

Ehlers-Danlos Syndrome (EDS) and Vascular / Arterial Rupture

Over fifty cases of bowel perforation have been documented in the literature. A type of Ehlers-Danlos Syndrome is vascular EDS and one case that was reported in the literature involved a 68-year-old man who presented with severe abdominal pain. It was due to a jejunal artery aneurysm rupture, that's the blood supply to the middle section of the small bowel. The man had to have part of the bowel removed and the two ends of the intestine put back together. The point is, that there has to be a high index of suspicion on the part of the doctor. As a patient, can assist with a detailed family history because the risk of multiple aneurysms, which are ballooning of the artery or vessel supplying oxygen to that part of the body, that may not be identified with screening that identifies inflammation or vasculitis.

The aneurysms are not restricted to the bowel and have been noted in the right hepatic, splenic, aortic, superior mesenteric, celiac and renal arteries. More alarming is most percent by the age of forty. Proper surgical management in vascular EDS patient, with its challenges, which may include hemorrhage (blood loss) and shock (loss of blood pressure and consciousness).

While the mechanism is controversial for what triggers the arterial accidents, it may involve inflammation or disruption of metabolic pathway for collagen, a protein that acts to keep the blood vessels intact. The individual with vascular EDS may not have the classic signs a skin hyper-extensibility or stretching but may just have easy bruising wrinkling or delayed wound healing.

While the details of collagen synthesis are beyond the scope of this blog, what is important is an accurate diagnosis to allow for rapid decision making by the treating physician which may require an MRI or CT scan as well as surgical intervention rather than medical treatment. A physician who is familiar with the implications of the family history becomes extremely important and preventing death due to ruptured arterial aneurysms. This may include using new treatments that increase survival in vascular EDS patients. Please note that research in this area is ongoing

