

Prenatal Diagnosis and Management of Umbilical Vein Varix of the Intra-amniotic Portion of the Umbilical Vein

Sean P. White, MD, Alexander Kofinas, MD

Varix of the umbilical vein is a relatively uncommon abnormality. It has been reported to cause hemorrhage through the amniotic sheath and subsequent fetal exsanguination.¹ We present a patient in whom the antenatal diagnosis of this condition was made by ultrasonography and describe the management of this case.

CASE

A 27 year old gravida 3, para 2 woman was referred for a sonogram at 29 3/7 weeks' gestation to rule out placenta previa. At that time, the patient was noted to have a total eccentric placenta previa with the cord insertion at the edge of the placenta just at the cervical os. Color Doppler investigation ruled out vasa previa. Moderate symmetric IUGR at the sixth percentile was noted. Reevaluation at 32 2/7 weeks demonstrated an unchanged placenta. A new

development relating to the umbilical cord was noted. Approximately 2 cm distal to the fetal abdomen, a varix measuring 2 × 2.3 cm was noted. Normal Doppler flow velocity waveforms were obtained from both umbilical arteries and the umbilical vein. Color flow Doppler sonography demonstrated bidirectional flow pattern inside the varix (Fig. 1). Improving fetal growth was noted, and observation was continued. Follow-up ultrasonography at 34 and 35 weeks' gestation revealed increasing size of the umbilical vein varix (2.3 × 2.4 cm and 2.7 × 2.4 cm, respectively).

At 35 weeks, NST was nonreactive and the biophysical profile score was 6 (normal movement, tone, and amniotic fluid, nonreactive NST, and no breathing activity). Lung maturity was ascertained and the patient was delivered because of concerns about rupture of the varix or thrombosis of the vein. Because of concurrent placenta previa, delivery of a viable female infant weighing 2041 grams with an Apgar score of 8 at 5 minutes was performed later that day by primary cesarean section. The placental pathology report demonstrated moderate villous edema, dilated and tortuous umbilical vein varix, and a recent partially occlusive umbilical arterial thrombosis adjacent to the varix. Compression injury of the umbilical artery from the expanding varix was suspected.

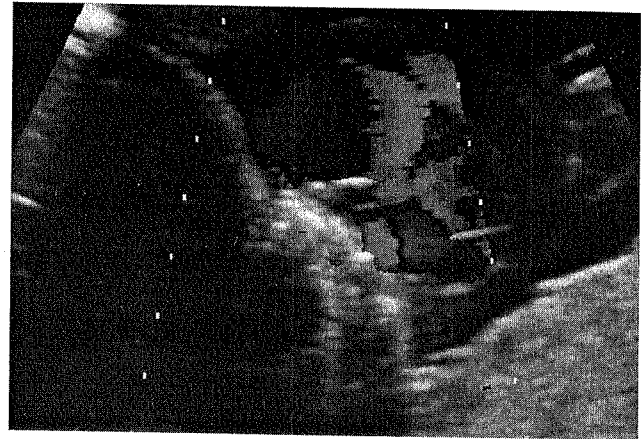
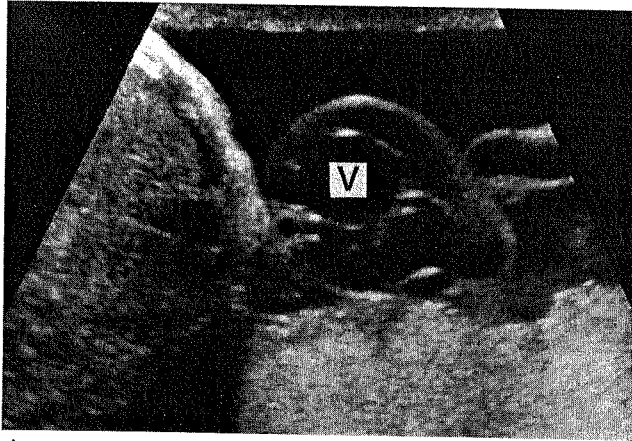
Between 1991 and 1994 we have had seven cases of antenatally diagnosed varix of the fetal intra-abdominal vein. Careful, extensive evaluation of the fetal anatomy and the remainder of the umbilical cord revealed no additional abnormalities. All patients were followed with sonographic and Doppler evaluations every 2 to 4 weeks until delivery. In three cases the varix decreased in size and the patients were released from further follow-up. In one case the varix was noted to be increased in size at the second visit but remained stable thereafter. In the other three cases

ABBREVIATIONS

IUGR, Intrauterine growth retardation; NST, Nonstress test; BPP, Blood pressure and pulse

Received March 7, 1994, from the Perinatal Diagnostic Center (S.P.W.), York, Pennsylvania; and the Department of Obstetrics and Gynecology, The Brooklyn Hospital Center (A.K.), Brooklyn, New York. Revised manuscript accepted for publication August 5, 1994.

Address correspondence and reprint requests to Alexander D. Kofinas, MD, Chief, Maternal-Fetal Medicine, The Brooklyn Hospital Center, 121 Dekalb Avenue, Brooklyn, NY 11201.



A Figure 1 A, Gray scale imaging of the umbilical vein varix (V). **B**, Color Doppler imaging documents bidirectional flow pattern inside the varix.

the varix remained stable in size with relative decrease, given the increasing size of the fetus. All seven fetuses were delivered at term without problems. The largest variceal diameter was 2.5 cm and the smallest was 1.4 cm. In all cases, the varix was grossly larger than the adjacent umbilical vein diameter. We utilized color Doppler imaging in all cases to evaluate the flow pattern within the varix and to rule out the development of thrombosis.

DISCUSSION

Siddiqi and coworkers² recently described a case of prenatal diagnosis of an umbilical artery aneurysm. Estroff and Benacerraf⁶ reported five cases of intra-abdominal umbilical vein varix with normal fetal outcomes. Our seven cases of antenatally diagnosed varix of the fetal intra-abdominal umbilical vein also had normal outcomes. This is in contrast to the findings of Mahony and colleagues,⁴ who reported increased perinatal loss of 44%. Additional reports described cases of intra-abdominal umbilical vein varices detected prenatally by ultrasonography.⁵⁻⁶ In contrast, the only reports we were able to identify on the diagnosis of intra-amniotic umbilical vein varix were postnatal placental and autopsy examinations.⁷⁻⁹ An autopsy series of 184 malformations of the cord reported that 3.8% were varices of the umbilical vein. In this series, all cases of varices and thrombosis of the umbilical vessels resulted in stillbirths.⁷

Ghosh and coworkers⁸ reported 12 pregnancies in which fetal distress and death were thought to be the result of umbilical cord anomalies. They noted that death tended to occur rapidly after an abnormal NST. Infants with cord anomalies also are at a higher

risk for thrombosis. Heifetz⁹ reported that cord thrombosis is seen in approximately 1 in 25 autopsies on patients with cord abnormalities. He noted that thrombosis has been associated with late gestational fetal distress or death and stated "Of 68 case reports on cord thrombosis in the literature, only two cases were not associated with fetal or neonatal morbidity or mortality." In his literature review, he found 17 cases of thrombosis with varicosity of the umbilical vein. He does not specify whether these were intra-abdominal or intra-amniotic. Hoag¹⁰ reported a fatal case of intra-amniotic umbilical vein thrombosis and fetomaternal hemorrhage.

In our case, the patient was referred for evaluation of an unrelated problem. The diagnosis of umbilical vein varix was an incidental finding. Because of this finding, the patient received careful antenatal monitoring. At the first indication of a change in the fetal well-being (the patient had a nonreactive NST), evaluation for fetal lung maturity was performed and delivery was recommended. The pathology report demonstrated a recent partially occlusive umbilical artery thrombosis in addition to the umbilical vein varix. If delivery had been delayed, a significant thrombosis could have formed and with the potential for an unfavorable outcome. The literature reports suggest that these patients are at high risk for late gestational fetal demise. We believe that based on the existing evidence, delivery of the fetus as soon as lung maturity is documented may be justifiable.

Prenatal diagnosis of umbilical vein varix and other cord anomalies should be more common with the addition of color Doppler imaging in most of the ultrasound systems residing in antenatal testing centers. In our experience, color Doppler imaging has

enhanced the ability to diagnose cord anomalies and improve accuracy. It is part of our imaging technique to always evaluate the cord insertion and intra-abdominal portion of the umbilical vessels with color imaging. The intra-amniotic portion of the cord is visualized carefully with gray scale imaging and color Doppler examination as indicated.

In conclusion, varix of the umbilical vein is a relatively uncommon anomaly. There are numerous reports of fatal outcomes in infants with umbilical vein varices and other vascular umbilical cord anomalies. These lesions can be detected antenatally, and delivery of the infant as soon as maturity is confirmed, even in the absence of documented fetal distress, may prevent adverse perinatal outcomes.

REFERENCES

1. Clausen I: Umbilical cord anomalies and antenatal fetal deaths. *Obstet Gynecol Surv* 44:841, 1989
2. Siddiqi T, Bendon R, Schultz D, et al: Umbilical artery aneurysm: Prenatal diagnosis and management. *Obstet Gynecol* 80:530, 1982
3. Estroff J, Benacerraf B: Fetal umbilical vein varix: Sonographic appearance and postnatal outcome. *J Ultrasound Med* 11:69, 1992
4. Mahony B, McGahan J, Nyberg D, et al: Varix of the intra-abdominal umbilical vein: Comparison with normal. *J Ultrasound Med* 11:73, 1992
5. Jeanty P: Fetal and funicular vascular anomalies: Identification with prenatal US. *Radiology* 173:367, 1989
6. Fuster JS, Benasco C, Saad I: Giant dilation of the umbilical vein. *J Clin Ultrasound* 13:363, 1985
7. Konstantinova BL: Malformations of the umbilical cord. *Acta Genet Med Gemellol* 26:259, 1977
8. Ghosh A, Woo JSK, MacHenry C, et al: Fetal loss from umbilical cord abnormalities—A difficult case for prevention. *Eur J Obstet Gynecol Reprod Biol* 18:183, 1984
9. Heifetz S: Thrombosis of the umbilical cord: Analysis of 52 cases and literature review. *Pediatr Pathol* 8:37, 1988
10. Hoag R: Fetomaternal hemorrhage associated with umbilical vein thrombosis. *Am J Obstet Gynecol* 154:1271, 1986