The physiological implications of absent ductus venosus during fetal and post-natal life

Shweta Bakhru, Nageswara Rao Koneti
Department of Pediatric Cardiology, Rainbow Children's Heart Institute, Hyderabad, Telangana, India

ABSTRACT

Background : **The ductus venosus (DV) is a pivotal component of fetal circulation. Absent DV (ADV) is associated with structural defects, portal vein (PV) anomalies, and chromosomal anomalies. This observational study aims to investigate the impact of ADV on fetal circulation and postnatal outcomes.**

Materials and Methods : **This observational study was conducted from August 2016 to January 2020 at a fetal and pediatric cardiac center. The DV was evaluated as part of routine fetal echocardiography. Cases of ADV were identified. Blood flow and exit points of the umbilical vein were studied. Cardiothoracic ratio, hydrops, and PV were assessed during the initial and follow-up scans. The postnatal evaluation included an ultrasound abdomen and computed tomography with triple-phase imaging to assess portosystemic shunts (PSSs). Serum ammonia levels were monitored.**

Results : **Twelve patients with ADV were identified. The median maternal age and median gestational age were 27.5 years and 22 weeks, respectively. Four patients had intrahepatic drainage, while eight had extrahepatic drainage. All patients (100%) exhibited cardiomegaly, but none developed hydrops. Four patients had persistent PSS postnatally. All four patients with PSS had asymptomatic hyperammonemia. Two patients underwent transcatheter closure of PSS. The intrahepatic variant showed good PV anatomy with no evidence of PSS.**

Conclusions : **DV evaluation should be performed during fetal echocardiography. ADV can lead to cardiomegaly and dilation of the right atrium and ventricle. PSS can be a potential sequela of the extrahepatic variant of ADV.**

Keywords : Fetal cardiomegaly, fetal echocardiography, portosystemic shunt

INTRODUCTION

The ductus venosus (DV) is an essential component of fetal circulation, regulating the flow of oxygenated blood from the umbilical vein (UV) to the heart through its sphincteric mechanism. Postnatally, the DV undergoes fibrotic transformation and forms the ligamentum venosum.[1] DV evaluation is often utilized as a screening marker for chromosomal anomalies, hydrops, or fetal growth restriction.[2,3] Literature discusses the association of absent DV (ADV) with chromosomal anomalies, structural defects, hydrops fetalis, and portal vein (PV) agenesis.[4-7] However, there is limited data on the physiological changes of ADV and its implications in prenatal and postnatal life.

Videos available on: https://journals.lww.com/aopc

Access this article online

Quick Response Code:

Website: https://journals.lww.com/aopc

DOI: 10.4103/apc.apc_93_24

How to cite this article: Bakhru S, Koneti NR. The physiological implications of absent ductus venosus during fetal and post-natal life. Ann Pediatr Card 2024;17:257-63.

Address for correspondence: Dr. Shweta Bakhru, Rainbow Children's Heart Institute, Hyderabad, Telangana, India.
E-mail: hrehaan2015@gmail.com

Submitted: 12-May-2024 **Revised:** 21-Jul-2024 **Accepted:** 07-Sep-2024 **Published:** 15-Nov-2024

The present study aims to delineate the physiological changes in fetal circulation in the absence of DV. In addition, our observations indicate that some patients exhibit an abnormal channel communicating between the portal and the systemic veins, known as portosystemic shunts (PSSs). These shunts may persist after birth and necessitate closure to prevent metabolic derangements.

MATERIALS AND METHODS

This prospective observational study was performed at a single fetal and pediatric cardiology center between August 2016 and January 2020. The study was carried out following relevant guidelines and regulations. An institutional review board waiver was obtained from "Rainbow Children's Medicare Pvt. Ethics Committee." Informed consent was obtained from subjects during fetal echocardiogram. All patients were referred for fetal echocardiography with various indications. DV was evaluated in all cases as a routine protocol. Cases with ADV were included in the study. Fetuses associated with other structural heart defects were excluded from the study.

Fetal assessment

A fetal echocardiogram was performed on Phillips iE33 (Koninklijke Philips N.V.) or GE Voluson sonography equipment (General Electric Company, Boston, MA 02210, United States). In all suspected cases of ADV, the UV course, drainage, and PV were evaluated. A detailed cardiac structural and functional assessment was performed. Every 4–6 weeks, periodic follow-up of fetal echocardiograms was done to look for cardiomegaly, chamber dilation, aortic, pulmonary velocity, and development of hydrops till delivery.

Steps for ductus venosus assessment

1. DV can be demonstrated in the thoracoabdominal midsagittal or oblique transverse planes. It is seen as a branching vessel from the UV with turbulent flow. Color flow mapping and speckled pulse wave Doppler were helpful in locating and confirming DV.[Figure 1a-d]
2. The umbilical drainage point was ascertained in the case of ADV. An attempt was made to look for PV to systemic vein communications. The midsagittal plane and the coronal plane of the fetus's abdomen were profiled to look for PSS connections
3. Dilation of systemic veins and cardiac chambers was assessed in all cases. The cardiothoracic ratio was measured periodically during follow-up. Cases were followed up for the development of hydrops fetalis. Umbilical artery and venous Dopplers were recorded.

Neonatal assessment

After birth, all babies underwent detailed transthoracic echocardiograms for cardiac function, pulmonary artery pressures, and identification of PSS. In suspected cases of PSS, a computed tomography (CT) angiogram with portal phase (triple phase) was done after 6 months of age. This was useful for assessing the PV and PSS to plan for further management. Periodic serum ammonia was monitored in all cases with PSS.

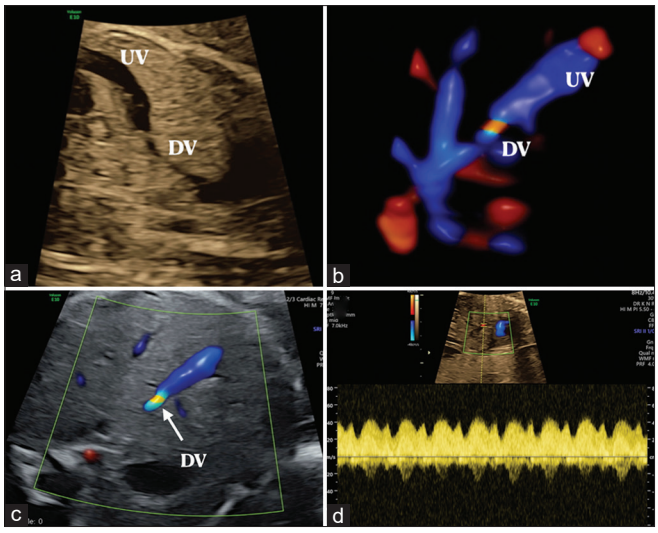

Figure 1: (a) Normal intra-abdominal umbilical vein and ductus venosus (DV) opening into portohepatic system, (b) color flow rendered image showing increased flow velocity at DV, (c) Abdominal cross section at the level of DV, (d) Spectral Doppler of DV showing the typical pattern with absent "a" reversal, DV: ductus venosus, UV: umbilical vein, IVC: Inferior vena cava

Management

Infants without PSS were asymptomatic and needed only medical follow-up. Patients with PSS and elevated ammonia levels were intervened to close the PSS. A balloon occlusion angiogram was initially done to check the portal pressures and intrahepatic portal radicles. A transcatheter device closure was done if portal pressures were <18 mmHg and portal radicles were adequate.

RESULTS

A total of 12 fetuses were diagnosed as ADV during the study period. The total number of fetal echocardiograms was 7552 during the study period. The median maternal age at diagnosis was 27.5 years (range: 22–36). The median gestational age was 22 weeks (range: 20–27). Amniocentesis was done in four patients and showed no abnormality. At least 3 settings of the follow-up fetal evaluation were done after diagnosis. The anatomical variants were divided into two groups.

Group 1: Intrahepatic variants (*n* = 4)

In this variety, UV was directly draining into PV. Blood flow from the UV was directed toward the PV and then to portal radicles and hepatic veins [Video 1]. The flow from the UV is naturally regulated by the portal system, and therefore, cardiomegaly was not noticed.

The PV was dilated in intrahepatic type, and the PV Doppler was similar to the UV without any pulsatile waveform [Figure 2].

Group 2: Extrahepatic variants (*n* = 8)

In this variety, UV drains into one of the systemic veins, namely, the inferior vena cava (IVC), coronary sinus, iliac vein, or right atrium (RA) [Figure 3]. Due to the absent sphincteric function of DV, unrestricted blood enters fetal circulation, which leads to cardiomegaly [Video 2]. All fetuses with the extrahepatic variant had cardiomegaly due to dilation of RA and right ventricle (RV). Grossly dilated IVC was seen in three patients where UV was draining into IVC [Figure 4]. Two patients with the extrahepatic variant had portosystemic communication (also called Abernethy malformation) due to side-to-side communication between PV and IVC [Figure 5 and Video 3].

None of the patients had hydrops fetalis or intrauterine death. Two pregnancies were terminated due to unknown reasons. The autopsy was not done.

Postnatal analysis [Table 1]

There were 10 (female 5) infants. The intrahepatic type (*n* = 4) had an uneventful outcome and a normal heart. Ultrasound of the abdomen did not reveal any PSS. The extrahepatic variant (*n* = 8) had a variable outcome. Two neonates had tachypnea soon after birth and needed hospitalization. One had hypertrophic cardiomyopathy, and the other had persistent pulmonary hypertension, which improved with medical therapy. The rest all had asymptomatic neonatal courses.

Cardiomegaly was resolved in all extrahepatic types (*n* = 6) after birth. PSS was seen in four patients. Two (cases 11 and 12) were diagnosed antenatally. The types of PSS found in these patients are as below:

i. PV to coronary sinus: 1 (case 1)
ii. PV to IVC side-to-side communication: 2 (cases 11 and 12). Case 11 had intrahepatic PV hypoplasia with a large communication of 14 mm from PV to IVC
iii. PV to the renal vein with multiple channels: 3 (case 3).

Serum ammonia levels were elevated in all PSS patients (median: 74 mmol; range: 72–400 mmol). Cases 1 and 3 underwent successful transcatheter closure of PSS at 1 year and 6 months of age, respectively [Figure 6]. Ammonia levels normalized soon after the closure of PSS. Case 3 had progressive hypertrophic cardiomyopathy along with generalized myopathy and succumbed to pneumonia. Case 11 has a severe degree of portal radicle hypoplasia and very high ammonia levels. Balloon occlusion angiogram showed high portal pressure (28–30 mmHg). Hence, he was planned for partial PSS closure. The case 12 is asymptomatic and waiting for the transcatheter closure. Case 5 had congenital IVC obstruction at the suprahepatic portion, detected on CT angiogram. No intervention was required as the patient was asymptomatic.

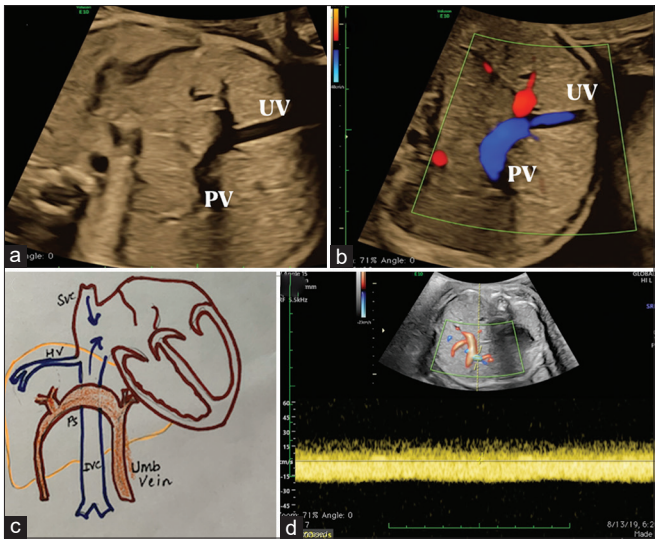

Figure 2: Ultrasound images of absent ductus venosus (DV) with intrahepatic variant: (a) Intrahepatic opening of umbilical vein (UV) into the portal vein (PV). DV is absent, (b) color flow mapping of the same showing UV is directly opening into PV, (c) schematic diagram showing absent DV, with intrahepatic drainage, (d) Doppler interrogation showing continuous flow across PV mimicking UV. PV: portal vein, UV: umbilical vein, DV: ductus venosus, HV: hepatic vein, SVC: superior vena cava, PS: portal sinus, IVC: Inferior vena cava

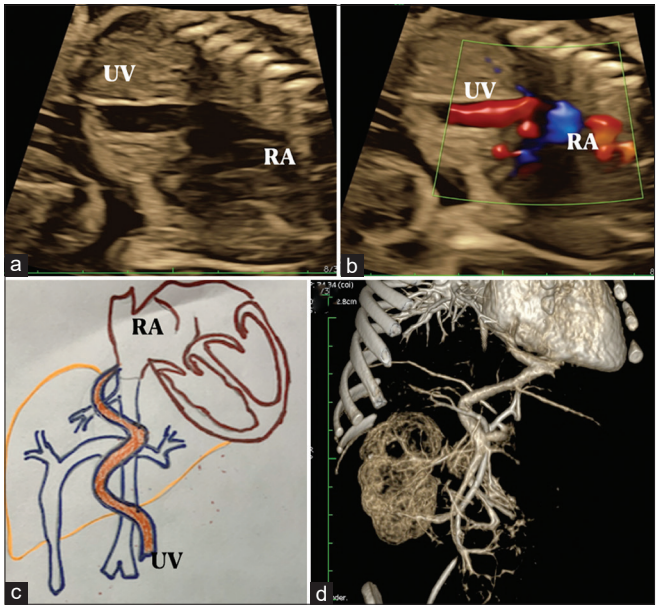

Figure 3: Ultrasound images of a 26-week fetus showing: (a) Direct opening of umbilical vein (UV) to the right atrium (RA), (b) color flow mapping showing the direct drainage of UV to RA, and (c) a schematic diagram showing intraabdominal UV to RA, (d) computed tomography angiogram showing persistent abnormal communication between portal vein to RA (portosystemic shunts). UV: Umbilical vein, RA: Right atrium

Table 1: Illustrated description of antenatal findings and postnatal outcome of absent ductus venosus cases

Gestational age (weeks)	Prenatal diagnosis	Cardiomegaly	Sex	Postnatal diagnosis	Outcome
24	UV to RA	Yes	Female	Portosystemic shunt	Device closure of PSS Hyperammonemia Doing well
23	UV to RA	Yes			Termination
20	UV to IVC	Yes	Female	PSS Myopathy HCM	Required prolonged oxygen Device closure of PSS Hyperammonemia Died
22	UV to RA	Yes			Termination
22	UV to RA	Yes	Male	Budd–Chiari syndrome IVC obstruction	Doing well
29	UV to PV	No	Female	Normal	Asymptomatic
22	UV to RA	Yes	Male	Normal	Asymptomatic
23	UV to PV	Yes	Female	Normal	Asymptomatic
27	UV to PV	No		Normal	Asymptomatic
22	UV to PV	No	Male	Normal	Asymptomatic
24	UV to IVC, PSS	Yes	Female	PSS	Hyperammonemia Predominantly asymptomatic
22	UV to IVC, PSS	Yes	Male	PSS	Had PPHN medically resolved

UV: Umbilical vein, RA: Right atrium, PSS: Portosystemic shunt, PV: Portal vein, IVC: Inferior vena cava, PPHN: Persistent pulmonary hypertension of the newborn, HCM: Hypertrophic cardiomyopathy

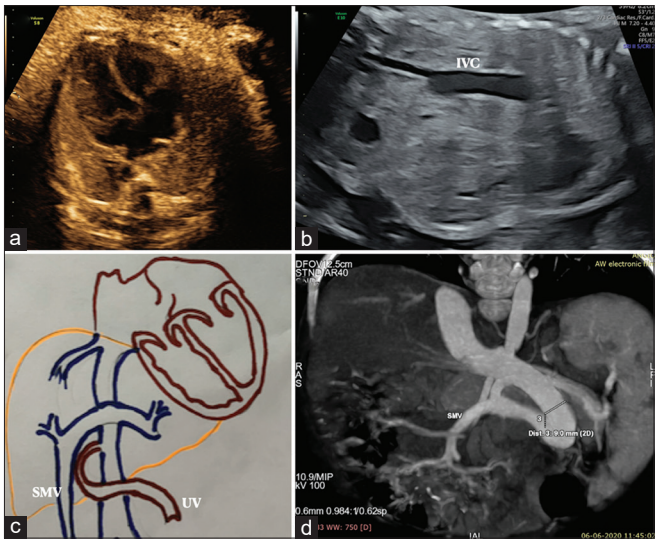

Figure 4: Fetal ultrasound images of a case of unexplained cardiomegaly in a 27-week fetus: (a) Cardiomegaly with no structural heart defect, (b) dilated mid-inferior vena cava (IVC), (c) schematic diagram showing direct drainage of umbilical vein to IVC, (d) computed tomography angiogram of the same patient after birth showing large fistula between the portal vein and IVC. Significant dilation of the intrahepatic portion of IVC is seen

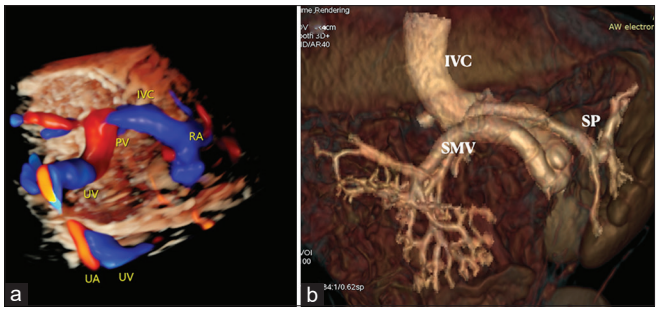

Figure 5: (a) Three-dimensional color flow volume rendered image of a fetus showing direct drainage of umbilical vein to inferior vena cava (IVC), (b) computed tomography angiogram of the same patient showing drainage of portal vein into IVC through the fistulous tract. UV: umbilical vein, PV: portal vein, RA: right atrium, IVC: inferior vena cava, SMV: superior mesenteric vein, SP: spleen, UA: umbilical artery

DISCUSSION

Three levels of shunts occur during fetal life, namely, the DV, foramen ovale, and ductus arteriosus. This arrangement enables preferential diversion of oxygenated blood from the placenta to the left atrium, left ventricle, and cerebral circulation of the fetus. The DV plays a pivotal role in fetal circulation by connecting the abdominal part of the UV to the IVC and carrying around 50% of cardiac output. Anatomically, the intra-abdominal portion of the UV reaches the hilum of the liver and provides branches to the left lobe of the liver. It further divides into the DV and a large arcuate branch, which travels to the right and joins the PV. The DV travels dorsally and cephalad through the liver parenchyma and joins the IVC just beneath the diaphragm, sharing a common entry into the IVC along with the left hepatic vein.[1]

The DV acts as a partial bypass of the hepatic circulation for umbilical venous blood. It is well documented that a sphincter at the outflow of the DV prevents unrestricted flow into the fetal circulation, possibly maintaining an impedance that prevents the complete mixing of umbilical venous and portal venous blood.[1,8]

The DV can be identified in fetal ultrasound either on a midsagittal plane or an oblique transverse plane at the thoracoabdominal level. Pulse Doppler at the isthmus of the DV demonstrates peak systolic velocity of 48–71 cm/s and peak diastolic velocity of 31–58 cm/s, depending on gestational age. The small reversal of waveform during atrial contraction is absent in a normal fetus.[9]

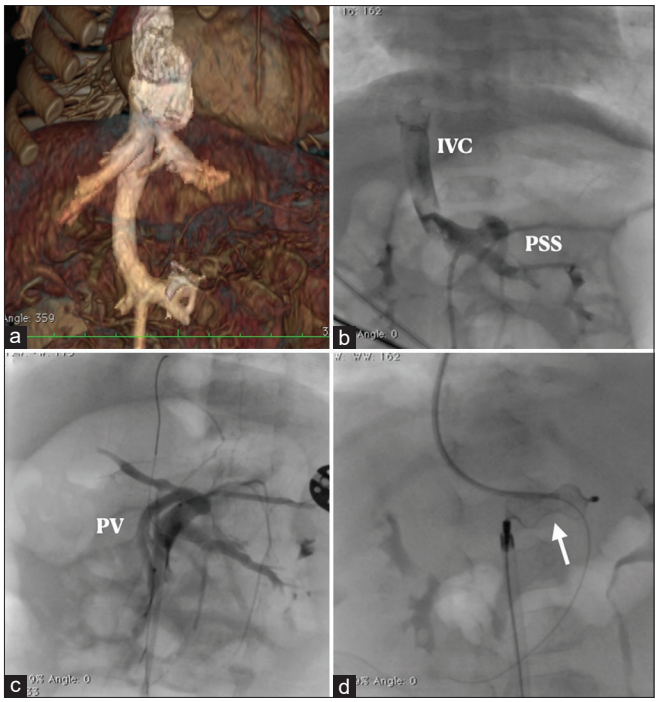

Figure 6: (a) Computed tomography angiogram of a child with absent ductus venosus during prenatal ultrasound examination showing portosystemic shunt (PSS), (b) cine angiogram showing PSS, (c) balloon occlusion angiogram showing intrahepatic portal radicals. (d) A vascular plug (white arrow) is being deployed in the channel of the PSS. PSS: portosystemic shunt, PV: portal vein, IVC: Inferior vena cava

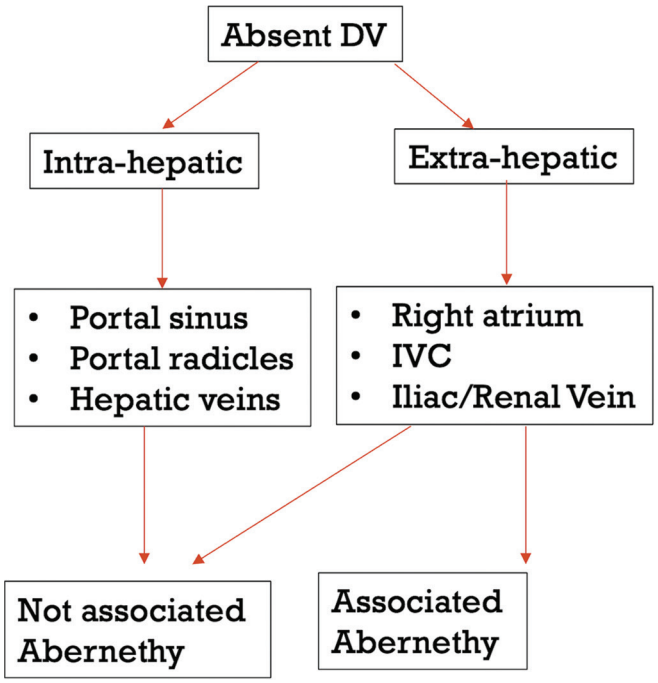

Figure 7: Flow chart showing the various possible outcomes of absent ductus venosus. IVC: Inferior vena cava, DV: Ductus venosus

The evaluation of the DV in the first trimester serves as a screening marker for chromosomal defects, fetal hydrops, or monitoring fetal growth restriction. However, routine evaluation of the DV during the second-trimester anomaly scan is not common practice, potentially leading to the oversight of DV agenesis. Consequently, the exact incidence of this anomaly remains uncertain.[4] Contratti *et al.* reported 10 cases of ADV and found that 24% were associated with chromosomal anomalies. They noted that 33% of cases had hydrops, with 20% experiencing mortality in their small cohort.[4] Agenesis of the PV was documented in some cases where the UV was connected to the RA or IVC. The high incidence of mortality might be biased findings in this study as patients presented with hydrops, polyhydramnios, and fetal anomalies. In our study, none of the fetuses developed hydrops. Similarly, other studies showed favorable outcomes where hydrops was not associated.[10-12] The consequences and implications of ADV during fetal and postnatal stages are understood, but our study offers further insight into this rare anomaly.

We encountered two types of presentations in this entity [Figure 7]:

1. Fetal presentation (progressive right heart dilation): All extrahepatic variants of ADV exhibited RA and RV dilation and cardiomegaly during the late second and third trimesters. As previously discussed, the sphincter and narrow diameter of the DV regulate blood flow entering the fetal circulation.[8] Therefore, in the absence of the DV, where the UV is directly connected to the RA or systemic vein, blood flow is diverted toward the RA. This results in an increase in preload and progressive dilation of the RA and RV, along with cardiomegaly. Occasionally, it can progress to hydrops.
   In cases of the intrahepatic variant, although unregulated blood flow from the UV enters the PV, cardiomegaly is not observed. This is due to the significant amount of blood flow distributed in the portal circulation and liver parenchyma
2. Postnatal presentation (portosystemic shunts in extrahepatic variant): The extrahepatic variant of ADV may present with PSS after birth.

In the postnatal presentation of the extrahepatic variant:

1. Metabolic consequences of portosystemic shunts: Out of eight cases of the extrahepatic type of ADV, 4 (50%) had PSS. It is an aberrant communication between the systemic vein and the PV. Gut metabolites, which are supposed to be detoxified in the liver, enter the systemic circulation due to PSS. This can manifest as hepatic encephalopathy, hyperammonemia, galactosemia, hypoglycemia, pulmonary artery hypertension, or cyanosis. Often, undiagnosed PSS patients may present with cyanosis or pulmonary hypertension due to portopulmonary syndrome, even in the first decade of life. This happens due to the formation of diffuse pulmonary

arteriovenous malformations and shear stress by the volume in the distal pulmonary vascular bed. PSS closure dramatically improves cyanosis and pulmonary hypertension. Therefore, it is an important entity where timely attention can prevent serious sequelae[13-16]

2. Portal vein hypoplasia: This is a spectrum of PSS where abnormal communication between the PV and systemic vein leads to the underfilling of portal radicals. This causes the steal of portal blood into the systemic circulation and further reduction of the portal circulation. Shen *et al.* showed that the diameter of a shunt in ADV might affect the development of the portal system. A wider shunt (larger than the size of the UV) is associated with underdevelopment or absence of the PV.[17] In our study, case 11 had a large (14 mm) PV to IVC communication, leading to hypoplasia of the portal system. This may cause a spurious diagnosis of PV agenesis, as shown in the literature[18-20]
3. Portosystemic shunts in ductus venosus agenesis-embryological attributes: It is unclear why some variants of extrahepatic ADV develop into PSS and others do not. In our study, all three cases (100%) of aberrant UV drainage to the IVC had PSS. Embryologically, there are three pairs of veins (vitelline, umbilical, and cardinal veins) connected. As development advances, some portions of these veins are fated to regress and others to continue. The PVs are derived from vitelline veins. The growth of the liver bud breaks off the portion of vitelline veins and a portion of the UV at its junction with the sinus venosus. When the terminal part of the left UV is broken by the liver bud, it gets connected with the right and left vitelline veins (future PV). The DV is a new channel that communicates between the junction of the right and left vitelline veins and the sinus venosus. Therefore, the UV gets connected to the sinus venosus through the DV. We presume that communications between the cardinal and vitelline veins persist whenever the DV is not formed, leading to various PSS. Unlike the DV, the PV to IVC communication does not close after birth.

Management

Serial fetal ultrasound is essential to monitor the progression of cardiomegaly and the development of hydrops. After birth, the UV tends to close naturally; hence, cardiomegaly disappears. Ultrasound of the abdomen and, after that, a CT angiogram should be performed in suspected cases of PSS. In our observation, neonates were asymptomatic at birth; therefore, a CT angiogram was done electively around 6 months of age. Besides routine neonatal care, blood sugar and ammonia levels should be monitored. The timing of closure of PSS is not universal, but pulmonary hypertension and cyanosis can be prevented if closed during early childhood.

Detailed steps for transcatheter PSS closure are out of the context of this article. However, in brief, the closure of PSS predominantly depends on its size, development of portal radicals, and postocclusion portal pressures. PSS can be closed successfully by a transcatheter technique using occluders. It gives rewarding results, such as normalization of ammonia levels, cyanosis, or regression of pulmonary artery hypertension.[20-24]

Some of the forms of PSS diagnosed postnatally may have had undetected ADV during fetal life. Acherman *et al.* demonstrated an association between PSS and ADV.[25]

In our study, the intrahepatic variant of ADV had a good fetal and neonatal outcome. They neither had cardiomegaly nor PSS. The portal system was well-formed. Further imaging, such as CT angiography, helps evaluate the portal system.

CONCLUSIONS

Portosystemic shunts are potential sequelae of extrahepatic ADV and should be evaluated after birth. It is possible that PSS detected postnatally might have undetected ADV during fetal life. Hence, routine prenatal evaluation of DV helps in the early identification of PSS.

Ethical standards

The authors assert that all procedures contributing to this work comply with ethical standards and are approved by the institutional ethics committee.

Acknowledgment

We acknowledge Dr. Tulika Dayal for antenatal care. We acknowledge Ms. Srilekha for the pictorial diagrams.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Rudolf A. Congenital Diseases of Heart. Clinical Physiological Consideration. 3rd ed. West Sussex, UK: Wiley-Blackwell; 2009. p. 1-14.
2. Bilardo CM, Müller MA, Zikulnig L, Schipper M, Hecher K. Ductus venosus studies in fetuses at high risk for chromosomal or heart abnormalities: Relationship with nuchal translucency measurement and fetal outcome. Ultrasound Obstet Gynecol 2001;17:288-94.
3. Figueras F, Benavides A, Del Rio M, Crispi F, Eixarch E, Martinez JM, *et al.* Monitoring of fetuses with intrauterine

growth restriction: Longitudinal changes in ductus venosus and aortic isthmus flow. Ultrasound Obstet Gynecol 2009;33:39-43.

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. Cohen SB, Lipitz S, Mashiach S, Hegesh J, Achiron R. In utero ultrasonographic diagnosis of an aberrant umbilical vein associated with fetal hepatic hyperechogenicity. Prenat Diagn 1997;17:978-82.
6. Hoppen T, Hofstaetter C, Plath H, Kau N, Bartmann P. Agenesis of the ductus venosus and its correlation to hydrops fetalis. J Perinat Med 2000;28:69-73.
7. Staboulidou I. Prevalances and outcome of absence of ductus venosus at 11-13 weeks. Fetal Diagn Ther 2011;30:35-40.
8. Gennser G. Fetal ductus venosus and its sphincter mechanism. Lancet 1992;339:132.
9. Kessler J, Rasmussen S, Hanson M, Kiserud T. Longitudinal reference ranges for ductus venosus flow velocities and waveform indices. Ultrasound Obstet Gynecol 2006;28:890-8.
10. Hofstaetter C, Plath H, Hansmann M. Prenatal diagnosis of abnormalities of the fetal venous system. Ultrasound Obstet Gynecol 2000;15:231-41.
11. 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.
12. Achiron R, Kivilevitch Z. Fetal umbilical-portal-systemic venous shunt: In-utero classification and clinical significance. Ultrasound Obstet Gynecol 2016;47:739-47.
13. Saad WE. Portosystemic shunt syndrome and endovascular management of hepatic encephalopathy. Semin Intervent Radiol 2014;31:262-5.
14. Azad S, Arya A, Sitaraman R, Garg A. Abernethy malformation: Our experience from a tertiary cardiac care center and review of literature. Ann Pediatr Cardiol 2019;12:240-7.
15. Weigert A, Bierwolf J, Reutter H, Gembruch U, Woelfle J, Ganschow R, *et al.* Congenital intrahepatic portocaval shunts and hypoglycemia due to secondary hyperinsulinism: A case report and review of the literature. J Med Case Rep 2018;12:336.
16. Mesquita RD, Sousa M, Vilaverde F, Cardoso R. Abernethy malformation: Beware in cases of unexplained hepatic encephalopathy in adults-case report and review of the relevant literature. BJR Case Rep 2018;4:20170054.
17. Shen O, Valsky DV, Messing B, Cohen SM, Lipschuetz M, Yagel S. Shunt diameter in agenesis of the ductus venosus with extrahepatic portosystemic shunt impacts on prognosis. Ultrasound Obstet Gynecol 2011;37:184-90.
18. Kwapisz L, Wells MM, AlJudaibi B. Abernethy malformation: Congenital absence of the portal vein. Can J Gastroenterol Hepatol 2014;28:587-8.
19. Morgan G, Superina R. Congenital absence of the portal vein: Two cases and a proposed classification system for portasystemic vascular anomalies. J Pediatr Surg 1994;29:1239-41.
20. Kobayashi N, Niwa T, Kirikoshi H, Fujita K, Yoneda M, Saito S, *et al.* Clinical classification of congenital extrahepatic portosystemic shunts. Hepatol Res 2010;40:585-93.
21. Bruckheimer E, Dagan T, Atar E, Schwartz M, Kachko L, Superina R, *et al.* Staged transcatheter treatment of portal hypoplasia and congenital portosystemic shunts in children. Cardiovasc Intervent Radiol 2013;36:1580-5.
22. Power AH, Bjarnason H. Large spontaneous intrahepatic portal-systemic venous shunt treated with coil and Amplatzer vascular plug embolization. Perspect Vasc Surg Endovasc Ther 2012;24:90-4.
23. Guneyli S, Cinar C, Bozkaya H, Parildar M, Oran I, Akin Y. Successful transcatheter closure of a congenital high-flow portosystemic venous shunt with the Amplatzer vascular plug II. Perspect Vasc Surg Endovasc Ther 2012;24:202-5.
24. Sharma S, Bobhate PR, Sable S, Kumar S, Yadav K, Maheshwari S, *et al.* Abernethy malformation: Single-center experience from India with review of literature. Indian J Gastroenterol 2018;37:359-64.
25. Acherman RJ, Evans WN, Galindo A, Collazos JC, Rothman A, Mayman GA, *et al.* Diagnosis of absent ductus venosus in a population referred for fetal echocardiography: Association with a persistent portosystemic shunt requiring postnatal device occlusion. J Ultrasound Med 2007;26:1077-82.