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Features and outcomes of cases with laterality defects diagnosed in fetal life

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Background: From the postnatal data we know that congenital heart defects (CHD) associated to laterality defects or heterotaxy syndromes have a worse prognosis. Therefore, prenatal counseling is difficult in these cases. In this study we aimed to analyse the outcomes of this category of CHD diagnosed already during the fetal life.

Material and methodology: Out of 5800 fetuses at risk for CHD examined between 1995 and Dec. 2010 by echocardiography, 1150 had CHD and 71 (6.2%) presented an abnormal visceroatrial situs: 19 – left isomerism (Lisom), 17 – right isomerism (Risom), 12 - situs inversus with dextrocadia (SVI-dx), 4 - SVI with levocardia (SVI-levo) and 19 situs solitus with dextrocardia (SSol-dx). Anatomical features of the fetal heart, association with chromosomal or extracardiac anomalies (CA, ECA), course in utero and after birth were analysed retrospectively from the data base and clinical documentation.

Results: Fifty nine fetuses (83.1%) had associated CHD, in 2 with CA (trisomy 18 and deletion 6, in SVI-dx and SSol-dx) and in 3 with ECA. Among CHDs the most frequent was complex atrioventricular defect - in 29/59 (49%) cases, in Lisom, Risom, SVI-dx and SSol-dx, with atrioventricular block (AVB) in 3, followed by complex DORV (9 cases) and UVH (7cases, 2 with AVB); 3 fetuses had corrected TGA, 3-VSD, 2 - pulmonary atresia+VSD, 2-tricuspid atresia, and one each had truncus, coarctation, partial pulmonary venous drainage and DOLV. Twelve fetuses presented isolated anomaly of the visceroatrial situs (4/12 SVI-Dx, 2 /4 SVI-Levo, 5/19 SVS-dx and 1 Lisom). Outcome: 18/59 cases (30,5%) opted for the termination of pregnancy, 4 died in utero (2 Lis, 1 SVI-dx, 1 SVS-dx), 15 died after birth post surgery or pacemaker implant, 2 spontaneously. Total mortality was 20/41 cases with CHD continuing pregnancy (48.8%), 56.25% (9/16) in Lisom, 45% (5/11) in Risom, 50% (5/10) in SSol-dx, 2/3 in SVI-dx and none in SVI-levo. One of 12 cases without CHD who had ECA, died after birth.

Conclusions: Our fetal cases with laterality defects and CHD presented a relevant mortality, mainly those with isomerisms and SSol-dx. This fact should be taken in account at prenatal counseling.