Early life Predictors for Major Adverse Events in Hypoplastic Left Heart Syndrome after Norwood Stage I Palliation: A 25-year Retrospective Study


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Objective: We aimed to identify perinatal risk factors for severe complications in infants with hypoplastic left heart syndrome (HLHS).

Methods: Ninety-three patients with HLHS who underwent stage I Norwood palliation between 1993-2017 were included. Fetal and postnatal echocardiograms, echo notes, demographic, surgical and other clinical data were reviewed. Fetal and postnatal risk variables included restrictive or intact atrial septum (RAS/IAS), left deviation of septum primum (LDSP), significant tricuspid regurgitation (sTR), right ventricle (RV) dysfunction, left ventricular (LV) morphology (aortic stenosis (AS)&mitral stenosis (MS), aortic atresia (AA)&MS and AA&MA), postnatal diagnosis and low preoperative weight (LPW; ≤2.5 kg). Major outcome variables were survival, need for ventricular assist device (VAD) and ECMO, protein-loosing enteropathy and heart transplant (Htx).

Results: The overall survival was 67% (interstage I: 76%; interstage II: 91%; post-TCPC 95%), rising from 54% during 1993-2002 to 81% during 2003-2017. Fetal diagnosis was available in 47%, rising from 22% during 1993-2002 to 64% during 2003-2017. Intrauterine sTR was linked to higher pulmonary pressure at stage II (15.7±2.3 vs. 11.8±2.8; p=0.03) and lower interstage II survival (67% vs. 96%, p=0.06). Postnatal diagnosis was associated with initial RV dysfunction (p=0.007), sTR (p=0.005) and delayed stage I surgery (p=0.001) but did not influence survival. LDSP was diagnosed in 71% of infants with RAS and was most often linked to AA&MA (p=0.0004), whereas AA&MS was associated with lower interstage I (60% vs. 84%; p=0.01) and overall survival (53% vs. 73%; p=0.06). RAS/IAS was linked to atrial septostomy prior to stage I (p<0.0001), longer mechanical ventilation (p=0.04), ICU (p=0.002) and overall stay in hospital (p=0.0002) after stage I as well as lower survival after stage III (p=0.006). LPW at stage I was associated with lower interstage I survival (45% vs. 80%; p=0.01), need of VAD/ECMO (2/11 vs. 2/82; p=0.02) and Htx (2/11 vs. 0/82; p<0.001).

Conclusion: In patients with HLHS, certain LV anatomical subtypes along with restrictive atrial septum and low body weight at stage I remain important risk factors for severe complications later in life.