What are Ehlers-Danlos syndromes?

These are Ehlers-Danlos syndromes

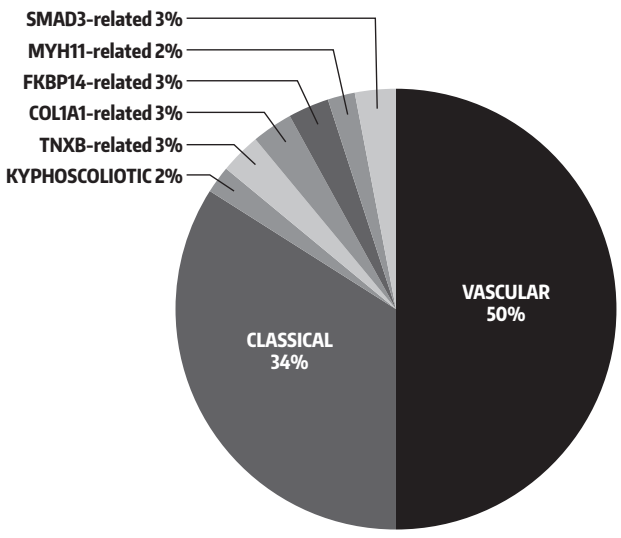
Ehlers-Danlos syndromes (EDS) are a group of connective tissue gene disorders that produce complex problems across multiple systems of the body. There are no cures.

Connective tissue disorders disrupt the most fundamental processes and structures of the body, so resulting problems can be widespread, in a wide range of severities, and affect areas that might seem to be otherwise unrelated.

There are at least six types of EDS; each type of EDS has certain physical traits and with notable exception to the most common form, the hypermobile type of EDS, most types have a known disease-causing gene. The physical characteristics that are common to all types of EDS include hypermobile joints (joints that move in greater amounts than expected) and skin involvement (soft, stretchy, saggy, too thin, easy bruising, easy wounding, poor wound healing, or atrophic scarring). EDS are known to affect more than one in 2,500 men and women, of every race and ethnicity.

Diagnostic testing

Frequency of EDS type genetic testing in a large laboratory



Visit **ehlers-danlos.com** for more information.

Our strength begins with hope.

Our hope begins with you.

The Ehlers Danlos Society™

These are the types of Ehlers-Danlos

Most but not all of the genes responsible are identified, and new genes are still being identified in rare forms of EDS. Current diagnostic information for each type can be found on ehlers-danlos.com/eds-types. Diagnostic criteria don't define what the experience of EDS will be, as there are many more symptoms than criteria.

HYPERMOBILE [distinctive cause unidentified]: Most prevalent EDS type; Hypermobile EDS may be the most common heritable disorder of connective tissue, perhaps affecting more than one in 1000 people.

CLASSICAL [COL5A1, COL5A2, COL1A1]:
Rare; second-most prevalent.

VASCULAR [COL3A1]: Rare. The most serious type due to the possibility of arterial or organ rupture.

KYPHOSCOLIOTIC [PLOD1]: Very rare.

ARTHROCHALASIA [COL1A1, COL1A2]: Very rare.

DERMATOSPARAXIS [ADAMTS2]: Extremely rare.

The six types identified above were categorized in the 1997 Villefranche nosology.

The new nosology in process (to be published after peer review in Spring 2017) refines the diagnostic criteria for these types, and includes newer known rare forms of EDS such as TNX-deficient, progeroid, musculocontractural, spondylocheirodysplastic, FKBP22-deficient, periodontitis, and brittle cornea syndrome.

For the first time, treatment guidelines are also being prepared, and will be published with the revised diagnostic criteria.

CONCLUSION

EDS have varied presentations and in some types can be life-threatening. EDS are multi-systemic disorders. Symptoms can be treated as they arise. Care is largely preventative, to support and manage EDS with the intent of keeping damage as minimal as possible. Specifics have to be tailored to those symptoms exhibited in the person with EDS. Even knowing what type EDS one has, each case of EDS is individual.

Early diagnosis is crucial to positive patient health.

The future of Ehlers-Danlos syndromes is a global community of patients, caregivers, medical professionals, and supporters, dedicated to saving and improving the lives of those affected by Ehlers-Danlos and related disorders.

We support collaborative research initiatives, awareness campaigns, advocacy, community-building, and care for the EDS population.

Our goals are worldwide awareness – and a better quality of life for all who suffer from these conditions. Research is at the center of what we do, so that one day we will have a cure.