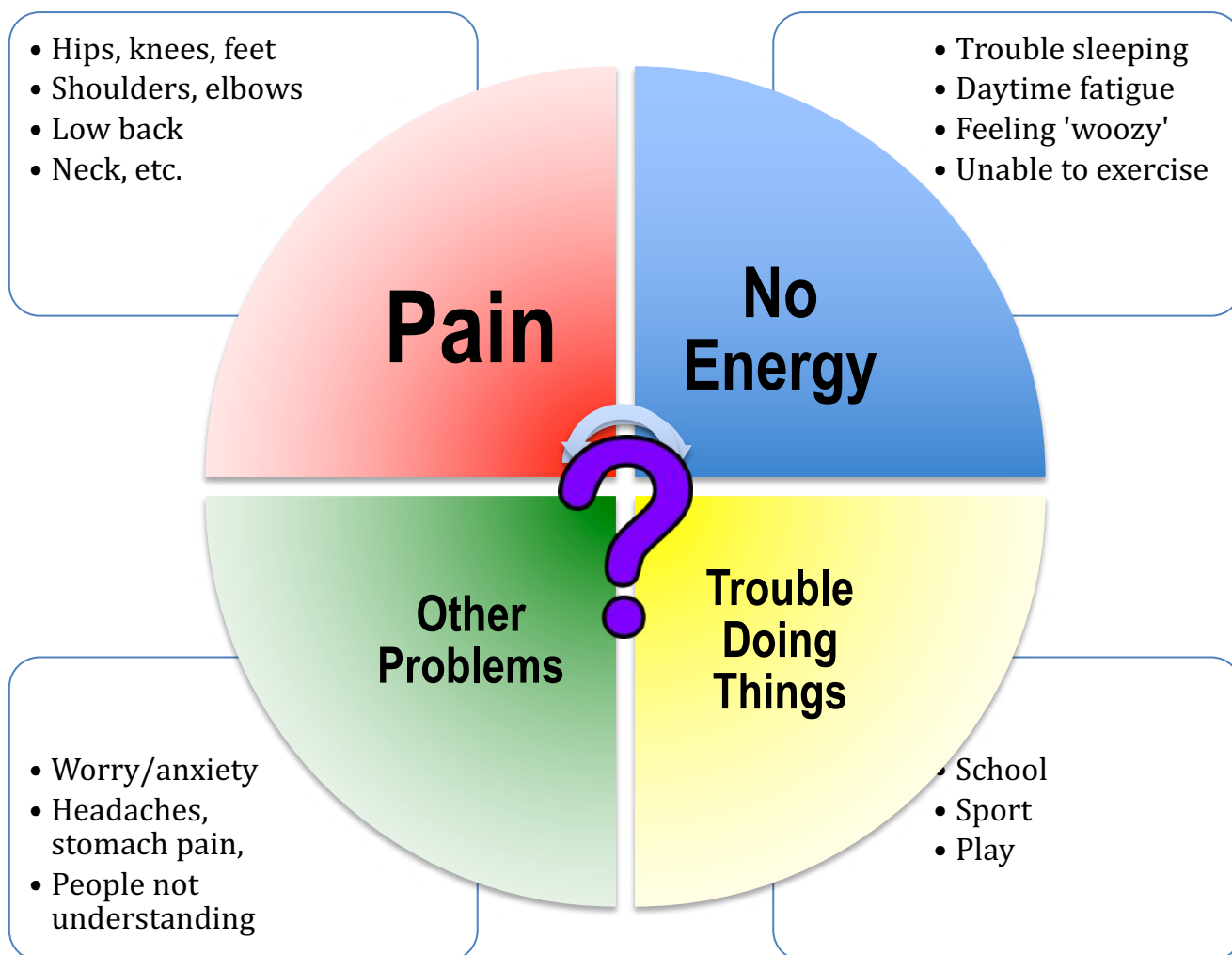


Self-Care for Kids & Teens with Hypermobility Spectrum Disorder

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What bothers you the most?



Hypermobile Ehlers-Danlos Syndrome (hEDS) is also known as *Joint Hypermobility Syndrome*, and *EDS-Hypermobility Type*. **Hypermobility Spectrum Disorder (HSD)** refers to the whole range of conditions that include joint hypermobility. This handout will use the term **HSD**, which is recommended for children. The official diagnostic checklist for children is included on the last page of this handout.

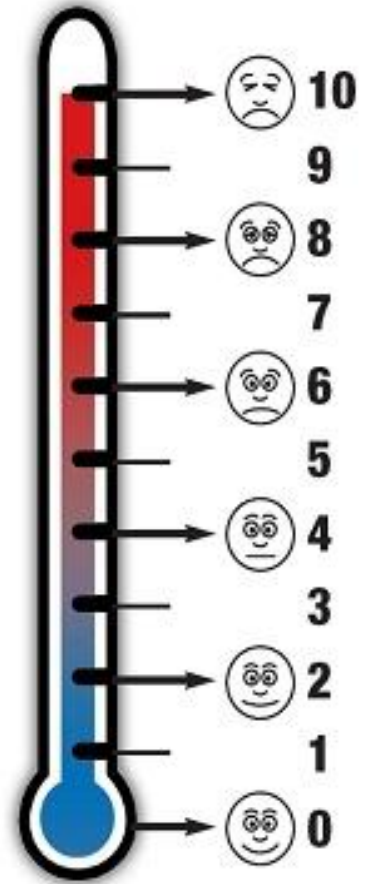
- England, which has a much more developed support system for people with hEDS, has an excellent organization, **Hypermobility Syndromes Association (HMSA)**, which has a terrific web site for children and adolescents: <https://www.hypermobility.org/kidz-zone>
- They also have a School Accommodations website: <https://theschooltoolkit.org/reasonable-adjustments/>
- The **Ehlers-Danlos Society** also has excellent on-line information at: <https://www.ehlers-danlos.com>

This handout is not medical advice. It is a work in progress. If you see errors, please let me know: Lrussek@clarkson.edu

Understanding Your Pain

How much does pain bother you? Use the thermometer scale to the right.

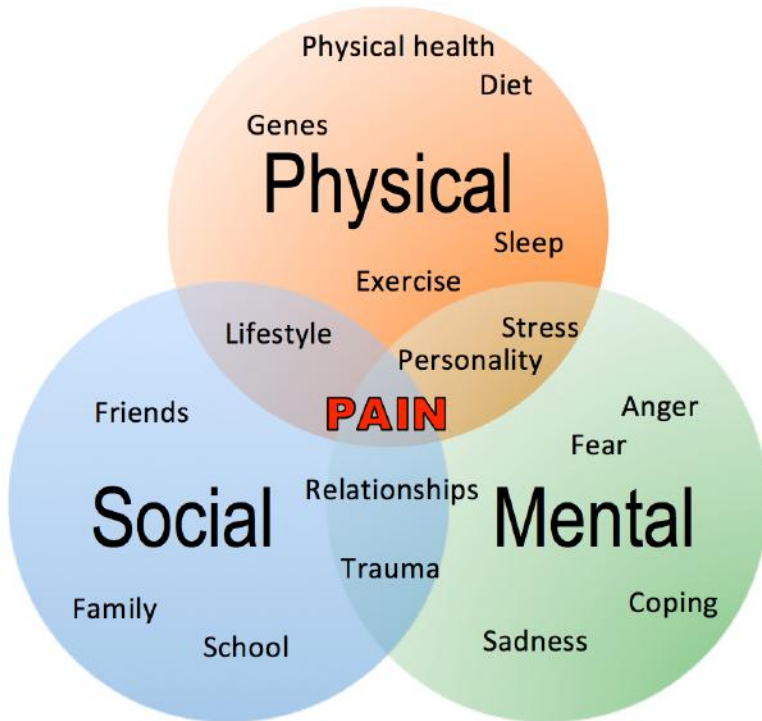
- **Where do you have pain, and what activities make each pain worse?** When you figure out what positions or activities make your pain worse, you can try to figure out why, and **fix the reason for your pain**. Here are a few examples.
 - a. Can you change how you do things that make your pain worse? Sometimes it takes time for the pain to decrease, and you won't notice things are better right away. For example:
 - i. If sitting at your desk at school makes your neck hurt or gives you headaches, can you sit with better posture?
 - ii. If walking, running, or ice-skating make your feet hurt, can you put arch supports into your shoes?
 - iii. If writing makes your fingers or hands hurt, it might help to use thicker pens or finger supports.
 - b. Do you have tight muscles that pull your joints out of position? You might need a physical therapist to help you figure this out.
 - i. If walking makes your low back hurt, you might have tight muscles at the hip, and might need to stretch them. It is important that you don't stretch joints.
 - ii. If you have pain around your kneecaps, especially when going up or down stairs, or after sitting for a while, you might need to stretch muscles on the outside of your leg, and strengthen muscles on the inside.
 - c. You might have muscles that are weak and don't provide support to the joints, or that are not coordinated enough to provide good control. Again, you might need a physical therapist to help you figure this out.
 - i. Weak neck or stomach muscles allow too much motion in the spine, which can cause headaches, neck or back pain.
 - ii. Weak muscles around the hip or shoulder can allow those joints to slip and feel unstable.
 - ii. Some exercises, sports, or even Wii-Fit Balance games can help improve strength and coordination.



It can take some detective work to figure out what activities or positions are causing pain. A physical or occupational therapist can help you figure these things out.

Controlling Your Pain

Pain can be caused by a combination of physical, mental, and social events that happen to us. If we can't change what happens to our bodies, sometimes we can change how the brain uses the information it gets by changing how we think, or relationships with people around us.



❖ This means that you can also feel pain because of thoughts (such as feeling sad or afraid), or social situations (such as loneliness or feeling left out).

❖ Things like stress or trouble at school can make pain from a physical injury hurt more.

❖ But this also means that you can sometimes use your thoughts to change how your brain interprets pain.

Try using one of the following methods to decrease your pain:

- ❖ Relaxation: Deep breathing, biofeedback or other ways to relax.
- ❖ Imagery: imagine you are someplace very pleasant. Use all of your senses to imagine this, including what you hear, smell, see, feel and taste.
- ❖ Distraction: Do something you enjoy to distract your brain from pain.
- ❖ Sensing: Experience different sensations, such as touch, sight, sound, smell or taste can decrease activity in nerves that process pain information. Take a short 'sensation vacation'.
- ❖ Channeling: Changing the "channel" in your brain, like changing a TV channel. When you have negative thoughts about pain, change to a 'channel' with good things – think about things that you like or are looking forward to.

Resources:

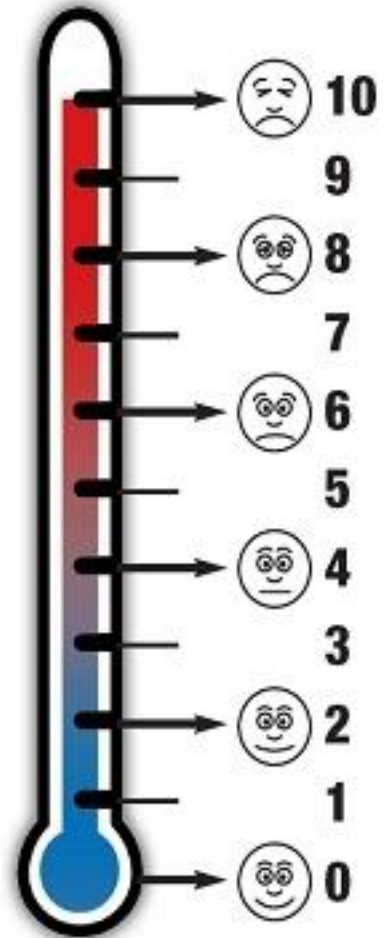
- **WebMAP** (free app): The WebMAP Mobileapp is designed to help adolescents manage chronic pain such as back pain, abdominal pain or headaches. Users imagine 'traveling' to different destinations (e.g., rain forest, desert, etc.) to learn skills for managing pain.
- **iBeatPain** (free app): The Mayo Clinic Center for Innovation created the iBeatPain for Teens app. The app focuses on 5 self-care strategies: 1) Practice relaxation; 2) Exercise; 3) Participate in school; 4) Get regular sleep; 5) Engage in self-care, such as basic hygiene and drinking enough water.
- A list of several kid/teen appropriate apps for pain management and relaxation:
<https://www.kidshealth.org.nz/relaxation-pain-management-apps>
- A pain management education website for kids and teens: <https://aci.health.nsw.gov.au/chronic-pain/painbytes>

Feeling Tired, No Energy

How much does it bother you to be tired or have no energy? Use the thermometer scale to the right, where not being tired at all is 0, and being so tired you can't do anything at all is 10.

There are several reasons for feeling tired when you have HSD:

1. **POTS:** HSD is often combined with a condition called POTS, which stands for Postural Orthostatic Tachycardia Syndrome. In HSD, your blood vessels are stretchy, like your joints. This means the blood vessels don't pump blood back to the brain properly, and the heart tries to pump extra fast to move your blood through your body. In addition to feeling tired, people with POTS often have a racing heartbeat, headaches, dizziness, light-headedness, nausea, trouble thinking clearly, and feeling anxious or scared for no good reason. (Actually, there is a good reason – your brain is screaming for more blood flow.) The test for POTS is not difficult, and involves having you lie still for a few minutes, then measuring your heart rate and blood pressure when you suddenly stand up (or the table you lie on tilts up).
 - a. **If you have POTS:**
 - i. Drink a lot of fluid and eat salty foods; this increases the amount of blood in your body so it easier for your heart to pump it around.
 - ii. Move your ankles up and down or tense and relax your other leg muscles to 'pump' blood back up to your brain.
 - iii. Lie down with your feet above your heart and head.
 - iv. Wear compression socks so blood doesn't pool in your legs and feet. There are some cool ones now.
 - v. Check out: <http://www.chop.edu/conditions-diseases/postural-orthostatic-tachycardia-syndrome-pots>
 - vi. Exercise protocol: http://www.dysautonomiainternational.org/pdf/CHOP_Modified_Dallas_POTS_Exercise_Program.pdf
2. **Not sleeping well.** People with HSD often don't sleep well because of pain or feeling restless.
 - a. Use good sleep habits, such as exercising and getting sunlight during the day, relaxing before bedtime,
 - b. Avoid things that will wake you up at bedtime: food with caffeine or sugar, activities that will wake you up, electronics (TV, phones, tablets) which produce blue light that can wake you up.
 - c. Make sure you are comfortable – your bed should be soft but supportive. You may need several pillows around you to make yourself comfortable.
 - d. More sleep suggestions at: <https://www.seattlechildrens.org/pdf/PE1066.pdf>
3. **Getting out of shape from not enough exercise.** If you don't get regular exercise (for example, if you have pain or your joints are injured), your body becomes less able to exercise, and you feel tired. It may seem backwards, but the solution is to get more exercise, even though you are tired. Since not getting enough exercise can also cause pain, regular exercise can help in a couple of ways, if it is done correctly.
 - a. Start with a little bit of exercise doing something you enjoy – such as walking, biking, dancing, or playing physical games. You might only be able to do a few minutes when you start, and you might need to do this several times during the day to get even 10-20 minutes of exercise. But gradually increase the number of minutes you exercise, even if you feel tired. It takes several weeks for this to work. People often try to do too much exercise too quickly; you can't rush this! "Start low; Go slow!"
 - b. Excellent self-help guide for chronic fatigue: [search the web for "graded exercise therapy bartscsfme"](#)
4. **Feeling exhausted by the pain.** Having pain can tire you out! Manage the pain using the techniques described on the first few pages of this handout.

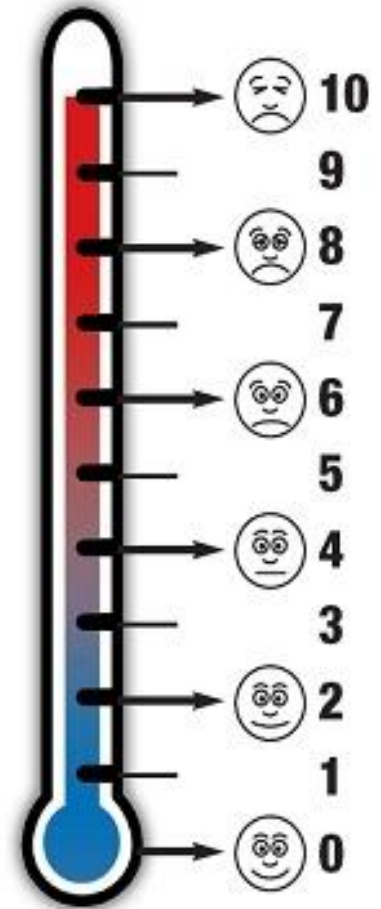


When You Can't Do The Things You Want To Do

How often do you feel like you just can't do the things you want to, because of your HSD? Use the thermometer scale, where having no limitation is 0, and being unable to do anything you want is 10.

Trouble at school because of pain, fatigue, difficulty thinking clearly, taking notes, walking from place to place, carrying books, etc.

- Try to use good posture, whether you are sitting or standing.
- You may need to modify things like desks, chairs, or pencils so that they don't put so much stress on your joints.
- You may need to move around more than most kids.
- Handwriting might be difficult or painful.
- Gym class can be hard for some kids, especially since the pain and fatigue can change a lot from day to day. Do what you can, so that your muscles and heart can stay strong. If you can't do what the class is doing, try to do something else, like walking, swimming, biking, yoga, tai chi or strengthening exercises.
- A physical or occupational therapist can help you figure out how to do things better at school.
- Missing school. Suggestions for school accommodations at: <https://theschooltoolkit.org>
- <https://www.hypermobility.org/kidz-zone/coping-at-school>



It may be hard to play or do the sports you want to do.

- Kids and teens with HSD have a wide range of exercise tolerance – some can barely do anything, while others can be quite active.
- See the section on pain for suggestions about how to figure out why you have pain. Sometimes you can do more with different shoes (or inserts), standing straighter, stretching tight muscles (without stretching loose joints), using good body mechanics.
- Some kids with HSD have problems with coordination or balance. Exercise is still important, but you might have trouble with sports that need a lot of coordination.
- You may need to change how you do things so you don't put as much stress on your joints. For example, you might not be able to do deep squats because of knee pain, or you might not be able to climb because your shoulders are too loose. You might need to be creative to figure out different ways to do things. Or you may need special equipment to help you with your sport or play activity.
- Pain and limitations can vary day to day. People need to understand that.
- Suggestions for exercise: <https://www.hypermobility.org/exercise>

Other Problems

How much do other problems related to HSD bother you? Use the thermometer scale to the right, where having no other problems is 0, and having so many problems that you can't enjoy life is 10.

1. Feeling clumsy or uncoordinated, having bad balance, or joints that 'slip out'

- The right exercise can improve your coordination and balance. The opposite is also true: if you stop exercising (because of pain or fatigue), your coordination and balance will get worse.
- People with HSD sometimes feel like their joints will 'slip out.' Strong muscles will make joints more stable. A simple exercise for shoulders and hips that slip: try 'sucking them into the joint' and holding 5 seconds.

2. Feeling scared or worried, sometimes for no good reason

- Find out if you have POTS, because that is a physical reason for feeling anxious, and there are physical ways to treat POTS.
- Not knowing how you will feel on any day can be scary. Try to keep a sense of humor and go with the flow! Talk to people about your concerns.
- Relaxation training, like deep breathing, meditation or yoga can help. See section on relaxation earlier in the handout.

3. Other people might not understand what it is like to have HSD

- Sometimes people don't believe you really have a real medical condition, because it is hard to see. You can give people information about HSD to read. The School Toolkit talks about problems you might have in school: <https://theschooltoolkit.org>
- Talking to other people who have HSD can help. If there is no support group in your area, there are several on-line. Check <https://www.ehlers-danlos.com/support-groups/> for support groups in your area and on-line groups.

4. Headaches:

- Headaches are common in people with HSD because the loose joints allow too much motion in the neck joints, which may cause the muscles in your neck to work too hard. Sometimes the neck muscles are not coordinated or strong enough. Poor posture can also stress the neck and cause headaches. Not sleeping well can also make headaches worse.
- One book that might help: V. DeLaune, *Trigger Point Therapy for Headaches and Migraines: Your Self-Treatment Workbook for Pain Relief*.

5. Stomachaches:

- Many people with HSD are sensitive to certain foods. This can be tricky to figure out. Sometimes a special diet can help. Consider:
 - FODMAP diet: <http://www.med.monash.edu/cecs/gastro/fodmap/>
 - Heidi Collins EDS diet: <https://docslide.net/documents/diet-supplement-guidelines-for-eds-heidi-a-collins.html>
- Some people with HSD have a condition called 'mast cell activation disorder', or MCAD. <http://www.mastattack.org/2015/05/mast-cell-disease-fact-sheet/>
- If you are constipated (if you don't have a BM most days), your gut might be moving food too slowly. Regular exercise can help. There is also a type of massage to your belly that can help. Abdominal propulsive massage is shown in the picture, here. (Harrington & Haskvitz, *Phys Ther.* 2006;86:1511-19)

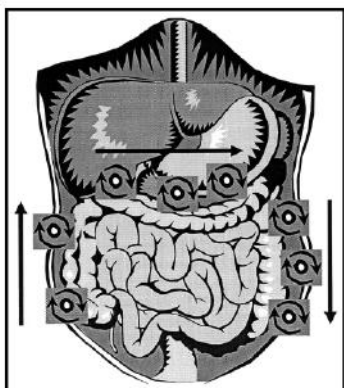
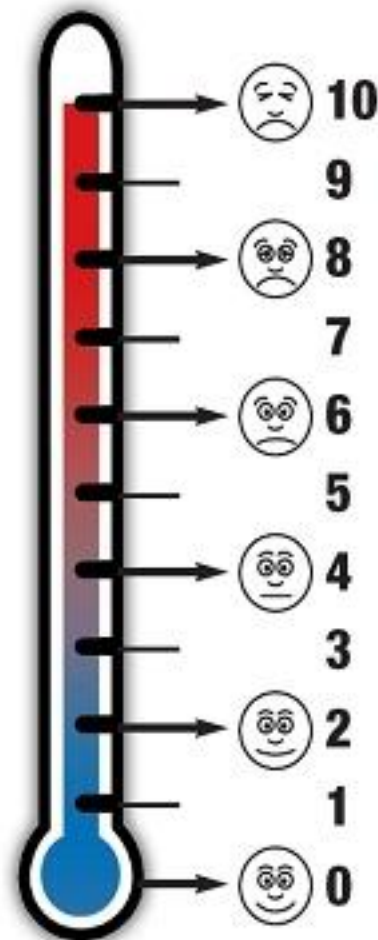
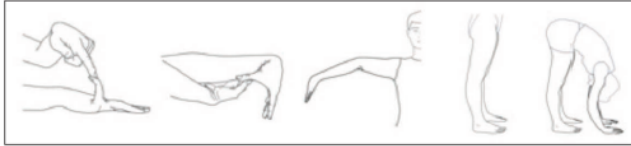


Figure.
Path of propulsive abdominal bowel massage taught to patient.



Patient name: _____ DOB: _____ DOV: _____ Evaluator: _____

Children From 5 Years Of Age Until Biological Maturity



Beighton Score: ____/9
 Must be a minimum of 6

L R L R L R L R

Skin and Tissue Abnormalities

- Unusually Soft Skin – unusually soft and/or velvety skin
- Mild Skin extensibility
- Unexplained striae distensae or rubae at the back, groin, thighs, breasts and/or abdomen without a history of significant gain or loss of body fat or weight
- Atrophic scarring involving at least 1 site and without the formation of truly papyraceous and/or haemosideric scars as seen in classical EDS
- Bilateral piezogenic papules in the heel
- Recurrent hernia, or hernia in more than one site (excludes congenital umbilical hernia)

Score: ____/6
 Must be a minimum of 3

Musculoskeletal Complications

- Episodic Activity related pain not meeting the chronic pain frequency and duration criteria
- Recurrent joint dislocations, or recurrent subluxations in the absence of trauma, and/or frank joint subluxation on physical exam in more than one joint (excludes radial head <2yrs)
- Soft tissue injuries – One major (needing surgical repair) and/or current multiple minor tendon, and/or ligament tears

Score: ____/3
 Must be a minimum of 2

Co-Morbidities

- Chronic primary pain
- Chronic fatigue
- Functional GI disorders
- Functional bladder disorders
- Primary dysautonomia
- Anxiety

Any number causing
 distressor disability?
 Y / N

Prerequisites:

1. This framework can only be used after exclusion of other Ehlers Danlos subtypes, heritable disorders of connective tissue, syndromic conditions, chromosomal microdeletions, skeletal dysplasia's, or neuromuscular disorders. From biological maturity or the 18th birthday, whichever is earlier, the 2017 Adult criteria should be used.

2. No genetic cause for hEDS has been identified at the time of publication of the checklist. In the future disease-causing genetic mutations may be identified in hEDS. In that scenario, if a child has a biological parent with an hEDS diagnosis and a confirmed disease-causing genetic mutation and the child also has the same mutation with GJH then the hEDS diagnosis should be used.

Fig. 1 A diagnostic checklist for pediatric joint hypermobility and hypermobility spectrum disorder

Tofts et al., 2023. Available at: <https://www.ehlers-danlos.com/diagnostic-criteria/new-diagnostic-framework-for-pediatric-joint-hypermobility-v2/> with full article at <https://ojrd.biomedcentral.com/articles/10.1186/s13023-023-02717-2>