ORİJİNAL ARAŞTIRMA ORIGINAL RESEARCH DOI: 10.24074/tjrms.2019-71585

Persistent Right Intrahepatic Umbilical Vein: Significance and Prognosis

Persiste Sağ İntrahepatik Umblikal Ven: Tanısı ve Önemi

Emine Ahu KOÇ[a], Erhan H. CÖMERT[a], Hidayet ŞAL[a], Turhan ARAN[a], Yasin S. EKİCİ[a], Gülseren DİNÇ[a], Cavit KART[a], Mehmet A. OSMANAĞAOĞLU[a]

[a]Karadeniz Technical University Faculty of Medicine, Department of Obstetrics and Gynecology, Trabzon, TURKEY

ABSTRACT

Objective: To investigate the incidence and clinical impact of prenatally diagnosed persistent right umblical vein in a referral population and to evaluate associated anomalies, neonatal prognosis. **Material and Methods:** All pregnants were taken to this single center retrospective study who admitted to the clinic with the diagnosis of persistent right umbilical vein between 2011 and 2018 in Karadeniz Technical University, Faculty of Medicine, Department of Obstetrics and Gynecology. The prenatal sonograms and neonatal outcome data of affected individuals were reviewed. **Results:** Persistent right umblical vein was detected in 7 fetuses during the study. All cases used regular folic acid from the beginning of pregnancy and the story of teratogenic drug use did not exist at all. Three of the 7 cases were pregnant with ivf therapy. Only one case was twin pregnancy. There were one umblical artery in two cases and meconium ileus with oligohidroamnios in one case. Persistent right intrahepatic umblical vein was isolated anomaly in the remaining 5 cases. All cases were evaluated by fetal echocardiography and no abnormality was detected. The median age of the cases was 32.8 (27-40), median gestational age at the time of diagnosis 21.7 (20-23) and the median gestational age at birth 37.7 (33-40). Five cases were nulliparous and two cases were multiparous. 2 cases terminated by preterm delivery. The causes of preterm deliveries were twin pregnancy and late onset intrauterin growth restriction. **Conclusion:** In summary, a persistent right umbilical vein is commonly an isolated finding but may be associated with other congenital anomalies. Therefore,consideration should be given to fetal echocardiography and detailed ultrasonography in cases of a persistent right umblical vein. Fetal caryotyping is not recommended in isolated persistent right umblical vein cases.

Keywords: Persistent right umbilical vein; prenatal ultrasound; fetal venous system anomaly; fetal anomaly

ÖZET

Amaç: Bu çalışmada kliniğimizde prenatal persiste sağ umbilikal ven tanısı alan olguların insidansı, eşlik eden diğer anomalilerin tespiti, gebeliğin prognozu ve doğum sonrası yenidoğanda gözlenen özellikler gibi postnatal sonuçların değerlendirilmesi amaçlandı. **Gereç ve Yöntemler:** Karadeniz Teknik Üniversitesi Tıp Fakültesi, Kadın Hastalıkları ve Doğum AD'da 2011-2018 tarihleri arasında persiste sağ umbilikal ven tanısı konan tüm gebeler bu tek merkezli retrospektif çalışmaya alındı. Hastalara ait verilere tıbbi kayıtlardan ulaşıldı. **Bulgular:** Çalışmaya toplam 7 persiste sağ intrahepatik umblikal ven tanısı alan olgu dahil edildi. Tüm olgular gebeliğin başından itibaren düzenli folik asit kullanan olgulardı. Teratojen ilaç kullanım öyküsü hiçbir vakada mevcut değil idi.Toplam 7 vakanın 3'ü in vitro tedavisi ile gebe kalan olgulardı. Sadece 1 vaka ikiz gebelik idi.2 vakada tek umblikal arter ve bir vakada mekonyum ileusu ve oligohidroamniyoz eşlik eden diğer anomalilerdi. Kalan 5 vakada persiste sağ intrahepatik umblikal ven izole anomali idi. Tüm vakalar fetal ekokardiyografi ile değerlendirildi ve herhangi bir anomali tespit edilmedi. Olguların ortalama yaşı 32.8 (27-40), tanı anında ortalama gebelik haftası 21.7 (20-23) ve doğum anında ortalama gebelik haftası 37.7 (33-40) idi. 5 vaka nullipar, 2 vaka multipar idi. 2 vaka preterm doğumla sonlandı. Biri ikiz gebelik, diğeri geç başlangıçlı intrauterin gelişme geriliği nedeni erken doğurtuldu. **Sonuç:** Yapılan çalışmalardan ve kliniğimizdeki olgulardan da anlaşılacağı gibi, persiste sağ intrahepatik umblikal ven izole bir bulgudur. Bu olgular iyi prognoza sahip olsalar da, eşlik eden çok sayıda konjenital anomali olabilir. Bu nedenle saptandığında fetal ekokardiyografi ve hedeflenmiş ultrasonografi ile araştırılmayı gerektirir. Ancak fetal karyotipleme açısından tek başına persiste sağ intrahepatik umblikal ven endikasyon oluşturmaz.

Anahtar Kelimeler: Persiste sağ umblikal ven; prenatal ultrasonografi; fetal venöz sistem anomalisi; fetal anomali

Correspondence: Emine Ahu KOÇ
Karadeniz Technical University Faculty of Medicine, Department of Gynecology and Obstetrics, Trabzon, TURKEY/TÜRKİYE
E-mail: emineahu52dr@hotmail.com

Peer review under responsibility of Turkish Journal of Reproductive Medicine and Surgery.

Received: 24 Sep 2019 ***Received in revised form:*** 06 Nov 2019 ***Accepted:*** 14 Jan 2020 ***Available online*:** 08 Jun 2020

Persistent right umbilical vein (PRUV) is a vascular anomaly in which the left umbilical vein is regressed and the right umbilical vein remains open.[1] It is the most common prenatally detected venous anomaly and the estimated incidence varies between 1/250 and 1/1250.[2,3] Although the etiology of PRUV is unknown, proposed mechanisms are the first trimester folic acid deficiency, exposure to teratogens such as retinoic acid, and early narrowing or obstruction of the left umbilical vein due to external pressure or placental thromboembolic events.[4,5] Persistent right umbilical vein may be an isolated anomaly or associated with congenital heart diseases, gastrointestinal, urinary and musculoskeletal malformations.[4] Single umbilical artery is the most common encountered associated anomaly.[6]

In our study, we aimed to present the anomalies associated with persistent right umbilical vein cases, the clinical importance, incidence and neonatal outcomes of the fetuses.

MATERIAL AND METHODS

This retrospective study included the cases of persistent umbilical vein which was diagnosed prenatally in our department, between 2011-2018. During the study period the anatomical evaluation and ultrasound measurements were performed in a standardized manner including color and spectral Doppler imaging of the fetal umbilical venous system in axial and sagittal sections. All ultrasound examinations were performed with a Voluson Expert machine (General Electric Healthcare Systems, Austria).

Ultrasonographic criteria for diagnosis of persistent right umbilical vein included: 1- Curvature of the portal vein towards the stomach rather than the right lobe of the liver, 2- Location of the fetal gallbladder is in the medial of umbilical vein, 3- The abnormal connection of the umbilical vein to the right, as opposed to the left portal vein.

Echocardiographic examination was performed in all cases. Fetal karyotyping was performed according to the obstetric indications including maternal age and abnormal screening test results or their combination with the presence of ultrasound findings. All patients were followed up to early childhood.

RESULTS

7 fetuses with PRUV were detected in 10.600 pregnant women (incidence %0.06 in our clinic). Persistent right umbilical vein was classified as intrahepatic type in all cases. The mean gestational week was 21,7 at diagnosis (the lowest 20, the highest 23). Amniocentesis was performed in 2 of 7 cases due to additional problems such as high risc of first trimester screening and maternal age risc. The results of amniocentesis were normal. The PSUV was the only finding in 4 of 7 cases. Single umbilical artery was accompanied with PSUV in 2 cases. Fetuses with PRUV, except for 2 cases, were born at term (Table 1). 5 of the fetuses were male, and others were female.

The fetal echocardiography was performed in all cases, no cardiac abnormality was detected. There were no complications in the neonatal period. None of the cases developed cardiac, gastrointestinal and musculoskeletal anomalies at the time of infancy. No disease developed during early childhood.

DISCUSSION

The intrahepatic form of PRUV occurs as an isolated anomaly according to described previous literature. The incidence of severe associated anomalies is very rare. In our study, the ratio of isolated persistent right umbilical vein was 57%. In addition, associated serious anomaly was not detected.

TABLE 1: General features of cases.

Number	Maternal Age	Gestation Week	Associated Anomalies	Amniosentesis	Gestational week at birth	Neonatal Birth-weight (g)	Result Neonaltly
1	35	20	Single umbilical artery	+	39	4150	Healty
2	33	21	None	-	40	3520	Healty
3	33	22	None	-	35	2550	Healty
4	30	21	Single umbilical artery	-	39	3200	Healty
5	32	23	None	+	33	1490	Healty
6	27	23	None	-	39	2700	Healty
7	40	22	None	-	39	3100	Healty

Umbilical veins are two separate structures at the beginning of normal embryonic development. The right umbilical vein begins to become obliterated around the fourth week of gestation, and disappears by the seventh week.[7] Persistence of the right umbilical vein reflects an abnormal embryonic vascular development in which the left umbilical vein obliterates and the right vein persists in remaining open. The cases of PRUV are usually subdivided into two groups; the intrahepatic type which is at the level of the sinus venosus isolated right umbilical vein joins with the portal system (such as in our cases) and the extrahepatic type which is PRUV bypasses the liver completely and drains into inferior vena cava or to the right atrium.[3,8] The distinction of these subtypes is important in assessing the risk of concomitant fetal anomalies. Extrahepatic variant is associated with severe cardiovascular and gastrointestinal anomalies in contrast to intrahepatic isolated type.[8] In our study, all cases had intrahepatic persistent right umbilical vein.

Martinez et al. evaluated 240 cases, in their study, PRUV-intrahepatik were isolated anomalies (76.3%), but the rest were accompanied by heart abnormalities such as atrioventricular canal with aortic stenosis and interrupted aortic arch, hypoplastic left heart syndrome,cardiovascular malformation with an unbalanced atrioventricular canal defect and right ventricle truncus arteriosus (7.9%), placental or umbilical cord anomalies (7%), genitourinary malformations (6.3%) or central nervous system malformations (3.8%). Genetic disorders were diagnosed in 1.3% of fetuses. A review of PRUV in the literature, Weichert et al. were reported cardiovascular anomalies in 60.3% cases.[3,9] The fetal echocardiography was performed to all PRUV cases in our clinic, in the light of this information. We suggest fetal echocardiography in terms of cardiac anomalies which may be possible on PRUV cases.

We could not determine the cause of persistent right umbilical vein in our study group. All cases were using regular folic acid and had no history of teratogen exposure. There were no cases of echogenic focus in the liver parenchyma suggesting a placental thromboembolic event. Chromosomal abnormality was not present in any case. There is no indication for karyotyping in isolated persistent right umbilical vein cases, but may be required in case of accompanying abnormality.[10,11] In our data, the result of 2 amniocentesis, as a result of other obstetric indications, was determined as normal karyotyping. We do not recommended routine genetic screening in cases with isolated PRUVs if there is no additional anomaly. The most common associated congenital anomaly in the persistent right umbilical vein cases is the single umbilical artery. We detected 2 cases with single umbilical artery. Neonates were evaluated carefully in terms of respiratory and nutritional problems. It was confirmed that there were no undetectable anomalies.

Current data shows that the intrahepatic type of PRUV occurs as an isolated anomaly more commonly. Our data also indicate that this type represents isolated finding and results in good prognosis. Nevertheless, it is still necessary to perform targeted fetal sonography and fetal echocardiography following prenatal sonographic diagnosis of an intrahepatic PRUV due to larger series of PRUV aren't available yet. Althougt PRUV is seen isolated, additional anomalies that may accompany the disease should be kept in mind. The fetal echocardiography should be considered. Genetic evaluation may be required according to the accompanying findings.

REFERENCES

1. Perez-Cosio C, Sheiner E, Abramowicz JS. Four-vessel umbilical cord: not always a dire prognosis. Journal of Ultrasound in Medicine. 2008;27:1389-91. [Crossref] [PubMed]
2. Hill LM, Mills A, Peterson C, Boyles D. Persistent right umbilical vein: sonographic detection and subsequent neonatal outcome. Obstetrics and Gynecology. 1994;84:923-5.
3. Weichert J, Hartge D, Germer U, Axt-Fliedner R, Gembruch U. Persistent right umbilical vein: a prenatal condition worth mentioning? Ultrasound in Obstetrics and Gynecology. 2010;37:543-8. [Crossref] [PubMed]
4. Jeanty P. Persistent right umbilical vein: an ominous prenatal finding? Radiology. 1990;177:735-8. [Crossref] [PubMed]
5. Monie IW, Nelson MM, Evans HM. Persistent right umbilical vein as a result of vitamin deficiency during gestation. Circulation Research. 1957;5:187-90. [Crossref] [PubMed]
6. Bell AD,Gerlis LM, Variend S. Persistent right umbilical vein-case report and review of the literature. Int J Cardiol. 1986;10:167-76. [Crossref]
7. Theander G, Karlsson S. Persistent right umbilical vein. Acta Radiol. 1978;19:268-74. [Crossref] [PubMed]
8. Claudia F. E. Kirsch, MD, Vickie A. Feldstein, MD, Ruth B. Goldstein, MD, Roy A. Filly, MD . Persistent intrahepatic right umbilical vein
9. Martinez R, Gamez F, Bravo C et al. Perinatal outcome after ultrasound prenatal diagnosis of persistent right umbilical vein. Eur J Obstet Gynecol Reprod Biol 2013;168:36-39. [Crossref] [PubMed]
10. Blazer S, Zimmer EZ, Bronshtein M. Persistent intrahepatic right umbilical vein in the fetus: a benign anatomic variant. Obstetrics and Gynecology 2000; 95:433-6 [Crossref]
11. Wolman I, Gull I, Fait G, et al. Persistent right umbilical vein: incidence and significance. Ultrasound in Obstetrics and Gynecology 2002;19:562-4. [Crossref] [PubMed]