Prospective validation of a prenatal score for prediction of postnatal outcome in cases with critical pulmonary stenosis

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Objective
Our group has showed that the postnatal outcome of fetuses diagnosed with pulmonary atresia - critical stenosis with intact ventricular septum (PA - CS/IVS) may be accurately established with a combination of three size-based parameters [tricuspid valve (TV)/mitral valve (MV) ratio, pulmonary valve (PV)/aortic valve (AoV) ratio, right ventricle (RV) length/left ventricle (LV) length ratio] and one functional marker (tricuspid inflow duration/cardiac cycle length). The objective of this study is to analyze the predictive capability for postnatal outcome of fetuses with PA - CS/IV, by prospectively applying this score.

Methods
This is an observational study of prospective cohorts undertaken at a tertiary care referral center of cases of PA - CS/VS, prenatally diagnosed between January 2010 - June 2016. Only liveborn fetuses and with complete postnatal follow-up information were included in the study. We applied our score during echocardiographic assessment to the diagnosis. We used the previously reported cut-off points of the parameters of our score (TV/MV ratio ≤ 0.83, PV/AoV ratio ≤ 0.75, RV/LV length ratio ≤ 0.64, and tricuspid inflow duration/cardiac cycle length ≤ 36.5%). We included three patients that underwent prenatal pulmonary valvuloplasty (PPV), in order to determine the discriminatory capacity of these cases with our score. According to our data if ≥ 3 of the four markers are present, there is a high probability of non-BV circulation. Therefore, we divided the score into three subgroups: ≤ 1 parameter (to predict those cases with a high probability of BV circulation), 2 (to select PPV candidates) and ≥ 3 (to predict cases highly likely to have a non-BV circulation after birth). We evaluated the predictive capability of the score for these three prognosis situations.

Results
The study group included 29 liveborn foetuses (22 with a BV circulation and 7 with non-BV circulation). According to the three prognosis settings, three subgroups (BV circulation, non-BV circulation and PPV required) and the predictive capability for these different situations were proposed. All cases that underwent a PPV had two markers of the score present.

Conclusion
The postnatal outcome of PA - CS/IVS fetuses can be predicted with good precision by the prenatal application of a score, based on four echocardiographic parameters that are easy to explore. This score also allows us to select those candidates for fetal therapy.