A case of of congenital partial agenesis of the fetal portal veins
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Objective
The agenesis of the portal vein or its branches known as ‘Abernity Malformations’ is a rarely diagnosed congenital anomaly in which a certain amount of portal blood flow is diverted into the systemic venous circulation. Here, we introduce a case of congenital partial agenesis of the portal vein (type 2) without associated congenital anomalies, that is prenatally diagnosed.

Methods
A 35-year-old G3P1 patient, referred to our division of Maternal Fetal Medicine at 29 weeks of gestation due to fetal growth retardation. On the first examination, she was afebrile (36.6 °C), and had normal blood pressure (100/70 mmHg). Past medical or family history revealed no significance and she had no drug, alcohol or tobacco use. Mild cardiomegaly, splenomegaly and dilatation of umbilical vein were found on the ultrasound scan and fetal growth was restricted for gestational age. The ductus venous was visible and accepted as normal with triphasic blood flow pattern on Doppler ultrasound. Amniotic fluid volume was within normal ranges. Fetal cardiac echocardiography was performed and normal fetal cardiac anatomy was demonstrated on multiple planes. The free cell DNA test was performed and results showed low risk for genetic abnormalities. At 38 weeks of gestation, a 1900 grams term male infant, with an APGAR score of seven at 1st minute and score of eight at 5th minute was delivered by cesarean section. Abdominal ultrasound was performed on the first day of life and it revealed splenomegaly and a dilated umbilical vein passing through the liver and then draining to vena cava inferior. The portal vein was interrupted at the portal hilum. There was no left portal vein branch and the right portal vein was observed as rudimentary. There were intrahepatic collateral veins between the umbilical and the portal vein. Therefore, patient was diagnosed with ‘Abernity Malformations’ (congenital partial agenesis of the portal veins- type 2).

Results
Our case has shown that congenital partial agenesis of the portal veins can be diagnosed in utero. Yet, in order to demonstrate this rare venous anomaly, the mid-trimester fetal anatomy scan should include the evaluation of the umbilical vein insertion, continuation and calibration of umbilical vein and its branches. In the case of umbilical vein dilatation, existence of portal vein and its branches must be evaluated. The spleen size and presence of ascites must be noted. Fetal growth restriction with cardiomegaly is usually associated with the ‘Abernity Malformations’. The outcome of congenital agenesis of the fetal portal venous system is clearly associated with the type of the disorder. Total agenesis is classified as Type 1 (classical Abernity) abnormality, in which there is complete diversion of the portal blood into the inferior vena cava (portosystemic shunt). This anomaly has been reported only in girls and is usually associated with other congenital abnormalities such as cardiac defects, biliary atresia and polysplenia. Partial agenesis is accepted as Type 2 abnormality, in which the portal vein may exist, but a certain amount of portal blood is diverted into the systemic venous circulation (portohepatic shunt). In Type 1 with additional anomalies, parents could choose termination of the pregnancy while Type 2 has a favorable prognosis. Parents should be informed about possible future complications and the various treatment options.

Conclusion
The dilatation of umbilical vein, splenomegaly, mild to severe cardiomegaly and lack of portal vein and its branches are alarming features in mid trimester fetal anomaly ultrasound scanning for ‘Abernity Malformations’.