



Varied Presentations of Absent Ductus Venosus, Their Associations, and Outcomes

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Abstract The present study aimed at identification of fetuses with absent ductus venosus at prenatal screening ultrasound, determination of the type of umbilical venous drainage, and follow-up to assess their outcomes. This prospective study was conducted at CIMAR Hospital, Kerala, in antenatal women with singleton pregnancy, who underwent screening at 11 to 13 + 6 weeks, from November 2013 to January 2015. Ultrasonography was done according to Fetal Medicine Foundation (FMF) protocol. Absence of ductus venosus was diagnosed, when ductus venosus was not visualized connecting the portal sinus to the subphrenic confluence. The type of umbilical venous drainage was determined. Karyotyping was offered in all diagnosed cases and follow-up was obtained. Absent ductus venosus was diagnosed in three out of the 3691 women screened. Two of these had intrahepatic and the third had extrahepatic type of umbilical venous drainage. Karyotype was normal in all the three. The first patient with intrahepatic type of drainage developed polyhydramnios and ended up in intrauterine fetal demise at 26 weeks of gestation. In the second patient, malaligned ventricular septal defect and intestinal obstruction was detected in the anomaly scan. She underwent termination of pregnancy. The fetus with extrahepatic type of umbilical venous drainage developed cardiomegaly on follow-up. This baby underwent ligation of patent ductus arteriosus at third month of life. Our study suggests that absence of ductus venosus is associated with variable fetal outcome. Careful

search of co-existing anomalies and follow-up is warranted in these cases.

Keywords Absent ductus venosus · Umbilical venous drainage · Cardiomegaly · Polyhydramnios · Intrauterine death

Introduction

Ductus venosus is a short vascular channel in the fetal abdomen connecting the intra-abdominal portion of the umbilical vein to the left side of the inferior vena cava. (Fig. 1) It is 5–17 mm long and trumpet shaped, the isthmus measurement increases from 0.5 mm at mid-gestation to 2 mm during the rest of the gestation and outlet width from 1.25 to 3 mm [1–4]. The reduction of diameter produces a pressure gradient between the umbilical vein and the atrium resulting in an acceleration in blood flow in the ductus [1, 2]. This velocity increases with gestational age [1]. It is estimated that 20 %–30 % of the oxygenated blood of the umbilical vein enters the ductus venosus and directly reaches the left heart across the foramen ovale [4]. Ductus venosus plays a major role in the regulation of fetal circulation by modifying the volume of its flow depending on the pressure gradient between the umbilical vein and the heart.

The DV waveform has three phases [1] (Fig. 2):

- S highest velocity peak, corresponding to ventricular systole
- D next peak reflecting early diastole
- a lowest velocity forward flow corresponding to atrial contraction

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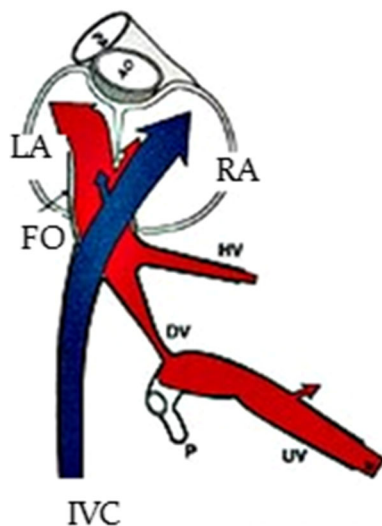


Fig. 1 Oxygenated blood from the umbilical vein (red color) directed through the ductus venosus (DV) and the left hepatic vein preferentially enters the left atrium (LA), while the deoxygenated blood (blue color) from the abdominal portion of the inferior vena cava enters the right atrium (RA) [5]

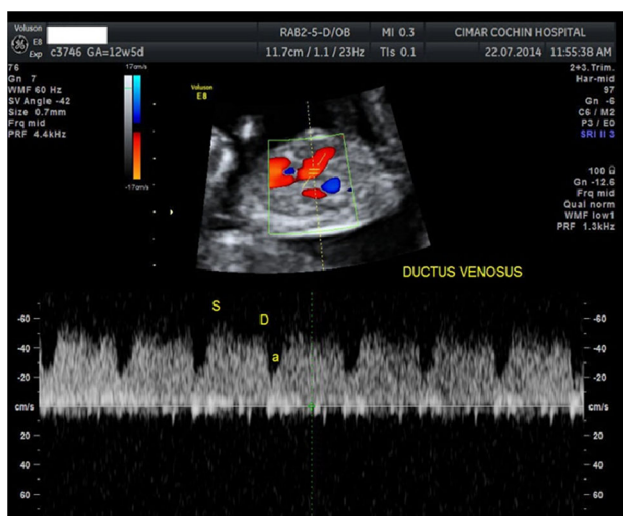


Fig. 2 Doppler assessment of ductus venosus showing the typical triphasic waveform

First trimester assessment of blood flow pattern across the ductus venosus is useful in screening for chromosomal abnormalities and major cardiac defects in the fetus [6, 7]. Absence of ductus venosus is a rare but well-described abnormality (Fig. 3). The incidence of absent ductus venosus in general population is unknown. It has been reported by Staboulidou et al. [8] that the incidence of absent ductus venosus was one in 2532, in a population presenting for the 11 to 13 + 6 weeks scan. Previously, studies have been conducted to assess the pregnancy outcome in absent ductus venosus diagnosed in second and

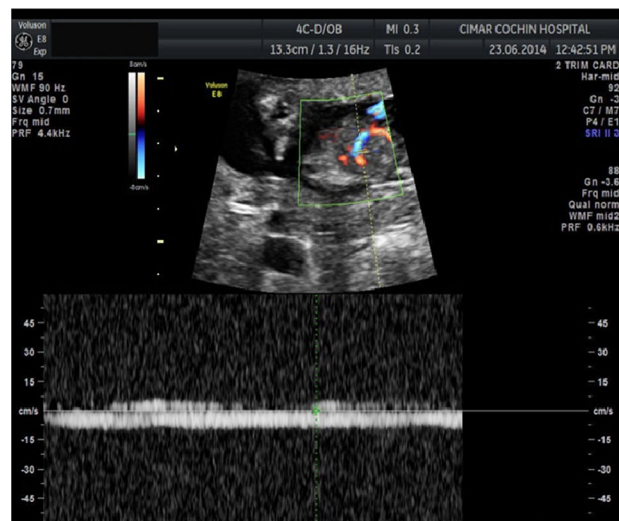


Fig. 3 Ultrasound image showing a case of absent ductus venosus

third trimester [9–14]. Analysis of previously reported cases of agenesis of ductus venosus shows that about 40 % had associated defects and aneuploidies, and in pregnancies with isolated absent ductus venosus, about 35 % resulted in termination or perinatal death [8].

Although many different varieties of abnormal venous circulation were documented, they can be mainly classified into extrahepatic and intrahepatic. In a retrospective study published in 2001, Contratti et al. [11] has mentioned three main patterns:

1. Extrahepatic variety with umbilical vein draining into the inferior vena cava by either one iliac or renal vein.
2. Extrahepatic variety with umbilical vein draining directly to the right atrium.
3. Intrahepatic variety with umbilical vein connecting to the portal circulation without giving rise to ductus venosus.

Materials and Methods

This was a prospective study conducted at CIMAR Hospital, Kerala from November 2013 to January 2015. All antenatal women with singleton pregnancy who underwent screening at 11 to 13 + 6 weeks gestation, crown rump length (CRL) within 45–84 mm, were included in the study. Multiple pregnancies were excluded. History was noted and clinical examination carried out and recorded. Ultrasound scan was performed transabdominally using 3–7.5 MHz curvilinear transducers on GE Voluson E8. Ultrasound was done according to Fetal Medicine Foundation (FMF) protocol and scans were done by sonographers who had obtained FMF certificate of competence in

the 11–13 weeks scan. The examination was undertaken during a period of fetal quiescence. The image was magnified such that the fetal thorax and abdomen occupied the whole screen. A right ventral mid-sagittal view of the fetal trunk was obtained and color flow mapping done to demonstrate the umbilical vein, ductus venosus, and fetal heart. Ductus venosus is identified as a yellowish aliasing area just above the umbilical sinus [15]. The pulsed Doppler sample was kept small (0.5–1.0 mm) and placed in the area of aliasing. The angle of insonation was maintained below 30° and filter frequency set low at 50–70 Hz. The sweep speed was high, around 2–3 cm/s.

If ductus venosus was not visualized connecting the portal sinus to the sub-phrenic confluence, absent ductus venosus was diagnosed. The type of umbilical venous drainage was also determined. These women were counseled, karyotyping was offered in all cases diagnosed, and they were followed up with monthly scans, including a detailed anomaly scan and fetal echocardiography.

Results

During the study period, 3691 women underwent screening. Out of them, three women were diagnosed to have absent ductus venosus (Table 1). One of them had extrahepatic type of umbilical venous drainage with the umbilical vein draining directly into the right atrium. The other two patients had an intrahepatic type of umbilical venous drainage. Karyotyping was done in all the three patients, all of them turned to be normal.

The patient who had a direct drainage of the umbilical vein into the right atrium developed cardiomegaly in the subsequent follow-up at 24 weeks of gestation. She delivered a term female baby of 2.5 kg birth weight. Echocardiography was done postnatally which was normal. This child underwent ligation of the patent ductus arteriosus at third month of life, and is doing well now.

The second patient in our study, who had an intrahepatic type of umbilical venous drainage, was diagnosed to have associated cardiac anomaly—malaligned ventricular septal defect and intestinal obstruction, in her follow-up scan. This patient underwent termination of pregnancy elsewhere.

The third patient who also had an intrahepatic type of umbilical venous drainage was followed up. She did not have any associated structural anomalies. She developed polyhydramnios at 24 weeks of gestation and had intrauterine fetal demise at 26 weeks of gestation. The fetus looked morphologically normal. The result of autopsy is awaited in this case.

Discussion

We were able to identify three positive cases of absent ductus venosus in our study. The incidence in the population studied comes to one in 1230. This is higher than that mentioned in a study by Staboulidou et al. [8] in a similar population. Considering the types of umbilical venous drainage, two types were identified, the intrahepatic type in two cases and in one, extrahepatic with direct drainage of umbilical vein into the right atrium. This is similar to the findings of Berg et al. [13] who suggested that intrahepatic drainage of umbilical vein is more frequent than the extrahepatic type of absent ductus venosus.

We did not identify any chromosomal abnormalities with any of our cases. This was in contrast to findings of Staboulidou et al. [8] who identified a high prevalence of aneuploidies in fetuses with absent ductus venosus.

Similar to the finding in the study by Berg et al. [13] who detected a significant association of cardiomegaly in fetuses with extrahepatic bypass, we identified cardiomegaly in a case of absent ductus venosus with umbilical vein draining directly into the right atrium. Persistence of patent ductus arteriosus in this case may be probably attributed to the cardiac overload and persistent pulmonary hypertension.

The second patient who had an intrahepatic type of absent ductus venosus had an associated cardiac anomaly and gastrointestinal malformation. Cardiac anomalies were the most common structural anomaly associated with absent ductus venosus [12, 13, 16–20].

Hyperdynamic circulation in absence of ductus venosus explains the mechanism behind the polyhydramnios developed in the third patient who had intrahepatic type of umbilical venous drainage. This has been documented in previous studies [11, 16, 17, 21, 22].

Table 1 Ultrasound findings, fetal karyotype, and pregnancy outcome in the cases diagnosed with absent ductus venosus

	Case Outcome	Type	Aneuploidy	Sonographic findings
1	Extrahepatic	Nil	Cardiomegaly	Normal delivery
2	Intrahepatic	Nil	Cardiac, GIT malformations	Termination of pregnancy
3	Intrahepatic	Nil	Polyhydramnios	Intrauterine demise

GIT gastrointestinal tract

Regarding the overall prognosis, the intrahepatic variety, which had associations like polyhydramnios, cardiac abnormality, and gastrointestinal malformation, had bad outcome in terms of intrauterine death and termination of pregnancy. The extrahepatic variety which was associated with cardiomegaly had a good outcome. Berg et al. [13] emphasized the importance of the type of umbilical venous drainage in predicting the outcome, where as Thomas et al. [23] did not support this. The latter concluded that the main determinant in the outcome in fetuses with absent ductus venosus was their associations with structural or chromosomal abnormality and functional cardiac overload. The study suggested that the fetuses with isolated agenesis of ductus venosus had a fairly good outcome. In the present study, it was difficult to draw a conclusion in this respect as we had limited cases only.

Conclusion

The present study suggests that absence of ductus venosus is associated with variable outcome. Determination of the type of umbilical venous drainage may help in prognostication, considering the impacts on fetal hemodynamics in each type. Absence of ductus venosus warrants careful search of associated chromosomal and structural anomalies and these fetuses need serial follow-up examinations.

Compliance with Ethical Standards

Conflict of interest None.

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