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Persistent right umbilical vein: incidence and significance

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ABSTRACT

Objectives *To conduct a prospective evaluation of the incidence and neonatal outcome of fetuses with persistent right umbilical vein. This condition had traditionally been considered to be extremely rare and to be associated with a very poor neonatal prognosis, but later evidence has raised some doubts about the veracity of these contentions.*

Methods *Between August 1995 and November 1998, 8950 low-risk patients were prospectively evaluated at two medical centers. The sonographic diagnosis of a persistent right umbilical vein was made in a transverse section of the fetal abdomen when the portal vein was curved toward the stomach, and the fetal gall bladder was located medially to the umbilical vein.*

Results *Persistent right umbilical vein was detected in 17 fetuses during the study. Four of them had additional malformations, of which three had been detected antenatally.*

Conclusions *We established that the incidence of persistent right umbilical vein in a low-risk population is 1 : 526. We believe that the sonographic finding of this anomaly is an indication for conducting targeted fetal sonography and echocardiography. When the persistent right umbilical vein is connected to the portal system and other anomalies are ruled out, the prognosis can generally be expected to be favorable.*

INTRODUCTION

Persistent right umbilical vein (PRUV) is a vascular pathology in which the left umbilical vein becomes occluded and the right vein persists in remaining open. In the normal fetus, the right umbilical vein begins to become obliterated around the 4th week of pregnancy, and disappears by the 7th week of gestation[1]. The persistence of an open right umbilical vein does not prevent the formation of the ductus venosus. Although the course of the blood within the liver is abnormal, it does not alter blood distribution to the fetus.

Persistent right umbilical vein was traditionally thought to be an uncommon event strongly associated with severe fetal anomalies, including congenital heart defects, and gastrointestinal, urinary and musculoskeletal system malformations[1,2], of which only a few cases had been reported in the literature up to 1995[1–6]. Since then, it has been suggested that this anomaly might not be as rare as could be implied by the paucity of the reports in the literature[7]. Furthermore, it has been suggested that the good outcome of babies with isolated PRUV indicates that this anomaly has only minor prognostic significance[7,8]. The incidence of this condition has never been established.

This prospective study was designed to determine the incidence and neonatal outcome of babies with PRUV in a large low-risk population.

MATERIALS AND METHODS

Between August 1995 and November 1998, a total of 8950 low-risk patients between 14 and 24 weeks of gestation was prospectively evaluated at two medical centers (Lis Maternity Hospital, Tel Aviv Sourasky Medical Center in Tel Aviv and the Herzylia Medical Center in Herzylia, Israel). A detailed structural evaluation of the fetus, including biometrical studies, was carried out. Special attention was given to the direction of the curve of the portal vein and the position of the gall bladder in each scan. These examinations were performed by two sonographers (I.W. and I.G.) using HDI 3000 and HDI-9 ultrasound machines (ATL, Bothell, WA, USA) both with a 2–4-MHz transabdominal transducer. For examinations at 14–16 weeks, a transvaginal transducer of frequency 4–8 MHz was used with the HDI 3000 and a 5–9-MHz transvaginal probe was used with the HDI-9.

The diagnosis of a PRUV was made in a transverse section of the fetal abdomen when the following criteria were met: (1) the portal vein curved towards the stomach (Figure 1), (2) the fetal gall bladder was located medially to the umbilical vein (i.e. between the umbilical vein and the stomach) (Figure 2), or (3) the umbilical vein was abnormally

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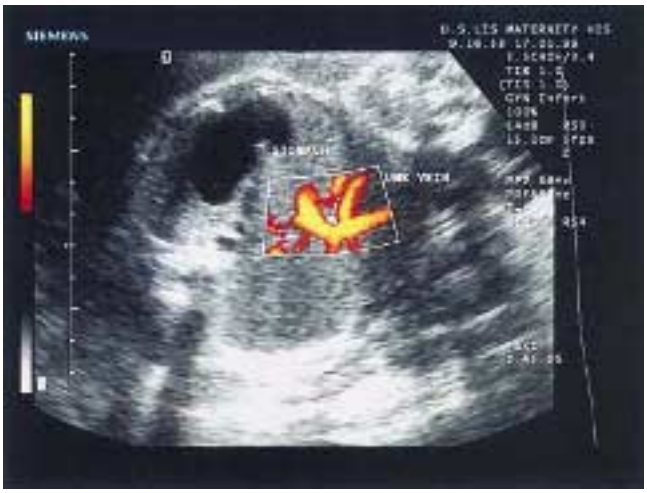

Figure 1 Umbilical vein curved towards the stomach.

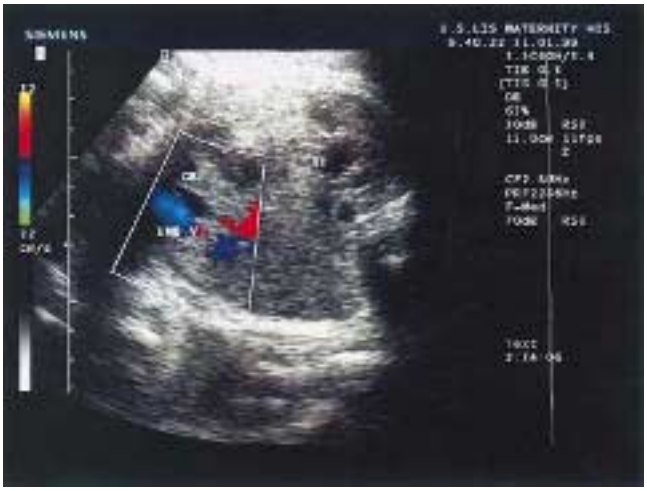

Figure 2 Fetal gall bladder (GB) located medially to the umbilical vein (UMB V). ST, stomach. Both figures were made with a color printer which is attached to a Sonoline Elegra ultrasound machine, (Siemens Medical System, Inc., Issaquah, WA, USA) with an abdominal 3.5-MHz curved array transducer.

connected to the right portal vein instead of to the left portal vein.

Once the diagnosis was made, it was confirmed by a second sonographer (A.J.J.) and the fetus underwent a more extensive evaluation. Fetal karyotyping was performed for obstetric indications including maternal age and abnormal triple-test result or their combination with the presence of a soft ultrasound marker. The umbilical cord was viewed in a transverse section and the number of vessels was determined. The fetuses also underwent echocardiographic evaluation. All neonates were carefully evaluated after birth for any additional congenital malformations that might have been missed by the sonographic examination.

RESULTS

Seventeen fetuses with PRUV were detected during the study period among the 8950 participants, yielding an incidence of 1 : 526. The mean ± standard deviation (SD) maternal age was 29 ± 5.2 years, fetal weight was 3470 ± 420 g, and gestational age at delivery was 38 ± 2.3 weeks. Of the 15 patients who delivered, 11 delivered spontaneously, three by vacuum extraction, and one by outlet forceps. The mean ± SD 5-min Apgar score was 9.6 ± 0.62.

Four of the 17 fetuses with PRUV had additional malformations of which three had been detected antenatally (Table 1). One fetus had anencephaly detected at 18 weeks of gestation and it was terminated. Another fetus had bilateral cleft lip and a complicated heart defect detected at 24 weeks of gestation: the pregnancy was terminated, and karyotyping revealed trisomy 18. The third fetus was born with bilateral atrophy of the optic nerve which was detected by magnetic resonance imaging only after birth. The fourth fetus had a single umbilical artery and agenesis of one kidney: the results of karyotyping were normal as were other screening procedures. The presence of a single kidney was confirmed after birth. The remaining 13 fetuses were born without any other malformation.

DISCUSSION

Persistent right umbilical vein was traditionally thought to be a rare finding because of the paucity of reports in the literature on its occurrence until 1995[1–6]. Since then, several larger series have been published and their results suggest that this anomaly is more common than had been widely upheld[7–9]. The incidence of PRUV has not previously been established, and the question of its significance remains controversial. Jeanty[2] reported that three of the six fetuses in his series of PRUV had anomalies ranging from benign to severe. Hill *et al.*[6] described 33 cases of PRUV detected between 15 and 37 weeks' gestation in a mixed high- and low-risk population monitored throughout pregnancy, and reported an incidence of 1 : 476. Additional malformations were discovered in 18.2% of fetuses, a figure which is in accordance with our finding of a 23% incidence of concomitant malformations. Their six patients were karyotyped and were all found to be normal. One case of trisomy 18 was detected amongst the four fetuses in our series which presented with additional malformations. However, of the 13 other cases with PRUV alone, nine underwent karyotyping and all were found to be normal. One fetus in the study of Hill *et al.*[6] was delivered at 34 weeks' gestation due to severe intrauterine growth restriction. Two of our pregnancies were terminated and the other

Table 1 The time of persistent right umbilical vein detection and outcome in the four cases of PRUV with additional anomalies

Detection (weeks)	*Additional anomaly*	*Outcome*
18	Anencephaly	TOP
24	Complicated heart malformation, bilateral cleft lip	TOP
17	Bilateral atrophy of optic nerve	Blindness
15	Single umbilical artery, single kidney	Confirmed after delivery

TOP, termination of pregnancy.

15 delivered at term. Although a single umbilical artery is the most common congenital anomaly associated with PRUV[4], there was only one such case in our series.

Shen *et al.*[7] described eight cases of PRUV detected on routine second-trimester ultrasound in an unselected population. Of the seven neonates that were delivered, one had a dextrocardia and a right-sided descending aorta but no other malformations. The outcome was favorable in all cases and all seven infants were well and thriving after 18 months of follow-up.

In the very early stages of intrauterine life, blood from the placenta is directed into the fetus via two umbilical veins. Both these veins bypass the liver through the splanchnic mesoderm and open into the sinus horns. By the 4th week of pregnancy, the right umbilical veins begin to become obliterated and completely disappears by the 7th week of gestation. The left umbilical vein undergoes similar processes at its distal part and then connects to the hepatic sinusoids, which participate in the development of the portal venous branches and the ductus venosus. When the left umbilical vein occludes and the right vein remains open, PRUV results. Persistent right umbilical vein can be divided into two forms[10]: the intrahepatic type, in which the aberrant right umbilical vein joins the portal system at the sinus venosus and proceeds to the ductus venosus (this type is not associated with other anomalies); the extrahepatic type, in which the tight umbilical vein drains to the right atrium, the inferior vena cava or the iliac vein. This type of PRUV is associated more frequently with central nervous system, heart and gastrointestinal malformations.

Several mechanisms have been proposed as a cause for PRUV. In a rat model[2], first-trimester folic acid deficiency or specific teratogens such as retinoic acid may cause the appearance of PRUV. Early obstruction of the left umbilical vein from external pressure or occlusion may also result in PRUV.

Persistent right umbilical vein can be reliably diagnosed on gray-scale ultrasound once the operator is alerted to the possibility of its existence. There are two simple sonographic landmarks which facilitate the diagnosis. One is that the portal vein is curved towards the stomach in a transverse section of the fetal abdomen (Figure 1) and the other is that the fetal gall bladder is located medially to the umbilical vein (between the umbilical vein and the stomach) (Figure 2). Color Doppler is needed only for demonstrating the type of PRUV according to its drainage.

It appears that the prognosis is good when the PRUV is connected to the portal system and other anomalies are ruled out. A similar observation was made by Kirsch *et al.*[9] who described nine cases of PRUV; other than one case of hypospadias, no additional malformations were detected before or after birth. The authors suggested that this vascular variant may not necessarily indicate an ominous finding.

Among the different anomalies reported in association with PRUV are gastrointestinal malformations, various cardiac anomalies, skeletal malformations, urinary tract malformations and single umbilical artery[1,2,4,5,11].

We believe that the sonographic finding of this anomaly is an indication for conducting targeted fetal sonography and fetal echocardiography, and that a close sonographic follow-up is needed throughout pregnancy. Due to the small size of this series it is hard to draw conclusions regarding the need for performing karyotyping once the diagnosis is made. However, having ruled out additional malformations, the likelihood is that the prognosis will be favorable in the majority of cases.

In summary, we have conducted a prospective study to estimate the incidence of PRUV in a low-risk population. Contradictory to long-held beliefs, we have been able to show that this condition seems to be fairly common, with an incidence of 1 : 526, and that it bears a good prognosis in cases in which no additional malformation exists.

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