Agenesis of ductus venousus in three fetuses: A rare reason of fetal cardiomegaly

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Objective: Agenesis of the venous duct (AGV) is a rare congenital anomaly resulting in abnormal drainage of the umbilical vein into the fetal venous circulation and is associated with a high incidence of fetal anomalies and adverse outcomes, including associated malformations, chromosomal aberrations and in utero heart failure. The umbilical venous flow may take various pathways. The clinical presentation and prognosis is variable, and may depend on the specific drainage pathways of the umbilical vein.

Methods. We described 3 fetuses with agenesis of ductus venousus. Indications for fetal echocardiography were cardiomegaly, extracardiac anomaly and suspicion of cardiac anomaly, respectively. First fetal echocardiography was performed 29-33th weeks of gestation. Echocardiograms showed that all of them have marked global cardiomegaly and dilated umbilical vein. In two fetuses the umbilical vein bypassed the portal sinus and the liver and connected to the right atrium directly. One of them had unilateral renal agenesis and the other had left renal hypoplasia. In the third fetus, absence of the ductus venosus with connection between the umbilical vein (UV) and a dilated inferior vena cava was detected. Early delivery was planned in all of them between 34-36th weeks because of hemodynamic compromise. All patients survived without any intervention. One patient had signs of congestive heart failure and required medication at early postnatal period.

Conclusions. An ADV should be ruled out in a third trimester fetus with unexplained cardiomegaly or dilatation of the umbilical vein, systemic veins, or portal sinus. The fetus with an ADV must be followed for signs of congestive heart failure due to volume overload, including cardiomegaly, tricuspid valve regurgitation, effusions, and hydrops. Worsening of fetal heart failure may be an indication for early delivery. Beside this, associated malformations and chromosomal aberrations must be investigated. Key words: absent ductus venosus; fetal echocardiography; prenatal diagnosis.

Figure: a. Absence of the ductus venosus with a direct connection between the umbilical vein and the right atrium. b. Absence of the ductus venosus with a direct connection between the umbilical vein and the right atrium.