

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

Fricke K.(1), Hanseus K.(1), Johansson Ramgren J.(1), Bergman G.(2), Mellander M.(3), Sunnegårdh J.(3), Rydberg A.(4), Odermarsky M.(1), Liuba P.(1)
Pediatric Heart Center, Skåne University Hospital, Lund, Sweden (1); Department of Women's and Children's Health, Karolinska Institutet, Stockholm, Sweden (2); Queen Silvia Children's Hospital, Göteborg University, Göteborg, Sweden (3); Department of Clinical Sciences, Pediatrics, Umeå University, Umeå, Sweden (4)

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-losing 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.