Prediction of biventricular repair by echocardiography in borderline ventricle

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Abstract

Objective: In recent years, attempting the biventricular pathway or biventricular conversions in patients with borderline ventricle has become a hot topic. However, inappropriate pursuit of biventricular repair in borderline candidates will lead to adverse clinical outcomes. Therefore, it is important to accurately assess the degree of ventricular development before operation and whether it can tolerate biventricular repair. This review evaluated ventricular development using echocardiography for a better prediction of biventricular repair in borderline ventricle.

Data sources: Articles from January 1, 1990 to April 1, 2019 on biventricular repair in borderline ventricle were accessed from PubMed, using keywords including "borderline ventricle," "congenital heart disease," "CHD," "echocardiography," and "biventricular repair."

Study selection: Original articles and critical reviews relevant to the review's theme were selected.

Results: Borderline left ventricle (LV): (1) Critical aortic stenosis: the Rhodes score, Congenital Heart Surgeons Society regression equation and another new scoring system was proposed to predict the feasibility of biventricular repair. (2) Aortic arch hypoplasia: the LV size and the diameter of aortic and mitral valve (MV) annulus should be taken into considerations for biventricular repair. (3) Right-dominant unbalanced atrioventricular septal defect (AVSD): atrioventricular valve index (AVVI), left ventricular inflow index (LVII), and right ventricle (RV)/LV inflow angle were the echocardiographic indices for biventricular repair. Borderline RV: (1) pulmonary atresia/intact ventricular septum (PA/IVS): the diameter *z*-score of tricuspid valve (TV) annulus, ratio of TV to MV diameter, RV inlet length *z*-score, RV area *z*-score, RV development index, and RV-TV index, etc. Less objective but more practical description is to classify the RV as tripartite, bipartite, and unipartite. The presence or absence of RV sinusoids, RV dependent coronary circulation, and the degree of tricuspid regurgitation should also be noted. (2) Left-dominant unbalanced AVSD: AVVI, LV, and RV volumes, whether apex forming ventricles were the echocardiographic indices for biventricular repair.

Conclusions: Although the evaluation of echocardiography cannot guarantee the success of biventricular repair surgery, echocardiography can still provide relatively valuable basis for surgical decision making.

Keywords: Borderline ventricle; Congenital heart disease; Echocardiography; Biventricular repair

Introduction

Borderline ventricle refers to a spectrum of left or right ventricular underdevelopment, usually associated with severe congenital heart disease (CHD). Typically, borderline left ventricle (LV) is common in the following CHD, including severe LV outflow tract obstruction, aortic arch dysplasia, right-dominant unbalanced atrioventricular septal defect (AVSD), mitral valve (MV) malformation, Shone's complex, and total anomalous pulmonary venous connection,[1,2] while borderline right ventricle (RV) often associates with severe RV outflow tract obstruction, pulmonary atresia/intact ventricular septum (PA/IVS), left-dominant unbalanced AVSD, and Ebstein's anomaly.[3,4]

In the patients with borderline ventricle, it is hard to make a decision between a biventricular repair and the Fontan-type operation. Patients being considered for the Fontan-type operation typically have only one effective ventricle, either left or right, due to a multitude of reasons. Univentricular repair is a relatively conservative option in the borderline ventricle. But, following Fontan-type operation, some complications may occur, including cardiac insufficiency, cyanosis and hypoxia, arrhythmia, protein-loss enteropathy, plastic bronchitis, hepatic insuf-

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ficiency, thrombosis and embolism, etc, which will lead to poor prognosis.[5-10] In recent years, attempting the biventricular pathway or biventricular conversions in patients with borderline ventricle has become a hot topic. However, inappropriate pursuit of biventricular repair in borderline candidates will lead to adverse clinical outcomes.[11] Therefore, it is important to accurately assess the degree of ventricular development before operation and whether it can tolerate biventricular repair. We reviewed the evaluation of ventricular development using echocardiography for a better prediction of biventricular repair in borderline ventricle.

Borderline Left Ventricle

Critical aortic stenosis

In critical aortic stenosis (AS), the aortic valve is severely narrowed such that a patent ductus arteriosus (PDA) is required to support the systemic circulation; it is frequently associated with LV hypoplasia and dysfunction.[4] Besides, endocardial fibroelastosis (EFE) is often seen in the patients with critical AS, further causing LV dysfunction.[4,12]

To predict the feasibility of biventricular repair in critical AS, several prediction models have been proposed. As early as 1991, Rhodes *et al*[13] put forward a biventricular repair predictive equation in critical AS: Score = 14.0 (BSA) + 0.943 (ROOTi) + 4.78 (LAR) + 0.157 (MVAi) − 12.03. Among which, BSA = body surface area, ROOTi = aortic root dimension indexed to BSA, LAR = ratio of the long-axis dimension of LV to long-axis dimension of heart, and MVAi = MV area indexed to BSA, with a discriminating score of less than –0.35 predictive of death after a biventricular repair. This predictive equation is well known as Rhodes score. In 2001, the Congenital Heart Surgeons Society (CHSS)[14] proposed a regression equation to predict 5-year survival benefit of univentricular repair *vs.* biventricular repair as follows: Survival benefit = Intercept + b1 (age at entry) + b2 (*z*-score of aortic valve at the sinuses) + b3 (grade of EFE) + b4 (ascending aorta diameter) + b5 (presence of moderate or severe tricuspid regurgitation) + b6 (*z*-score of the LV length). The CHSS regression equation incorporated EFE and tricuspid regurgitation into the scoring system. The presence of EFE with consequent diastolic dysfunction may be more important than LV volume in determining the outcome.[15] It is worth noting that EFE can be suspected by echocardiography as echo brightness in the LV wall but is better imaged by magnetic resonance imaging (MRI). On MRI, EFE manifested at the endocardial surface as a rim of hypointense signal in the perfusion sequences and as a rim of hyperintense signal in the myocardial delayed-enhancement sequences.[16] Later on, another scoring system was devised by Colan SD[17]: Score = 10.98 (BSA) + 0.56 (aortic valve annulus z-score) + 5.89 (LAR) – 0.79 (presence of grade 2 or 3 EFE) – 6.78. Taking a discriminant cutoff of –0.65, this model accurately predicted outcome in 95% of survivors and 80% of events. Based on these predictive models, the size and shape of aortic annulus and MV, size and function of LV, tricuspid regurgitation, and presence of EFE should be taken into account when planning biventricular repair in critical AS. Though these equations and scores are helpful in the decision-making process, because of the complexity of the disease, no clear-cut echocardiographic criteria guarantee success of biventricular repair.[4]

Aortic arch hypoplasia

In aortic arch hypoplasia, the aortic arch is diffusely small, often with coarctation of the aorta; it is frequently associated with LV hypoplasia but without EFE.[4,18]

Because of the different anatomy, the Rhodes score or CHSS regression equation may be not suitable for aortic arch hypoplasia.[19,20] Mart and Eckhauser[18] proposed a predictive model for biventricular repair of aortic arch dysplasia:

$$2\text{V}-\text{score} = \left(\frac{\left(\frac{(\text{MV}_{4\text{C}}/\text{AV}_{\text{PSLA}})}{(\text{LVL}_{4\text{C}}/\text{RVL}_{4\text{C}})} + \text{MPA}\right)}{\text{BSA}}\right)$$

where MV_{4C} is the MV annulus measured in the apical 4-chamber sectional view, AV_{PSLA} is the aortic valve annulus measured in the parasternal long axis sectional view, LVL_{4C} and RVL_{4C} are the left or right ventricular length measured in the apical four-chamber sectional view, MPA is the main pulmonary artery diameter, and BSA is the body surface area. Using a 2V-score cutoff value of ≤16.2 in their cohort, biventricular repair predicted with both sensitivity and specificity of 100%. But the scoring system needs to be prospectively validated. Another study showed that the combination of MV to tricuspid valve (TV) ratio ≤0.66 with an aortic valve annulus *z*-score ≤–3 had the power to predict biventricular repair failure with the sensitivity 71% and specificity 94%.[21] From these studies, the diameter of aortic and MV annulus and LV size should be taken into considerations for biventricular repair in aortic arch hypoplasia. For all that, there are still clinical uncertainties.

Right-dominant unbalanced atrioventricular septal defect

In right-dominant unbalanced AVSD, the RV is larger, the LV is hypoplastic, and the common atrioventricular valve is located on the right side relative to the interventricular septum. Moreover, the LV outflow tract is vulnerable to obstruction.

To evaluate the degree of ventricular hypoplasia in this situation, several measurement indices have been proposed. Atrioventricular valve index (AVVI) was introduced by Cohen MS[22]: AVVI = left valve area/right valve area. They mentioned that if AVVI was <0.67 in the presence of a large ventricular septal defect, a univentricular repair should be considered. To evaluate the feasibility of biventricular repair, the flow of LV inflow tract is also an important factor. Szwast *et al*[23] proposed the LV inflow index (LVII): LVII = color flow width of the LV inflow/left atrioventricular valve annulus diameter. In cases of mild or moderate LV hypoplasia, a greater LVII predicted survival after biventricular repair. Cohen *et al*[24] proposed the RV/LV inflow angle as another echocardiographic index, a wider RV/LV inflow angle predicted possibility of success after biventricular repair. However, even if the above echocardiographic criteria are met, biventricular repair may still fail because of left atrioventricular valve stenosis or regurgitation, LV outflow obstruction, and other clinical situations.

Borderline Right Ventricle

Pulmonary atresia/intact ventricular septum

In PA/IVS, there is luminal discontinuity between the RV outflow tract (RVOT) and the main pulmonary artery in the absence of ventricular septal defect. There is significant morphologic heterogeneity among patients with PA/IVS,[25,26] from RV hypoplasia to relatively normal RV, even RV dilation.

In PA/IVS with borderline RV, preoperative imaging evaluation is vital for clinical strategy. Several morphologic indices are important for evaluation of RV development, including the diameter z-score of tricuspid annulus, ratio of tricuspid to MV diameter, RV inlet length z-score (tricuspid annulus to apex at end-diastole), RV area z-score at end-diastole, RV development index, RV-TV index, etc.[26,27] Less objective but more practical description is to classify the RV as tripartite (mild hypoplasia), bipartite (moderate hypoplasia), and unipartite (severe hypoplasia). The presence or absence of RV sinusoids, RV dependent coronary circulation, and the degree of tricuspid regurgitation should also be noted.

The features of the PA/IVS with borderline RV include tricuspid annulus z-score –2.5 to –4.5, usually bipartite RV (absent or markedly attenuated trabecular component), patent infundibulum, commonly small pulmonary valve annulus and subvalvar stenosis, commonly minor sinusoids (maybe major sinusoids). In this group of patients, radiofrequency valvotomy and PDA stenting is recommended as the primary procedure. Meanwhile, balloon atrial septostomy may be helpful in reducing venous congestion. Some patients may achieve RV well growth without further interventions, or just requiring device closure of atrial shunt to relieve cyanosis. Others need re-interventions, such as bidirectional Glenn shunt (1½ ventricle repair) for inadequate RV growth, RVOT reconstruction for subvalvar obstruction or small pulmonary annulus, TV repair for severe or progressive regurgitation.[26,27] Conditions of biventricular repair includes those without RV dependent coronary circulation, tricuspid annulus z-score >-3, and ratio of tricuspid to MV diameter >0.5.[28,29] Maskatia *et al*[30] found that the baseline RV area ≥ 6 cm^2/m^2 had 93% sensitivity and 80% specificity for identifying patients with PA/IVS who ultimately achieved biventricular circulation, while all patients with RV area ≥ 8 cm^2/m^2 achieved biventricular circulation.

Left-dominant unbalanced atrioventricular septal defect

In left-dominant unbalanced AVSD, the LV is larger, the RV is hypoplastic, and the common atrioventricular valve is located on the left side relative to the interventricular septum.

As a discriminator of balanced and unbalanced forms of complete AVSD, AVVI is a useful echocardiographic indicator to judge the degree of ventricular hypoplasia, as mentioned in right-dominant unbalanced AVSD. Jegatheeswaran *et al*[31] classified the patients as unbalanced if AVVI ≤ 0.4 (right dominant) or ≥ 0.6 (left dominant). The majority of patients with balanced AVSD ($0.4 < \text{AVVI} < 0.6$) underwent biventricular repair, while the patients with AVVI < 0.19 uniformly underwent univentricular repair. As for those with $0.19 < \text{AVVI} < 0.39$, it is a challenge to make the appropriate decision. Nathan *et al*[32] defined the unbalanced AVSD with mild hypoplasia of ventricle or atrioventricular valve as LV or RV volumes >30 mL/m^2, 60% to 80% overriding, $0.19 < \text{AVVI} < 0.39$ or $0.61 < \text{AVVI} < 0.80$, and apex forming ventricles. In this group of patients, primary biventricular repair or initial pulmonary artery banding/shunting with subsequent biventricular conversion was recommended. The moderate hypoplasia of ventricle or atrioventricular valve was defined as LV or RV volumes 15 to 30 mL/m^2, 60% to 80% overriding, $0.19 < \text{AVVI} < 0.39$ or $0.61 < \text{AVVI} < 0.80$, and near apex forming ventricles. In this group, single ventricular palliation and ventricular recruitment with staged biventricular conversion was recommended.

Summary

It is vital to judge the degree of ventricular development using echocardiography before surgical decision making for this borderline ventricle population. However, even if the above echocardiographic criteria are met, biventricular repair may still fail because of clinical complexity, available surgical options, personal and institutional experience, and other confounding factors and unpredictable events. Despite many uncertainties, the improvement of disease cognition and the introduction and application of new echocardiographic indicators can provide a relative reference for the surgical decision making of this population.

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Conflicts of interest

None.

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