

# PRENATAL DIAGNOSIS OF PERSISTENT RIGHT UMBILICAL VEIN USING THREE-DIMENSIONAL SONOGRAPHY WITH POWER DOPPLER

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## SUMMARY

**Objective:** To investigate the incidence and the importance of isolated persistent right umbilical vein (PRUV) in our obstetric population and to determine the role of three-dimensional (3D) ultrasound in prenatal diagnosis of isolated PRUV.

**Material and Methods:** A total of 1,302 women who received regular antenatal care by a sole obstetrician at our hospital were prospectively evaluated between July 2003 and April 2005. Detailed anatomical evaluation of the fetus was performed by one sonographer. When the diagnosis of PRUV was made, it was confirmed by a senior obstetrician. 3D ultrasound with power Doppler was applied to delineate local anatomy. Echocardiography was performed in all the newborns by pediatric cardiologists to confirm the prenatal diagnosis and to evaluate for the presence of associated anomalies.

**Results:** Six fetuses with PRUV were detected among the 1,302 study subjects. The incidence of PRUV in our population was 0.46% (1:217 live births). Vascular anatomy was easy to demonstrate using 3D power Doppler. The ductus venosus (DV) was present in all six fetuses. An atrial septal defect was shown to exist in four newborns by neonatal echocardiography, but spontaneous closure had occurred in the follow-up scan.

**Conclusion:** PRUV is a common vascular anomaly that is easy to be overlooked. Reconstruction of the portal system in the affected fetuses using 3D ultrasound facilitated the identification of the DV. If the DV is present, and other anomalies are excluded, the fetus with PRUV has a good outcome. [*Taiwanese J Obstet Gynecol* 2007; 46(1):43-46]

**Key Words:** ductus venosus, persistent right umbilical vein, three-dimensional ultrasound

## Introduction

Persistent right umbilical vein (PRUV) is a pathologic vascular anomaly occurring in embryonic development in which the right umbilical vein persists and the left vein is occluded. Prior to 1995, PRUV was thought to be an uncommon event and was often associated with

severe fetal anomalies [1,2]. Thereafter, several large retrospective studies were conducted from which it was concluded that the fetus with isolated PRUV has a good prognosis [3-5]. Here, we report 6 cases with isolated PRUV diagnosed from July 2003 to April 2005. We also report the usefulness of three-dimensional (3D) ultrasound to reconstruct the portal venous system in fetuses with PRUV.

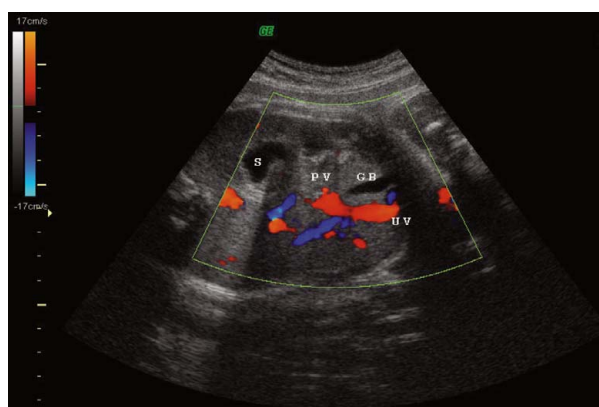
## Material and Methods

A total of 1,302 pregnant women who received regular antenatal care by the sole obstetrician at our hospital

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Accepted: July 20, 2006

were prospectively evaluated between July 2003 and April 2005. Detailed anatomical evaluation of the fetuses was performed by one sonographer using a GE Voluson 730 Expert ultrasound machine (GE Medical Systems, Milwaukee, WI, USA). Sonographic criteria used to diagnose PRUV included: (1) the portal vein curving toward the stomach rather than the right lobe of the liver, (2) location of the fetal gallbladder medial to the umbilical vein, and (3) an abnormal connection of the umbilical vein to the right, as opposed to the left portal vein (Figure 1). When the diagnosis of PRUV was made, it was confirmed by a senior obstetrician who is familiar with targeted scans.

Three-dimensional ultrasound with power Doppler was utilized to delineate local anomalies. A commercially available Voluson 730 scanner and a transabdominal convex volume transducer with isonation frequencies of 4–8 MHz were used. Three-dimensional power Doppler imaging to visualize the fetal portal vein system was similar to that described in previous studies [6]. Briefly, the fetal axial plane used to measure the abdominal circumference was chosen for data acquisition. Then, the 2D power Doppler was initiated. The



**Figure 1.** Two-dimensional sonograph of fetus with persistent right umbilical vein in the axial plane to measure abdominal circumference with color Doppler. UV = umbilical vein; S = stomach; PV = portal vein; GB = gallbladder.

color area was set at an appropriate size to cover the area of umbilical vein and portal system. The volume angle was set at 55°C. During data acquisition, transducer movement was avoided and the women were asked to hold their breath for 7.5–10 seconds. We used a medium wall filter and a gain of 50% to avoid movement artifacts. The volume data set was made automatically and could be used for post-scan analysis.

Echocardiography was performed on all newborns by pediatric cardiologists to confirm the diagnosis of PRUV and ascertain the presence of any associated anomalies.

## Results

Six fetuses with PRUV were detected among the 1,302 study subjects. The incidence of PRUV in our population was therefore 0.46% (one in 217 live births). The mean maternal age was 31.2 years old and the mean gestational age at diagnosis was 34 weeks (Table).

The ductus venosus (DV) was present in all six fetuses (Figure 2). An atrial septal defect (ASD) was observed in four newborns by neonatal echocardiography, but spontaneous closure had occurred in the follow-up scan. Three-dimensional power Doppler ultrasonography clearly showed the umbilical vein turning to the left side, then in a cranial direction to join with the right portal vein, through the DV, and entering the inferior vena cava (IVC). From a longitudinal view, we simultaneously visualized the presence of branches of the hepatic vein connecting separately to the IVC (Figure 3). The DV was demonstrated in all cases.

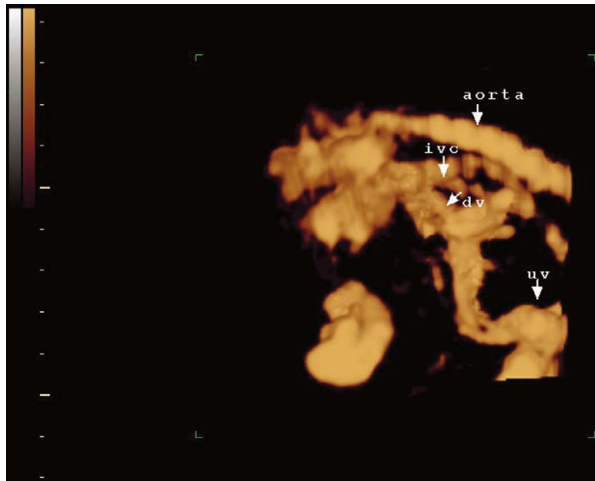
## Discussion

During the 4<sup>th</sup> week of embryonic development, regression of the right umbilical vein usually occurs while the left umbilical vein remains patent between the derivative of the vitelline veins and the DV. In those cases

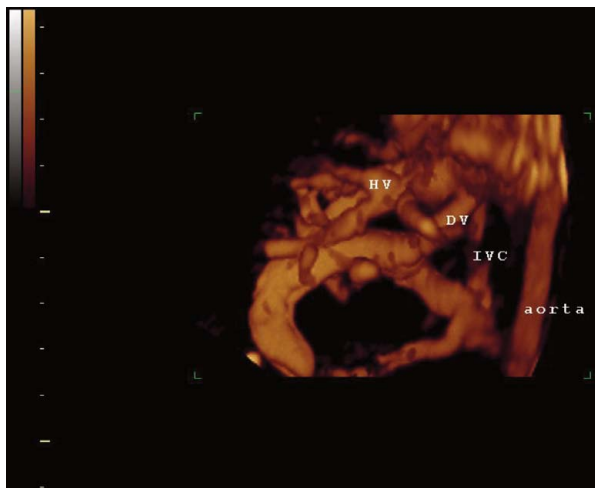
**Table.** Characteristics and outcome of persistent right umbilical vein (PRUV) in our 6 cases

Case no.	Maternal age (yr)	Para	Detection age (wk)	Ultrasound finding, type	Additional malformation	Outcome
1	34	2	28	IH, PRUV	ASD secundum	Healthy
2	36	3	37	IH, PRUV	ASD secundum	Healthy
3	31	2	33	IH, PRUV	Nil	Healthy
4	34	2	34	IH, PRUV	Nil	Healthy
5	25	1	37	IH, PRUV	ASD secundum	Healthy
6	28	1	37	IH, PRUV	ASD secundum	Healthy

IH = intrahepatic; ASD = atrial septal defect.



**Figure 2.** Surface rendering of three-dimensional ultrasound with power Doppler. ivc = inferior vena cava; dv = ductus venosus; uv = umbilical vein.



**Figure 3.** Longitudinal view of three-dimensional ultrasound with power Doppler. HV = hepatic vein; DV = ductus venosus; IVC = inferior vena cava.

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. The right umbilical vein may also completely replace the left umbilical vein or bypass the liver to create an aberrant drainage of blood into the IVC or the right atrium, the so-called extrahepatic type of PRUV [1,3,7,8]. There are some case reports of extrahepatic types of PRUV without the DV. The affected fetuses had severe hemodynamic burdens that resulted in hydrops fetalis [1,7,9].

The cause of failure of normal regression of right umbilical vein is unknown. Specific teratogens, maternal folic acid deficiency in the first trimester, or obstruction of the left umbilical vein by a thrombus have been proposed [10]. Traditionally, PRUV was thought to be

a rare finding that was associated with cardiovascular, gastrointestinal, urinary, musculoskeletal, and central nervous system anomalies [1–5,7,9,11,12]. Several recent studies have suggested that PRUV is more common than had been previously thought [3–5]. The incidence of PRUV in our population was 0.46%, slightly higher than that has been previously reported (i.e. 0.19–0.4%). This higher incidence may be because our obstetric population included both private service and women transferred to our department in the third trimester of pregnancy in preparation for delivery at our hospital. A vascular anomaly of this nature is easy to be overlooked. This may also explain why the diagnosis of PRUV was delayed in our unit.

In previous reports, most fetuses with PRUV have no concomitant obvious anomalies [3–5]. In the study reported by Blazer et al [4], of 69 fetuses with PRUV, only nine fetuses (1.4%) had other anomalies. In another recent report by Wolman et al [5], 13 of 17 cases had isolated PRUV without other abnormal findings. In our cases, none of the fetuses had other malformations, with the exception of ASD secundum, which is difficult, even not impossible, to diagnose antenatally. Four of our 6 cases with PRUV had ASD secundum, which was detected by postpartum echocardiography. All of the fetuses were uneventful without surgical intervention.

Of the 3 types of PRUV, the intrahepatic type (type 1) is the most prevalent in the fetus with isolated PRUV. In the intrahepatic type, the umbilical vein passes lateral to the right side of the gallbladder, and fuses with the right portal vein, then bends toward the stomach. After passing through the DV, the umbilical vein connects with the hepatic vein and drains into the IVC. Because there is little interference in hemodynamics, this type of PRUV has a good prognosis. It is estimated that just 20–30% of the blood of the umbilical vein enters the DV and reaches the heart. In type 2, the umbilical vein connects to the iliac veins or caput medusae directly with absence of the DV. In type 3, the umbilical vein connects directly to the right atrium or infracardiac portion of the inferior vena cava without by way of DV [1,8]. In contrast 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 absence of the DV. There are several case reports of agenesis of the DV with hydrops fetalis [13,14]. If the DV is absent, the blood of the umbilical vein will return to the heart directly. We proposed that this might increase hemodynamic burden and lead to hydrops fetalis. The prognosis of extrahepatic types of PRUV is thus worse than that of the intrahepatic type. When the DV is observed, it carries a

better prognosis [7]. All the fetuses with PRUV in our study had the DV. The DV can easily be visualized with 3D ultrasound because vascular anatomy is easily discriminated with a 3D power Doppler scan. It is our opinion that the prognosis of the fetus with intrahepatic type PRUV is good when the DV is present. To the best of our knowledge, this is the first report using 3D ultrasound to assist in the prenatal diagnosis of PRUV.

In summary, we confirm that PRUV is not as rare as previous reports. The incidence in our population was 1:217 live births. If the DV is found and other anomalies are excluded, the fetus with PRUV carries a good prognosis.

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