Echocardiography-Derived Left Ventricular Outflow Tract Gradient and Left Ventricular Posterior Wall Thickening Are Associated with Outcomes for Anatomic Repair in Congenitally Corrected Transposition of the Great Arteries

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Background: Congenitally corrected transposition of the great arteries is a rare form of congenital heart disease. Management is controversial; options include observation, physiologic repair, and anatomic repair. Assessment of morphologic left ventricle preparedness is key in timing anatomic repair. This study's purpose was to review the modalities used to assess the morphologic left ventricle preoperatively and to determine if any echocardiographic variables are associated with outcomes.

Methods: A retrospective review of patients with congenitally corrected transposition of the great arteries eligible for anatomic repair at Lucile Packard Children's Hospital from 2000 to 2016 was conducted. Inclusion criteria were (1) presurgical echocardiography, magnetic resonance imaging, and cardiac catheterization and (2) clinical follow-up information. Echocardiographic measurements included left ventricular (LV) single-plane Simpson's ejection fraction, LV eccentricity index, LV posterior wall thickening, pulmonary artery band (PAB)/LV outflow tract (LVOT) pressure gradient, and LV and right ventricular strain. Magnetic resonance imaging measurements included LV mass, ejection fraction, eccentricity index, and LV thickening. LV pressure, PAB/LVOT gradient, right ventricular pressure, pulmonary vascular resistance, and Qp/Qs constituted catheterization data. Outcomes included achieving anatomic repair within 1 year of assessment in patients with LVOT obstruction or within 1 year of pulmonary artery banding and freedom from death, transplantation, or heart failure at last follow-up.

Results: Forty-one patients met the inclusion criteria. PAB/LVOT gradients of 85.2 ± 23.4 versus 64.0 ± 32.1 mm Hg (P = .0282) by echocardiography and 60.1 ± 19.4 versus 35.9 ± 18.9 mm Hg (P = .0030) by catheterization were associated with achieving anatomic repair and freedom from death, transplantation, and heart failure. Echocardiographic LV posterior wall thickening of 35.4 ± 19.8% versus 20.6 ± 15.0% (P = .0017) and MRI LV septal wall thickening of 37.1 ± 18.8% versus 19.3 ± 18.8% (P = .0306) were associated with achieving anatomic repair. Inter- and intraobserver variability for echocardiographic measurements was very good.

Conclusions: PAB/LVOT gradient and LV posterior wall thickening are highly reproducible echocardiographic measurements that reflect morphologic LV performance and can be used in assessing patients with congenitally corrected transposition of the great arteries undergoing anatomic repair. (J Am Soc Echocardiogr 2017;■:■-■.)

Keywords: Congenitally corrected TGA, Echocardiography, Anatomic repair, Outcomes

Congenitally corrected transposition of the great arteries (CCTGA) is a rare form of congenital heart disease with an incidence of one in 33,000 live births, accounting for 0.5% of all congenital heart malformations.[1] CCTGA is characterized by double discordance, a combination of atrioventricular and ventriculoarterial discordance. Presentation in these patients is variable, and symptoms may be secondary to associated malformations, heart block, or systemic right ventricular (RV) failure.[2] Management of this patient population is also variable, with some advocating physiologic repair and others preferring anatomic correction to avoid the anticipated long-term dysfunction of the systemic right ventricle.[3] Success of anatomic repair is dependent on the morphologic left ventricle being prepared to sustain the systemic circulation. Training of the morphologic left ventricle is therefore necessary and has been achieved by temporary banding of the pulmonary artery to increase the left ventricular (LV)

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Abbreviations

AUC = Area under the curve

CCTGA = Congenitally corrected transposition of the great arteries

LV = Left ventricular

LVEF = Left ventricular ejection fraction

LVOT = Left ventricular outflow tract

LVPW = Left ventricular posterior wall

MRI = Magnetic resonance imaging

PAB = Pulmonary artery band

RV = Right ventricular

TGA = Transposition of the great arteries

VSD = Ventricular septal defect

afterload in the absence of adequate pulmonary outflow tract obstruction.[4] Assessment of LV preparedness has been used to determine the timing of anatomic repair, although exact indications and selection criteria remain controversial. At present there is no established imaging standard for determining LV preparedness, because of the rarity of the cardiac lesion as well as center-specific approaches. Cardiac magnetic resonance imaging (MRI) is commonly used to assess the adequacy of the morphologic left ventricle in patients with CCTGA.[5] MRI allows quantification of morphologic LV mass and function. Cardiac catheterization has been used to obtain useful hemodynamic parameters. Echocardiography is commonly used for the assessment of LV structure and function. The abnormal position of the morphologic left ventricle in CCTGA, however, makes conventional methods of echocardiographic evaluation challenging.

The purpose of this study was to review the various modalities used to assess LV preparedness before anatomic repair. This includes echocardiography, MRI, and cardiac catheterization. In addition, we sought to determine the utility of echocardiography as a modality to evaluate the morphologic left ventricle and enhance our understanding of LV mechanics in patients with CCTGA undergoing anatomic repair.

METHODS

Patient Population

A retrospective review of patients with CCTGA, eligible for anatomic repair, at Lucile Packard Children's Hospital, Stanford University, between 2000 and 2016 was conducted. Anatomic repair was defined as (1) the double-switch operation, which combines an atrial baffle procedure with an arterial switch operation or combines hemi-Mustard, Glenn, and arterial switch procedures in patients without LV outflow obstruction,[6,7] or (2) an atrial baffle procedure combined with a Rastelli procedure in patients with ventricular septal defects (VSDs) and LV outflow tract (LVOT) obstruction. Patients were identified from the echocardiography database using Syngo Dynamics workstation (Siemens Medical Solutions USA, Mountain View, CA; Syngo Dynamics Solutions, Ann Arbor, MI) and a prospectively maintained surgical database. Inclusion criteria were (1) a diagnosis of CCTGA, (2) availability of preanatomic repair echocardiographic and magnetic resonance images as well as cardiac catheterization data, and (3) availability of clinical follow-up information. Patients were excluded if CCTGA anatomy precluded anatomic repair and necessitated single-ventricle palliation, imaging was inadequate, or patients were lost to follow-up. The study was approved by the Stanford University institutional review board.

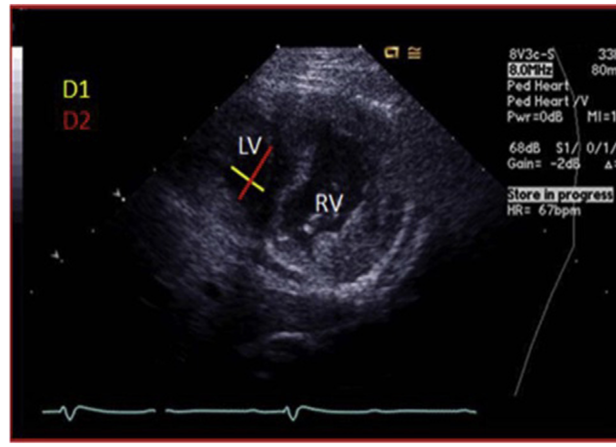

Figure 1 Echocardiography: parasternal short-axis measurements of the morphologic left ventricle to determine LV eccentricity index during systole (D1/D2).

Preanatomic Repair Assessment

Criteria used at our center to determine suitability for anatomic repair are based on echocardiographic, cardiac catheterization, and MRI data. This includes (1) normal LV function and mitral valve function by echocardiography, (2) near systemic LV pressure by cardiac catheterization, and (3) achievement of normal LV mass by cardiac MRI.[8]

Echocardiography

All echocardiographic images were acquired on the Siemens Sequoia C512 (Siemens Medical Solutions) or the Philips iE33 (Philips Medical Systems, Bothell, WA). Postprocessing of echocardiographic images was performed by a single investigator to obtain measurements of morphologic LV structure and function. Measurements included eccentricity index, single-plane Simpson's LV ejection fraction (LVEF),[9] LV posterior wall (LVPW) dimensions, continuous-wave Doppler–derived pulmonary artery band (PAB)/LVOT pressure gradient, and LV and RV strain measurements. Eccentricity index was measured during systole as the ratio of the septolateral distance (D1) and anteroposterior distance (D2) (Figure 1). Two dimensional tracing of the LV endocardial surface in the apical four-chamber view during systole and diastole was done to determine morphologic LV single-plane function using Simpson's method of disks. Because two-chamber views of the morphologic left ventricle were not recorded or not possible to obtain, biplane Simpson's measurements could not be performed. In the parasternal view, two-dimensional measurements were taken of the LVPW using an extension of the line D2 (Figure 1) during diastole and systole. The LVPW systolic-to-diastolic ratio was then calculated to establish thickening. The highest and mean PAB/LVOT gradient by continuous-wave Doppler was determined (Figure 2). Postprocessing using Syngo Velocity Vector Imaging 3.0 software (Siemens Medical Solutions USA), compatible with both the Philips iE33 and Siemens Sequoia C512 ultrasound machines, was used to calculate LV strain. This was performed on echocardiographic loops from the parasternal short-axis view and four-chamber apical view to obtain longitudinal, radial, and circumferential systolic strain imaging of the morphologic left ventricle. Two- and three-chamber apical views of the morphologic left