EHLERS-DANLOS SYNDROMES

Background

Ehlers-Danlos syndromes (EDS) are a group of inherited connective tissue disorders. Connective tissue is a complex mixture of proteins and other substances that provide strength and elasticity to the underlying structures in the body. An essential building block called collagen is responsible for providing a stable structure for the body's muscles, ligaments and organs. Collagen also provides flexibility in the body; the "glue "responsible for providing a stable structure. In children with EDS, there is either not enough collagen or weak collagen leading to an unstable structural system. As a connective tissue disorder, EDS can cause hypermobility or overly flexible joints, loose "stretchy" skin and fragile tissue.

Joints can easily exceed the normal range of motion causing dislocation. Complications such as low muscle tone, delayed gross and fine motor development, and proprioceptive dysfunction may cause the individual to appear uncoordinated or clumsy. Skin may stretch more than normal and feel velvety to touch. Fragile blood vessels may cause excessive bruising and predisposition to bleeding episodes including nosebleeds and heavy menstrual cycles.

There are 13 types of EDS, each with its own genetic cause and unique sub features. The most common types include hypermobile EDS, classical EDS, and vascular EDS.

- Hypermobile EDS causes large and small joints to be especially loose (e.g., elbows, knees, fingers, toes). Other joints like the shoulder, knee, and jaw dislocate frequently resulting in chronic pain.
- Classical EDS is characterized by very stretchy skin and scarring of pressure points. Tissue damage ranging from hiatal hernia to anal prolapse can also occur. Sprains, strains, and dislocations are more common in classical EDS.
- Vascular EDS is the most serious and life-threatening due to potential organ or artery rupture. Skin tends to be translucent with visible veins and unique facial features that include large eyes, thin nose, lobe less ears and thin scalp hair. Increased joint mobility is most seen in the fingers in this subtype.

Pain caused by the characteristic joint instability of EDS can become more chronic and widespread. Fatigue, headaches, digestive problems, abnormal function of the autonomic nervous system (e.g., dizziness, changes in blood pressure) and dental problems may also occur. Life-threatening complications related to rupture of blood vessels, internal bleeding, and stroke may occur specific to the type of EDS.

Top Takeaways for School Considerations

Ehlers-Danlos syndromes (EDS) are a group of connective tissue disorders with characteristic features including joint hypermobility, loose skin and tissue fragility.

Side effects such as pain, fatigue, headaches, and dizziness have also been associated with cognitive effects like brain fog and inability to concentrate.

Students with EDS may be known to fidget to manage persistent joint pain and discomfort. Simple positioning adjustments and/or use of specialized equipment to support posture is recommended.

Joint elasticity and pain can cause issues with handwriting. The student may benefit from writing utensil aids or assistive communication devices.

Daily physical school activities (e.g., carrying heavy books, note taking, distance between classes) may cause damage to joints or fragile tissues. Rest breaks and activity precautions/restrictions for some physical activities (e.g., contact sports) may be recommended.

Kennedy Krieger Institute's Specialized Health Needs Interagency Collaboration

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

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Considerations for the Individualized Healthcare Plan (IHP)

- Nursing diagnosis of impaired physical mobility, risk for injury, fatigue and pain (acute/chronic)
- Current diagnosed health condition including date of diagnosis, progress of disease process and other chronic health conditions
- Current medication and treatment orders (consider schedule, equipment needs and side effects)
- Use of specialized equipment, adaptive equipment, and orthotics
- Activity, positioning, transferring (consider precautions and/or restrictions)
- Skin check, pressure relief techniques
- Consider emergency care plan(s) (ECP) and emergency evacuation plan(s) (EEP) as related to medical needs in the school setting, and staff education/training, as appropriate

Discussion Starters for Educational Team

1. Would the student benefit from evaluations or assessments in any of the following areas: physical therapy, occupational therapy, speech and language therapy, assistive technology, adapted physical education, functional behavior, psychology, hearing and vision?
2. Would the student benefit from additional academic support and/or modified education (e.g., copies of notes, extra time, reduced workload, simplified instructions, alternative formats for presentation of material, 504/IEP)?
3. Would schedule flexibility support the student?
4. Can rest breaks, safe spaces or reduced stimulation times be built into the student's schedule?
5. Does the student need support with gross or fine motor skills in the classroom?

   Does the student require activity precautions to prevent injury?
6. Is the physical school environment safely accessible for the student's mobility needs (e.g., entry and exit, ramps, location of classes, access to elevator, doorways)?

Resources

Kennedy Krieger Institute: Pediatric Pain Rehabilitation Program
kennedykrieger.org

The Ehlers Danlos Society
ehlers-danlos.com/

The Marfan Foundation
marfan.org/

Scan QR code or visit
KennedyKrieger.org/HealthInformation for more information.

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