

Outcome after the Norwood operation for Hypoplastic left heart syndrome and related malformations – a 22 years single centre experience

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Introduction: Survival for Hypoplastic left heart syndrome (HLHS) and related single-ventricle lesions improved over the past decades. However, the Norwood circulation with its vulnerable balance between pulmonary and systemic blood flow is still associated with relatively high mortality. We aimed to identify risk factors for adverse outcome after the Norwood procedure and subsequent superior cavopulmonary anastomosis (SCPA).

Methods: Medical records of 379 patients (HLHS, n=304; non-HLHS, n=75) who underwent a Norwood procedure between 1996 and 2018 were reviewed. Follow-up to SCPA was completed in all survivors. Important changes in management included the utilization of antegrade cerebral perfusion (January 2000) and the introduction of an interstage surveillance program (October 2005). Three groups were analysed (Group 1: 1996-1999, n=50; Group 2: 2000-2005, n=103; Group 3: 2006-2018, n=226).

Results: Median age at surgery was 6 (IQR 4-9) days. A modified BT-Shunt was the standard source of pulmonary blood flow (n=357, 94.2%); 3 mm or 3.5 mm shunts were most commonly used. Early mortality (within the first 30 postoperative days or before discharge) was 10.6% and declined over time (Group 1: 22.0%; Group 2: 12.6%; Group 3: 7.1%, p=0.008). With introduction of the home-monitoring program interstage mortality of discharged patients decreased (12/117 vs. 5/147, p=0.041), but inpatient treatment before SCPA became more common (10/130 vs. 63/209, p<0.001).

Survival to SCPA was 83.2% and improved over time (Group 1: 70.0%; Group 2: 79.2%; Group 3: 90.7%, p<0.001). Weight at surgery <3 kg (p=0.041), ascending aorta <2 mm (p=0.006) and use of a 3 mm shunt (p=0.017) were identified as risk factors for mortality prior to SCPA. The indexed shunt diameter was not different between survivors and non-survivors (1.05 ±0.13 mm/kg vs. 1.08 ±0.14 mm/kg, p=0.123). Early mortality after SCPA was 6.4%. Age at surgery <90 days (p=0.014) and inpatient treatment before SCPA (p<0.001) were related to increased mortality.

Conclusions: The postoperative course after the Norwood operation and the interstage period before SCPA still carry a relatively high risk of mortality. The introduction of home-monitoring programs reduces interstage death, but children not fulfilling discharge criteria have a poorer prognosis in terms of survival after SCPA.