

PubMed

**Format:** Abstract[Full text links](#)Am J Med Genet C Semin Med Genet. 2017 Mar;175(1):168-174. doi: 10.1002/ajmg.c.31543. Epub 2017 Feb 4.

Cardiovascular autonomic dysfunction in Ehlers-Danlos syndrome-Hypermobile type.

Hakim A, O'Callaghan C, De Wandele I, Stiles L, Pocinki A, Rowe P.

Abstract

Autonomic dysfunction contributes to health-related impairment of quality of life in the hypermobile type of Ehlers-Danlos syndrome (hEDS). Typical signs and symptoms include tachycardia, hypotension, gastrointestinal dysmotility, and disturbed bladder function and sweating regulation. Cardiovascular autonomic dysfunction may present as Orthostatic Intolerance, Orthostatic Hypotension, Postural Orthostatic Tachycardia Syndrome, or Neurally Mediated Hypotension. The incidence, prevalence, and natural history of these conditions remain unquantified, but observations from specialist clinics suggest they are frequently seen in hEDS. There is growing understanding of how hEDS-related physical and physiological pathology contributes to the development of these conditions. Evaluation of cardiovascular symptoms in hEDS should include a careful history and clinical examination. Tests of cardiovascular function range from clinic room observation to tilt-table assessment to other laboratory investigations such as supine and standing catecholamine levels. Non-pharmacologic treatments include education, managing the environment to reduce exposure to triggers, improving cardiovascular fitness, and maintaining hydration. Although there are limited clinical trials, the response to drug treatments in hEDS is supported by evidence from case and cohort observational data, and short-term physiological studies. Pharmacologic therapy is indicated for patients with moderate-severe impairment of daily function and who have inadequate response or tolerance to conservative treatment. Treatment in hEDS often requires a focus on functional maintenance. Also, the negative impact of cardiovascular symptoms on physical and psycho-social well-being may generate a need for a more general evaluation and on-going management and support.

© 2017 Wiley Periodicals, Inc.

KEYWORDS: Ehlers-Danlos; autonomic; hypotension; orthostatic; tachycardiaPMID: 28160388 DOI: [10.1002/ajmg.c.31543](https://doi.org/10.1002/ajmg.c.31543)

[Indexed for MEDLINE]

Publication type, MeSH terms, Supplementary concept**LinkOut - more resources**

