PRENATAL DIAGNOSIS OF INTRAHEPATIC PERSISTENCE OF THE RIGHT UMBILICAL VEIN USING THREE DIMENSIONAL SONOGRAPHER WITH COLOR DOPPLER – AN ANATOMICAL VARIANT. CASE REPORT

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PRENATAL DIAGNOSIS OF INTRAHEPATIC PERSISTENCE OF THE RIGHT UMBILICAL VEIN USING THREE DIMENSIONAL SONOGRAPHER WITH COLOR DOPPLER – AN ANATOMICAL VARIANT. CASE REPORT (Abstract): Persistent right umbilical vein (PRUV) is a vascular pathology in which the left umbilical vein becomes occluded while the right umbilical vein persists and remains open. The persistent right umbilical vein may replace the normal left umbilical vein or be supernumerary. When observing fetal anatomy during a routine 20-week scan (or earlier), it is important to notice the direction of the vasculature for possible fetal anomalies such as PRUV. Prenatal diagnosis of anomalies of the umbilical veins is usually straightforward and may involve supernumerary vessels, abnormal course, size and insertion and persistence of embryological vascular structures. The development and widespread use of color Doppler interrogation as well as the advent of three-dimensional ultrasound applications has facilitated early in-utero diagnosis of abnormalities of the fetal umbilico-portal circulation. Following the identification of the PRUV, the fetus is assessed for the possibility of other associated anomalies. Clearly, when an anomaly is observed, it is crucial to assess the fetus due to its associated with numerous and occasionally lethal malformations. This condition had traditionally been considered to be rare and associated with a poor neonatal prognosis. Later evidence has raised some doubts about the veracity of these observations. Key words: UMBILICAL VEIN, PORTAL VEIN, UMBILICAL CORD

INTRODUCTION

In the normal fetus, the right umbilical vein begins to become obliterated around the 4th week of pregnancy, and disappears by the seventh week of gestation (1,2,3). During the sixth week, a critical anastomosis occurs between the left UV (umbilical vein) and the hepatic sinusoids, while the cranial portion of the both UV is programmed to regress, thereby losing the direct communication with the sinus venosus. Simultaneously, the liver bud progressively extends and incorporates the cranial part of the vitelline veins, which are involved in the formation of the hepatic sinusoids, portal vein and the ductus venosus (4,5).

In certain cases the anastomosis between the left UV and the right vitelline vein fails to develop and subsequent degeneration of both UVs occurs. As a result, oxygenated venous blood return from the placenta is blocked and redirected via aberrant vessels towards systemic veins bypassing the liver (iliac vein, inferior vena cava or direct communication with the right atrium). Partial failure to form critical anastomosis seems to be more common than...
Based on these findings, two major paths of blood flow towards the heart have been established:

1. The most common of which is PRUV, which is characterized by failure of the right-sided UV to form an anastomosis with the right vitelline vein and degeneration of the left UV.

2. The second, less frequent path involves direct drainage of the UV into the right atrium.

Therefore, cases of proven PRUV are usually subdivided into two groups:

1. the intrahepatic type (isolated right UV joining the portal system at the level of the sinus venosus, giving rise to the ductus venosus) and

2. the extrahepatic type in which the PRUV bypasses the liver completely (absent ductus venosus) [6, 7].

In addition, a third variant has occasionally been described as a supernumerary intracorporeal (intrahepatic) or extracorporeal vascular structure (four-vessel umbilical cord) as a result of patency of both UVs.

The intrahepatic form of persistent right umbilical vein might present as an isolated finding, with no additional anomalies, whereas all the cases of persistent extrahepatic right umbilical vein were associated with significant fetal abnormalities [8].

The exact underlying mechanism causing a PRUV remains unclear, although several pathophysiologic explanations have recently been suggested. Jeanty proposed that the reason for patency of the right-sided vessel might be related to streaming of the early flow traversing the UV. Early obstruction of the left UV from external pressure or occlusion by thromboembolic events arising from the placenta may also result in the appearance of PRUV [9].

In an experimental approach teratogenic agents such as retinoic acid and folate deficiency were found to induce similar developmental alterations as described in primary failure to form an anastomosis, resulting in a patent right UV [10].

The persistence of an open right umbilical vein does not prevent the formation of the ductus venosus. Although the course of the blood within the liver is abnormal, it does not alter blood distribution to the fetus. Persistent of right umbilical vein was traditionally thought to be an uncommon event strongly associated with severe fetal anomalies, including congenital heart defects, and gastrointestinal, urinary and musculoskeletal system malformations [11, 12, 13, 14].

A single umbilical artery is the most common congenital anomaly associated with a PRUV. But the most fetuses with PRUV was proved to be normal.

The prevalence of PRUV ranges from 0.1% to 0.7%, according to some reports [15, 16, 17, 18, 19].

The occurrence of concomitant atrial septal defects in cases with an abnormal UV might be attributed to an increase in flow via the inferior vena cava, thereby interfering with developmental processes during atrial septation [20].

Differential diagnoses of PRUV include umbilical vein varix, gallbladder duplication, abnor-
mal course of the portal vein and its branches and intrahepatic cysts \[21, 22, 23, 24\].

**MATERIALS AND METHODS**

A 34-year-old woman was referred for assessment at 20 weeks of gestation. She had no significant pathology in the past. A detailed structural evaluation of the fetus, including biometrical studies, was carried out. The sonographic diagnosis of a persistent right umbilical vein was made in a transverse section of the fetal abdomen. Special attention was given to the direction of the curve of the portal vein and the position of the gall bladder. The following criteria were met:

1. the portal vein curved towards the stomach (Fig. 1)
2. the fetal gall bladder was located medially to the umbilical vein (between the umbilical vein and the stomach) (Fig. 2)

On ultrasound examination the heart was normally sited with normally positioned great vessels. High definition color flow Doppler was used to examine the fetal portion of the umbilical vein from the cord insertion. The umbilical cord had normal structure (3 vessels) with normal abdominal insertion (Fig. 3). A diagnosis of persistence of the right umbilical vein (PRUV) was made. The fetal heart rate and rhythm appeared normal. No other fetal abnormality was identified.

**CONCLUSION**

Three sets of veins drain into the primitive heart and join at the sinus venosus, these are the common cardiac veins, the vitelline veins and the umbilical veins which drain the body, yolk sac and placenta, respectively. During normal embryological development the right umbilical vein occludes at approximately 28–32 days (embryological age). The cranial portion of the vitelline veins are incorporated into the hepatic sinusoid thus forming a portal system that drains blood from the abdominal foregut, midgut, and upper anorectal canal. During this time, the critical anastomosis occurs between the remaining left umbilical vein and the hepatic sinusoids. Failure of this critical anastomosis leads to aberrant drainage of the umbilical vein together with lack of formation of the ductus venosus. The umbilical vein may then form connections to any of the major vessels such as the IVC, iliac veins or even directly into the right atrium. Persistence of the right umbilical vein is rare.

Two subtypes have been described. In the first, the PRUV exists as an intrahepatic variant (our case) and appears to carry a good prognosis if no other abnormality is seen. The second type when the PRUV bypasses the liver completely.

In conclusion, vascular abnormalities within the fetal umbilico portal venous system are relatively common prenatal findings. Persistent intrahepatic right umbilical vein is a fetal anatomic variant that is not so rare and usually is associated with a favorable outcome. The correct antenatal diagnosis of PRUV as well as its intra- and extra hepatic connections is of particular importance owing to its potential association with additional malformations. Nevertheless, the overall prognosis for fetuses with isolated PRUV and proper connection to the portal system is good.

**BIBLIOGRAPHY**


