

Contemporary Outcomes and Factors associated with Mortality after a Fetal or Neonatal Diagnosis of Tricuspid Valve Disease

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Background: Ebstein's anomaly (EA) and tricuspid valve dysplasia (TVD) are rare anomalies and data on outcomes after a fetal or neonatal EA/TVD diagnosis is conflicting.

Methods: To examine the outcome and identify markers predictive of mortality, we reviewed our single center experience with 79 cases of EA/TVD from 2000-2014. Variables were analyzed separately for cases diagnosed in-utero without pregnancy termination (n=5) and for all live-born patients.

Results: Of 47 fetal cases with an intention-to-treat, 8 (17%) died in-utero and 10 (21%) as neonates. Independent predictors of in-utero demise included severe tricuspid regurgitation with a Doppler gradient <40 mmHg (odds ratio (OR) 1.22 per mmHg deduction; $p=0.003$) and pulmonary regurgitation (OR 11.4; $p=0.03$) at the baseline exam. A novel prognostic score (range 0-10) combining the severity of 5 echocardiographic findings shown in the Table was independently associated with overall mortality (hazard ratio (HR) 1.39 per point increase; $p=0.01$).

Variable	Weighting		
	0	1	2
Cardiothoracic ratio	<0.65	0.65-0.75	>0.75
Right atrial area index	<0.75	0.75-1.0	>1.0
Pulmonary forward flow	Normal	Reduced	Absent
Tricuspid regurgitation	No or mild	Moderate or severe	Moderate or severe
- TR Doppler gradient	-	≥ 40 mmHg	<40 mmHg
Pulmonary regurgitation	No	Yes	Yes
- Umbilical artery flow	-	Antegrade	Absent or reversed

Survival rates of 66 live-births at 1-month, 1-year and 5-years were 86%, 82% and 80% respectively, while 75%, 60% and 55% remained free from surgery at the same points in time. Factors associated with postnatal death by multivariate analysis included a younger gestational age at birth (HR per week: 1.59; $p<0.001$), tricuspid annulus diameter (HR per z-score increase: 1.76; $p=0.004$), and no pulmonary forward flow (HR 4.63; $p=0.03$).

Conclusion: Our experience with fetal and neonatal EA/TVD shows better survival rates than previously reported. Mortality after a fetal diagnosis was significantly associated with hemodynamic changes indicative of a circular shunt, including pulmonary and tricuspid regurgitation severe enough to cause diastolic umbilical arterial flow reversal.