

Fact Sheet

Ehlers-Danlos Syndrome Pain

About

Ehlers-Danlos Syndrome (EDS) is a group of hereditary connective tissue disorders, characterized by joint hypermobility, skin extensibility and tissue fragility. Chronic, early onset, debilitating musculoskeletal pain is a symptom of EDS. Individuals with EDS have a defect in their connective tissue, the tissue that provides support to many body parts such as the skin, muscles and ligaments. The fragile skin and unstable joints found in EDS are the result of faulty collagen. Collagen is a protein, which acts as a “glue” in the body, adding strength and elasticity to connective tissue.¹

“ About 1 in 5,000 people has EDS. There is no cure.⁴ ”

Patients with Ehlers-Danlos Syndrome (EDS) are often undiagnosed or misdiagnosed for years. They may be told they are merely seeking attention or pain medications.² Clinicians may attribute their difficulties to “depression” and refer them to psychiatrists.³

When they seek an answer regarding their child’s pain or other symptoms, parents may hear they are simply “overprotective” of their undiagnosed child or may be suspected of child abuse because of frequent bruising or dislocations.¹

Because management of EDS-related pain has not been studied in depth, no treatment protocols exist. Even after correct diagnosis, unsuccessful trials of multiple medications or modalities may take their toll on patients and those who support them physically, emotionally, and financially.¹

Facts

About 1 in 5,000 people has EDS. There is no cure. Treatment involves managing symptoms. Individuals with EDS often experience both acute and chronic discomfort/pain related to a variety of causes.⁴

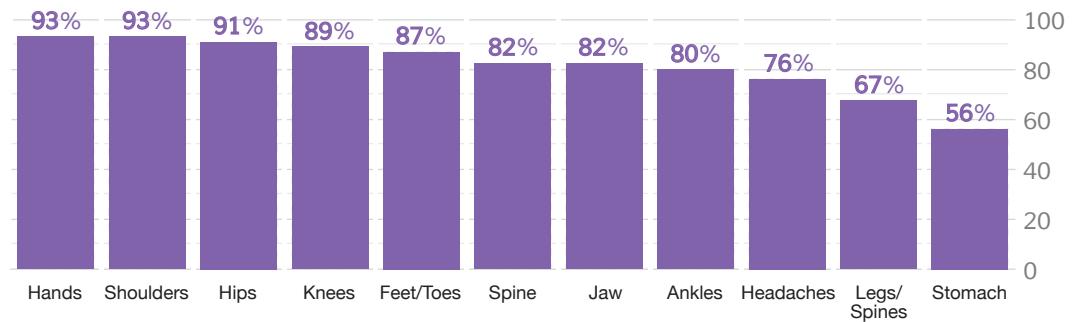
Pain is common and severe in EDS. The results of a large Danish study showed that:⁵

- chronic pain in EDS is highly prevalent and associated with regular use of medications;
- pain is more prevalent and more severe in the hypermobility type than in the classic type;

- pain severity is correlated with hypermobility, dislocations, and previous surgery;
- pain is correlated with low nocturnal sleep quality; and
- pain contributes to functional impairment in daily life, independently of the level of fatigue.

Acute discomforts from EDS are often the results of fractures, dislocations, subcutaneous bleeding or bruising after falls.²

Chronic pain from EDS may be constant yet changeable throughout the day. It interferes with sleep or activities of daily living. Distribution of chronic pain in individuals with EDS:²



There are six major types of EDS. The different types of EDS are classified according to the signs and symptoms that are manifested. Each type of EDS is a distinct disorder that “runs true” in a family. This means that an individual with Vascular Type EDS will not have a child with Classical Type EDS. The types of EDS include:⁶

- **Hypermobility** (Formerly EDS Type III) where joint hypermobility is the dominant clinical manifestation. Chronic joint and limb pain is a common complaint among individuals with the Hypermobility Type.
- **Classical** (Formerly EDS Types I & II) where marked skin hyperextensibility (stretchy) with widened atrophic scars and joint hypermobility are found.
- **Vascular** (Formerly EDS Type IV) is generally regarded as the most serious form of EDS due to the possibility of arterial or organ rupture. The skin is usually thin and translucent with veins being seen through the skin. This is most apparent over the chest and abdomen. There are certain facial characteristics present in some affected individuals. These manifestations include large eyes, thin nose, lobeless ears, short stature and thin scalp hair.
- **Kyphoscoliosis** (Formerly EDS Type VI) is characterized by general joint laxity and severe muscle hypotonia (weak muscle tone) at birth are seen in this type of EDS. The muscular hypotonia can be very pronounced and leads to delayed gross motor development. Individuals with the Kyphoscoliosis Type present with Scoliosis at birth that is progressive.

- **Arthrochalasia** (Formerly EDS Type VII A&B). Congenital hip dislocation has been present in all biochemically proven individuals with this type of EDS. Severe generalized joint hypermobility with recurrent subluxations are seen in individuals with this type of EDS.
- **Dermatosparaxis** (Formerly EDS Type VIIC). Individuals with Dermatosparaxis Type EDS have severe skin fragility and substantial bruising.

Other classifications

The current EDS type V (X-linked) has been described in a single family. It is a rare variant and the molecular basis of which remains unknown. The current EDS type VIII is similar to the Classical Type except that in addition it presents with periodontal friability. This is a rare type of EDS. The existence of this syndrome as an autonomous entity is uncertain. The EDS type IX was previously redefined as “Occipital Horn syndrome,” an X-linked recessive condition allelic to Menkes syndrome. This was previously removed from the EDS classification. The current EDS type X has been described in only one family. The EDS type XI termed “Familial Joint Hypermobility syndrome” was previously removed from the EDS classification. Its relationship to the EDS is not yet defined.

Additional Resources

The Ehlers-Danlos National Foundation

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www.ednf.org

Ehlers-Danlos Syndrome Network

C.A.R.E.S. Inc.
 P.O. Box 66
 Muskego, WI 53150
 Phone: (203) 514-2851
 Fax: (203) 514-2851
 E-mail: edsnetwork@wi.rr.com
www.ehlersdanlosnetwork.org

Ehlers-Danlos Syndrome Support Group

PO Box 748
 Borehamwood WD6 9HU
 Phone: 0208-736-5604
 E-mail: contact via website
www.ehlers-danlos.org

National Institute of Arthritis and Musculoskeletal and Skin Diseases

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Resources verified July 2011.

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