

# THE VEDS MOVEMENT

*Charging forward. Saving lives.*

## VEDS for the Primary Care Physician

### Considerations for the Primary Care Physician

1. Care is generally local and under the supervision of the patient's primary care physician.
2. Your patient with VEDS needs a care team that includes their primary care physician, a vascular surgeon, cardiologist, general surgeon, and a geneticist. To the extent possible an expert in VEDS should be available (even at a distance) to help guide this care team. If your patient does not already have a care team established, assist in creating a collaborative care team.
3. Pediatric care is typically managed by a pediatrician or family care practitioner in concert with the care team.
4. Emergency care can be a reality of having VEDS. To ensure your patient gets the proper care at the emergency department, consider giving your patient a phone number where an on-call physician can be reached to call the emergency department to notify them about the life-threatening risks your patient faces.
5. A patient care coordination note should be added to the patient's electronic health record to assist in proper care in the emergency setting. For instructions in EPIC, see these instructions from The Marfan Foundation.  
[marfan.org/wp-content/uploads/2021/09/PCCN-Instructions\\_5.15.2020.pdf](http://marfan.org/wp-content/uploads/2021/09/PCCN-Instructions_5.15.2020.pdf)
6. There is a new ICD-10 Code for VEDS: Q79.63. This should be added to their electronic health record.
7. Discuss emergency preparedness with your patient. If the county or city offers a premise file for first responders, advocate for your patient to have one.

### BACKGROUND ON



- **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.
- **VEDS is caused by a genetic mutation in the gene** called *COL3A1* that tells the body how to make type III collagen, a major constituent of arterial, bowel, and uterine walls.
- **There are different kinds of mutations in *COL3A1* that cause VEDS.** Some people with VEDS make faulty type III collagen, while others may have a reduction in the amount of normal type III collagen made.
- **People with VEDS are prone to life-threatening emergencies**, including arterial (including the aorta) dissections and ruptures, spontaneous pneumothorax, hemothorax, bowel perforations, and organ ruptures, including gravid uterine rupture.

8. Because of the fragile vessels and tissues associated with VEDS, your patient may experience spontaneous life-threatening emergencies. Spontaneous or severe pain must be taken seriously and these emergencies must be ruled out definitively.
9. Pregnancy should be managed with a high-risk obstetric team.
10. Exercise is still recommended for people with VEDS. Intense exercise, such as heavy weightlifting, should be avoided. Favor a non-competitive activity performed at a pace that permits conversation, such as brisk walking, leisurely bicycling, slow jogging, shooting baskets, leisurely tennis or swimming, and use of light weights without straining. The Marfan Foundation Physical Activity Guidelines for people with Marfan syndrome may be useful.

[marfan.org/wp-content/uploads/2021/09/FINAL-Physical-Activity-Guidelines-11\\_17.pdf](http://marfan.org/wp-content/uploads/2021/09/FINAL-Physical-Activity-Guidelines-11_17.pdf)

## Suggested Care Modifications

1. Colonoscopies and endoscopies can cause life-threatening damage to tissues 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. 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 because of the bowel inflation used for clarity of images.
3. Constipation should be avoided. To treat constipation, 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. Enemas should only be done under supervision, as they can cause increased pressure on tissue and lead to rupture.
4. Avoid aspirin and NSAIDs (if possible, because of increased bruising) and be aware that narcotic pain medications can lead to severe constipation.
5. Catheter angiography should generally be avoided for individuals with VEDS unless absolutely necessary to localize source of bleeding, due to the risk of arterial ruptures and dissections associated with the high pressure injections used during the invasive procedure.
6. Fluoroquinolone antibiotics should be avoided in people with VEDS due to the increased risk of aneurysms and dissections. See FDA warning.

[www.fda.gov/drugs/drug-safety-and-availability/fda-warns-about-increased-risk-ruptures-or-tears-aorta-blood-vessel-fluoroquinolone-antibiotics?utm\\_campaign=Local%20%26%20Community%20Events&utm\\_source=hs\\_email&utm\\_medium=email&utm\\_content=2&\\_hsenc=p2ANqtz-87CgYUwwWfl-PYkFjOfmyuLhGholR7txFlsTUWZtFypG1OIQ82MWsgtZFCd\\_GYsevQZlwnULznabx0JcXW7tPIQoGX4w&\\_hsmi=2](http://www.fda.gov/drugs/drug-safety-and-availability/fda-warns-about-increased-risk-ruptures-or-tears-aorta-blood-vessel-fluoroquinolone-antibiotics?utm_campaign=Local%20%26%20Community%20Events&utm_source=hs_email&utm_medium=email&utm_content=2&_hsenc=p2ANqtz-87CgYUwwWfl-PYkFjOfmyuLhGholR7txFlsTUWZtFypG1OIQ82MWsgtZFCd_GYsevQZlwnULznabx0JcXW7tPIQoGX4w&_hsmi=2)

***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](http://TheVEDSMovement.org/ask).***

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