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A case of right isomerism

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Objective

Systematic segmental analysis of the heart helps in diagnosis of many complex cardiac defects antenatally.

Methods

Sequential segmental analysis of the fetal heart was done as per ISUOG guidelines.

Results

20 year old, G1P0, 22 weeks of gestational age was referred for fetal echocardiography due to suspected cardiac abnormality. Unremarkable personal and obstetrical history apart from consanguinity. The following findings were noted: Abdominal Situs: Ambiguous, Stomach: Left sided, liver in midline, gallbladder in midline, azygous vessel in the vicinity of aorta, IVC draining into the right atrium, Heart Size: cardiomegaly with minimal pericardial effusion. Apex: Right (Dextrocardia), FHR: 147 bpm, Atria were symmetric, Atrial septum primum deficient, normal foramen ovale with Normal flow (from right to left), AV Junction: concordant, normal AV connection, Ventricles: enlarged right. ventricle, left ventricle hypoplastic, Interventricular Septum: VSD, No flow seen across mitral valve. Stenotic ?, Outflow Tracts: pulmonary artery arising from RV, enlarged; aorta small narrow, 3 vessel view shows dilated MPA, absent aorta. Extra vessel seen on the left of MPA (persistent LSVC). The findings suggest complex cardiac anomalies - Heterotaxy syndrome (right isomerism), VSD, , hypoplastic left heart syndrome, persistent LSVC. The findings were confirmed on autopsy.

Conclusion

In heterotaxy syndrome, the normal symmetry of the thoracic and abdominal organs is lost, resulting in an unusual degree of asymmetry of organs and vessels. The term “isomerism”, derived from Greek (iso, meaning "equal, " and meros, meaning "part"), refers to this abnormal developmental symmetry in which morphologic structures that normally develop on one side are found on both sides of the body, and is the currently accepted term used to describe hearts with isomeric atria and atrial appendages. Prognosis depends upon the extent of visceral and cardiac involvement. In affected patients, instead of a distinct left and right side, individuals with isomerism will have either two right sides or two left sides resulting in either two right atria or two left atria (atrial isomerism). Systemic, segmental analysis of the fetal heart is mandatory for the diagnosis of heterotaxy syndromes which helps in prediction of neonatal outcome.