Neurodevelopmental outcome in Fontan patients – is the Norwood procedure a prognostic factor?

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Objective: Patients with single ventricle physiology are prone to neurodevelopmental abnormalities. Complex surgery within the neonatal period may be a risk factor for brain injury and neurodevelopmental impairment. Neurocognitive outcome at pre-school age was compared between Fontan patients who underwent a Norwood procedure and other single ventricle patients who did not require complex neonatal surgery.

Methods: Verbal IQ, performance IQ and full scale IQ were evaluated with the Hannover-Wechsler-Intelligence scale. The German “Kognitiver Entwicklungstest für das Kindergartenalter” (KET-KID), which is composed of a global scale for cognitive development, a verbal and nonverbal scale, was applied to assess cognitive functions. To identify potential risk factors for adverse neurodevelopmental outcome, patient and procedural variables were evaluated.

Results: Neurocognitive assessment was completed in 90 Fontan patients at a median age of 4.1 (3.5-7.1) years. 62 underwent a Norwood procedure, among the 28 remaining patients 16 had placement of a systemic-to-pulmonary artery shunt, 10 underwent pulmonary artery banding and 2 did not require any intervention in the neonatal period. Overall, IQ-scores and percentile ranks of the KET-KID were in the normal range and, except for the KET-KID verbal scale, did not differ between patients who underwent a Norwood procedure and those who did not (verbal IQ: 96 ±14 vs. 92 ±11, p=0.095; performance IQ: 93 ±10 vs. 92 ±10, p=0.408; full scale IQ: 93 ±12 vs. 91 ±10, p=0.177; KET-KID global: 42 ±29 vs. 33 ±30, p=0.102; KET-KID verbal: 47 ±30 vs. 31 ±30, p=0.016; KET-KID nonverbal: 38 ±27 vs. 37 ±26, p=0.385). Full scale IQ was below average in 14 (23%) of the Norwood patients compared to 10 (36%) of the remaining cases (p=0.192). Global KET-KID scores were below average in 17 (27%) and 10 (36%) patients (p=0.427), respectively. Gestational age, lower birth weight, smaller head circumference and increasing number of complications during staged palliation were associated with worse neurocognitive test results.

Conclusion: Neurocognitive outcomes of Fontan patients at pre-school age were in the normal range, but performance was lower compared to population norms. Surprisingly, the Norwood procedure was not associated with neurocognitive test results.