Ultrasound Obstet. Gynecol. 8 (1996) 31–33

Prenatal diagnosis of persistent right umbilical vein

*O. Shen, O. P. Tadmor and S. Yagel**

Department of Obstetrics and Gynecology, Shaare Zedek Medical Center; *Department of Obstetrics and Gynecology, Hadassah Mount Scopus, Jerusalem, Israel

Key words: ULTRASOUND, PRENATAL DIAGNOSIS, FETUS, UMBILICAL VEIN, ANOMALY

ABSTRACT

A persistent right umbilical vein is thought to be a rare anomaly, frequently associated with other life-threatening malformations.

Eight cases of prenatal diagnosis of persistent right umbilical vein are presented. All cases were discovered on a routine second-trimester sonogram in an unselected population. Of the seven babies who were delivered, one had a dextrocardia and right-sided descending aorta, but none had other associated major malformations. The outcome was favorable in all cases and all seven infants are well and thriving with up to a year and a half of follow-up.

Our findings suggest that persistent right umbilical vein might not be as rare as the paucity of reports in the literature implies. Provided that no other malformations are present, this anomaly is probably of little prognostic significance, requiring no further evaluation or follow-up.

INTRODUCTION

By the seventh gestational week, three pairs of major veins connect to the primitive heart: the vitelline veins, draining blood from the yolk sac and primitive midgut[1]; the cardinal veins, collecting blood from the embryo; and the umbilical veins, carrying the oxygen-rich blood from the placenta to the embryo. The umbilical veins pass initially on each side of the liver bud. In the course of embryogenesis, the umbilical veins connect to the hepatic sinusoids. Subsequently, in the 6-mm embryo, the right umbilical vein degenerates, leaving the entire placental venous return to the left umbilical vein, which connects to the left portal vein. Later on, a direct connection is established between the left portal vein and the systemic circulation – the ductus venosus. Still later, the umbilical vein shifts to the midline and occupies the free edge of the falciform ligament.

The reason for the right umbilical vein remaining patent in some cases is unknown. When it does, it might connect to the right portal vein or it might bypass the liver and connect to the inferior vena cava or the right atrium.

Only 20 cases of persistent right umbilical vein have been reported, the majority in the pathological literature. Not surprisingly, all cases diagnosed postnatally have been associated with various other congenital anomalies that dominated the clinical presentation[2–5]. There has been a single report, of six cases, diagnosed prenatally[6].

Jeanty, describing the first six cases of sonographic prenatal detection of this anomaly, expected half of them to have a poor outcome due to associated anomalies. In three of his cases, in which the anomalous vein was the only abnormality, the outcome was good. We add eight of our own cases, none with associated major malformations. Seven of our cases have been delivered, all with a favorable outcome.

CASE REPORTS

In all eight cases presented, the diagnosis of persistent right umbilical vein was made on a routine targeted organ scan at the Hadassah Mount Scopus ultrasound unit between January 1994 and October 1995. We view, as a matter of routine, the face, brain, spine, lungs and diaphragm, perform an echocardiogram, including color Doppler, and scan the abdomen, also using color Doppler, for the umbilical cord insertion and the long bones, fingers and toes. Scans were performed using the Acuson color 128 XP-10 with a 3.5-MHz transducer. In all cases the umbilical vein's course was lateral to and on the right side of the gallbladder, connecting to the right portal vein and curving toward the stomach rather than away from it (Figure 1). Table 1 summarizes the demographic characteristics of the eight cases. In all cases, pregnancy and delivery were uncomplicated, except in case 2, in which the patient required treatment for premature contractions, and eventually delivered at term. In case 8, at the time of submission of

Correspondence: Dr O. Shen, Department of Obstetrics and Gynecology, Shaare Zedek Medical Center, Jerusalem, 91031 Israel

Received 6–3–95
Revised 27–2–96
Accepted 29–3–96

Table 1 Summary of eight cases of persistent right umbilical vein

Case	*Age* (years)	*Scan week*	*Birth weight* (g)	*Gravida*	*Para*	*Associated anomalies*	*Sex*	*Remarks*
1	25	22	2.8	2	1	none	F	see Figure 1
2	38	20	3.9	7	4	none	M	
3	29	21	3.2	6	4	none	M	
4	24	20	3.6	2	1	dextrocardia	F	see text
5	27	22	3.4	9	5	none	M	
6	25	21	3.4	4	3	none	F	
7	22	24	3.2	2	1	none	F	
8	29	22		2	0	undelivered on submission		

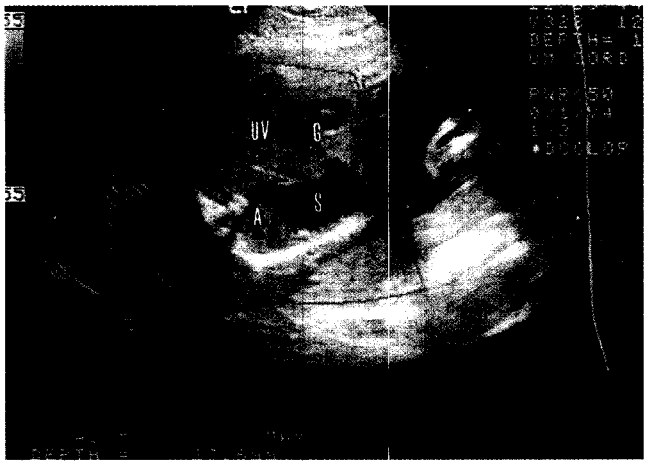

Figure 1 Case 1: persistent right umbilical vein. UV, umbilical vein; S, stomach; G, gallbladder; A, aorta

this paper, the patient is still pregnant. In all cases with the exception of case 4, persistent right umbilical vein was the only abnormality detected. In case 4, dextrocardia and right-sided descending aorta were diagnosed as well. The karyotype, performed because of the dextrocardia, was 46,XX. In the other six cases, the neonatal examination was normal. In case 4, the prenatal diagnosis of dextrocardia was confirmed. The seven infants are well and thriving with up to a year and a half of follow-up.

DISCUSSION

The sonographic prenatal diagnosis of persistent right umbilical vein is straightforward. The umbilical vein passes lateral to and to the right side of the gallbladder, where it connects to the right portal vein. It then curves leftward towards the stomach instead of curving away to the right. This picture is usually easily visible on the section obtained to measure the abdominal circumference. Color Doppler further facilitates the diagnosis by accentuating the aberrant intrahepatic course of the umbilical vein and distinguishing it from the gallbladder. This might be especially helpful in cases where image resolution is poor.

This anomaly is often associated with serious malformations, most of which are readily identifiable on the prenatal sonogram (Table 2). It follows that the diagnosis should prompt a detailed targeted study of the fetus. Twelve of the total 18 reported cases of persistent right umbilical vein have been published in the surgical, radiological or pathological literature. The diagnosis had been made either on umbilical vein catheterization, or as an incidental finding on autopsy. The indication for catheterization or the cause of death was related, in all cases, to the associated defects. An isolated anomalous umbilical vein might present as bowel obstruction due to the abnormal location of the falciform ligament[7].

Table 2 Anomalies associated with persistent right umbilical vein

Anomaly	*Estimated incidence (per number of live births)*
Duodenal atresia[6]	1 : 10 0008
Imperforate anus[2]	1 : 5000[9]
Echogenic bowel[6]	1 : 500[10]
Bowel malrotation[3]	1 : 6000[11]
Annular pancreas[2]	2.5–10 : 100 000[12]
Situs inversus[3,4]	1 : 5000[13]
Atrial septal defect[5]	1 : 2000[13]
Total anomalous pulmonary venous return[5]	1 : 14 000[13]
Ventricular septal defect[5]	1 : 500[13]
Mitral atresia[6]	—
Coarctation of the aorta[6]	1 : 3500[13]
Double outlet right ventricle[6]	1 : 15 000[13]
Tracheoesophageal fistula[3]	1 : 2500[14]
Non-immune hydrops[6]	1 : 1600–70 000[15]
Phocomelia[5]	—
Hemivertebra[5]	—
Sirenomelia[4]	1 : 60 000[16]
Unilateral agenesis of the kidney[5]	1–5 : 1000[17]
Bilateral hydronephrosis[4]	—
Ectopic kidney[4]	1 : 1200[18]
Unicornuate uterus[5]	—
Hypospadias[6]	1 : 300[19]
Single umbilical artery[3–6]	2–10 : 1000[20]

All ten cases reported to date (including this report) of infants with isolated persistent right umbilical vein have had a favorable outcome. It should be remembered, however, that some of the possible associated malformations might not be recognized on the prenatal sonogram. We suggest that, in cases of persistent right umbilical vein, a meticulous search for other anomalies should be carried out. If none are detected, the patient should be reassured that the condition is usually benign, and requires no further evaluation. The need to obtain a karyotype should be dictated by the nature of any associated malformations. The rarity of reported persistent right umbilical vein precludes any attempt, at this time, to estimate any added risk of chromosomal abnormality in such cases. In none of the

previously reported cases was an abnormal karyotype found.

Special attention should be given to any respiratory or feeding problems in the neonatal period, resulting possibly from a previously undetected malformation.

Jeanty[6], scanning a selected population, detected six cases in 16 months. We agree with Jeanty[6] that this anomaly might not be as rare as was previously believed, and proper alertness might uncover its true prevalence. An accurate estimation is difficult to make, at this time, given the small numbers. During the above-mentioned period, we have performed some 3600 targeted scans, putting the local incidence during that specific time at 2.2 : 1000. An increased awareness in our department, arising after the first case, is a plausible explanation for the possibly increased detection rate.

In Jeanty's series one pregnancy was terminated because of multiple malformations (none of them lethal). A second fetus with severe growth retardation was delivered at 27 weeks, and died several weeks later. A third has a guarded prognosis due to a complex cardiac malformation. The three other fetuses were either normal other than the persistent right umbilical vein, or had associated minor malformations. The overall worse prognosis in that report might be related to the more selected nature of the target population. Only additional reporting of this anomaly will disclose its true incidence and prognostic significance.

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