Clinical course and the impact of interstage monitoring after Norwood I and Hybrid procedure

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Objectives: Infants with hypoplastic left heart syndrome (HLHS) are at risk for interstage morbidity and mortality especially between the first and second surgical stage after Norwood and Hybrid procedure. Methods: We compared morbidity (defined as hospitalization/reinterventions) and mortality in patients treated for HLHS between the first and second stage, and the impact of interstage monitoring on outcome.

Results: Between June 2008 and December 2011 twenty-six (male=14) infants with HLHS (n=16) and other univentricular heart malformations with aortic arch anomaly (n=10) were planned for interstage monitoring after Norwood I (n=12) and Hybrid procedure (n=14). Three (11.5%) infants died after first stage palliation (Hybrid n=1; Norwood n=2), 3 (11.5%) after second stage palliation (Hybrid n=2; Norwood n=1), all of them after early second stage (<90 days). There were no difference between Norwood I and Hybrid procedure regarding over-all mortality (23%) (Hybrid n=3; Norwood n=3). 7 (26.9%) infants could not be discharged from hospital due to hemodynamic instability and were referred to early second stage (<90 days). After first stage invasive re-evaluation rate before discharge was high with catheterizations in 14 (53.8%) (Hybrid n=8; Norwood n=6), with the need for re-interventions in 8 (30.7%) (Hybrid n=5; Norwood n=3) or cardiac surgery in 3 (11.5%) (Hybrid n=2; Norwood n=1).

14 infants had interstage monitoring for 89 (10-177) days. One infant (3.9%) died during interstage monitoring. Interstage monitoring was positive in 7 (50%) infants after 10 (4-68) days (Hybrid n=5; Norwood n=2), leading to rehospitalization and catheterization in 6 (Hybrid n=4; Norwood n=2), requiring interventions in 2 (PDA stent dilatation, ASD stenting, all in Hybrid). Overall, 3 of the 7 patients with positive interstage monitoring were candidates for an early second stage.

Conclusions: Morbidity in infants after first stage before discharge is characterized by a high reintervention rate (30.7%). After discharge interstage monitoring is positive in 50% indicating the need for catheter treatment in 29%. Despite retrograde aortic flow in infants with HLHS after Hybrid procedure mortality is comparable in both groups. Mortality after second stage is associated with early age (<90 days). This highlights the importance of interstage monitoring in both treatment groups.