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Partial and total anomalous pulmonary venous connection in the fetus: two-dimensional and Doppler echocardiographic findings

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KEYWORDS: anomalous pulmonary venous connection; fetal echocardiography; pulmonary venous flow pattern

ABSTRACT

Objective *Prenatal diagnosis of total (TAPVC) or partial (PAPVC) anomalous pulmonary venous connection in isolation or associated with other cardiac disease is important for appropriate prenatal counseling and perinatal management. We sought to assess the echocardiographic clues to the fetal diagnosis of TAPVC and PAPVC in a cohort of affected fetuses.*

Methods *We retrospectively reviewed 29 fetal echocardiograms performed in 16 pregnancies with fetal TAPVC or PAPVC, systematically analyzing heart chamber size, presence of a confluence behind the left atrium or of a vertical vein, and Doppler flow patterns.*

Results *Prenatal diagnosis was made at a mean gestational age of 27 ± 7 weeks. TAPVC was found in 11 cases; five cases for each of supracardiac and infracardiac types and one mixed type. PAPVC was diagnosed in five fetuses, four of which had scimitar syndrome. Ten fetuses had an additional major cardiac defect, including hypoplastic left heart syndrome and right atrial isomerism. In three cases the prenatal diagnosis was only made at follow-up assessment. Among TAPVC cases, visualization of a confluence behind the left atrium (10/11) and a vertical vein (11/11) were the most consistent echocardiographic clues. Dextrocardia and a small right pulmonary artery suggested scimitar syndrome. The diagnosis was confirmed postnatally or at autopsy in 12 cases. In six fetuses with TAPVC and obstruction confirmed postnatally, continuous turbulent flow in the vertical vein and monophasic continuous flow in the pulmonary veins were demonstrated by color and spectral Doppler.*

Conclusions *Fetal echocardiography permits prenatal diagnosis of TAPVC or PAPVC. Spectral and color Doppler provide clues to the presence of an obstructed pulmonary venous pathway. Copyright © 2003 ISUOG. Published by John Wiley & Sons, Ltd.*

INTRODUCTION

Both total (TAPVC) and partial (PAPVC) anomalous pulmonary venous connections may occur in isolation or in combination with other cardiac defects. Their clinical presentation depends on number of pulmonary veins involved, site of anomalous connection, presence of pulmonary venous (PV) obstruction and additional cardiac defects[1]. When TAPVC is complicated by PV obstruction, the patient's condition can deteriorate rapidly after birth, with severe respiratory distress and cyanosis. As its clinical signs and echocardiographic findings are similar to those observed in pulmonary hypertension, obstructed TAPVC may escape correct diagnosis[2]. Isolated TAPVC, if detected and corrected surgically early in life, has an excellent outcome[3,4], but the prognosis is poor when TAPVC is associated with other cardiac lesions, such as right atrial isomerism[5] and PV obstruction[6].

Prenatal diagnosis of an anomalous PV connection with appropriate perinatal management may allow neonatal transfer and early decompensation to be avoided, facilitate surgical planning, and enable accurate counseling of the family. Prenatal parental counseling is particularly important when the condition is associated with a complex congenital heart defect.

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Table 1 Fetal diagnoses, clinical profiles and outcomes

Case	GA at diagnosis	Pulmonary venous connection site	Obstruction	Associated CHD	Extracardiac malformations	Diagnosis at postnatal echo* or autopsy†	Outcome
1	NA	TAPVC infracardiac	None	None	None	Confirmed*	Neonatal death
2	32	TAPVC infracardiac	None	None	Microcephaly, microphthalmia, translocation 16/22	NA	TOP
3	29	TAPVC innominate vein	Suspected	RAI, AVSD, PA, bilateral SVC	None	Confirmed*	Died aged 7 months
4	18	TAPVC azygous vein, LSVC	Suspected	RAI, AVSD, PA, right aortic arch	None	Confirmed*	Neonatal death
5	18	TAPVC mixed: LSVC and innominate vein	Suspected	RAI, AVSD, DORV	None	NA	TOP
6	19	TAPVC infracardiac	None	RAI, VSD, PA, bilateral SVC	None	Confirmed†	TOP
7	34	TAPVC infracardiac	None	RAI, DORV, VSD, PS	VACTERL association	Confirmed*	Died aged 3 months
8	30	TAPVC LSVC	Suspected	RAI, AVSD, PA	None	NA	TOP
9	39	TAPVC innominate vein	None	RAI, AVSD, DORV, PS, bilateral SVC	None	Confirmed*	Alive
10	38	TAPVC infracardiac	Suspected	RAI, AVSD, DORV, PA	None	Confirmed*	Neonatal death
11	30	TAPVC SVC	Suspected	HLHS	None	Confirmed*	Neonatal death
12	25	PAPVC innominate vein	None	HLHS	None	Confirmed*	Died aged 1 month
13	19	PAPVC IVC/RA junction	None	Scimitar syndrome	None	Confirmed†	TOP
14	26	PAPVC IVC/RA junction	None	Scimitar syndrome	Right CDH	Confirmed*	Died aged 8 months
15	20	PAPVC IVC/RA junction	None	Scimitar syndrome	Vertebral, rib anomalies, hypoplastic right kidney	Confirmed†	TOP
16	26	PAPVC IVC/RA junction	None	Scimitar syndrome	None	NA	TOP

AVSD, atrioventricular septal defect; CDH, congenital diaphragmatic hernia; CHD, congenital heart defect; DORV, double-outlet right ventricle; GA, gestational age; HLHS, hypoplastic left heart syndrome; IVC, inferior vena cava; LSVC, left-sided superior vena cava; NA, not available; PAPVC, partial anomalous pulmonary venous connection; PA, pulmonary atresia; PS, pulmonary stenosis; RA, right atrium; RAI, right atrial isomerism; SVC, superior vena cava; TAPVC, total anomalous pulmonary venous connection; TOP, termination of pregnancy; VSD, ventricular septal defect.

Only a few papers, mostly case reports, have described prenatal diagnosis of an anomalous PV connection[7–11]. The low incidence of PV anomalies in fetal series may reflect the difficulty in detecting them *in utero*[12]. The objectives of our study were to identify two-dimensional (2D) and Doppler echocardiographic clues to the fetal diagnosis of TAPVC and PAPVC with and without obstruction in a larger cohort of affected fetuses and to correlate the findings with those of postnatal echocardiograms or fetal autopsy.

METHODS

We identified all the pregnancies with a fetal diagnosis of anomalous PV connection made over the past 6 years by searching the Cardiology computer database of our institution. We retrospectively reviewed the prenatal and postnatal echocardiograms and the medical records including the autopsy reports when available. Two of the authors (S.J.Y. and E.R.V.) reviewed the videotaped fetal echocardiograms simultaneously to reach consensus on the findings and analyzed them systematically for the features previously described for postnatal diagnosis of anomalous PV connection[13–16]. We scrutinized the site of PV connection and looked for the presence of a confluent vein behind the left atrium or of a vertical vein. We measured the widths of the ventricles in a four-chamber view and the diameters of the right (RPA) and left (LPA) pulmonary arteries. The size of the chambers was not measured when an interatrial or interventricular communication was present, except for a foramen ovale. The dimensions of the cardiac structures were compared with the normal values established by Tan *et al.*[17]. We also calculated the ratios between the right and the left heart structures (described as mean ± SD). We analyzed the PV flow patterns in the pulmonary veins, in the vertical vein and at its connection to the systemic veins as shown by spectral Doppler when available.

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Prenatal echocardiographic features were compared with the findings of postnatal echocardiograms or findings at fetal autopsy.

This study was approved by the Research Ethics Board of our institution.

RESULTS

From December 1995 to October 2001, a total of 23 fetuses were diagnosed with anomalous PV drainage. Seven cases with heterotaxy syndrome, in which the pulmonary veins were connected to the posterior wall of the common atrium, were excluded from the study. The fetal diagnoses and clinical profiles for the 16 cases assessed are summarized in Table 1. The indications for fetal echocardiography were suspected cardiac defect at obstetric ultrasound in 15 pregnancies, including asymmetry between the two ventricles in one and abnormal position of the heart in four, and presence of multiple extracardiac anomalies in one pregnancy. The mean gestational age at time of correct diagnosis was 27 ± 7 (range, 18–39) weeks.

Diagnosis of TAPVC was made in 11 fetuses, including five with supracardiac connection, five with infracardiac connection and one mixed type. PAPVC was present in five fetuses, including four cases of scimitar syndrome and one case with the left-sided pulmonary veins connected to the innominate vein. Additional cardiac defects were present in 10 fetuses, including right atrial isomerism in eight and hypoplastic left heart syndrome in two. Four fetuses had extracardiac malformations (Table 1).

The diagnosis of TAPVC or PAPVC was made at initial prenatal assessment in 12 fetuses. In three cases the diagnosis was made at the subsequent follow-up examination. The echocardiograms of these three cases showed that the anomalous pulmonary veins were considered normal in the initial study because they were in close proximity to the posterior wall of the left atrium. In one case (Case 14) scimitar syndrome was diagnosed because two veins from the right thorax were connected to the inferior vena cava. After birth, however, both veins were shown to be the hepatic veins draining the right lobe of the liver, which was herniated through a right diaphragmatic defect. Postnatal echocardiography and surgery revealed a small pulmonary vein, abnormally connected to the inferior vena cava at its right atrial junction, draining the compressed and hypoplastic right lung. Thus in this case the diagnosis of scimitar syndrome was made prenatally but incorrectly described.

Echocardiographic findings in fetal anomalous PV connection

A PV confluence was identified in 10/11 fetuses with TAPVC. The venous confluence was seen as an oval- or star-shaped vessel behind the posterior atrium in transverse or coronal views. A PV confluence could not be recognized in one fetus with suspected mixed type of TAPVC, where the right-sided pulmonary veins connected to the innominate vein and the left-sided pulmonary veins to the left-sided superior vena cava (Case 5). In all 10 fetuses the PV confluence joined a vertical vein, which could be visualized in its entire course to a supracardiac or infracardiac systemic vein by sweeping the transducer in transverse or coronal planes (Figure 1). In 6/11 fetuses with TAPVC, including the mixed type, and in a case of PAPVC (Case 12) a separation between the posterior wall of the atrium and the descending aorta, which often contained either the PV confluence or the vertical vein, was observed (Figure 2).

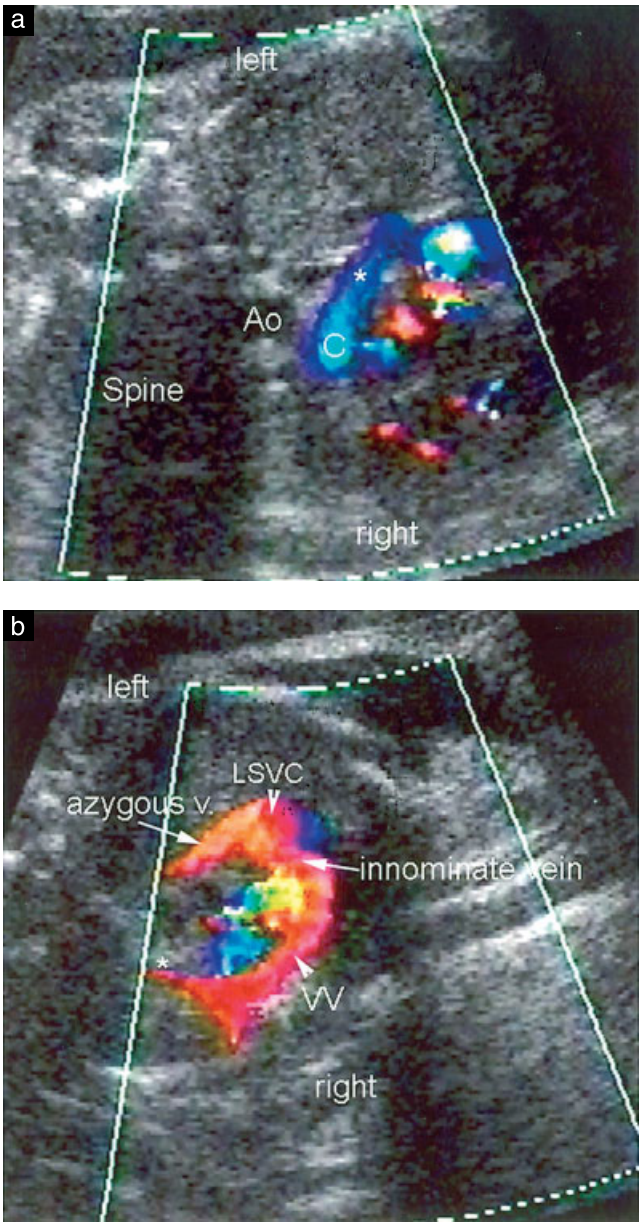

Figure 1 Total anomalous pulmonary venous connection to the innominate vein in a fetus with right atrial isomerism, dextrocardia, atrioventricular septal defect and double-outlet right ventricle (Case 9). (a) Color Doppler image in transverse view shows the left pulmonary vein (*) joining the pulmonary venous (PV) confluence (C). (b) Color Doppler image in oblique coronal plane demonstrates that the PV confluence is connected to the innominate vein through the vertical vein (VV). Ao, descending aorta; LSVC, left superior vena cava.

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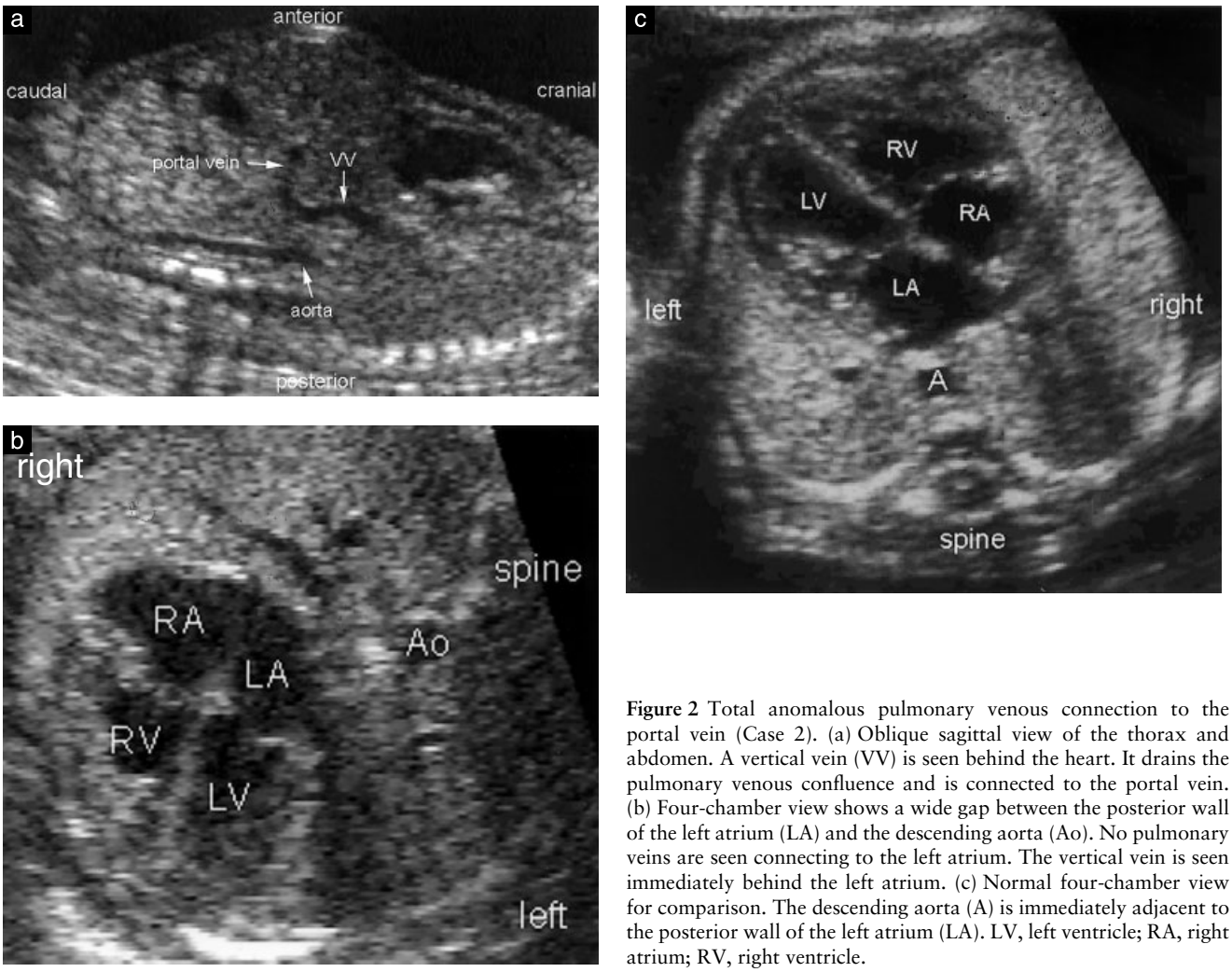

Figure 2 Total anomalous pulmonary venous connection to the portal vein (Case 2). (a) Oblique sagittal view of the thorax and abdomen. A vertical vein (VV) is seen behind the heart. It drains the pulmonary venous confluence and is connected to the portal vein. (b) Four-chamber view shows a wide gap between the posterior wall of the left atrium (LA) and the descending aorta (Ao). No pulmonary veins are seen connecting to the left atrium. The vertical vein is seen immediately behind the left atrium. (c) Normal four-chamber view for comparison. The descending aorta (A) is immediately adjacent to the posterior wall of the left atrium (LA). LV, left ventricle; RA, right atrium; RV, right ventricle.

Scimitar syndrome was diagnosed in four fetuses. Dextroposition was present in three cases (Figure 3). In one fetus with right-sided diaphragmatic hernia the heart was normally located in the left thorax. Hypoplasia of the RPA was observed in three cases. In one fetus the RPA could not be visualized because it was severely hypoplastic. In those three cases with measurable RPAs and LPAs the mean (± SD) ratio between the two was 0.65 ± 0.3.

Among the other cases, progressive discrepancy in size of the branch pulmonary arteries was observed in serial follow-up studies only in the fetus (Case 12) with hypoplastic left heart syndrome and unilateral PAPVC to a left vertical vein and to the innominate vein. Postnatal examinations confirmed hypoplasia of the LPA and showed progressive obstruction of the left pulmonary veins.

The widths of the right and left ventricles were measured in six fetuses, including four with scimitar syndrome and two with TAPVC. Absolute values were within the normal ranges[17] in all studies, but the right ventricle was disproportionately larger than the left ventricle in 5/6 cases[18]. The mean (± SD) ratio between the right and left ventricular widths was 1.36 ± 0.19.

Doppler flow pattern in fetal anomalous PV connection

Stenosis along the anomalous PV pathway was identified in four fetuses with supracardiac TAPVC, one fetus with mixed TAPVC and another with infracardiac TAPVC. In all of these cases color Doppler showed flow turbulence at the site of obstruction. Spectral Doppler demonstrated high-velocity continuous flow (Figure 4c) at the junction of the vertical vein to the systemic veins or at the site of anatomic vice, where the vertical vein was impinged, in all six cases of obstruction. Flow patterns were also available for the pulmonary veins in 5/6 cases with obstruction. In all pulmonary veins with downstream obstruction there was abnormal, monophasic, continuous low-velocity pattern (Figure 4b). An identical flow pattern was demonstrated in a vertical vein with downstream obstruction in another case. In contrast, two cases with unobstructed PV pathway showed biphasic flow pattern with cessation of flow in the end-diastolic phase, similar to the pattern seen in the normal fetal pulmonary veins[19]. Additionally a large respiratory variability of the PV flow was observed in one fetus with infracardiac TAPVC without evidence of obstruction. Color Doppler evaluation did not raise any

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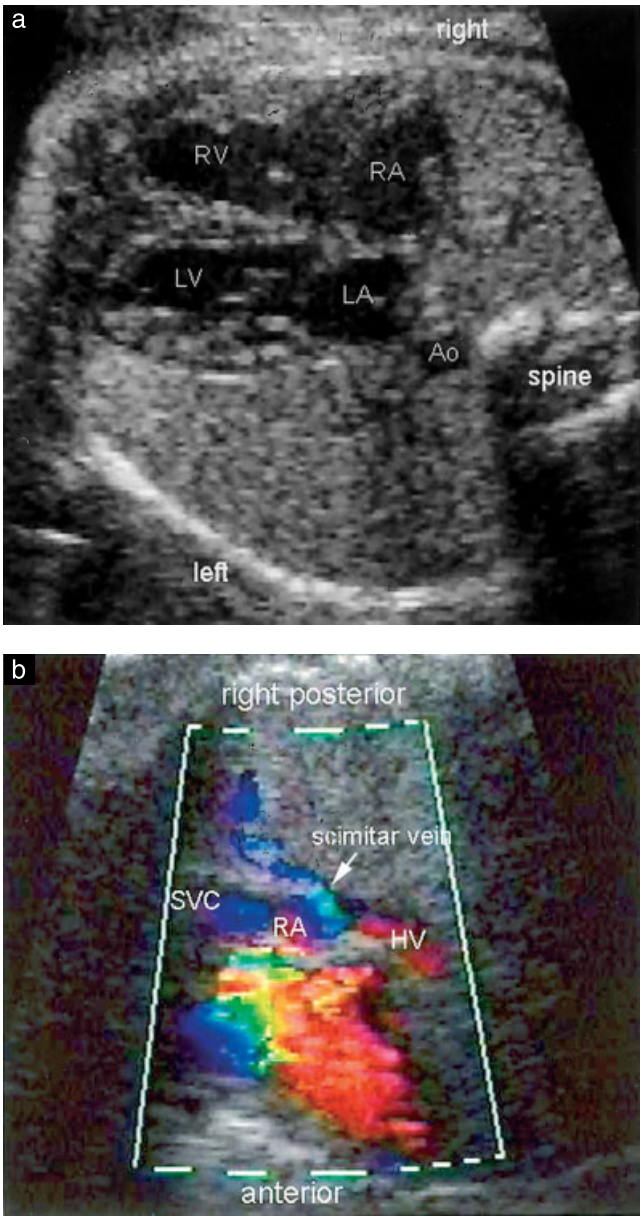

Figure 3 Scimitar syndrome (Case 13). (a) Four-chamber view showing dextroposition of the heart. (b) Color Doppler image in oblique sagittal view shows the scimitar vein. It drains the pulmonary veins from the right lung and enters the lower part of the right atrium (RA) at its junction with the inferior vena cava and hepatic veins (HV). AO, descending aorta; LA, left atrium; LV, left ventricle; RV, right ventricle; SVC, superior vena cava.

suspicion of obstruction in four fetuses with infracardiac TAPVC.

Postnatal findings and outcome

Nine fetuses were born at term and seven pregnancies were terminated (Table 1). There was no intrauterine spontaneous death. The fetal diagnosis regarding site of anomalous PV return and presence of obstruction along the PV pathway was confirmed by postnatal echocardiography in nine cases. In 3/7 terminated cases, autopsy confirmed the prenatal diagnosis. Autopsy was not performed in the remaining cases.

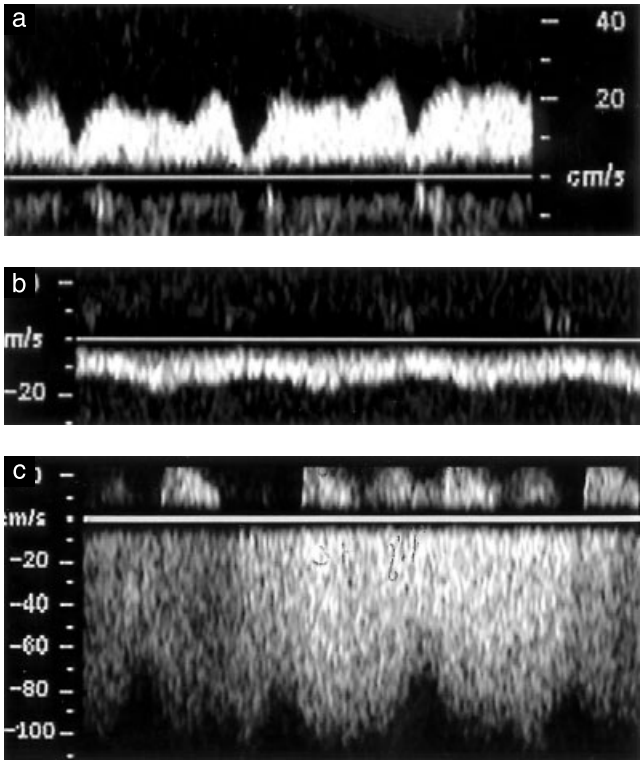

Figure 4 (a) Doppler waveform from a normal pulmonary vein in the fetus. There is biphasic flow with a systolic and diastolic peak. Very little or no forward flow is seen at the end of diastole. (b) Doppler waveform from a pulmonary vein upstream from a stenosis (Case 15). There is continuous monophasic flow with low velocity. (c) Doppler waveform from the vertical vein downstream from the stenosis (Case 7). There is continuous forward flow with increased velocity.

Compassionate care was decided for one patient with hypoplastic left heart syndrome and two with right atrial isomerism and complex heart defect. One patient with hypoplastic left heart syndrome and progressive, severe stenosis of the anomalous pulmonary veins died while awaiting heart transplantation (Case 12). Two patients died after developing progressive stenosis of the pulmonary veins after surgical repair of TAPVC. One patient succumbed to severe pulmonary hypertension at the age of 8 months after surgical repair of a congenital diaphragmatic hernia and right pulmonary lobectomy (Case 14). Another patient died at the age of 3 months after surgical repair of the anomalous PV connection due to significant residual cardiac pathology (Case 7). Only one patient was alive at time of data collection. This 4-week-old infant with right atrial isomerism, mild pulmonary stenosis and unobstructed TAPVC of supracardiac type is currently awaiting PV repair and single ventricle palliation (Case 9).

DISCUSSION

The diagnosis of anomalous PV connection can be made accurately after birth by combining two-dimensional echocardiography with color Doppler mapping[20–22]. However, only a few papers and case reports have

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described successful prenatal diagnosis of TAPVC and PAPVC[8,10,11]. This retrospective study performed in a larger cohort of affected pregnancies demonstrates that the presence of an anomalous PV connection can be identified by targeted fetal echocardiography and provides important 2D and spectral Doppler echocardiographic clues to the diagnosis. In this patient group the correct diagnosis was made prenatally in 15/16 cases. Additionally, we are aware of two fetuses that were assessed by fetal echocardiography in the period of our study but in whom the correct diagnosis of anomalous PV connection was not recognized. Both cases showed complex intracardiac anatomy; one with hypoplastic left heart syndrome and TAPVC to a dilated coronary sinus and the other with crisscross heart and PAPVC with the right upper pulmonary vein connected to the superior vena cava. Some more cases may have been potentially missed during this 6-year period. However, improved image resolution[11] and high level of suspicion in the presence of an associated cardiac anomaly clearly contributed in the course of time to our ability to make the diagnosis.

The fetal echocardiographic clues to the diagnosis of TAPVC we observed included the inability to demonstrate a direct PV connection to the left atrium, the presence of a PV confluence behind the atrium, a separation between the posterior wall of the atrium and the descending aorta, and the visualization of an ascending or descending vertical vein. These features were present not only in the fetuses with complex intracardiac anatomy but also in the two cases with isolated TAPVC. Both PV confluence and vertical vein were identified by careful evaluation of the space behind the heart. On axial images through the fetal chest, the space between heart and spine should contain only the descending aorta, usually located left of, and anterior to, the spine. The presence of any additional vessel in the retrocardiac space indicates anomalous PV connection. However, a dilated esophagus may mimic a vertical vein or a PV confluence. Color Doppler interrogation provides additional information about the direction of flow through the vertical vein and differentiates it from other vascular and non-vascular structures[23].

Fetal cardiac dextroposition was present in 3/4 cases and hypoplasia of the RPA in all cases with scimitar syndrome we assessed. These findings have also been previously reported as indicative of right lung hypoplasia[24]. Once these clues were detected, we scrutinized the inferior vena cava at its junction with the right atrium, where the scimitar vein draining all or part of the right lung is usually connected. In another case the asymmetric size of the branch pulmonary arteries was a useful clue to the presence of PAPVC.

The degree of ventricular disproportion we observed in two cases with TAPVC and in three cases with scimitar syndrome was mild. This observation is similar to the experience of Allan and Sharland, who retrospectively showed a disproportion in ventricular size only in late pregnancy in about half their cases with TAPVC[10]. They explained that the right ventricle does not become disproportionately large until late pregnancy, when pulmonary blood flow increases from 20% to 25% of the combined ventricular output. As an alternative explanation, they suggested that the presence of a large atrial septal defect allows some of the excessive venous return in the right atrium to reach the left atrium. Similarly, if an infracardiac type of anomalous PV connection is present, such as in scimitar syndrome, the increased venous return from the hepatic veins may flow directly into the left atrium through the patent foramen ovale, allowing a normal growth of the left-sided cardiac structures[23].

The PV pathway can be obstructed anywhere from the individual pulmonary veins to the site of abnormal connection to the systemic veins[1]. Color and spectral Doppler assessment facilitated the identification of the fetuses with PV pathway obstruction and therefore at risk for acute postnatal deterioration. In this study we demonstrate that in the fetus, despite reduced pulmonary blood flow, the PV flow patterns in the presence of obstruction are similar to those described after birth, with flow acceleration and high-velocity turbulent flow at the site of obstruction, and monophasic continuous low-velocity flow curve in the vertical vein and in the pulmonary veins upstream from it[13,25]. In one fetus with TAPVC of infracardiac type and no evidence of obstruction we noticed a strong influence of fetal respiration on the flow in the PV pathway, which corresponds to the influence of fetal respiration on systemic venous flow and filling mechanisms of the right atrium. In infradiaphragmatic TAPVC a significant obstruction may develop only after birth as the PV flow increases and the ductus venosus, widely patent in the fetus, constricts in the first few hours of life[26]. In fact the descending vein may connect either directly to the ductus venosus or to the portal venous system that prenatally communicates with the ductus venosus.

The majority of the cases we describe in this study had additional significant cardiac defects or abnormal cardiac position that were the main indication for referral for fetal echocardiography. However, despite our ability to accurately define PV connections in the fetuses that had been referred for echocardiography, there was no case with uncomplicated anomalous PV connection. This suggests that the majority of neonates with TAPVC or PAPVC without any associated anomalies may continue to go unrecognized before birth. Therefore, it is crucial that the connection of the pulmonary veins is looked for as a routine during a normal scan and not only in high-risk settings such as complex congenital defects. This paper should serve to acquaint fetal sonographers with the clues to the diagnosis of anomalous PV connection, even as an isolated defect, and to refer suspected cases to a tertiary center for further imaging.

The outcome of our cases of anomalous PV connection was dismal, with a mortality of 88% among live births. This high morbidity and mortality were largely due to the presence of severe additional cardiac pathologies, similarly to previous reports[5]. In these patients fetal

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diagnosis did not significantly improve the prognosis, although it was critical for parental counseling and perinatal management. Nevertheless, in the isolated type of TAPVC the outcome might be good if the diagnosis is made prenatally.

We conclude that TAPVC and PAPVC can be correctly diagnosed in the fetus. PV pathway obstruction can also be predicted prenatally by assessing PV flow pattern with color and spectral Doppler mapping. Detailed assessment of the PV connection, both at initial examination and in serial fetal studies, is important for the prenatal detection of anomalous PV connection in isolation and when additional lesions are found.

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