Case Report

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Gynaecology & Obstetrics Case report
ISSN 2471-8165

2021
Vol.7 No.6:143

Umbilical Vein Connection to Right Atrium with Absent Ductus Venosus: A Rare Congenital Defect

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Citation: Osei FA, Brewer J (2021) Umbilical Vein Connection to Right Atrium with Absent Ductus Venosus: A Rare Congenital Defect. Gynecol Obstet Case Rep Vol.7 No.6:143

Abstract

Introduction: An abnormal course of the Umbilical Vein (UV) is a rare anomaly and this can be associated with the congenital absence of the Ductus Venosus (DV), cardiac and extra-cardiac anomalies. The major clinical importance of these anomalies is the need to recognize them during prenatal and postnatal diagnostic studies and at the time of cannulation or catheterization of the umbilical vein in sick neonates. Through targeted examination of the fetal heart and the venous system, a correct prenatal diagnosis is possible.

Case Report: A 25-year-old gravida 1 para 0 with type 1 diabetes mellitus at 21+6 weeks gestation referred for fetal echocardiogram because abnormal vessel was noted connecting the UV to the right atrium on the obstetric ultrasound by her maternal fetal medicine specialist. A diagnosis of drainage of the UV directly into the right atrium with absent DV and other minor cardia defects were made on the fetal echocardiogram. Preterm delivery occurred at 31+3 weeks gestation and had a favorable clinical course.

Conclusion: An abnormal course of the umbilical vein is a rare anomaly and this can be associated with the congenital absence of the Ductus Venosus (DV), cardiac and extra-cardiac anomalies. Through a thorough and meticulous ultrasonographic assessment, the diagnosis can be made and that will be helpful in guiding the management of the rest of the pregnancy and counsel the family what to expect.

Keywords: Umbilical vein; Ductus venosus; Right atrium

Received: May 28, 2021; **Accepted:** June 21, 2021; **Published:** June 28, 2021

Introduction

An abnormal course of the Umbilical Vein (UV) is a rare anomaly and this can be associated with the congenital absence of the Ductus Venosus (DV) [1]. DV connects the portal and embryonic venous circulation into the inferior vena cava [2]. Through targeted examination of the fetal heart and the venous system, a correct prenatal diagnosis of rare anomalies, like abnormal course of the umbilical vein and absent ductus venosus, is possible. Usually the primary finding is an abnormal course of the UV, and the secondary finding is often an absent DV. Close follow-up of fetuses with abnormal venous systems but no other associated malformations can help to diagnose early signs of fetal decompensation and also help to determine the optimal time for delivery [1]. Early diagnosis and the associated abnormalities may also help the family to have an idea of what to expect postnatally. We present a case that was diagnosed at 21+6 weeks gestation and had a relatively favorable outcome.

Case Report

A 25-year old gravida 1 para 0 with type 1 diabetes mellitus at 21+6 weeks gestation referred for fetal echocardiogram because a vessel was noted connecting the UV to the right atrium on the obstetric ultrasound by her maternal fetal medicine specialist. The fetal echocardiogram confirmed the drainage of the UV directly into the right atrium with absent DV **(Figures 1-3)** and the other main cardiac findings included cardiomegaly, levorotation of the cardiac axis **(Figure 4)**, and a very small muscular ventricular septal defect. There was periodic follow up during the rest of the pregnancy. She had issues associated with her diabetes mellitus and because she wanted to move

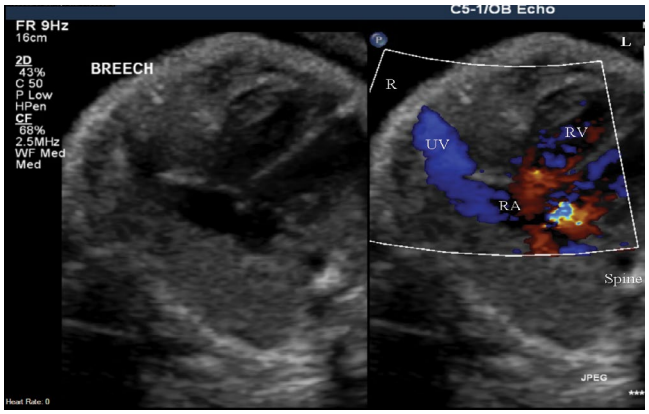

Figure 1 Cross-sectional image through the fetal thorax demonstrating the 4-chamber view.

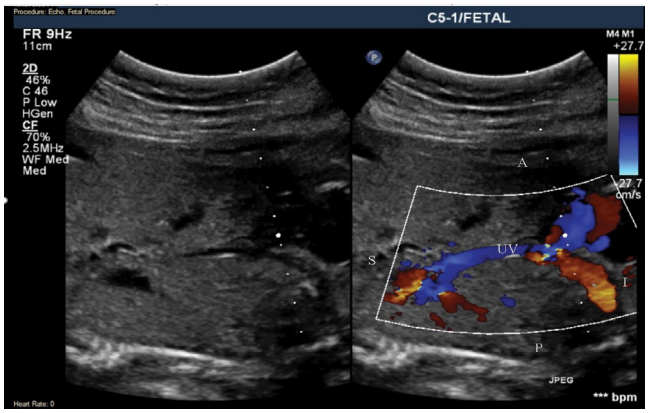

Figure 2 Sagittal view demonstrating the more distal section of Umbilical Vein (UV) which runs relatively close to the anterior wall of the abdomen with absent ductus venosus.

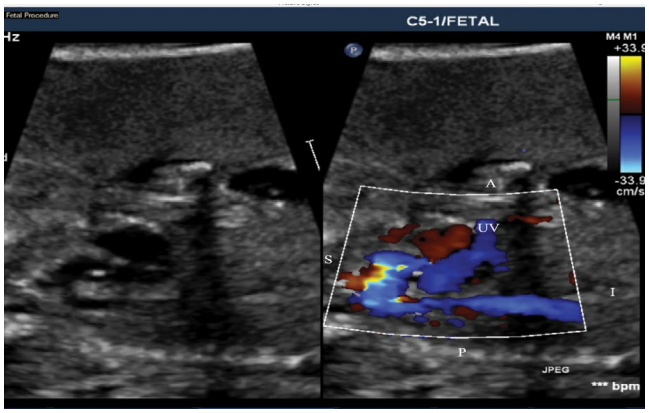

Figure 3 Sagittal view demonstrating the Umbilical Vein (UV) attaching to the right atrium.

closer to her endocrinologist, she relocated her obstetric care to our main campus in a different city. She went into prenatal labor and delivered at 31+3 weeks gestation. The main findings on the postnatal echocardiogram were a small muscular ventricular

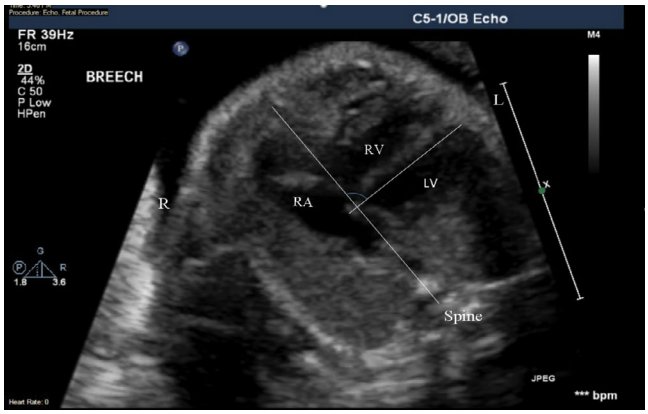

Figure 4 Transverse view image through the fetal thorax demonstrating the 4-chamber view.

septal defect and patent foramen ovale. Physical examination was unremarkable and the hospital stay was uneventful except for the repair of bilateral inguinal hernia repair, left orchiopexy, and right hydrocele drainage at 47 days old. The baby was seen in pediatric cardiology clinic at 2 months of age, he was otherwise reportedly asymptomatic from the cardiac standpoint, and his follow up echocardiogram continued to show the small muscular ventricular septal defect and patent foramen ovale.

Discussion

Embryologically the paired umbilical veins appear between 2 and 4 weeks of gestational age and after passing through the septum transversum, drain into the sinus venosus. As the liver primordium grows into the septum transversum, the umbilical veins lose their connection to the sinus venosus and develop connections to the hepatic sinusoids. Normally, the right UV regresses by 4½ weeks and only the left UV persists, which enters the hepatic sinusoids or the portal system [3]. Later, the normal in utero drainage of the UV into the portal venous system and then through the DV into the right atrium occurs. However, if the normal connection between the umbilical veins and the sinusoids is not formed, the right UV may persist. The persistent right UV may retain a number of primitive channels, including direct connection of the UV with the right atrium. Other aberrant connections include UV drainage into the suprahepatic or Inferior Vena Cava (IVC), infrahepatic IVC, iliac veins, or subcutaneous collateral vessels into the superior vena cava [4].

The explanation of the direct inflow into the right atrium is that the right UV, which normally totally disappears during the 7th week, persists. If the right UV is not really affected by the development of the liver and the septum transversum, it does not terminate in the developing liver but runs relatively close to the front wall of the abdomen in the falciform ligament, and enters the right atrium directly on the diaphragmatic surface of the liver [1].

If the anastomosis between the hepatic sinusoids and the umbilical veins is lost, a number of different aberrant drainages of the umbilical vein or umbilico-systemic connections may occur and may be associated with different malformations. The

anomalies of the umbilical vein within the fetal abdomen that may be detected with prenatal sonography may be divided into three groups: a direct inflow into the right atrium, a direct connection with the infrahepatic part of the IVC (these two bypass the portal system) and a direct connection with the vena porta system (in this latter case the whole blood mass coming through the umbilical vein has to pass the portal system) [5,6].

The ductus venosus, which develops in the 5th to 6th gestational week on the visceral surface of the liver, connects the persistent caudal part of the left umbilical vein with the IVC, and due to this anastomosis, normally about 30% of the blood arriving from the placenta can bypass the hepatic sinusoids and enter the heart directly. The well-oxygenated blood from the ductus venosus preferentially passes through the foramen ovale into the left atrium to supply the arteries leading to the brain directly. The rest of the blood from the UV passes through the left portal vein into the left lobe of the liver [7,8]. The constricting sphincter mechanism around the origin of the DV from the UV is believed to protect the fetal brain from excessive placental flow. Excessive placental blood returning to the fetal heart occurs when the DV is bypassed, as with a persistent right UV entering the inferior vena cava or right atrium [9]. This explains why there was cardiomegaly in our case (CTR=0.6).

In the absence of DV, the normal streaming of highly oxygenated umbilical venous blood through the foramen ovale to the left atrium is absent. However, entire oxygenated blood from the placenta returns directly into the heart via the UV, so that fetal arterial blood oxygen concentration may not be affected [10]. It has been shown that the cardiothoracic ratio of the fetus correlate with the central venous pressure. The presence of cardiomegaly indicates that direct drainage of the umbilical vein into the heart leads to high central venous pressure [11,12]. This raised central venous pressure is most likely due to the volume overload as a result of loss of the DV regulatory mechanism [13]. This chronic volume overload may lead to increased demands on the fetal myocardium with the risk of high-output heart failure, leading to fetal hydrops [10]. Fetuses with DV agenesis appear to be at risk of additional cardiac and extra-cardiac anomalies [12]. Anomalies that commonly occur involve the gastrointestinal, cardiovascular, genitourinary and musculoskeletal systems [3]. Cardiovascular anomalies may include atrial septal defect, ventricular septal defect or more complex cardiac malformations. The most commonly associated genitourinary tract anomalies include bilateral hydronephrosis, ectopic kidney and unilateral renal agenesis. Musculoskeletal anomalies such as hemivertebrae have also been reported. A single umbilical artery or two-vessel cord has also been noted in some cases. Fortunately our patient did not have any significant congenital anomalies apart from the small muscular ventricular septal defect which was noted during the prenatal period and the inguinal hernia and the hydrocele noted postnatally.

Conclusion

An abnormal course of the umbilical vein is a rare anomaly and this can be associated with the congenital absence of the Ductus Venosus (DV), cardiac and extra-cardiac anomalies. Through a thorough and meticulous ultrasonographic assessment, the diagnosis can be made and that will be helpful in guiding the management of the rest of the pregnancy and counsel the family what to expect. In the absence of any other major abnormality, the prognosis of babies with direct umbilical venous return into the heart can be good.

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