

Use of echocardiographic subxiphoid five-sixth area length (bullet) method in evaluation of adequacy of borderline left ventricle in hypoplastic left heart complex

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ABSTRACT

Indexed left ventricular end-diastolic volume (ILVEDV) is commonly used in evaluating “borderline left ventricle (LV)” in hypoplastic left heart complex (HLHC) to determine if the LV can sustain adequate systemic cardiac output. Commonly used quantification methods include biplane Simpson or the traditional five-sixth area length “bullet” methods, which have been shown to underestimate true LV volumes, when septal position is mildly abnormal. Subxiphoid five-sixth area length method is proposed as a more accurate estimate of true LV volume in the evaluation of borderline LV.

Keywords: Echocardiography, hypoplastic left heart syndrome, left ventricle

INTRODUCTION

Hypoplastic left heart complex (HLHC) describes a wide spectrum of small left heart structures. Patients with severe hypoplasia of left heart structures are managed with univentricular repair (UVR), while those with mild hypoplasia of left heart structures undergo some form of biventricular repair (BVR). Patients in the “gray zone”, with so-called “borderline left ventricle (LV)”, present a dilemma forcing a dichotomized management decision of UVR versus BVR early in life. The inappropriate pursuit of BVR in borderline LV has been shown to be more consequential than is inappropriate pursuit of UVR,^[1] emphasizing the importance of an accurate predictive model of borderline LV.

Existing criteria to evaluate borderline LV in HLHC do not reliably identify neonates who will thrive with a BVR,^[2] with many criteria based on studies of infants with critical aortic stenosis.^[3,4] Echocardiography is the main diagnostic tool used to predict the adequacy of the LV in supporting systemic circulation. Established criteria used

in deciding the adequacy of the LV to sustain BVR include morphometric and functional parameters, hemodynamic data, available surgical options, along with results of institutional experience.^[5] Indexed left ventricular end-diastolic volume (ILVEDV) is commonly used in this decision-making process, with many studies quoting an ILVEDV less than 20 mL/m² predicting poor survival with BVR.^[6-8] LVEDV is commonly calculated using the biplane Simpson or traditional five-sixth area length “bullet” methods, using apical and parasternal views.^[9] These methods in patients with HLHC tend to underestimate true LV volumes as the volume-loaded RV may compress the under-filled LV.^[10] Subxiphoid “bullet” method has been validated against cardiac magnetic resonance imaging (CMR), showing excellent reproducibility and agreement with established normal values in pediatric patients aged 0-3 years.^[11,12] We describe a case of borderline LV who underwent successful BVR, for whom preoperative echocardiographic LV volumes estimation by the traditional biplane Simpson and standard “bullet” methods showed prohibitive size, but had significantly improved estimation by both subxiphoid “bullet” method and CMR, with the latter methods having close agreement.

CASE REPORT

The patient is a full-term newborn with suspicion of congenital heart disease on a technically limited late gestation fetal echocardiogram. Initial transthoracic

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echocardiogram showed a left sided aortic arch with aberrant right subclavian artery, hypoplastic transverse aortic arch (4 mm, Z-score: -4.4), and discrete coarctation of the aortic isthmus (2 mm). There were mildly hypoplastic unobstructed mitral and bicuspid aortic valves (8 mm, Z-score: -2 and 4.6 mm, Z-score: -4.1, respectively). The LV was moderately hypoplastic and not apex forming, with LV-to-RV long axis ratio of 0.8:1. There was no evidence of endocardial fibroelastosis. LV assessment by Simpson's biplane method showed an ILVEDV of 13 mL/m². Traditional "bullet" method showed an ILVEDV of 21 mL/m². Retrospectively, subxiphoid "bullet" method showed an ILVEDV of 30 mL/m² [Figure 1].

The patient subsequently underwent CMR to better define the borderline LV. Again seen was a significant discrepancy between the LV-to-RV long axis dimension, with a ratio 0.7:1. However, the ILVEDV measured normal at 32 mL/m² (Z-score: -1.1) [Table 1]. CMR LVEDV was reassuring, given the prior underestimation of LVEDV on echocardiogram, and with the available data and clinical picture, it was decided to pursue BVR. The patient underwent aortic arch reconstruction with a homograft patch. Postoperatively, the left ventricle showed evidence of diastolic dysfunction (which subsequently improved) and the LV was adept to handle the systemic circulation.

DISCUSSION

Accurate assessment of infants with borderline LV is crucial in determining the optimal management strategy. Echocardiography derived ILVEDV is commonly used to aid in decision-making in these infants.^[6-8] While standard echocardiographic assessment of LV volume in our patient raised questions about its ability to sustain systemic output with BVR, the subxiphoid "bullet" method and CMR demonstrated much improved estimation of LV size.

A study by Grosse-Wortmann *et al.*,^[10] showed that echocardiography consistently and significantly underestimated LVEDV and correlated poorly with CMR. If they had made decisions based off of standard echocardiographic LVEDV assessment, more than half of their patients who had successfully undergone BVR would have undergone UVR. They also evaluated the concept of observed versus potential preoperative volumes in the setting of volume-loaded RV, which was introduced by Phoon and Silverman,^[13] calculating an average potential volume increase of 23% in their borderline LV patients with HLHC.^[10]

The subxiphoid "bullet" method of estimating LVEDV may have advantages in the setting of mildly abnormal ventricular septal position or limited parasternal acoustic windows.^[12] The subxiphoid window tends to have better

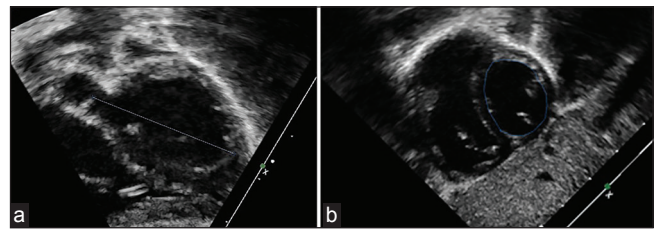


Figure 1: Subxiphoid five-sixth area length echocardiographic assessment of left ventricular (LV) volume. (a) Subxiphoid long axis diastolic frame with aortic annulus to LV apex measurement. (b) Subxiphoid short axis diastolic frame at the midventricular level with area measurement

Table 1: Comparison of Indexed LVEDV

	CMR	Subxiphoid fifth-sixth area length	Traditional fifth-sixth area length	Biplane Simpson
Indexed LVEDV (mL/m ²)	32	30	21	13

LVDEV: Left ventricular end-diastolic volume, CMR: Cardiac magnetic resonance imaging

definition of the LV endocardial border. The ability to estimate the true LV length is much better by the subxiphoid "bullet" method, especially in the setting of a large right ventricle wrapping the LV apex. Moreover, because of the saddle shape of the mitral valve, Nielsen *et al.*,^[11] proposed and validated that the use of apex-to-LV outflow length, yielded a larger LVEDV than the apex to mitral valve measurement used in the traditional "bullet" method. Our patient's ILVEDV by subxiphoid "bullet" method not only was larger and more reassuring than traditional methods, it also correlated better with CMR.

In conclusion, the subxiphoid "bullet" method may be more accurate in assessing true LV volumes than other more commonly used echocardiographic methods, especially in the setting of HLHC with borderline LV. Even so, if discrepant or borderline measurements are obtained by echocardiographic assessment, CMR should be obtained to help determine the best treatment pathway.

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