

Perinatal care of fetuses with heterotaxy syndromes

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Objectives: To evaluate the spectrum of prenatal findings and obstetric perinatal care of fetuses with heterotaxy syndromes.

Methods: A retrospective review of fetuses with heterotaxy syndrome. Fetal echocardiography findings, perinatal care and obstetric outcomes were evaluated.

Results: 37 fetuses with heterotaxy syndrome, 21(57%) with left isomerism(LAI) and 16(43%) with right isomerism(RAI) were diagnosed and delivered in our institution between January 2006 and December 2014. The main cardiac pathologies were AVSD 49%(18/37), pulmonary stenosis 32%(12/37) and pulmonary atresia 22% (8/37). Dextrocardia was in 22%(8/37). Other abnormalities in RAI: total or partially anomalous pulmonary venous drainage 44%(7/16), TGA 31% (5/16) and SV 31% (5/16). In LAI: common atrium 24% (5/21), DORV 19%(4/21), hypoplastic left ventricle 19%(4/21), interrupted inferior vena cava with azygos continuation in 52% (11/21). Rhythm disturbances: bradyarrhythmias were only observed in fetuses with LAI 57% (12/21). Situs inversus was the most common visceral anomaly in 41%(15/37) of all fetuses. Asplenia was in 22% (8/37) of fetuses, mainly in RAI 37%(6/16). Polysplenia was only seen in 14%LAI patients (3/21). The median gestational age at delivery was 38.6 weeks. 73% (27/37) were born by vaginal delivery, 16%(6/37) by elective cesarean section and 11%(4/37) by urgent CC. Excluding the parameter of bradycardia, abnormal intrapartum fetal heart rate patterns were observed only in two cases: reduced variability lasting more than 80 minutes (5,4%) and high variability lasting more than 10 minutes (5,4%). There were only 3 cases of Apgar score less than 7 at 5 minutes, and no cases of umbilical cord blood pH lower than 7.0. Out of 37 fetuses 20(54%) children, 13 with LAI (35%) and 7 with RAI (19%) alive at the time of publication.

Conclusions: The spectrum of cardiac and extracardiac abnormalities of HS is wide. Vaginal delivery is safe for this fetuses so CC should be reserved only for traditional obstetric indications. Electronic fetal monitoring is not always possible due to rhythm disturbances, especially in LAI.