

Ebstein's anomaly and tricuspid valve dysplasia: prognosis after diagnosis in utero.

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Introduction: Tricuspid valve malformations are rare congenital heart diseases. The prenatal diagnosis of Ebstein's anomaly (EA) and Tricuspid valve dysplasia (TVD) are associated with high mortality.

There are conflicting reports concerning accurate prognostication after diagnosis in utero. The aim of our study was to assess prognostic factors based on our experience.

Methods: We retrospectively reviewed echocardiography of 37 fetuses, 26 EA and 11 TVD, between 1984 and June 2010, from 7 cardiopediatric centers. There were 11 terminations, 5 intrauterine deaths, 7 neonatal deaths and 14 survivors to age over 2 years.

Results: We found that the major prognostic factor for outcome was the pattern of pulmonary valve flow. Retrograde pulmonary valve flow had a predictive positive value (PPV) for death of 83 %, and when associated with significant pulmonary insufficiency, PPV was 100% for death. An anatomic pulmonary atresia was also associated with 100 % of death. By contrast, cardiothoracic index, right to left ventricular ratio, Celermajer index were not useful prognostic markers. Compared with retrograde pulmonary valve flow, antegrade flow in utero predicted good outcomes with a PPV for survival of 86%.

The SAS score, more complex, was less correlated to our series with 79 % PPV of death when the score was ≥ 5 and a PPV survival of 79 % when the score was < 5 .

Conclusion: Pulmonary artery valve flow is a simple and excellent prognostic factor when major tricuspid valve disease is diagnosed in utero. Nevertheless, these fetuses should be monitored throughout the pregnancy because several hemodynamic factors may change the prognosis.

Key Words: Ebstein, Tricuspid valve dysplasia, Prognosis, Fetal.