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PERINATAL OUTCOMES AFTER FETAL DIAGNOSIS OF EBSTEIN ANOMALY OR TRICUSPID VALVE DYSPLASIA IN THE CURRENT ERA: A MULTI-CENTER STUDY

Moderated Poster Contributions
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Background: Ebstein anomaly and tricuspid valve dysplasia (EA/TVD) are rare congenital tricuspid valve malformations associated with high perinatal mortality. The literature to date consists of small, single center case series often spanning several decades. We performed a multi-center study to assess perinatal outcomes after fetal diagnosis of EA/TVD in the current era.

Methods: Twenty-three centers contributed to this retrospective study, which included fetuses diagnosed with EA/TVD from January 1, 2005 to September 1, 2011. Fetuses with complex associated lesions, such as congenitally corrected transposition or left heart obstruction, were excluded. Among live-born patients, the primary outcome was survival to neonatal hospital discharge.

Results: We included 272 fetuses with EA/TVD diagnosed at a median gestational age (GA) of 25 weeks (interquartile range: 21-30). Nearly one-third developed hydrops, 15 (6%) had documented arrhythmias in utero, and 61 (22%) were known to have extracardiac anomalies and/or a genetic diagnosis. There were 17 elective terminations (6%), 48 fetal demises (18%) at a median GA of 30 weeks (26-33), and 12 fetuses (4%) lost to follow-up. Of the 195 live-born patients (72% of the initial cohort), 65 (33%) died prior to discharge. Eighteen (9%) patients died within 1 day of life, and among those surviving >1 day, the median age at death was 11 days (6-24). Neonatal non-survivors had lower GA and weight at birth than survivors (35.6 vs. 37.5 weeks; 2.4 vs. 3.0 kg; both $p<0.001$). Hydrops, intubation in the delivery room, inotropic support, and ECMO were associated with mortality ($p<0.001$), while neonatal interventional catheterization and cardiac surgery were not ($p=0.9$).

Conclusion: In this large, contemporary cohort of fetuses with EA/TVD, there was a higher proportion of live-born patients compared to prior series. However, neonatal mortality remained significant and was associated with prematurity, lower birth weight, and the need for more intensive medical therapy. Anatomic and physiologic characteristics as assessed by fetal echocardiography will likely help further delineate predictors of poor outcome in this population.