Persistent Extrahepatic Right Umbilical Vein

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Abstract

Persistent right umbilical vein (PRUV) is a vascular pathology in which the left umbilical vein becomes occluded while the right umbilical vein persists and remains open. When observing fetal anatomy during a routine 18-week scan, it is important to notice the direction of the vasculature for possible fetal anomalies such as PRUV. In this case, a full detailed sonogram of the fetus was conducted and an extrahepatic PRUV was observed using 2D imaging and further confirmed using color Doppler to identify the type of PRUV. Following the identification of the PRUV, the fetus was thoroughly assessed for the possibility of other associated anomalies. Clearly, when an anomaly is observed, it is crucial to assess the fetus for associated congenital variances to identify risk and potential outcome.

Keywords

persistent right umbilical vein (PRUV), prenatal diagnosis, anomaly, fetus

Persistent right umbilical vein (PRUV) is considered a vascular pathology in which the left umbilical vein becomes occluded and the right vein persists and remains open. Normally, the right umbilical vein regresses in fetuses of 6 mm, around the fourth week of pregnancy, and completely disappears by the seventh week of gestation (Figure 1). In two-thirds of cases, the PRUV may replace the normal left umbilical vein or, more rarely (one-third of cases), be supernumerary.[1] If only the right umbilical vein is present, blood from the placenta passes through the right branch of the portal vein, the ductus venosus, the hepatic veins, and finally the inferior vena cava before entering the heart.[2] However, if both the left and right umbilical veins are present, the left umbilical vein provides the fetal circulation with placental blood through the portal system, and the right umbilical vein empties directly into the right atrium.[2] Of the PRUV, there are two types, intrahepatic and extrahepatic. The intrahepatic variant is seen when the right umbilical vein joins the portal system at the sinus venosus and proceeds to the ductus venosus. In the extrahepatic type, the right umbilical vein drains into the right atrium, the inferior vena cava, or the iliac vein. PRUV was once believed to be a rare occurrence that was strongly associated with severe fetal anomalies, including congenital heart defects as well as gastrointestinal, urinary, and musculoskeletal malformations.[1,2] Following these findings, several large retrospective studies were carried out from which it was found that a fetus with isolated PRUV has a good prognosis.[3,4]

Case Presentation

A woman in her late 20s was referred for a detailed routine 18-week sonogram, which included the brain, face, spine, heart, diaphragm, abdominal cavity, and limbs. During the scan, the amniotic fluid level, cord, placenta, uterus, and cervix were also assessed. Conventional methodology was used to measure the biparietal diameter, head circumference, abdominal circumference (AC), and femur length. The direction of the umbilical vein was determined on the AC measurement.

The 2D sonogram revealed persistent extrahepatic right umbilical vein, which was further confirmed using color Doppler (Figures 2–4). The PRUV appeared to drain into the inferior vena cava or the right atrium, with color Doppler confirming the specific type.

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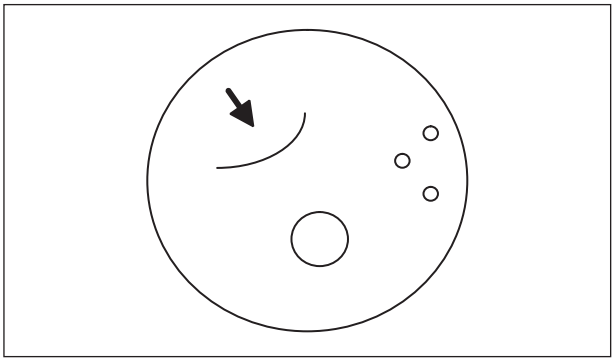

Figure 1. Normal appearance of the fetal abdomen at the level of the left umbilical vein.

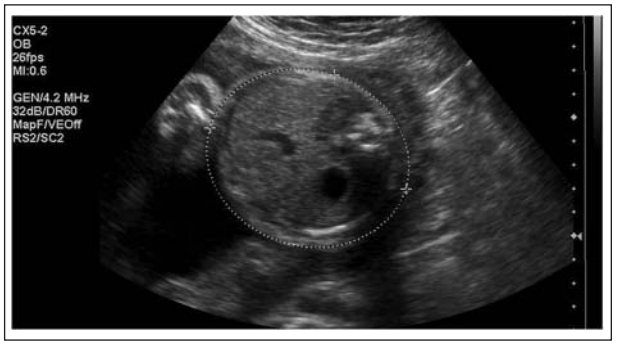

Figure 2. Fetus with persistent right umbilical vein in which the portal vein curves toward the stomach.

Sonographic criteria associated with PRUV include the portal vein curved toward the stomach (Figure 2); the fetal gallbladder located medially to the umbilical vein, between the umbilical vein and the stomach; and abnormal connection of the umbilical vein to the right portal vein instead of to the left portal vein.

Following the detection of PRUV, the fetus was carefully assessed to exclude associated and potentially more serious congenital malformations, none of which were found. Two, more extensive, follow-up sonograms were also performed in addition to fetal echocardiography at 23 weeks and 28 weeks, respectively, in addition to regular fetal monitoring.

Both follow-up scans along with the fetal echocardiography showed normal growth, fluid, and Doppler with no structural anomalies identified. The heart was situs solitus and appeared structurally normal. Thus, chromosomal testing was not carried out as no indications were found. The PRUV found in the fetus appeared to be an isolated finding, likely a normal variant of no clinical consequence after birth.

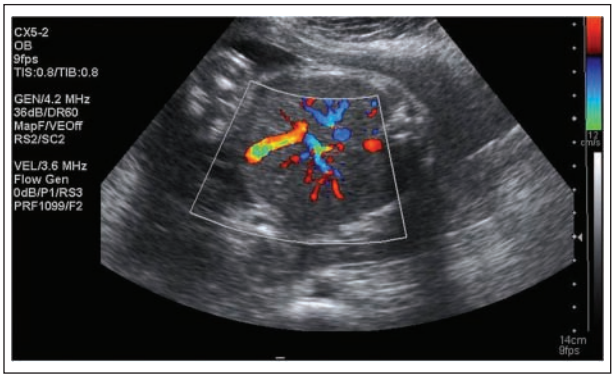

Figure 3. Color Doppler used to confirm the type of persistent right umbilical vein according to its drainage.

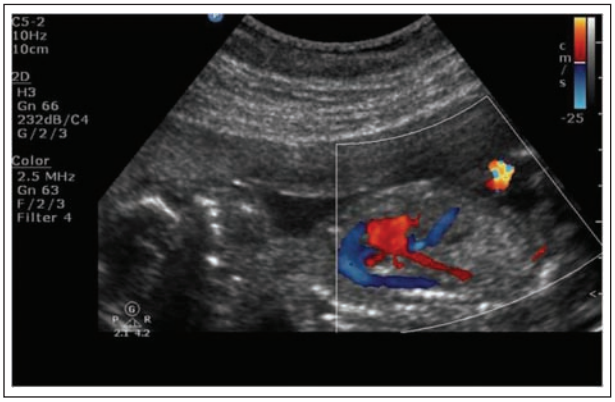

Figure 4. Color Doppler used to confirm the type of persistent right umbilical vein (PRUV) according to its drainage. Arrow, PRUV.

Discussion

At around the fourth week of gestation, the chorionic veins drain into the paired allantoic veins to form the primordial double umbilical vein. Three sets of veins—the common cardiac veins, viteline veins, and umbilical veins—drain into the primitive heart and join at the sinus venosus, which serves to drain the body, yolk sac, and placenta, respectively. During the sixth gestational week, the hepatic bud enlarges, and the right umbilical vein between the liver and the sinus venosus degenerates, leaving only a left umbilical vein to carry all the blood from the placenta to the fetus. The ductus venosus forms in the liver and connects the left umbilical vein and the inferior vena cava. The ductus allows blood to bypass the liver and flow directly from the placenta to the heart. Postnatally, the left umbilical vein becomes the ligamentum teres.

PRUV was traditionally thought to be a rare occurrence because of the lack of reports in the literature on this

variant until 1990.[1,2,5] Since then, several larger studies have been published, and the results suggest that this anomaly is more common than previously believed.[5]

In cases involving PRUV, the right umbilical vein is persistently open and may coexist with the left umbilical vein as an intrahepatic supernumerary structure or connect separately to the right portal vein.[6] The right umbilical vein may also completely replace the left umbilical vein or bypass the liver to create an aberrant drainage into the inferior vena cava or the right atrium, which is known as the extrahepatic type.[1,4,6,7]

Of the three types of PRUV, the intrahepatic type (type 1) is the most common in the fetus with isolated PRUV.[6] With this type, the umbilical vein passes lateral to the right side of the gallbladder and fuses with the right portal vein, then curves toward the stomach. Once passing through the ductus venosus, the umbilical vein connects with the hepatic vein and drains into the inferior vena cava. As there is little interference in hemodynamics, this type of PRUV has a good prognosis as it is estimated that only 20% to 30% of the blood of the umbilical vein enters the ductus venosus and reaches the heart.[6] In the second type, the umbilical vein connects to the iliac veins or caput medusa directly without the ductus venous. Last, in the third type, the umbilical vein connects directly to the right atrium or infracardiac portion of the inferior vena cava in the absence of the ductus venosus.[1,7] As opposed to type 1, the other two types of PRUV are extrahepatic and have a higher frequency of associated anomalies and greater hemodynamic effects due to the absence of the ductus venosus.[6,7] There are case reports of the extrahepatic type of PRUV without the ductus venosus in which these fetuses had severe hemodynamic stresses that resulted in hydrops fetalis.[1,8]

The precise causes of failure of normal regression of the right umbilical vein are unknown; however, several potential etiologies have been suggested. In the rat model, specific teratogens such as retinoic acid and first-trimester folic acid deficiency may result in persistence of the right umbilical vein.[1,3,4,6] Obstruction of the left umbilical artery by thrombus, embolus, or external pressure early in the pregnancy might also cause the right umbilical vein to remain patent to maintain placental blood supply to the fetus.[3,6,9]

The prevalence of PRUV was not well established until more recently, and now the incidence and significance have become the focus to put aside the controversy on the occurrence of this vascular pathology. Jeanty[1] gathered only a dozen reported cases in the literature and added six new cases.[2,3,5] Associated anomalies were found in all published cases and in three of Jeanty's own six patients, which suggested that PRUV might be an ominous prenatal finding.[1] Results from several retrospective studies demonstrate the incidence of PRUV to be more frequent than previously reported. Blazer et al.[3] found the incidence to be 1:438, which was similar to ratios of 1:476 reported by Hill et al.,[2] 1:450 detailed by Shen et al.,[5] and 1:526 reported by Wolman et al.[4] However, these were significantly lower than the rate of 1:217 recently reported by Yang et al.,[6] who detected six fetuses with PRUV among 1302 study subjects. Yang et al.[6] attributed the higher incidence of PRUV to the fact that their obstetric population included both private service and women transferred to their department in the third trimester of pregnancy.

The true incidence of PRUV may be much higher, considering the false-negative rate of sonographic detection. Through several studies, it has been found that the intrahepatic form of PRUV might present as an isolated finding, with no other anomalies, whereas those cases demonstrating the extrahepatic type were associated with fetal abnormalities.[3,7] PRUV with no intrahepatic portion is an uncommon finding, with a review of the literature revealing only five other cases.[7] There is an association with congenital cardiac malformation, and the presence of PRUV can sometimes be the only clue to alert sonographers and clinicians to the presence of these malformations.[10] Other associated conditions include abnormal systemic venous connections[11] and Noonan syndrome.[7]

Conclusion

In agreement with Jeanty,[1] PRUV might not be as rare as was previously believed, and proper alertness might uncover its true prevalence. Of all imaging modalities, sonography is the most likely to facilitate detection of the anomaly because the section used to demonstrate PRUV is routinely used in all obstetrical scanning. In addition, sonography can depict the anomaly at a stage when it may be of clinical significance, and color Doppler can be applied to confirm if the direction of flow is toward the stomach. If such an anomaly is identified, an extensive targeted sonographic examination should be carried out. Depending on the significance of the abnormalities detected via sonography, a chromosomal study may be warranted and offered to the patient. This case in particular proves to be interesting as the anomaly detected was an extraheptic PRUV, which is commonly associated with more severe congenital malformations. On the basis of three extensive sonograms coupled with echocardiography, the fetus appeared normal with no associated anomalies in utero and was likely to have no clinical consequence after birth.

Declaration of Conflicting Interests

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