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Two Rare Variants of Partial Portal System Agenesis Involved Fetal Portal Sinus Without Porto-Systemic Shunt

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ABSTRACT

Background & Objective: Congenital partial agenesis of the portal venous system is a very rare anomaly in which part of the portal blood bypass the liver, and is usually associated with portosystemic shunt (PSS). The prognosis is good and they usually have excellent hemodynamic status with normal fetal growth and a survival rate.

Case report: In this study we report two rare cases of fetal portal system anomalies at second trimester which mainly involved portal sinus. They are the first cases that have ever been reported without any systemic shunt. Both fetuses were born normally and have had normal outcomes ever since.

Conclusion: Partial portal system agenesis with only minor changes in portal system anatomy and without PSS can be considered as variants of portal system with good outcome.

Keywords: Fetus, Portal agenesis, Portal sinus, Ductus venosus

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Introduction

The fetal umbilical–portal venous system has a complex anatomy **(1,2)**. The normal anatomy of the system can be seen in Figure 1a. Left portal vein (PV) has two branches: umbilical segment and pars transversa (portal sinus), which connects to main PV.

Congenital agenesis of the portal venous system (CAPVS) is a very rare anomaly, in which the portal blood bypass the liver and a portosystemic shunt (PSS) between the spleno-mesenteric system and the systemic circulation is made. In 1994, Morgan *et al.* attempted to classify the several types of CAPVS **(5)** into two categories: type I which is a total agenesis with a PSS, and type II (partial agenesis) in which a portion of the portal blood is delivered into the systemic venous circulation (porto-hepatic shunt). The anomaly is sometimes accompanied by ductus venosus (DV) agenesis **(1-4)**.

Recently, a few studies have been conducted showing a great amount of interest in assessing portal system abnormalities. In this study, we report two rare cases of fetal portal system anomalies which mainly involved portal sinus without any systemic shunt.

Case Report

The first case was detected at 22 weeks of gestation in a 25-year-old healthy, para 1, and gravid 1 woman with no remarkable family history. Fetal ultrasound revealed a congenital absence of fetal portal sinus, in which the medial branch of the left PV was elongated and connected to the connection between main PV and the right PV branches in a trifurcated pattern based on the connection pattern proposed by Kivilevich *et al.* **(5)**. The DV was present with normal course (Figure 1b,c,d).

The second case was detected at 23 weeks of gestation in a 31-year-old, para 2, and gravid 2 woman with no remarkable past history. In this case, the medial branch of the left PV was dilated and connected to the anterior branch of the right PV via an atypical portal sinus located more posteriorly rather than midline.

Again, the DV was normal and no PSS was identified (Figure 2).

No associated anomaly, growth restriction, or evidence of hydrops fetalis were found during pregnancy. Both fetuses were born normally via vaginal delivery and have had normal outcomes ever since. The first infant is a 6-month-old baby and the second one is 8 months old now. None of the babies have had any complications at birth or afterwards, and they have normal development.

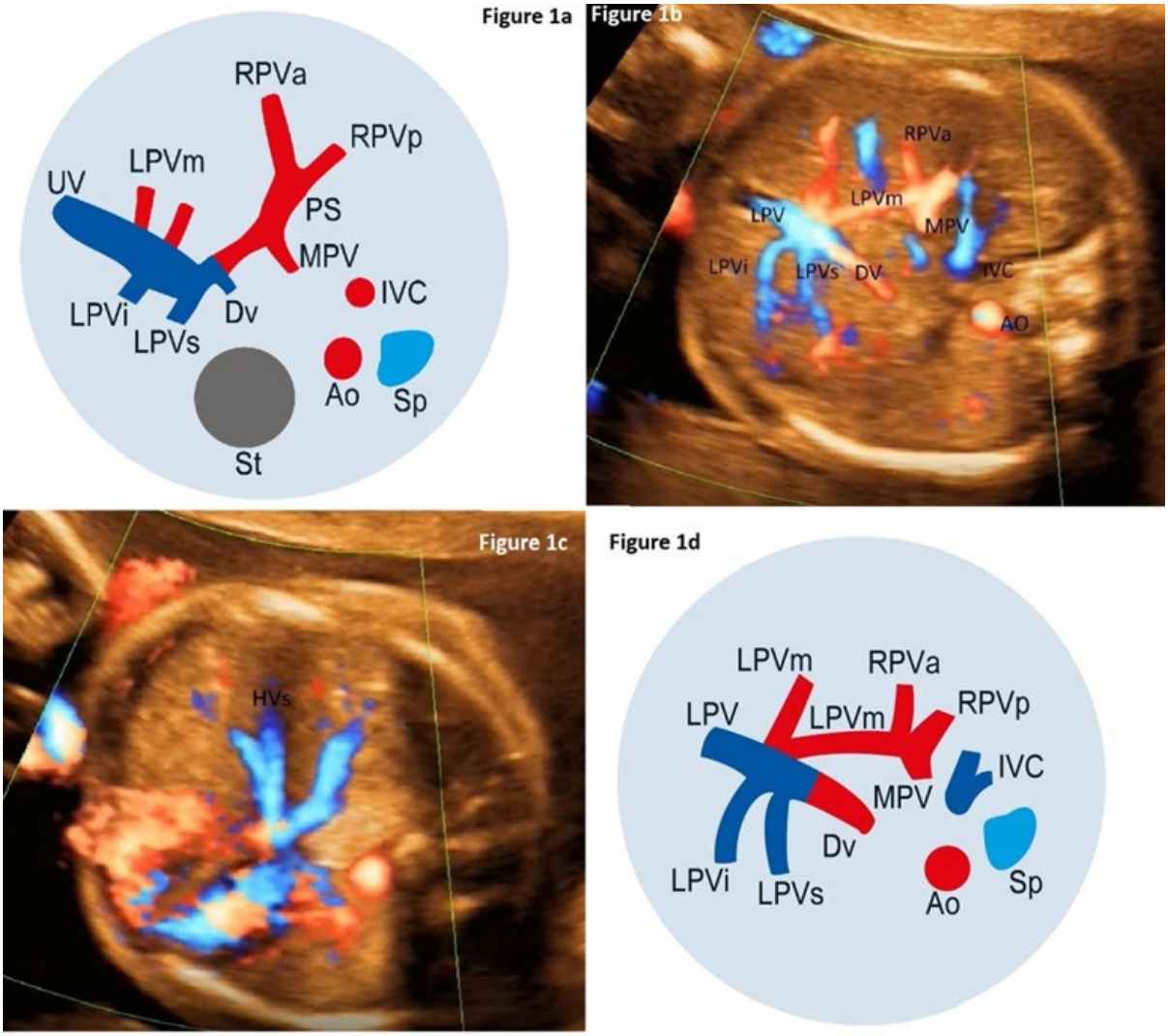
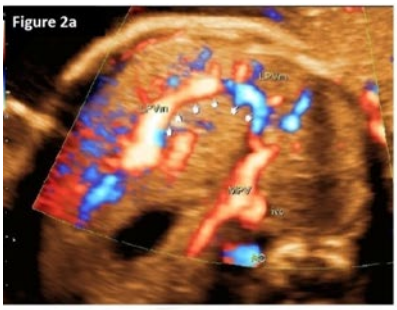
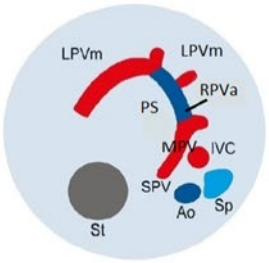
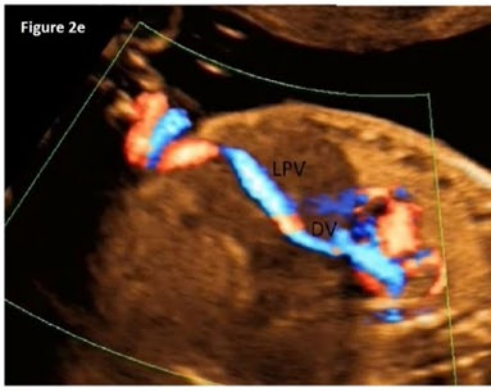


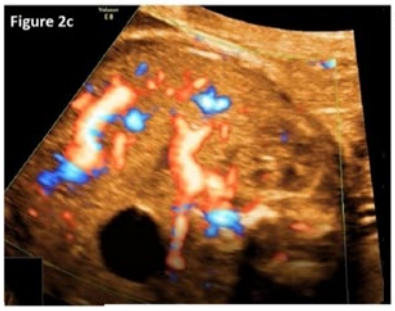
Figure 1a: The schematic view of the normal portal system anatomy. The normal portal system anatomy can be seen in which the umbilical vein (UV) transports oxygen rich blood from the placenta and as it passes in a upward direction into the liver, where it is joint by the left portal vein (LPV), thus, perfusing the left lobe of the liver and giving rise to the Ductus Venosus (DV), which then by-passes the liver and shunts blood into the heart of the fetus, directly. At the point of the median fissure of the liver it connects to the right portal vein (RPV). The main portal vein (MPV) then joins the RPV which transports de-oxygenated blood from the spleen and fetal gut, therefore contributing to the blood supply of the right liver lobe. Figure 1b: In ultrasound view, fetal portal sinus is absent, in which the medial branch of the left portal vein was elongated and connected to the connection between main PV and the right PV branches in a trifurcated pattern. DV is located in normal position; Figure 1c: Normal hepatic veins. Figure 1d: The same findings of figure 1b are shown by the schematic view.

UV: Umbilical vein, PV: Portal vein, LPVm: Left portal vein medial branch, LPVi: Left Portal vein inferior branch, LPVs: Left Portal vein superior branch, DV: Ductus Venosus, PS: Portal sinus, RPVa: Right portal vein anterior branch, RPVp: Right portal vein posterior branch, MPV: Main portal vein, IVC: Inferior Vena Cava, Ao: Aorta, St: Stomach, Sp: Spine.









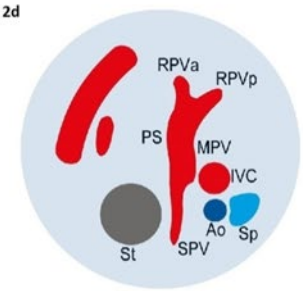


Figure 2a and b, In ultrasound and schematic views respectively, an atypical location of fetal portal sinus is noted in which the medial branch of LPV connects to the anterior branch of the RPV directly. Figure 2c and d: The atypical portal sinus is located more posterior and lateral rather than midline. This figure is obtained slightly lower at the level of the RPV branches. Figure 2e: In an axial oblique view, there is a normal DV. SPV: Splenic vein, RPV: Right portal vein, LPVm: Left portal vein medial branch.

Discussion

CAPVS is a rare anomaly and less than 100 cases have been reported so far worldwide **(3,6)**. These anomalies are often reported and discussed in the literature in association with DV agenesis; but they can exist with normal DV structure **(6)**. All the partial portal agenesis cases reported in the literature had PSS, which was not present in our cases. The presence of any shunt is caused by disruption in the normal anatomy of portal system or DV. Cardiac decompensation develops more commonly in extrahepatic shunts. Achiron *et al.* (2016) classified the umbilical–portal–systemic venous shunt in fetuses into 4 groups; among PSS groups, only partial portal agenesis was found in 25% of intrahepatic type and both partial and complete portal agenesis were discovered in most cases of extrahepatic types **(6)**.

Similar to our results, previous studies reported that fetuses with partial portal system agenesis have excellent hemodynamic status with normal fetal growth and a survival rate of 100% **(1)**. As far as the researchers investigated, both cases reported in this study are the first variants of partial portal agenesis that have ever been reported without PSS and with normal DV, and regarding only minor changes in portal system anatomy without shunt can be considered as variants of portal system.

Conclusion

Partial portal system agenesis with only minor changes in portal system anatomy and without systemic shunt can be considered as variants of portal system with good outcome.

Acknowledgments

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Conflict of Interest

The authors declare no conflicts of interest

References

1. Achiron R, Gindes L, Kivilevitch Z, et al. Prenatal diagnosis of congenital agenesis of the fetal portal venous system. Ultrasound Obstet Gynecol. 2009;34(6):643-652. [DOI:10.1002/uog.7460] [PMID]
2. Wu H, Tao G, Cong X, et al. Prenatal sonographic characteristics and postnatal outcomes of umbilical-portal-systemic venous shunts under the new in-utero classification: A retrospective study. Medicine (Baltimore). 2019;98(2):e14125. [DOI:10.1097/MD.0000000000014125] [PMID] [PMCID]
3. Yagel S, Kivilevitch Z, Cohen SM, et al. The fetal venous system, Part II: ultrasound evaluation of

the fetus with congenital venous system malformation or developing circulatory compromise. Ultrasound Obstet Gynecol. 2010;36(1):93-111. [DOI:10.1002/uog.7622] [PMID]

4. Morgan G, Superina R. Congenital absence of the portal vein: Two cases and a proposed classification system for portosystemic vascular anomalies. J Pediatr Surg 1994; 29: 1239- 1241. [DOI:10.1016/0022-3468(94)90812-5]

5. Z. Kivilevitch L. Gindes H. Deutsch R. Achiron. In-utero evaluation of the fetal umbilical-portal venous system: two- and three-dimensional ultrasonic study. Ultrasound Obstet Gynecol. 2009 Dec;34(6):634-42 [DOI:10.1002/uog.7459] [PMID]

6. R Achiron, Z Kivilevitch. Fetal umbilical-portal-systemic venous shunt: in-utero classification and clinical significance. Ultrasound Obstet Gynecol 2016 Jun;47(6):739-47. [DOI:10.1002/uog.14906] [PMID]

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