

Prevalence of Gastrointestinal, Cardiovascular, Autonomic, and Allergic Conditions in Hospitalized Patients with Ehlers-Danlos Syndrome: A Case-Control Study

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OBJECTIVE

The aim of this study is to assess the prevalence of gastrointestinal, cardiovascular, autonomic, and allergic manifestations in hospitalized patients with Ehlers-Danlos syndrome (EDS). Previous observations suggest an association between EDS and dysfunction of the digestive, cardiovascular, immune, and autonomic nervous systems. We sought to determine whether a hospital diagnosis of EDS, any type, is associated with a higher prevalence of gastrointestinal conditions, autonomic dysfunction, food allergies, and cardiovascular complications compared to hospitalized patients without EDS. We also evaluated hospital outcomes, which have not been previously explored.

METHODS

EDS cases and controls were acquired from the 2016 National Inpatient Sample (NIS) of the Healthcare Cost and Utilization Project (HCUP). Patients with EDS were identified using the International Classification of Diseases, Tenth Revision (ICD-10) diagnosis code Q79.6. In 2016, there were 2,007 discharges with a diagnosis of EDS. EDS cases were matched 1:2 to controls according to 5-year age intervals, gender, and month of admission. The study population included 6,021 individuals, 2,007 of which were EDS patients. Bivariate analyses were performed for case-control comparisons, and multivariate logistic regression models were used to assess and adjust for confounders.

RESULTS

GI abnormalities were found in 44% of EDS patients compared to 18% of controls (odds ratio [OR] = 3.57, confidence interval [CI]: 3.17-4.02, $P < 0.0001$), with irritable bowel syndrome, gastroparesis, and celiac disease strongly associated with EDS. Autonomic dysfunction, including postural orthostatic tachycardia syndrome (POTS), neurocardiogenic syncope, and orthostatic hypotension, was found in 20% of EDS patients versus 6% of controls (OR = 4.45, CI: 3.71-5.32, $P < 0.0001$). Several conditions were found almost exclusively in EDS patients, such as POTS (OR = 223.77, CI: 31.21-1604.46, $P < 0.0001$) and other disorders of the autonomic nervous system (OR = 54.72, CI: 7.43-403.00, $P < 0.0001$). EDS patients were 3.88 times more likely to have a food allergy (CI: 2.65-5.66, $P < 0.0001$) and 6.16 times more likely to have cardiovascular complications, including mitral valve disorders, aortic aneurysm, and cardiac dysrhythmias (CI: 4.60-8.23, $P < 0.0001$). On multivariate logistic regression analysis, GI conditions, autonomic dysfunction, food allergies, and cardiovascular complications remained highly-associated with EDS after considering potential confounders. EDS patients were 76% more likely to have longer-than-average hospital stays, defined as > 4 days (OR = 1.76, CI: 1.54-2.02, $P < 0.0001$).

CONCLUSIONS

GI, cardiovascular, autonomic, and allergic disorders are significantly more prevalent in EDS patients compared to the general population of hospitalized patients without EDS. EDS patients may be more likely to face adverse complications during hospitalization, requiring longer hospital stays than those unaffected by EDS.

Physicians should consider EDS in hospitalized patients presenting with multiple unexplained GI, cardiovascular, autonomic, or allergic manifestations and exercise precautions when treating EDS patients in a hospital setting.

DECLARATIONS OF INTEREST

None