

Intermittent' restrictive ventricular septal defect in Tetralogy of Fallot

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ABSTRACT

Ventricular septal defect (VSD) in Tetralogy of Fallot (TOF) is usually large and non-restrictive with equalization of right and left ventricular pressures. Restrictive VSD in TOF is rare. We present an unusual case of TOF with restriction to VSD caused by accessory tricuspid valve tissue that varied with respiration.

Keywords: Accessory tricuspid tissue, restrictive ventricular septal defect, Tetralogy of Fallot

CLINICAL CASE

A 10-year-old boy presented with cyanosis (oxygen saturation 78%) and progressive dyspnea on exertion since childhood. The jugular venous pulse showed prominent 'a' wave and had left parasternal lift. A grade IV/VI harsh ejection systolic murmur was heard in second left intercostal space, which increased on expiration and decreased on inspiration. A transthoracic echocardiogram (TTE) diagnosed Tetralogy of Fallot (TOF) [Videos 1 and 2]. Electrocardiogram and chest roentgenogram were typical of TOF. However unlike classical TOF, the ventricular septal defect (VSD) was restrictive. Restriction was caused by a flap-like accessory septal tricuspid valve (TV) tissue that varied with respiration. On expiration, the right ventricle (RV) size decreased bringing the accessory tissue close to interventricular septum (IVS) causing restriction to the VSD flow with peak velocity of 3 m/sec and a right to left gradient of 36 mmHg [Figure 1a and 1c, Video 1, Video 3]. On inspiration RV size increased, separating the accessory tissue from the IVS, and thus reducing the restriction [Figure 1b]. Cardiac catheterization confirmed TOF with supra-systemic RV pressures at 190 mmHg,

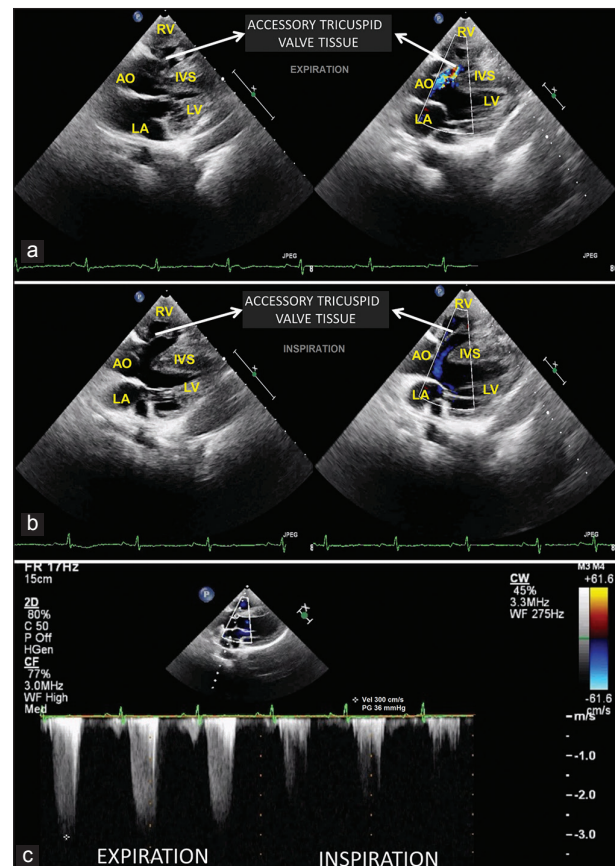


Figure 1: (a) During expiration, the right ventricular (RV) decreases in size (compared with 2D image during inspiration). This brings the accessory tricuspid tissue close to interventricular septum (IVS) leading to restriction of ventricular septal defect (VSD) flow seen as turbulence across VSD. (b) During inspiration the RV size increase, separating the accessory tissue from IVS, thus decreasing the restriction (decreased turbulence across the VSD). (c) Continuous wave Doppler at the VSD showing right to left jet with maximum velocity of 3 m/sec and gradient of 36 mmHg during expiration. This gradient is reduced on inspiration

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10.4103/0974-2069.149533

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simultaneous left ventricular (LV) systolic pressure being 130 mmHg.

Restrictive VSD in TOF is rare. Restriction is most commonly caused by accessory TV tissue, and less frequently by hypertrophied septal band.^[1] Echocardiographic features of this rare variant have been described.^[1] Two types of accessory TV tissue have been described, “Mobile” and “fixed”.^[2] The mobile type, by virtue of its greater mobility has a greater propensity of protrusion in to sub-aortic region causing restriction to VSD as well as left ventricular outlet obstruction.^[2] To the best of our knowledge, such intermittent restriction of VSD by TV apparatus, presenting with unusual increase in murmur on expiration in patient with TOF, has not been previously described.

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How to cite this article: Shetkar SS, Kothari SS. Intermittent' restrictive ventricular septal defect in Tetralogy of Fallot. *Ann Pediatr Card* 2015;8:80-1.

Source of Support: Nil, **Conflict of Interest:** None declared

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