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Features and outcomes of cases with agenesis/dysplasia of the pulmonary valve diagnosed in fetal life

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Background: Agenesis of the pulmonary valve (APV) or dysplastic pulmonary valve (DPV) is a rare anomaly, occurring mostly as a variant of Tetralogy of Fallot (TF). Its features are pulmonary stenoinsufficiency of a variable degree with dilated pulmonary branches, causing cardiomegaly and potential lung hypoplasia.
Material – methods: Out of around 800 cases with congenital heart disease examined by fetal echocardiography during the past 10 years, 8 were found to have a pattern of APV/DPV at 21–32 weeks' gestation (w.g.). Six cases (group 1) had TF, while in 2 other cases (group 2) the anomaly was isolated. A global score was introduced to summarize the echocardiographic findings: z-score of PV anulus + degree of other findings in points 1,2,3.
Results: Group 1: Three fetuses had microdeletion 22q11.2, they developed at 25-26 polyhydramnios, progressing to fetal hydrops in 2; all died after preterm birth, one after operation. All these cases had a large pulmonary anulus (z-score >4), marked pulmonary insufficiency (PI), large dilated pulmonary branches, cardiomegaly and high global score. Three other cases had smaller pulmonary anulus (z-score 1.5 -3), major degree of pulmonary stenosis (PS), lesser degree of PI and cardiomegaly and lower global score; they were delivered at term and operated at 5 and 6 months, surviving. The ductus was absent in all.
Group 2: One case had a moderate PS and PI and is well, stable at 5 years. The second case had a peculiar form of valve dysplasia, with a marked PI and a markedly enlarged right ventricle, improved after birth, was operated at 2 years and is well at 5 years. The ductus closed spontaneously in both.
Conclusions: The data of our small series show a better outcome in the fetuses with APV associated to TF who had a major degree of PS than in those with a large pulmonary anulus and a marked PI. The isolated form presents probably a better prognosis.