Persistent right umbilical vein: a prenatal condition worth mentioning?

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KEYWORDS: fetus; persistent right umbilical vein; prenatal; ultrasound; venous system

ABSTRACT

Objectives To investigate the incidence and clinical impact of prenatally diagnosed persistent right umbilical vein (PRUV) in a referral population and to evaluate the findings together with those from previous publications.

Methods This was a retrospective analysis of 39 cases with PRUV diagnosed in three tertiary referral centers for prenatal medicine between 1996 and 2009. Fetuses with situs inversus, situs ambiguous and heterotaxy (left and right isomerism) were excluded. During the study period 46,653 consecutive high- and low-risk pregnancies were examined. The prenatal sonograms and neonatal outcome data of affected individuals were reviewed. Our findings were analyzed together with findings retrieved from the scientific literature.

Results Twenty-nine fetuses had an isolated PRUV as a single anomaly, whereas 10/39 (25.6%) were found to have PRUV accompanied by additional minor and major abnormalities. These anomalies comprised mainly cardiovascular, genitourinary and gastrointestinal malformations. In common with our series, previously published cases of isolated PRUV (n = 197) displayed an uneventful course of pregnancy and a favorable postnatal outcome. Sixty-six previously reported cases of PRUV with additional anomalies were identified in the literature. Intrahepatic umbilical drainage is the most frequent type of PRUV. Including our cases, there were 16 cases (5.3%) with extrahepatic drainage of PRUV, all of which had additional anomalies.

Conclusions Consistent with previous reports, in the majority of cases (74.8%) PRUV is an isolated finding. While these cases carry an excellent prognosis, PRUV can be associated with severe congenital anomalies, so this finding should prompt detailed prenatal assessment of the fetus. Copyright © 2011 ISUOG. Published by John Wiley & Sons, Ltd.

INTRODUCTION

Prenatal diagnosis of anomalies of the umbilical veins is usually straightforward and may involve supernumerary vessels, abnormal course, size and insertion and persistence of embryological vascular structures. The development and widespread use of color Doppler interrogation as well as the advent of three-dimensional ultrasound applications has facilitated early in-utero diagnosis of abnormalities of the fetal umbilicoportal circulation as described here.

Persistence of the right umbilical vein (PRUV) as an uncommon antenatal finding reflects an altered embryonic vascular development in which the left umbilical vein regresses and the right-sided vein persists in remaining open. PRUV may be an isolated anomaly or may be part of more complex anomalies of the viscera and venous system, particularly in fetuses with situs inversus visceralis, situs ambiguous and heterotaxy (left and right isomerism)1–5. PRUV is variably associated with other congenital abnormalities such as genitourinary, gastrointestinal, cardiac and skeletal disorders, but to a lesser extent than previously thought1,2.

The objective of the present study was to investigate the antenatal course, associated anomalies and clinical impact of PRUV not associated with situs abnormalities diagnosed in fetal life. We reviewed the perinatal databases of three tertiary referral centers for prenatal medicine in order to tabulate antenatal and perinatal characteristics of fetuses with PRUV and to discuss our findings together with what is known from the current literature.

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with extrahepatic course had additional anomalies and a worsened prognosis.

**DISCUSSION**

Embryologically, the right branch of the initially paired umbilical veins (UV) begins to become obliterated within the fourth week of gestation and tends to completely disappear by the seventh week. During the sixth week, a critical anastomosis occurs between the left UV and the hepatic sinusoids, while the cranial portion of the UV is programmed to regress, thereby losing the direct communication with the sinus venosus. Simultaneously, the liver bud progressively extends and incorporates the cranial part of the vitelline veins, which are involved in the formation of the portal venous branches and the ductus venosus. In certain cases the anastomosis between the UV and the vitelline vein fails to develop and subsequent degeneration of both UVs occurs. As a result, oxygenated venous return from the placenta is blocked and redirected via aberrant vessels towards systemic veins bypassing the liver (iliac vein, inferior vena cava or direct communication with the right atrium). Partial failure to form critical anastomosis seems to be more common than complete primary failure. Based on these findings, two major paths of blood flow towards the heart have been established: the most common of which is PRUV, which is characterized by failure of the right-sided UV to form an anastomosis with the right vitelline vein and degeneration of the left UV. The second, less frequent path involves direct drainage of the UV into the right atrium. Therefore, cases of proven PRUV are usually subdivided into two groups: the intrahepatic type (isolated right UV joining the portal system at the level of the sinus venosus, giving rise to the ductus venosus) and the extrahepatic type, in which the PRUV bypasses the liver completely (absent ductus venosus). In addition, a third variant has occasionally been described as a supernumerary intracorporeal (intrahepatic) or extracorporeal vascular structure (four-vessel umbilical cord) as a result of patency of both UVs.

The exact underlying mechanism causing a PRUV remains elusive, although several pathophysiologic explanations have recently been suggested. Jeanty proposed that the reason for patency of the right-sided vessel might be related to streaming of the early flow traversing the UV. Early obstruction of the left UV from external pressure or occlusion by thromboembolic events arising from the placenta may also result in the appearance of PRUV. In an experimental approach teratogenic agents such as retinoic acid and folate deficiency were found to induce similar developmental alterations as described in primary failure to form an anastomosis, resulting in a patent right UV.

Our study documents the incidence and spectrum of additional anomalies of a large cohort of fetuses with PRUV evaluated at three tertiary referral centers for prenatal medicine. Although several prior publications have reported on fetal PRUV, detailed information regarding its true frequency, impact of associated pathologies and clinical outcome data were derived from only a few larger series. The incidence of PRUV in our series was lower than those reported by Wolman et al. (1 : 1228 compared with 1 : 526 and 1 : 719, respectively). However, the exact incidence of PRUV might be somewhat higher, due to the fact that an aberrant UV may be easily overlooked during standard sonography or in transverse planes other than the appropriate section for measuring the abdominal circumference. On the other hand, the cases with fetal abnormalities are certainly over-represented because of the referral nature of our study population. This may be the case in other series as well.

As indicated by our findings, extrahepatic drainage of the UV may have deleterious consequences for the fetus. Unrestricted umbilical flow entering systemic veins may result in congestive heart failure with subsequent hydrops and fetal demise due to increased cardiac preload. The occurrence of concomitant atrial septal defects in cases with an abnormal UV might be attributed to an increase...
in flow via the inferior vena cava, thereby interfering with developmental processes during atrial septation. However, 77.2% (233/302) of all documented cases of PRUV showed a favorable postnatal outcome.

Differential diagnoses of PRUV include umbilical vein varix, gallbladder duplication, abnormal course of the portal vein and its branches, and intrahepatic cysts. There are no consistent data in the literature to indicate whether PRUV is associated with heterotaxy syndromes or whether the right-sided vessel constitutes a 'normal' UV as part of the visceral rearrangement. In our series, fetuses with situs anomalies and heterotaxy were excluded from the analysis. Some authors, however, included these cases of PRUV in their series. It has been noticed, ascertaining the spatial orientation is an important step for confirming the diagnosis and differentiating among subtypes in order to predict the most frequent associated anomalies. In agreement with previous findings, the most frequent associated anomalies were cardiovascular (60.3%), gastrointestinal (12.8%), renal (15.4%), cerebral (15.4%) and limb abnormalities (7.7%).

Recently, a number of studies have examined the exact morphological and functional relationship between the umbilical and portal venous systems using novel three- and four-dimensional ultrasound techniques in addition to gray-scale and color Doppler ultrasoundography to enhance our understanding of the complex vascular network within the fetal liver. Once a vascular anomaly has been noticed, ascertainment of the spatial orientation is an important subsequent step for confirming the diagnosis and differentiating among subtypes in order to predict potential complications or plan surgical interventions postpartum. Achiron et al. proposed a classification system of the spectrum of abnormalities of the fetal central veins (cardinal veins, pulmonary venous connection) and umbilicoporal veins (umbilical, vitelline veins) based on embryologic precursor and etiologic considerations. However, conclusive information regarding clinical correlations is difficult to derive due to the paucity of reliable data on venous anomalies in the fetus, and clearly depends on the affected vascular connection.

In conclusion, vascular abnormalities within the fetal umbilicoporal venous system are relatively common prenatal findings. The correct antenatal diagnosis of PRUV as well as its intra- and extrahepatic connections is of particular importance owing to its potential association with additional malformations. Nevertheless, the overall prognosis for fetuses with isolated PRUV and proper connection to the portal system is excellent.

REFERENCES


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SUPPORTING INFORMATION ON THE INTERNET

The following supporting information may be found in the online version of this article:

Table S1 Clinical and outcome data of published series of PRUV.
Persistent right umbilical vein: incidence and significance


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KEYWORDS: Incidence, Outcome, Persistent right umbilical vein

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. 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, of which only a few cases had been reported in the literature. 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. Furthermore, it has been suggested that the good outcome of babies with isolated PRUV indicates that this anomaly has only minor prognostic significance. 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.

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...
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 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.

ACKNOWLEDGMENT

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Persistent Intrahepatic Right Umbilical Vein in the Fetus: A Benign Anatomic Variant

SHRAGA BLAZER, MD, ETAN Z. ZIMMER, MD, AND MOSHE BRONSHTEIN, MD

Objective: To evaluate outcomes of fetuses with antepartum sonographic diagnoses of persistent intrahepatic right umbilical veins.

Methods: A detailed fetal sonographic examination was done in 30,240 consecutive pregnancies at 14–26 weeks’ gestation. High- and low-risk pregnancies were included and persistent right umbilical veins specifically were recorded.

Results: Sixty-nine fetuses had persistent intrahepatic right umbilical veins, of which 60 had no additional sonographic abnormalities, four had transient nuchal findings, and four had minor anomalies or anatomic variants. Only one of the 69 fetuses had a major anomaly (diaphragmatic hernia), and died after surgery. The remaining 68 fetuses were normal and healthy after birth.

Conclusion: Persistent intrahepatic right umbilical vein is a fetal anatomic variant that is not rare and usually associated with a favorable outcome. (Obstet Gynecol 2000;95:433–6. © 2000 by The American College of Obstetricians and Gynecologists.)
venous forms in the liver and connects the left umbilical vein and the inferior vena cava. The ductus enables blood to bypass the liver and flow from the placenta to the heart. Postnatally, the left umbilical vein becomes the ligamentum teres.

Alterations of normal anatomy result from failure of the right umbilical vein to become obliterated and involute. A persisting patent right umbilical vein might coexist with the left umbilical vein as an intrahepatic supernumerary structure, or it might connect to the right portal vein. The intrahepatic right umbilical vein also might completely replace the left umbilical vein, which then regresses. The right umbilical vein might further persist and bypass the liver, causing an aberrant drainage of blood into the inferior vena cava or right atrium. The precise causes of failure of normal regression of the right umbilical vein are unknown, but several potential etiologies have been proposed. Obstruction of the left umbilical artery by thrombus, embolus, or external pressure early in pregnancy might cause the right umbilical vein to remain patent to maintain placental blood supply to the fetus. Specific teratogens and maternal folic acid deficiency in the first trimester also have been considered to increase risk of persistent right umbilical vein in a rat model.

Alteration in umbilical venous anatomy was hitherto considered rare, particularly persistent right umbilical vein. Earlier reports were based on incidental pathologic findings from autopsies, detection during surgery, or upon insertion of umbilical vein catheters. Presently, prenatal sonographic detection is the main method of diagnosing persistent right umbilical vein. That widespread technique enables us to determine the true frequency of the condition, and shows that it is more frequent than reported. The incidence of 1:438 in our series was similar to the incidence of 1:476 reported by Hill et al, or 1:450 reported by Shen et al, and was lower than the rate of 1:250 reported by others. The true incidence of persistent right umbilical vein might be even higher, considering the false-negative rate of sonographic detection. The intrahepatic form of persistent right umbilical vein might present as an isolated

### Table 2. Reported Series of Persistent Right Umbilical Vein

<table>
<thead>
<tr>
<th>Study</th>
<th>No. of cases</th>
<th>Incidence</th>
<th>Type</th>
<th>Severe anomalies (%)</th>
<th>Associated congenital malformations</th>
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<tbody>
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<td>Case reports</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>Shryock et al13</td>
<td>1</td>
<td>EH</td>
<td></td>
<td>Situs inversus, sirenomelia, SUA, ectopic kidney, bilateral hydronephrosis</td>
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<tr>
<td>Leonidas and Fellows2</td>
<td>1</td>
<td>EH</td>
<td></td>
<td>Noonan syndrome, SUA</td>
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<tr>
<td>Theander and Karlsson3</td>
<td>1</td>
<td>EH</td>
<td></td>
<td>Right-sided cardiac apex, SUA, bowel malrotation, left-sided liver</td>
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<tr>
<td>Fliegel and Nars4</td>
<td>1</td>
<td>EH</td>
<td></td>
<td>Hydrocephalus</td>
<td></td>
</tr>
<tr>
<td>Bell et al7</td>
<td>1</td>
<td>EH</td>
<td></td>
<td>TAPVR, unilateral renal agenesis, phocomelia, SUA</td>
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</tr>
<tr>
<td>Ricklan et al6</td>
<td>1</td>
<td>IH</td>
<td></td>
<td>PDA</td>
<td></td>
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<tr>
<td>Artiuku et al14</td>
<td>1</td>
<td>IH</td>
<td></td>
<td>Healthy</td>
<td></td>
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<tr>
<td>Moore et al15</td>
<td>1</td>
<td>IH</td>
<td></td>
<td>Lateral ventricular enlargement, mild dilatation of third ventricle</td>
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<tr>
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<td>ASD, dilated IVC, SUA</td>
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<td>De Catte et al12</td>
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<td>EH</td>
<td></td>
<td>Trisomy 18</td>
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<td>Lal17</td>
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<td></td>
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<td>IH</td>
<td>50</td>
<td>Healthy</td>
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<tr>
<td>Jeanty1</td>
<td>6</td>
<td>IH</td>
<td></td>
<td>Normal (3); SUA, atrial septum aneurysm, goiter (mother treated for hyperthyroidism) (1); thickened nuchal fold, ascites, hypospadias, allantoic duct remnant (1); mitral atresia, DORV, COA (1)</td>
<td></td>
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<tr>
<td>Kirsch et al7</td>
<td>9</td>
<td>1:250</td>
<td>IH</td>
<td>11</td>
<td>Normal (8); Hypospadias (1)</td>
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<td>Hill et al8</td>
<td>33</td>
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<td>IH</td>
<td>18.2</td>
<td>Normal (27); dysplastic kidney, dysmorphic kidneys (1); multicystic kidney, VSD, hemivertebrae (1); caudal regression, absent fibula, club foot, mild aortic stenosis, SUA (1); truncus arteriosus, clubfoot, cleft lip, SUA, severe IUGR (1); asplenia syndrome (1); anencephaly (1)</td>
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<td>Shen et al9</td>
<td>8</td>
<td>1:450</td>
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<td>13</td>
<td>Normal (7); dextrocardia (1)</td>
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<td>IH</td>
<td>1.4</td>
<td>Data presented in Table 1</td>
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</table>

SUA, single umbilical artery; EH, extrahepatic; TAPVR, total anomalous pulmonary venous return; IH, intrahepatic; PDA, patent ductus arteriosus; ASD, atrial septal defect; IVC, inferior vena cava; DORV, double outlet right ventricle; COA, coarctation of the aorta; VSD, ventricular septal defect; IUGR, intrauterine growth restriction.

Figures in parentheses represent number of cases.
finding, with no additional anomalies, whereas all the cases of persistent extrahepatic right umbilical vein were associated with significant fetal abnormalities (Table 2).7

Our sonographic findings indicated severe malformations in 1.4% of cases with persistent intrahepatic right umbilical veins, compared with up to 50% of cases in the literature (Table 2). Our study was not a population-based study; therefore, bias was possible in incidence of associated anomalies. However, all our cases were detected in low-risk pregnancies. Despite the suggested association of persistent right umbilical veins with increased frequency of severe malformations and “transient” fetal anomalies (such as increased nuchal translucency), only one report of chromosomal abnormality could be found.12 Further studies are needed to clarify the relationship of persistent right umbilical vein and chromosomal defects, or to deem it purely coincidental. Karyotype analysis still might be indicated, especially when persistent right umbilical vein is accompanied by other sonographic anomalies. Reported cases of persistent right umbilical vein show a variety of cardiovascular, gastrointestinal, urinary, musculoskeletal, and central nervous system malformations. A summary of the literature (Table 2) showed that single umbilical artery was the most consistent finding. Two reported cases3,13 had situs inversus, as in three of our fetuses. It is unclear whether those cases represent genuine persistent right umbilical vein, or feature a normal “left” umbilical vein on the right side of the fetus, due to organ inversion. All those fetuses had malformations. Lai17 reported two cases referred for persistent right umbilical vein on obstetric ultrasonography examination. Careful fetal echocardiogram and Doppler examination showed an abnormal left umbilical vein in the setting of visceral situs inversus, with an interrupted inferior vena cava andazygous continuation.

Our data and those from other series indicate that persistent intrahepatic right umbilical vein is frequent. An extensive targeted ultrasonographic examination should be done whenever the condition is identified, and chromosomal studies are warranted in every fetus in which ultrasonographic abnormalities are detected. In the absence of associated malformations, a persistent intrahepatic right umbilical vein is usually an innocuous condition and carries a good prognosis.

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Persistent right umbilical vein: an ominous prenatal finding?

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Abstract

The persistence of a right umbilical vein is an uncommon finding, with only a dozen cases reported since 1826. The persistent right umbilical vein may replace the normal left umbilical vein or be supernumerary. The anomaly is associated with numerous and occasionally lethal malformations. In this series, only three of six fetuses (and another two in the literature) had no associated anomalies. All the others had a variety of associated lesions ranging from minor to lethal. The appearance at ultrasound is easy to recognize: The intrahepatic portion of the umbilical vein is lateral to the gallbladder, and the portal vein curves toward the stomach, instead of parallel to it. Since the recognition of the persistent right umbilical vein is simple and does not require additional scanning (it is visible in the section used to measure the abdominal perimeter), the author suggests using it as an indicator for more in-depth scanning.