Genetic Education Materials for School Success

Ehlers-Danlos Syndrome (EDS)

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Disclaimer:

This is not intended to be medical or clinical advice. Any recommendations here should be reviewed for the appropriateness for your individual child and their unique situation. Also, please note that each document should include the date that it was last reviewed or updated. Research and guidelines that have been released after that date may represent more up-to-date recommendations.

Ehlers-Danlos Syndrome (EDS)

At a Glance

Ehlers-Danlos Syndrome (EDS) is a group of *inherited* connective tissue disorders. (Note - these are different from *inflammatory* connective tissue disorders.) Connective tissue supports, protects, and gives structure to other tissues and organs in the body. When there is a genetic disorder of the connective tissue, there is increased flexibility and fragility of the body. This will be most obvious in the skin and joints but can affect mosts organs and tissues.

Meet Bayla on page 11

There are several types of EDS that each have their own charateristics. The most common type is *hypermobile EDS,* followed by *classical EDS*, and the rarer, most serious form is *vascular EDS*.

Common Features of EDS:

- Loose, unstable joints that can extend farther than normal (*hypermobility*)
- Frequently injured joints – sprain or dislocate easily (child may be able to relocate joint themselves)
- Extreme fatigue
- Soft, stretchy, fragile skin that can bruise or split easily
- Wounds that are slow to heal and leave wide or unusual appearing scars
- Digestive problems – heartburn, nausea, irritable bowel syndrome, constipation
- Dizziness and rapid heart rate when standing (may become nauseated, faint, or collapse)
- Difficulty handling temperature extremes
- Hernias and problems with the heart valves
- Some forms of EDS involve problems with the eyes, bone formation, or organ rupture
- Problems with bladder control possible (stress incontinence)

Because there are overlapping symptoms among the various forms of EDS, it is important to be aware of the type affecting the child in your care. Within each type, the severity of the symptoms can vary among individuals even in the same family.

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In Depth

Medical and Dietary Considerations

What you need to know

The list of *possible* medical problems in each type of EDS can be extensive. However, each individual usually has only some of these problems. Also, the severity of any one of these medical problems varies widely among individuals. Therefore, it is important to ask the parents about the medical issues in their child. School age children with EDS may have annual doctor and specialist visits to monitor medical conditions. No special diet is required for EDS, although a well-balanced diet is important.

Hypermobile EDS – the most common type:

- Joint hypermobility in multiple joints
- Other joint problems
  - Recurrent dislocations/subluxations (altered position or partial dislocation)
  - Joint pain with or without injury
- Skin findings
  - Soft velvety skin that is injured easily
  - Stretch marks not related to weight gain
  - May have some easy bruising
- Other findings
  - Chronic pain – other than joints
  - Functional bowel disorders
    - Irritable bowel syndrome
    - Slowed motility can cause nausea, vomiting, and constipation
  - Postural orthostatic tachycardia (POTS) - very rapid heart rate when standing. Can be accompanied by dizziness, fainting, collapsing, blurred vision, or nausea/vomiting
  - Problems regulating temperature
  - Migraines and sleep disorders common
  - Mild heart valve regurgitation

Classical EDS

- Joint issues as above
- Skin findings similar to above except
  - Incredibly stretchy, like rubber
  - Small harmless bumps under skin
- Other findings
  - Similar to above though tend to have less systemic findings

Vascular EDS – much less common but more potentially fatal

- Skin findings
    - Thin, translucent – can see veins easily
    - Easy bruising
    - Early onset varicose veins
    - Aged appearance, especially in hands
- Joint problems
    - Hypermobility of small joints
    - Chronic subluxations/dislocations
    - Congenital hip dysplasia
- Vascular
    - Arterial rupture
    - Intestinal rupture
    - Uterine rupture during pregnancy
    - Abnormal connections between arteries and veins

What you can do

- Encourage a yearly check-up and studies as needed in the child's Medical Home. Obtain copies of any updated care plans as appropriate.
- Encourage regular eye exams
- Discuss pain management plan with parents
    - Use anti-inflammatories as appropriate
    - Avoid aspirin as it can increase bleeding/bruising
    - Allow storage of ice pack/gel packs at school
- Have a plan to quickly address frequent stomach aches due to reflux, delayed emptying, irritable bowel, etc.
- Take complaints of pain seriously – these children may be injured with minimal trauma
- Address *all* injuries quickly - children with dysautonomia may not report pain.
- Encourage good hydration
- Allow rest break for children who become fatigued.
- Be aware of any changes in behavior or mood that seem out of line with the situation and notify the parents.
- Report any suspected injuries to parent.
- Be aware of any academic changes. Contact parents when any differences are noticed.

Education Supports

It is important to have high learning expectations for children who have Ehlers-Danlos syndrome, utilizing their strengths and interests as a foundation. Encourage use of the core educational curriculum and modify it to meet the child's individual needs.

What you need to know

There is no evidence that EDS causes learning complications. However, premature birth is a complication associated with EDS and this may cause learning differences. Individuals may have speech, visual, and hearing problems. There also may be motor difficulties due to joint

hypermobility – difficulties holding a pencil is a common issues. Children may become uncomfortable sitting in one position too long. Watch for signs of pain or fatigue that may interefere with the child's ability to learn.

What you can do

- Consider formal IQ and performance testing
- If there are verbal performance discrepancies, make sure it is not because of motor difficulties
    - Motor difficulties due to hand hypermobility can affect written test scores.
    - May require alternative ways to assess performance or allow extra time
- Consider OT consult to assess grip and make recommendations to help handwriting
- Pair student with a note taker
- Use pencil grips
- Consider finger ring splints in individuals with Hypermobile EDS
- Use Computers
- Provide two sets of textbooks: one for home and one for school
- Allow the use of an elevator and/or extra time to go to and from classrooms
- Provide a locker at eye level with a digital lock
- Offer priority seating
- Provide a cushion for sitting on the floor and for a chair seat. Allow child to reposition during class as needed
- Music can be helpful - Select instruments to minimize stress to joints
- If POTS (postural orthostatic tachycardia) is present
    - Provide extra fluids prior to rising from a rest period
    - Allow frequent breaks as sitting for a long time in one position is difficult
- Consider allowing child to keep water bottle at desk – staying hydrated diminshes symptoms of dysautomonia
- Allow rest break for children who become fatigued. In extreme instances, child may have difficulty participating in full day activities

Behavioral and Sensory Support

What do you need to know

Individuals with EDS may have psychological and psychosocial difficulties. Fatigue and pain may make psychological problems worse and psychological distress may increase pain. Depression, anxiety, affective disorder, low self-confidence, negative thinking, hopelessness and desperation may occur. Individuals may feel misunderstood, disbelieved, or alone. Children may be self-conscious of physical differences

Pain can be a major part of EDS. The fear of pain and joint instability may lead to avoidance behaviors, and make dysfunction and disability worse. This can further isolate the child.

What you can do

- Suggest counseling and support for pain
- Encourage meditation and gentle physical exercise routines
- Help them with adaptation and acceptance of issues and potential limitations
- Believe the child when they verbalize the symptoms and experiences
- Have a range of inclusive activities in the classroom and during recreational time

Physical Activity, Trips, Events

What you need to know

EDS is different for each person. An individual with EDS may require no accommodations or many. Individuals may require a 504 plan. An alternative health-related credit should be provided if PE accommodations are not possible. Children with dysautonomia may have difficulty in extreme temperatures and may require air conditioning on very hot days and may need to avoid very cold temperatures. These children can participate in field trips and special events with appropriate precautions.

With Hypermobile EDS:

- Position sense may be inaccurate in some individuals with EDS
- Learning to protect the joints will help prevent further injury and keep individuals active
- Individuals should participate in activities that do not cause joint pain and look for less painful ways to move and perform certain tasks
- Frequently overextending or locking joints can lead to traumatic arthritis
- High-impact activity increases risk for
    - Subluxation/dislocation
    - Chronic pain
    - Osteoarthritis
- Joint stability can be improved through exercise programs to strengthen muscle
    - Low resistance exercise may increase muscle tone
    - Important to increase both core and extremity strength
- Non weight bearing exercise promotes strength and coordination

What you can do

Consider Non-Weight Bearing Exercises

- Swimming/water exercises
- Walking
- Biking
- Low impact aerobics
- Core toning exercises

Physical Therapy Can Be Helpful

- Low resistance muscle toning exercise may improve joint stability
- Exercises that promote improved proprioception (position sense) or balance are important

Consider Assistive Devices

- Braces to improve joint stability
- Wheelchairs/scooter may help relieve stress
- May wear soccer pads or ski stocking to protect skin from bruising during activities

Involve Nurse if Pain Medication is Used to Facilitate Exercise

- Mild to moderate - as needed medication may be sufficient
- More significant pain - may require higher doses or multiple medications
- Prevention and control of pain is important!

Arrange Occupational Therapy

- Teach joint protection strategies
- Adjust chair/desk
- Ring splints to stabilize finger joints
- Wrist or wrist/thumb braces for small joint instability
- Neck collar - may help with neck pain and headaches
- Wheelchair/scooter
- May require accommodations in school
    - Use of elevator
    - Extra time in halls

Avoid the following:

- Joint hyperextension/locking joints
- Resistance exercises or isometric exercises, which can be problematic with too much resistance
- Certain activities
    - Weightlifting
    - Running or other high impact activity
    - Contact/ fighting sports ,e.g., football
- Extreme temperatures. Keep hydrated and in shade on sunny days.

Note: With Classical EDS, have higher vigilance for injury because of the skin fragility. Vascular EDS will require even more vigilance because of risk of organ rupture.

School Absences and Fatigue

What you need to know

- Chronic absences may be an issue because of frequent injury and multiple medical

appointments.
- Pain, joint instability, and dysautonomia may cause fatigue

What you can do

- Provide assignments and notes to facilitate student keeping up with class.
- Consider assistive devices
- Allow frequent breaks to manage fatigue
- Contact parents if changes in fatigue are noticed

Emergency Planning

Develop an emergency plan if necessary, depending on the needs of individual children. Specific emergency plans may be required for injuries especially with vascular form of EDS.

Resources

Medline Plus

MedlinePlus is an online health information resource for patients and their families and friends. It is a service of the National Library of Medicine. This link will provide a good medical overview of the condition without being too technical.
https://medlineplus.gov/genetics/condition/ehlers-danlos-syndrome/

Ehlers-Danlos Society

International organization with a lot of support resources for individuals impacted by EDS. Their website also contains information about the various types of EDS and links to educate healthcare professionals about the condition. https://www.ehlers-danlos.com/

Ehlers-Danlos Syndrome Research Foundation

This is a fairly new nonprofit that funds and disseminates research on EDS and related conditions. https://www.edsrf.org/

EDS Awareness

This site is run "by EDSers for EDSers." You will find lots of resources here. Type "school" in the search bar, and it will provide several school related resources for helping students with EDS.
https://www.chronicpainpartners.com/

Info for School Nurse and Primary Care Staff

Ehlers-Danlos Syndrome (EDS) is a complex condition that will be best managed by good collaboration between the school nursing staff and primary care staff as part of the student's team. Work with the parents to get signed releases to share information at the school nurse's office and the primary care office.

Gene Reviews is a peer-reviewed point-of-care reference accessible via the National Library of Medicine. Each entry provides a comprehensive summary of the condition and describes some ongoing surveillance and management issues. It is a great resource for medical providers. Here are the links for the two most common forms of EDS and the most fatal form of EDS.

GeneReviews – Hypermobile EDS http://www.ncbi.nlm.nih.gov/books/NBK1279/

GeneReviews – Classical EDS http://www.ncbi.nlm.nih.gov/books/NBK1244/

GeneReviews – Vascular Type https://www.ncbi.nlm.nih.gov/books/NBK1494/

HIPAA Concerns

Sometimes concerns by primary care staff about violating HIPAA regulations can hamper care by limiting access to necessary medical information. Please note that if care is intended to be delivered by school staff, clarifying medical orders or medication instructions represents continuation of medical care and is not a violation of HIPAA. To avoid any issues like this, remind the parents to sign a release of information form at the primary care office. The school nurse should keep a copy of that release in the student's file, if possible, to facilitate sharing of information in the event that there is difficulty obtaining needed information.

Example letter for physician to provide to school

"My patient_____ has been diagnosed with Ehlers-Danlos Syndrome. This is a group of genetic connective tissue disorders that are characterized by unstable joints, fragile skin, chronic pain and fatigue. Patients with EDS may have frequent injuries and multiple medical complications that can impact their participation in school activities. This condition may necessitate a 504 plan."

Meet a Child with Ehlers-Danlos Syndrome :

Being Bayla

The pieces of the puzzle started to come together for Bayla and her family after little Bayla tripped and fell on a family camping trip. This resulted in an extensive injury which probably would not have been serious in most children. A plastic surgeon was needed to repair the injury, and he suggested a follow up with her physician because of the extensiveness of the injury and the great elasticity of her skin. The plastic surgeon remembered learning about Ehlers-Danlos Syndrome in medical school but never had seen anybody with it. He felt her symptoms aligned with the diagnosis of Ehlers-Danlos Syndrome. Further investigation with a genetics team confirmed the suspicion and a blood test showed that she had the 'classic type' of the syndrome (there are 5 other types). Suddenly, all the bruises that appeared from small events, the significant scarring after cuts, and her overly frail skin made sense with this new diagnosis.

Bayla is a vibrant, fun-loving four-year-old who her mother describes as "outgoing, sassy, and full of life!" She will soon be starting kindergarten. Plans are underway to make sure the staff knows Bayla's needs, her symptoms, and her abilities well before the first day of school. Bayla had been in preschool for a short while. However, her mother Kate felt home-school communication needs were so important and her own anxiety was high with this new diagnosis, so Bayla withdrew. Kate notes how important it was to hear *every* physical complaint Bayla made in a timely way so her medical needs could be addressed. This experience makes Kate a little nervous about kindergarten but she wants to work hard to make sure everyone is on the same page.

Bayla loves to play sports and, with this diagnosis, contact sports pose more of a risk for injury. Though Bayla has to choose sports like swimming that have less direct contact, her family encourages her to explore all that interests her. Bayla and her family take the precautions needed to keep her body free from injury without limiting the exploration of new activities. Bayla's family wants to make sure she can experience a full and active life. Bayla's diagnosis also gives her an edge in some areas! She is much more limber and flexible than the average person. Sprains, dislocations, bruising, and skin tears happen often with Bayla. For most parents, this would be associated with a lot of anxiety, but Bayla's parents realize that this is their "new normal" for life with Bayla and do not panic with every injury. They have learned to be comfortable with managing whatever injury may arise and any care she may need.

There is anxiety that goes along with sending Bayla off to play in the care of someone else but Kate makes sure everyone knows about Bayla's condition. She is careful not limit Bayla's ability

to “spread her wings and fly.” Bayla has two siblings who know that she must be treated a little more gingerly. Her parents have helped her older sister learn that Bayla is more sensitive and, consequently, her sister has developed a good sense of what might be dangerous.

Kate is a nurse administrator and has some solid advice and strategies for both parents and teachers.

For parents:

- Trust your instincts as a parent! Kate felt as if something was not right 2 years prior to the diagnosis. She had questioned the significant bruising, thin skin, and prolapsed rectum at her wellness visits but the work ups did not happen until sometime later.
- Let your child find their own boundaries. Know that as they age, they will become more knowledgeable about their bodies and aware of potential challenges and restrictions.
- Make connections with others who have EDS. The best resource Kate has found are those people who are older (even adults) and have lived with this syndrome. They have offered great tips for how to preserve and protect Bayla’s body and keep it free from injury.
- Spread awareness, to doctors, friends, and family. Bayla’s parents have found that there are many people, including physicians, that are not aware of this syndrome.

For teachers:

- Keep the lines of communication open with the child and parents so that everyone is aware of what is happening. Watch for cues and listen to the child’s complaints with care.
- Report even minor complaints to the parents as they are monitoring this carefully.
- Children with EDS have faulty collagen in their body. They bruise more than other children and are prone to greater injury than other children. A simple grab of the arm as a child runs away can result in a severe bruise. Be aware of this and make sure the children playing with a child with EDS know this as well. Classmates need to know that play has to be gentle.
- Working with a child with EDS should be a collaborative approach in the school setting. The teachers on the playground, the gym teacher, the school nurse, and the child’s classmates all need to be aware of the child’s health condition.

Thank you so much to Bayla and family for sharing their story! And thank you to the team at the Institute on Disability at UNH for assisting in collecting these stories.