

Fetal Aorta-Portal Vein-Umbilical Vein Anastomosis: Prenatal Diagnosis and Brief Review of Literature

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Abstract

Keywords

- ▶ congenital hepatic arteriovenous fistulae
- ▶ fetal aorta-portal vein- umbilical vein anastomosis
- ▶ fetal growth restriction
- ▶ hepatic arteriovenous malformation
- ▶ pseudo ductus venosus waveforms

Congenital hepatic arteriovenous fistulae (AVFs) are rare and occur in less than 1:100,000 live births and are linked with poor perinatal outcome. Hepatic arteriovenous malformation can be of three types, out of which we present a case of hepatic AVF with AV connection in a case of primigravida at 26 weeks gestation presented with fetal growth restriction and Doppler changes.

Case Details

A 26-year-old Rhesus (Rh) positive, primigravida was referred at 26 weeks of gestation for a fetal growth and Doppler study. High-resolution ultrasound examination (Voluson E 10, Wipro GE Healthcare Private Limited, Tiefenbach 15 4871 Tiefenbach, Austria) showed a single intrauterine fetus with early onset fetal growth restriction (estimated fetal weight on second centile) and oligohydramnios (single largest pocket 2.6 cm). Detailed echocardiographic examination showed a structurally normal heart with normal ventricular function and no pericardial effusion. Doppler examination of the umbilical artery showed high-resistance flow; the middle cerebral artery showed an increase in diastolic flow and

brain sparing effect (cerebro placental ratio <1) and the ductus venosus showed normal flow.

We noted an intrahepatic arterial connection between the portal vein and descending aorta showing high-velocity flow in the connecting vessel. There were no other associated gross anomalies (▶ **Fig. 1**, ▶ **Video 1–4**).

Video 1

Aorta portal umbilical vein shunt. Online content including video sequences viewable at: <https://www.thieme-connect.com/products/ejournals/html/10.1055/s-0043-57019>.

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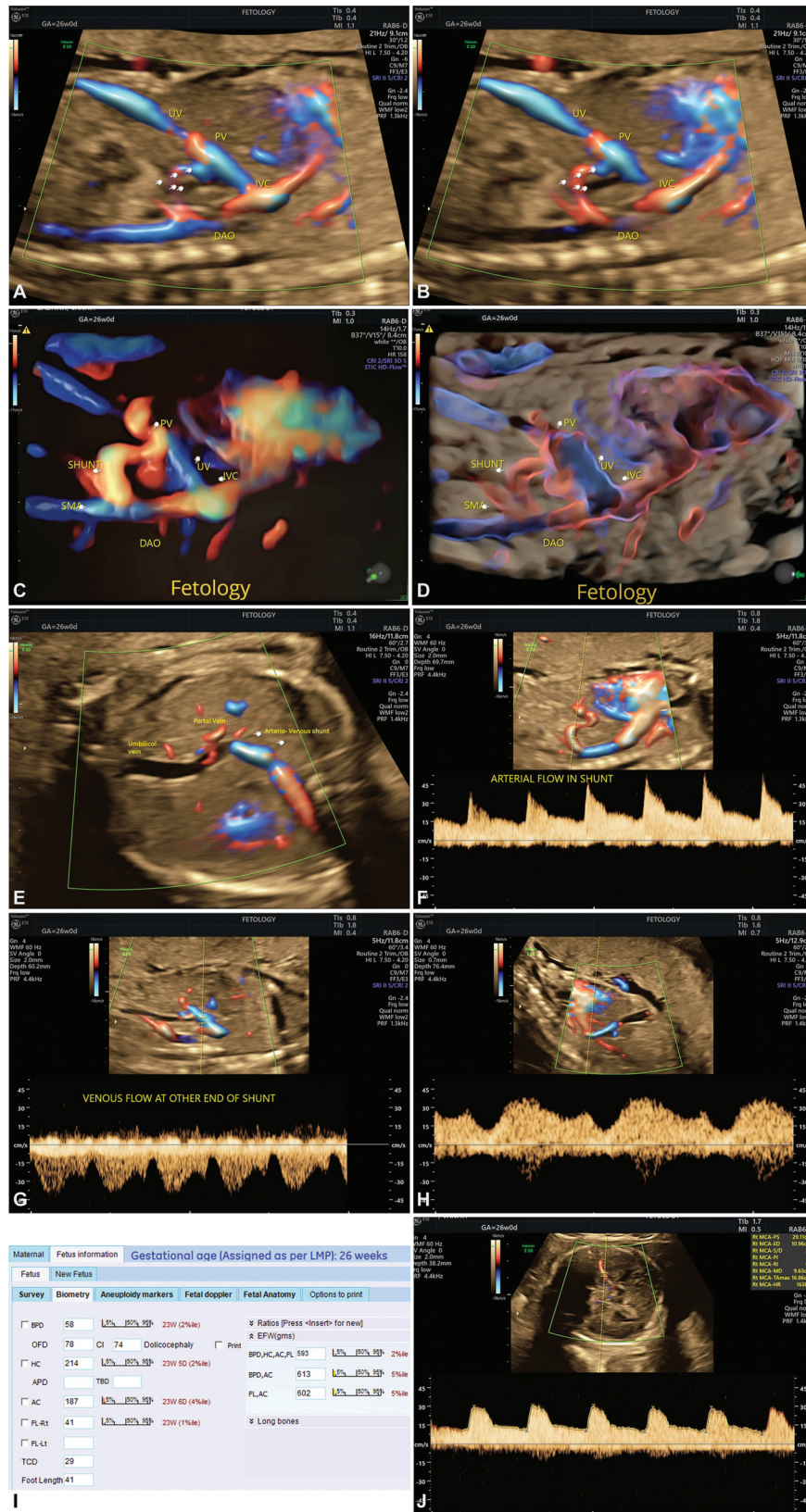


Fig. 1 Ultrasound images of fetal abnormal aorta-portal-umbilical vein (UV) shunt. (A) Color Doppler showing UV directly opening into the inferior vena cava (IVC) instead of opening into the vestibule. (B) Color Doppler image of arteriovenous malformation between descending aorta and UV. (C, D) Four-dimensional rendered image of arteriovenous malformation between descending aorta and UV. (E) Transverse section of fetal abdomen showing anomalous arteriovenous connection with high-velocity flow. (F) Color Doppler showing pseudo venous waveforms at UV insertion into IVC. (G) Color Doppler showing venous flow at the other end of the shunt. (H) Color Doppler showing pseudo ductus venosus waveforms at UV insertion into IVC. (I) Biometry parameters showing early onset fetal growth restriction pattern. (J) Middle cerebral artery Doppler showing low-resistance flow with cerebro placental ratio (CPR) <1. AVM, arteriovenous malformation; DA, descending aorta; DV, ductus venosus; PSV, peak systolic volume; PV, portal vein; UA, umbilical artery.

Video 2

Aorta portal umbilical vein shunt. Online content including video sequences viewable at: <https://www.thieme-connect.com/products/ejournals/html/10.1055/s-0043-57019>.

Video 3

Aorta portal umbilical vein shunt. Online content including video sequences viewable at: <https://www.thieme-connect.com/products/ejournals/html/10.1055/s-0043-57019>.

Video 4

Aorta portal umbilical vein shunt B mode. Online content including video sequences viewable at: <https://www.thieme-connect.com/products/ejournals/html/10.1055/s-0043-57019>.

Brief Review of Literature

Congenital hepatic arteriovenous fistulae (AVFs) occur in less than 1:100,000 live births and are linked with poor perinatal outcome.^{1,2} Prenatal diagnosis has been described, mostly secondary to nonimmune hydrops. 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.

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.” At birth, when the systemic vascular resistance rises, more blood flows to and through the AVM low-resistance shunt worsening the clinical condition.

At present, invasive in utero treatment is not available. 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.³

Conflict of Interest

None declared.

References

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