PERINATAL JOURNAL

Case Report

Perinatal Journal 2012;20(3):150-152

Agenesis of ductus venosus: a case report

Talat Umut Kutlu Dilek[1], Burcu Dilek[2]

[1]*Department of Obstetrics and Gynaecology, Faculty of Medicine, Mersin University, Mersin, Turkey*
[2]*Radiology Clinic, Private Mersin Yenişehir Hospital, Mersin, Turkey*

Abstract

Objective: Agenesis of ductus venosus is a rare vascular abnormality. We report a of case ductus venosus agenesis, with multiple abnormalities.

Case: Atrioventricular septal defect, right multicystic dysplastic kidney, rocker bottom foot, and single umbilical artery were detected in the anatomic evaluation. It was found that ductus venosus did not exist and umbilical vein was opening directly to right atrium. Karyotype result was reported as normal constitutional karyotype and the pregnancy of the case was terminated due to multiple congenital anomalies.

Conclusion: Coincidental fetal abnormalities determine prognosis of ductal agenesis.

Key words: Ductus venosus, agenesis, extrahepatic drainage.

Ductus venosus agenezisi: Olgu sunumu

Amaç: Ductus venosus agenezisi nadir bir fetal vasküler anomalidir. Bu olgu sunumunda, multiple anomalilerle beraber olan ductus venosus agenezisi olgusunu bildirdik.

Olgu: Yapılan anatomik değerlendirmede atrioventriküler septal defekt, sağ multikistik displastik böbrek ve ayaklarda rocker bottom foot, tek umbilikal arter saptandı. Ductus venosus'un olmadığı umbilikal venin direkt sağ atriyuma açıldığı gözlendi. Karyotip sonucu normal konstitüsyonel karyotip olarak raporlanan olgunun gebeliği multiple konjenital anomaliler nedeniyle sonlandırıldı.

Sonuç: Ductus venosus agenezisi vakalarında, eşlik eden fetal anomalilerin varlığı prognozu belirlemektedir.

Anahtar sözcükler: Ductus venosus, agenezis, ekstrahepatik drenaj.

Introduction

Circulation system in fetal life is different than postnatal circulation by 3 shunts included. These are ductus venosus, foramen ovale and ductus arteriosus. During embryonal development period, while right umbilical vein regresses, an area of left umbilical vein from entrance point into abdomen to inferior vena cava endures and is named as ductus venosus. Ductus venosus is the shunt responsible for carrying blood, which has with high oxygen saturation carried by umbilical vein, into inferior vena cava. Blood transferred to inferior vena cava by this shunt is carried on left dorsal part and reaches right atrium.

The development appeared in ultrasonography technique helped to understand anatomy and anomalies of fetal venous system better.[1] Ductus venosus agenesis is a rare fetal vascular anomaly. Its prevalence is reported as 1/2500 according to screening programs performed between 11 and 14 gestational weeks.[2] In this report, ductus venosus agenesis case accompanying other congenital anomalies found in a pregnant referred due to abnormal second trimester screening test has been discussed.

Case Report

A 27-year-old patient with second pregnancy who had no obstetric and medical risk factor and referred for urinary anomaly suspect was evaluated. In the second trimester screening test of the patient, the risk for neural tube defect was calculated as 1/50 and alpha-feto

Correspondence: Talat Umut Kutlu Dilek, MD. Mersin Üniversitesi Eğitim ve Araştırma Hastanesi, Zeytinlibahçe, Mersin, Turkey.
e-mail: umutdilek@gmail.com

Received: November 13, 2012; **Accepted:** December 11, 2012

Available online at:
www.perinataljournal.com/20120203006
doi:10.2399/prn.12.0203006
QR (Quick Response) Code:

protein MoM was reported as 3.21. In the prenatal evaluation of the patient performed when she was pregnant for 19 weeks and 2 days according to her last menstrual period, atrioventricular septal defect, right multicystic dysplastic kidney, rocker bottom foot deformity on foot, and single umbilical artery were detected in fetus of which biometric measurements were consistent with 18 weeks and 5 days. It was observed that ductus venosus did not exist and umbilical vein was directly opened to right atrium by displaying extrahepatic drainage (**Figs. 1 and 2**). Genetic amniocentesis was done to the patient due to these diagnoses.

Karyotype result was reported as normal constitutional karyotype. Due to the existing multiple congenital anomalies, termination of pregnancy was recommended to the patient. Upon the acceptance of the patient, the pregnancy was terminated at 22 gestational weeks. Postmortem examination was not done since the patient did not accept.

Discussion

Ductus venosus is an anatomic shunt in intrauterine life providing connection between umbilical vein and inferior vena cava. It turns into ligamentum venosum after delivery. In the existence of ductus venosus, umbilical vein may be opened directly into intrahepatic left portal vein, iliac vein, inferior vena cava, and right atrium.[3] It was reported that progressive cardiac decompensation and hydrops fetalis are seen much more in extrahepatic insertions (right atrium, vena cava inferior etc.) of ductus venosus.[3,5]

Shen et al.[6] reported that in ductus venosus agenesis cases where extrahepatic drainage is seen, portal system is developed if shunt diameter is small and portal system is not developed if shunt is has a diameter same with or wider than umbilical vein. Non-existence of ductus venosus may be accompanied with hydrops, cardiac, (ASD, VSD, major vascular transposition, right ventricle with dual outlet, non-existence of inferior vena cava, pulmonary arterial anomalies), renal anomalies (hypoplasia, agenesis), cleft palate, hypospadias, mesomelic karyotype anomalies (Turner, Noonan, Trisomy 22, other subchromosomal disorders).[3,7-9] Since associated karyotype anomaly reaches up to 25% in some series, it can be recommended to carry out karyotyping in cases with ductus venosus agenesis.[10]

In a series of 22 cases, it was reported by Thomas et al.[7] that perinatal loss and pregnancy termination rate was 45% in cases accompanied by structural anomaly, and it was 75% in cases accompanied by chromosomal anomaly while survival rate was 100% in isolated cases.

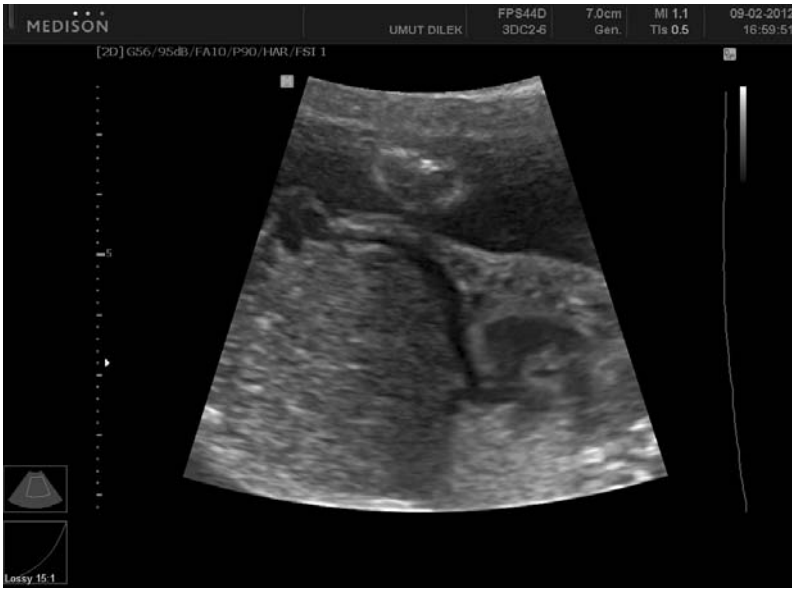

Fig. 1. The course of umbilical vein beginning from the inlet point into abdomen and direct opening to right atrium.

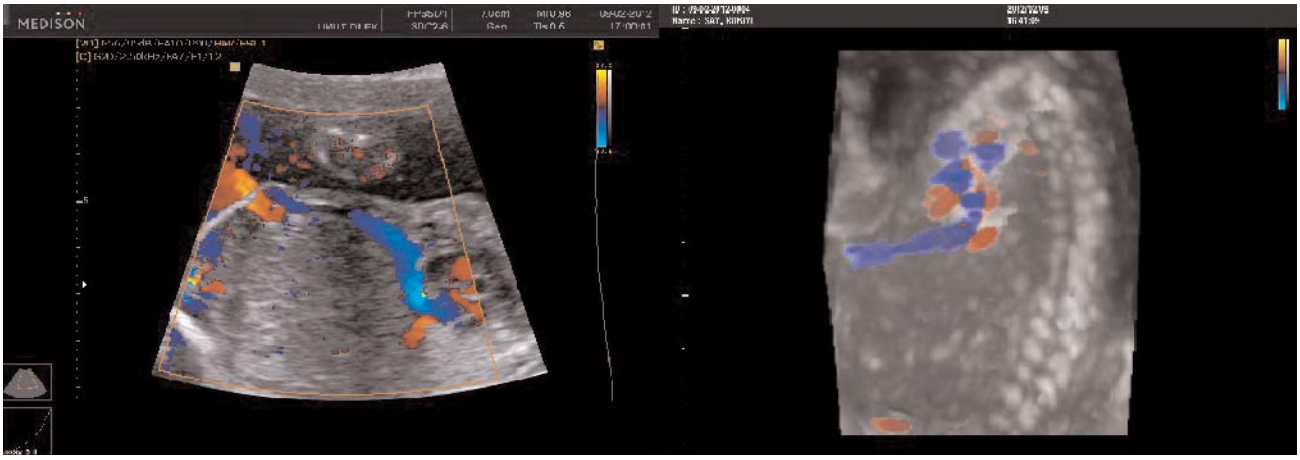

Fig. 2. Direct opening of umbilical vein into right atrium and inferior vena cava joining to the same point from posterior in *color-flow* mode.

Berg et al.[3] reported that prognosis varied according to intrahepatic or extrahepatic drainage of umbilical vein and it was worse in those with extrahepatic drainage. Survival rate was 100% in ductus venosus agenesis cases where isolated, hydrops and pleural effusion did not accompanied; otherwise, the rate was reduced to 50%.[7] In our case, hydrops was not observed; however, accompanying cardiac and renal anomalies and existence of single umbilical artery were consistent with the case reports and cases series reported in the literature.

Conclusion

Increasingly examining ductus venosus by ultrasonographic evaluations performed during aneuploidy screenings between 11 and 14 gestational weeks enabled to establish ductus venosus agenesis diagnosis more. Yet, it is a malformation rarely observed. Portal venous system should also be evaluated in case of extrahepatic drainage of umbilical vein as well as full anatomic evaluation and karyotyping in cases diagnosed ductus venosus agenesis. Existence of hydrops and accompanying anomalies are major factors determining prognosis.

Conflicts of Interest: No conflicts declared.

References

1. Yagel S, Kivilevitch Z, Cohen SM, Valsky DM, Messing B, Shen O, et al. The fetal venous system, part I:normal embriyology, anatomy, hemodynamics, ultrasound evaluation and doppler investigation. Ultrasound Obstet Gynecol 2010;35:741-50.
2. Stabulidau I, Pereira S, de Jesus Cruz J Syngelaki A, Nicolaides KH. Prevalence and outcome of absence ductus venosus at 11(+0) to 13(+6) weeks. Fetal Diagn Ther 2011;30:35-40.
3. Berg C, Kamil D, Geipel A, Kohl T, Knöpfle G, Hansmann M, et al. Absence of ductus venosus–importance of umbilical venous drainage site. Ultrasound Obstet Gynecol 2006;28:275-81.
4. Contratti G, Banzi C, Ghi T, Perolo A, Pilu G, Visentin A. Absence of the ductus venosus: report of 10 new cases and review of the literature. Ultrasound Obstet Gynecol 2001;18:605-9.
5. Jaeggi ET, Fouran JC, Hornberger LK, Proulx F, Oberhänsli I, Yoo SJ, et al. Agenesis of the ductus venosus that is associated with ex-trahepatic umbilical vein drainage: prenatal features and clinical outcome. Am J Obstet Gynecol 2002;187:1031-7.
6. Shen O, Valsky DV, Messing B, Cohen SM, Lipzschuetz M, Yagel S. Shunt diameter in agenesis of the ductus venosus with extrahepatic portosystemic shunt impact on prognosis. Ultrasound Obstet Gynecol 2011;37:184-90.
7. Thomas JT, Petersen S, Cincotta R, Lee-Tannock A, Gardener G. Absent ductus venosus -outcomes and implications from tertiary center. Prenat Diagn 2012;32:686-91.
8. Corbacioglu A, Aslan H, Dagdeviren H, Ceylan Y. Prenatal diagnosis of abnormal course of umbilical vein and ductus venosus agenesis: report of three cases. J Clin Ultrasound Prenat Diagn 2012:40;590-3.
9. Clerici G, Rosati A, Di Renzo GC. Absent ductus venosus associated with skeletal anomalies of the ulna and radius. Prenat Diagn 2010;32:83-5.
10. Volpe P, Marasini M, Caruso G, Lituania M, Marzullo A, Volpe G, et al. Prenatal diagnosis of ductus venosus agenesis and its association with cytogenetic/congenital anomalies. Prenat Diagn 2002;22:995-1000.