Case Report

Prenatal 3D-ultrasound diagnosis of isolated intrahepatic portal-systemic shunt with intact ductus venosus: A case report and literature review

Maxime Van Houdt, MD\textsuperscript{a,b,*}, Johannes van der Merwe, MD, PhD\textsuperscript{a,b}, Marc Gewillig, MD, PhD\textsuperscript{a}, Luc De Catte, MD, PhD\textsuperscript{a,b}

\textsuperscript{a}Department of Development and Regeneration, Cluster Woman and Child, Group Biomedical Sciences, KU Leuven University of Leuven, Belgium
\textsuperscript{b}Department of Obstetrics and Gynaecology, Division Woman and Child, University Hospitals Leuven, Leuven, Belgium
\textsuperscript{c}Department of Paediatric Cardiology, University Hospitals Leuven, Leuven, Belgium

Abstract

We present a case of isolated intrahepatic portosystemic shunts with an aorto-hepatic-umbilical connection; prenatally diagnosed with 3D Doppler flow in a 27-year-old patient at 27 weeks of gestation. Fetal karyotyping was normal, detailed evaluation of the venous and cardiovascular system showed a patent ductus venosus and no other abnormalities. These fetuses with isolated intrahepatic shunts are at risk for intra-uterine growth restriction (27%), all cases had a good prognosis with live births. Postnatal follow-up to control shunt closure is vital as persistent shunts may lead to subsequent complications.

Introduction

Congenital portosystemic venous shunts (CPSS) have been described in numerous cases, most commonly diagnosed in childhood or adult life. High resolution sonography and 3D Doppler flow applications have enabled the prenatal detection of liver arterio-venous malformations involving the umbilical-portal venous system (UPVS). The UPVS is unique in the sense that it is derived from two different embryological precursors namely the portal and/or vitelline and umbilical veins.

Prenatally diagnosed isolated intrahepatic portosystemic venous shunts (IPSS) have been widely scattered among different case series with diverse presentations and variable outcomes. Most commonly they have been diagnosed in late pregnancy and involving the left portal vein and the left hepatic vein. We present a case and reviewed the literature with respect to potential applications of 3D ultrasound for prenatal diagnosis.

Keywords:
Intrahepatic portosystemic shunt
Prenatal ultrasound
Umbilical-Portal Venous System

* Competing Interests: The author declares that there is no conflict of interest that could be perceived as prejudicing the impartiality of the research reported.
* Corresponding author.

E-mail address: maxime.vanhoudt@student.kuleuven.be (M. Van Houdt).

\textcopyright 2021 The Authors. Published by Elsevier Inc. on behalf of University of Washington. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/)
gards to prenatally detected isolated IPSS cases and their outcomes.

Case report

A 27-year-old primigravida was referred to our centre for a possible umbilical varicose vein dilatation (UVD) at 26 weeks and 6 days. She was in good health and had an uncomplicated pregnancy with a low risk NIFTI test. The UVD of 6.9 mm was confirmed and on UPVS review multiple intrahepatic portal-systemic arterio-venous malformations were noted. Shunts were present between the portal- hepatic system incorporating the right portal vein and right hepatic vein; left inferior portal and middle hepatic vein and lastly the left inferior portal vein and left hepatic vein. The portal system, inferior vena cava and ductus venosus (DV) were present and drained separately and directly into the right atrium. Upon 3D Doppler reconstruction another shunt between the hepatic artery and the umbilical vein was also noted (Fig. 1).

The mesenteric and caval system appeared normal and no other abdominal anomalies were noted. In addition, there was mild cardiomegaly with a cardiothoracic ratio of 0.68, normal cardio-pulmonary vascular connections, grade 1 tricuspid regurgitation, and absence of any hydrops features. Just distal to the fetal umbilical cord insertion an UVD of 10 mm (normal average 6,0 mm, SD 1,0) [1] was noticed without turbulent flow (Fig. 2).

During weekly follow up visits, the fetus remained hemodynamically stable and no further anomalies were noted. At 29 weeks the dilatation of the UVD increased to 17.7 mm (normal average at 29 weeks, 6.6 mm SD 0.9 mm) [1] with presence of a small thrombosis of 5 x 6 mm (Fig. 2). Flow in the umbilical artery and vein remained laminar and non-obstructed.

After multidisciplinary discussion the patient was admitted for daily CTG and Doppler monitoring and administration of antenatal corticosteroids. All features subsequently remained stable, and she was induced at 37 weeks. An uneventful delivery resulted in a girl of 2800 g being born who only needed short term CPAP support (<24 hours). Postnatal conventional cytogenetic analysis revealed a normal female karyotype.

Postnatal ultrasound and MRI angiography confirmed the prenatal findings noting multiple vascular malformations involving liver segments II (4 mm and 3 x 7 mm), IV (6 mm), VII (5,5 mm), VIII (6 mm). The hepatic veins and portal veins were of normal caliber with triphasic- and uniphase flow respectively. There was no arterial hypertrophy nor hyperdynamic arterial flow in the liver (PSV 45 cm/s). Echocardiography noted a normal structured and functional heart with a mild left-right shunt and a patent foramen ovale, a persistent open arterial duct closed by day 21. Pathology of the umbilical cord confirmed the presence of a small thrombus.

The mother and child were discharged in good condition after 10 days. The hyperammonemia (maximal 138 mmol/l) partially resolved spontaneously by 3 weeks. But during her weekly follow ups the shunt persisted with mildly elevated hyperammonemia. At no point was any cardio- pulmonary or hemodynamic dysfunction noted. After 6 months the most

Fig. 1 – Twenty-seven-year-old primigravida at 27 weeks of gestation. (A) 3D multiplanar reconstruction & (B) 2D cross sectional sagittal colour doppler ultrasound evaluation of the portosystemic shunts (*). UV, umbilical vein; DV, ductus venosus; SMA, superior mesenteric artery; HA, hepatic artery; SDV, subdiaphragmatic vestibulum; RA, right atrium; Ao, aorta; PV, portal vein.
prominent shunt in segment IV was coiled percutaneously (Fig. 3).

**Discussion**

CPSS are rare vascular anomalies connecting portal branches directly to systemic veins, bypassing the liver. The CPSS have been classified depending on the presence of intra and/or extrahepatic features and the presence or absence of the DV with most literature coming from the pediatric surgery field [2,3]. Achoron et al., presented a prenatal classification system with three subtypes: Type I, umbilical-systemic shunt; Type II, DV-systemic shunt; Type III, portal-systemic shunt [4]. Therefore, our case fits the description of a type IIIa, with an intrahepatic shunt between the portal venous system and the hepatic veins.

Review of the literature identified 22 prenatally detected cases of isolated IPSS with intact DV and normal karyotype, as summarized in Table 1. Median gestational age at diagnosis was 31 weeks (range 26-34 weeks). All resulted in a live birth but 27% (6/22) was associated with intrauterine growth restriction (IUGR). Delle Chiaie et al., suggested that IUGR without hypoxia can be explained in these cases due to the impaired umbilical blood perfusion of the liver parenchyma [5]. In view of this finding, not only should IPSS be actively excluded in cases of unexplained IUGR, but inversely all CPSS cases should be monitored for altered fetal growth.

Since the majority of CPSS cases have been described in the context of multiple major abnormalities and syndromes, especially in the context of extrahepatic shunts [6], common associated cardiovascular anomalies must be excluded. In five cases of intrahepatic portosystemic shunts an additional aorto-portal-systemic connection was described. Two in context of chromosomal abnormalities, being trisomy 21 [7] and trisomy 17 [8]. While in those with a normal chromosomal profile, one had a left sided inferior vena cava [9], and two cases had an intrahepatic porto-systemic shunt with a coincidental shunt between the hepatic artery and de vena porta [10,11].

In 23% (5/22) there was no data reported on postnatal examinations to confirm the shunt. And in one case the shunt was not observed on postnatal ultrasound. In our case, the aorto-hepatic-umbilical connection was not confirmed post-
Table 1 – Review of prenatally diagnosed isolated intrahepatic portal-systemic shunt cases with DV present and normal karyotype.

| Case | Maternal Age (years) | GA (weeks) at diagnosis | Shunt origin and drainage | DV | Prenatal additional anomalies | Pregnancy outcome | Postnatal confirmation | Shunt outcome or resolution |
|------|----------------------|-------------------------|--------------------------|----|-------------------------------|-------------------|-----------------------|---------------------------|
| 1    | Achiron et al. [4]   | 32.2 ±5.3               | 21                       | LPVm–RHV, LPV–MHV, Absent RPV | Present | SUA, SVC to coronary sinus, Right clubfoot | Live birth          | Confirmed              | Spontaneous closure       |
| 2    |                     | 28                      | 28                       | LPVs–LHV, Absent RPV          | Present | None                          | Live birth          | Confirmed              | Spontaneous closure       |
| 3    |                     | 27                      | 35                       | LPVm–MHV                        | Present | None                          | Live birth          | Confirmed              | Spontaneous closure       |
| 4    |                     | 35                      | 35                       | LPV–LHV                         | Present | None                          | Live birth          | Confirmed              | Spontaneous closure       |
| 5    |                     | 36                      | 35                       | LPV–LHV                         | Present | None                          | Live birth          | Confirmed              | Spontaneous closure       |
| 6    |                     | 35                      | 35                       | LPVI–MHV                        | Present | None                          | Live birth          | Confirmed              | Spontaneous closure       |
| 7    |                     | 31                      | 35                       | LPV–LHV                         | Present | None                          | Live birth          | Confirmed              | Spontaneous closure       |
| 8    |                     | 33                      | 28                       | LPV–HV cavernous                | Present | Early IUGR, Dilated echogenic Bowel. | Live birth          | Confirmed              | Spontaneous closure       |
| 9    |                     | 26                      | 34                       | PV–HV cavernous                 | Present | Early IUGR, Hepatic calcifications, Right Hydronephrosis. | Live birth          | Confirmed              | Spontaneous closure       |
| 10   | Delle et al. [5]     | 32                      | 23                       | PV–HV aneurysm                  | Present | IUGR, oligohydramnios          | Live birth, 35 weeks | Confirmed (US)         | Spontaneous closure at 3 m coiling after 1 month hyperammonaemia |
| 11   | Han et al. [10]      | 36                      | 36                       | LPV–LHV shunt HA-PV             | Present | SGA                           | Live birth, 39,3 weeks | Confirmed (US)         | Spontaneous closure 2 m |
| 12   |                     | 28                      | 33                       | LPV-LHV                         | Present | IUGR, Prominent umbilical vein | Live birth, 30 weeks | Confirmed (US)         | Spontaneous closure 28d |
| 13   | Jerabek-klestil et al. [13] | 27                      | 34                       | LPV–LHV                         | Present | Early IUGR, DCDA twin          | Live birth, 36w5d    | Confirmed (US)         | Spontaneous closure 6 m |
| 14   |                     | 20                      | 33                       | LPV–RHP/LHV/MHV                 | Present | IUGR in affected fetus        | Live birth, PPROM    | Confirmed (US)         | Spontaneous closure 6 m |
| 15   | Achiron et al. [14]  | 31                      | 22                       | RPV–HV                          | Present | None                          | Live birth          | Confirmed (US)         | Spontaneous closure 6 m |
| 16   | Wu et al. [11]       | 27.8 (20-40)            | 31                       | PV–LHV                          | Present | Cardiomegaly                  | Live birth          | Not reported            | Unknown                   |
| 17   |                     | 27.8 (20-40)            | 25                       | PV–HV With HA-PV                | Present | Cardiomegaly                  | Live birth          | Not reported            | Unknown                   |

(continued on next page)
Table 1 (continued)

| Case | Maternal age (years) | GA (weeks) at Shunt origin | Pregnancy outcome | Shunt additional anomalies | DV | Prenatal additional anomalies | Postnatal confirmation | Shunt outcome or drainage | Shunt resolution or additional diagnosis | Shunt outcome or drainage |
|------|----------------------|---------------------------|-------------------|---------------------------|-----|-------------------------------|-----------------------|-------------------------|------------------------------------------|-------------------------|
| Wang et al [15] | 18 | 43 | 31 | Present | None | None | Not confirmed (US) | Live birth | No data reported | Not confirmed | Unknown |
| | 19 | 26 | 26 | Present | None | None | Confirmed (US) | Live birth | No data reported | Persistent after 7 m | Unknown |
| | 20 | 39 | 26 | Present | None | None | Unknown | Live birth | No data reported | No postnatal examinations | Unknown |
| | 21 | 31 | 32 | Present | None | None | Unknown | Live birth | No data reported | Unknown | Unknown |

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:10.1016/j.radcr.2021.02.037.

References

[1] Weissman, et al. Sonographic measurements of the umbilical cord and vessels during normal pregnancies. J Ultrasound Med 1994;13(11–14).
[2] Park JH, Cha SH, Han JK, Han MC. Intrahepatic portosystemic venous shunt. AJR Am J Roentgenol 1990;155:527–8.
[3] Lautz TB, Tantemsapya N, Rowell E, Superina RA. Management and classification of type II congenital portosystemic shunts. J Pediatr Surg 2011;46:308–14.
[4] Achiron R, Kivilevitch Z. Fetal umbilical-portal-systemic venous shunt: in-utero classification and clinical significance. Ultrasound Obstet Gynecol 2016;47:739–47.
[5] Delle Chiaie L, Neuberger P, Von Kalle T. Congenital intrahepatic portosystemic shunt: prenatal diagnosis and possible influence on fetal growth. Ultrasound Obstet Gynecol 2008;32:223–5.
[6] Papamichael M, Pizanias M, Heaton N. Congenital portosystemic venous shunt. Eur J Pediatr 2018;177:285–94.
[7] Hartung J, Chaoui R, Kalache K, Tenstedt C, POLLMAN R. Prenatal diagnosis of intrahepatic communications of the umbilical vein with atypical arteries (A-V fistulae) in two cases of trisomy 21 using color Doppler ultrasound. Ultrasound Obstet Gynecol 2000;16:271–4.
[8] Iliescu DG, Ruican D, Nagy R, Burada F. Aorta-umbilical vein fistulae in fetus with trisomy-17 mosaicism. Ultrasound Obstet Gynecol 2020;55:419–21.
[9] Zhou J, Zhou Q, Peng Q, Zeng S. Prenatal diagnosis of Aorta-Porto-Umbilical vein fistulas with left-sided inferior vena cava. AJR Rep 2016;6:e59–61.
[10] Han BH, Park SB, Song MJ, Lee KS, Lee YH, Ko SY, et al. Congenital portosystemic shunts: prenatal
manifestations with postnatal confirmation and follow-up. J Ultrasound Med 2013;32:45–52.

[11] Wu Y, Zhou L, Chen L. Correlations among congenital hepatic shunt, absent ductus venosus, and umbilical vein shunt revealed by prenatal ultrasound. Fetal Diagn Ther 2020;47:237–44.

[12] Sokollik C, Bandsma RH, Gana JC, van den Heuvel M, Ling SC. Congenital portosystemic shunt: characterization of a multisystem disease. J Pediatr Gastroenterol Nutr 2013;56:675–81.

[13] Jerabek-Klestil S, Brantner C, Nehoda R, D’Costa E, Campei S, Scheier M. Prenatal sonographic diagnosis of intrahepatic portosystemic shunts. J Ultrasound Med 2014;33:543–6.

[14] Achiron R, Gindes L, Kivilevitch Z, Kuint J, Kidron D, Boyanover Y, et al. Prenatal diagnosis of congenital agenesis of the fetal portal venous system. Ultrasound Obstet Gynecol 2009;34:643–52.

[15] Wang Y, Yan Y, Yang Z, Wei J, Liu G, Pei Q. Prenatal diagnosis of congenital portosystemic shunt: a single-center study. J Obstet Gynaecol Res 2020;46:1988–93.