

Neurodevelopment outcomes following fetal diagnosis of Ebstein Anomaly or Tricuspid Dysplasia

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Background: Data concerning neurodevelopmental outcome in subjects with Ebstein Anomaly (EA) or severe Tricuspid Valve Dysplasia (TVD) are lacking. New cardiovascular magnetic resonance (CMR) technology allowing hemodynamic assessment in the fetus could provide insights into the mechanisms of brain injury and dysmaturation as well as prognostic information. We aimed to investigate the clinical and neurodevelopment outcome of patients following fetal diagnosis of EA/TVD. **Population and methods:** From 2013 to 2017, we identified 15 fetuses with EA and 6 fetuses with TVD at our institution at a median gestational age of 23 weeks. Of this cohort, 12 had fetal CMR at a median gestational age of 36 weeks. Fetal biometry and hemodynamics were compared with those in 30 normal fetuses. **Results:** Two fetuses underwent early termination of pregnancy. Three fetuses died in utero, four developed hydrops but survived to birth, five had circular shunts, 11 had functional or anatomical pulmonary atresia, and five had pericardial or pleural effusions. The tricuspid regurgitation was severe in 17, moderate in three and mild in one. CMRI revealed significantly lower combined ventricular output ($p<0.001$), superior vena caval flow ($p<0.01$) and fetal oxygen delivery ($p<0.01$) in EA/TVD fetuses compared to controls. EA/TVD fetuses had increased oxygen extraction fraction ($p<0.01$), but body and brain weight z-scores were still reduced ($p<0.01$ and $p<0.001$). Of the 12 fetuses having had fetal CMR, 9 survived and 8 had neurodevelopment assessment. Brain MRI, neurodevelopmental and clinical outcomes are summarized in Figure 1. **Conclusions:** Neurodevelopmental delay appears to be very common in all severe forms of EA and TVD regardless of the mode of management. This might be explained by the severity of cerebral hypoxia ischemia in this form of congenital heart disease, which is already present in fetal life, and which persists postnatally, both in subjects undergoing palliative surgery and in subjects managed conservatively. Cerebral lesions are generally present on brain MR at birth, even when patients are born at term and managed conservatively. These preliminary data might be of help in prenatal counselling and strongly support the need for early neurodevelopmental assessment and life-long follow-up, irrespective of the surgical approach.

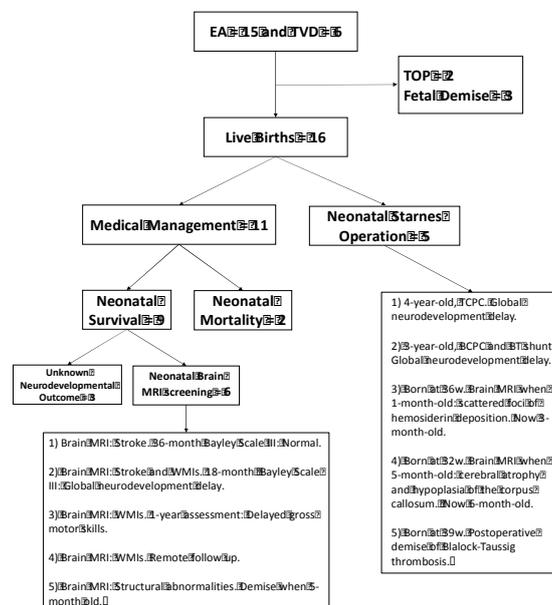


Figure 1. BCPC: bidirectional cavopulmonary connection; EA: Ebstein anomaly; TCPC: total cavopulmonary connection; TVD: Tricuspid Valve Dysplasia; TOP: termination of pregnancy.