

Title: Type IV Ehlers-Danlos Syndrome and a Fragile Uterus

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Background: Ehlers-Danlos Syndrome (EDS) encompasses a group of connective tissue disorders with various inheritance patterns as well as a constellation of patient symptoms, physical characteristics and maladies. The vascular subtype, known as type IV, is caused by a mutation affecting type III procollagen and is typically inherited in an autosomal dominant manner. EDS type IV patients do not characteristically exhibit the classic symptoms such as joint hypermobility and skin elasticity seen in more common EDS subtypes, rather arterial and digestive system morbidity as well as obstetrical complications not typically seen in other forms of EDS. Patients are often undiagnosed with EDS type IV until experiencing complications such as vascular or organ rupture. The sigmoid colon is the most common site of intestinal perforation, however the small intestine is occasionally involved. Uterine rupture or vascular rupture during pregnancy, particularly in the 3rd trimester, is associated with a significant risk for maternal mortality as both the intestinal and uterine walls contain an abundance of type III collagen. Non-gravid *uterine* complications in patients with EDS type IV are rare. This case report presents a female with a history of EDS type IV who presented with a defect in her uterine fundus, possibly due to unhealed perforation or spontaneous rupture in the context of a weakened uterine wall.

Objective: A case of diagnosis and treatment of spontaneous uterine rupture versus delayed wound dehiscence in the non-gravid uterus of a patient with Ehlers-Danlos Syndrome, type IV. We share this case to encourage that greater consideration of potential risks associated with gynecologic procedures in this patient population is imperative to improve long-term outcomes.

Case Report: A 30-year-old female with a history of EDS type IV presented to the emergency department with abdominal pain and dark-tinged vaginal discharge 2 weeks after an elective surgical abortion. The patient had a history of 1 prior cesarean section and 2 prior D&Cs. Physical examination and transvaginal ultrasound were unremarkable other than for bacterial vaginosis. Three months later, she returned with worsening right-sided abdominal pain. A CT scan revealed an ill-defined collection of blood products communicating with the endometrial canal. The patient was admitted for observation. Further escalation of abdominal pain with peritoneal signs and declining hematocrit prompted a diagnostic laparoscopy. A whole uterine fundal defect and hemoperitoneum were encountered. The defect was repaired laparoscopically and the patient recovered appropriately.

Results: This case highlights a uterine insult in a patient with EDS type IV in the absence of pregnancy. Dehiscence of a prior perforation and spontaneous uterine rupture should be considered in such patients, especially with a history of gynecological procedures. The diagnosis of uterine dehiscence in a non-gravid uterus may be challenging and transvaginal ultrasound may be inconclusive. CT imaging may have more diagnostic value and identify hemo- and pneumo-peritoneum. The clinical presentation of uterine rupture in non-gravid patients is variable and mimics other pathologies. A high index of suspicion is crucial. The literature on gynecological complications in EDS type IV patients is limited other than for vaginal mucosal injury, and more research is needed to assess the risks associated with gynecological procedures and the ability of uterine tissue to heal.

Conclusion: This case report emphasizes the need to consider uterine tissue and vascular quality in patients with EDS type IV outside of pregnancy. It highlights the challenges in diagnosing uterine dehiscence or rupture in non-gravid patients and the importance of maintaining high clinical suspicion with patients with this disease, especially in the setting of prior gynecologic and obstetric procedures. Awareness of uterine and vascular fragility in patients with EDS type IV may lead to decreased morbidity and mortality.