P-76

Prenatal diagnosis of pulmonary atresia and intact ventricular septum – can we predict rationale for prenatal intervention ?

Dangel M.M.(1), Kolesnik A. (2), Brzezinska-Rajszys G. (3), Dangel J.H.(4)
Perinatal Cardiology Unit, 2nd Department of Obstetrics and Gynecology, Students Scientific Club, Medical University of Warsaw (1)
Department of the Clinical Anatomy, Medical University of Warsaw and The Children's Memorial Health Institute (2)
Cardiac Catheterization Laboratory, The Children's memorial health Institute, Warsaw (3)
Perinatal Cardiology Unit, 2nd Department of Obstetrics and Gynecology, Medical University of Warsaw (4)

OBJECTIVE: To evaluate anatomy, intrauterine evolution and outcome of fetuses with pulmonary atresia and intact ventricular septum (PA&IVS) diagnosed in tertiary center for fetal cardiology and look for the indication for prenatal intervention.
METHOD: 28 fetuses with confirmed PA&IVS between 1995–2010. Anatomy of the RV, pulmonary artery, tricuspid regurgitation(TR),sinusoids and neonatal outcome was evaluated. Measurements of the right and left heart structures were repeated, Z-sores were calculated. Fetal measurements were compared with the results of treatment.
RESULTS: The mean age of diagnosis was 27+/-6weeks, 10 was made before24 weeks. In 16karyotype was normal, not signs of chromosomal anomalies in others. One pregnancy was terminated, the fetus had skeletal dysplasia. There was one late intrauterine death. 3neonates were born prematurely, two were hydropic, all 3 died in the neonatal intensive care unit. 23 children were treated. Radiofrequency was not performed. In two neonates critical pulmonary atresia was diagnosed and pulmonary valvuloplasty following by BT shut was performed. In the rest 21 BT shunt was done in the neonatal period. 10 children died after surgical treatment: 7 after BT shunt, all had sinusoids, 3-additional TR, 1 died after hemi-Fontan and 2 after Glenn. None of them had TR. 13 children survived, 11 with holosystolic TR with high velocity. Survival rate was 46% for all,50% for live-born, 56% for operated neonates. RVOT opening was performed in 2,both had Z-score of TV within normal limits. There were not children after biventricular operation. Ther was not significant difference in Z-scores of the diameter of TV annulus and the size of RV between fetuses with and without TR, and those who died and survived. The medium value of Z-scores for RV diameters about -4, medium Z-score for the pulmonary artery branches was normal.
CONCLUSIONS
The prognosis for fetuses with AP&IVS in our experience is poor. Prematurity is the important risk factor for neonatal death. Due to the lack of biventricular repair and high rate of death after surgical treatment prenatal intervention should be considered to change the prenatal natural history of this severe disease, even if the right ventricle is hypoplastic.