Right Ventricular Assessment in Adult Congenital Heart Disease Patients with Right Ventricle–to–Pulmonary Artery Conduits

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Background: There is little data on right ventricular (RV) remodeling patterns in complex congenital heart disease (CHD) patients with right ventricle to pulmonary artery (PA) conduits, and novel RV imaging modalities have not been explored in this population. Knowledge of the RV remodeling process is an important first step to future understanding and tracking of the RV response to pressure and volume overload in this diverse population. Three-dimensional knowledge-based reconstruction (3DKBR) derived from two-dimensional transthoracic echocardiography (TTE-3DKBR) is a novel approach to RV assessment. The aims of this study were twofold: (1) to assess the feasibility and accuracy of 3DKBR in patients with CHD with RV to PA conduits and (2) to characterize the three-dimensional shape of the RV across the spectrum of CHD with RV to PA conduits.

Methods: Seventeen patients with tetralogy of Fallot, pulmonary atresia with ventricular septal defect, or truncus arteriosus (mean age, 29 ± 8 years; 24% women) and a conduit referred for cardiac magnetic resonance imaging (CMR) were prospectively recruited and underwent TTE-3DKBR. TTE-3DKBR echocardiographic image acquisition was performed using a standard ultrasound scanner linked to a Ventripoint Medical Systems unit. The surface RV volumetric reconstruction was performed by transmitting two-dimensional data points to an online database and comparing these with a lesion-specific catalog to derive the RV reconstruction. Parameters analyzed were end-diastolic volume (EDV), end-systolic volume, and ejection fraction. Intertechnique agreement was assessed using Pearson's correlation analysis, coefficients of variation, and Bland-Altman analysis. Three-dimensional shape comparisons of RV surface reconstructions were performed via automated validation testing of CMRs from 43 patients (mean age, 30 ± 8 years; 32% women) with RV to PA conduits (tetralogy of Fallot, n = 15; pulmonary atresia, n = 19; and truncus arteriosus, n = 9) distinct from patients in the 3DKBR comparison.

Results: There was good correlation and agreement between the two modalities: EDV, R = 0.77, P = .0004; end-systolic volume, R = 0.93, P < .0001; ejection fraction, R = 0.75, P < .0005. On Bland-Altman analyses, CMR EDV was slightly larger TTE-3DKBR, while EF was slightly higher by 3DKBR. Qualitative and quantitative assessment both demonstrated RV shape diversity based on surface reconstructions.

Conclusion: This study demonstrates that TTE-3DKBR is an alternative technology that can be used to assess the RV in patients with complex CHD with a conduit. A novel method was used to compare RV shapes in this important population, and our results draw specific attention to the fact that the RV both within and outside diagnostic groups has very different unpredictable shapes and should not be treated equally. Our findings should set into motion future work focused on indices of RV shape and their impact on overall RV function and clinical outcomes, hence defining optimal timing of conduit revision, which at the current time is very unclear. (J Am Soc Echocardiogr 2015;28:522-32.)

Keywords: Right ventricle, Congenital heart disease, Conduit, Magnetic resonance imaging

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Abbreviations

AVT = Automated validation testing

CHD = Congenital heart disease

CMR = Cardiac magnetic resonance imaging

EDV = End-diastolic volume

EF = Ejection fraction

ESV = End-systolic volume

PA = Pulmonary artery

RV = Right ventricular/ventricle

RVOT = Right ventricular outflow tract

TA = Truncus arteriosus

3D = Three-dimensional

3DKBR = Three-dimensional knowledge-based reconstruction

TOF = Tetralogy of Fallot

TTE = Transthoracic echocardiography

TTE-3DKBR = Three-dimensional knowledge-based reconstruction derived from two-dimensional transthoracic echocardiography

2D = Two-dimensional

VMS = Ventripoint Medical Systems

VSD = Ventricular septal defect

Surgical placement of a right ventricle (RV)–to–pulmonary artery (PA) conduit plays an important role in the repair of complex congenital heart disease (CHD), specifically in conotruncal abnormalities, in which there is abnormal development of the right ventricular outflow tract (RVOT).[1] Insertion of a RV-PA conduit has improved pulmonary blood flow in defects such as pulmonary atresia with ventricular septal defect (VSD), tetralogy of Fallot (TOF) and allows for a biventricular repair in truncus arteriosus (TA). The critical limitation of RV-PA conduits is the high failure rate over time, leading to symptoms and progressive RV dilatation and dysfunction.[2-5] RV assessment is thought to be important to timing of reintervention, although there are little data to support this notion or to define optimal timing of intervention based on RV parameters. Furthermore, the RV has been assumed to be the same in patients with conduits regardless of the underlying CHD diagnosis. Knowledge of the RV remodeling process is an important first step to future understanding and tracking the RV response to pressure and volume overload in this diverse population. This line of investigation has been pursued in patients with TOF without conduits.[6,7] This work has led to many novel studies, including the investigation of surgical techniques that address RV shape abnormalities to optimize RV function post-surgery.[8] The RV in the complex conduit population has not been studied to date.

Two-dimensional (2D) transthoracic echocardiography (TTE) is limited in providing a quantitative assessment of RV volumes or ejection fraction (EF), as a result, cardiac magnetic resonance imaging (CMR) has become the reference standard. Quantitative three-dimensional (3D) echocardiography has been evaluated as a more economical and facile alternative to CMR. However, several limitations have been observed, including poor endocardial definition and consistent underestimation of RV volumes related to severe enlargement and abnormal remodeling such that the RV extends to outside the transducer imaging volume.[9,10] Generation of a 3D RV model from 2D TTE images is possible with 3D knowledge-based reconstruction (TTE-3DKBR). This method has been validated in vitro and against CMR in patients with TOF, transposition of the great arteries, and in pulmonary hypertension.[6,11,12] Specific anatomic landmarks are identified, and the proprietary reconstruction algorithm uses these landmarks to fit specific regions of the RV to various hearts in a catalog of patients with similar pathology. The algorithm then generates a 3D model based on all the subregions.

The aims of this study were twofold: (1) to assess the feasibility and accuracy of TTE-3DKBR in CHD patients with RV to PA conduits and (2) to characterize the 3D shape of the RV across the spectrum of patients with CHD with RV-PA conduits.

METHODS

Study Design and Population

This was a single-center, prospective, observational study. The study was approved by the institutional research ethics board. Written informed consent was obtained from all patients prior to study enrollment. All patients were ≥18 years of age, had an RV-PA conduit, and had an underlying diagnosis of TOF, pulmonary atresia with VSD, or TA. Patients who were scheduled to undergo CMR for a clinical indication were eligible for inclusion. Exclusion criteria were any contraindication to CMR, including pacemaker or defibrillator, inability to comply with breath-hold instructions, and claustrophobia. Patients with a surgical history of a Rastelli procedure were excluded as the RV-PA conduit 3DKBR catalog did not extend to include this group. Patients were not screened for 2D transthoracic image quality prior to enrollment. Just before or after the CMR, an echocardiographic study was performed, including image acquisition for TTE-3DKBR. A total of 17 patients were prospectively enrolled for this TTE-3DKBR comparison. To determine if RV shape among patients with RV-PA conduit varied by underlying diagnosis, 3D CMR shape comparisons were performed. CMR studies of patients from our program were used for this purpose, for a total of 43 studies from patients with a conduit (TOF, $n = 15$; pulmonary atresia with VSD, $n = 19$; and TA, $n = 9$). These were patients who had a CMR and were not clinically eligible for another CMR and hence were not eligible for enrollment into the TTE-3DKBR comparison as described above. Details pertaining to conduit type, implantation approach, and conduit location were obtained and recorded from operative notes.

3D Knowledge-Based Reconstruction Method

Image Acquisition. Two-dimensional TTE images were acquired with standard ultrasound equipment (iE33 system and S5 transducer; Philips Medical Imaging, Andover, MA). This was connected to a specialized console and used with a magnetic field generator, located underneath the patient bed (Ventripoint Diagnostics Ltd, Seattle, WA). A standard 2D probe was used, with the addition of the magnetic field localizing system (Ascension Technology Corporation, Andover, MA) to track the transducer movements and position relative to the magnetic field transmitter located under the examination bed. The position and orientation of the receiver and thus the plane of the 2D picture can be computed and placed within the volume created by the magnetic transmitter. Images were recorded from the parasternal long-axis, RV inflow and outflow views, parasternal short-axis at both the papillary muscle and apical level apical four-chamber and focused RV apical views, with a focus on endocardial definition of the RV. Additional views were performed to demonstrate the RV-PA conduit. Each image was obtained at end-expiration with the patient in a fixed position throughout the study. The ultrasound scanner is linked to a computer (Ventripoint Medical Systems [VMS]; Ventripoint, Inc, Seattle, WA) through the