Utility of three-dimensional echocardiography and magnetic resonance imaging in the diagnosis of double-orifice tricuspid valve

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ABSTRACT

Duplication of atrioventricular valves involves the mitral valve more often than the tricuspid valve and is often associated with other cardiac defects. Double-orifice tricuspid valve (DOTV) is often identified in surgery or autopsy and missed on echocardiography, as the two orifices are orthogonal to the imaging plane. If suspected on echocardiography, it masquerades as mild tricuspid hypoplasia. Three-dimensional echocardiography and magnetic resonance imaging of a DOTV are presented.

Keywords: Double-orifice tricuspid valve, duplication of atrioventricular valves, magnetic resonance imaging, three-dimensional echocardiography, tricuspid hypoplasia, ventricular septal defect

Double-orifice tricuspid valve (DOTV) is often associated with additional heart defects.^[1,2] It is often missed on echocardiography and commonly diagnosed on autopsy or surgery. Three-dimensional echocardiography and magnetic resonance imaging have helped in diagnosing many rare anomalies.^[3]

A 7-year-old asymptomatic girl with normal growth was diagnosed on echocardiography to have a perimembranous ventricular septal defect partially restricted by the septal tricuspid leaflet and mild tricuspid hypoplasia on apical view [Figure 1 and Video 1]. The electrocardiogram showed normal sinus rhythm, normal QRS axis, no preexcitation, and left ventricular dominance. An orthogonal rightward sweep of a parasternal long-axis view toward the right ventricular inflow demonstrated double orifice [Figure 2 and Video 2a and b]. A large anterosuperior and a small septal leaflet whose edges partially closed the ventricular septal defect formed the anterior larger orifice. A smaller posteroinferior second orifice formed by a broad anterior leaflet and

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smaller apically displaced posterior leaflet showed a small atrialized right ventricle and mild regurgitation.

Three-dimensional echocardiography confirmed the presence of two separate tensor apparatuses for both these orifices [Figure 3 and Video 3a and b]. The combined area of the two orifices maintained adequate right ventricular inflow, contrary to the initial impressions of tricuspid hypoplasia. Magnetic resonance imaging confirmed DOTV, adequate right ventricular volumes, shunt ratio of 1.5, and insignificant tricuspid regurgitation [Figure 4 and Video 4]. No interventions were planned as there was no significant shunt or chamber enlargement.

DOTV is commonly associated with septal defects, mitral and pulmonary valve malformations, Ebstein's anomaly, and tetralogy of Fallot.^[1,2] It is missed on echocardiogram as the two orifices often have orthogonal orientation to the apical four-chamber view.^[4] It may masquerade

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Figure 1: Four-chamber apical view shows mild tricuspid annular hypoplasia. A small right ventricular inflow should alert the clinician to carefully look for double-orifice tricuspid valve



Figure 3: Two- and three-dimensional echocardiogram from subxiphoid window shows the two orifices with separate tensor apparatus

as a hypoplastic tricuspid valve.^[5] It is classified into commissural, central, and hole types.^[1] The relatively rare commissural type has a smaller accessory orifice at the end of a valve commissure.^[2] The more common central type has a band dividing the valve into equal or nearly equal orifices as in our patient. The hole type, where an accessory orifice appears as a hole within a large leaflet, is also very rare. DOTV is distinguished from a simple fenestration or cleft by a lack of subvalvular tensor apparatus support for the latter. The difficulty to identify this subvalvular tensor apparatus on preoperative echocardiography is one of the reasons why most cases are identified only on surgery or autopsy.^[6]

Three-dimensional echocardiography and magnetic resonance imaging improve spatial resolution to enable preoperative diagnosis. They additionally quantify right ventricular volumes, as DOTV often presents on echocardiography as hypoplastic tricuspid valve.^[5] Magnetic resonance imaging also helps the assessment of the shunt ratio and excludes other extracardiac associations.



Figure 2: Parasternal long-axis view (a) with a rightward sweep to image the right ventricular inflow with color flow imaging (b) demonstrates the two orifices of the tricuspid valve with larger anterior orifice and smaller posterior orifice. There is minimal atrialization of the posterior basal right ventricular wall due to apical displacement of the posterior leaflet



Figure 4: Magnetic resonance imaging in sagittal plane (a) and right ventricular inflow–outflow view (b) demonstrates the two orifices, restrictive ventricular septal defect, and mild tricuspid regurgitation and confirms the adequacy of right ventricular inflow

IMPACT ON CLINICAL PRACTICE

A small tricuspid annulus on four-chamber view on echocardiography should alert the clinician to look for DOTV. An orthogonal projection in a right swept parasternal long-axis view with color flow imaging clinches the diagnosis. Three-dimensional echocardiography demonstrates separate tensor apparatus for both the orifices. Magnetic resonance is a unique multiutility tool in the confirmation of diagnosis, assessing the adequacy of the right ventricle and quantifying the shunt and regurgitation.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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