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Original Article

Persistent Right Umbilical Vein: Clinical Outcomes and Prognostic Factors in Prenatal Diagnosis

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ABSTRACT

Objective: To evaluate clinical outcomes and associated anomalies in fetuses diagnosed with persistent right umbilical vein (PRUV) during routine prenatal ultrasound at a tertiary perinatology clinic.

Methods: This retrospective study included 11 cases of PRUV diagnosed between October 2022 and January 2024. Data were collected on maternal demographics, gestational age at diagnosis, associated anomalies, and neonatal outcomes. Ultrasound examinations were performed using B-mode and color Doppler, with fetal echocardiography to assess cardiac abnormalities. Cases were classified as isolated PRUV or PRUV with associated anomalies.

Results: PRUV was detected in 11 out of 10,176 pregnancies (0.1%). Seven cases were isolated PRUV, while four cases had associated anomalies, including cardiovascular and genitourinary defects. One case with extrahepatic PRUV and severe cardiovascular abnormalities was discontinued. The remaining 10 cases, including those with isolated PRUV, resulted in healthy live births. Six births were by cesarean section, and four were spontaneous deliveries. The presence of additional malformations was associated with more complex prenatal management and a poorer prognosis.

Conclusion: Isolated PRUV is usually associated with favorable outcomes, but the presence of additional anomalies, particularly cardiovascular defects, has a significant impact on management and prognosis. Comprehensive prenatal imaging, including echocardiography, is essential in PRUV cases to inform clinical decisions. Larger studies are needed to further elucidate the long-term outcomes of PRUV.

INTRODUCTION

A rare vascular anomaly that occurs when the left umbilical vein regresses during embryonic development and the right umbilical vein remains open is called Persistent Right Umbilical Vein (PRUV).[1] Normally, the left umbilical vein does not close and carries oxygenated blood to the fetus, but in PRUV, the left umbilical vein closes and the right umbilical vein continues to function. This condition is usually detected during routine prenatal ultrasound examinations in the second trimester.[2]

The incidence of PRUV varies across studies but is generally estimated to be between 1/250 to 1/1250 pregnancies.[3] However, the actual prevalence may be higher as the anomaly can easily be missed on standard ultrasound examinations. Advances in imaging techniques, including color Doppler and 3D ultrasound, have improved the ability to diagnose PRUV.[3]

PRUV is often considered an isolated finding, meaning it occurs in the absence of other fetal anomalies, and in such cases the prognosis is generally favorable.[4] However, PRUV may also be associated with other malformations, in particular cardiac defects and gastrointestinal, genitourinary, and skeletal anomalies.[1] This association emphasizes the importance of a detailed anatomical examination of the fetus and fetal echocardiography as soon as a PRUV

is detected.[1]

The etiology of PRUV remains unclear, although some studies suggest possible factors such as folic acid deficiency or teratogenic exposure.[3] In addition, some researchers suggest that thrombosis or external pressure on the left umbilical vein could lead to persistence of the right umbilical vein.[3]

In this study, we aim to evaluate the prognostic outcomes of fetuses diagnosed with PRUV, including any associated fetal malformations or chromosomal abnormalities, using a retrospective analysis of cases diagnosed by routine prenatal ultrasound examinations.[3,5]

MATERIALS AND METHODS

This study was conducted to retrospectively evaluate the clinical outcomes of fetuses diagnosed with PRUV during routine prenatal ultrasound examinations. Data were collected from all patients who underwent prenatal ultrasonography between October 2022 and January 2024 at the Perinatology Clinic of our institution.

Study Participants

In this study, we included pregnant women who were diagnosed with PRUV during routine ultrasound examinations in the last two trimesters. Cases with chromosomal abnormalities or other major fetal malformations were excluded.

Ultrasound Evaluation

In all patients, the venous system was examined using a Voluson E8 GE ultrasound machine (GE Medical Systems, Milwaukee, WI, USA) equipped with a convex 4-8 MHz transabdominal transducer. Two-dimensional color Doppler imaging was used to assess the target vessels. Three basic criteria were considered in the diagnosis of PRUV:

1. Abnormal course of the portal vein towards the stomach (Fig. 1),

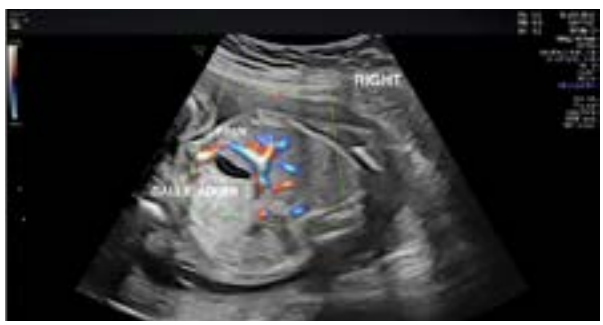

Figure 1. Prenatal ultrasound image showing a persistent right umbilical vein (PRUV) in a transverse section of the fetal abdomen. The PRUV can be seen in the direction of the right portal vein as indicated by the color Doppler flow. The gallbladder can be seen medial to the PRUV. This finding is characteristic of PRUV in which the umbilical vein remains on the right side instead of the usual left side. The use of Doppler imaging helps visualize the abnormal vascular flow pattern and aids in diagnosis.

2. Presence of an umbilical vein on the right side of the gallbladder,

3. Connection of the umbilical vein with the portal veins.

In addition, the medial location of the gallbladder in relation to the umbilical vein served as a further diagnostic marker. To detect possible cardiac anomalies, fetal echocardiography was performed in all cases.

Data Collection

For each case, maternal demographic characteristics (age, gravidity, parity), gestational age at diagnosis, and any associated fetal anomalies were recorded. Cases were classified as either isolated PRUV (without other anomalies) or PRUV with associated anomalies. All cases were followed up until delivery, and neonatal outcomes, including gestational age at delivery, mode of delivery, and postnatal findings, were documented. The results of invasive diagnostic tests for fetal chromosome analysis were recorded, if available. Information about newborns was obtained from the families by telephone.

Ethical Approval

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional research committee at which the studies were conducted (Clinical Research Ethics Committee of Ankara Etlik City Hospital No. 1 [Decision No.: AEŞH-EK-2024-008, date: 10/01/2024]) and with the 2013 Helsinki Declaration and its later amendments or comparable ethical standards.

Statistical Analysis

Descriptive statistics were used to present the data, including the average gestational age at diagnosis and the incidence of associated malformations. The incidence of PRUV was calculated as a percentage of the total number of pregnancies examined. Statistical analyses were performed using SPSS software (version 26.0), and results with a p-value below 0.05 were considered statistically significant.

RESULTS

Among 10,176 pregnancies evaluated at the perinatology clinic, PRUV was detected in 11 cases, which corresponds to an incidence of 0.1%. The general week of diagnosis of these patients was the same as the week of admission to our hospital and ranged from 16 to 33 weeks. One out of 11 patients had a twin pregnancy, while the others had singleton pregnancies (Table 1). One out of 11 patients had an extrahepatic PRUV, while 10 had an intrahepatic PRUV. The ductus venosus was present in all patients except for the extrahepatic PRUV.

Seven of the 11 patients had isolated PRUV, and four had concomitant abnormalities. One patient had cardiovascular abnormalities (extrahepatic PRUV), two had genitourinary system anomalies, and one had a single umbilical

Table 1. Characteristics of the cases, malformations associated with PRUV

Age (years)	GA at diagnosis (weeks)	Additional findings	GA at delivery (weeks)	Birth Weight (gr)	Delivery Type	Gender	Neonatal Outcome	Type of PRUV	Presence of Ductus Venosus	Karyotype
35	16	Isolated	34.6	2500	Caesarean section	Male	Healty	Intrahepatic	Yes	46 XY
23	21.6	Bilateral renal pelvis dilatation	35.0	2540	Caesarean section	Male	Healty	Intrahepatic	Yes	
29	33.1	Isolated (FGR)	38.0	2170	Caesarean section	Male	Healty	Intrahepatic	Yes	
32	20.6	Tetralogy of Fallot (Dextrocardia, Inlet VSD, Pulmonary Artery Atresia)	21.0	425	Terminated	Male	Terminated	Extrahepatic	None	46 XY
26	20	Isolated	35.0	2660	Caesarean section	Male	Healty	Intrahepatic	Yes	
32	25	Isolated	38.0	3050	Spontaneous	Female	Healty	Intrahepatic	Yes	
28	21	Isolated	39.0	3100	Spontaneous	Female	Healty	Intrahepatic	Yes	
34	27	Isolated	37.0	2750	Caesarean section	Male	Healty	Intrahepatic	Yes	
28	22	SUA	39.0	2950	Spontaneous	Female	Healty	Intrahepatic	Yes	
36	20	Bilateral renal pelvis dilatation	38.0	3400	Spontaneous	Female	Healty	Intrahepatic	Yes	46 XX
34	21	Isolated	36.0	2460	Caesarean section	Female-Male	Healty	Intrahepatic	Yes	

Abbreviations: GA; Gestational age; PRUV: Persistent right umbilical vein; FGR: Fetal growth restriction; VSD: Ventricular septal defect; SUA: Single umbilical artery.

artery. None of the patients had teratogenic exposure, and only one patient did not take folic acid in early pregnancy.

Four of the 11 patients were delivered by normal spontaneous delivery (NSD), and six by cesarean section (C/S). One patient with concomitant dextrocardia was also terminated. Six patients delivered at term, while four had a premature delivery. All fetuses were healthy, with the exception of the fetus that was terminated in neonatal follow-up.

DISCUSSION

This study provides valuable insight into the clinical relevance of PRUV, a rare vascular anomaly commonly detected on routine prenatal ultrasound. The incidence of PRUV in our cohort remains low, consistent with previous findings, but its clinical significance cannot be underestimated. PRUV can occur either as an isolated anomaly or together with other congenital abnormalities, which has a significant impact on prognosis and management.

Our findings are consistent with previous literature on the incidence of PRUV, which ranges from 0.08% to 0.5% of pregnancies.[6-8] In our study, the majority of PRUV cases were isolated, and the prognosis in such cases tends to be favorable, as also found in other studies. When PRUV is isolated, it is often considered a benign variant of normal vascular anatomy.[8,9] However, when it is associated with other anomalies, particularly cardiovascular malformations, the prognosis becomes more complex.[3,9] While our findings align with previous studies, the incidence of abnormalities associated with intrahepatic PRUV in our study is higher (36%) than in other studies.[6,8,10] This result may be related to our being a referral center.

PRUV was defined by Jeanty et al.[11] in two types: intrahepatic and extrahepatic. If the umbilical vein is directly connected to the inferior vena cava and the right atrium, it is called an extrahepatic PRUV. In this type, there is usually no ductus venosus, and the prognosis is generally poor.[8,12] The most common type of PRUV is the intrahepatic type (95%), and there is usually a ductus venosus.[12,13] In agreement with the literature, in our study only one of 11 cases had an extrahepatic PRUV that did not have a ductus venosus. This case was terminated because of severe cardiovascular abnormalities that had a poor prognosis, which is consistent with the studies.

The presence of a single umbilical artery (SUA) alongside PRUV further complicates fetal outcomes. As Mohapatra et al.[10] point out, the co-occurrence of SUA and PRUV increases the likelihood of additional malformations, particularly in the cardiovascular system. These findings emphasize the need for detailed fetal echocardiography in all PRUV cases to detect possible cardiovascular abnormali-

ties.[5,9] This correlation was not observed in our findings. Among the 11 cases, only one involved SUA, and no other congenital abnormalities were identified. A larger sample size may be needed to align our results with those reported in the literature.

The most common hypotheses for the etiology of PRUV are early thrombosis of the left umbilical vein, teratogenic exposure, and folic acid deficiency in the first trimester.[5,6] Interestingly, most of our patients had no history of teratogenic exposure, and all but one had taken folic acid in the first trimester. This supports previous research suggesting that folic acid deficiency does not play a major role in the development of PRUV in all cases.[5,8]

The management of PRUV should be based on the presence or absence of additional anomalies. Isolated cases of PRUV generally do not require invasive procedures as they are associated with a good prognosis.[6,7,9] However, when PRUV is detected together with other malformations, especially in the cardiovascular or urinary systems, further genetic testing and detailed anatomical scans are indicated to better predict the outcome of the newborn.[5,10]

Conclusion

In summary, although isolated PRUV is usually a benign finding, the presence of associated anomalies significantly alters the management and prognosis of affected pregnancies. Comprehensive prenatal imaging, including detailed ultrasonography and echocardiography, is essential to accurately diagnose PRUV and determine the best course of action for each individual case. Further studies with larger cohorts are needed to better understand the pathophysiology of PRUV and its long-term impact on neonatal health.

Ethics Committee Approval

The study was approved by the Ankara Etlik City Hospital Ethics Committee (Date: 10.01.2024, Decision No: AEŞH-EK-2024-008).

Informed Consent

Retrospective study.

Peer-review

Externally peer-reviewed.

Authorship Contributions

Concept: S.Ö., A.A., F.B.F., M.L.D., Ş.Ç.; Design: S.Ö., A.A., F.B.F., M.L.D., Ş.Ç.; Supervision: S.Ö., A.A., F.B.F., M.L.D., Ş.Ç.; Data collection &/or processing: F.B.F.; Analysis and/or interpretation: M.L.D.; Literature search: A.A.; Writing: S.Ö., A.A.; Critical review: S.Ö., Ş.Ç.

Conflict of Interest

None declared.

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Persistan Sağ Umbilikal Ven: Prenatal Tanıda Klinik Sonuçlar ve Prognostik Faktörler

Amaç: Bu çalışmanın amacı, üçüncü basamak bir perinatoloji kliniğinde yapılan rutin prenatal ultrason sırasında persistan sağ umbilikal ven (PSUV) tanısı konan fetüslerde klinik sonuçları ve ilişkili anomalileri değerlendirmektir.

Gereç ve Yöntem: Bu retrospektif çalışma, Ekim 2022 ile Ocak 2024 tarihleri arasında PSUV tanısı konan 11 vakayı içermektedir. Anne demografik verileri, tanı anındaki gestasyonel yaş, ilişkili anomaliler ve neonatal sonuçlarla ilgili veriler toplanmıştır. Kardiyak anomalileri değerlendirmek amacıyla B-mod ve renkli Doppler ile ultrason muayeneleri yapılmış ve fetal ekokardiyografi ile desteklenmiştir. Vakalar, izole PSUV ya da ilişkili anomalili PSUV olarak sınıflandırılmıştır.

Bulgular: PSUV, 10.176 gebeliğin 11'inde (%0.1) tespit edilmiştir. Yedi vaka izole PSUV iken, dört vakada kardiyovasküler ve genitoüriner defektler dahil olmak üzere ek anomaliler görülmüştür. Ekstrahepatik PSUV ve ciddi kardiyovasküler anomalilere sahip bir vaka sonlandırılmıştır. İzole PSUV vakaları dahil olmak üzere kalan 10 vaka sağlıklı canlı doğumlarla sonuçlanmıştır. Altı doğum sezaryen ile, dört doğum ise spontan gerçekleşmiştir. Ek malformasyonların varlığı, daha karmaşık doğum öncesi yönetim ve daha kötü bir prognoz ile ilişkilendirilmiştir.

Sonuç: İzole PSUV genellikle olumlu sonuçlarla ilişkilendirilirken, özellikle kardiyovasküler defektler gibi ek anomalilerin varlığı, yönetim ve prognoz üzerinde önemli bir etkiye sahiptir. PSUV vakalarında kapsamlı prenatal görüntüleme, ekokardiyografi dahil olmak üzere, klinik kararları yönlendirmek için hayati öneme sahiptir. PSUV'nun uzun vadeli sonuçlarını daha iyi anlamak için daha geniş kapsamlı çalışmalara ihtiyaç vardır.

Anahtar Sözcükler: Fetal anomaliler; perinatal sonuçlar; persistan sağ umbilikal ven; prenatal ultrason.