Journal of Gynecology Obstetrics and Human Reproduction xxx (2019) xxx–xxx

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Case Report

Fetal umbilical-systemic shunt with a positive issue

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ARTICLE INFO

Article history:
Received 17 June 2019
Received in revised form 8 October 2019
Accepted 17 October 2019
Available online xxx

Keywords:
Fetal umbilical-portal-systemic venous shunts
Agenesis ductus venosus
Postnatal outcome

ABSTRACT

We herein report the case of abnormal umbilical-venous return in which the antenatal ultrasound enabled us to establish the diagnosis of *umbilical-systemic shunt* (Type 1 according to Achiron (Achiron and Kivilevitch, 2016)). Due to the concomitant associations of cardiomegaly, intrauterine growth retardation, oligohydramnios, and left-lobe hypoplasia with agenesis of the *intrahepatic umbilical vein – left portal vein – ductus venosus,* a poor prognosis (11.1% survival) was to be expected. In spite of development of pulmonary arterial hypertension at birth, which was promptly treated, the evolution was nevertheless good, both on clinical and ultrasound follow-up.

Introduction

Umbilical venous return abnormalities are rare [2,3]. The incidence of such abnormalities, particularly those involving *fetal umbilical-portal-systemic venous shunts*, is as yet undocumented [1,4].

In the last few years, high-definition ultrasound devices have been developed that, along with new color Doppler techniques and the growing expertise of technicians, facilitate the non-invasive diagnosis of fetal umbilical venous return abnormalities [4–9].

In this article, we present the case of a portal-systemic shunt. Next, we will review and discuss the published literature on such cases.

Case summary

The patient was 33 years old and in her first pregnancy. An abnormality in the fetus' umbilical vein course was detected upon ultrasound imaging in the second trimester of pregnancy, performed at 22 weeks and 6 days of gestation.

The patient had a medical history of prolactinoma treated using quinagolide (dopamine agonist) until 12 weeks of amenorrhea. She was obese (BMI: 40.5), a non-smoker, and consumed no alcohol. The father of the fetus was healthy and neither smoked nor consumed alcohol either.

The pregnancy was normal until the second-trimester ultrasound. This ultrasound imaging revealed that the umbilical vein was running along the anterior surface of the liver without passing through it, instead directly feeding into the systemic circulation, specifically the right atrium (Fig. 1). This umbilical-systemic shunt was associated with agenesis of the ductus venosus. The pulsed doppler of the shunted umbilical vein shows a typical triphasic systemic waveform.

Though the portal vein was clearly observed to be joining the right hepatic lobe (Fig. 2), we were unable to see a normal left portal vein- ductus venosus complex. In addition, the left liver lobe appeared to be smaller (Fig. 3).

Fetal biometrics revealed intrauterine growth retardation (in the 5th percentile). The amniotic fluid index (AFI) was 8.8 cm, *i.e.,* at the lower end of the normal amniotic fluid quantity range. Doppler imaging of the mother's uterine arteries and the fetus' umbilical and cerebral arteries presented normal resistance indexes. Apart from a slightly hyperechogenic bowel, no fetal morphological abnormality were revealed. Amniocentesis was performed at 24 weeks, revealing a normal fetal molecular karyotype of 46 XY.

Throughout close ultrasound follow-up, the patient presented normal cardiac function, with just a slight cardiomegaly observed (cardiothoracic index: 0.55).

While growth continued, oligohydramnios developed at 35 weeks (AFI: 3 cm), requiring hospitalization. At 35 weeks, 3/7th, the patient elected for a caesarian section due to the severe oligohydramnios and potential risk of *in utero* death.

The newborn was male, presented an Apgar score of 1/8/8, and a weight of 2350 g (p40). He underwent neonatal resuscitation for

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http://dx.doi.org/10.1016/j.jogoh.2019.101656
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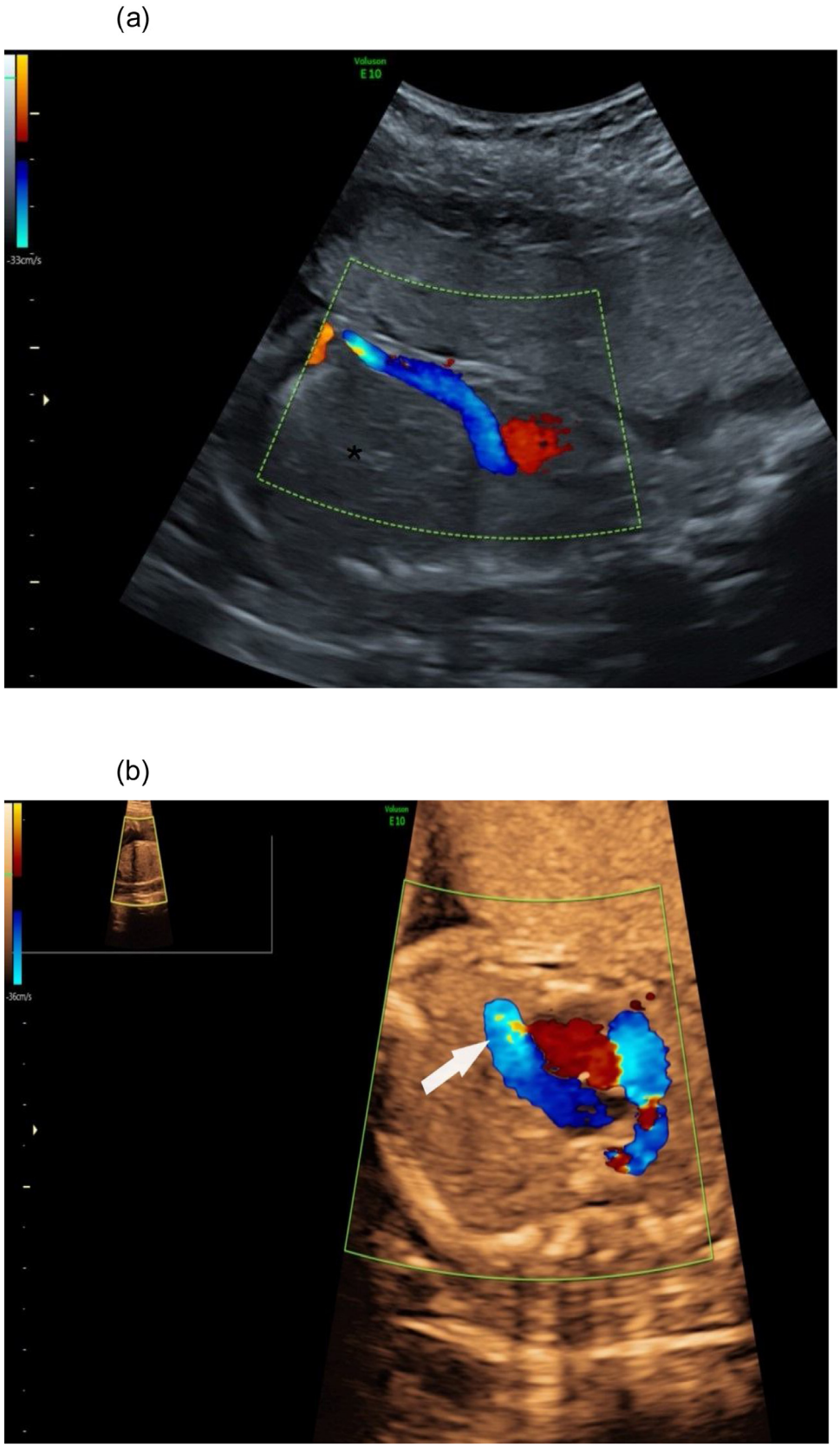

Fig. 1. Ultrasound imaging of the fetal umbilical systemic shunt (a) Sagittal view of the umbilical vein running among the anterior surface of the liver. (b) Direct connection between umbilical vein and right atrium.

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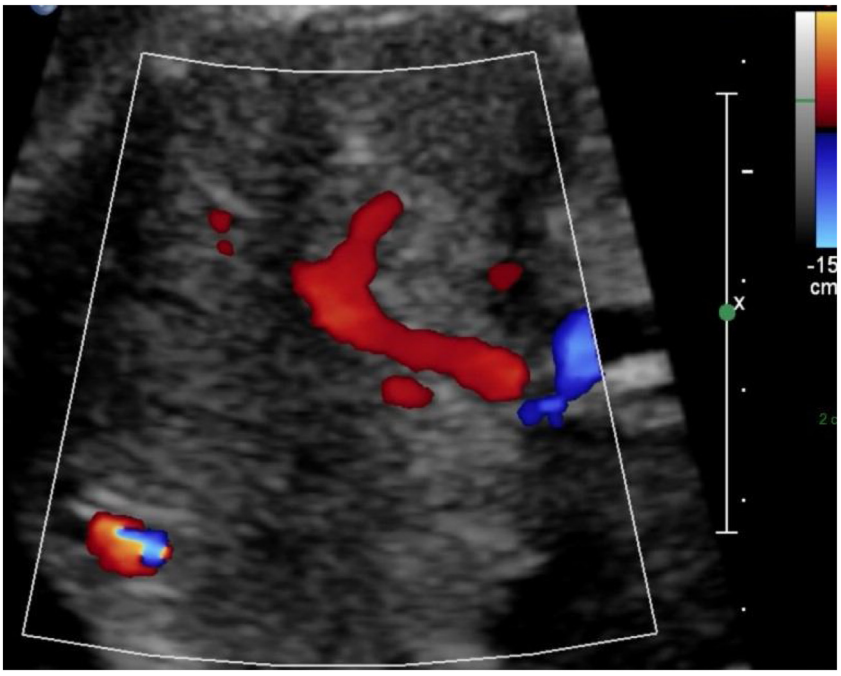

Fig. 2. Color Doppler image showing intrahepatic right portal vein.

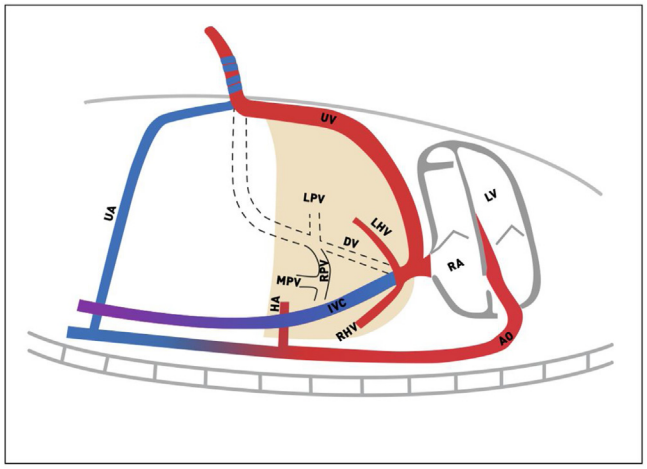

Fig. 3. Case's diagram: AO: Aorta - DV: ductus venosus - HA: hepatic artery - IVC: inferior vena cava - MPV: Main portal vein - LHV: Left hepatic vein - LPV: Left portal vein - LV: Left ventricle - RA: Right atrium - RHV: Right hepativ vein - RPV: Right portal vein - UA: Umbilical artery - UV: Extrahepatic umbilical vein.

bradycardia and fitted with CPAP in neonatal care, where he was administered surfactants for hyaline membrane disease.

Neonatal cardiac ultrasound performed on Day 1 revealed pulmonary arterial hypertension (HTAP) due to patent ductus arteriosus and foramen ovale (FO) with a bidirectional shunt primarily oriented left/right. In addition, hypoplasia of the left liver lobe and good portal perfusion of the right lobe were confirmed. An ultrasound check-up on Day 5 revealed that the FO and ductus arteriosus were now closed, along with a left-ventricular defect, which was treated using captopril (an angiotensin-converting enzyme inhibitor). The patient then showed positive evolution.

Another ultrasound on Day 8 demonstrated good left ventricular function. The extra-hepatic umbilical vein was still observed as having weak bidirectional flow. On Day 15, the extra-hepatic umbilical vein was observed as having shrunk, and the newborn was discharged.

The infant was re-admitted for pediatric cardiology check-up on Day 22. Ultrasound revealed good ventricular function and normal heart anatomy, except for a persistent small FO of 2 mm with a shunt of the left heart towards the right.

The extra-hepatic umbilical vein had disappeared. At 1 year, the infant presented normal evolution.

Discussion

The development of the venous system can be disrupted at any stage [5]. The failed occlusion of a primitive vein, development of anastomoses [9] or new vessels [5] or even a secondary occlusion of a vein [1,9], any of these malformations can cause abnormalities in the umbilical-portal circulation [4,5]. These malformations are not always accompanied by agenesis of the ductus venosus [5,8], but do typically cause the development of fetal umbilical-portal-systemic shunts [1,5].

Umbilical-portal-systemic abnormalities can be classified in many different ways, based on etiology or on whether or not there is an associated agenesis of the ductus venosus.

For our case study, we opted to refer to the classification proposed by Achiron et al. [1] based on the embryological and anatomical origins of the shunt. This method defines four types: (1) umbilical-systemic shunt; (2) ductus venosus-systemic shunt; (3a) intrahepatic portal-systemic shunt; (3b) extrahepatic portal-systemic shunt.

Thus classification is not based on whether there is agenesis of the ductus venosus or not, but on the type of shunt, which itself determines the fetus prognosis [1].

If the umbilical vein feeds directly into the right heart, a characteristic of Achiron's Type 1 abnormality, this is often accompanied by cardiomegaly due to the increased central venous pressure [4], especially when the vein joining the heart displays a diameter equal or superior to that of the umbilical vein itself [10,11]. This pressure leads to volume overload due to the loss of the regulatory mechanism typically ensured by the ductus [4,11]. The consequences can go beyond just cardiomegaly, too, as this chronic overload can lead to cardiac decompensation and hydrops fetalis [5,12], typically resulting in *in utero* death.

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Abnormalities of the venous system can be isolated [3] or accompanied by a wide range of other malformations [3,4,13]. Umbilical-systemic shunts are, in over half of all cases [1] associated with such abnormalities. These include: (1) cardiovascular defects; (2) digestive problems; (3) musculo-skeletal abnormalities; (4) genital-urinary or renal malformations [13]. Similarly, a strong association between fetal death [5,10] and hydrops fetalis [5,8,10] has been reported by several authors.

Abnormalities that affect only the umbilical, hepatic, or portal circulations typically result in minor clinical repercussions. Our case is a clear example of such Type 1 abnormality. The prognosis for this type of abnormality depends on what the defect is itself, *i.e.,* what hemodynamic issues could result from the abnormal connections created, along with any associated malformations [3,4] and the integrity of the intrahepatic portal venous system [1,10]. These prognostic factors typically point towards poor prognoses: 11.1% survival (1 case) in the series (9 cases) of umbilical-systemic shunts described by Achiron [1].

Other authors, on the other hand, have demonstrated that the prognosis can be poor even when there is no associated abnormality. This could be, in their opinion, due to a greater possibility of the infant developing heart failure [10]. In all of their cases, Shen et al. [10] described there being a poor prognosis when the shunt was of a diameter equal or superior to that of the umbilical vein, typically leading to reduced flow in the portal vein and incomplete development of the portal system. The size of a shunt could thus present an indicator of poor development of the portal system (something that is difficult to accurately assess) and poor prognosis for the fetus.

In our case, despite the hypoplasia of the left liver lobe, agenesis of the intrahepatic umbilical vein – left portal vein – ductus venosus complex, intrauterine growth retardation, and oligohydramnios, the child was still progressing well at the time of writing.

Detecting this under-diagnosed pathology in utero can thus only be achieved if the intrahepatic umbilical vein – left portal vein –ductus venosus complex can be fully visualized on standard abdominal ultrasound in transverse planes [1].

Conclusion

When an abnormality is observed in a fetus' venous system, particular attention must be paid to meticulously ruling out any associated abnormalities [9] and performing a detailed anatomical assessment of the *umbilical- portal –systemic* and ductus venosus circulation [1,8]. The Achiron classification [1] can be used to establish a prognosis, and it is also essential to analyze the karyotype, particularly if another malformation were to be diagnosed [10].

If a fetus presents with agenesis of the ductus venosus, close ultrasound follow-up must be carried out in order to check for signs of cardiac decompensation, particularly when accompanied by direct umbilical- right atrium shunt [9]. A cardiac decompensation can be observed even when the umbilical-systemic shunt is isolated.

Conversely, should an unexplained cardiomegaly be discovered in the fetus, a rigorous assessment of the ductus venosus and umbilical vein must be conducted [4]. Similarly, if the umbilical vein is found to be wide in diameter, with defective draining or presenting with biphasic flow on Doppler at the intra-abdominal umbilical vein connection, screening must be performed.

In addition, post-natal ultrasound follow-up using Doppler is necessary to detect any occlusion of intra-hepatic shunts, as, if not closed, this could lead to portal hypertension [14].

Our case (Type 1) has rarely described in the scientific literature and is of particular interest given our subject's good evolution.

Source funding

None.

Declaration of Competing Interest

The authors declare no conflict of interest.

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