Cerebral MR Morphometry and Neurodevelopmental Outcome in Children before Fontan procedure at 2 years of age – White and grey matter volumes are larger after Hybrid procedure

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Objectives: Advances in cardiac surgery and intensive care have reduced mortality in patients with congenital heart disease (CHD). Improvement of neurodevelopmental outcome is one of the most important goals for children with complex CHD. Delayed brain development has been described preoperatively in newborns with CHD, affecting mainly white matter (WM) structures. Perioperative complications may additionally cause brain injury. We set out to study brain volumes in children with hypoplastic left heart syndrome (HLHS) or univentricular heart (UVH) at the age of 2 years prior to Fontan surgery.

Methods: Prospective two-center cross-sectional study. Cerebral MRI (21 Zurich patients: 3 Tesla GE MR 750 scanner; 23 Giessen patients: 3 Tesla Magnetom Verio B17 Siemens scanner; FreeSurfer image analysis) was performed before Fontan. Morphometric findings were correlated with results from the Bayley Scales of Infant and Toddler Development III (Bayley III).

Results: 44 patients (male 28; age mean 26.7±3.9 months) with HLHS (25/44) and non-HLHS (19/44) were included, treated by Hybrid or Norwood procedure. The average brain volumes derived were: 909.1±83ml for total brain volume, 277.1±30ml for WM, 610.5±59ml for gray matter (GM), and 42.7±5ml for subcortical GM. Ventricular CSF volume was larger in patients with abnormal neurologic examination (i.e. tonus and/or abnormal reflexes) and correlated with poorer Bayley III scores (CSS: rSpearman’s Rho=-.31, p=.043; LCS: rSpearman’s Rho=-.40, p=.009, MCS: rSpearman’s Rho=-.31, p=.04). Brain volumes did not differ between type of UVH (HLHS vs. non-HLHS; p values all > .07). Subcortical GM (p=0.01) and WM (p= .05) volumes were larger in patients receiving Hybrid procedure as first surgery. However, patients after Hybrid did not show significantly higher Bayley III scores (p> .69). Brain volumes were not different in infants with cerebral lesions compared to those without (p-values all > .38).

Conclusions: Cerebral MR morphometry is a good tool to measure brain volumes in children with CHD. In children undergoing Fontan procedure, larger ventricular CSF volume correlates negatively with neurodevelopmental outcome. WM brain volume loss was associated with the surgical approach, but not with the subtypes of CHD. Perioperative management in combination with routine neurodevelopmental follow up programs are needed to further improve outcome.