Atrial and Ventricular Mechanics in Patients after Fontan-Type Procedures: Atriopulmonary Connection versus Extracardiac Conduit

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Background: Differences in systemic venous flow dynamics and energy losses exist in various Fontan-type procedures, which may affect atrial and ventricular filling. The aim of this study was to test the hypothesis that atrial and ventricular mechanics differ between two types of Fontan procedures, atriopulmonary connection (APC) and extracardiac conduit, which have distinctly different systemic venous hemodynamics.

Methods: This was a cross-sectional, case-control study of 28 Fontan patients (13 with APC, 15 with extracardiac conduit) aged 19.8 ± 6.5 years and 26 healthy controls. Atrial and systemic ventricular myocardial deformation was determined using speckle-tracking echocardiography, while ventricular volumes and systolic dyssynchrony index were assessed using three-dimensional echocardiography.

Results: Compared with controls, patients had significantly lower values of global ventricular longitudinal, circumferential, and radial systolic strain in all three directions, reduced systolic and early diastolic strain rates (SRs) in more than one dimension, lower ejection fractions, and worse ventricular dyssynchrony. For atrial deformation, patients had lower global and positive strain and conduit and reservoir SRs and delayed electromechanical coupling. Among patients, those with APC had significantly lower ventricular longitudinal strain and early diastolic SRs, worse ventricular dyssynchrony, and reduced atrial positive and negative strain and conduit and active contractile SRs. Atrial global strain (r = 0.60, P = .001) and conduit SR (r = 0.49, P = .008) correlated positively with systemic ventricular early diastolic SR.

Conclusions: Atrial and ventricular mechanics are impaired in patients after Fontan-type operation, which is worse with APC than extracardiac conduit. (J Am Soc Echocardiogr 2014;■:■-■.)

Keywords: Atrium, Ventricle, Cardiac mechanics, Speckle-tracking echocardiography, Fontan procedure

Fontan-type procedures involve palliative surgery for patients with functional single ventricles.[1] Since the first report of success in 1971,[2] the initially described operation has undergone several modifications, from atriopulmonary connection (APC) through lateral tunnel to extracardiac conduit (EC) procedures.[3,4,5]

Long-term complications of Fontan-type procedures, in particular in patients with APC, are well documented.[1,6,7] These include atrial arrhythmias, venous thromboembolism, compression of pulmonary veins, protein-losing enteropathy, atrioventricular valvar regurgitation, and progressive systemic ventricular dysfunction. Notwithstanding the relatively smaller number of patients with APC compared with those with lateral tunnel or EC in the present era, these patients are frequently encountered among patients who had functional single ventricles palliated in an earlier surgical era with longer term follow-up. In these symptomatic patients, conversion of the Fontan circuit from APC to EC has been reported to improve functional class, reverse protein-losing enteropathy, and enable better control of refractory cardiac arrhythmias,[8,9,10,11] although these outcomes are not guaranteed. The atrial and ventricular performance in the two different types of Fontan procedures, however, is unclear.

Differences in systemic venous flow dynamics and energy losses exist in various Fontan-type procedures. In APC, progressive dilation of the right atrium causes greater fluid energy losses and increases the energy required to move blood from the caval vein to the pulmonary arteries. By contrast, flow in total cavopulmonary connection and EC is hemodynamically more organized and uniform and hence more efficient.[12,13,14] Differences in systemic venous flow dynamics in various Fontan-type procedures may have implications for atrial and ventricular filling and mechanics. In the present study, we tested the hypothesis that atrial and ventricular mechanics differ between two types of Fontan procedures, APC and EC (Figure 1), which have distinctly different systemic venous hemodynamics. We further explored potential relationships between atrial and ventricular mechanics in Fontan patients.

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Abbreviations

APC = Atriopulmonary connection

EC = Extracardiac conduit

SR = Strain rate

SRcon = Atrial conduit strain rate

METHODS

Subjects

This was a cross-sectional, case-control echocardiographic study of patients who had undergone Fontan-type procedures and healthy, age-matched controls. Of the 32 patients recruited, three with APC were excluded because of a loss of sinus rhythm, and one with EC was excluded because of a suboptimal acoustic window. A total of 28 patients, 13 with APC and 15 with EC, were hence included in the final analysis. The following data were retrieved from the case notes: cardiac diagnosis, age at surgery, history of shunt operation, type of Fontan procedure, existence of fenestration, history of cardiac arrhythmias, current medications, and duration of follow-up since operation. Twenty-six healthy subjects were recruited as controls. These included healthy volunteers; subjects identified in the cardiac clinic with functional murmurs, nonspecific chest pain, and palpitation without an organic cause; and healthy siblings of patients. Body weight and height were measured, and body mass index and surface area were calculated accordingly. In patients, pulse oximetry and electrocardiography were performed. All subjects underwent echocardiographic assessment as described below. The institutional review board approved the study, and all adult subjects and parents of minors gave informed written consent.

Doppler Echocardiographic Assessment

Echocardiographic assessments were performed using the Vivid 7 ultrasound system (GE Vingmed Ultrasound AS, Horten, Norway). The averages of echocardiographic indices measured from three cardiac cycles were obtained for statistical analysis. Color Doppler flow mapping was performed to assess semiquantitatively the degree of systemic atrioventricular valvar regurgitation.[15] Regurgitation was graded as trivial to mild if the regurgitant jet did not cross two thirds of the systemic atrium, moderate if beyond, and severe if the jet reached the posterior wall. Pulsed-wave Doppler examination was performed to obtain the systemic atrioventricular valvar early (E) and late (A) diastolic inflow velocities and E/A ratio. Tissue spectral Doppler imaging was performed with the sample volume positioned at the base of the right-sided and left-sided free wall–atrioventricular valve annular junction in patients and at the ventricular septum and left ventricular free wall–mitral valve annular junction in controls to obtain the following indices: the peak myocardial velocities at systole (s), early diastole (e), and late diastole (a); the e/a ratio; and the E/e ratio.

Assessment of Ventricular Mechanics

Mechanics of the systemic ventricle were assessed using two-dimensional speckle-tracking echocardiography.[16] Two-dimensional echocardiographic recordings were analyzed offline using two-dimensional strain software (EchoPAC; GE Vingmed Ultrasound AS). Deformation of the dominant functional single ventricle in Fontan patients and the left ventricle in control subjects was assessed (Figure 2). Global systemic ventricular longitudinal systolic strain and systolic and diastolic strain rate (SR) were determined from the apical four-chamber view, while circumferential and radial strain and SR were assessed from the midventricular short-axis. The right-sided and left-sided ventricular free walls for functional single ventricle and left ventricular free wall and ventricular septum in structurally normal hearts were divided respectively into three segments (basal, middle, and apical) for quantification of regional longitudinal strain.

Assessment of Atrial Mechanics

Mechanics of the pulmonary venous atrial chamber were determined also using speckle-tracking echocardiography, with tracing of the entire atrial contours.[17] Left atrial deformation was assessed in patients with APC and in controls, while the two atria were regarded as a common atrium in patients with EC because these patients either had right atrial isomerism with almost complete deficiency of atrial septum or had undergone atrial septectomy. The onset of the P wave was used as the reference point for the determination of peak positive strain, peak negative strain, and total strain (Figure 2). The atrial active contractile SR, atrial conduit SR (SRcon), and reservoir SR, were then derived accordingly.[18] Atrial electromechanical coupling was evaluated by measuring the time from onset of the P wave to peak negative strain[19] and normalized to the square root of the RR interval.

Real-Time Three-Dimensional Echocardiography

Full-volume ventricular data sets were acquired from the apical four-chamber view using the matrix-array transducer and analyzed offline with commercial four-dimensional analysis software (TomTec Imaging Systems, Unterschleissheim, Germany). The endocardial borders were traced and used to derive the end-systolic and end-diastolic volumes and ejection fraction. The systolic dyssynchrony index of the systemic ventricle was calculated as the standard deviation of time taken to reach minimum regional volume for each of the 16 ventricular segments as a percentage of the cardiac cycle.[20]

Statistical Analysis

All data are expressed as mean ± SD. Absolute values of strain and SR were used to facilitate interpretation. Ventricular volumes were indexed to body surface area. Intraobserver and interobserver variability was assessed in 10 patients and reported as the coefficients of variation, calculated by dividing the standard deviation of differences between measurements by the mean and expressed as a percentage. Differences in demographic, clinical, and echocardiographic parameters between groups were compared using unpaired Student's t tests, Fisher's exact tests, and χ^2 test as appropriate. Pearson's correlation analysis was used to study relationships between atrial and ventricular deformation parameters and ejection fraction. P values $< .05$ were considered statistically significant. All statistical analyses were performed using SPSS version 16.0 (SPSS, Inc, Chicago, IL).

RESULTS

Subjects

The 28 patients (19 men) aged 19.8 ± 6.5 years were studied 13.4 ± 6.2 years after undergoing Fontan-type procedures. Situs solitus was present in 14 patients, while an abnormal situs was present in the other 14 patients (situs inversus in seven, right atrial isomerism in six, and left atrial isomerism in one). Ventricular-arterial connection was single outlet with pulmonary atresia in 14 patients, concordant in seven, discordant in three, double outlet from the right ventricle in three, and double outlet from the left ventricle in one. Before