

Cerebral MRI findings and neurodevelopmental outcome in children before Fontan procedure at 2 years of age – Enlargement of liquor spaces influences outcome

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Objectives: Children with complex congenital heart disease undergoing neonatal cardiac surgery are at risk for neurodevelopmental impairment, especially in univentricular heart malformations. Neonatal cerebral MRI findings show pre- and postoperative cerebral injuries such as white matter injury and cerebral stroke. We focussed on the long term impact of cerebral injuries comparing cerebral MRI findings and neurodevelopmental outcome at two years of age.

Methods: In a prospective multicenter cross-sectional study we analysed consecutive patients with hypoplastic left heart syndrome (HLHS) or with UVH before Fontan procedure. Patients were palliated either by Hybrid or Norwood procedure. Neurodevelopmental outcome was investigated by Bayley Scales of Infant and Toddler Development III (Bayley III) and compared to findings of cerebral MRI scan before Fontan procedure.

Results: 48 patients (male 32; age mean 26.6±3.8 months) with HLHS (n=26) and non-HLHS (n=22) were included. 44 patients were treated first by Hybrid procedure (n=25), Norwood procedure (n=7) and shunt or banding procedure without cardiopulmonary bypass (n=12) before bidirectional cavopulmonary anastomosis (n=48). In Bayley III Scales, median cognitive scale (CCS) 100 (range 65-120), language scale (LCS) 97 (68-124) and motor composite scale (MCS) 97 (55-124) were only inferior compared to norm data for LCS (Z=-2.2, p=.025). 36.2% (17/47) of patients showed disease-related intracranial lesions: isolated ventriculomegaly in 3, minimal (<2mm) white matter lesions in 5, cerebral stroke (less one third of vascular supply territory) in 9. The latter two findings were additionally associated with liquor spaces enlargement in 7 patients. Therefore, the enlargement of inner and/or outer liquor spaces was the most frequent pathologic MRI finding and was associated with lower scores in all subscales of Bayley III (CCS: p=.020; LCS: p=.002; MCS: p=.013).

Conclusions: Despite an overall good neurodevelopmental outcome before Fontan procedure 36.2% of patient exhibit intracranial lesions on cerebral MR Scan. Enlargement of inner or outer liquor spaces seems to be the major contributing factor of poor neurodevelopmental outcome. Improved perioperative management and neurodevelopmental follow up programs are needed in this patient group to detect neurological deficit early and to further improve quality of life.