Left atrial isomerism associated with lung agenesis
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Objective
To report a case of unilateral lung agenesis and left atrial isomerism/polysplenia syndrome diagnosed by ultrasound and magnetic resonance imaging (MRI).

Methods
Case report.

Results
We report about a 24 years old Gravida II with non-consanguine marriage who presented for her first scan at 25 weeks gestation. The ultrasound examination showed right renal agenesis, single umbilical artery and polyhydramnios. Fetal echocardiography disclosed a complex heart disease with left atrial isomerism, pulmonary atresia, right ventricular hypoplasia, and dysplasia and hypoplasia of the tricuspid valve. Fetal MRI confirmed agenesis of the left lung. A female baby was born at 38 weeks gestation (2840g) and died minutes after birth. No external abnormality or dysmorphism was noted at the postmortem examination. Autopsy showed umbilical cord with two vessels; left pulmonary agenesis with complete absence of lung tissue, bronchus and vascular supply; tracheoesophageal fistula and right hypoplastic bronchus and lung with subpleural and interstitial emphysema. Heart analysis disclosed, beyond the findings described above persistent left superior vena cava draining into the coronary sinus and absence of supra-hepatic segment of inferior vena cava; direct drainage of the hepatic veins to the atrium on the right and drainage of two pulmonary veins into the atrium located on the right; ambiguous atrioventricular connection; hypoplastic and imperforate pulmonary valve; pulmonary trunk with absence of left pulmonary ramus and severe hypoplasia of the right branch which communicates to the hypoplastic lung; absence of atrial septum and patent ductus arteriosus. There was also polysplenia and right renal agenesis. The left kidney had an ureteropelvic dilatation. The liver was located in the midline and the biliary tract was permeable.

Conclusion
In our review of the literature, we did not find association between lung agenesis and left atrial isomerism/polysplenia syndrome. Pulmonary findings usually observed in this condition consist of abnormalities in lung segmentation. More reports will be important to define a possible association.