



## **Self-Reported Changes in Health Status During the Coronavirus Pandemic: A Survey of Individuals with Ehlers-Danlos Syndrome and Hypermobility Spectrum Disorder.**

Hakim A, Bloom L, Weber S, Gluck R, Bergfield M, Malfait F, Francomano C A, Gandy W

The Ehlers-Danlos Society

### **Background**

People living with the Ehlers-Danlos Syndromes (EDS) and hypermobility spectrum disorders (HSD) experience many associated conditions that cause significant ill-health and reduced quality of life <sup>1,2</sup>.

From the onset of the Coronavirus (COVID-19) pandemic, the Ehlers-Danlos Society learned from its Helpline and Support Group program that members of the EDS and HSD community were experiencing a deterioration in well-being considered to be a consequence of disruption to normal levels of care, and, imposed social restrictions.

The Global Registry for EDS, HSD, related symptoms, and associated conditions was established in 2016<sup>3</sup>. To date, the Registry houses diagnostic, demographic, and medical history data on over 10,000 participants of all ages worldwide with either EDS or HSD. Using the Global Registry, the aim of the study was to explore the impact of the COVID-19 pandemic on the EDS and HSD community.

### **Methods:**

A survey was constructed and placed on the Registry for all registrants to participate in of their free will. The study gained IRB approval from Genetic Alliance (EDSRegistry001). The survey included questions on diagnosis, demographics, economic status and impact of the pandemic, Coronavirus symptoms and testing, access to ongoing routine/elective and urgent/emergency care, changes in symptoms related to a number of associated conditions, and, the overall impact of changes on well-being.

The survey was run in June and July 2020, during the international height of the current phase of the pandemic.

### **Results:**

The survey was completed by 1334 registrants. The average age of responders was 40 years (range 6 to 81). 91% of responders were female, 5% male, and 4% non-binary. By race or ethnicity 89% self-reported being White, 5% Hispanic, 3% Alaskan or American Indian, 1% Black American or Afro-Caribbean, 1% Asian, and 1% other.

Figure 1 shows the distribution of responders by country or region of residence. 69% of responders were from the USA, and 20% from Great Britain. Two-thirds (67%) had a diagnosis of hEDS (Figure 2).

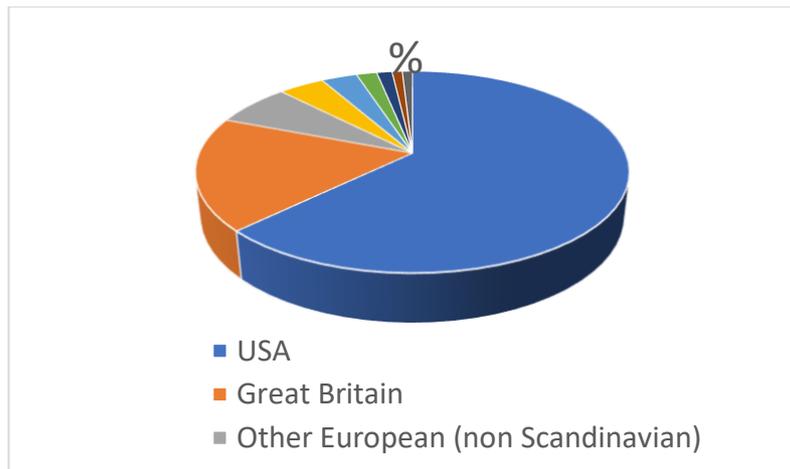


Figure 1. Distribution of survey responders by country or region of residence

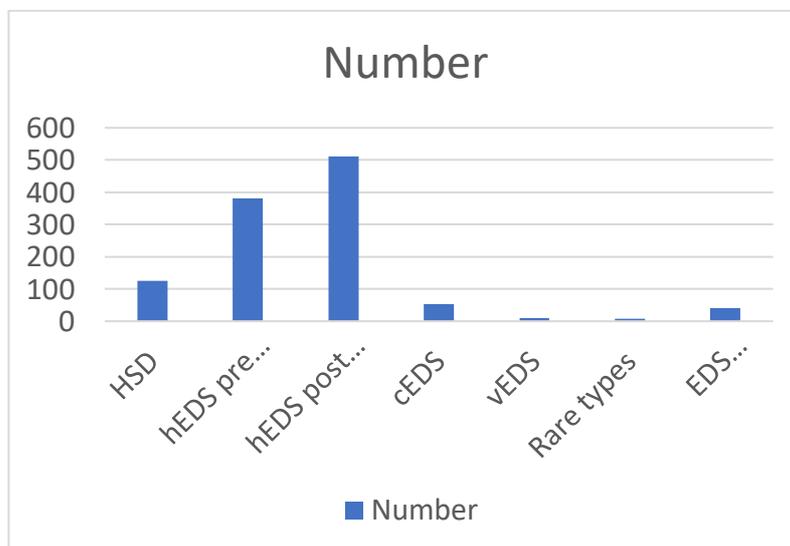


Figure 2. The distribution of diagnoses (n=1344)

The cohort had a high burden of associated symptoms and conditions that were reported as present prior to the COVID-19 pandemic. Widespread musculoskeletal chronic pain, and fatigue were almost universal. Figure 3 shows the distribution of conditions and concerns across all responders at the time of the survey.

With regard to symptoms related to COVID-19 infection, 70% reported no symptoms, 15% reported symptoms but were negative for COVID-19 on testing, 12.5% reported symptoms but had not been tested for COVID-19, and 2.5% had confirmed infection.

Three quarters (75%) of responders reported disruption to their doctors' clinic services; 50% disruption to physiotherapy, occupational therapy, or psychotherapy services; and, 20% disruption to pharmacy services.

When asked overall how disruptions to services had affected well-being, 51% reported a moderate to severe, and 31% a mild negative impact on their health (Figure 4) i.e. 82% of the cohort.

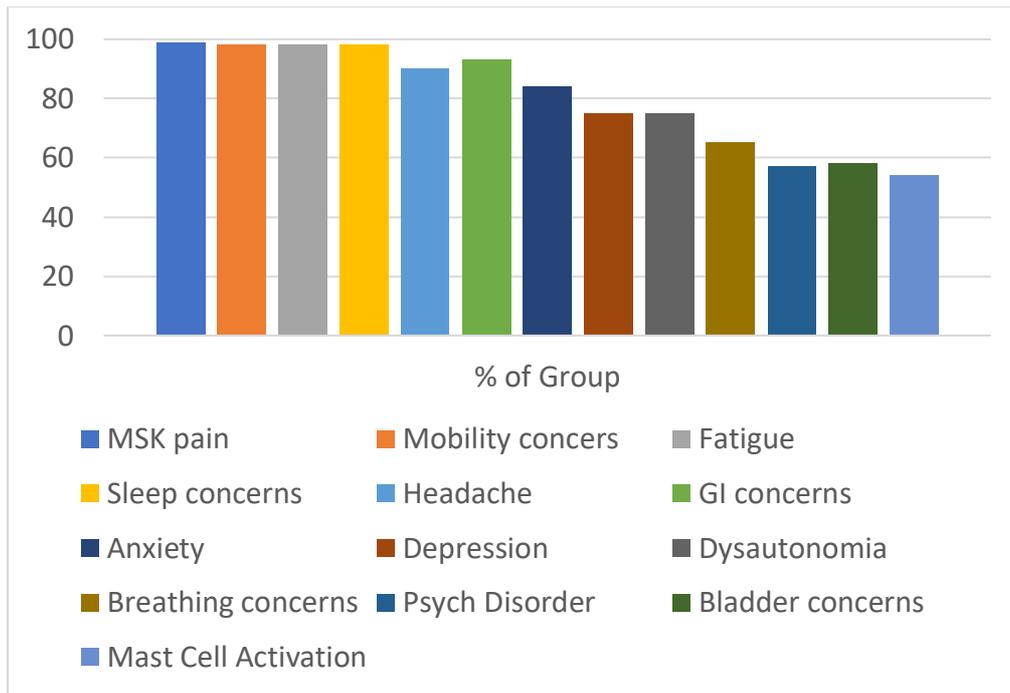


Figure 3. Prevalence of associated symptoms and concerns (n=1344)

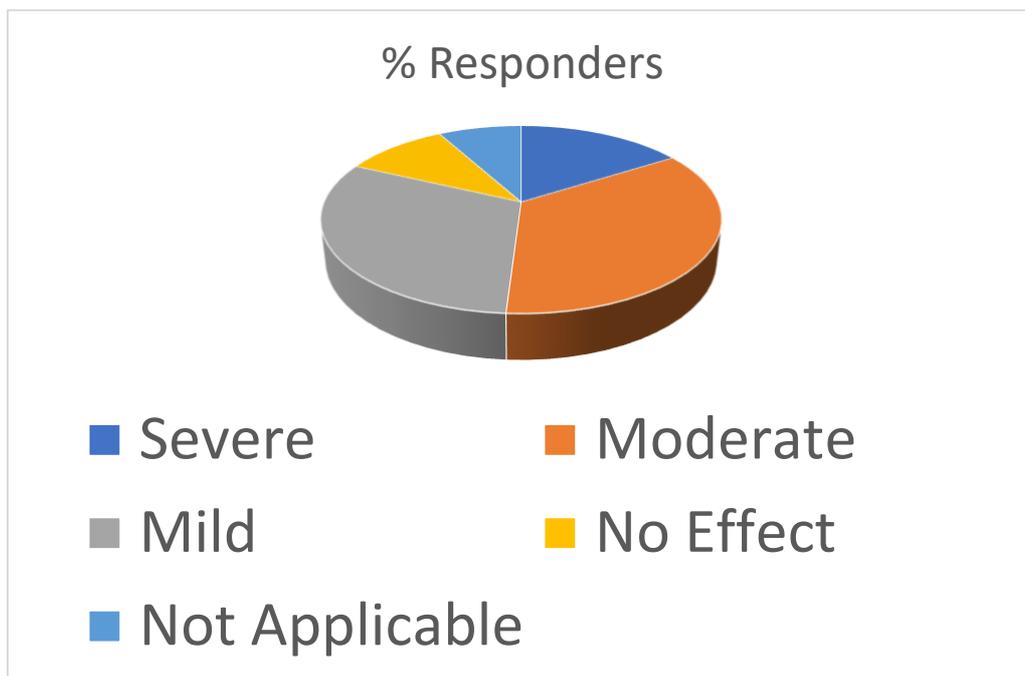


Figure 4. Perceived negative impact on well-being as consequence of changes in access to healthcare (n=1344).

With regard to symptoms and associated concerns, two-thirds or more reported somewhat worsening or significant worsening of musculoskeletal pain and mobility, fatigue, quality of sleep, headache, anxiety, and depression.

Among other areas of health, half reported their gastrointestinal concerns to be the same or better, and half worse. 60% reported symptoms of dysautonomia, or mast cell activation, or

respiratory concerns to the same or better, and 40% worse. 75% reported bladder concerns to be the same or better.

### **Conclusions**

The cohort reported a significant burden of disease that was present prior to the current pandemic and restrictions placed on society. Across the conditions explored, at least 2 or 3 out of every 5 individuals reported a moderate to severe deterioration in at least 1 of their health concerns since the onset of the pandemic.

Given that at least 85% of responders had either no symptoms or had a negative test for COVID-19, infection with COVID-19 cannot explain the change in symptoms in the majority of cases.

It could be argued that individuals would be expected to experience worsening of symptoms as part of the natural history of such conditions. However, we are not aware of studies that have reported such changes over a period of 3-6 months in any situation or in a population of this size with previously stable symptoms, let alone report such a degree of change in terms of the proportion of individuals affected.

The data will be explored further for correlations; looking, for example, at relationships between symptom severity and access to care; and, correlations between anxiety and depression and the number and severity of other symptoms and concerns.

### **References**

1. Tinkle B, Malfait F, Francomano CA and Byers PH (Eds.). (2017) The Ehlers-Danlos Syndromes: Reports from the International Consortium on the Ehlers-Danlos Syndromes [Special Issue]. *Am J Med Genet Part C*. 175 (1), 1 -245
2. Hakim A J, Severity classes in adults with hypermobile Ehlers-Danlos syndrome/hypermobility spectrum disorder, *Rheumatology*, 2019; 58 (10): 1705–1706
3. <https://www.ehlers-danlos.com/eds-global-registry/> (accessed September 2020)

### **Conflict of Interest**

The Ehlers-Danlos Society owns the Global Registry in EDS and HSD.