

Intrauterine treatment of critical pulmonary stenosis/atresia with intact septum – preliminary experience of intrauterine and postnatal course

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Critical pulmonary stenosis/atresia with intact septum (CPS/IVS, PA/IVS) remains a therapeutic challenge with significant morbidity and mortality.

Between 10/2000 and 12/2013 a total of 10 attempts of in-utero pulmonary valvuloplasties have been performed in 8 fetuses (3 PA/IVS, 5 CPS/IVS). All had hypoplastic RVs with suprasystemic RV pressures. Median TV z-score was -2,48 (-1,26 - -2,94). Median gestational age at intervention was 27+4 weeks (range: 24+6 - 32+2 weeks). Technical success was achieved in 6/10 procedures or 6/8 fetuses (75%). All procedures were carried out under general anesthesia of the mother. No maternal side effects were observed. Median procedure time was 1:51 hours (range 1:04-4:00 hours). There was no fetal death, no fetal bradycardia or significant pericardial effusion. Balloons with diameters of 4,3 – 4,8 mm were inserted through 17G (2 fetuses) and 18 needles. In all successful cases the pulmonary valve was perforated with the sharp needle before inserting the catheter. After successful intervention there was always a better (longer and biphasic) RV filling. A high gradient across the pulmonary valve remained in 5/6 fetuses with increased flow through the RV, in 1 case there was severe pulmonary regurgitation with almost no residual gradient. Continuous, but slower than normal growth of TV and RV was observed in all successfully treated fetuses. After birth, 5/6 newborns received a modified BT Shunt, in 1 newborn a balloon valvuloplasty alone was sufficient and still is at the age of 3 years. In 3 children the BT Shunt was successfully removed at 8 months of age, 1 patient received a Glenn Shunt (1,5 ventricle repair) and 1 BT shunt is still in place 6 months after surgery. At a median follow-up time of 3,8 years (0,6 – 13,2), 4/6 children are biventricular with acceptable RV size and function.

Conclusions: in-utero pulmonary valvuloplasties are technically more challenging, but seem to be better tolerated than LV interventions. Successful intervention resulted in improved flow through the right heart, which might have led to better RV growth and function at birth. Postnatal transient RV support with a systemic to pulmonary artery shunt was necessary in most cases.