

Outcome after Fetal Diagnosis of Ebstein's Anomaly and Dysplasia of the Tricuspid Valve: A Single Center Experience

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Background: Ebstein's anomaly (EA) and tricuspid valve dysplasia (TVD) are serious fetal observations with survival rates to infancy below 25% in all major studies. There is little doubt that the worst disease spectrum is detected in utero, with many affected fetuses notable for significant cardiomegaly, severe tricuspid regurgitation and absent antegrade pulmonary blood flow. To survive, the fetal left ventricle needs to compensate for the dysfunctional right ventricle (RV) while postnatal survival rather depends on the RV to restore adequate pulmonary flow. Potential treatment strategies to improve outcomes include preterm delivery at signs of heart failure, and postnatal pharmacological lowering of the pulmonary vascular resistance and ligation of the arterial duct to avoid circular shunts. We reviewed the institutional experience with this entity.

Methods: Fetuses diagnosed with EA/TVD between 1999 and 2010 were identified from our prospectively acquired database. Echocardiography data were correlated with the established outcome of all cases.

Results: EA was diagnosed in 33 and TVD in 13. There were 4 pregnancy terminations with EA, 5 intrauterine deaths with EA, 8 neonatal deaths (mean age 10.1 days) and 29 survivors >1 months. Survival of the entire cohort was 80% at birth and 63% at 1 month, or 88% and 69%, respectively, on an intention-to-treat basis. Parameters associated with mortality (n=15) included earlier gestational age at diagnosis (25.5 ± 4.7 vs. 31.4 ± 6.8 weeks; $p < 0.005$), larger right atrial area index (0.95 ± 0.1 vs. 0.57 ± 0.2 ; $p < 0.0001$), increased cardio-thoracic circumference ratio (0.6 ± 0.1 vs. 0.49 ± 0.1 ; $p < 0.005$), absent pulmonary forward flow (73% vs. 22%; $p < 0.003$) and severe tricuspid regurgitation (67% vs. 30%; $p = 0.03$). No associations were found between EA vs. TVD (11/33 vs. 4/13; $p = 0.74$), with fetal hydrops (33% vs. 27%; $p = 0.73$), additional cardiac abnormalities (13% vs. 33%; $p = 0.27$) and era of diagnosis (1999-2004: 4/12 (33%) vs. ≥ 2005 : 11/30 (33%); $p = 1.0$).

Conclusion: Survival rate of prenatally diagnosed EA/TVD in our experience is significantly better than previously reported likely because less severe forms were more commonly detected in our catchment area during the past decade. Previously established predictors associated with adverse outcomes were confirmed in this study.