Perinatal outcome after ultrasound prenatal diagnosis of persistent right umbilical vein

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ABSTRACT

Objective: Our aim was to describe ultrasound findings and perinatal outcome after prenatal diagnosis of persistent right umbilical vein (PRUV).

Study design: We performed a retrospective analysis of fetuses with an ultrasound-based prenatal diagnosis of PRUV on record at 2 tertiary centers in Madrid, Spain. We describe clinical, maternal, fetal and perinatal variables for all cases.

Results: A total of 20,426 fetuses were delivered between the study centers. We detected 22 cases (0.1%) of PRUV. The male-to-female ratio was 1:1. All cases were intrahepatic type and diagnosed during the second and third trimesters (median, 21 weeks; IQR, 20–29 weeks). Doppler ultrasound revealed normal flow in the ductus venosus in all cases. Nine fetuses (40.9%) had additional ultrasound anomalies but no chromosomal abnormalities. Cardiovascular malformations were the most frequently associated congenital anomalies (4/9), followed by neurological malformations (2/9). In 5 of the cases with no concomitant anomalies, the weight of the newborn was below the tenth percentile for gestational age. Gestational development was normal for the remaining newborns. Delivery was unremarkable, and post-natal evolution was favorable.

Conclusion: Our results point to a potential association between PRUV and other fetal malformations and a very low rate of chromosomal abnormalities. Prenatal diagnosis of PRUV should be followed by detailed anatomical evaluation and echocardiography in order to rule out other structural malformations. The indication for a fetal karyotype study must be made on an individual basis considering PRUV type and other ultrasound findings.

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1. Introduction

Recent advances in ultrasound techniques, combined with a systematic approach to fetal morphology, have revealed an increase in the number of prenatally diagnosed vascular anomalies [1–3]. Persistent right umbilical vein (PRUV) is the most common prenatally detected venous anomaly, with an estimated prevalence of between 1 in 250 and 1 in 1250 [4–9].

The embryonic venous system consists of 3 symmetrical pairs of veins that drain into the heart, namely the cardinal, umbilical, and yolk sac veins. The umbilical and yolk sac veins are the major afferent vessels of the fetal liver. The right umbilical vein and the cranial portion of the left umbilical vein usually atrophy and disappear, making the left umbilical vein the main conduit of blood from the placenta. During the eighth week of development, the intrahepatic portion of the vitelline vein, and more specifically the left portal branch, form an anastomosis between the intrahepatic segment of the left umbilical vein and the ductus venosus, which arises from the coalescence of the hepatic sinusoids, and drains into the hepatocardiac segment of the inferior vena cava [10–12].

In most cases of PRUV, the left umbilical vein regresses and the right umbilical vein remains open [13]. Although the etiology of PRUV is unclear, proposed mechanisms include teratogenic agents and early obstruction of the left umbilical vein secondary to external pressure or thromboembolic events from the placenta, in which case echogenic foci can be observed in the liver parenchyma [14,15]. Prenatal ultrasound-based diagnosis of PRUV can be made during measurement of abdominal circumference in the routine fetal morphology scan [15]. Unlike the distribution in a normal fetus (Fig. 1A and Fig. 2A), the aberrant vein passes laterally to the right of the gallbladder (Fig. 1B, Fig. 2B and C).
Recently published studies have shown that PRUV can occur in association with other congenital anomalies or in isolation, in which case recommending additional tests such as chromosomal study remains open to debate [4–9]. We analyzed perinatal maternal and fetal variables after diagnosis of PRUV in order to determine their usefulness in the management of this ultrasound finding.

2. Materials and methods

We performed a descriptive observational study of patients with a prenatal ultrasound–based diagnosis of PRUV at two tertiary centers in Madrid: Hospital General Universitario Severo Ochoa (January 2002–December 2005) and Hospital General Universitario Gregorio Marañón (October 2010–July 2011). In these periods our sonographer (FGA) performed all the ultrasound examinations and collected perinatal information of the cases in both centers. All ultrasound examinations were performed using a 4–8 MHz abdominal probe (GE Logic 9 and Voluson Expert). Written informed consent was obtained from all parents. PRUV was diagnosed using the ultrasound criteria described by Jeanty [14], as follows: (1) aberrant course of the portal vein toward the stomach (instead of being roughly parallel); (2) position of the fetal gallbladder medial to the umbilical vein; and (3) umbilical vein not connected to the left portal vein, but abnormally to the right portal vein (Fig. 1B, Fig. 2B and C).
Gestational age was calculated from the date of the last menstrual period or using crown-rump-length during the first trimester of pregnancy. Exhaustive fetal ultrasound and echocardiography were performed after the diagnosis of PRUV was confirmed in order to detect associated anomalies. A karyotype study was offered in all cases of associated abnormalities. After diagnosis, and depending on the nature and progress of each case, specialists from both centers monitored the pregnancies with additional ultrasound scans according to the protocols of the Spanish Society of Obstetrics and Gynecology. Following delivery, all live born infants were evaluated by a pediatrician, and neonatal outcome data were obtained from medical charts.

Maternal and fetal and perinatal variables were analyzed retrospectively by two obstetricians (RM and CB).

3. Results

The study population comprised 20,426 fetuses of which 22 had PRUV (incidence of ≈1/1000). All cases were diagnosed during the second and third trimesters (median, 21 weeks; IQR, 20–29 weeks). Table 1 describes maternal, fetal, and perinatal variables for each case. In all cases, PRUV was classified as intrahepatic type with normal blood flow in the ductus venosus. The male-to-female ratio was 1:1.

Nine cases (40.9%) had additional anomalies (non-isolated cases), although the results of their chromosomal studies were normal. A cardiovascular malformation was identified as the leading anomaly in 4 of the 9 cases, and 2 of these had hypoplastic left heart syndrome. Another fetus had an atroventricular canal with aortic stenosis and interrupted aortic arch, and the fourth presented a complex cardiovascular malformation with an unbalanced atroventricular canal defect and right ventricle truncus arteriosus (right isomerism was suspected). One fetus with non-isolated PRUV had clinodactyly, and a further two had umbilical vein dilatation and hydrocele, respectively, as the only associated ultrasound finding. The second most common associated malformations were neurological anomalies, with a case of cerebellar vermis hypoplasia and a ventriculomegaly.

Consecutive ultrasound scans demonstrated intrauterine growth restriction in 3 cases of non-isolated PRUV; newborn weight was below the tenth percentile in 5 of the isolated cases. Perinatal outcome in non-isolated PRUV was unfavorable in 5 cases (4/5 associated with cardiovascular malformation and one case of ventriculomegaly, short long bones and normal karyotype). Delivery was uneventful and postnatal outcome was favorable in all cases of isolated PRUV (Table 1).

4. Comment

Prenatal diagnosis of PRUV is unusual, with a prevalence of ≈1/1000 in our series [5–9]. In general, most of the cases (95%) reported in the literature involve PRUV with intrahepatic drainage (PRUV-I), where the right umbilical vein connects to the portal system at the portal venous sinus to form the ductus venosus (Fig. 1B, Fig. 2B and C); in some cases, however, the ductus venosus may not have formed [9]. Drainage can also be extraportal (PRUV-E), that is, blood from the right umbilical vein flows directly into the inferior vena cava or right atrium; this finding is associated with agenesis of the ductus venosus and poor prognosis [14]. Depending on the type of PRUV, associated anomalies vary and include cardiac, musculoskeletal, genitourinary, and gastrointestinal malformations [16].

In 1990, Jeanty [14] reported six cases of PRUV, of which 50% were associated with structural malformations (compared with...
frequent fetal intrauterine abnormalities. PRUV-I. Recommended to be further studied, as abnormalities are frequently associated with increased flow in the systemic venous system. In this group, the prevalence of chromosomal abnormalities was 18.8%, and structural abnormalities were present in all cases. The remaining 286 cases (94.7%) were PRUV-I. An associated abnormal karyotype was detected in 0.3% of cases in the isolated PRUV-I group. Structural abnormalities were identified in 60 cases (21%), and chromosomal abnormalities were found in 1.6%. In our series, however, we found no chromosomal abnormalities.

Since cases with PRUV-I are occasionally accompanied by structural abnormalities that are rarely associated with chromosomal abnormalities, the recommendation for a chromosomal study in cases of non-isolated PRUV should be made on an individual basis [5,6,8]. Consecutive ultrasound demonstrated intrauterine growth restriction in 30% of our cases (46% in isolated PRUV versus 22% in non-isolated PRUV). All isolated PRUV had a favorable perinatal outcome. Today, most cases of PRUV-I appear in isolation and have a good postnatal prognosis [9], although the nature of associated anomalies is the most important indicator of perinatal outcome [17,18]. Consistent with the results of previous studies, the most frequent associated anomalies in our series were heart defects.

In summary, prenatal diagnosis of PRUV should be followed by an enhanced fetal morphology scan and echocardiography. In addition, consecutive ultrasound should be performed to monitor fetal growth. Information given to parents on prognosis in cases of isolated PRUV-I should be reassuring. Fetal karyotyping is not recommended in isolated PRUV-I, and further fetal studies should be recommended in non-isolated PRUV-I on an individual basis, depending on the anomaly or the presence of ultrasound markers of aneuploidy. Nevertheless, invasive studies should be considered in both isolated and non-isolated PRUV.

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