

The association between the head circumference and the central venous pressure in children undergoing the Fontan procedure

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Introduction: High central venous pressure (CVP) may cause communicating hydrocephalus and macrocephaly. We examined if higher pulmonary pressures (PAP) in children undergoing bidirectional cavopulmonary anastomosis (BDCPA) leads to an increased growth of the head circumference (HC) as the central venous pressure (CVP) interferes with cerebral venous drainage.

Methods: Seventy-four consecutive children with single-ventricle physiology (24 HLHS, 11 DILV, 10 DORV, 9 unbalanced AVSD, 7 PA IVS, 5 TA, 4 PA VSD and 4 unbalanced ccTGA) were included in a retrospective analysis. Patients with other causes of a macrocephaly were excluded. CVP and PAP were measured during the routine catheterization before BDCPA and Fontan completion. The occipital-frontal circumference was measured using a flexible non-stretchable measuring tape at the same time and at the routine clinical visits and compared with longitudinal age dependent percentiles in normal children measured in our own institution.

Results: Median age at BDCPA and Fontan were 4.8 (1.6-12) and 27.9 (7-40.6) months respectively. HC was more than 1 SD below 50th percentile at the time of the BDCPA and showed significant growth between pre-BDCPA- and pre-Fontan catheterization (7 (0-100) vs. 36 (3-56)th percentile, $p < 0.001$), while PAP decreased significantly (14.8 ± 2.7 vs. 13.2 ± 2.1 mmHg, $p = 0.012$). There was no correlation between PAP and HC pre-Fontan ($R^2 = 0.001$). Children with lower differences in CVP pre-BDCPA and PAP pre-Fontan showed increased growth of HC. Higher cerebral venous drainage may explain this weak correlation ($R^2 = 0.19$, $p = 0.012$).

Conclusions: Moderately elevated CVP in children with single ventricle physiology does not lead to macrocephaly. Between BDCPA and Fontan, HC increased significantly and achieved values close to norm for age. The lack of direct correlation between PAP before Fontan and HC may be explained by a catch-up growth of HC in patients with better pulmonary vascular bed. Further studies with focus on high PAP are needed to exclude or prove a direct correlation.