A case of left isomerism

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Objective
Heterotaxy (or situs ambiguous) is defined as the abnormal arrangement of thoracic or abdominal organs, or both, across the left-right axis. Right and left atrial isomerism, is found in between 2.2% and 4.2% of infants with congenital heart disease. Left atrial isomerism is associated with the presence of ‘double’ left-sided structures and right-sided structures are underdeveloped or absent. In a fetal series, left isomerism has been reported to be more common than right isomerism. Here we present a case of antenatally detected left isomerism.

Methods
This is a case report.

Results
A 36 year-old patient G2P1 was referred to our perinatology clinic due to fetal anomaly at 21 week gestation. She had no history of consanguinity and her personal and family history was unremarkable. In our ultrasound examination, a right-sided stomach was confirmed. Interruption of the inferior vena cava with azygos continuation was demonstrated as a ‘double sign’ in the upper abdomen in the axial plane. A parasagittal view of the abdomen and chest also demonstrated the azygos vein posterior to the descending aorta. Fetal echocardiography showed a double outlet right ventricle with an ativoventricular septal defect. Right aortic arch, persistent left superior vena cava and total anomalous pulmonary vein drainage were also detected. The patient was offered karyotyping and termination of pregnancy. But the patient refused invasive testing. She is currently at 30 weeks gestation and continues her antenatal follow-up. Left isomerism generally has poor prognosis, depending on the severity of the cases. Fetuses with left isomerism and heart block, are at risk of hydrops and intrauterine death.

Conclusion
Risk of recurrence in subsequent pregnancies have been reported up to 10% in some series. In postnatal survivors, associated cardiac defects determine morbidity and mortality in the neonatal period.