Prenatal Sonographic Diagnosis of Rare Fetal Anomaly

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Section 2 – Answer

Case report

A 24-year-old primi-gravida female came for routine prenatal ultrasound at 26 weeks of gestation. Ultrasonography showed polyhydramnios (Amniotic fluid index: 26 cm) and a cardiac anomaly as evaluated on gray scale [Figures 1 and 2] and color Doppler images [Figure 3]. There was no other structural anomaly seen. What is your diagnosis?

Interpretation

Ultrasonography showed dilated fetal umbilical vein (UV), crossing the diaphragm and directly draining into the right atrium (RA). The RA was dilated [Figures 1-3]. There was no other structural anomaly seen in this case; hence, a diagnosis of isolated absent ductus venosus (DV) was made.

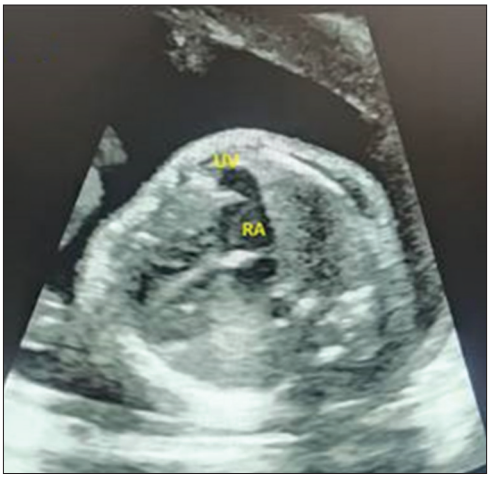

Figure 1: Gray scale image of fetal heart shows mild dilated right atrium

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Discussion

DV agenesis is a rare anomaly which is significantly associated with cardiac, extracardiac, and chromosomal anomalies.[1,2] Associated anomalies ranging from isolated cardiac anomalies like ventricular septal defect to complex anomalies like Dandy-Walker syndrome, corpus callosum agenesis, and chromosomal anomalies like trisomy 18 and 21 have been documented.[3,4] With or without associated anomalies, risk of developing fetal hydrops and intrauterine cardiac failure remains high.[3,4] DV is an important fetal shunt connecting intra-abdominal UV to fetal inferior vena-cava near its entry to heart.[3,5] In case of agenesis of DV, the blood from UV flows through alternative vasculature that may either be extrahepatic or intrahepatic system (via the portal venous system). Multiple variable connections have been seen in extrahepatic shunts. Bypassing the liver, the blood from UV is directly shunted to the heart via one of the multiple channels viz., inferior vena cava, iliac vein, renal vein, or to the RA and rarely to left

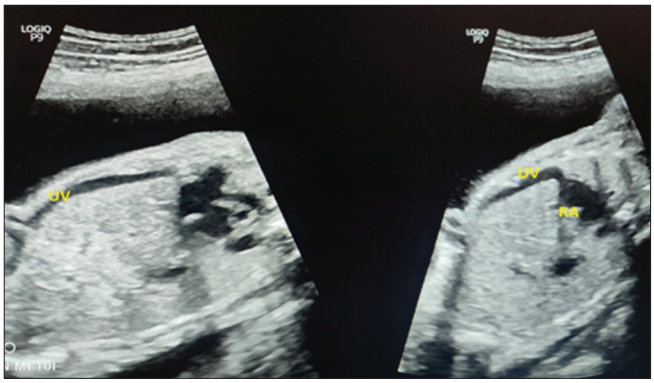

Figure 2: Gray scale image showing dilated fetal umbilical vein (UV), crossing the diaphragm and directly draining into the right atrium (RA)

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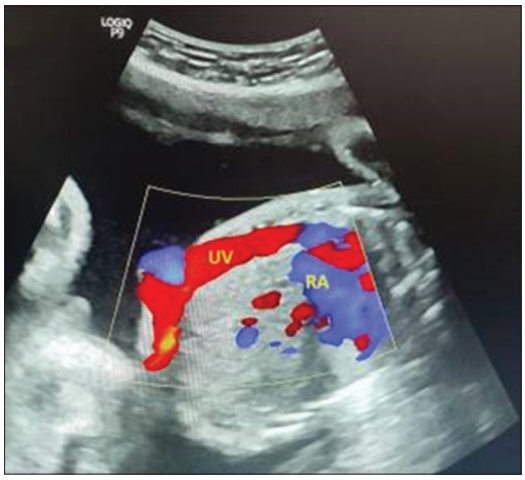

Figure 3: Colour Doppler image showing dilated fetal umbilical vein (UV), crossing the diaphragm and directly draining into the right atrium (RA)

atrium or coronary sinus. In intrahepatic shunts, the UV shows usual connection through portal sinus to hepatic sinusoids but without giving rise to the DV.[1,3-6] Prenatal ultrasound may be helpful in early diagnosis.[2] In our case, the route for umbilical venous return was extrahepatic umbilical venous drainage, where the UV was seen shunting blood directly into the RA. Although no other structural anomaly was seen, associate findings of polyhydramnios increased the risk of poor prognosis. Review of literature of absent DV suggests poor intrauterine as well as perinatal prognosis. The would-be parents in our case were thoroughly counseled about poor prognosis and they opted for termination of pregnancy.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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