

Klippel-Trenaunay-Weber Syndrome-Associated Arterial and Venous Malformations in the Lower Uterine Segment

Geneviève Bouchard-Fortier, MD, MSc, Darine El-Chaar, MD, Peter Hawrylyshyn, MD, John Kingdom, MD, Elliott Lyons, MD

Department of Obstetrics and Gynaecology, University of Toronto, Toronto ON

A 41-year-old multigravid woman with Klippel-Trenaunay-Weber syndrome was seen at 36 weeks' gestation. This rare congenital vascular disorder is associated with port-wine stains, soft tissue swelling, and vascular malformations.¹ She had a previous vaginal delivery complicated by severe postpartum hemorrhage.

The presentation was breech, and a detailed ultrasound of the uterus was performed to plan a Caesarean section because of the risk of vascular malformations.² The lower uterine segment was thick with multiple venous and arterial vessels (Figure 1). A midline laparotomy with classical uterine incision was performed, followed by tubal ligation. Large vessels were noted on the lower uterine segment at the time of surgery (Figure 2).

The baby did well, and estimated blood loss was 900 mL. This management averted potentially severe complications associated with this syndrome.³

Consent to publish these images was obtained from the patient.

REFERENCES

1. Atis A, Ozdemir G, Tuncer G, Cetincelik U, Goker N, Ozsoy S. Management of a Klippel-Trenaunay syndrome in pregnant women with mega-cisterna magna and splenic and vulvar varices at birth: a case report. *J Obstet Gynaecol Res* 2012;38(11):1331-4.
2. Andreasen KR, Tabor A, Weber T. Klippel-Trenaunay-Weber syndrome in pregnancy and at delivery. *J Obstet Gynaecol* 1999;19(1):78-9.
3. Sivaprakasam MJ, Dolak JA. Anesthetic and obstetric considerations in a parturient with Klippel-Trenaunay syndrome. *Can J Anaesth* 2006;53(5):487-91.

Figure 1

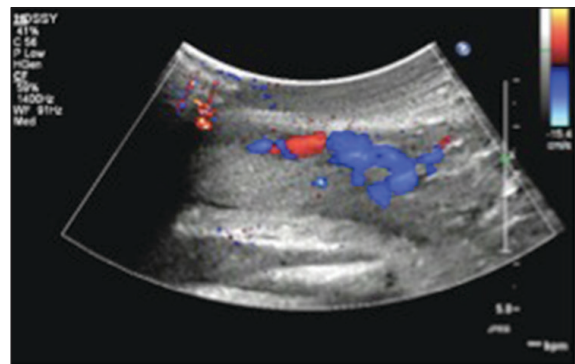


Figure 2

