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# Research Week 2024

## Venous Manifestations in Vascular Ehlers Danlos Syndrome

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### Keywords

Ehlers-Danlos syndrome, vascular EDS, venous disease, varicose veins

### Abstract

Vascular Ehlers-Danlos syndrome (vEDS) is a heritable connective tissue disorder that predisposes to serious cardiovascular disease including aortopathy and arteriopathy. Many patients are initially diagnosed following severe complications such as aortic dissection, with delays in diagnosis associated with increased risk of morbidity and death. To facilitate early diagnosis, major and minor criteria are used to prompt further workup in patients with suspected vEDS. Early-onset varicose veins are one of the minor criteria used in diagnosis of vEDS. However, venous disease continues to be an under-described component, leading to poorly-defined incidence and management. To evaluate the current state of research on venous disease in vEDS, a review was performed by searching published literature in PubMed until November 21, 2023, and the resulting 156 publications were screened based on abstracts. Varicose veins were the most commonly described manifestation, and numerous treatment modalities were utilized, including vein stripping and endovenous laser ablation, each exhibiting differing degrees of success. Additionally, some cases identified vEDS patients who developed an arteriovenous fistula (AVF) or aneurysm, while other patients with unconfirmed vEDS underwent surgery for AVF or pseudoaneurysm, and subsequently received a post-operative vEDS diagnosis. Deep vein thrombosis and pulmonary embolism are the most feared manifestations of venous disease, but our review found no evidence of increased vulnerability to venous thromboembolism. The spectrum of presentations, ages, and timing of vEDS diagnosis following venous disease treatment emphasizes the importance of establishing evidence-based guidelines for more standardized diagnosis. Given the diverse factors that influence venous disease in vEDS, patient-specific care is critical for managing the complexity of an individual case. Future steps will involve using a large vEDS database to correlate various venous disease manifestations to associated risks, treatments, and outcomes in a clinical population. This groundwork will strengthen utility of venous disease as a diagnostic criterion for vEDS.