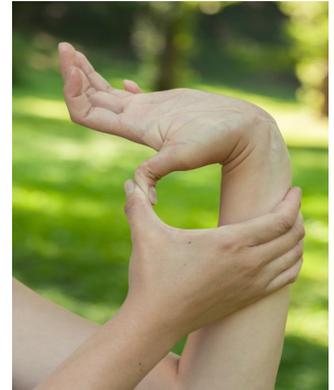


What is it?

Ehlers-Danlos syndrome (EDS) is a group of disorders affecting connective tissue that supports skin, muscles, ligaments, blood vessels, organs, eyes and even gums. This syndrome is thought to alter the body's composition of collagen. As a protein, collagen is supposed to provide a stable structure. When the body's muscles, ligaments and organs are built on defective collagen, the entire structural system is weak and unstable. As a connective tissue disorder, people with Ehlers-Danlos syndrome characteristically have flexible joints with stretchy, yet fragile skin.



What are the signs and symptoms?

Defects in connective tissue cause the signs and symptoms of EDS. There are several classifications and unique features of the disease. Each type is defined by a distinct problem in making or using the type of collagen affected. About 1 in 5,000 people may be affected in some degree, regardless of gender or race. Note that tiredness and fatigue described as “brain fog” is also often reported.

- Joints: Hypermobility, increased or excessive range of motion, unstable, frequent dislocations, pain, early onset osteoarthritis, impaired mobility, poor coordination
- Skin: Fragile, stretchy, soft, saggy, thin, easily bruised, easily wounded, poor healing, excessive scarring
- Other (less common): Early onset and debilitating musculoskeletal pain, scoliosis, poor muscle tone, gum disease, arterial/intestinal/uterine rupture

What are the types?

- ⇒ Classic: Very stretchy skin with scarring of pressure points and even tumors. Other tissue damage can occur, ranging from hiatal hernia to anal prolapse. Common for sprains, strains, dislocations to occur.
- ⇒ Hypermobility: Joints are especially loose in large and small joints like elbows, knees, fingers and toes. Other joints like the shoulder, knee, and jaw dislocate frequently resulting in chronic pain. Skin bruising may occur but will vary in severity.
- ⇒ Vascular type: Most serious and life-threatening form of EDS as caused by potential organ or artery rupture. Skin tends to be translucent with visible veins. Unique facial features include large eyes, thin nose, lobeless ears, and thin scalp hair. Increased joint mobility is most commonly seen in the fingers.
- ⇒ Kyphoscoliosis type: Characterized by severe muscle hypotonia leading to delays in motor development. Scoliosis is almost immediately diagnosed as it continues to progress.
- ⇒ Arthrochalasia Type: Congenital hip dysplasia present with other joints suffering hypermobility as well.
- ⇒ Dermatosparaxis type: Severely fragile, saggy skin and bruising. Wound healing is not affected.

Suggested school accommodations

- Consider 504
- PT/OT considerations
- Keep safe from predictable physical injury
- Emergency procedure for injury
- Plan for absences, make-up work
- Consider motor difficulties
- Elevator use
- Hallway considerations and extra time
- Adapted PE
- Rest breaks as needed
- Extra set of book for home
- Use of computer or assistive devices
- Additional work time
- Priority seating
- Cushioned seating
- Handwriting accommodations
- Support the child's acceptance and self-esteem

SHNIC school nurses information:

Specific health issues for individual health care plans

- Complete diagnosis documentation
- Current medication orders for school and home
- Orders and/or considerations for PRN pain meds, cool packs, heat packs, etc.
- Nutrition orders including feeding protocol or positioning for digestive issues
- Skin assessment
- Skin protection considerations (safety and sun)
- Orders for orthotics and braces
- Open communication to understand a full list of symptoms (they can vary day to month to year)
- Rest periods
- Emergency procedure for dislocation
- Emergency procedure for vascular event

Resources & Manuals

Ehlers-Danlos Society
<http://ehlers-danlos.com>

EDS Awareness

<http://www.chronicpainpartners.com/can-schools-better-understand-ehlers-danlos-syndrome/>

The Marfan Foundation
<http://www.marfan.org/>

Life as a Zebra Foundation

<http://www.zebration.org/>