Fetal pulmonary valvuloplasty – how sufficient it is?

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OBJECTIVE: Evaluation of fetuses diagnosed in our unit with pulmonary atresia or critical stenosis and intact ventricular septum (PA/PS&IVS) between 1995-2010 (group1) showed poor outcome. Basing on this results prospective study was undertaken to establish indications and results of fetal pulmonary valvuloplasty (FPV) between 2011-2014 (group2).

METHOD: Review the data of 59 fetuses with PA/PS&IVS. Group1 (28 fetuses) was presented previously. Data and outcome of group2 (31 fetuses) was analyzed. 5 underwent FPV.

RESULTS: The mean age of diagnosis in group1 was 27+/-6 weeks, for group2 25+/- 6 weeks. Previous evaluations of group1 showed survival rate of 46% and no biventricular (BV) repair. Out of 59 fetuses, 25 were diagnosed before 24 weeks, 6 had abnormal karyotype. 5 were terminated, just one with isolated heart defect. In 5 fetuses who had FPV, RV outflow tract was patent and TV annulus was just slightly smaller, but RV major diameter was decreased. Severe TR with high velocity was in 4, in one it was absent and sinusoids were suspected. All FPV were successful. In one fetus PA closed completely just before delivery, so radiofrequency was performed in a neonate. In all 5 neonates TV annulus was within normal limits and right ventricle enlarged significantly. One neonate died due to post-operative complications. The size of TV and RV in this neonate was within normal limits, PA was patent with 50mmHg RV-PA gradient. Other 4 most likely will have biventricular circulation (BV). Out of rest 26 fetuses, FPV would probably be possible in 8 fetuses. It was discussed in 3 couples, who refused. 1 family decided to terminate the pregnancy. In 5 the diagnosis was too late for FPV. None of those 7 who were life-born had BV circulation. None of fetuses whose anatomy was not suitable for FPV had BV repair, there were 2 IUD and 3 neonatal death.

CONCLUSIONS
The prognosis for fetuses with PA/PS&IVS after fetal intervention is much better than in those with natural history. FPV is possible in fetuses with patent RVOT, good size of pulmonary valve and branches. More precise criteria for FPV are necessary. Fetuses with hypoplastic RV and atretic RVOT had poor prognosis.