

IMAGING VIGNETTE

INTERMEDIATE

CLINICAL VIGNETTE

Dynamic Right Ventricular Outflow Tract Obstruction in Straight Back Syndrome



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ABSTRACT

Straight back syndrome is a congenital skeletal abnormality of the upper dorsal spine. This clinical case describes a 29-year-old woman with atypical chest pain and a changing murmur that was attributed to dynamic right ventricular outflow tract obstruction on echocardiography. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2019;1:436-7) © 2019 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

Straight back syndrome is a congenital abnormality of upper dorsal spine characterized by loss of natural curvature of spine (1). In some patients it can mimic a structural heart disease. Diagnosis is typically made on frontal and lateral chest radiographs using Davies or De Leon criteria (2,3).

Here we describe a 29-year-old thin lady, having atypical chest pain for last 2 years. On examination she had a heart rate of 70 beats/min and blood pressure of 110/70 mm Hg. The first and second heart sounds were loud with a normally split S₂, and there was a grade III/VI ejection systolic murmur in left second intercostal space in sitting posture. Her electrocardiogram was normal and supine echocardiography did not reveal any abnormality (Figures 1A and 1B, Video 1). Her subsequent visits would reveal the same murmur but repeated echocardiography was normal. At this point we performed simultaneous auscultation and echocardiography in supine and sitting positions. The murmur was absent in the supine position and reappeared on sitting. Dynamic echocardiography in the supine and sitting positions unveiled the mechanism responsible. During echocardiography in sitting position, we found a dynamic turbulent flow through the right ventricular outflow tract (RVOT) with a peak gradient of 90 mm Hg corresponding with murmur in sitting posture (Figures 1C and 1D, Video 2). Because she was a lean, thin, young lady, the possibility of straight back syndrome was suspected as the cause responsible for this changing murmur. The frontal and lateral chest radiographs showed an anteroposterior (AP) diameter of 6.4 cm, and lateral (L) diameter of 21.6 cm and the ratio of AP/L was 0.3 (De Leon criteria of AP/L: 1/3) (Figures 1E and 1F). On lateral chest radiograph, the distance from the middle of the anterior border of T8 to the line joining the top of the anterior border of T4 to the bottom of the anterior border of T12 was 1.17 cm (Davies criteria of <1.2 cm) (Figure 1G). She did not have any other skeletal or cardiac abnormalities. There was no limitation of her activity. She was reassured and advised to have a yearly follow-up.

The mechanism of this murmur has been related to the compression of RVOT and kinking of great vessels as the heart comes nearer to sternum in upright posture (1).

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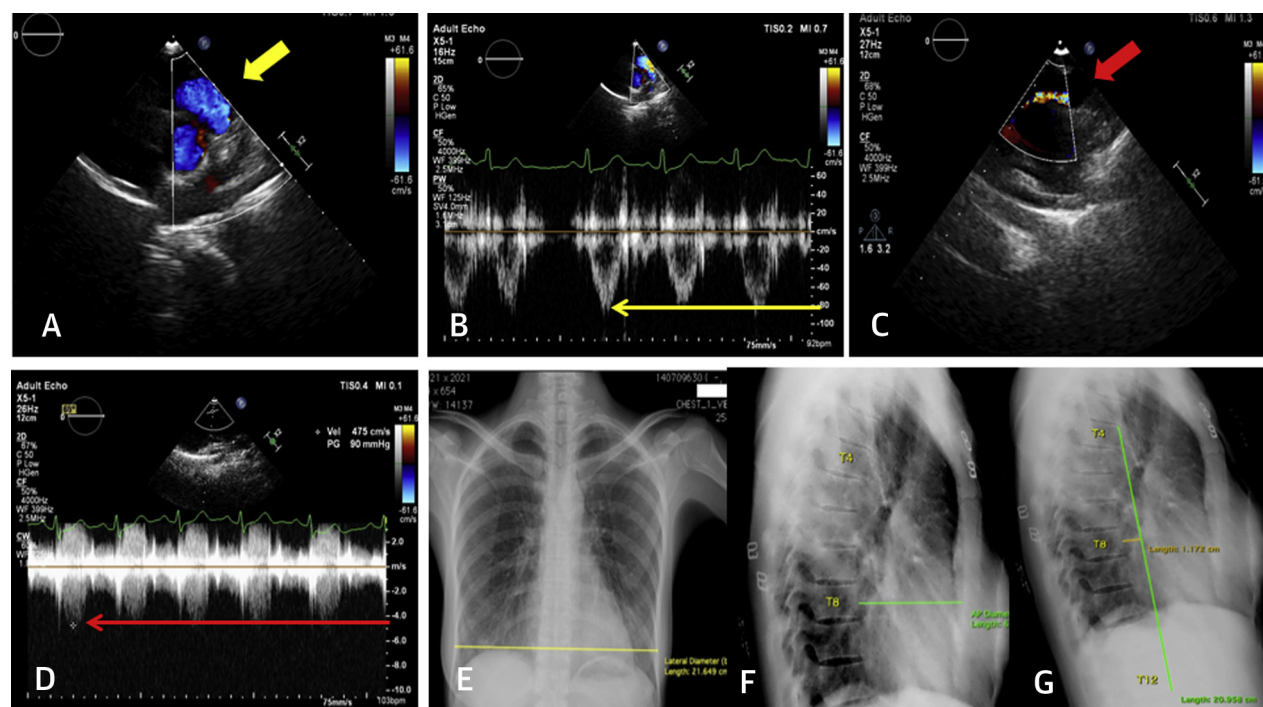
This murmur lacks the typical Rivero-Carvalho sign, and we further propose that the decrease in size of the right ventricular cavity on standing leads to a turbulent flow through RVOT (Figure 1C) similar to that in hypertrophic cardiomyopathy. The other causes of a murmur in straight back syndrome are mitral valve prolapse and bicuspid aortic valve. This case highlights the value of detailed clinical examination in making a diagnosis and optimal use of technology beyond routine indication such as dynamic echocardiography in this case.

ABBREVIATIONS AND ACRONYMS

AP/L = antero-posterior/
lateral diameter

RVOT = right ventricular
outflow tract

FIGURE 1 Echocardiography in Supine and Sitting Posture Showing Dynamic Flow Across Right Ventricular Outflow Tract and Frontal and Lateral Chest Radiographs Confirming the Diagnosis of Straight Back Syndrome



(A) Transthoracic 2-dimensional color Doppler echocardiographic images in left parasternal short-axis view in supine position showing normal flow across right ventricular outflow tract (yellow arrow, Video 1). (B) Pulsed-wave Doppler with a peak flow velocity of 0.9 m/s (horizontal yellow arrow). (C) Two-dimensional echocardiography in sitting position showing turbulence across right ventricular outflow tract (red arrow, Video 2). (D) Continuous-wave Doppler showing a peak gradient of 90 mm Hg. (E) Frontal chest radiograph showing a lateral diameter of 21.65 cm (yellow line). (F) Lateral view showing the anteroposterior diameter of 6.4 cm at the level of T8 vertebra (green line). (G) Lateral chest radiograph showing the distance of 1.17 cm between anterior border of T8 vertebrae and line joining T4 to T12. RVOT = right ventricular outflow tract.

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KEY WORDS echocardiography, murmur, right ventricle

APPENDIX For supplemental videos, please see the online version of this paper.