THE VEDS MOVEMENT

Charging forward. Saving lives.

VEDS for the Gastroenterologist

Considerations for the Gastroenterologist

1. Because of the fragile vessels and tissues associated with VEDS, your patient may experience spontaneous bowel perforations.
2. Risk of perforation during endoscopic procedures are higher in individuals with VEDS.
3. Some GI related clinical concerns include hiatal hernias, postoperative hernias, gastritis, abdominal pain or distension, nausea, vomiting, GERD, constipation, evacuation disorders, pelvic floor dysfunction, and intestinal rupture, typically in the colon. These GI symptoms are also seen in the general population.
4. Patients may experience early or delayed gastric emptying, as well as delays or accelerations in colonic transit.
5. Your patient with VEDS may have an existing care team that includes a vascular surgeon, cardiologist, geneticist, primary care doctor, and/or a general surgeon. Please collaborate.

Suggested Care Modifications

1. Colonoscopies and endoscopies can cause life-threatening bowel perforation in people with VEDS. If these procedures are unavoidable, they should be done with a team aware of the underlying diagnosis, including a vascular surgeon, at a center with expert knowledge. Input should be obtained from a VEDS specialist prior to proceeding.
2. If endoscopy is needed, luminal distension should be minimized to degree possible to achieve clinical goal. Water insufflation can be considered over CO2. CO2 insufflation is likely still safer than previously used air insufflation.
3. Routine colonoscopy for cancer screening is discouraged in the absence of concerning symptoms or a strong family history of colorectal cancers. If there is no strong family history of colorectal cancers, consider non-invasive testing, such as Cologuard testing, first. CT colonography also increases risk of perforation.
4. Avoid stimulant laxatives and opt for osmotic laxatives first. Many people with VEDS take osmotic laxatives and/or stool softeners daily to avoid constipation and reduce strain.
5. Enemas should only be done under supervision, as they can cause rupture.
6. Physical therapy should be the first line of treatment for pelvic floor dysfunction.
7. Avoid NSAIDS when possible because of possible increased bleeding risks, including gastrointestinal bleeding.
8. Narcotic pain medications can lead to severe constipation.

BACKGROUND ON VEDS

- **VEDS is also known as Vascular Ehlers-Danlos Syndrome**, Ehlers-Danlos Type IV, vEDS, and previously known as Sack-Barabas Syndrome or the arterial form of Ehlers-Danlos Syndrome. It is a distinct genetic condition and is NOT the same as hypermobile Ehlers Danlos syndrome (HEDS).
- **VEDS is caused by genetic alteration in the gene** called *COL3A1* that encodes the chains of type III collagen. This protein is an important component of tissue in the bowels.
- **There are different kinds of mutations in *COL3A1* that cause VEDS.** As a consequence, some people with VEDS have faulty type III collagen, while others may have a reduction in the amount type III collagen, but what is made is normal.
- **People with VEDS are prone to life-threatening emergencies**, including arterial dissections and ruptures, pneumothorax, hemothorax, bowel perforations, and organ ruptures, uterine rupture during pregnancy, and recurrent pulmonary problems.

If a consult is needed, reach out to the Help and Resource Center at The VEDS Movement to be connected with a physician familiar with VEDS on The Marfan Foundation's Professional Advisory Board.
The Help and Resource Center can be reached at TheVEDSMovement.org/ask