EHLERS-DANLOS SYNDROME

What is it?

Ehlers-Danlos syndrome (EDS) is a group of inherited disorders affecting connective tissue primarily in skin, joints, and blood vessel walls. Connective tissue is a complex mixture of proteins and other substances that provide strength and elasticity to the underlying structures in the body. As a protein, collagen is responsible for providing a stable structure. When the body's muscles, ligaments and organs are built on defective collagen, the entire structural system becomes weak and unstable. As a connective tissue disorder, EDS is characterized by joint hypermobility, loose "stretchy" skin, and fragile tissues. These complex syndromes can affect many body systems at once and most are less visible than the skin and joint differences. Chronic pain and fatigue are hallmark complications of EDS.

What are the signs and symptoms?

Physical characteristics common for EDS include:

Joints	Hypermobile, unstable, increased or excessive range of motion, 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	Early onset and debilitating musculoskeletal pain, poor muscle tone, scoliosis, gum disease, arterial/intestinal/uterine rupture, vision problems

What are the types?

The major types of EDS are classified by clinical criteria, with a significant overlap between features. There are 13 total. Connective tissue disorders can range from mildly loose joints to life-threatening complications. Some of the types of EDS include:

Classic	Very stretchy skin, scarring of pressure points. 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 most present, other joints suffering hypermobility.
Dermatosparaxis type	Severely fragile, saggy skin and bruising.

Kennedy Krieger Institute

The **Specialized Health Needs Interagency Collaboration (SHNIC)** program is a collaborative partnership between the Kennedy Krieger Institute and the Maryland State Department of Education.

Suggested school accommodations

There is no evidence to indicate that EDS causes learning difficulties but speech, hearing, and vision problems have been associated. Students with EDS could require different needs or restrictions for their physical environment. It is important to guide students and staff that each activity should be accessed based on the individual student's abilities including impaired mobility, poor coordination, and/or weak hand control. Supporting students with this condition in the school require educators and parents/guardian to work as a team. Some accommodations to consider for a 504/IEP could include:

- PT/OT evaluation
- Use of computer or assistive devices
- Monitor fatigue, offer rest breaks
- Extra time for assignments
- Evaluate best method to assess for understanding of classroom material
- Handwriting accommodations
- Consider motor difficulties (physical layout of school, changes in flooring, adapted PE, use of elevator, extra time to transition between classes, rolling backpack)
- Extra set of books for home
- Modified school day
- Plan for absences and make up work
- Rest breaks as needed
- Priority seating (consider consult with OT regarding seating accommodations)
- Adjusted chair/table height
- Support the child's acceptance and self-esteem
- Communication/plan with student for chronic pain
- Staff education/training as appropriate
- Emergency Evacuation Plan (EEP)

Specific health issues for Individualized Healthcare Plan

- 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
- Plan/protocol for chronic pain
- Skin assessment, skin protection considerations
- Orders and protocol for orthotics and braces
- Open communication to understand a full list of symptoms (they can vary day to month to year)
- Communicate with school staff, parents/guardian, and provider any changes or concerns about the disease
- Emergency Care Plan(s) (ECP) related to medical needs in the school setting and staff education/training as appropriate for each

Resources & Manuals

Kennedy Krieger Institute: Pain Rehabilitation Program
https://www.kennedykrieger.org/patient-care/centers-and-programs/pain-rehabilitation-clinic-outpatient

The Ehlers Danlos Society
http://ehlers-danlos.com

The Ehlers Danlos Society: An Educator's Guide
https://ehlers-danlos.com/wp-content/uploads/Educator-Parent-Guide-2016.pdf

The Marfan Foundation
https://www.marfan.org/