Absent ductus venosus: different perinatal outcome related to anatomy

ABSENT DUCTUS VENOSUS : DIFFERENT PERINATAL OUTCOME RELATED TO ANATOMY

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Congenital absence of the ductus venosus (ADV) is a rare condition which can present with several anatomic settings and associated to congenital anomalies of other systems. Different clinical patterns in the fetus and the newborn can emerge therefore. We report two cases of ADV with opposite perinatal outcomes. **Case #1.** A 28 y.o. black woman came at 22 weeks' gestation (GA) for mid-pregnancy evaluation. She showed polyhydramnion (amniotic fluid index (AFI)= 261mm) associated to normal cardiac anatomy and normal karyotype (46,XX). The enlarged umbilical vein (UV) showed a pulsatile pattern at echoDoppler with direct connection to the RA. A fistula between the UV and the iliac artery was evidenced by colorDoppler as well. Despite normal ventricular contractility (EF) and diastolic function (E/A ratio) at echocardiographic monitoring, fetal cardiac enlargement progressively occurred with mild pericardial effusion. At 28 GA placental detachment occurred and a female infant (BW 915g, <10°p) was born with severe perinatal asphyxia. The infant died at 5 hours of life from severe acidosis refractory to intensive care and resuscitation efforts. Postmortem evaluation confirmed the anatomic pattern and absence of the portal vein (PV) was demonstrated as well. **Case #2.** A 35 y.o. white woman was admitted to our tertiary care at 33 GA because of monolateral renal agenesis and unique umbilical artery. Despite ADV, mesocardia and mild cardiac enlargement the fetus was stable (normal diastole and contractility). The UV echoDoppler showed a normal flat pattern at the beginning of its abdominal course but it progressively became pulsatile as the UV run cephalad to the heart. The infant was born at 38 GA from a planned cesarean delivery (BW 3080g). The perinatal adaptation, karyotype and phenotype were normal. Echocardiography in the newborn showed normal diastole and contractility (E/A ratio, LVEF, LVDd). The associated congenital anomalies were confirmed.

DISCUSSION. ADV is a rare anomaly in which perinatal prognosis is difficult to predict and clinical presentation can vary greatly due to the different patterns, i.e. fetal cardiac failure, associated congenital anomalies, polyhydramnion. We reported 2 cases of ADV with opposite clinical course. As it often occurs, case #2 was detected occasionally, late in pregnancy, with a good hemodynamic status despite some malformative features. The normal perinatal transition shifted the cardiovascular system to a setting which did not need any DV activity in regulating venous return and the neonatal course was asymptomatic.

Conversely case #1 showed fetal hemodynamics impaired since the beginning of the 3rd trimester. Maybe this could be the consequence of a huge hemodynamic overload due to the presence of both ADV and veno-arterial fistulas emphasizing the diastolic overload of a direct connection of the UV to the RA. The PV was also absent and this has been decribed as being related to a negative prognosis.

It is still difficult to completely understand why some fetuses can tolerate the missing function of the DV in regulating the systemic venous return while others do not. To monitorate fetal cardiac function by echoDoppler can be helpful but it is not a standard yet. So case-by-case detailed evaluation of complete anatomy and analysis of both diastole and contractility remains the better choice. Finally the Obstetrician will be mandatory as any modification in the course of pregnancy can be life-threatening due to the thin hemodynamic balance of these fetuses.