Prenatal Diagnosis of Aorta-Portal Vein-Umbilical Vein Anastomosis

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Section 2 – Answer

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 (PSV) [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.

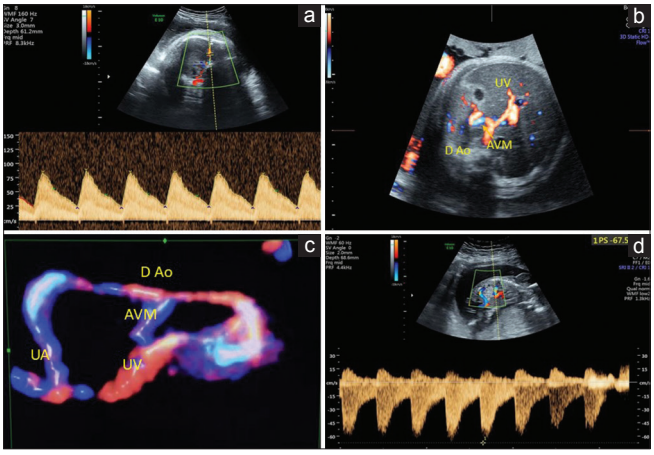

Figure 1: Ultrasound images of intrahepatic arteriovenous malformation. (a) Middle cerebral artery Doppler showing low-resistance flow. (b) Color Doppler showing anomalous connection between descending aorta and inferior vena cava. (c) Four-dimensional rendered image of arteriovenous malformation between 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

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Discussion

Congenital hepatic arteriovenous fistulae (AVFs) occur in less than 1:100,000 live births and are characteristically associated with poor perinatal outcome.[1,2] Prenatal diagnosis has been described, mostly secondary to nonimmune hydrops.[3,4] Hepatic arteriovenous malformation (AVM) can be of three types: a direct communication between a systemic artery and a hepatic vein (hepatic AVF), a communication between the hepatic artery and the portal venous system (hepatoportal fistula), and multiple AV microfistulae as part of the hereditary hemorrhagic telangiectasia or hemangioma.[5]

Our case was a hepatic AVF with AV connection. As systemic blood pressure is higher on the arterial side, there is a progressive dilation of the venous drainage, resulting in the characteristic sonographic findings of dilated vascular channels within the liver. As more blood is shunted through this low-resistance, high-flow outlet, fetal cardiac output rises concomitantly to meet the increasing and competing demands of fetal growth and the AVM "steal." This process might result in high-output heart failure, hydrops, or even Kasabach–Merritt sequence (microangiopathic hemolytic anemia, thrombocytopenia, and consumptive coagulopathy).[6]

An important finding in both cases was a high PSV in MCA with low-resistance flow, which prompted the search for an abnormal AV connection. This finding is seen in fetal anemia caused by parvovirus infection, Rh alloimmunization, or metabolic disorders. This might be utilized as a cue to look for abnormal AV connections in cases with nonimmune hydrops.

At birth, when the systemic vascular resistance rises, more blood flows to and through the AVM low-resistance shunt worsening the clinical condition.[7,8] When our case was diagnosed *in utero*, polyhydramnios, fetal cardiomegaly, and fetal distress were already present, suggesting significant hemodynamic compromise.

At present, invasive *in utero* treatment is not available, and hence, prenatal diagnosis allows for referral and delivery at a tertiary center with invasive interventional radiology and pediatric surgery services. Ideally, the baby should be delivered at term. Postnatal treatment of hepatic AVMs consists of obliteration of the feeder vessels with surgical ligation or percutaneous transcatheter coil embolization.[7]

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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Conflicts of interest

There are no conflicts of interest.

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