as measured by cycle ergometer, has been observed in seated pectus excavatum patients compared with both supine pectus excavatum patients and seated controls. Compression of the right-sided cardiac chambers, exacerbated when seated, interrupts the ability to appropriately maintain the cardiac output.² The pericardial sac is suspended in the chest cavity, and its positioning is susceptible to the effects of gravity. A case report on a dog with pectus excavatum described dynamic RVOT stenosis and a prominent systolic murmur.³ Another case report described a patient with pectus excavatum noted to have severe hypotension when placed in the prone position for elective spinal surgery; echocardiogram confirmed an elevated peak right ventricular inflow gradient of 17 mm Hg

that resolved when supine and could be reproduced by direct compression of the anterior chest.⁴ Our patient was completely asymptomatic when supine but demonstrated an increased RVOT velocity, murmur, tachycardia, and decreased blood pressure when seated. Echocardiography confirmed a dramatic degree of dynamic RVOT obstruction.

Disclosures

The authors have no conflicts of interest to disclose.

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