

Case Report

The Use of 3-Dimensional Echocardiography for Tricuspid Valve Surgery in Hypoplastic Left Heart Syndrome

Andrea Pisesky, MD,^a Osami Honjo, MD, PhD,^b Olivier Villemain, MD, PhD,^a and Andreea Dragulescu, MD, PhD^a

^a The Labatt Family Heart Centre, Department of Pediatric, the Hospital for Sick Children, Toronto, Ontario, Canada

^b Division of Cardiovascular Surgery, the Hospital for Sick Children, Toronto, Ontario, Canada


We present a case of a patient with hypoplastic left heart syndrome requiring tricuspid valve repair due to anterior leaflet prolapse. The use of 3-dimensional echocardiography enabled delineation of the primary and secondary mechanisms responsible for valvar regurgitation: an abnormally positioned, dominant posteromedial papillary muscle and a tethered septal leaflet. With this information, the surgical team modified their approach, proceeding with the placement of 10 artificial chords in the anterior leaflet. At the 6-month follow-up appointment, there was no valvar regurgitation or stenosis.

Tricuspid valve regurgitation is associated with significant morbidity and mortality for patients with hypoplastic left heart syndrome.¹ In this population, tricuspid valve regurgitation is often complex and multifactorial, involving both structural and functional abnormalities.^{2,3} Thus, the preoperative assessment of the mechanisms of valvar dysfunction is critical to personalizing the surgical strategy and achieving an optimal result. Three-dimensional echocardiography has emerged as an invaluable tool in valvar heart disease enabling dynamic evaluation of the spatial and functional relationship between all of the supporting valvar structures.⁴ It has not yet been widely adopted in the single ventricle population with previous studies primarily demonstrating technical feasibility.⁵

Case Report

A 6-month-old boy with hypoplastic left heart syndrome after stage 1 palliation with the Norwood procedure presented with recurrent symptoms of low cardiac output due to severe tricuspid valve regurgitation. After stabilization, cardiac catheterization and magnetic resonance imaging demonstrated

favourable haemodynamics and anatomy. We thus proceeded with a bidirectional cavopulmonary connection in conjunction with a tricuspid valve repair.

The patient's 2-dimensional echocardiogram had identified annular dilatation and anterior leaflet prolapse with elongated chords, which were thought to be the primary and secondary mechanisms of regurgitation, respectively. Consequently, the tentative surgical plan was to assess the valve and perform a commissuroplasty and an annuloplasty. Before starting the operation, a perioperative 3-dimensional echocardiogram was performed with the surgical team using a Philips 5X-1 (Philips, Bothell, Washington) 3-dimensional probe to delineate these mechanisms further. We visualized that the primary mechanism of regurgitation was the redundant and prolapsing anterior leaflet with elongated chords and bright papillary muscles and a tethered septal leaflet (Fig. 1; Video 1,  view video online). Intraoperatively, the surgical team additionally identified dysplasia of the septal-posterior commissure. Integrating the 3-dimensional echocardiograph and intraoperative findings, the surgical team modified their approach, proceeding with the placement of 10 artificial chords along in the free edge of the anterior leaflet using 7-0 polytetrafluoroethylene sutures (GORE-TEX Suture; W. L. Gore & Associates, Inc. DL) to prevent prolapse, partial closure of the dysplastic septal-posterior commissure, and aggressive annuloplasty on the posterior annulus, along the anteroseptal and anteroposterior commissures. Recovery was uncomplicated, and there was no significant tricuspid regurgitation or stenosis at the 6-month follow-up appointment.

Comment

The use of intraoperative 3-dimensional echocardiography has been shown to be valuable in complex and rheumatic mitral valve repair;⁴ however, its role in the single ventricle population and in tricuspid valve surgery has not been elucidated. We describe a case of hypoplastic left heart syndrome with severe tricuspid regurgitation whereby the intraoperative 3-dimensional echocardiogram identified additional structural abnormalities necessary to address surgically. Fully

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Corresponding author: Dr Andrea Pisesky, Division of Cardiology, the Hospital for Sick Children, 555 University Ave, Toronto, Ontario M5G 1X8, Canada. Tel.: +1-416-813-7500.

E-mail: Andrea.pisesky@sickkids.ca

Novel Teaching Points

- In hypoplastic left heart syndrome, atrioventricular valvar regurgitation is typically multifactorial.
- Surgical planning should address all mechanisms responsible for valvar regurgitation.
- Three-dimensional echocardiography should routinely be used in surgical planning for atrioventricular valvar regurgitation in hypoplastic left heart syndrome.

assessing the tricuspid valve is likely the best when it involves multiple modalities due to the specific blind spots of each modality and the anatomic complexity of the tricuspid valve and right ventricle. First, our patient's preoperative 3-dimensional echocardiogram identified anterior leaflet prolapse; however, it was not possible to understand its mechanism. As such, the value of 3-dimensional echocardiography was intuitive because we could capture the entire annulus, all 3 leaflets, and the supporting structures in one view. By visualizing the entire perivalvar apparatus, we observed that the abnormally positioned papillary muscle caused tethering of the septal leaflet and laxity of the anterior leaflet, both of which were responsible for the prolapsing anterior leaflet. Before 3-dimensional imaging techniques, understanding 3-dimensional structures, such as papillary muscles, required extrapolation from 2-dimensional images, a strategy inherently inaccurate and challenging to communicate. Our case highlights how visualizing the abnormally shaped and positioned papillary muscle in one 3-dimensional volume facilitated an understanding of the implications of this pathology and a collaborative approach to surgical planning. Furthermore, surgical assessment has inherent limitations because it occurs in the setting of an unloaded heart; it is thus not surprising that there is poor agreement between echocardiographic and surgical assessment. In our case, surgical assessment would not have been able to understand the functional

implications of the tethered leaflet and abnormal papillary muscle; however, the surgeons were able to identify additional anomalies that required repair: elongated anterior leaflet chords and a dysplastic septal-posterior commissure, neither seen using 2- or 3-dimensional echocardiography. Atrioventricular regurgitation in the single ventricle population is typically complex, involving multiple structural abnormalities in addition to the invariably present annular dilatation. In our experience, addressing only one valvar mechanism does not typically achieve a competent valve. As such, we aim to identify and surgically address all primary and secondary mechanisms of valvar regurgitation, a strategy that relies on using multiple diagnostic modalities.

Our case highlights how the most accurate assessment of the tricuspid valve required the integration of all available modalities. We had initially considered several surgical options based on the preoperative 2-dimensional imaging; however, visualization of the entire valve structure in 3-dimensional volumes with a beating heart allowed the surgical team to individualize their approach to the anatomy found on direct inspection. Given the poor outcomes associated with hypoplastic left heart syndrome with atrioventricular valve regurgitation, we believe that 3-dimensional echocardiography should be considered part of routine surgical planning, particularly when complete in the operating room in close collaboration with the surgical team.

Ethics Statement

The research reported has adhered to the relevant ethical guidelines.

Patient Consent

The authors confirm that a patient consent form has been obtained for this article.

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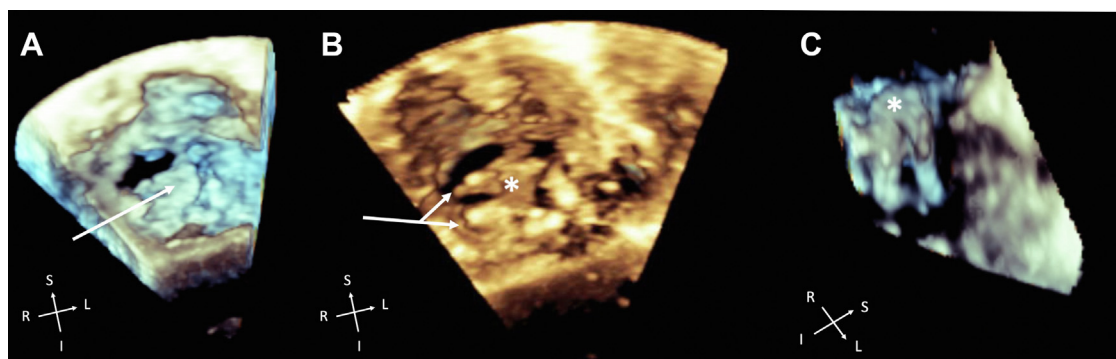


Figure 1. (A) En face view of the tricuspid valve from the right atrial side demonstrating the prolapse of the anterior tricuspid valve leaflet (arrow). The incomplete circle delineates the previous ring annuloplasty. (B) En face view of the tricuspid valve from the ventricular side demonstrating the dichotomous, papillary muscle (arrows) supporting the anterior tricuspid valve leaflet and the commissure between the anterior and inferior leaflets; the chords appear elongated resulting in the prolapse of the anterior tricuspid valve leaflet (*). (C) Right lateral view of the tricuspid valve focusing on the anterior leaflet support apparatus demonstrating the dichotomous papillary muscle (arrows) supporting the anterior tricuspid valve leaflet with elongated chords resulting in the prolapse of the anterior tricuspid valve leaflet (*). I, inferior; L, left; R, right; S, superior.

Disclosures

The authors have no conflicts of interest to disclose.

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