Objective
Human anatomy shows asymmetry of structures, especially internal organs, between the left and right side of the body. The establishment of the right and left axes in human embryogenesis is done early in development. The complete left-right reversal of all structures is situs inversus totalis. However, a combination of correct and incorrect-sided structures is situs ambiguous or more commonly referred to as heterotaxy syndrome. According to the development of right-sided and left-sided organs, heterotaxy syndromes are categorized in two types: left atrial isomerism (polysplenia) and right atrial isomerism (asplenia). We present a case of left atrial isomerism.

Methods
This is a case report.

Results
22-year-old G2P1L1. First examination was performed in our clinic at 12 gestational weeks. Mid-trimester ultrasound scan showed absence of inferior vena cava, presence of azygos vein and persistent left superior vena cava. The fetal bladder was seen in the midline. The stomach bubble was observed on the left side but much closer to the midline than normal. The fetus had no additional anomaly and was followed up to term, as the family requested the continuation of pregnancy. At 38 weeks 4 days, a caesarean section was performed due to previous caesarean section. A live female fetus 3240 gr with 8/10 APGAR was born. There was no early complication in a neonatal period and the neonate continues to grow healthy.

Conclusion
The identification of heterotaxy syndrome prenatally is very important. Fetal diagnosis helps us better inform expecting parents and plan appropriately the delivery. While early survival is possible, long-term outcomes are still uncertain. We need more management strategies for this unique cases.