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Norwood I type procedure for Hypoplastic Left Heart Syndrome at time of Fontan completion: does it make any difference?

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Background: Stage one reconstruction (Norwood operation) for Hypoplastic Left Heart Syndrome (HLHS) can be performed with either a modified Blalock-Taussig shunt (mBTS) or a right ventricle-pulmonary artery (RV-AP) conduit. Hemodynamic and physiological differences between the 2 surgical palliations have been described before stage 2 surgical reconstruction. However, few data are available about differences at time of Fontan.

Aim: To evaluate hemodynamic and physiological differences between the 2 surgical palliations for HLHS at time of Fontan procedure.

Methods: The clinical records of consecutive 17 patients with HLHS who completed Fontan between 2007 and 2011 were retrospectively reviewed. Following first and second stage palliation, serial clinical evaluations and echocardiograms were performed in all patients, followed by cardiac catheterization before Fontan. Pre-catheterization data included the degree of right ventricular dysfunction and atrioventricular valve regurgitation (AVVR). Significant ventricular dysfunction was defined “a priori” by pre-catheterization echocardiographic examination or evidence of hemodynamic compromise (ventricular end-diastolic pressure >14 mmHg).

Results: Of 17 patients undergoing Fontan at a mean age of 50.5 months (range 38.2-57.5 months), 16 had an extracardiac conduit and 1 had a fenestrated lateral tunnel. At stage 1 palliation, 6 patients had a mBTS (group 1) and 11 had a RV-AP conduit (group 2). Follow-up echocardiograms and cardiac catheterization data were available in all patients. Significant right ventricular dysfunction was present only in 1 patient, in group 2. Moderate AVVR was present in 2 patients (11.7 %), in group 2. The hemodynamic results did not showed statistically significant differences. For mBTS versus RV-AP shunt there was a pulmonary artery pressure (PAP) of 10.8 ± 1.3 versus 11.8 ± 1.4 (p= 0.21), a Qp/Qs ratio of 0.66 ± 0.09 versus 0.65 ± 0.06 (p=0.83), a right ventricular end-diastolic pressure (RVEDp) of 8 ± 1.58 versus 8.9 ± 1.64 (p=0.31), a coronary perfusion pressure of 64.2 ± 9.73 versus 60.6 ± 8.39 (p=0.47).

Conclusions: At time of Fontan, the degree of right ventricular dysfunction, AVVR and hemodynamic data were not statistically different between the two groups. Status of patients with HLHS at time Fontan completion is not influenced by Norwood I type of initial surgical palliation.