

Echocardiographic assessment of coronary flow in the diagnosis of right ventricular-dependent coronary circulation in pulmonary atresia with intact ventricular septum

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Background: Right ventricular-dependent coronary circulation (RVDCC) is associated with pulmonary atresia with intact ventricular septum and is defined by two or more epicardial coronary arteries with atresia or severe stenosis resulting in the dependency of coronary supply by retrograde flow. The hypothesis of this study is that coronary Doppler flow patterns on echocardiography can be used to distinguish patients with RVDCC.

Methods: Between 2007 and 2016, we reviewed 16 patients with pulmonary atresia or critical pulmonary stenosis. Patients were divided into two groups, those with RVDCC (determined by angiography or pathology evaluation) and those without. Echocardiographic evaluation of the coronary arteries included 2-dimensional measurements and pulse wave Doppler flow pattern in 3 epicardial coronary arteries. Velocity-time integral (VTI) and maximal velocity (Vmax) were measured and compared between the two groups.

Results: Three coronary flow patterns were demonstrated: (1) all antegrade flow, (2) antegrade to retrograde VTI flow ratio >1 , and (3) antegrade to retrograde VTI flow ratio ≤ 1 . Of the 7 patients with RVDCC, 6 (86%) had evidence of flow pattern 3 in ≥ 2 of the 3 coronary arteries in contrast to 0 (0%) of the non-RVDCC patients ($p=0.005$). Higher retrograde Vmax was associated with RVDCC ($p=0.001$) and coronary artery dilatation with Z-score $\geq +3$ was also associated with RVDCC ($p=0.02$).

Conclusions: Echocardiographic evaluation of the coronaries can be useful in identifying RVDCC. More retrograde flow in at least two coronary arteries is strongly suggestive of RVDCC. Dilatation of the coronary arteries is also supportive evidence.