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Prenatal diagnosis of congenital agenesis of the fetal portal venous system

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KEYWORDS: fetal; portal venous system; ultrasound evaluation

ABSTRACT

Objective *To describe the prenatal diagnosis and review our experience of fetal congenital agenesis of the portal venous system (CAPVS) and to review the current literature on this poorly documented vascular malformation.*

Methods *This was a retrospective survey covering the 12-year period between 1996 and 2008. The database of a single, large, ultrasonographic tertiary academic referral center in Israel was analyzed and cases with a prenatal diagnosis of CAPVS were identified. All fetuses underwent detailed biometric and structural ultrasound examinations and a precise anatomical description of the fetal umbilical, portal and hepatic venous system was noted, as well as the presence of aberrant vessels, shunt location and the presence or absence of the DV. Results of fetal echocardiography, karyotyping and toxoplasma, rubella, cytomegalovirus and herpes evaluations were determined. Medical records were evaluated. Diagnosis was confirmed by pathology, postmortem venography or neonatal ultrasound or venography. Liveborns were examined by a certified neonatologist and long-term follow-up from pediatric gastroenterology units was determined.*

Results *Nine cases with CAPVS were studied. In all cases an aberrant umbilical–portal vein was the primary indication for detailed portal system evaluation. Five fetuses demonstrated total CAPVS (Type I) and four showed partial agenesis of the portal vein (Type II). Among the five Type I fetuses, there was a shunt from the umbilical vein to the inferior vena cava in three (60%), to the right atrium in one and to the coronary sinus in one. In this group, in only one case could we delineate a common confluence between the splenic vein and the superior mesenteric vein shunting to the inferior vena cava. In four cases termination of pregnancy was performed due to additional findings: one case with hydrothorax, ascites and mitral atresia, one with cleft lip/palate and one with trisomy 21. One case had no additional anomalies, but the parents elected to terminate the pregnancy. All four of the Type II fetuses had a portosystemic shunt: in two cases to the right atrium, in one to the iliac vein and in one to the right hepatic vein. In three, the shunt resolved spontaneously. In only one case was abnormal liver function present over a follow-up period of 2–10 years.*

Conclusion *CAPVS can be detected prenatally. An abnormal course of the umbilical vein necessitates prompt sonographic evaluation of the umbilical–portal venous system and meticulous investigation for additional anomalies. Complete CAPVS may be associated with remote clinical consequences of which the parents should be informed. Partial CAPVS has a favorable prognosis.*

INTRODUCTION

The fetal umbilical–portal venous system (UPVS) is unusual in being derived from two different embryological precursors, the portal/vitelline and the umbilical veins (UV). It comprises the UV, the portal system and the ductus venosus (DV). The UV conveys oxygenated blood from the placenta. As it courses in a cephalad direction into the liver it joins the left portal vein (LPV), perfusing the left liver lobe and giving rise to the DV, which bypasses

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the liver and shunts blood directly to the fetal heart. At the median fissure of the liver it joins the right portal vein (RPV). At this point the main portal vein (MPV) joins the RPV which carries deoxygenated blood from the spleen and fetal gut, thus supplying the right liver lobe[1].

Sonographic evaluation of the fetal UPVS was described in the early 1980s[2,3]. However, in recent years, meticulous pathological studies[4,5] and high-resolution sonography combined with three-dimensional (3D)-4D applications have advanced our knowledge and ability to identify the normal UPVS and its abnormalities[6–9].

Congenital agenesis of the portal venous system (CAPVS) is a rare anomaly, in which the portal blood bypasses the liver and a splenomesenteric shunt to the systemic circulation is created. Although congenital portosystemic shunt was first described 200 years ago by Aberenthy[10], it was only in 1994 that Morgan and Superina attempted to classify the various types of CAPVS[11]. They proposed two types, each further classified into two subtypes: total agenesis was classified as Type I (classical Aberenthy) abnormality, in which there is complete diversion of the portal blood into the inferior vena cava (portosystemic shunt), and partial agenesis as Type II abnormality, in which the portal vein may exist, but a certain amount of portal blood is diverted into the systemic venous circulation (portohepatic shunt). In Type Ia the splenic vein and the superior mesenteric vein (SMV) do not come together to form a confluence, and thus there is no anatomical portal vein, while in Type Ib, they do form a confluence that may shunt to the inferior vena cava (IVC), renal vein, iliac vein, azygous vein or right atrium. Type IIa was defined as a congenital variant, while Type IIb was defined as an acquired one[11].

During recent years numerous publications have appeared showing tremendous interest in this rare anomaly. However, most describe neonates or adults and were published in pediatric, radiology or pathology journals[12]. A detailed search of the literature yielded only four cases with a prenatal diagnosis of CAPVS[13–16].

The objectives of our study, therefore, were to describe the prenatal diagnosis and review our experience of CAPVS, and to review the literature on this peculiar vascular malformation.

METHODS

In this retrospective survey covering the 12-year period between 1996 and 2008, the database of a single, large, ultrasonographic academic center in Israel was analyzed. An interim analysis of fetuses with central venous and UPVS anomalies was reported by our group in 2006[6]; however, in the present study we included only cases with a diagnosis of CAPVS. In all patients gestational age was determined by last menstrual period and confirmed by first-trimester ultrasound at 6–8 gestational weeks.

Ultrasound examinations during the 12-year study period were performed with a variety of ultrasound equipment: Acuson 128 XP10 (Acuson, Mountain View, CA, USA), ATL HDI 9 (Advanced Technology Laboratories, Bothell, WA, USA) or Synergy (Diasonics, Haifa, Israel)

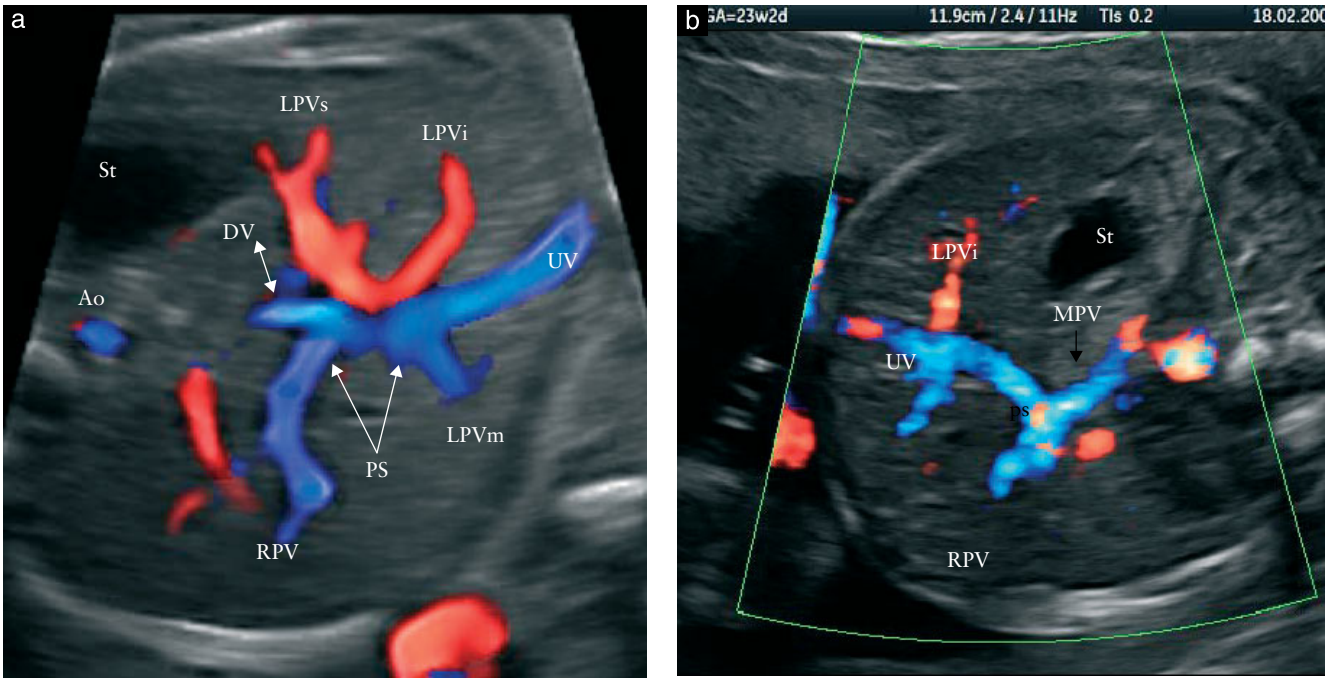

Figure 1 Transverse fetal upper abdominal sections at mid-gestation, showing details of normal umbilical–portal venous system. (a) Three-dimensional ultrasound using render mode with high-definition flow showing how the umbilical vein (UV) joins the left portal vein (LPV), dividing into three branches: the inferior (LPVi) and superior (LPVs) branches, which supply the left lobe, and the medial branch (LPVm). Thereafter the LPV continues as the L-shaped portal sinus (PS and arrows). The ductus venosus (DV) emerges (double-headed arrow) just before the LPV turns to the right to continue as the right portal vein (RPV). In (b) the main portal vein (MPV, arrow) is seen joining the PS from left to right. Ao, aorta; St, stomach.

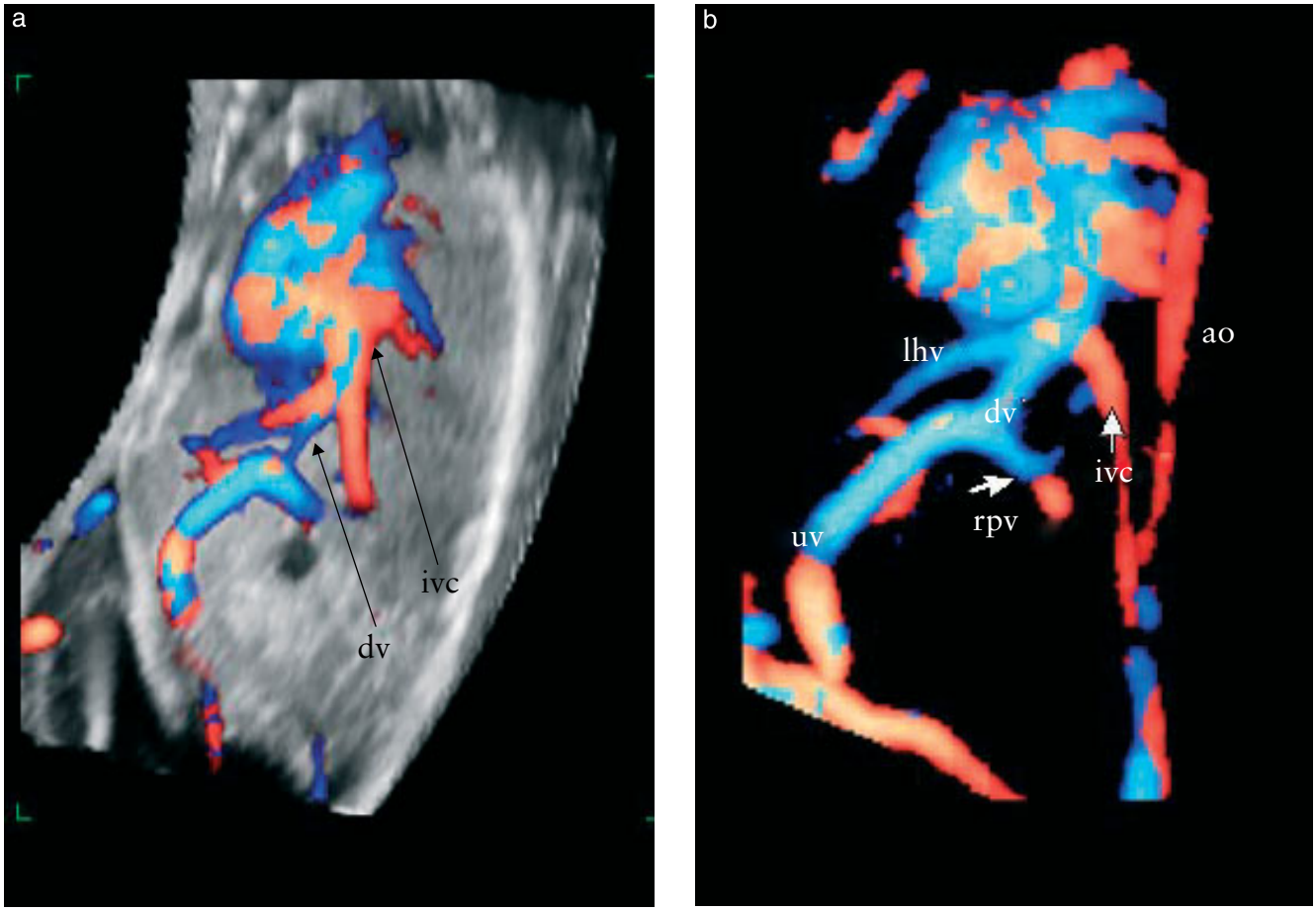

Figure 2 Longitudinal view of the normal fetal umbilical–portal venous system. (a) Three-dimensional ultrasound using glass body render mode with high-definition flow; (b) same image as in (a) but using color Doppler only. Note the separate insertion of the ductus venosus (dv) and the inferior vena cava (ivc) into the right atrium. ao, aorta; lhv, left hepatic vein; rpv, right portal vein; uv, umbilical vein.

systems, equipped with 3.5-, 5.0- or 6.0-MHz transducers, and Voluson 730 Expert or E8 (GE Medical Systems, Zipf, Austria) systems equipped with RIC 5–9-MHz or RAB 4–8-MHz transducers. The study was approved by our institutional review board.

In our center, evaluation of the fetal abdominal veins is an integral part of the routine antenatal anatomical survey. Ultrasonographic assessment of the UPVS is performed at the time of measurement of the fetal abdominal circumference in the standard transverse plane of the fetal upper abdomen, according to the Chinn method[2] and as described recently by our group[1]. Normally at this level the UV courses in a cephalad direction in the inferior margin of the liver. It joins the umbilical portion of the LPV, which is an intrahepatic segment. The inferior branch of the left portal vein (LPVi) then emerges, supplying the inferior segment of the left liver lobe. From this point, the LPV continues as the L-shaped portal sinus that courses abruptly to the right, communicating with the MPV and becoming the RPV (Figure 1). The DV originates from the LPV at the bend in the L-shape of the portal sinus, just before the communication of the LPV with the MPV, which courses from left to right. The DV is also evaluated in all cases in the sagittal fetal view as described by Kiserud *et al.*[17] (Figure 2).

During the 12-year period reviewed, any abnormality of this typical venous configuration was investigated further, in order to classify the anomaly into complete or incomplete type according to Morgan and Superina[11]. Failure to detect any branch of the portal venous system was defined as Type I (total agenesis), and the existence of even a small branch of the portal veins was defined as partial agenesis or Type II CAPVS. For each case we determined a precise anatomical description of the fetal umbilical, portal and hepatic venous system. In addition, the presence of aberrant vessels was investigated with color Doppler sonography, and in the last 4 years with 3D-4D technology and high definition flow (GE Medical Systems). Shunt location was recorded as was the presence or absence of the DV. All fetuses underwent detailed biometric and structural examinations by a single experienced operator in fetal ultrasound (R.A.), and fetal echocardiographic evaluation. Fetal karyotype and toxoplasma, rubella, cytomegalovirus and herpes (TORCH) evaluations were performed. The medical records comprising reports, prints, videotapes of scans and computerized files were evaluated. Diagnosis was confirmed by pathology in two cases, postmortem venography in two, and neonatal imaging, ultrasound or venography in five cases. All live newborns were examined

Table 1 Summary of nine cases of fetal congenital agenesis of the portal venous system (CAPVS)

Case	MA (years)	GA (weeks)	Indication for ultrasound	DV	Type of shunt	Type of CAPVS*	Additional ultrasound findings	Outcome
1	27	22	Hydrothorax	No	UV–IVC	Total I	Hydrothorax	TOP
2	29	24	Ascites	No	UV–CS	Total I	No IVC, rudimentary left cardinal vein, mitral atresia	TOP
3	31	25	Cleft lip	No	UV–IVC	Total Ib	Cleft lip	TOP
4	36	14	Routine	No	UV–IVC	Total I	NT 4 mm	TOP, Down syndrome
5	30	22	Routine	No	UV–RA	Total I	None	TOP
6	31	22	Routine	Yes	RPV–HV	Partial IIa	None	A&W, portohepatic shunt resolved spontaneously
7	30	15	Routine	Yes	UV–RA	Partial IIa	Liver echogenicities	A&W, LPV obliteration
8	33	16	Routine	Yes	UV–RA	Partial IIa	Liver echogenicities	A&W, LPV obliteration
9	28	22	Enlarged IVC	No	UV–iliac vein	Partial IIa	Enlarged IVC	Mild hepatic dysfunction, elevated ammonia, portal hypertension

*Following definitions of and adapted from Morgan and Superina[11]. A&W, alive and well; CS, coronary sinus; DV, ductus venosus; GA, gestational age; HV, hepatic vein; IVC, inferior vena cava; LPV, left portal vein; MA, maternal age; NT, nuchal thickness; RA, right atrium; RPV, right portal vein; TOP, termination of pregnancy; UV, umbilical vein.

Figure 3 Abnormal insertion of the umbilical vein (UV) into the inferior vena cava (IVC) in Case 3. (a) Three-dimensional ultrasound using glass body render mode with high-definition flow; (b) same image as in (a) but using color mode only. In (c), note the normal insertion of the UV into the right atrium (arrow). Absence of the ductus venosus is evident from the lack of high velocity flow in (a) and (b).

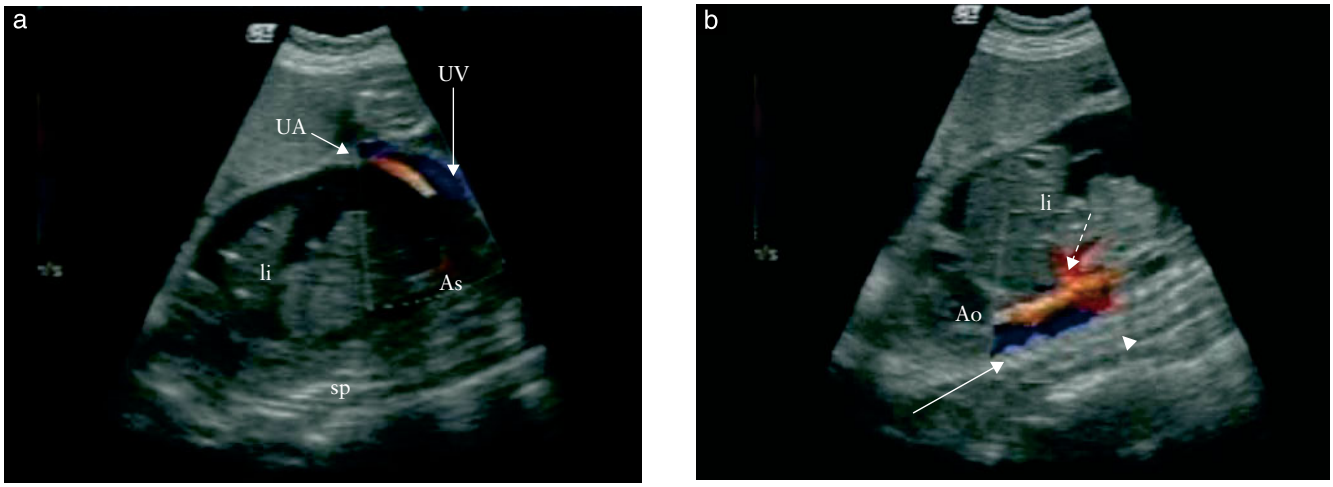

Figure 4 Longitudinal section of Case 2 (with head to the left and spine (sp) down) showing: (a) the umbilical vein (UV) coursing caudally (blue color and arrow) instead of in the normal cephalad direction. The umbilical artery (UA) is seen (red color) flowing towards the transducer. As, ascites; li, liver. (b) The UV is seen advancing towards the heart (blue color; solid arrow) beneath the aorta (Ao; red color) and the spine (arrowhead). Note liver (li) perfusion is mainly from the systemic arterial circulation (dashed arrow). (c) Venography post-termination confirming the unusual channel (supposedly the rudimentary cardinal vein) communicating between the UV and the left atrium. Note the poorly perfused liver with neither umbilical–portal venous system nor inferior vena cava.

by a certified neonatologist and long-term follow-up from pediatric gastroenterology units was determined. A detailed literature search in the PubMed bibliographic database using the terms 'fetal', 'prenatal', 'congenital', 'absence/agenesis', 'portal vein' and 'portosystemic shunt' was undertaken, and the relevant publications were reviewed.

RESULTS

We were able to retrieve from the database nine fetuses with a prenatal diagnosis of CAPVS made during the period between 1996 and 2008. The various details of the patients are summarized in Table 1. The average gestational age at diagnosis was 20.2 (range, 14–25) weeks, and the earliest gestational age at which it was first suspected was 14 weeks.

According to the Morgan and Superina classification[11], five fetuses (Cases 1–5) showed total agenesis (Type I), and four (Cases 6–9) had partial agenesis (Type II). All nine fetuses demonstrated an abnormal UPVS connection manifested by an aberrant shunt from the UV to the systemic venous circulation.

There was, however, some variation in the UV drainage site. Among the five Type I cases, with total agenesis of the portal system, Cases 1, 3 and 4 showed direct communication of the UV with the intrahepatic portion of the IVC (Figure 3). In Case 2 there was total absence of the UV, portal vein and IVC, replaced by an unusual vessel which coursed from the umbilical ring caudally and to the left, continuing dorsally beyond the aorta to join the coronary sinus (Figure 4). This vessel was hypothesized to be a rudimentary cardinal vein. In Case 5 the UV shunted directly to the right atrium (Figure 5). None of the cases with Type I anomaly had a DV, and in all cases prominent hepatic artery replaced poor and scant venous perfusion of the liver (Figure 5). In only one fetus of this type (Case 3) could be identified by way of pulsed Doppler a systemic, triphasic, venous pattern in the MPV (Figure 6), thus indicating a total portosytemic shunt (Type Ib). This was confirmed (Figure 7) by venography performed post-termination, when the splenic vein was found to form a confluence with the renal vein, and the SMV to form one conduit that bypassed the liver and shunted directly into the IVC.

Among the five Type I fetuses, four (80%) showed additional anomalies: hydrothorax (Case 1), ascites and mitral atresia (Case 2), cleft lip (Case 3) and trisomy 21 with increased nuchal translucency (Case 4). Only one case (Case 5) did not show associated anomalies. In all cases a multidisciplinary team counseled the parents. In the four Type I cases with additional anomalies the parents opted to terminate the pregnancy for this reason.

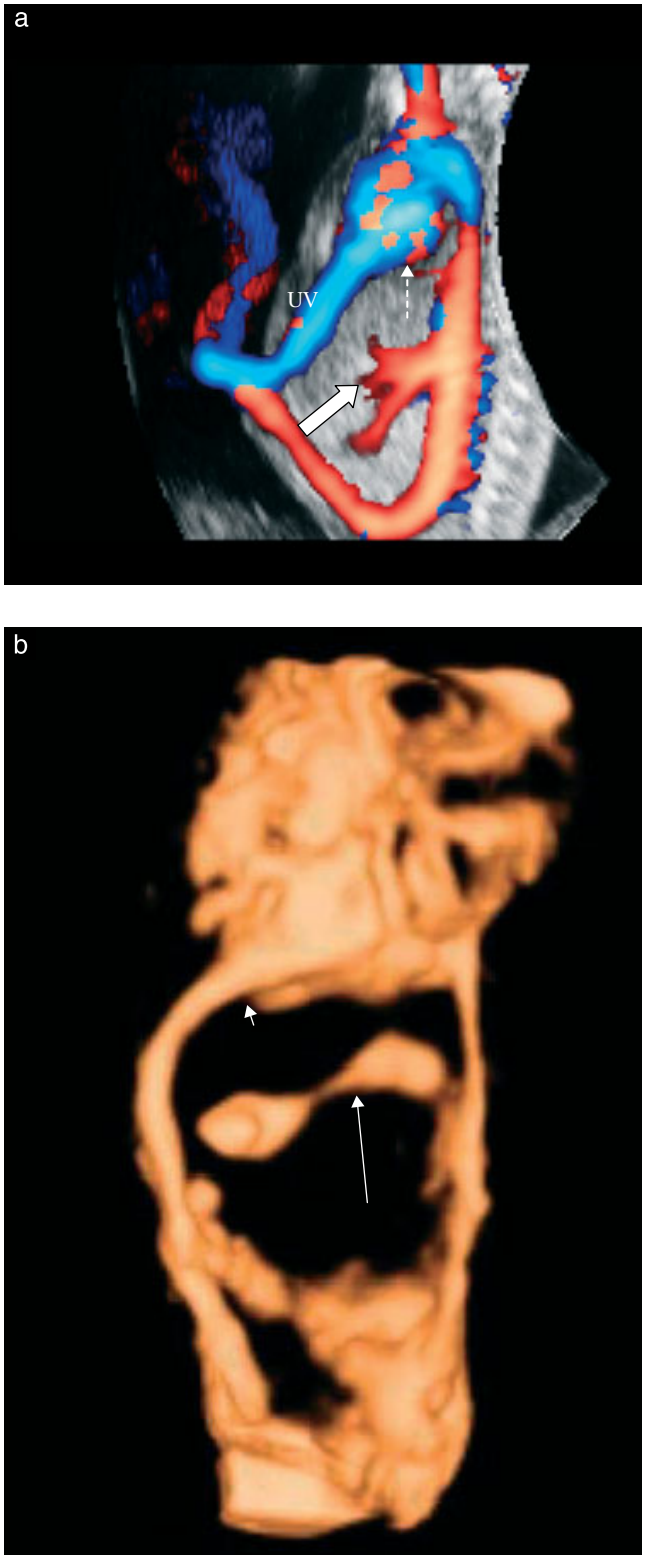

Figure 5 (a) Transvaginal three-dimensional ultrasound image (longitudinal view) at 14 weeks' gestation (Case 5) using glass body render mode and high-definition flow, demonstrating abnormal insertion of the umbilical vein (UV) (blue color) into the right atrium (dashed arrow). Note the prominent celiac and hepatic artery (solid arrow) without afferent liver venous perfusion. (b) The same fetus as in (a) before termination at 24 weeks, visualized using inversion mode and confirming the aberrant insertion of the UV (short arrow) and the prominent hepatic artery (long arrow).

Case 5 was terminated at the request of the parents due to the gastroenterologist's inability to predict prognosis (which, according to the literature, includes significant risk for consequences which may not manifest until much later in life). Pathological examination was performed in two cases (Cases 2 and 3), in both of which was found an abnormal portal space consisting of prominent branches of the hepatic arteries with scant, inconspicuous venous channels representing poor portal venous perfusion of the liver (Figure 8).

Among the four Type II cases, with partial agenesis of the portal system, Case 6 had absence of the RPV detected *in utero* and a portohepatic shunt was verified by postnatal ultrasound (Type IIa). Case 7 showed direct communication of the UV to the right atrium with intact DV. Since we detected small echogenic foci in the liver in this case, we assumed it to be Type IIb (acquired). Both cases were included in our previous publication[6]. However, a few years later the same woman (of Case 7) presented with another pregnancy (Case 8) that showed the same features as those of her previous one; therefore, the classification was changed to Type IIa (congenital) in both cases. In Case 9, following detection of an enlarged IVC at routine examination, an abnormal UV was detected shunting to the left iliac vein (Figure 9). Since *in-utero* evaluation revealed some remnants of the RPV it was classified as Type IIa. The neonate's course was uneventful; however, due to elevated levels of postprandial ammonia, venography was performed 3 weeks after delivery, verifying splenorenal shunt with retrograde flow from the portal vein to the shunt, suggesting a hypoplastic or absent portal system. At the 2-year follow-up the infant's development was reasonable, except for a high level of ammonia that necessitated a dietary regime. In all Type II cases the visible portal branches showed a normal monophasic flow pattern on Doppler examination.

All four Type II fetuses were liveborn and none had additional anomalies. In Case 6 the left portohepatic shunt had resolved spontaneously by the 6-month follow-up. Postnatal ultrasonography confirmed the diagnosis of LPV obliteration in Cases 7 and 8. Both children at ages 9 and 11 years, respectively, at the time of writing, were alive and healthy. Only one of this group showed hepatic dysfunction (Case 9), observed in a neonate with abnormal ammonia level at 3 days of age.

DISCUSSION

Our study has shown, for the first time, that CAPVS can be diagnosed confidently *in utero* and confirmed by various postnatal imaging modalities. However, in order to understand this rare venous anomaly, it is essential to review the embryological development of the portal system[18].

Although anomalies of the UV have been reported widely[6,9], anomalies of the vitelline veins are extremely rare, and there have been only a few reports in the fetus. Complete agenesis of the fetal portal system is an extreme example of total failure of the vitelline

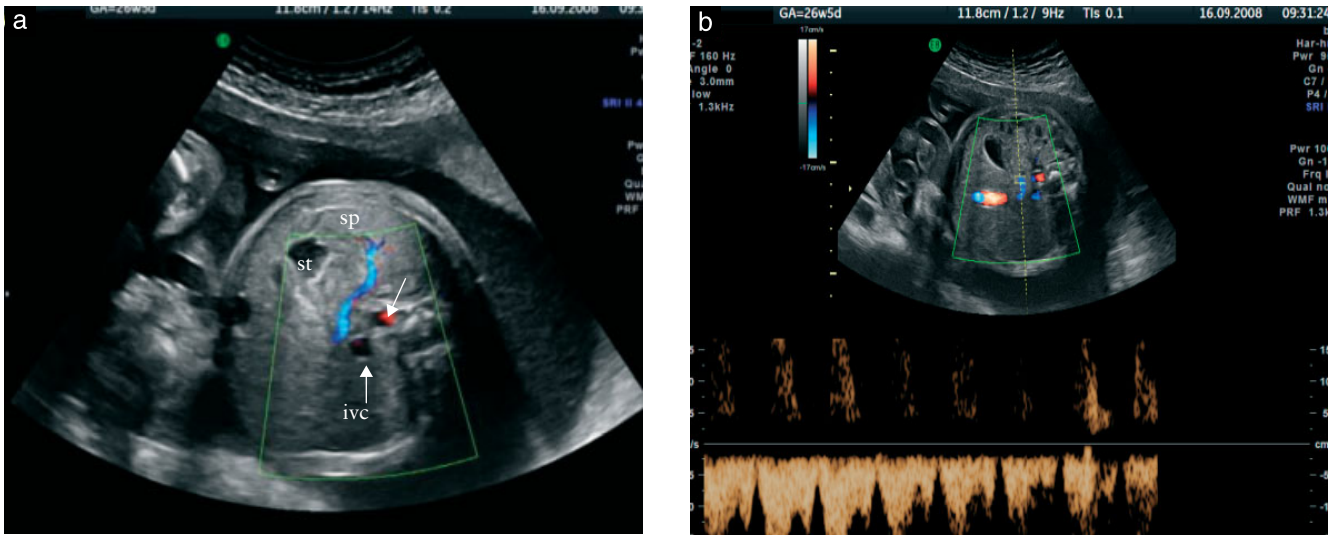

Figure 6 (a) Transverse view of the fetal abdomen (Case 3) showing the spleen (sp) beyond the stomach (st), with the splenic vein coursing towards the right side of the body (blue color) to form a confluence with the main portal vein. Note the absence of the afferent intrahepatic venous system and the enlarged inferior vena cava (ivc, arrow) when compared with the aorta (red color, arrow). (b) Pulsed Doppler insonation of the splenic vein–portal system junction depicting triphasic flow compatible with a systemic venous shunt.

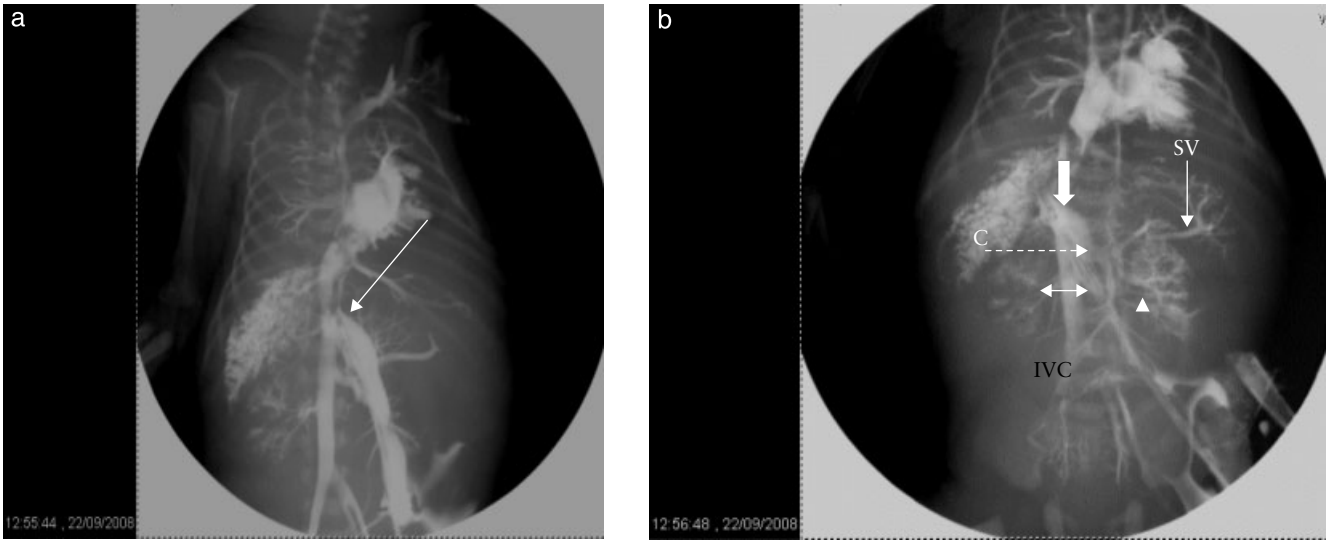

Figure 7 Venography post-termination in Case 3. (a) Note the umbilico-inferior vena cava (IVC) shunt (arrow). One minute later (b) the splenic vein (SV, thin solid arrow) formed a confluence with the renal vein (small arrow) and with the superior mesenteric vein (double-headed arrow) to form a conduit (C, dashed arrow) which became enlarged (thick white arrow) before shunting into the IVC.

veins to transform into the portal system, i.e. there is a primary failure to form the anastomosis with the hepatic sinusoids or with the UV. The most convincing embryological explanation for this rare anomaly was offered by White *et al.*, who introduced the term 'critical anastomosis'[19]. They speculated that through unknown causes of embryological maldevelopment, anastomosis between the UV and vitelline veins fails to form and resorption in early embryonic life of the UV occurs. As a result, the umbilical flow and the enterohepatic circulation are disturbed, and the portal as well as the placental venous blood is shunted systematically. The complex development of the vena cava and its close relationship with the vitelline veins likely explains the occurrence of the extrahepatic portosystemic shunt in cases with CAPVS.

Smith[20] showed that in a 7-mm pig embryo there is a connection between the primitive pelvic venous plexus that drains the lower limb bud and the embryonic UV. It is therefore not surprising that in all our cases in which primary failure to form a critical anastomosis occurred, the placental venous return was rerouted to the IVC, right atrium and iliac vein. We have shown that although the UV had various insertion sites, a common feature of the Type I group in our study was agenesis of the DV. From an embryological point of view, agenesis of the DV under these circumstances is part of a more fundamental maldevelopment process. However, it is most interesting that fetal agenesis of the DV has been reported extensively in the literature[21–23], while CAPVS has been reported in only four cases[13–16]. This discrepancy indicates that

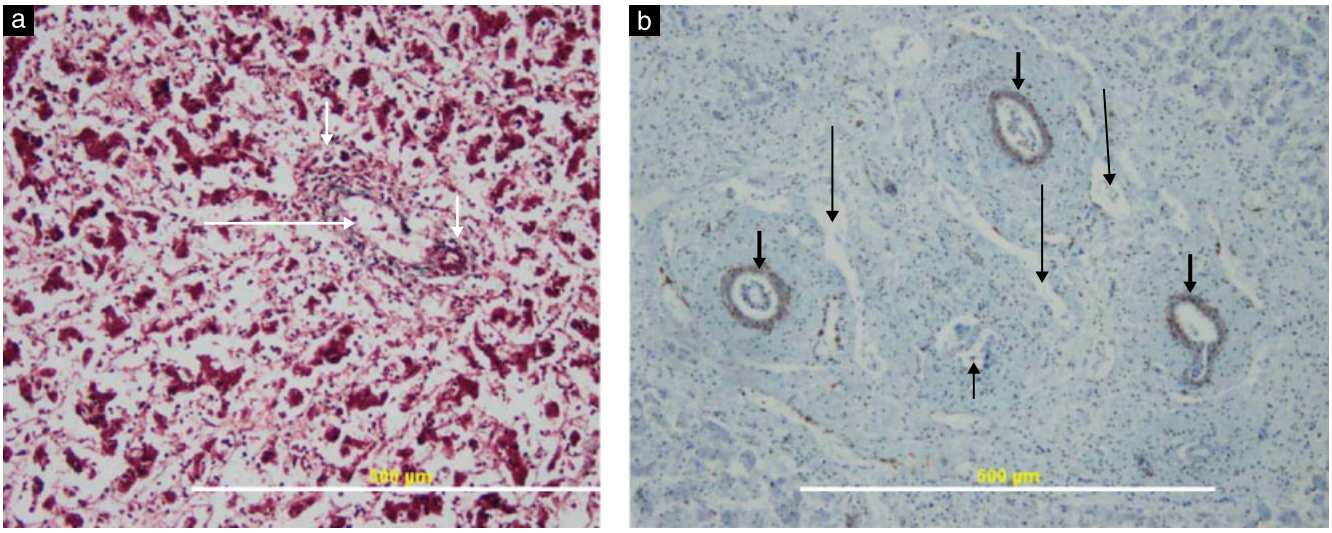

Figure 8 (a) Microscopic examination of normal portal space (a) showing a prominent, centrally located portal vein (long arrow), peripherally placed hepatic artery (thick arrow) and bile duct (short arrow). (b) Abnormally large portal space in Case 3, consisting of several prominent, large branches of thick-walled hepatic arteries (short, thick arrows), a relatively small bile duct (short thin arrow) and a dilated, inconspicuous venous channel (long thin arrows). Immunohistochemical stain for actin, original magnification, ×100.

CAPVS is a cryptic condition and therefore should be explored further.

The recent review of agenesis of the DV by Berg *et al.* in 2006[24] accumulated a total of 42 cases, including those of previous series[21–23], and careful inspection of the data revealed that nine of 20 (45%) cases that had extrahepatic and one of 13 (33%) that had intrahepatic UV drainage suffered from portal vein agenesis. However, Berg *et al.*[24] did not clarify the clinical significance and outcome in fetuses with portal vein agenesis mainly because their emphasis was on cardiac function rather than hepatic follow-up[25].

Thus, in an attempt to better understand the clinical significance of CAPVS, since it represents a developmental abnormality of the liver we reviewed the pediatric and gastroenterology literature. Morgan and Superina's[11] classification of CAPVS into two types was as a result of their observations from pediatric surgery. Their classification into complete and partial agenesis, and particularly into subtypes Ia and Ib, is of enormous significance if patients with CAPVS require liver transplantation. In our series the outcome of CAPVS was clearly associated with this typing. In Type I, 80% of our cases had additional anomalies and in all cases the parents chose to terminate the pregnancy, while in Type II all delivered and all except one with mild liver dysfunction were healthy over a long-term follow-up period which ranged between 2 and 10 years. It is obvious that those who present with Type I and with additional anomalies such as Down syndrome and severe hydrops are likely to terminate the pregnancy. However, in cases without additional anomalies counseling is far more difficult, due mainly to the limited data in the literature.

Regarding the four previously reported cases of complete CAPVS diagnosed prenatally, all had a good outcome[13–16]. However, one should bear in mind that most of the clinical consequences of CAPVS with systemic shunt may not manifest until much later in life. These include hypergalactosemia[26], hyperammonemia[27] and increased incidence of hepatoma, nodular hyperplasia and hepatocellular carcinoma[28,29]. The parents should be informed about the possibility of future complications and the various treatment options. If there is severe liver failure, transplantation will be needed. In mild liver dysfunction, as in our Case 9, only a low-protein diet is indicated.

Of interest in this rare anomaly is the scarcity of portosystemic encephalopathy which is common among patients with hyperammonemia. It was found that patients with CAPVS have markedly low blood ammonia in the SMV. This is as a result of decreased urease activity in their intestinal bacterial flora, which appears to function as a homeostatic mechanism[30].

In contrast to those with total CAPVS, we found that those with partial CAPVS may have an almost normal outcome; they should therefore be encouraged to continue with the pregnancy. However, meticulous fetal observation is needed in order to detect early hemodynamic deterioration, the neonate should be investigated for postprandial ammonia levels, and ultrasound is mandatory for confirming the diagnosis. Venography should be considered by the pediatric gastroenterologist to verify the shunt and in some cases occlusion of the shunt may be needed.

In conclusion, it is likely that CAPVS occurs more frequently than was previously thought. Fetuses with abnormal insertion of the UV should be investigated meticulously for presence of the DV and the UPVS should be explored. Doppler ultrasound is very useful for the detection of systemic venous flow patterns in the shunt. If a triphasic systemic venous flow pattern exists in the spleen or SMV, a portosystemic shunt is evident. Any remnant of portal vein branches indicates partial CAPVS, which carries a favorable prognosis. Additional

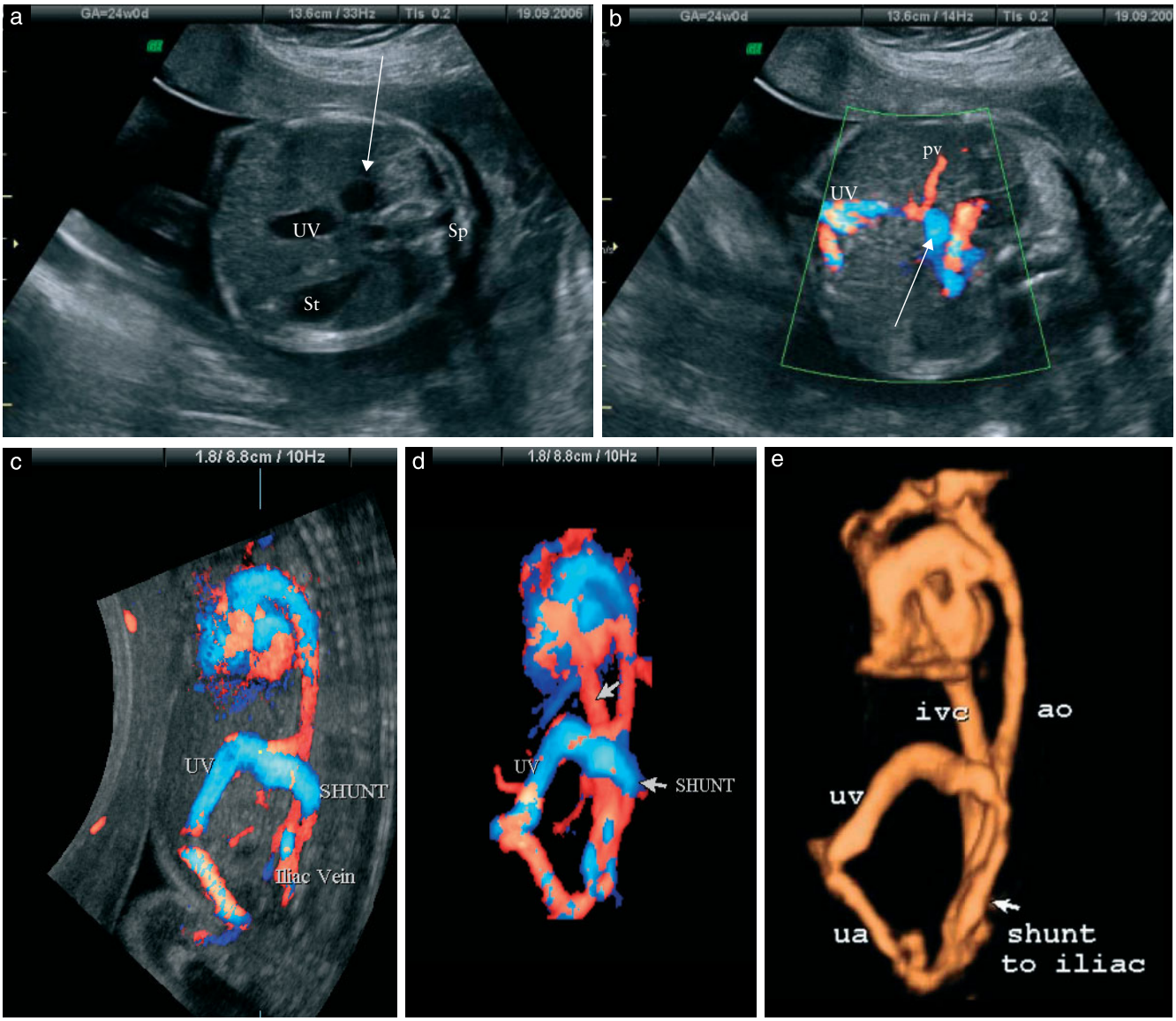

Figure 9 Partial congenital agenesis of the portal venous system (CAPVS) Type IIa (Case 9) at 24 weeks' gestation. (a) Transverse abdominal scan showing enlarged inferior vena cava (arrow). Note the large segment of the umbilical vein (UV). Sp, spleen; St, stomach. (b) Same section as in (a), with high-definition flow showing part of the right portal vein (PV, red color) perfusing the right liver lobe and the abnormal UV coursing to the left (arrow, blue color) to join the lower plane of the iliac vein. (c–e) Sagittal views; (c) demonstrates the level of the shunt. (d) Three-dimensional image using render mode with high definition flow; note the enlarged inferior vena cava (ivc) (upper arrow). (e) Three-dimensional image using inversion mode demonstrating the umbilico-iliac shunt. Ao, aorta; ua, umbilical artery.

structural anomalies, as well as chromosomal aberrations, should be ruled out[23]. Parents of fetuses with complete CAPVS should be informed about the possibility of future clinical consequences, and if termination of pregnancy is performed, postmortem angiography can be used to confirm the diagnosis.

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