A systematic method for using 3D echocardiography to evaluate tricuspid valve insufficiency in hypoplastic left heart syndrome

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ABSTRACT

With surgical palliation of hypoplastic left heart syndrome (HLHS), the tricuspid valve (TV) becomes the systemic atrioventricular valve and moderate/severe TV insufficiency (TVI), an adverse risk factor for survival to Fontan, has been reported in up to 35% of patients prior to stage I palliation. Precise echocardiographic identification of the mechanism of TVI cannot be determined by two-dimensional echocardiography. Three-dimensional echocardiography (3DE) can provide significant insight into the mechanisms of TVI. It is the intent of this report to propose a systematic method on how to evaluate and display 3DE images of the TV in HLHS which has not been done previously.

TV anatomy, function, and the known mechanisms of insufficiency are reviewed. We defined three regions of the TV (anterior, posterior, septal) that can help define valve "leaflets" that incorporates the many variations of TV anatomy. To determine how the surgeon views the TV, a picture of a pathologic specimen of the TV was placed on a computer screen and rotated until it was oriented as it appears during surgery, the "surgeons view."

We have proposed a systematic method for evaluating and displaying the TV using 3DE which can provide significant insight into the mechanisms causing TVI in HLHS. This has the potential to improve both the surgical approach to repairing the valve and, ultimately, patient outcomes.

Keywords: Insufficiency, three-dimensional echocardiography, tricuspid valve

BACKGROUND

Surgical palliation of hypoplastic left heart syndrome (HLHS) results in the tricuspid valve (TV) becoming the systemic atrioventricular valve and development of moderate/severe TV insufficiency (TVI) has been reported in up to 35% of patients prior to stage I palliation.^[1]

The mechanisms of TVI in HLHS can be divided into two broad categories, structural and functional. Structural abnormalities include dysplastic leaflets, clefts, or fissures (indentations) in the valve leaflets,

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leaflet tethering, leaflet prolapse, accessory orifices, Ebstein's anomaly, bileaflet, and quadraleaflet valves.^[2-4] Functional abnormalities include ventricular dilation, annular dilation, and right ventricular (RV) dysfunction.^[2,3]

The development of moderate/severe TVI has been shown to be a significant adverse risk factor for survival to Fontan^[1,2] and TV surgery is necessary in up to 25% of those who survive stage I palliation.^[2,5] Understanding the principal cause of the TVI improves the chances of a successful surgical repair and patient outcome.^[6]

Precise echocardiographic evaluation of the TV has been advocated even when performing stage I palliation.^[1] Two-dimensional echocardiography (2DE) is sensitive for detecting leaflet thickening, distortion, and motion abnormalities but is inadequate for identifying structural abnormalities and how extensively the various valvar components contribute to the insufficiency.^[3]

Three-dimensional echocardiography (3DE) can display the TV in a manner that is not possible using standard

Address for correspondence: Dr. Christopher Robin Mart, University of Utah – Primary Children's Medical Center, 100 North Mario Capecchi Drive, Salt Lake City, Utah 84113, USA. E-mail: christopher.mart@imail2.org 2DE.^[7,8] Recent advances in 3DE technology have provided superior ability to describe valve anatomy leading to a better understanding of the topographical aspects of valvar pathology^[7,8] and has provided important insights into the mechanisms of TVI in patients with HLHS.^[9] However, a systematic method on how to use 3DE to assess or display the TV in HLHS has not been implemented.

The intent of this report is to propose a systematic method for using 3DE to identify and display both the pathologic anatomy and mechanism of insufficiency of the TV in children with HLHS.

Tricuspid valve anatomy and function

3DE imaging of the TV requires an understanding of valve anatomy, including the annulus, leaflets (cusps), chordal apparatus, and papillary muscles. However, there is no consensus in the literature regarding the actual anatomy of the TV.^[10] Although commonly believed to have three cusps (anterior, posterior, and septal), the TV is a heterogeneous structure with great variability^[11] and some have contended that the numerous variations in the cusps and support apparatus (chordae/papillary muscles) make the anatomy of the TV as unique as one's fingerprint.^[12]

Normal tricuspid valve anatomy

Annulus

The TV is a nonplanar structure with an elliptical^[13] saddle-shaped annulus^[7,14] with the highest points located in an anterior-posterior orientation and the lowest points in a mediolateral orientation,^[7] [Figure 1].

The annulus is a dynamic structure that decreases both in circumference and area during ventricular contraction.^[15-17] When the valve is placed on the clockface, the annulus contracts roughly from the 12:30 to the 6 o'clock position [Figure 2].

Valve leaflets/cusps

The valve is divided into what are called leaflets or cusps by "commissures." The TV is traditionally considered to have three commissures.^[14] The anteroseptal separates the anterior and septal leaflets, the anteroposterior lies between the anterior and posterior leaflets, and the posteroseptal divides the posterior and septal leaflets. Generally, the free edge of the commissures forms a smooth arch although small projections, "miniscallops" or commissural cusps, may be present^[14] and are most frequently located between the anterior and posterior cusps^[18] [Figure 3].

The TV is described as having three leaflets/cusps. However, a universally accepted definition of what constitutes a valve commissure/cusp, and how to distinguish one cusp from another, has not been



Figure 1: The tricuspid valve (TV) has a saddle shape because of anterior and posterior high points and mid septal and lateral wall low points



Figure 2: During ventricular systole, the annulus of the tricuspid valve (TV) contracts roughly from the 12:30 to 6:00 o'clock position, red arrows. A: Anterior leaflet, P: Posterior leaflet, S: Septal leaflet



Figure 3: Although free edge of the valve commissure generally forms a smooth arch (blue and red arrows), there may be a small projection of leaflet tissue, the "miniscallop" or commissural cusp (white arrow), in the free edge of the commissure. The blue arrow points to the anteroseptal commissure, the red arrow to the anteroposterior commissure, and the white arrow to the posteroseptal commissure. A: Anterior leaflet, P: Posterior leaflet, S: Septal leaflet. Right atrial view

defined.^[10-12,19] Subdivided, accessory, and commissural cusps^[19] have been described and the number of cusps found in a normal "tricuspid" valve varies from 2 to 8.^[10,13,17-25] Alternatively, others have proposed that

all TVs are bicuspid with a sepal cusp (generally with no clefts) and a mural cusp with a varying number of clefts which artificially divide it into multiple segments.^[12]

The anterior leaflet is generally the largest^[10,14,17,21] and the most mobile.^[13] The septal leaflet is generally the second largest and the least mobile.^[10,12,13,17,18,21] The posterior leaflet is typically triangular and is generally the smallest.^[10,11,13,18]

Papillary muscles and chordae tendineae

The number of papillary muscles varies considerably ranging from two to nine.^[18,19] The large anterior papillary muscle (APM) is attached to the RV anterior wall with chordae to the anterior/posterior cusps. The posterior papillary muscle consists of two or more parts attached to the inferior wall of the RV with chordae to the posterior/septal cusps. The septal papillary muscle, Lancisi's muscle, is a variable group of small papillary muscles attached to the ventricular septum with chordae to the anterior/septal cusps.^[17,19,20,22,26] Not all of the papillary muscles are always present, some are fused and the number of heads found on each muscle differs considerably.^[10,12,20]

The enormous variability in the anatomic description of the TV highlights the importance of a consistent and reproducible method of describing the valve.

Tricuspid valve function

Coaptation of the TV leaflets is a dynamic, complex process involving the entire valve apparatus.^[22,27] Optimal coaptation occurs when the annulus has an elliptical shape due to the limited mobility of the septal leaflet. This is maintained by atrial, annular, and papillary muscle contraction,^[16] along with the ventricular septum bending toward the RV which decreases the annular diameter in the lateral direction.^[16,27] The TV is not designed to close a circular orifice, circular geometry results in the septal leaflet being pulled away from the anterior/posterior leaflets resulting in valvar incompetence.^[16]

Mechanisms of TVI

Normal heart

In normal hearts, the most common cause of TVI is left heart disease leading to RV dilation/dysfunction^[7,22] with secondary TVI. The tricuspid annulus dilates, becomes more circular, and the leaflets are tethered, all of which lead to inadequate leaflet coaptation.

Hypoplastic left heart syndrome

In contrast, the mechanisms of TVI in HLHS are multifactorial and represent a complex interaction between the right atrium (RA), annulus, leaflets, papillary muscles, and the RV.^[3,15,28]

These include right atrial dilation, annular dilation, abnormal annular geometry, leaflet abnormalities,

chordal/papillary muscle variation, dilation/ geometric changes of the RV due to chronic volume overload, abnormal LV-RV interaction, RV dysfunction, myocardial damage/ischemia, and increased afterload created by inadequate arch reconstruction after stage 1 repair.^[2-4,9,15]

Valve leaflets

TVI in HLHS is frequently associated with structural anomalies of the valve. Bileaflet, trileaflet, and quadraleaflet valves have been described along with: leaflet prolapse, leaflet restriction or "tethering," accessory orifices, dysplastic leaflets, clefts, parachute malformation, and Ebstein's anomaly. TV dysplasia is more common in those with a patent mitral valve (62%) and least common in those with both mitral and aortic valve atresia (15%).^[3,4]

Chordae/papillary muscles

The location of the anterior papillary muscle (APM) plays a key role in the development of TVI. In the normal heart, the angle (lateral tethering angle) between the plane of the TV leaflets and the chordae/APM at mid/end systole is approximately 90°, allowing optimal coaptation between the valve leaflets [Figure 4a].

As TVI develops in HLHS, the APM may shift leftward creating a lateral tethering angle of approximately 100° [Figure 4b]. If this compensatory shift of the APM does not occur, or reverses [Figure 4c], the lateral tethering angle remains close to, or becomes less than, 90°.^[9,15,27]

Echocardiographic evaluation of the tricuspid valve Optimal understanding of the TV anatomy and function is achieved by combining the information obtained from both 2DE and 3DE interrogation.



Figure 4: (a) In the normal heart, the lateral tethering angle at end systole is approximately 90° which allows optimal coaptation of the valve leaflets. (b) In hypoplastic left heart syndrome (HLHS) with mild tricuspid valve insufficiency (TVI), the lateral tethering angle is approximately 100° which tethers the anterior/posterior leaflets more medially optimizing leaflet coaptation. (c) In HLHS and severe TVI, the lateral tethering angle is approximately 90° or less pulling anterior/posterior leaflets away from the septal leaflet

2DE imaging

To maximize the information obtained from 3DE interrogation, a thorough 2DE evaluation of the TV should be performed. During 2DE imaging, the TV should be interrogated from all standard imaging planes (parasternal long axis, parasternal short axis, apical four chamber, subcostal). Particular attention should be given to determining the annular diameter, leaflet mobility, anatomic leaflet abnormalities (leaflet tethering, prolapse, flail leaflet, chordal prolapse/ rupture), valvar stenosis, degree of valvar insufficiency where the insufficiency jet/jets are coming through the valve, and the lateral tethering angle of the APM.

3DE imaging

The understanding of the TV gleaned from the 2DE analysis serves as a guide to maximize the information obtained from the 3DE interrogation. Contemporary 3DE interrogation of the TV in HLHS is performed using transthoracic 3DE probes since most patients are too small to have 3D transesophageal echocardiography. Accurate 3DE recognition of the mechanism of TVI is facilitated by knowing where the insufficiency jet courses through the valve and can be determined with color Doppler interrogation using both 2DE/3DE.

Obtaining the 3DE images

Methods for obtaining quality 3DE images have been previously described^[29,30] and only a brief description of how the valve should be imaged will be given here. 3DE interrogation of the TV in HLHS should be performed from four different imaging planes: Parasternal long axis, parasternal short axis, apical four chamber, and subcostal sagittal [Figure 5].

We begin by imaging the TV with 2D interrogation in the parasternal long axis or parasternal short axis



Figure 5: 3DE interrogation of the tricuspid valve (TV) should be performed in (a) the parasternal long axis imaging plane with the sound beam directed toward the TV, (b) the parasternal short axis imaging plane at the level of the TV, (c) the apical four-chamber imaging plane, and (d) the subcostal sagittal imaging plane with the sound beam directed toward the TV

imaging plane [Figure 6 — Top Panel]. 3D images are then obtained using Live 3D, 3D zoom, or a full volume data set. The 3D image is then cropped to just above the level of the tricuspid annulus and rotated (from the bottom of the image toward the top of the screen) along the X-axis until the valve is seen en face; the valve is then oriented (see discussion below) such that the plane of the septal leaflet lies along a line running from 9:00 o'clock to 6:00 o'clock when viewing the valve from the RA (surgeon's view).

This is followed by 2D interrogation in the apical fourchamber imaging plane [Figure 6 — Bottom Panel] and 3D images are again obtained using Live 3D, 3D Zoom, or full volume acquisition. The 3D image is then cropped to just above the level of the TV annulus. The cropped image is then rotated (from the top of the image toward the bottom of the screen) along the X-axis until the TV is seen en face. The image is then rotated 180° along the Z-axis and the rotation is adjusted such that the plane of the septal leaflet lies along a line running from 9:00 o'clock to 6:00 o'clock when viewing the valve from the RA (surgeon's view).

In any of the imaging planes discussed above, the valve can be viewed from the right ventricular aspect by rotating the image 180° along the Y-axis; the plane of the septal leaflet is then oriented so that it lies along a line running from 3:00 o'clock to 6:00 o'clock. When using Live 3D acquisition, the valve will initially be seen on the half-edge and the interrogation beam will need to be steered using the Elevation Position and Lateral Position so that the entire valve can be seen en face.

In our experience, the best images of the TV are generally obtained from either the parasternal long axis or apical four-chamber imaging plane with either Live 3D imaging or full volume acquisition with subsequent cropping.



Figure 6: The tricuspid valve (TV) is first imaged with 2D interrogation; 3D images are then obtained using Live 3D, 3D Zoom, or a full volume data set. The top panel shows how the 3D image is oriented when the valve is interrogated from the parasternal long axis or parasternal short axis imaging plane. The bottom panel shows how the 3D image is oriented when the valve is interrogated from the apical four-chamber imaging plane. See text for complete explanation

Displaying and analyzing the 3DE images of the tricuspid valve

Leaflet identification

Given the numerous and varying anatomic descriptions of the TV, adequately trying to describe the valve leaflets during 3DE appears to be a formidable task. Describing the valve leaflets can be simplified by understanding that the TV has been shown to open and close with three zones of apposition. Since these zones of apposition serve as junctions between the valve cusps, most TVs can be considered to be functionally trileaflet.^[10] We divide the valve leaflets into three regions during 3DE imaging: Anterior region — occupied by the anterior leaflet, and the septal region — occupied by the septal leaflet [Figure 7].

Not all valves will have three distinct regions. There may be no discernible division between the posterior/ septal leaflets and this should be described as a bileaflet TV [Figure 8a]. If there is a deep cleft in the apex of the anterior leaflet,^[14] it is essentially divided into two separate leaflets (anterior leaflet, anteroseptal leaflet) and this should be described as a quadraleaflet TV [Figure 8b]. We consider all other TVs to be trileaflet with clefts (discontinuation in the leaflet that extends to the annulus^[29]) or indentations (discontinuation of the leaflet (s).

Displaying the 3DE image

Spatial orientation with 2DE is determined by external landmarks and transducer position. Orientation with 3DE is determined by the operator and is largely independent of these factors.

In the literature published about 3DE imaging of the TV in congenital heart disease and HLHS, there has been no consensus of how the TV should be displayed. Some authors display the valve with markedly different orientation, even within the same article.^[7,9,15,28]

In HLHS, we feel that the TV is best displayed in the right atrial or "surgeons view." To determine this, we placed a picture of a pathologic specimen of the TV on a computer screen and rotated until it was oriented as it appears during surgery. In this view, the plane of the septal leaflet runs from 9 o'clock to 6 o'clock [Figure 9]. This orientation, which differs from a normal heart, is, in our opinion, due to clockwise rotation of the RV secondary to the paucity of left ventricular mass and suspension of the right atrial walls.

Analyzing the 3D images to determine the mechanism of TVI To determine the mechanism of TVI, we recommend systematically evaluating the 3DE images of the TV from the annulus to the chordae as follows.

The annular shape, dimensions, and area should is determined [Figure 10].



Figure 7: Two different pathologic specimens of the tricuspid valve (TV) with the anterior, posterior, and septal regions outlined. During 3DE imaging, we consider the TV to have three regions: Anterior (A: Red outline), posterior (P: Black outline), and septal (S: Blue outline). Note that although the regions may vary in size, most valves will have three distinct regions. Right atrial view



Figure 8: 3DE image (right atrial, RA, view) of a bileaflet (a) and quadraleaflet (b) tricuspid valve (TV). Key: A: Anterior leaflet, AL: Anterolateral leaflet, P: Posterior leaflet, S: Septal leaflet



Figure 9: Surgical view (right atrial view) of the tricuspid valve (TV) in hypoplastic left heart syndrome (HLHS). See text for details

The valve leaflets are identified to determine the number of leaflets and any anatomic abnormalities (clefts/ indentations) that may be present [Figures 7 and 11].

We recommend observing leaflet motion from the RA and RV views in both real time and frame by frame to determine:

- 1. Leaflet anatomy,
- 2. Identification of any clefts or indentations, and
- 3. The location of the coaptation defect [Figure 12].

Chordal length and thickness is evaluated to determine if there is any leaflet tethering by examining the valve



Figure 10: 3DE imaging allows determination of the annular shape (a: Circular, b: Elliptical, c: Irregular), area, and the anteriorposterior, septal to free wall dimensions (b)



Figure 11: 3D echocardiographic images of two different tricuspid valve's (TV's) demonstrating a cleft in the anterior leaflet (arrow)



Figure 12: Leaflet anomalies resulting in coaptation defects. In (a), there is a cleft in the anterior leaflet (arrow); when the valve closes there is a coaptation defect where the cleft in anterior leaflet contacts the posterior leaflet (b,c black arrow); (b) right atrial (RA) view, (c) right ventricular (RV) view. This patient had mild to moderate insufficiency through the defect caused by the cleft (b, black arrow) and a trivial insufficiency jet through a small coaptation defect at the anteroseptal commissure (b, white arrow). (d) demonstrates a quadraleaflet tricuspid valve (TV). In systole, there is a large diamond-shaped coaptation defect in the central aspect of the valve between the four leaflets (e,f white arrow); (e) right atrial (RA) view, (f) right ventricular (RV) view. The patient had moderate to severe insufficiency through this coaptation defect. A: Anterior leaflet, P: Posterior leaflet, S: Septal leaflet, AL: Anteriolateral leaflet

support apparatus from the right ventricular view [Figure 13].

The APM lateral tethering angle is determined. This can be approximated from a 2DE image of the RV from the apical four-chamber imaging plane [Figure 14].

The information obtained from the 2DE/3DE analysis of the TV can be used to determine the mechanism of TVI and this understanding can be very valuable in planning the surgical repair for significant TVI. For example, if the mechanism of TVI is limited to annular dilation resulting in inadequate coaptation of the valve leaflets, then a tricuspid valvuloplasty may be all that is needed to repair the valve.

On the other hand, if there are leaflet anomalies (indentations, clefts), these may need to be addressed with or without a valvuloplasty in order to achieve a competent valve. Chordal abnormalities may necessitate an even more complicated surgical repair.

The surgical repair for significant TVI in quadracuspid valves, in our experience, has been challenging with unsatisfactory results obtained from a straightforward tricuspid annuloplasty. In our opinion, surgical repair in these complex valves should be undertaken only after very careful planning.

An understanding of the mechanism of significant TVI in HLHS allows for adequate surgical preparation which should optimize valve repair and patient outcome.

DISCUSSION

Significant TVI occurs frequently in patients with HLHS and TV repair may be required at any stage of the palliation. Although the data obtained from 2DE provide important information, it is often inadequate in accurately describing valvar pathology. These deficiencies can be overcome with 3DE which substantially improves imaging of the TV and may provide a more in-depth understanding of valve anatomy and greater insight into the mechanism of insufficiency.

Our orientation of the 3DE image of the TV differs slightly from that proposed by Lang *et al.*^[29] where the septal leaflet is located at the 6:00 o'clock position with the plane of the leaflet running from 3:00 o'clock to 9:00 o'clock. Although this orientation is adequate in the normal heart, in HLHS we prefer orienting the valve as the surgeon actually sees it (9:00 o'clock to the 6:00 o'clock position). In our opinion, this orientation more accurately reflects the anatomic orientation of the TV in HLHS leading to better understanding of valvar pathology.

We have proposed a systematic method for evaluating and displaying the TV using 3DE which can provide significant



Figure 13: 3D echocardiographic image (right ventricular view) of normal tricuspid valve (TV) chordae (a: Red arrows). Short, thickened chordae (b: Red arrows) that significantly tethered and decreased the mobility of the septal leaflet (red line runs along the ventricular septum) resulting in a large coaptation defect (white arrow) between the septal and anterior/posterior leaflets. A: Anterior leaflet, P: Posterior leaflet, S: Septal leaflet

insight into the mechanisms causing TVI in HLHS. This has the potential to improve both the surgical approach to repairing the valve and ultimately, patient outcomes.

The images of the tricuspid valve used in Figures 2,3,7 (second image), and 9, were generously provided by Paul A. Iaizzo from the University of Minnesota and are available at the website: http://www.vhlab.umn.edu/atlas/right-atrium/tricuspid-valve/index.shtml. Used with permission.

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Figure 14: The lateral tethering angle is the angle between the plane of the tricuspid valve (TV) and the anterior papillary muscle (APM) at end systole. This patient, with a lateral tethering angle of 71°, had moderate TV insufficiency (TVI) requiring surgical repair

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