

CASE REPORT

ABSENT DUCTUS VENOSUS WITH SINGLE UMBILICAL ARTERY: A RARE ASSOCIATION

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ABSTRACT: Absent ductus venosus is a rare anomalous venous connection in fetus where the fetal outcome depends on type of venous connection and associated anomalies. Poor outcome is reported where the umbilical vein directly drains in to Rt atrium bypassing liver. Better resolution of ultrasound with Color Doppler makes possible to detect the anomaly early. Here we are presenting a case of antenataly diagnosed absent ductus venosus with umbilical vein directly draining in to Rt atrium bypassing liver. Case was diagnosed by Ultrasonography with color Doppler and MRI study.

INTRODUCTION: Normally in fetal circulation Oxygen-rich blood is carried by the umbilical vein from the placenta to the fetus. The umbilical vein enters at the umbilicus and gives two branches: right umbilical vein passing to the right side of the liver, is joined by the portal vein, and finally drains into the inferior vena cava (IVC) via the hepatic veins. The left umbilical vein gets shunted via the ductus venosus into the IVC bypassing the hepatic circulation and shunting the oxygenated blood primarily to fetal brain and myocardium. Due to such a critical role ductus venosus has an upper hand in conjunction to other fetal shunts. Here we present a case of absent ductus venosus with single umbilical artery and other associated anomalies which was diagnosed during routine ultrasound examination and subsequently examined by 3 Tesla MRI.

CASE REPORT: A 27 year old female with obstetric history gravida-4 para-3, came to our hospital for routine obstetric ultrasound check-up at 18 wk of gestation. She has no any complain and no any significant obstetric history. On ultra sound finding there was no fusion of amniotic membrane with chorion (Fig. 1). There was bilateral hydronephrosis more on left kidney (Fig. 2). Umbilical cord showed single umbilical artery and vein (Fig. 3). There was absent ductus venosus and umbilical vein drains directly into the right atrium (Fig. 4). Mild cardiomegaly was also noted. Musculoskeletal anomalies were seen in the form of mal-alignment of spine (Fig. 7). There was early onset IUGR. Fetal MRI was done in 3 Tesla MRI machine which shows hypointense bend from cord insertion to cardiac shadow, in correlation with the ultra sound finding it was interpreted as anomalous venous course and insertion of umbilical vein into the right atrium. Further biochemical investigation were done, which showed significant raised levels of triple marker i.e. alpha feto-protein (81.5ng/ml), beta HCG (13540mIU/ml), un-conjugated Estriol (3.58ng/ml) and Estradiol (4300H). After 15 days she delivered a dead male fetus by vaginal delivery. There was no external abnormality on fetus except small for gestational age.

DISCUSSION: Ductus venosus connect the portal and embryonic venous circulation into the inferior vena cava and plays an important role in fetal circulation. Because of the narrow lumen of the ductus venosus measures one third of the diameter of the umbilical vein, the blood is shunted to the IVC under pressure. This increased pressure pushes the blood through IVC into the right atrium and directly into the left atrium via, foramen ovale. The blood then passes into the systemic circulation through the left ventricle and aorta. The blood flow pattern seen on pulse wave Doppler shows a

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systolic peak, a diastolic peak and a nadir during atrial contraction. It allows identification of ductus venosus from adjacent hepatic veins (Showing triphasic flow). Absence of ductus venosus is a rare anomaly with a prevalence of 1 in 2532,¹ in which the umbilical vein connection to the venous system may be extrahepatic, bypassing the liver or intrahepatic via the portal venous system.

There can be at least four main patterns of abnormal venous circulation when the ductus venosus is absent.^{2,3} (1) The umbilical vein drains into the right atrium directly or through a dilated coronary sinus: considered the commonest type³ and carries worst prognosis due to risk of developing congestive cardiac failure. (2) The umbilical vein shows direct connection into the inferior vena cava. (3) The umbilical vein drains into the iliac or renal vein. (4) Umbilical vein drains into the portal vein. First case of absent ductus venosus was reported by Paltauf in 1888, an infant born with severe hydrops, portal congestion and absent ductus venosus.⁴ Fetuses with absence of the ductus venosus are at risk of other congenital anomalies including facial clefts, hemi vertebrae, cardiac, genitourinary, gastrointestinal anomalies; affected infants also have a poorer prognosis.⁵ It has been seen in various studies that patient with extra hepatic umbilical venous drainage have poorer outcome and association with other structural defects make the postnatal prognosis worst. As in a cohort study by Berg et al, a significant association was demonstrated between extra hepatic umbilical venous drainage, portal vein agenesis and cardiomegaly, leading to severe post natal complication.

The prognosis of isolated absent ductus venosus is more favorable in the presence of intra-hepatic venous drainage.⁶ However some studies also propose better outcome in patients with intra-hepatic venous drainage in absent ductus venosus, if no associated anomalies are present. As in the study by Berg et al⁶ the fetus with no or minor associated anomalies along with intra-hepatic drainage showed better postnatal status and had no long term sequelae. Postnatal complication may be seen in the form of pulmonary edema, hepatic tumor, and congestive heart failure. Cardiomegaly is seen mostly in cases showing direct drainage of the umbilical vein into heart suggesting high central venous pressure. This might be due to volume overload because of loss of regulatory mechanism by ductus venosus.^{3,7,8,9,10} Single umbilical artery has not shown any co-relation with absent ductus venosus in the literature; however multiple aneuploidies have been reported.¹¹ Here we are presenting a case of absent ductus venosus with single umbilical artery which is extremely rare and other anomalies in form of mal-aligned spine, hydronephrotic kidneys. Further we add visualization of anomalous venous drainage of umbilical vein in to the Rt atrium in fetal MRI study.

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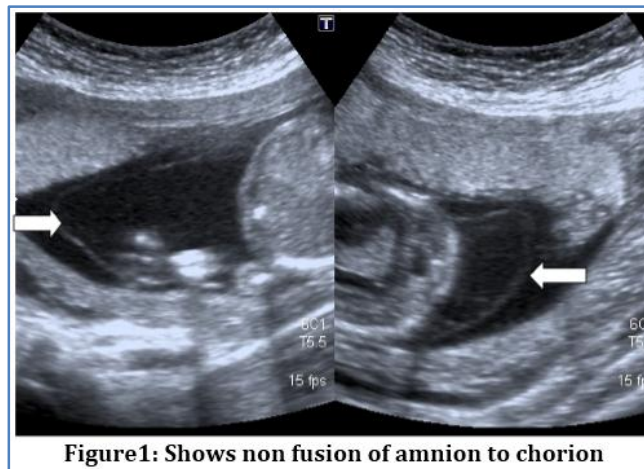


Figure1: Shows non fusion of amnion to chorion

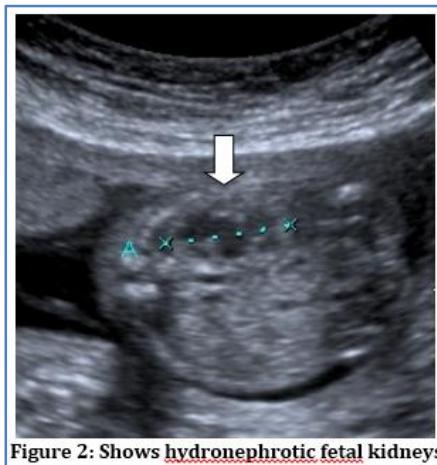


Figure 2: Shows hydronephrotic fetal kidneys

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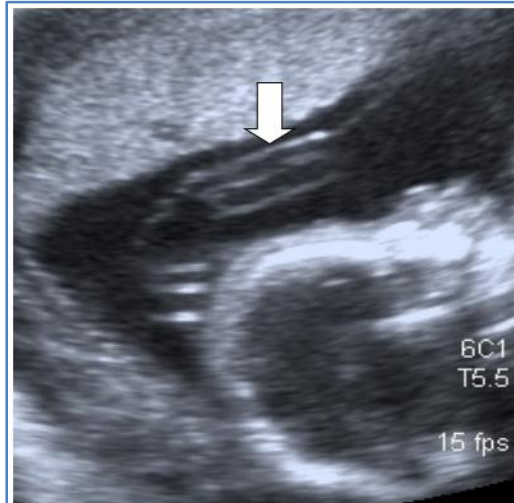


Figure 3: Shows single umbilical artery

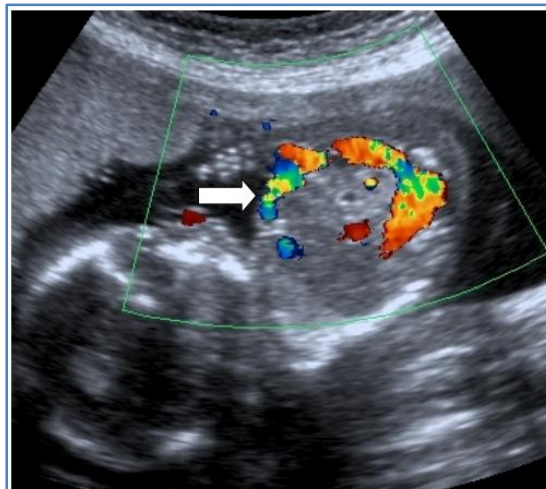


Figure 4: Shows color Doppler picture of umbilical vein draining in to Rt atrium

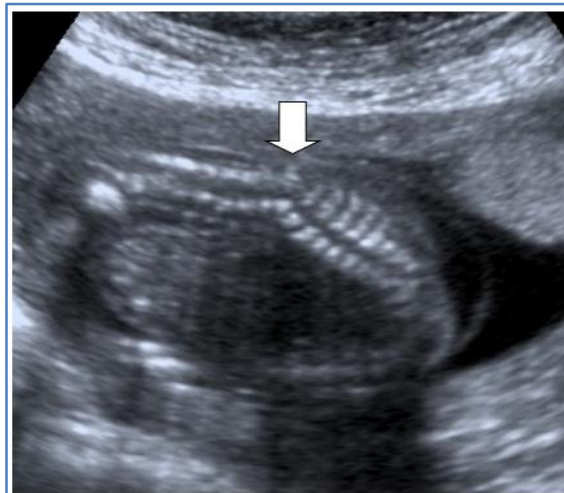


Figure 5: Shows mal-alignment of fetal spine

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Figure 6: Show MRI T2 WI extra-hepatic Umbilical vein draining in Rt atrium

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