Comparison of neurodevelopmental outcome at 1 and 4 years in children undergoing Hybrid and Norwood procedures for hypoplastic left heart syndrome

Knirsch W. (1,5), Liamihi R. (1,2,5), Prêtre R. (3), Dave H. (3,5), Bernet V. (4,5), Latal B. (2,5), Kretschmar O. (1,5)

Divisions of (1) Cardiology, (2) Child Development Center, (3) Congenital Cardiovascular Surgery, (4) Neonatology and Pediatric Intensive Care Unit, (5) Children’s Research Center, University Children’s Hospital Zurich, Switzerland

Background: Data on long-term neurodevelopmental outcome of children treated for hypoplastic left heart syndrome (HLHS) comparing Hybrid and Norwood procedure are needed.

Methods: Neurodevelopmental outcome assessment at 1 and 4 years of age of infants undergoing Hybrid (2006-2008) and Norwood procedure (2004-2008) for HLHS after implementation of both treatment options in a single centre setting. Assessment included at 1 year of age, psychomotor (PDI) and mental developmental index (MDI) of Bayley Scales of Infant Development III, at 4 years of age the Wechsler Primary Preschool Intelligence Scale – III, and Movement-ABC 2. Furthermore, we compared outcome of early (within first year after implementation) vs. late period (after first year).

Results: Thirty-one (18 males) infants were treated with Hybrid (n=13) and Norwood (n=18) procedure. Twenty children survived until age of 1 year. Motor (PDI) and cognitive (MDI) outcome at 1 year were significantly impaired compared to the norm [PDI 57(49–99), p<0.001; MDI 91(65–109), p=0.002], but not different comparing Hybrid (n=9) and Norwood (n=11) procedure [PDI: Norwood 56.5(49–81) vs. Hybrid 65(50–99), p=0.18; MDI: Norwood 93(65–109) vs. Hybrid 88(71–102), P=1.0] (1). At 4 years, 16 of 20 children were reevaluated (late death n=1, lost to follow up n=3). Overall, both cognitive and motor performance was poorer than the norm [IQ: 89(76-116), p=0.02; motor outcome: p=0.002], but not different between Hybrid (n=7) and Norwood (n=9) procedure [IQ: Norwood 92(80–104) vs. Hybrid 88(76–116), p=1.0; motor outcome: p>0.8]. However, cognitive performance of children treated in the early learning period was significantly lower compared to children treated in the late period independent of performed procedure [IQ early period 87(76-101) vs. late period 96(80-116), p=0.03]. Motor outcome was comparable between early versus late treatment period (p>0.8).

Conclusions: Overall, cognitive and motor functions are impaired at 1 and 4 years of age in children with HLHS. As described for the 1 year outcome, at 4 years of age there are no evident differences between Hybrid and Norwood procedure, but we could observe that an institutional learning curve affects cognitive outcome.

(1) Knirsch W et al., Eur J Cardiothorac Surg 2012