Prenatal diagnosis of coarctation of the aorta in a fetus with asplenia and right atrial isomerism

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Objective
To present a case of prenatal diagnosis of coarctation of the aorta in a fetus with asplenia and right atrial isomerism.

Methods
This is a case report.

Results
The authors reported a case of a patient at 21 weeks of gestation. She was referred to Maternal Fetal Medicine (MFM) unit, Thammasat University hospital because of the diagnosis of dextrocardia. Ultrasonographic findings demonstrated a male fetus with stomach located on left side and the juxtaposition of the descending aorta and inferior vena cava on the right side of the spine. Fetal echocardiography revealed dextrocardia with obviously abnormal four-chamber view. Common atrium with common atrio-ventricular valve and common ventricle were identified. There was no connection between the pulmonary veins and the atriums thus total anomalous venous connection was diagnosed. Coarctation of the aorta, which is a rare condition in right isomerism, was found in the case while the pulmonary artery was normal. After counseling the prognosis of the fetus, the parents decided to terminate pregnancy. Autopsy findings revealed a male fetus with left-sided stomach, midline liver, left-sided gall bladder, asplenia and left-sided appendix. Both lungs had three lobes. Autopsy of fetal cardiovascular system confirmed echocardiographic findings.

Conclusion
This is a rare case of left-sided obstructive lesion in right atrial isomerism and is confirmed with pathological report. Hetrotaxy syndrome is a rare condition, the incidence is approximately 1 per 10,000 to 40,000 total births. In fetal series, atrial isomerism accounts for around 3 to 6 percent of all congenital heart defects. Several fetal echocardiographic studies report that right isomerism is less common than left isomerism and has tendency to affect males, while for left isomerism to affect females. Cases of asplenia and right atrial isomerism (RAI) are associated with congenital heart disease though cases with polysplenia and left atrial isomerism (LAI) may be found incidentally with no cardiac defect. Common congenital cardiac defects in right and left isomerism are dissimilar. Right atrial isomerism results in two right sides with bilateral right atria and atrial appendages and an absence of left-sided structures. Patients with RAI have a pattern of cardiac lesions that consist of anomalous pulmonary venous drainage, pulmonary artery stenosis or atresia and single ventricle physiology. Left atrial isomerism results in bilateral left atria and atrial appendages. The typical pattern of LAI is an interrupted inferior vena cava. Otherwise, there is greater variability of cardiac anomalies in LAI compared with RAI. Cardiac malformations are a major component of hetrotaxy syndrome, resulting in significant morbidity and mortality. Prenatal diagnosis with routine ultrasonography and echocardiography is worth.