THE VEDS MOVEMENT

Charging forward. Saving lives.

VEDS for the Pulmonologist

Considerations for the Pulmonologist

1. Because of the fragile vessels and tissues associated with VEDS, your patient may experience lung hemorrhage, lung cavities, lung cysts, pneumothorax, blebs, and sleep apnea.
2. About 16% of people with VEDS will experience pulmonary hemorrhage (hemoptysis, hemothorax), lung nodules (solid or cavitary), thin-walled cysts, or pneumothorax
3. Your patient can experience spontaneous lung laceration and spontaneous vascular injury.
4. In many people with VEDS, recurrent low-grade hemoptysis is common. Thresholds for clinical intervention should be established collaboratively between the pulmonologist and the knowledgeable geneticist and discussed in sufficient detail with the patient and family.
5. Pneumothorax is a feature of VEDS and often precedes a diagnosis of VEDS. For young people who present with a pneumothorax, VEDS, as well as other genetic disorders such as Marfan syndrome, should be considered as a possible cause, especially with recurrence.
6. Sleep apnea can be life-threatening, with high clinical impact. Experts think that the pressure changes in the chest caused by obstructive sleep apnea can stress soft tissues causing some trauma. However, more data is needed to validate this concern in VEDS.
7. 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. Collaborate.

Suggested Care Modifications

1. Angiography can cause life-threatening damage to vascular tissues in people with VEDS. If angiography is unavoidable, it should be done with a team aware of the underlying diagnosis, including a vascular surgeon, at a center with expert knowledge.
2. If a bronchoscopy or lung biopsy is necessary, obtain input from a VEDS specialist prior to proceeding. A second opinion of the lung biopsy may be needed to confirm diagnosis.
3. If pleurodesis is required, mechanical pleurodesis is preferred over chemical pleurodesis for people with VEDS because there is less thoracic trauma and less engagement of pleural wound repair cascades compared with chemical pleurodesis. In addition, any tissue problem that occurs is under direct visualization with mechanical pleurodesis.
4. Efforts at durable re-expansion should observe surgical considerations of tissue fragility.
5. For individuals with asthma, discourage use of bronchodilators that are beta-agonists, as they can increase heart rate and blood pressure. Consider steroid inhalers and anticholinergics as first-line medications before proceeding to beta-agonists.

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 lung.
- **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