Two ductus venosus: a previously unreported anomaly

Mohammed Firdouse · Arnav Agarwal ·
Bharadwaj Pindiprolu · Tapas Mondal

Received: 18 December 2013 / Accepted: 17 March 2014 / Published online: 16 April 2014
© Società Italiana di Ultrasonologia in Medicina e Biologia (SIUMB) 2014

Abstract In early fetal life, the ductus venosus (DV) connects the umbilical vein, carrying nutrient-rich and oxygenated blood from the placenta to the inferior vena cava (IVC). We present the first ever case of a fetus with two ductus venosus. The fetus presented with a four-vessel cord and a dilated bowel at 31 weeks of gestation. Ultrasonography showed a persistent right umbilical vein. Echocardiographic investigation revealed normal cardiac anatomy with no major malformations. Two DVs with slightly different Doppler patterns were visualized anastomosing with the IVC. The baby was born uneventfully at 39 weeks of gestation with stable hemodynamics. Assuming every supernumerary umbilical vein should be

connected to a DV for balanced circulation, a fetus with supernumerary umbilical veins lacking a corresponding number of DV connections is likely to be predisposed to complications such as hydrops fetalis and poor perinatal outcomes. The possibility of one or more umbilical veins lacking a DV connection warrants significant attention and regular monitoring from feto-maternal specialists, given the severity of the associated morbidity and mortality.

Keywords Two ductus venosus · Four-vessel umbilical cord

Riassunto Nei primi mesi di vita fetale, il dotto venoso (DV) collega la vena ombelicale, portando il sangue ossigenato, ricco di sostanze nutritive dalla placenta alla vena cava inferiore (IVC). Presentiamo il primo caso in assoluto di un feto con due dotti venosi. Il feto, alla 31 settimane di gestazione, si presentava con quattro vasi nel cordone ombelicale e un intestino dilatato. L'ecografia mostrava la persistenza di una vena ombelicale destra. L'indagine ecocardiografica rilevava un'anatomia cardiaca normale, senza malformazioni maggiori. Sono stati evidenziati due DV con tracciati Doppler leggermente diversi, che si anastomizzavano con l'IVC. Il bambino è nato senza complicazioni a 39 settimane di gestazione, con emodinamica stabile. Supponendo che ogni vena ombelicale soprannumeraria deve essere collegato ad un DV per una circolazione equilibrata, un feto con vene ombelicali soprannumerarie, che manchi di un corrispondente numero di DV rischia di essere predisposti a complicanze come l'idrope fetale e difficoltà perinatali. La possibilità di una o più vene ombelicali prive di DV merita notevole attenzione e un regolare monitoraggio feto- materno, data la gravità della morbidità e la mortalità associate

M. Firdouse · A. Agarwal · B. Pindiprolu
Faculty of Health Sciences, McMaster University, Hamilton,
ON, Canada
e-mail: mohammed.firdouse@learnlink.mcmaster.ca

M. Firdouse
McMaster University, 4077 Tea Garden Circle, Mississauga,
ON L5B 2W5, Canada

A. Agarwal
McMaster University, 252 Macedonia Crescent, Mississauga,
ON L5B 3R7, Canada
e-mail: arnav.agarwal@learnlink.mcmaster.ca

B. Pindiprolu
McMaster University, 4371 Alta Court, Mississauga,
ON L4Z 1N6, Canada
e-mail: bharadwaj.pindiprolu@learnlink.mcmaster.ca

T. Mondal (✉)
Department of Pediatrics, McMaster Children's Hospital,
McMaster University, 1280 Main Street West, Hamilton,
ON L8S 4K1, Canada
e-mail: mondalt@mcmaster.ca

Introduction

Considering that gaseous exchange of fetal blood occurs in the placenta and not in fetal lungs, the cardiovascular system and its associated hemodynamic properties are significantly different in prenatal and postnatal life. The placenta receives deoxygenated systemic blood from the fetus via umbilical arteries and returns oxygenated blood to the fetus via umbilical veins. In contrast to the adult cardiovascular system, the fetal cardiovascular system is designed in such a way that it preferentially streams oxygenated blood to the brain and heart via intracardiac and extracardiac shunts.

The ductus venosus (DV) is one such extracardiac shunt that helps in fetal development. Anatomist Giulio Cesare Aranzio from the sixteenth century was the first to describe DV as a short trumpet or funnel shaped vessel in the fetal liver. Its primary function is to connect the intra-abdominal umbilical vein returning from the placenta via the umbilical cord to the inferior vena cava (IVC). The presence of a DV is crucial during fetal development as it helps the nutrient-rich and oxygenated blood returning from the placenta to bypass the liver. The degree of shunting through the ductus venosus in the human fetus varies from 30 % of umbilical venous blood in the second trimester and reduces to about 20 % towards the end of the third trimester [1]. As this oxygenated blood bypasses the liver and travels up the IVC, the Eustachian valve tends to push it from the right to the left atrium (LA) across the foramen ovale, an intracardiac shunt in the atrial septum. This oxygenated blood then travels from the LA to the LV, from where it is pumped to the heart and brain. This mechanism helps to ensure that the blood with the highest oxygen concentration is supplied to the most vital organs first. Within 1–4 days following birth, the DV begins to close and eventually transforms into the ligamentum venosum in the infant's liver [2] (Fig. 1).

Advancements in imaging technologies such as ultrasonography and Doppler have made it possible to examine the DV both prenatally and postnatally. The umbilical cord usually contains two arteries and one vein. The commonest reported abnormality in the cord is the presence of a single umbilical artery; however, multiple arteries and veins have also been reported [3]. Furthermore, the most commonly reported anomaly of the DV is its agenesis, which is often associated with chromosomal as well as fetal cardiac and extracardiac malformations often manifesting as hydrops fetalis [4, 5]. Hypothetically each umbilical vein in cords with supernumerary vessels should have a DV to regulate fetal circulation. In cases of multiple umbilical veins in the cord, there has been no mention in previous literature regarding the number of DV. To the best of our knowledge, this is the first antenatal description of a fetus with a two

Fig. 1 After entering the liver, a portion of the left umbilical vein connects to the hepatic sinusoids, whereas majority of the oxygenated blood bypasses the liver through a trumpet-shaped ductus venosus (DV) in order to drain into the inferior vena cava (IVC)

functional ductus venosus and a four-vessel cord containing two umbilical arteries and two umbilical veins.

Case report

We report the case of a fetus at 31 weeks of gestation who presented to us with questions of a multiple vessel umbilical cord and dilated bowel following routine fetal ultrasonography. The estimated weight of the fetus was 1,578 g, which is between the 10th and 50th percentile for the given gestational age. The ultrasound revealed a persistent right umbilical vein in the cord. The descending colon appeared to be somewhat prominent with a maximum diameter of 7.6 mm, however, it was within the normal range (6.3–9.05 mm). The DV was not assessed in this examination. Overall, the surveillance for fetal well-being was promising with normal biophysical profile, umbilical artery Dopplers and amniotic fluid volume.

A follow-up with an echocardiographic investigation noted a normally placed heart with a normal cardiothoracic ratio. A normal four-chamber view was visualized with no signs of any pericardial effusion. The superior and inferior vena cavae were noted to drain normally into the RA. The great vessels were also normally positioned with the aorta posteriorly and pulmonary artery anteriorly. The foramen ovale was patent and blood flowed from the RA to the LA. There were no signs of mitral or tricuspid valve

Fig. 2 **a** Color flow mapping showing laminar flow in the inferior vena cava (IVC) and turbulent flow in the two ductus venosus (DV). **b** Doppler flow pattern of the left ductus venosus. **c** Doppler flow pattern of the right ductus venosus

regurgitation. No obstructions were noted in the left or the right ventricular outflow tract either. Overall, the fetus had a normal cardiac anatomy with no obvious malformations. Two umbilical veins and two umbilical arteries were visualized in the cord. The umbilical artery had a pulsatility

Fig. 3 Cross section of the umbilical cord at birth showing four vessels—two umbilical veins (*dashed arrows*) and two umbilical arteries (*dashed circles*)

index of 0.9 and resistance index of 0.6. Two DV were also noted to anastomose with the IVC, each one of them having slightly different Doppler flow patterns (Fig. 2a–c). Doppler assessment of blood flow in the IVC at this level and in the region between the IVC–RA junction was normal with no dilation. No other prominent vessels were visualized in this area to suggest absence of DV venous in either of the umbilical veins, which might cause volume overload.

The baby was born uneventfully at 39 weeks of gestation. An inspection of the umbilical cord during delivery confirmed the presence of a four-vessel cord, with two arteries and two veins (Fig. 3). The baby was monitored for any potential cardiovascular complications, had an uneventful neonatal period and remained hemodynamically stable. A postnatal echocardiogram on day 2 was normal. Consequently, the baby was discharged without any concern.

Discussion

The development of venous system in fetal life occurs 21–35 days following fertilization and implantation of the embryo. In the early stages of intrauterine life, the embryo has three different sets of veins—vitelline veins, umbilical veins and cardinal veins. The vitelline veins drain blood from the yolk sac into the sinus venosus (SV) of the developing heart. The umbilical veins drain oxygenated and nutrient-rich blood from the placenta to the SV. The cardinal veins drain deoxygenated blood from the rest of the body to the SV. Each of the aforementioned sets of veins are present bilaterally (right and left). As the fetus

develops, the vitelline veins become the IVC, portal and hepatic veins, whereas the cardinal veins become the SVC and the azygous vein. Furthermore, the entire length of the right umbilical vein as well as the proximal part of the left umbilical, which is connected to the SV, undergo degeneration by 5 weeks. Consequently, they cannot be visualized during antenatal imaging. The distal part of the left umbilical vein is redirected into the hepatic sinusoids where it helps in the development of the portal system as well as in the formation of a special channel called the DV which helps the oxygenated blood to bypass the hepatic parenchyma and drain into the developing IVC [6]. Following birth, the umbilical vein undergoes fibrosis to form the ligamentum teres while the ductus venosus becomes the ligamentum venosum.

Occasionally, as seen in cords containing supernumerary vessels, the right umbilical vein does not degenerate. The presence of a persistent right umbilical vein (PRUV) can be classified into the intrahepatic type (PRUV merges with the hepatic portal system and drains into the DV) or the extrahepatic type (PRUV drains directly to either the RA, IVC or the iliac veins). The intrahepatic type is often asymptomatic due to the presence of the DV, whereas the extrahepatic type, which likely lacks a DV, has been associated with malformations in the cardiovascular, nervous and gastrointestinal system [7]. Agenesis or the absence of DV has also been frequently reported in the literature. This results in an anomalous venous drainage from the placenta can be classified into three distinct types depending on its site of drainage [8]:

1. The left umbilical vein drains only into the hepatic portal vein.
2. The left umbilical vein connects directly the IVC (suprahepatically or intrahepatically).
3. The left umbilical vein drains directly into the RA.

Based on our understanding of the hemodynamics in the fetal circulatory system, as long as each umbilical vein drains into the IVC through its respective DV, we do not anticipate any cardiac complications, as the circulation remains balanced. The absence of a DV, however, often presents with hydrops fetalis, generalized skin edema, ascites, pleural effusion and heart failure which could have detrimental consequences on the fetus such as premature delivery and perinatal mortality [8]. Thus, fetomaternal specialists should pay attention to the fact that even if one DV is present, in the rare situation where there are multiple

umbilical veins, there is a risk of hydrops fetalis due to the absence of corresponding numbers of DV.

Conclusion

To the best of our knowledge, we present the first case of a fetus with two ductus venosus and a corresponding four-vessel cord, with two arteries and two veins. Due to the significant hemodynamic role that the DV plays during fetal life, perinatologists and imaging specialists must pay greater attention to and constantly monitor fetuses with supernumerary cords containing multiple umbilical veins but only one DV.

Conflicts of interest Mohammed Firdouse, Arnav Agarwal, Bharadwaj Pindiprolu, Tapas Mondal declare that they have no conflict of interest.

Ethics standards The authors confirm that all human and animal studies have been approved by the appropriate ethics committee and have therefore been performed in accordance with the ethical standards laid down in the Helsinki Declaration of 1975 and its late amendments. Additional informed consent was obtained from all patients for which identifying information is not included in this article.

References

1. Kiserud T, Rasmussen S, Skulstad SM (2000) Blood flow and degree of shunting through the ductus venosus in the human fetus. *Am J Obstet Gynecol* 182:147–153
2. Rychick J, Zhiyun T (2012) Fetal cardiovascular imaging: a disease-based approach. Saunders, Philadelphia, pp 397–402
3. Wolman I, Gull I, Fait G, Amster R, Kupferminc MJ, Lessing JB et al (2002) Persistent right umbilical vein: incidence and significance. *Ultrasound Obstet Gynecol* 19:562–564
4. Staboulidou I, Pereira S, de Jesus CJ et al (2011) Prevalence and outcome of absence of ductus venosus at 11 + 0 to 13 + 6 weeks. *Fetal Diagn Ther* 1:35
5. Berg C, Kamil D, Geipel A et al (2006) Absence of ductus venosus-importance of umbilical venous drainage site. *Ultrasound Obstet Gynecol* 3:275
6. Sadler TW (2012) Langman's medical embryology, 12th edn. Lippincott Williams & Wilkins, Philadelphia, pp 192–195
7. Blazer S, Zimmer EZ, Bronshtein M (2000) Persistent intrahepatic right umbilical vein in the fetus: a benign anatomic variant. *Obstet Gynecol* 95:433–436
8. Contratti G, Banzi C, Ghi T, Perolo A, Pilu G, Visentin A (2001) Absence of the ductus venosus: report of 10 new cases and review of the literature. *Ultrasound Obstet Gynaecol* 18(6):605–609