Ductus venosus agenesis: prenatal diagnosis and feto-neonatal outcome of 35 cases

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Objective
To evaluate the prenatal sonographic findings associated with absent ductus venosus (ADV) and the feto-neonatal outcome.

Methods
This is a multicentre observational study, including both low risk patients recruited by first and second trimester ultrasound screening examination, and referral patients sent to our centres for suspected fetal anomaly. In all cases of ADV, prenatal sonographic findings, associated anomalies and feto-neonatal outcome have been evaluated.

Results
35 cases of ADV have been diagnosed: 20 had intrahepatic drainage of the umbilical vein and 15 fetuses had extrahepatic drainage; 23 cases were diagnosed during the first trimester examination and the remaining 12 cases in the II/III trimester. The anatomical arrangements of the extrahepatic umbilical vein drainage (UVD) was as follows: 5 drained directly into the right atrium; 9 drained into the inferior vena cava and 1 drained into the right iliac vein. Seven of these fetuses developed cardiac failure. Fetal karyotyping was abnormal in 7/26 (20%) cases that underwent invasive procedure: in all these fetuses other major anomalies and/or increased NT were associated. Six patients opted for termination of the pregnancy. One case of spontaneous intrauterine demise and 2 cases of neonatal death occurred. One case was lost at the follow up. Among the 25 surviving fetuses, 20 cases were asymptomatic after birth: most of them had UV intrahepatic drainage pattern.

Conclusion
Increased NT and major anomalies represent risk factors for chromosomal anomalies in fetuses with ADV. Association with structural and chromosomal anomalies are the most important poor prognostic signs. In cases of extrahepatic UVD, even if isolated, the risk of congestive heart failure is significant.