Perinatal Risk Factors for Mortality after Norwood Palliation of Hypoplastic Left Heart Syndrome

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Background: Since 1993, pediatric cardiology in Sweden has been centralized to Lund and Gothenburg, which together serve a population of nearly 10 million people. We sought to determine the impact of several perinatal variables on survival in patients with HLHS.

Methods: A retrospective survey of 90 consecutive HLHS cases who underwent stage I Norwood palliation (S1P) between 1993 and 2013 at our center. Data on prenatal diagnosis (PND), birth characteristics, preoperative variables, type of shunt at S1P (Blalock-Taussig (BT) or Sano (S)) and early postoperative variables were collected from the hospital’s database. Death occurring between S1P and stage 2 palliation (interstage I mortality; ISM-I), between stage 2 and 3 (ISM-II) and overall mortality were chosen as outcome variables.

Results: During the study period, there was a relatively steep decline in ISM-I from nearly 60 % in the beginning of the 1990s to a plateau < 10 % during the last decade. PND did not improve survival but was associated with earlier S1P (p=0.001) and with less preoperative tricuspid regurgitation (TR; p=0.04). Those who underwent S1P of the Norwood procedure after the 1st week of life had higher risk for death within the first month of life (Odds Ratio (OR) 7.2, p=0.002). Restrictive atrial communication (p = 0.005; OR 6.5) was associated with increased ISM-I, while TR increased risk for ISM-II (p = 0.02, OR 2.4). Norwood procedure with BT shunt was exclusively performed during 1993-2002, and was linked to higher ISM in patients with aortic atresia-mitral stenosis (AA-MS; p=0.06, OR 2.8). During 2003-2013, in which the majority received S shunt, the only predictors for ISM-I were prematurity and low birth weight (p<0.05 and OR > 5 for both).

Conclusion: Lower preoperative weight and restrictive atrial communication remain important risk factors for mortality, whereas Norwood palliation within the 1st week of life improves survival. Patients with AA-MS appear to be at higher risk for ISM-I during Norwood surgery with BT shunt, but no such trend could be observed in those palliated with Sano shunt.