



DATA • INCLUSION • COLLABORATION • EXCELLENCE

DICE Global Registry Data Guide

Version 1. 2023

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Introduction

The Ehlers-Danlos Society DICE Global Registry is an extensive repository of demographic, morbidity, and co-morbidity data from people of all ages across the Ehlers-Danlos syndromes (EDS) and hypermobility spectrum disorders (HSD). Providing rapid and cost-free access to a large community, the registry is rich with data for cohort and nested case-control studies and is suited to interpopulation comparisons. In addition, it provides access to specific groups, identified as required by their demographics and health concerns, for invited participation in new studies. We encourage you to consider the possibilities the DICE Registry can bring to your research, and we look forward to exploring opportunities to assist and collaborate with you in the support of the EDS and HSD community.

- Associate Prof. Dr Alan Hakim
*Chief Medical Officer and Co-PI DICE Global Registry
The Ehlers-Danlos Society*

We are pleased to invite researchers to use this data guide as a starting point for requesting data from the DICE Global Registry. The following sections outline the data available, the details of which are available after a request has been initiated.

Replication and unauthorized distribution of this guide is prohibited.

Data use

Acceptable uses

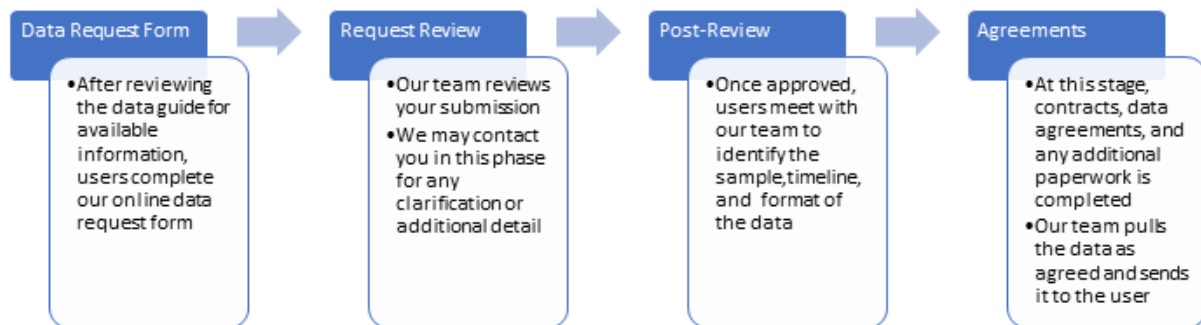
DICE Registry data is available to approved researchers with the aim of advancing research in EDS, HSD, and their related comorbidities. All data must be cited to accurately credit The Ehlers-Danlos Society, and at all times used in accordance with our [Privacy Policy](#), as well as any agreements made prior to sharing the data.

Under no circumstances should data provided by The Ehlers-Danlos Society be used for any purpose other than the purpose approved by The Ehlers-Danlos Society. Sequential studies using the same dataset must be made known to The Society in writing prior to initiation.

Data Access Tiers

| DICE Third Party Access | | |
|---|--|--|
| Tier 1 | Tier 2 | Tier 3 |
| <p>All de-identified data except:</p> <ul style="list-style-type: none"> • Age* • Postal code • Redacted genetic letters • Year of diagnosis** | <p>All Tier 1 data and any of:</p> <ul style="list-style-type: none"> • Age • Postal code • Redacted genetic letters • Year of diagnosis | <p>All Tier 1 & 2 data and any of:</p> <ul style="list-style-type: none"> • Sensitive and/or identified data (Tier 3a)*** • Data generated through collaborative data collection (Tier 3b) |
| <p>* Age range only ** Can offer ranges and/or pre- post- 2017 categories if needed</p> | | <p>***Examples include contact information, participant-uploaded documents</p> |
| <ul style="list-style-type: none"> • Data request agreement required • Data Sharing Agreement may be required in some scenarios | <ul style="list-style-type: none"> • Data request agreement required • Data Sharing Agreement may be required in some scenarios | <ul style="list-style-type: none"> • Data request agreement required • Data Sharing Agreement required • Collaborator contract required (Tier 3b) |

Overview of data request process



All users of DICE Global Registry Data are required to comply with the relevant data sharing, data protection, and data use guidelines.

Registry Sections:

The following sections provide a general outline of the data found in the DICE Global Registry sections. Please note that all data is de-identified, and direct contact is only facilitated between third parties and DICE participants via Tier 3.

Participant contact and re-contact

We recognize the need in some studies for re-contacting of registry participants for further data acquisition specific to the study. The DICE Global Registry team, complying with data sharing regulation, will work with the researcher to facilitate this.

Demographics

| Availability by Data Access Tier | | | |
|----------------------------------|------------|--------|--------|
| Datapoint | Tier 1 | Tier 2 | Tier 3 |
| Age | Range only | X | X |
| Country | X | X | X |
| Province/State | X | X | X |
| Postal code/zipcode | | X | X |
| Race | X | X | X |
| Employment status | X | X | X |
| Education level | X | X | X |
| Sex at birth | X | X | x |
| Gender | X | X | X |
| Sexual orientation | X | X | X |

EDS or HSD Diagnosis

| Availability by Data Access Tier | | | |
|--|------------|--------|--------|
| Datapoint | Tier 1 | Tier 2 | Tier 3 |
| Diagnosis status (diagnosed, suspected, etc) | X | X | X |
| Year of diagnosis | Range only | X | X |
| Diagnosis | X | X | X |
| Genetic testing confirmation status | X | X | X |
| Genetic testing results* | | X | X |

Family History

| Availability by Data Access Tier | | | |
|--|--------|--------|--------|
| Data | Tier 1 | Tier 2 | Tier 3 |
| Diagnosis by generation (great-grandparents through grandchildren) | X | X | X |
| Genetic confirmation status by generation (great-grandparents through grandchildren) | X | X | X |
| Symptomatic status by generation (great-grandparents through grandchildren) | X | X | X |
| Paternal/maternal patterns | X | X | X |
| Symptomatic patterns within generations (siblings affected vs. unaffected) | X | X | X |
| Multiple types of EDS diagnosed within families | X | X | X |

General Health

| Availability by Data Access Tier | | | |
|----------------------------------|--------|--------|--------|
| Data | Tier 1 | Tier 2 | Tier 3 |
| Fatigue | X | X | X |
| Sleep issues | X | X | X |
| Weight fluctuation | X | X | X |
| Rare-type signals | X | X | X |

Pain Symptoms

| Availability by Data Access Tier | | | |
|---|--------|--------|--------|
| Data | Tier 1 | Tier 2 | Tier 3 |
| Frequency of pain | X | X | X |
| Pain interference with daily activities | X | X | X |
| Pain medications and interventions | X | X | X |
| Use of dietary changes for pain reduction | X | X | X |
| Use of physical and manual therapies for pain reduction | X | X | X |
| Use of cognitive and mindful therapies for pain reduction | X | X | X |
| Supports and assistive devices for pain reduction | X | X | X |

Joints, Muscles, and Bones

| Availability by Data Access Tier | | | |
|--|--------|--------|--------|
| Data | Tier 1 | Tier 2 | Tier 3 |
| Scoliosis diagnosis | X | X | X |
| Joint pain frequency and location | X | X | X |
| Muscle pain frequency | X | X | X |
| Muscle weakness | X | X | X |
| Hypermobility and instability in joints | X | X | X |
| Frequency and location of dislocated joints | X | X | X |
| Frequency and location of joint subluxations | X | X | X |

Hands and Feet

| Availability by Data Access Tier | | | |
|--|--------|--------|--------|
| Data | Tier 1 | Tier 2 | Tier 3 |
| Presence of foot issues (flat/high arches, ankle sprains, club foot, etc.) | X | X | X |
| Presence of piezogenic papules | X | X | X |
| Presence of Steinberg sign | X | X | X |
| Presence of Walker sign | X | X | X |

Gynecological

| Availability by Data Access Tier | | | |
|-------------------------------------|--------|--------|--------|
| Data | Tier 1 | Tier 2 | Tier 3 |
| Pregnancy related questions | X | X | X |
| Uterine prolapse history | X | X | X |
| Uterine rupture history | X | X | X |
| Polycystic Ovary Syndrome diagnosis | X | X | X |

Urinary

| Availability by Data Access Tier | | | |
|---|--------|--------|--------|
| Data | Tier 1 | Tier 2 | Tier 3 |
| Presence of urinary symptoms (urgency, incontinence, etc) | X | X | X |

Gastrointestinal

| Availability by Data Access Tier | | | |
|---|--------|--------|--------|
| Data | Tier 1 | Tier 2 | Tier 3 |
| Presence of gastrointestinal symptoms (abdominal pain, bloating, etc) | X | X | X |
| Frequency of GI symptoms | X | X | X |
| Bowel perforation history | X | X | X |
| Abdominal hernias | X | X | X |

Allergy, Autoimmune, and Endocrine

| Availability by Data Access Tier | | | |
|--|--------|--------|--------|
| Data | Tier 1 | Tier 2 | Tier 3 |
| Autoimmune diagnoses (Addison's, Graves' disease, Pernicious Anemia, etc) | X | X | X |
| Severe infection frequency | X | X | X |
| Hormone-related symptoms (temperature instability, menstrual disruptions, etc) | X | X | X |
| Allergy diagnoses (asthma, allergic rhinitis) | X | X | X |
| Allergy symptoms (anaphylaxis, angioedema, rash, etc) | X | X | X |
| Mast Cell Activation Syndrome diagnosis | X | X | X |

Mental Health

| Availability by Data Access Tier | | | |
|---|--------|--------|--------|
| Data | Tier 1 | Tier 2 | Tier 3 |
| Mental illness diagnoses (anxiety, depression, bipolar disorder, etc) | X | X | X |
| Neurodevelopmental disorders (ADHD, Autism spectrum disorders, cerebral palsy, etc) | X | X | X |
| Disability and impairments (hearing impairment, visual impairment, etc) | X | X | X |

Neurological & Spinal

| Availability by Data Access Tier | | | |
|---|--------|--------|--------|
| Data | Tier 1 | Tier 2 | Tier 3 |
| Presence of neurological and spinal musculoskeletal conditions (atlantoaxial instability, carpal tunnel, Chiari, etc) | X | X | X |
| Presence of neurological symptoms (confusion, fainting, headache, double vision, etc) | X | X | X |

Cardiovascular

| Availability by Data Access Tier | | | |
|---|--------|--------|--------|
| Data | Tier 1 | Tier 2 | Tier 3 |
| Cardiovascular conditions diagnosis (arrhythmia, chest pain, POTS, etc) | X | X | X |
| Blood vessel conditions (ruptures, aneurysms, dissections) | X | X | X |
| Carotid-cavernous sinus fistula diagnosis | X | X | X |
| Presence of enlargement/aneurysm at aortic root | X | X | X |
| Mitral valve prolapse diagnosis | X | X | X |

Head, Ears, Eyes, Nose, and Teeth

| Availability by Data Access Tier | | | |
|---|--------|--------|--------|
| Data | Tier 1 | Tier 2 | Tier 3 |
| Vision (hyperopia, myopia, floaters/spots) | X | X | X |
| Hearing loss | X | X | X |
| ringing in ears | X | X | X |
| Oral features (dental crowding, tooth abnormalities, high palate) | X | X | X |
| Oral symptoms (dry mouth, gum disease) | X | X | X |

Respiratory

| Availability by Data Access Tier | | | |
|----------------------------------|--------|--------|--------|
| Data | Tier 1 | Tier 2 | Tier 3 |
| Chronic cough | X | X | X |
| Difficulty breathing | X | X | X |
| Shortness of breath | X | X | X |
| Wheezing | X | X | X |

Hair and skin symptoms

| Availability by Data Access Tier | | | |
|--|--------|--------|--------|
| Data | Tier 1 | Tier 2 | Tier 3 |
| Bruising at unusual sites | x | x | x |
| Easy bruising | x | x | x |
| Hair falling out | x | x | x |
| History of skin tears with minimal or no trauma | x | x | x |
| Hives | x | x | x |
| Itching | x | x | x |
| Poor wound healing | x | x | x |
| Rashes | x | x | x |
| Scars that are wide or sunken (at least two) | x | x | x |
| Severe scarring | x | x | x |
| Stretch marks NOT relate to weight gain | x | x | x |
| Thin hair | x | x | x |
| Unexplained bruising | x | x | x |
| Unexplained stretch marks | x | x | x |
| Unusually soft or velvety skin | x | x | x |
| Unusually stretchy skin (that can be pulled out more than usual) | x | x | x |

Blood and lymphatic symptoms

| Availability by Data Access Tier | | | |
|----------------------------------|--------|--------|--------|
| Data | Tier 1 | Tier 2 | Tier 3 |
| Prolonged bleeding | x | x | x |
| Frequent nose bleeds | x | x | x |
| Unexplained swollen lymph nodes | x | x | x |