Agenesis of ductus venosus: what's the real clinical relevance?

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Background: Anomalies of the Ductus Venosus (ADV) are highly associated with poor fetal outcome (aneuploidy, structural malformations and fetal hydrops). Prognosis is variable and depends on such anomalies and the type of drainage (to portal sinus: intrahepatic or extrahepatic). Objective: describe real clinical consequences of this anomaly in two tertiary referral centers.

Methods: Prospective study (January 2005 to June 2012) from two tertiary centers where a complete first trimester scan (DV Doppler assessment) has been performed. All cases underwent serial fetal echocardiography and Necropsy or postnatal follow-up.

Results: we report 51 cases of ADV (3 twin pregnancies), 42% of them were diagnosed in first trim scan. 33(65%) had intrahepatic (IH) umbilical venous drainage and 18(25%) had extrahepatic (EH) drainage, either directly into right atrium (5), Inferior Vena Cava (7) or the Iliac Vein (2). We registered development of severe hydrops (IH 6%, EH 22% (P<0.05)), associated anomalies (IH 24%, EH 39%) and increased NT (IH 24%, EH 16%, among the IH we had 1 CATCH and 1 Noonan Sd.). The overall survival was 69% (IH 85%, EH 39% (P<0.01)) but Isolated cases of ADV had 97% survival. We had 2 fetal demise and 12% underwent TOP (IH 6%, EH 22%). Survivors are healthy children (1 Aortic Coartation and 1 Ventricular septal defect operated on, 1 left isomerism without CHD and 1 renal polycystosis). 2 hydropic newborn had persistent porto-systemic shunt.

Conclusions: Fetuses with IH umbilical venous drainage have better prognosis than EH drainage. The absence of associated malformations, hydrops or increased NT has excellent prognosis.