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Incidental Recognition of Umbilical-Portal-Systemic Venous Shunt Diagnosed During BPP for Decreased Fetal Movement

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Case Report Title: Incidental recognition of Umbilical-Portal-Systemic Venous Shunt diagnosed during BPP for decreased fetal movement.

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Background: The prevalence of prenatally diagnosed umbilical-portal systemic venous shunts (UPSVS) is reportedly rare, ranging from 0.003-0.209% [1]. Disruption in the normal development of the fetal venous system can be caused by primary failure of a system or by secondary occlusion of an already transformed system [2]. While UPSVS's are rare, the downstream effects are potentially fatal, including intrauterine growth restriction, high output cardiac failure, associated neonatal morbidity and mortality, and thus necessitate early detection with appropriate monitoring.

Introduction: The patient was a 21-year-old G2P1001 who presented to the Maternal Fetal Medicine Ultrasound Genetics (MFMUG) clinic at 36w2d for a scheduled Biophysical Profile (BPP) as a follow up after a non-reactive NST in the office. During the encounter, the sonographer noted hypervascularity within the liver, abdominal circumference <10%, and a BPP of 8/8. After further evaluation, the preliminary diagnosis was a portal-systemic shunt involving the portal system and the left and middle hepatic veins, with uncertain presence or absence of a ductus venosus. It was recommended to return weekly for AFI's, twice weekly NST's, delivery at 38w for fetal growth restriction and neonatal evaluation.

Discussion: This case demonstrates recognition of an UPSVS in an otherwise uncomplicated pregnancy with previously normal anatomy ultrasound at 20 weeks' gestation. The role in ultrasonography and BPP for fetal wellbeing is a luxury to facilities with MFM specialists readily available. The patient described in this report had decreased fetal movement, a non-reactive NST in the office, and was referred to MFMUG for a formal BPP. Currently, there are approximately one MFM specialist per 14 OBGYNs, and one MFM per 2,277 births [3]. The states with the leading number of MFM specialists include California, New York, Texas, Florida, and Pennsylvania [3]. Identification of this UPSVS may not have been readily identified by the untrained eye, but growth ultrasounds and Biophysical Profiles are key skills of any obstetrician. This case report aims to raise awareness for identification of these abnormalities to help facilitate proper evaluation and management.

References

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Basic Embryology – USOB Part I (reference 4)

- The heart begins beating by d23
- 3 symmetric paired veins form the basis of the early venous system draining into the heart
    - Umbilical veins (UVs) – drain the chorion
    - Vitelline veins (VVs) – drain the yolk sac
    - Cardinal veins (CVs) – drain the body of the embryo
- All 3 pairs open to the right & left horn of the sinus venosus
- Simultaneously, the liver buds begin to form from the ventral endodermal wall of the foregut
    - These cells invade the mesenchymal tissue (septum transversum) → connective tissue of the future liver
- Between weeks 4-6 the vessels undergo complex growth, anastomosis and degeneration, resulting in
        - Right proximal VV → hepatocardiac segment of the IVC
        - Intrahepatic efferent veins → LHV, MHV, RHV
        - Distal left & right VV → portal vein
    - The left UV becomes the dominant conduit of blood from the placenta
- Ductus venosus forms in the 8th week of development: formed via coalescence of hepatic sinusoids and drains into the hepatocardiac segment of the IVC
- The IVS is formed via 4 different embryonic sources in a caudocranial order
    - Most caudal, Sacrocardinal segment via posterior CV
    - Prerenal, sacrorenal segment from right supracardinal vein
    - Suprarenal, hepatic renal segment from right subcardinal vein
    - Most cranial, hepatocardiac segment from right VV

Fetal venous anatomy - USOB Part I

- Oxygen/nutrients from the placenta to the heart via the UV and ductus venosus
- Intraabdominal segment of the UV courses via falciform ligament to MERGE w/ the left portal vein (LPV)
    - LPV gives rise to the ductus venosus (DV)
    - The DV connects to the IVC distally WITH the LHV and MHV just proximal to the RA
- Junction of the MPV and the PS is a continuum of morphologic variation in the angle of communication
    - T-shaped MC (68%)
    - X-shaped (12%)
    - H-shaped (15%)
- “The venous system represents one of 4 that comprise the fetomaternal vascular system, the others being the heart, placenta, and arterial system. Their function individually and as part of an integrated system depends on the healthy performance of each component.”
    - “a pressure gradient is created between the atria and ventricles which reduces the preload in the venous circulatory system and allows blood to flow toward the

heart. This pressure gradient is further accentuated by the physiologic stenosis of the DV."
    - "Intrathoracic pressure variations caused by fetal breathing movements have been shown to affect both venous return to the heart and the arterial system."
    - "The placenta therefore is a system capable of transmitting pressure changes occurring within the heart and thorax."
    - "The fetal liver is thus divided into two physiologically different lobes, the left, supplied by blood rich in oxygen and nutrients, and the right, which receives a mixed supply of blood."

Congenital venous system malformation (Part 2)

- Disruption in the normal development of the fetal venous system can either be due to "(1) primary failure of a system or part of a system to form or to create critical anastomoses, or (2) by secondary occlusion of an already transformed system"
- "Cardiac output progressively increases and resistance of the placental vascular bed decreases while the umbilicocaval pressure gradient is maintained steady during the third trimester of pregnancy. Any situation hampering this hemodynamic balance initiates compensatory mechanismsin thee arterial and venous systems."
    - The changes that are made to compensate for these situations are "gradual and sequential, in correlation with the severity of the hypoxic insult…"
- Uteroplacental insufficiency initiates compensatory mechanisms in fetal circulation
    - The right ventricle/placenta has high resistance
    - Left ventricle/brain has low resistance
    - Reduced blood supply from placenta to liver, increased proportion of cardiac output diverted to fetal body/IVC, increased blood flow through SVC and increased afterload to right ventricle
        - "This leads to increased preload that hampers the supply of high quality blood flow from the placenta"
    - ***Kiserud et al (90) "found that the degree of shunting in IUGR fetuses is positively correlated with the severity of placental insufficiency as reflected by the UA diastolic flow"
        - "preferential flow through the DV results from reciprocal hemodynamic changes inside the liver."
        - "Reduced liver perfusion may provide the first clinical evidence of placental insufficiency, restricted fetal growth, and decreased fetal weight."
- Doppler waveform abnormalities are sensitive tools for assessing fetal wellbeing before 32w EGA
    - <u>Umbilical vein</u>: pulsatile flow in the UV is a simple and reliable marker of circulatory compromise (marker for asphyxia)
    - <u>Ductus Venosus</u>: normally at 20w 30% of blood is shunted through DV and at 30w 20%
        - for SGA fetuses have higher shunting and can be exacerbated by earlier placental compromise

        - reversed or 0 flow in the A-wave is a sign of disordered cardiac function (decreased in IUGR)
    - Hepatic/Portal Veins: left portal vein is the watershed of the fetal venous circulation – simple marker of circulatory compromise.
        - Normal distribution of blood flow btw left and right lobes is 60%/40%
        - Blood flow in the UV is directed to the left liver lobe and the DV with only a minority of flow diverted to the LPV and right liver lobe

FUPSVS 2016 (Reference 5)

- Types of shunts
    - Type I: umbilical-systemic shunt – USS (20.4%)
        - The UV failed to form the normal intrahepatic connection with the left PV-DV due to agenesis of both the left branch of the PV and DV, and connected to the systemic circulation
        - USS earliest mean gestational age at diagnosis (16w +/- 7.2w), highest incidence of absence of normal IHPVS and highest incidence of associated major anomalies, lowest rate of live births
    - Type II: ductus venosus-systemic shunt – DVSS (43.2%)
        - The ductus venosus blood flow was shunted from its normal path towards the left heart into systemic veins
        - DVSS normal IHPVS, the ONLY group in which aneuploidy was detected
            - Trisomy 21 – all four terminated
            - Of the remaining, 73.7% cases of isolated DVSS survived with normal outcomes
            - Male gender 93%
    - Type III: portal-systemic shunt – PSS (intra-hepatic 27.2%; extrahepatic portal-systemic shunt 9.1%)
        - IHPSS highest incidence of IUGR and lowest mean birth-weight centile
        - Main prognostic factor of EHPSS group was complete absence of the IHPVS – no effect on fetal growth pattern or association with major anomalies were observed (2 of 2 cases reported)
        - One case presented with signs of heart volume overloading (cardiomegaly with tricuspid regurgitation)
        - Male gender 75% EHPSS
- "Most favorable prognostic factors were absence of associated major malformation and presence of IHPVS - thus intrahepatic portal-systemic shunt had the best outcome"
- "Mapping of the IHPVS is paramount for optimizing prenatal counseling and postnatal care"
- "Only within the last 10 years or so that larger series have emerged in the literature and several classifications have been suggested…"
- Overall postnatal live-birth rate was 70.5%
    - Shunt not seen in DVSS cases after birth
- "The DVSS and the IHPSS, when isolated, had the best prognosis, due to the presence of the IHPVS with spontaneous closure of the shunt after birth in all cases."

- “USS and the EHPSS were characterized by an absence of or abnormal IHPVS and had worse prognosis.”
- Incidence of UPSVS is unknown, and identification is likely underreported due to low “level of awareness.”