Prenatal Diagnosis of Aorta-Portal Vein-Umbilical Vein Anastomosis

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Section 1 – Quiz

Case 1

A 36-year-old Rhesus (Rh)-positive, gravida 2, para 1 woman was referred at 36 weeks of gestation in view of hepatomegaly noted in the fetus during third-trimester scan. High-resolution ultrasound examination (Voluson E 10, Wipro GE Healthcare Private Limited, Austria) showed single intrauterine fetus with normal growth parameters, hepatosplenomegaly (liver span 75 mm [95th centile = 62 mm], spleen transverse diameter 63 mm [95th centile = 55 mm]), placentomegaly, and a uniformly echogenic bowel. There was polyhydramnios; however, there were no signs of hydrops. Detailed echocardiographic examination showed structurally normal heart with cardiomegaly (cardiac circumference to thoracic circumference ratio: 0.66), normal ventricular function, and pericardial effusion. Doppler examination of umbilical artery showed high-resistance flow; middle cerebral artery (MCA) showed increase in diastolic flow with increased peak systolic velocity [Figure 1a]; ductus venosus showed normal flow. The picture was suggestive of a hyperdynamic circulation. We noted an intrahepatic arterial connection between portal vein and descending aorta showing high-velocity flow in the connecting vessel [Figure 1b]. Mesentery was echogenic depicting mesenteric steal phenomenon. There were no other associated gross anomalies. The patient had spontaneous onset of labor at 36 weeks 4 days and had a vaginal delivery. The baby expired 3 h after delivery, due to high output cardiac failure.

Case 2

A 31-year-old Rh-positive, gravida 2 para 1 woman was referred at 15 weeks 5 days of gestation with cystic hygroma for the second opinion. Ultrasound examination showed single intrauterine fetus corresponding to gestational age. The fetus had cystic hygroma, pleural effusion, generalized skin edema, and hepatomegaly of 25 mm (more than 95th centile). Detailed echocardiography revealed tricuspid as well as mitral regurgitation, otherwise normal heart with no pericardial effusion. Doppler examination showed an abnormal vascular shunt connecting descending aorta and umbilical vein with high-velocity flow [Figure 1c and d]. MCA showed increased systolic flow.

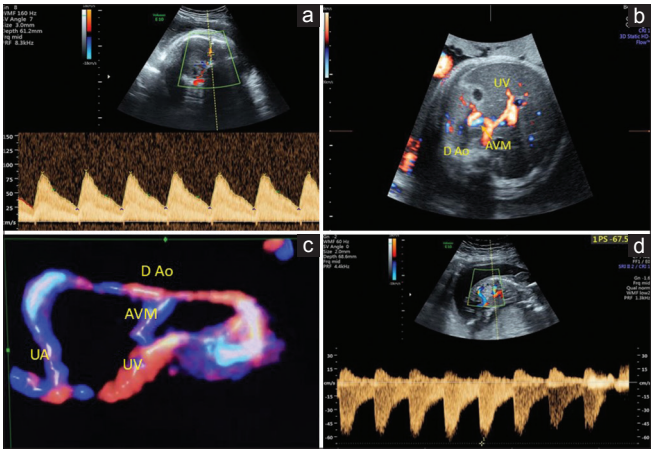

Figure 1: Ultrasound images of intrahepatic arteriovenous malformation. (a) Middle cerebral artery Doppler showing low resistance flow. (b) Color Doppler showing anomalous connection between descending aorta and inferior vena cava. (c) Four-dimensional rendered image of arteriovenous malformation between descending aorta and umbilical vein. (d) Longitudinal section of fetal abdomen showing anomalous arteriovenous connection with high-velocity flow. D Ao: Descending aorta, UV: Umbilical vein, UA: Umbilical artery, AVM: Arteriovenous malformation, PSV: Peak systolic volume

Declaration of patient consent

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

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