

# Hypermobility 105: Ehlers-Danlos Syndrome in Children



Leslie N Russek, PT, DPT, PhD, OCS  
Clarkson University, Canton-Potsdam Hospital, Potsdam, NY

Slide handouts are now available at: <https://webpace.clarkson.edu/~lrussek/hsd.html>



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## Objectives

1. Describe common problems children experience
  2. Outline a process for PT (or OT) to evaluate children
  3. Describe treatment approaches PT may use with children
  4. Explain how PT or OT should empower children and their families to manage EDS/HSD
- Podiatrists and psychologists may also be important members of the health-care team

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## DISCLAIMER

- I cannot provide individual medical advice in this presentation
- The information provided here is generally applicable to HSD/hEDS, but your child's situation may be different.
- You should discuss options with your healthcare provider before starting a new management approach.

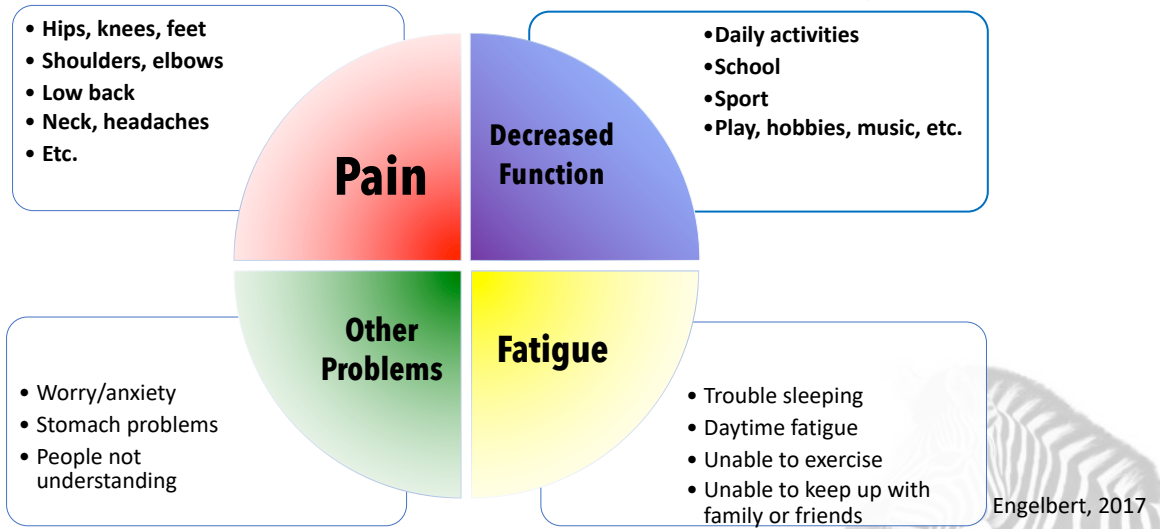


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## Common Problems for Children/Teens



Engelbert, 2017

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# Managing Pain & Instability

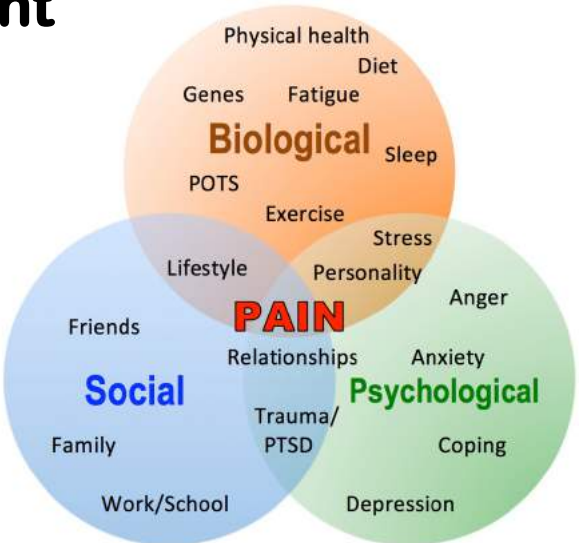
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## Pain Assessment

- Pain is biopsychosocial: it involves biological, psychological, and social factors
- Look for things that increase pain:
  - Physical activities or postures
  - Deconditioning from decreased activity or exercise
  - Anxiety, depression, stress..



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## Emotional Factors Affecting Pain

- Stress
- Anxiety
- Depression
- Poor self-image
- Feeling out of control
- Fatigue
- Not being believed
- Trauma (physical, emotional, sexual)..



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## Physical Factors That Increase Pain

1. Bad postures and joint alignment
2. Unstable joints, chronic or subluxed/dislocated
3. Tight muscles pulling on loose joints, especially after growth spurts
4. Poor muscle control and coordination
5. Puberty also aggravates POTS, which may amplify problems

Engelbert, 2017

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# Sitting Positions

- Slouched
- Cross-legged
- W-sitting



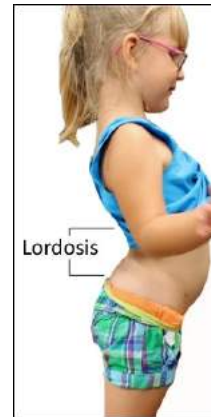
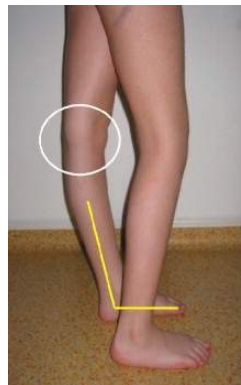
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# Standing Posture

- Flat feet
- Hyperextended knees
- Over-extended spine



Cottalorda, 2012;  
<https://doi.org/10.1016/j.otsr.2012.04.015>


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## Posture Scenarios

**Match posture to likely pain:**


- A. Headaches
- B. Neck pain
- C. Shoulder pain
- D. Low back pain
- E. Hip pain
- F. Knee pain
- G. Ankle/foot pain




A,B,  
E,F




A,B,  
C,D



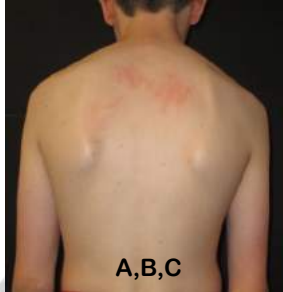
C,D,F



B,D



E,F,G




A,B,C

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## Managing Instability & Posture

- Choosing appropriate postures, activities and exercises
- Provide proper support to the body
  - Good shoes, chairs, desks, pencils, etc.
- Education to use good posture and body mechanics
- Adaptive tools, assistive devices, orthotics
- Motor control and coordination exercises..
- Strengthening exercises



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**BEWARE OF IPOSTURE**

**BAD**

**GOOD**

<http://thepowerofposture.net/children/>  
 Also: <https://www.missjaimeot.com/positioning-in-the-classroom/#more-3024>

**PROPER POSTURE**

Stabilize the paper with your non-dominant hand.

On the desk, the paper should be angled. Angle it so that the right side of the paper is slightly higher for right handed students.

Desk should be 1-2" higher than bent elbows.

Hips bent at a 90 degree angle.

Knees bent at a 90 degree angle.

Feet flat on the floor.

Go to [www.YourTherapySource.com/thwstation](http://www.YourTherapySource.com/thwstation) for the complete download.

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Questions?

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## Select Appropriate Activities

But each person is different!

### Easier for EDS/HSD

- Low impact: walking, biking, yoga, weights, horseback riding, swimming, aquatic exercise, low impact dance
- Non-contact: Tai Chi, Pilates, dance, karate forms

### More Challenging for EDS/HSD

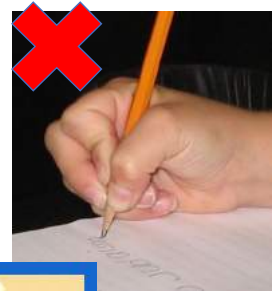
- High impact: running, basketball, tennis, ballet
- Contact: football, soccer, karate
- Weight-bearing on hands: gymnastics, some yoga or Pilates
- Forceful gripping: weight-lifting, some musical instruments



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## Pens, Pencils, and Gripping

- Forceful gripping of pencils, etc. can cause finger, wrist, arm pain, fatigue, and difficulty with handwriting.
- Grippers can help
- Good info about grippers:
  - <https://theanonymousof.com/2018/09/11/pencil-grip-101-updated/>
  - <https://www.ot-mom-learning-activities.com/pencil-grips-for-kids.html>



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### Device-Complaint Matching

Which devices are likely to help each of the following joints?

- Neck
- Shoulder
- Low back
- Hip
- Knee
- Feet

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# Exercises

- Strengthen muscles around joints
- Improve joint alignment
- Stretch tight muscles (while stabilizing joints)
- Improve control and coordination
- Both generalized fitness and patient-specific exercises help
- Exercises must be appropriate, not be too difficult, kids must be able to do them correctly, must have time & support
  - Kemp, 2010; Birt, 2014



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# Exercise Options



THEINSPIREDTREEHOUSE.COM



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## Make Exercises Fun!

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## ”But Exercise Makes Me Worse”

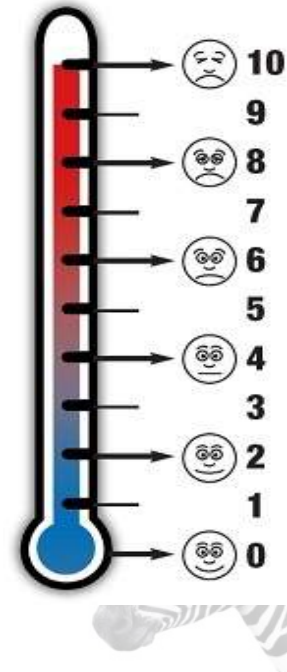
- Reasons why exercise/PT might have failed in the past:
  - Exercises too difficult or progressed too quickly:
    - Using what works for other kids, not accounting for deconditioning or fragile tissues
  - Not for long enough:
    - 4-6 weeks might not be enough, people with HSD/EDS progress slowly
  - Too much too often:
    - Too often can cause flares. Maybe 1-2x/week better than 3-5x/wk
  - Gym teachers who don't believe the child really has pain
  - Failing to treat the whole person, other HSD/EDS issues
    - (Simmonds 2018; Scheper, 2013; Engelbert, 2006)

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## Managing Pain in HSD/EDS

- Use a scale to rate pain severity
- Identify what increases pain
  - Physical factors
  - Psychological and social issues, stress, anxiety...
- Try to avoid things that cause or increase pain
- Self-care:
  - Physical strategies: ice packs, hot packs, topical rubs, braces, exercise, TENS machines, massage, etc.
  - Psychological strategies: relaxation, meditation, visualization, yoga breathing, biofeedback, etc.



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## Resources for Managing Pain in Kids

- FREE pain management phone apps for kids – kid-friendly format and language
  - **WebMAP Mobile**
  - **iBeatPain**
- For teens: [www.growingpains.org](http://www.growingpains.org) is a website created for teens living with pain to share with other teens experiencing pain.
- University of Michigan Medicine: Pain and Your Child or Teen:
  - <http://www.med.umich.edu/yourchild/topics/pain.htm>
  - Info about pain in children, links to resources such as biofeedback, breathing techniques, art therapy, CBT, distraction, acupuncture, massage etc.
- Relaxation apps for kids:
  - <http://kidsrelaxation.com/uncategorized/7-apps-to-help-kids-relax/>
  - <http://parentingchaos.com/anxiety-apps-kids/> .
- Palermo & Law. *Managing Your Child's Chronic Pain*. 2015. How to use cognitive behavioral skills with kids.

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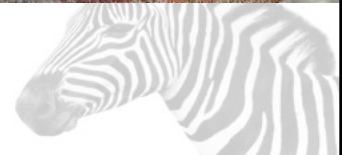
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## Fatigue and Sleep Problems

- Fatigue is the second leading cause of problems for children and teens with HSD/EDS (Mu, 2019)
- Factors that increase fatigue:
  - POTS
  - Pain/discomfort from HSD/EDS
  - Poor sleep habits or pain interfering with sleep
  - Being out of shape from lack of exercise
  - Stress, anxiety, depression
  - Poor nutrition

Kizilbash, 2014



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## Managing Fatigue In Children/Teens

- Exercise, “Start low, Go slow” - very gradually progressed
  - POTS-specific exercise, if appropriate (Kizilbash, 2014)
  - [http://www.dysautonomiainternational.org/pdf/CHOP\\_Modified\\_Dallas\\_POTS\\_Exercise\\_Program.pdf](http://www.dysautonomiainternational.org/pdf/CHOP_Modified_Dallas_POTS_Exercise_Program.pdf)
- Get enough sleep: use good sleep habits (“sleep hygiene”)
- POTS care ([https://www.ehlers-danlos.org/wp-content/uploads/2017/07/MAYO\\_Clinic\\_Teens\\_Dysautonomia\\_copy.pdf](https://www.ehlers-danlos.org/wp-content/uploads/2017/07/MAYO_Clinic_Teens_Dysautonomia_copy.pdf))
- Try to maintain normal activities, including school, friends, hobbies, etc.
- Good nutrition; avoid caffeine and sugar
- Behavioral approaches such as prioritizing, pacing, & planning
  - Stress management, meditation, yoga, managing depression or anxiety..



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## Good Sleep Habits

- Make sleep a high priority
- Make the room a sleep haven: cool, quiet and dark.
- Don't use the bed for anything but sleep (no studying, TV, games, etc.)
- No electronics (TV, phone, computer) for 1 hr before bedtime
- Keep a regular sleep schedule, including on weekends
- Don't eat, drink, or do vigorous exercise within a few hours of bedtime
- Have a bedtime routine: warm bath/shower, read, yoga, etc.
- Suggestions for kids: <https://childmind.org/article/encouraging-good-sleep-habits/>
- Suggestions for teens: <https://www.sleepfoundation.org/articles/teens-and-sleep>




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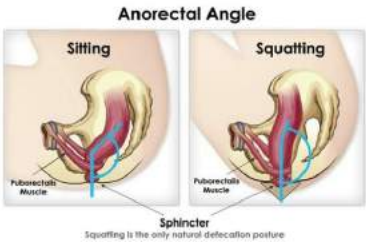
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## Toileting

- Managing constipation
  - Posture on the pot: 35° hip flexion
  - Relax the tummy
  - Proper diet (varies)
  - Regular physical activity
  - Tummy massage
    - [https://pwr4life.org/forms/wellness-series-docs/Diana\\_BLOG\\_ABDOMINAL\\_MASSAGE.pdf](https://pwr4life.org/forms/wellness-series-docs/Diana_BLOG_ABDOMINAL_MASSAGE.pdf)
  - Managing constipation in children: [www.bladderbowelhealth.org.au/children-and-young-people/constipation-in-children](http://www.bladderbowelhealth.org.au/children-and-young-people/constipation-in-children)
- Managing incontinence/wetting
  - Avoid constipation
  - Planned fluid intake and voiding
  - Pelvic floor exercises
  - [www.mottchildren.org/health-library/hw220056](http://www.mottchildren.org/health-library/hw220056)





<https://www.pinterest.ca/pin/191684527862553378/>



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## Questions?





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## Common Problems for Kids in School

- Pain sitting still for long periods of time
  - Uncomfortable, unsupportive chairs
  - Discomfort sitting on the floor
  - Pain leaning forward to read, take notes, or look at the board
- Difficulty with handwriting
- Trouble getting around school, carrying books, etc.
- Trouble participating in physical education
- Fatigue
- Missing school due to injuries, medical care, fatigue, etc..

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# Sitting Suggestions



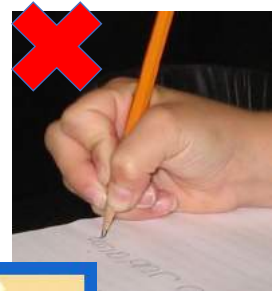
<https://www.missjaimeot.com/positioning-in-the-classroom/#more-3024>



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# Handwriting Problems

- Grippers can help (see above)
- May benefit from typing into a computer rather than writing by hand
- Occupational Therapists can assist with manual skills



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## Adapting Physical Education

- Encourage children/teens to participate in activities they can do safely
- Adapt regular gym activities, when possible
- Find alternative activities, preferably for the whole class (e.g., Pilates or yoga rather than kickball and running)
  - Solo alternative activity, if necessary: walking, yoga, Pilates, PT exercise, etc.
- Arrange for your child to 'opt out' when not feeling well
- Good resources:
  - EDS: A Parent's Guide Helping Your Child Succeed at School
  - WrightsLaw: <https://www.wrightslaw.com/info/pe.index.htm>
  - Center for Parent Information and Resource: <https://www.parentcenterhub.org>



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## Other Possible School Adaptations

Students may need:

- Help getting around school:
  - Extra time to get from one class to another
  - A bookbag on wheels to carry books
  - Access to elevators
- Rest breaks during the day
- To move around during class, rather than sit still
- A special chair
- Some kids may only be able to do part of the day

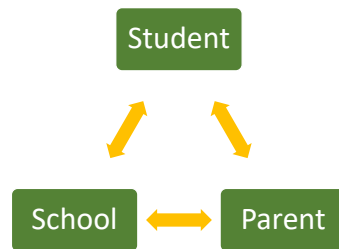


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## School Accommodations

- Difficulties with school activities can be addressed 3 ways:
  1. Informal/unofficial modifications
  2. Accommodations through a 504 plan
  3. Accommodations through an Individualized Educational Program (IEP)



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## Informal Modifications

- If your child does not need a formal 504 plan or IEP
- You, your child and the teacher come up with solutions to address issues so that your child can participate in school activities
- Examples (but may vary by school district):
  - Using a pencil grip or taking notes on a laptop computer
  - Sitting on a cushion or stool rather than cross-legged on the floor
  - Playing the goalie in soccer rather than a running role
  - Doing Pilates or PT exercises instead of the regular gym class
  - Scheduling a study hall to rest during the school day
  - Drinking extra fluids (especially for POTS) & extra bathroom breaks

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## 504 Plan

- A formal arrangement for accommodations that will allow your child to meet educational core objectives.
- Changes the learning environment or resources to allow children to remain in their regular classes
- Examples (but may vary by school district):
  - Being allowed to take standardized exams on a computer rather than writing answers by hand
  - Having a special ergonomic desk and chair
  - Being allowed to replace gym with alternative activities or classes
- 504 plan does not cover extracurricular activities



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## Individualized Educational Plan (IEP)

- When your child requires special services to be able to meet the core educational objectives
  - Extracurricular activities are not included in determining need
- The disability must limit the child's ability to learn and benefit from the standard curriculum
- Decisions about accommodations are made by the IEP team
- Children may receive PT, OT, speech therapy, etc. as part of the IEP
  - Only if learning objectives cannot be met with accommodations



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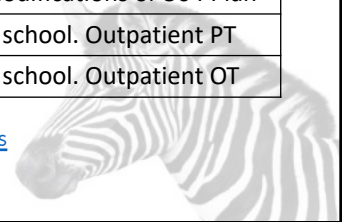
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## 504 Plan vs. IEP

Issue	Probable Solution
Poor handwriting, but able to use a laptop	504 Plan or informal modification
Unable to write or use a laptop, even with grippers or finger splints	IEP with OT
Needs extra time and elevator pass to get around school	504 Plan
Unable to play on the basketball team	Informal mods, nothing or outpt PT
Needs to take standardized exams on computer with extra time	504 Plan
Has back pain after sitting through a full day of classes	Informal modifications or 504 Plan
Has hip and knee pain sitting on floor, but able to sit on stool	Informal modifications or 504 Plan
Wants to take dance classes, but knees hurt too much	Nothing in school. Outpatient PT
Wants to improve poor handwriting, but able to use laptop	Nothing in school. Outpatient OT

[www.understood.org/en/school-learning/special-services/504-plan/the-difference-between-ieps-and-504-plans](http://www.understood.org/en/school-learning/special-services/504-plan/the-difference-between-ieps-and-504-plans)

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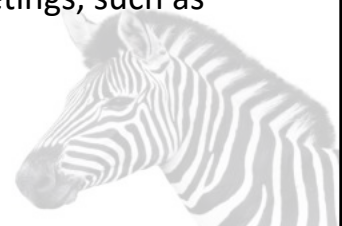
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## How Do You Figure This Out?

- Every school has a Chair of Special Education, who oversees IEPs and 504 plans; this person's job is to help you.
- Every school has a list of local advocates who can advise you and guide you through the process. Assistance from an advocate is free, but you typically must request one.
  - You can ask that your advocate be part of the IEP/504 team.
- You can ask anyone else to be part of the IEP/504 meetings, such as your PT or OT, or a friend to take notes for you.
- There are web sites with advice

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## Potential Accommodations

- Sources of information on potential problems and accommodations:
  - <https://ehlers-danlos.com/wp-content/uploads/Educator-Parent-Guide-2016.pdf>
  - <http://hypermobility.org/help-advice/kids-teens/coping-at-school/>



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## Additional Resources for School

- Help developing a 504 plan for your child in school:
  - <https://www2.ed.gov/about/offices/list/ocr/docs/504-resource-guide-201612.pdf>
  - [www.understood.org/en/school-learning/special-services/504-plan/the-difference-between-ieps-and-504-plans](http://www.understood.org/en/school-learning/special-services/504-plan/the-difference-between-ieps-and-504-plans)
- IEP: [www2.ed.gov/parents/landing.jhtml](http://www2.ed.gov/parents/landing.jhtml)
  - [www.specialeducationguide.com/pre-k-12/individualized-education-programs-iep/the-iep-process-explained/](http://www.specialeducationguide.com/pre-k-12/individualized-education-programs-iep/the-iep-process-explained/)
  - [www.parentcenterhub.org/iep-overview/](http://www.parentcenterhub.org/iep-overview/)
- Center for Parent Information and Resource:
  - <https://www.parentcenterhub.org>



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## Other Resources

- Ehlers-Danlos Society: An Educator's Guide Meeting the Needs of the Ehlers-Danlos Child & A Parent's Guide to Helping Your Child Succeed at School:
  - <https://ehlers-danlos.com/wp-content/uploads/Educator-Parent-Guide-2016.pdf>
- Hypermobility Syndromes Association (UK)
  - <https://www.hypermobility.org/Pages/Category/children-and-young-people/Tag/child>
  - <http://hypermobility.org/help-advice/kids-teens/pe-and-exercise/>
- Self-Care for Kids & Teens with HSD:
  - <https://webspaces.clarkson.edu/~lrussek/docs/hypermobility/> "HSD in children and adolescents"
- Review of relaxation apps for kids:
  - <https://www.simplepractice.com/blog/anxiety-relaxation-mindfulness-apps-kids/>
- WebMAP mobile app for pain management
- iBeatPain app for pain management



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## Summary

- Identify the biggest problems for the child/teen
  - Prioritize what is most important to him or her
- Try to figure out what causes or aggravates those problems
  - This requires thinking, trial and error, and sometimes professional advice
- Find ways to prevent or manage pain, instability, fatigue & stress
  - Involve the child/teen, family, school, and appropriate health care providers
- Work with the school to identify accommodations
- Be patient! Even once you are doing all the right things, improvement takes time
  - It may take many months for things to get better; expect flares along the way.



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## Questions?



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