Restrictive Right Ventricular Physiology, Right Ventricular Fibrosis as Suggested by Cardiac Magnetic Resonance, and Exercise Capacity After Biventricular Repair of Pulmonary Atresia and Intact Ventricular Septum in Children

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Objective:

Little is known of the prevalence of restrictive RV physiology, extent of RV fibrosis, and the functional implications of RV diastolic dysfunction in long-term survivors of pulmonary atresia and intact ventricular septum (PAIVS) with a biventricular circulation. We tested the hypothesis that right ventricular (RV) restrictive physiology is prevalent and related to RV fibrosis and exercise capacity in patients after biventricular repair of PAIVS.

Materials and Methods:

Twenty-seven patients aged 16.5±5.6 years after biventricular repair of PAIVS were recruited. All patients underwent echocardiography, cardiac magnetic resonance (MR) and treadmill exercise tests. Restrictive RV physiology was defined by presence of antegrade diastolic pulmonary flow and RV fibrosis assessed by late gadolinium enhancement (LGE) cardiac magnetic resonance. All MR examinations were performed by a 1.5T scanner (General Electric). Analysis of the RV and left ventricular (LV) function was obtained by fastcard-SPGR cine on axial and short axis planes, respectively. Flow analysis of the pulmonary artery was obtained by using fast 2D phase contrast and taking a plane perpendicular to the flow direction of the RV outflow tract just above the pulmonary valve but before bifurcation. The ventricular function and flow analyses were performed by using the software in Advantage Window version 4.2. Late gadolinium enhancement imaging was performed from 10 minutes after intravenous injection of gadolinium-DTPA 0.2mmol/kg. The planes included short-axis views from base to apex, standard LV long-axis planes, standard RV long-axis planes, RV oblique planes, and 4-chamber views. To analyze the extent of RV LGE, a scoring system based on division of the right ventricle into 7 segments was adopted. The maximum RV LGE score was 20 (3 for 6 segments and 2 for 1 segment). For the left ventricle, a standard 17-segment model was used. Each of the LV segments was scored on a 5-point scale (0 to 4), and the maximum LV LGE score was 68. The patient's RV function was compared with that of 27 healthy controls and related to RV LGE score and exercise capacity.

Results:

PAIVS patients had lower tricuspid annular systolic and early diastolic velocities, RV global longitudinal systolic strain, systolic strain rate, and early and late diastolic strain rates (all $p<0.05$). Twenty-two (81%, 95%CI 62-94%) PAIVS patients demonstrated restrictive RV physiology. Compared with those without (n=5), these 22 patients had lower RV global systolic strain, lower RV systolic and early diastolic strain rates, higher RV LGE score, and greater % predicted maximum oxygen consumption (all $p<0.05$). The RV LGE score correlated negatively with RV global longitudinal early diastolic strain rate (r=-0.40, p=0.038), and positively with exercise duration (r=0.45, p=0.029) and % predicted maximum oxygen consumption (r=0.43, p=0.042). In terms of cardiac magnetic resonance-derived ventricular volumes and flow, patients with restrictive RV physiology compared to those without had greater forward flow per cardiac cycle across the pulmonary trunk (90.2±33.1 ml vs 60.3±24.5 ml, p=0.045). The forward flow per cardiac cycle further correlated with percent predicted VO_2 max (r=0.42, p=-0.049). Their RV end diastolic volume tended to be smaller (111±44 ml vs 154±55 ml, p=0.10), and LV end diastolic volume (84.5±18.6 ml vs 68.9±14.9 ml, p=0.08) and LV stroke volume (57.5±11.0 ml vs 46.3±12.5ml, p=0.05) tended to be larger than those without restrictive RV physiology.

Table: Late gadolinium enhancement scores of the right ventricle

Ventricular segment	Total score of 27 patients	Mean ± SD
Right ventricle		
anterior wall of outflow	29	1.0 ± 1.0
anterior wall	46	1.6 ± 0.9
inferior wall	33	1.2 ± 1.1
RV surface of septum	4	0.1 ± 0.4
membranous region	1	0.04 ± 0.2
trabecular bands	1	0.04 ± 0.2
RV insertion points	21	0.8 ± 0.5

Conclusion:

Restrictive RV physiology is prevalent in patients after biventricular repair of PAIVS, while the corresponding RV diastolic dysfunction is related to the magnitude of RV fibrosis and associated with better exercise capacity. RV LGE score was a good indicator of clinical outcome of the patients.