Long term follow-up of fetal cases with tricuspid valve anomalies

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Objectives of the study: retrospective-prospective study of the characteristics and long-term outcomes of fetal cases with tricuspid valve anomalies, aiming to assess negative prognostic factors.

Material and methods: The echocardiographic data and long term outcome (6m-27yrs) of 41 fetuses diagnosed between 1986 – June 2012 as Ebstein (Ebst) or non-Ebstein anomaly (NE), at 21-37 week’s gestation (wg), median 29, were analysed. Nineteen cases had Ebst and 22 NE, one with mitral dysplasia and one with coarctation. Two fetuses had extracardiac anomalies, 3 Ebst had familial history of congenital heart disease, 1 mother was taking lithium and 1 gardenal. Following variables were compared in cases who died and in survivors: grade of tricuspid regurgitation (TR), Celermajer index (CInd), cardiothoracic ratio (CR), fetal hydrops (FH), pulmonary stenosis/ atresia (PS, PAtr).

Results: Echocardiographic features: 13/19 fetuses with Ebst had a moderate-severe displacement of the TV and moderate-severe TR at presentation, 3 had pulmonary stenosis (PS) and 5 pulmonary atresia (PAtr), 13/22 cases with NE had severe TR, 3 had PS and 5 PAtr. Seven had fetal hydrops (FH)–5 Ebst, 2 NE.
Outcome: Two Ebst and 1 NE opted for the termination of pregnancy, 3 Ebst died in utero (2 FH, 1 supraventricular tachycardia). Thirty six cases were delivered at 31-39 wg. Six neonates died spontaneously at 1-7days (3 Ebst, 3 NE). Five neonates with Ebst and 6 with NE were operated: 7 died, 4 NE survived. One Ebst died late at 3 yrs for resistant complex arrhythmias and severe worsening.
Total mortality was 17/38 cases (44.7%), 12/17 Ebst (70.6%), 5/21 NE (23.8%). Five of cases that died had FH, 2 severe arrhythmias, all a higher grade of TR, CInd>1, CR >0.65 and 9 had PAtr; the variables TR, CInd and PAtr were highly significantly different with respect to the survivors (p=0.002-0.006). Twenty one cases with milder forms are alive at 6m-27yrs, stable or improved (5 Ebst, 16 NE).

Conclusions: Our data confirm a relevant mortality of severe tricuspid valve anomalies diagnosed in utero, main negative prognostic factors being the grade of TR, CInd and PAtr. Milder forms of both variants stabilized after birth.