Characteristics and outcomes of fetuses with laterality defects. Comparison of the newer outcomes data with the more remote ones.

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Background: Congenital heart defects (CHD) associated to laterality defects or heterotaxy syndromes have potentially a poor prognosis. Aim of this study was to analyse whether the outcomes of these defects diagnosed in fetal life modified in more recent years with respect to the previous era.

Material and methodology: Out of around 7000 fetuses at risk for CHD examined between 1995 and Dec. 2013 by echocardiography, 1320 had CHD and 81 (6.1%) presented an abnormal visceral atrial situs: 42 in the period till Dec. 2002 and 39 afterthen. In total - 19 had left isomerism (Lisom), 21 right isomerism (Risom), 12 - situs inversus with dextrocardia (SVI-dx), 6 - SVI with levocardia (SVI-levo) and 24 situs solitus with dextrocardia (SVS-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. The mortality results in two periods were compared.

Results: Thirteen cases (6.8%) showed normal intracardiac anatomy: 5/12 SVI-dx, 2/6 SVI-levo, 5/24 SVS-dx and 1 Lisom with an isolated anomaly of the systemic venous return. Out of the remaining 68 cases with CHD, the most frequent was complex atrioventricular defect (25 cases) - in Lisom, Risom, SVI-dx and SVSdx, with atrioventricular block (AVB) in 3, complex DORV and UVH, with AVB in one. Other fetuses had corrected TGA, VSD, pulmonary atresia+VSD, tricuspid atresia, truncus, coarctation, partial pulmonary venous drainage and DOLV. Six had associated chromosomal or extracardiac anomalies (in SVI-dx, SVI-levo, SVS-dx and Lisom). Outcome: 20/68 cases (29.4%) opted for the termination of pregnancy, 4 died in utero, 8 after birth spontaneously and 10 after operation or pacemaker implant. Total mortality in all cases with CHD continuing pregnancy was 22 (45.8%) Comparing the 1st and the 2nd period it was 50% vs. 28.6% in SVS-dx, 25% vs. 45.4% in Risom and 71.4% vs. 33.3% in Lisom.

Conclusions: Our fetal cases with laterality defects and CHD presented a relevant mortality, in both periods, mainly in cases with isomerisms and SVS-dx. The outcomes seem slightly better in more recent period, despite the limits due to the small numbers.