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ORIGINAL ARTICLE

Foetal Venous Anomalies: Experience in a Primary Referral Unit

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Abstract Congenital heart disease (CHD) is one of the most common congenital anomalies reported. Incidence of CHD is 8 to 9 per 1000 live births in published data worldwide (Chaudhary and Patil in J Fetal Med 5:221, 2018; Hoffman and Kaplan in J Am Coll Cardiol 39(12):1890–1900, 2002; Khalil et al. in Indian Pediatr 31(5):519–527, 1994; Wanni et al. in Heart India 2(3):76–95, 2014). Association of venous anomalies is known and has a significant impact on the perinatal and postnatal outcome. There is no published data about the incidence and spectrum of foetal venous anomalies in India. We tried to find out the incidence and spectrum of foetal venous anomalies in second and third trimester low risk population during routine ultrasound examination in our primary referral unit. 61 foetuses out of 19,929 were found to have venous anomalies with an incidence of 3 per 1000 s and third trimester pregnancies. Persistent left superior vena cava was the most common anomaly seen followed by umbilical vein varyx and absent ductus venosus. 46% of cases showed associated cardiac and extracardiac anomalies.

Keywords Ultrasound (US) examination · Foetal venous anomalies · Incidence and spectrum · India

Introduction

The foetal venous system has been investigated intensively in the past few years. It plays a very important role in the foetal circulation by transporting oxygenated blood from the placenta to the fetal heart. Any abnormality in development of fetal venous system will hamper the flow of oxygenated blood going to the foetal heart. There are few case reports on abnormalities of the venous system in the literature and their intrauterine detection by ultrasonography. The outcome of the pregnancies and of the child development varies depending on the abnormality and the association with a cardiac defect [1]. Increasing use of high-resolution and color Doppler ultrasonography along with targeted examination of foetal cardiovascular system in routine ultrasonography has enabled the prenatal detection of congenital anomalies of the foetal veins [1].

Few case studies have been recently published which focus on individual veins like persistent left superior vena cava, hepatic or umbilical vein etc [2–4]. However, no antenatal study has been done in India which covers abnormalities of entire foetal venous system.

In this study, we tried to find out antenatal incidence and spectrum of foetal venous anomalies along with their association with intracardiac and extracardiac anomalies in our primary referral unit. This is the first largest antenatal study done ever on a low-risk Indian population.

Materials and Methods

We retrospectively analyzed the data of 19,929 US examinations of pregnant patients in second and third trimester over the period of 5 years and 4 months from January 2014 to April 2019 in our primary referral center. We looked for anomalies of venous system which include

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umbilical vein (UV), ductus venosus (DV), caval veins and pulmonary veins (PV) along with all major and minor cardiac anomalies and extracardiac anomalies. We tried to find out the incidence, spectrum and association of venous anomalies with intra and extracardiac anomalies. Association with chromosomal abnormalities is not included in our study. Venous anomalies were divided into the following groups:

1. Umbilical vein and ductus venosus abnormalities.
2. Caval venous (superior vena cava—SVC and inferior vena cava—IVC) abnormalities.
3. Pulmonary venous abnormalities.

Foetal cardiac evaluation was carried out as per the ISUOG, AIUM, and American Society of echocardiography screening guidelines [5–7].

Systematic cardiac examination with targeted examination of venous system was done in all second and third trimester cases using cardiac preset using gray scale and color imaging using convex probes (C1-4, C1-5 MHz). The endocavitatory (TV 3–9 MHz) and linear (9 MHz and 3–12 MHz) probes were also used if needed. Foetal position preferred was cardiac apex pointing anteriorly and laterally in most of the cases.

The quick cardiac screening was done by taking caudocranial sweep from foetal abdomen to the base of neck to look for situs, position and axis of the heart and for any major deviation from normal. Venous connections were specifically looked for.

Sections documented and studied were—four chamber (4 CH), foetal upper abdomen, left ventricular outflow tract (LVOT), right ventricular outflow tract (RVOT), three vessel trachea (3VT) and confluence of arches. Other sections such as short axis, aortic and pulmonary arches view, longitudinal view and caval view were also documented.

Targeted venous examination included course of the umbilical vein, presence of ductus venosus, position of IVC in upper abdominal section, coronary sinus and pulmonary venous connections in 4CH view, position of SVC in 3VT view, SVC and IVC in longitudinal section and azygos vein in upper mediastinal section. Two dimensional and colour mode examination was done in all sections preferably in dedicated venous color preset. Ductus venosus was also documented on pulsed wave examination. Anomalies of the portal, hepatic veins and left brachiocephalic vein were not included.

Results

We retrospectively analyzed data of 19,929 US examinations of pregnant patients. About 355 cases of foetal cardiac anomalies were diagnosed over the period of 5 years and 4 months (January 2014 to April 2019). This includes all major and minor cardiac abnormalities including venous anomalies. We were able to diagnose venous abnormalities in 61 cases. This gives incidence of 3 per 1000 (second and third trimester) cases. Isolated venous anomalies were seen in 33 cases (54%) and associated intra cardiac and extra cardiac abnormalities were seen in 28 cases (46%). Venous anomalies were termed isolated with respect to their association with intra and extra cardiac defects. Chromosomal association is not included in our study.

Persistent left superior vena cava (PLSVC) (57%) was the most common anomaly found, followed by umbilical vein varyx (UVV) (10%), absent ductus venosus (DV) (10%), total anomalous pulmonary venous connection (TAPVC) (8%), unusual pulmonary vein confluence (2%), portal sinus (PS) thrombosis (2%), isolated left SVC (2%) and persistent right umbilical vein (PRUV) (2%) respectively. Majority of the isolated anomalies were primarily suspected on 4CH view which showed right dominant heart.

The classification of abnormalities is set out in Tables 1, 2, 3.

Table 1 General classification of abnormalities

Type of vein	No. of cases	Percentage	Per 1000 cases
Umbilical vein and ductus venosus	14	22.9	0.7
Caval veins (SVC/IVC)	42	68.8	2.1
Pulmonary veins	5	8.3	0.2

Table 2 Gestational week wise distribution

Gestational weeks	No. of cases	Percentage	Per 1000 cases
12–20	23	37.7	1.1
21–28	21	34.5	1.1
> 28	17	27.8	0.8

Discussion

Structural cardiac anomalies are the most frequently missed anomalies in prenatal US examination [2]. Reported post natal incidence of CHD varies worldwide from 2.5 to 15 per 1000 cases [8–11]. Recently published data on Indian low risk population shows the antenatal incidence of cardiac anomalies up to 8.8 per 1000 s and third trimester cases [8]. Foetal venous system is least studied till recent times due to technical limitations. With the availability of high resolution ultrasound and color Doppler

Table 3 Spectrum of specific abnormalities

Abnormality	No. of cases	Percentage	Per 1000 cases
PLSVC	37	57	1.8
UV varyx	6	10	0.3
Absent DV	6	10	0.3
TAPVC	4	7.5	0.2
Interrupted IVC	4	7.5	0.2
Unusual PV confluence	1	2	0.05
Portal sinus thrombosis	1	2	0.05
PRUV	1	2	0.05
Isolated left SVC	1	2	0.05

equipment, it is now possible to do the detailed evaluation of foetal venous system. There are few studies in literature about the foetal venous anomalies which describe anomalies of individual veins. There are no reported data available about the incidence of foetal venous anomalies in India which covers the entire spectrum. This study was done to assess the prenatal incidence and spectrum of venous anomalies in low risk Indian population referred for routine US examination in our primary referral unit. From our analysis, the incidence of venous anomalies is 3 per 1000 in s and third trimester prenatal US examination which covers anomalies of umbilical, caval and pulmonary veins. If a systematic basic protocol of cardiac screening is followed along with targeted venous examination, most of the spectrum of venous anomalies can be detected in prenatal examination. Important clues to diagnosis or suspicion of venous anomalies are right dominant heart, absence of normal vein, presence of additional vein, dilated but otherwise normally located vein, increased retro cardiac space and fetal hydrops etc.

The diagnostic criteria used for a few commonly seen anomalies are as follow:

- UV varyx—caliber of umbilical vein greater than 9 mm or with a ratio of more than 50% between the dilated and a more distal normal intra-abdominal portion of the vein.
- Absent DV—non-visualization of DV on color and grey scale.
- PLSVC—fourth vessel on left to pulmonary artery in 3VT view and along the left border of left atrium in 4CH view, dilated coronary sinus (normal size 1 to 3 mm and dilated when measures between 3 to 7 mm or more) and right dominant heart.
- TAPVC—increased retro cardiac space, non-demonstrable PV connections with LA, retro-cardiac confluent vein, right dominant heart.
- IVC interruption—non-demonstrable intra-hepatic IVC in upper abdominal cross-section dilated azygos vein which is almost equal in size to the aorta (seen as a double vessel sign in cross section), absence of an IVC connecting to right atrium, hepatic veins joining the right atrium directly.

We found that PLSVC (57%) was the most common venous anomaly in fetus followed by UVV (10%), absent DV (10%) and TAPVC (7.5%) respectively. Most of the PLSVC's were isolated and were picked up easily on 3VT view, especially in late second and third trimester (Fig. 1). Other PLSVC's were associated with other cardiac anomalies like Fallot's tetralogy and transposition of great vessels etc. We found that PLSVC is the second most common anomaly in fetus after minor muscular ventricular septal defect (VSD). All UVV were isolated (Fig. 2). Right dominant heart with prehepatic, subhepatic (Fig. 3) or intrahepatic course of umbilical vein was seen in absent DV. One case of absent DV showed unusual intrahepatic umbilico-caval shunt with DV like waveform at the shunt site (Fig. 4). One case of isolated left SVC was diagnosed at around 32 weeks.

Spectrum of abnormal pulmonary venous drainage comprised of infracardiac (Fig. 5), cardiac and supracardiac venous drainage. In two cases unusual pulmonary venous drainage was seen into left atrium, which showed left pulmonary vein traversing in retrocardiac space to right and then joining the right pulmonary vein subsequently draining in left atrium (Fig. 6).

Other anomalies included interrupted inferior vena cava (7.5%) (Fig. 7) which were associated with situs anomalies like left atrial isomerization, complete situsinversus. One case of isolated interrupted IVC was seen. Portal sinus thrombosis was seen at 14 weeks and presented with foetal hydrops (Fig. 8). 46% of cases were associated with intra and extra cardiac anomalies. Common intracardiac anomalies associated were Fallot's tetralogy, transposition of great vessels and heterotaxies. Extracardiac anomalies associated were as central nervous system, spinal, renal, limb anomalies, two vessel cord and abnormal nuchal translucency (NT) etc. Chromosomal associations could not be studied due to limited resource availability as a primary referral unit. We found that venous anomaly detection rate is more in the early second trimester when they are associated with other intracardiac anomalies and in late second and third trimester especially when they are isolated. Based on these findings systemic venous evaluation is crucial even in late stages of pregnancy.

In our study, we could diagnose most of the spectrum of venous anomalies in the prenatal examination. Anomalies of the portal, hepatic veins and left brachiocephalic vein (LBCV) are not included as visualization of these veins in routine screening is difficult due to limited resolution of equipment used in our centre. Fetal venous anomalies are

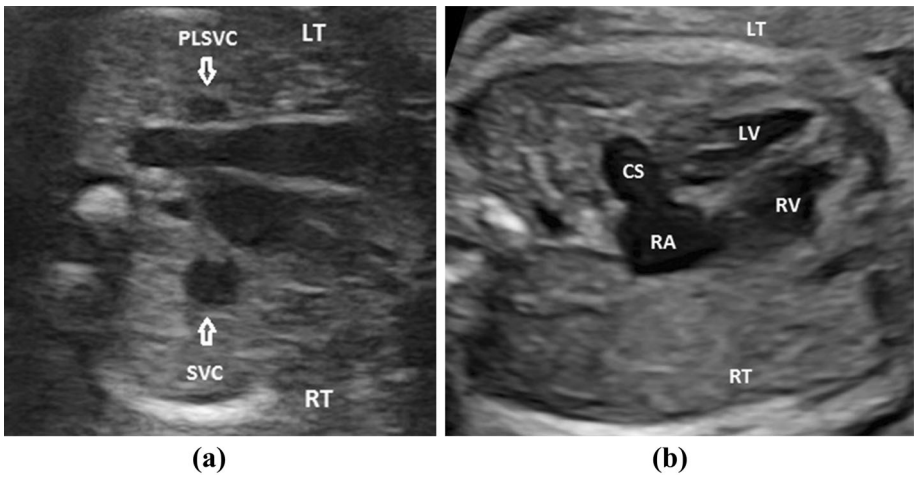

Fig. 1 PLSVC (**a**) and (**b**)

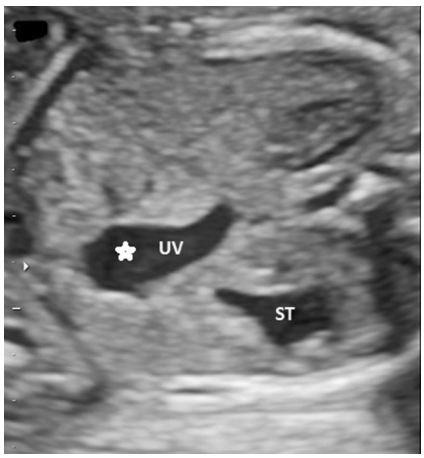

Fig. 2 UV varyx

largely unrecognized and underestimated worldwide and previously thought to be rare. This is mainly because of lack of awareness and unavailability of uniform prenatal diagnostic facilities. Knowing prenatal incidence and exact spectrum of cardiac anomalies along with venous anomalies is very crucial in parental counseling and planning of postnatal treatment. Cases of venous anomalies like PLSVC can be carefully evaluated for associated chromosomal abnormality and intracardiac anomalies as well as these can be followed up in the post natal period for evolving coarctation of aorta as these associations are known [3]. This will definitely increase the postnatal detection rate of coarctation of aorta. If the repairable abnormalities like TAPVC, especially those draining above the diaphragm which have favorable outcomes, are diagnosed in the prenatal period, post natal treatment can be done at earliest saving the crucial time. Cases of interrupted IVC and intrahepatic umbilico-caval shunts may need long term follow ups for their late consequences. In our study post natal long term follow up is not included. To know exact association of PLSVC with coarctation and types of antenatally missed venous anomalies, larger study may be needed with detailed postnatal evaluation and long term follow up.

Our study shows that incidence of foetal venous anomalies in Indian population is significant. The study

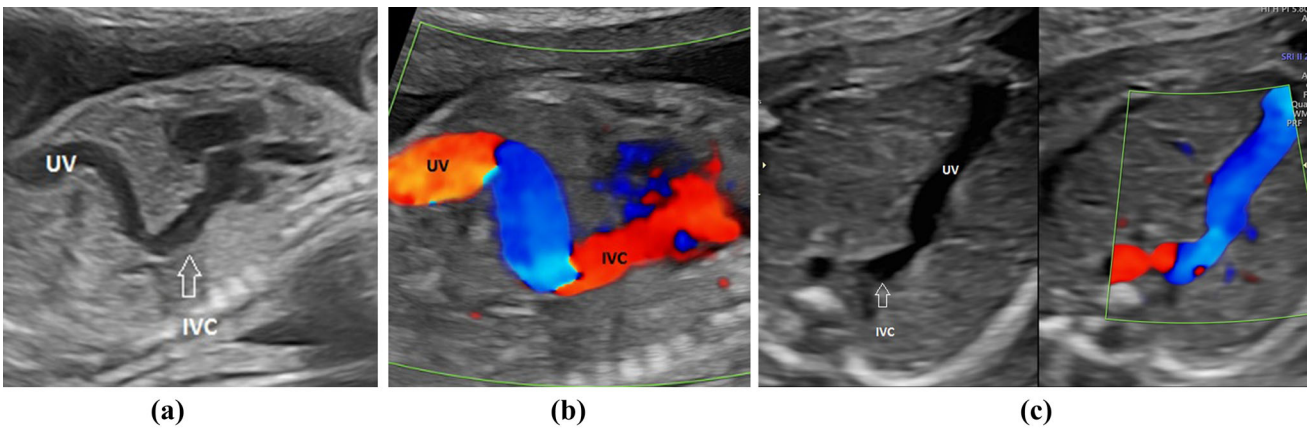

Fig. 3 Absent DV (**a**), (**b**) and (**c**)

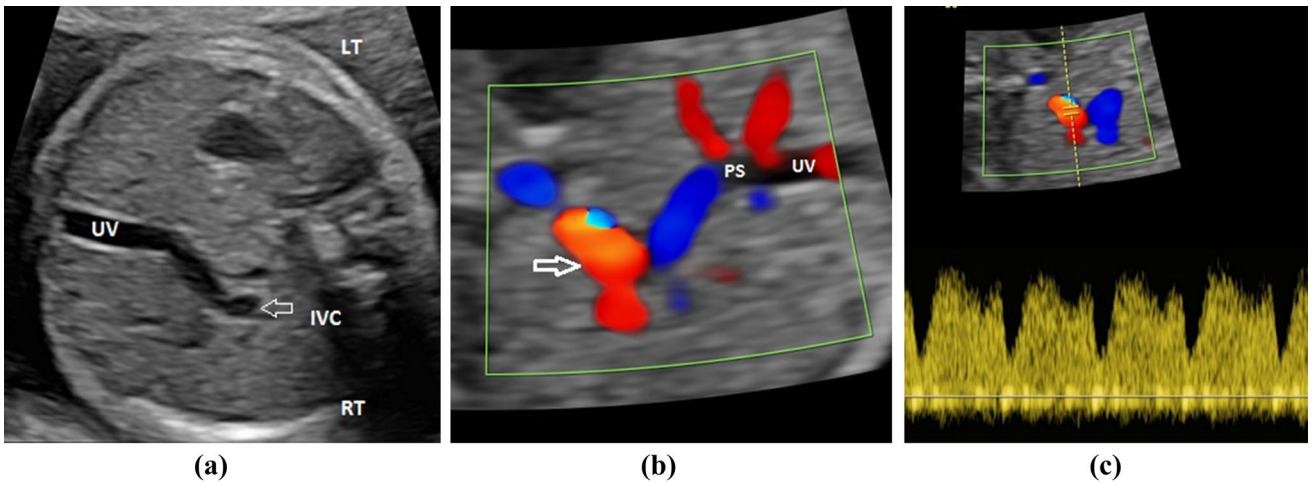

Fig. 4 Intrahepatic umbilico-caval shunt (**a**), (**b**) and (**c**)

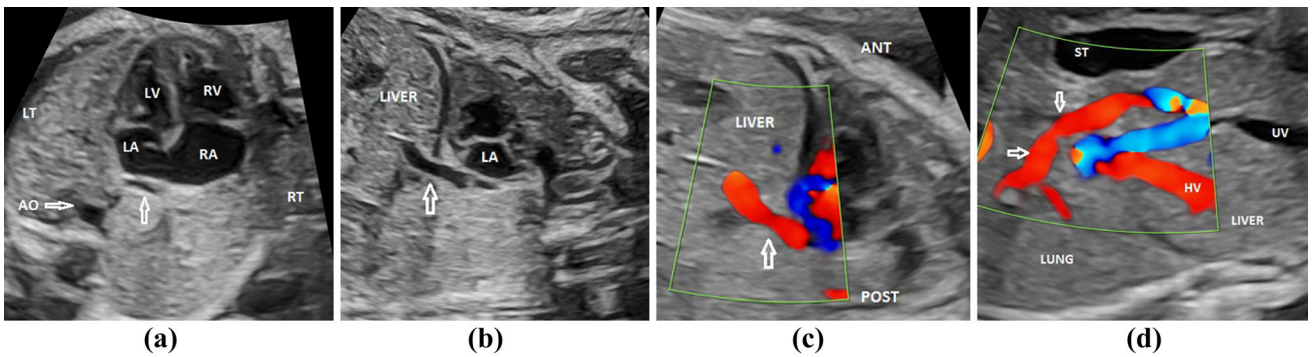

Fig. 5 TAPVC with infra diaphragmatic drainage (**a**), (**b**), (**c**) and (**d**)

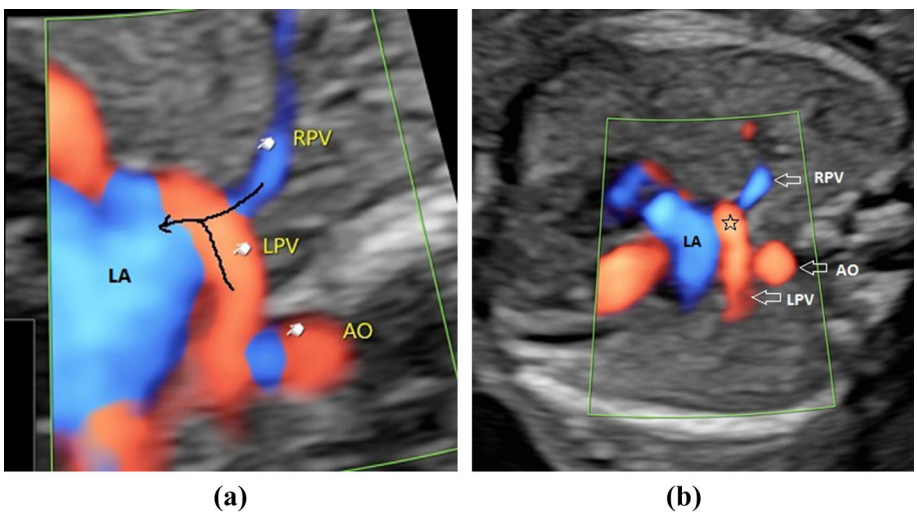

Fig. 6 Unusual pulmonary venous confluence (**a**) and (**b**)

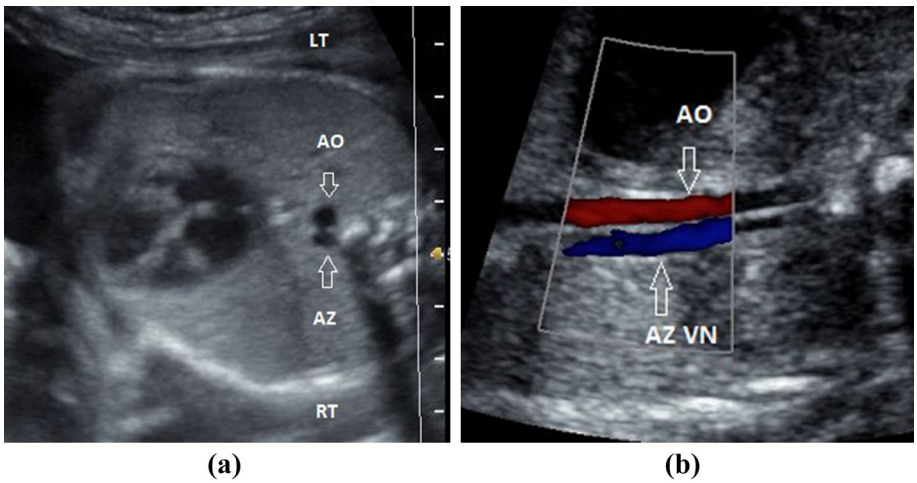

Fig. 7 Interrupted IVC (**a**) and (**b**)

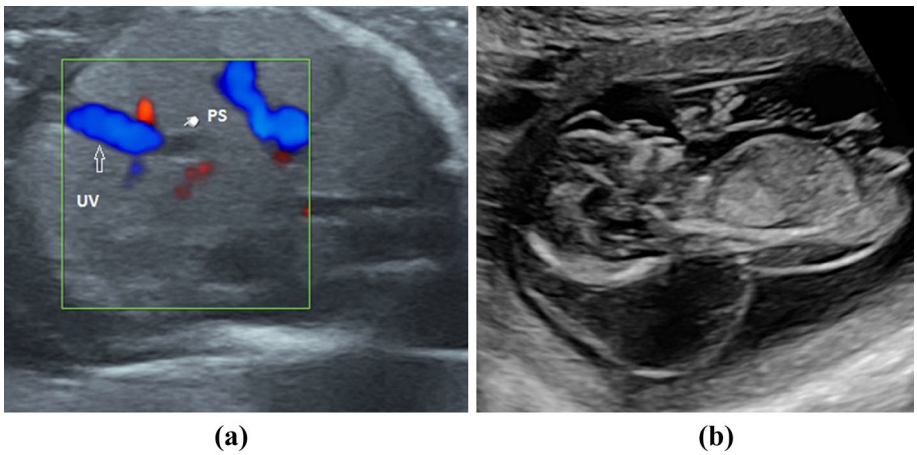

Fig. 8 Portal sinus thrombosis (**a**), foetal hydrops (**b**)

also shows importance of systematic approach to foetal cardiac and venous evaluation. Hence our study will definitely be helpful to imaging specialists and clinicians in India to improve the prenatal diagnosis of venous anomalies.

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