

OT School-Based Management of Hypermobility in Children

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OCCUPATIONAL THERAPISTS

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PEDIATRIC UPDATE SYMPOSIUM

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Disclosures

We have no conflicts of interest to declare



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Agenda

- What is Joint Hypermobility
- Assessment of Joint Hypermobility
- Ehlers-Danlos Syndromes vs Hypermobility Spectrum Disorders
- Associated Conditions and Symptoms
- Implications
- Evidence
- Existing and New Resources & Handouts
- Questions

Poll – Knowledge Check

How knowledgeable are you about OT management considerations (both broader and targeted) for joint hypermobility?

1. Not that knowledgeable
2. Knowledgeable but not sure how to go about it
3. Familiar
4. Have experience with some children and youth
5. Very familiar with this population and OT management

Poll

*How important is occupational therapy for this population?
(both broader and targeted management and intervention)*

1. No opinion
2. Not that important
3. Slightly important
4. Important
5. Fairly important
6. Very important

Poll

How satisfied are you with your current knowledge and skills in occupational therapy management and intervention for this population? (both broader and targeted management and intervention)

1. No opinion
2. Not that satisfied
3. Slightly satisfied
4. Satisfied
5. Fairly satisfied
6. Very satisfied

Learning Objectives

- Thorough review of the current state of evidence around joint hypermobility (HSD)
- Occupational therapy assessment of joint hypermobility
- The impact of joint hypermobility on participation in day to day activities
- Occupational therapy modifications and adaptations to improve function and participation
- Implications for future research

Joint Hypermobility Misconceptions



"Double jointed"



"Only" part of Ehler's Danlos Syndrome



"Just" very flexible



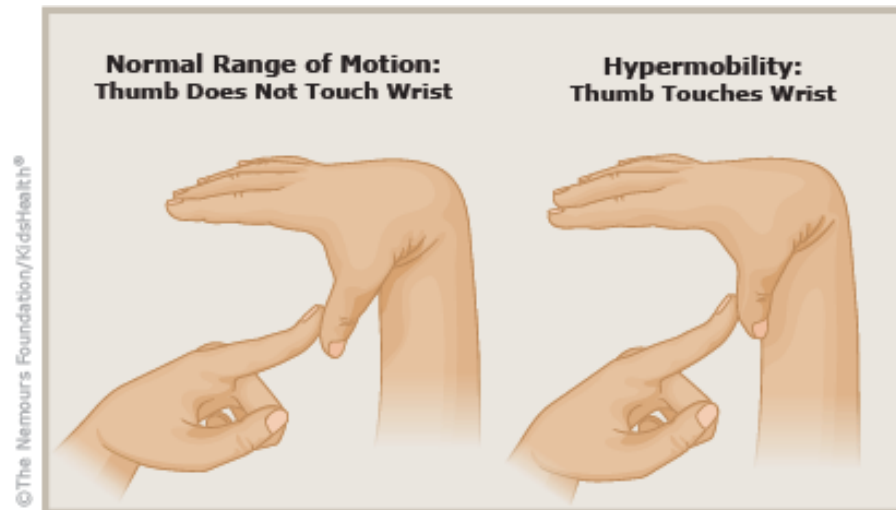
"Benign"



Not painful

Joint Hypermobility

- Joint Hypermobility is the capability of joints to move beyond normal limits
 - JH in <5 joints = localized joint hypermobility
 - JH in 5+ joints = generalized joint hypermobility



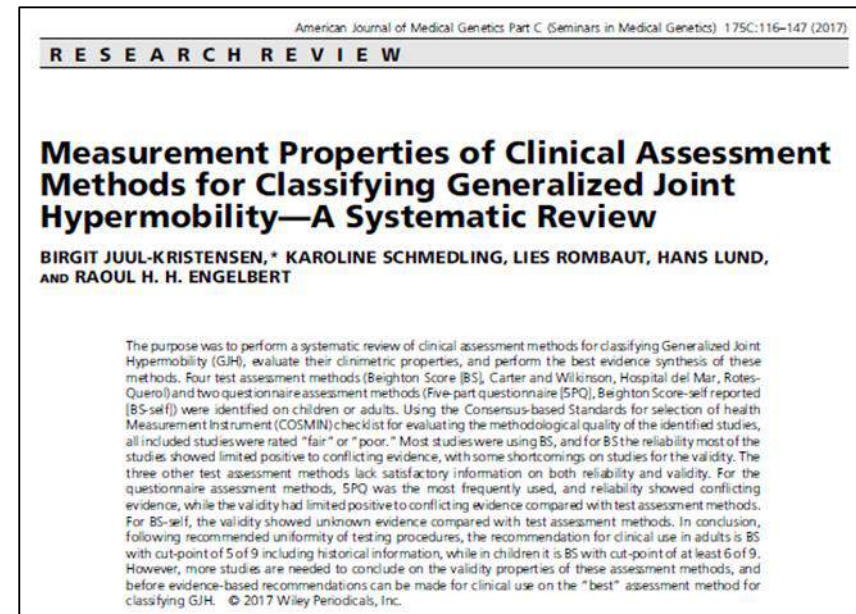
Joint Hypermobility Types

- **Generalized** joint hypermobility may be inherited or can be acquired
 - Training
 - Inflammatory or degenerative diseases
 - Hypothyroidism
 - Endocrine disorders
- **Peripheral** joint hypermobility affects only the hands/or feet
- **Historical** joint hypermobility is found in older adults who have progressively lost JH

Assessment of Joint Hypermobility

Assessment

- The **Beighton** was found to be the most used assessment with reliability – limited, positive to conflicting findings and some short comings re validity
- Recommendations for clinical use:
 - Adults: cut-point of **5/9** (including historical information)
 - Children: cut-point of at least **6/9**



Beighton Scoring System

- The Beighton Scoring System measures joint hypermobility on a 9-point scale
- Where applicable range of motion is measured using a goniometer
- The ROM assessed:
 - ☐ Dorsiflexion of the fifth MCP
 - ☐ Opposition of thumb to forearm
 - ☐ Hyperextension of the elbow
 - ☐ Hyperextension of the knee
 - ☐ Forward flexion of trunk
(knees extended/palms on the floor)

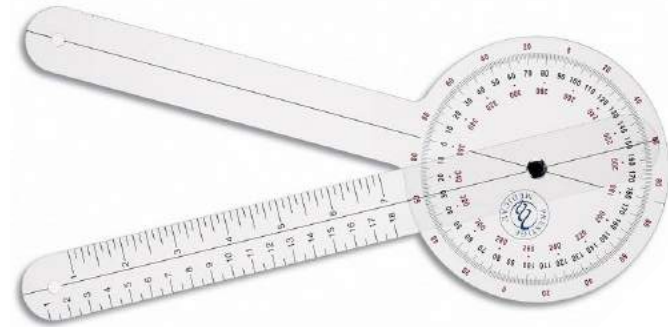


Image credit:

<https://www.prestigemedical.com/products/web/64.jpg>

Beighton Scoring System

1) I can bend my little finger up at 90 degrees (right angles) to the back of my hand



Right hand

yes ☐ no ☐

Left hand

yes ☐ no ☐

2) I can bend my thumb back on the front of my forearm



Right thumb

yes ☐ no ☐

Left thumb

yes ☐ no ☐

3) I can bend my elbow more than 10 degrees



Right elbow

yes ☐ no ☐

Left elbow

yes ☐ no ☐

4) I can bend my knee backwards



Right knee

yes ☐ no ☐

Left knee

yes ☐ no ☐

5) I can put my hands flat on the floor with my knees straight



yes ☐ no ☐

Total Yes (0-9): ____

Beighton Scoring System

- **Age is important** – results of the Beighton should be interpreted with caution for children under age 9; more research is needed on this topic

Table 1. The distribution (%) of children with different cut-off points for general joint laxity estimated by the Beighton score. Figures in bold indicate the closest upper limit for 95 % of the study population (n = 1845).

Age (y)	Gender	Beighton score					
		≥4	≥5	≥6	≥7	≥8	9
9	Boys	37.6	20.2	16.1	5.6	5.0	0.3
	Girls	47.9	26.4	21.8	7.3	5.7	1.2
12	Boys	21.0	8.6	6.2	0.6	0.3	0.0
	Girls	37.8	19.9	14.9	5.4	3.0	0.9
15	Boys	15.5	7.9	7.2	2.8	1.7	0.3
	Girls	53.0	33.4	24.0	13.9	7.4	2.0

A Jansson et al. General joint laxity in 1845 Swedish school children of different age- and gender- specific distributions. Acta Pndiatr 2004. 94: 1202-06.

Ehlers-Danlos Syndromes & Hypermobility Spectrum Disorders

INFORMATION FROM THE EHLERS-DANLOS
SOCIETY WEBSITE

<https://www.ehlers-danlos.com/ehlers-danlos-info/>

EDS & HSD



The Ehlers-Danlos Society

Ehlers-Danlos Syndromes and
Hypermobility Spectrum Disorders
2017 International Classification

<https://www.ehlers-danlos.com/ehlers-danlos-info/>

EDS & HSD



The **Ehlers-Danlos** Society.

[About Us](#)[EDS/HSD Info](#)[Community Resources](#)[Get Involved](#)[Professionals](#)[Research](#)[News](#)[Events](#)[Giving](#)

MEDICAL ARTICLES

Anesthesiology/Surgery

- **Effective plexus anesthesia in a patient with Ehlers-Danlos syndrome type III** [full paper]
- **Ehlers-Danlos Syndrome: Complications and Solutions Concerning Anesthetic Management** [full paper]
- **Local Anesthetic Failure in Joint Hypermobility Syndrome** [full paper]
- **Recommendations for anesthesia and perioperative management in patients with Ehlers-Danlos syndrome(s)** [full paper]

Autonomic

- **Postural tachycardia syndrome: a UK occupational therapy perspective** [full paper]
- **Antiadrenergic autoimmunity in postural tachycardia syndrome** [full paper]
- **Pathogenesis and Individualized Treatment for Postural Tachycardia Syndrome in Children** [full paper]
- **Effects of intermittent intravenous saline infusions in patients with medication-refractory postural tachycardia syndrome** [full paper]
- **Joint Hypermobility Syndrome and Postural Orthostatic Tachycardia Syndrome – HyPOTS** [full paper]

Cardiology

- **Abnormalities of the Spine and Reduced Bone Density in Vascular Ehlers-Danlos Syndrome** [program abstract]
- **Celiprolol: A Unique Selective Adrenoceptor Modulator** [full paper]
- **Vascular phenotypes in nonvascular subtypes of the Ehlers-Danlos syndrome: a systematic review** [full paper]

Classical EDS

- **Spectrum of mucocutaneous, ocular and facial features and delineation of novel presentations in 62 classical Ehlers-Danlos syndrome patients** [abstract]

EDS & HSD - Terminology

Old	New
Ehlers-Danlos syndrome	the Ehlers-Danlos syndromes
Hypermobility EDS	Hypermobile EDS
HEDS/VEDS/CEDS	hEDS/vEDS/cEDS
Type 3, III, 4, 2, etc.	Please do not use numeral descriptors. It will now be Hypermobile, Classical, Vascular, etc. or hEDS, cEDS, vEDS, etc.

<https://www.ehlers-danlos.com/ehlers-danlos-info/>

(Castori, Tinkle, Levy, Grahame, Malfait & Hakim, 2017)

EDS & HSD

Ehlers Danlos Syndromes (EDS)

- Group of connective tissue disorders that can be inherited
- Varied in how they affect the body and genetic causes
- Characterized by joint hypermobility, skin hyperextensibility, and tissue fragility
- Can cause symptoms throughout the body, requiring medical attention and validation

Hypermobility Spectrum Disorders (HSD)

- Group of conditions involving joint hypermobility
- Diagnosed after all other conditions that cause joint hypermobility including all EDS types have been excluded
- Can cause symptoms throughout the body, requiring medical attention and validation

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Significance of HSD

(Scheper et al., 2016)

- Scheper, M. C., Juul-Kristensen, B., Rombaut, L., Rameckers, E. A., Verbunt, J., & Engelbert, R. H. (2016). Disability in adolescents and adults diagnosed with hypermobility-related disorders: a meta-analysis. *Archives of physical medicine and rehabilitation*, 97(12), 2174-2187.
- Meta-analysis of 21 articles that included associated symptoms, impact on disability, and treatment of generalized joint hypermobility (GJH), hypermobility syndrome (HMS), and Ehlers-Danlos syndrome – hypermobility type (EDS-HT)

Significance of HSD

(Scheper et al., 2016)

Key Findings from the meta-analysis:

- **Similar levels of disability and comparable levels of pain severity** were present among both HMS and EDS-HT groups
- Subgroups of patients with EDS-HT have been identified as presenting with different types of complaints and severity. Children diagnosed with HMS appear to show similar subgroups.
- There is large variability and insufficient differentiation among HMS and EDS-HT, therefore health care providers must recognize the clinical variability within these diagnoses and ensure that assessment incorporates all aspects of the ICF model

EDS & HSD



Types of hypermobility spectrum disorders

NAME OF HSD	BEIGHTON SCORE	MUSCULOSKELETAL INVOLVEMENT
Asymptomatic GJH	Positive	Absent
Asymptomatic PJH	Usually negative	Absent
Asymptomatic LJH	Negative	Absent
Generalized-HSD	Positive	Present
Peripheral-HSD	Usually negative	Present
Localized-HSD	Negative	Present
Historical-HSD	Negative*	Present
hEDS	Positive	Possible

* Historical presence of joint hypermobility (*e.g.*, positive 5-point questionnaire)

<https://www.ehlers-danlos.com/ehlers-danlos-info/>

Associated Conditions & Symptoms

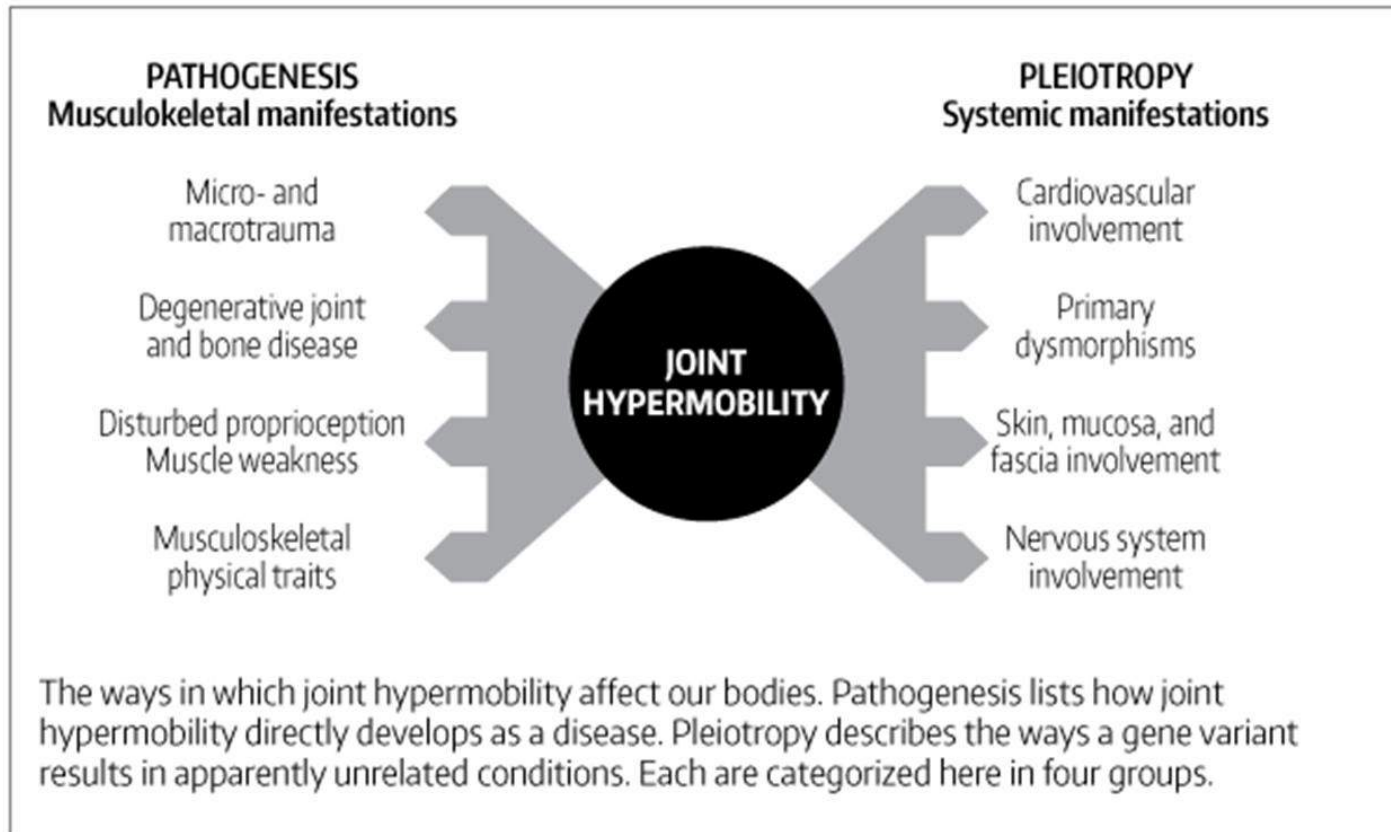
Associated Conditions & Symptoms

- Trauma
 - Macrotrauma (dislocation, subluxation, and connected tissue damage)
 - Microtrauma (injuries too small to be noticed as they happen)
- Chronic Pain
 - Occasional, recurring pain is a natural result of the trauma
 - Chronic pain can develop from sensitivity to pain, impaired connective tissue
- Disturbed Proprioception
- Musculoskeletal traits
 - Flat feet (flexible type), misaligned bones in the elbow and big toes, mild to moderate scoliosis, kyphosis of upper spine, lordosis of the lower spine

Associated Conditions & Symptoms

- Issues not the direct result of the mechanics of JH:
 - Very real and seriously affect quality of life
 - Must be managed as part of treatment
- The strongest (but not only) associations:
 - Anxiety disorders
 - Orthostatic tachychardia
 - Functional gastrointestinal disorders
 - Pelvic and bladder dysfunction
- **These additional problems need to be evaluated and treated when an HSD is diagnosed**

Associated Conditions & Symptoms



Associated Symptoms & Conditions

Chronic fatigue and gastrointestinal disorders are common in hypermobility conditions

- **Chronic fatigue**
 - Symptoms overlap with chronic fatigue syndrome
 - Problems that worsen fatigue include: sleep disorders, autonomic dysfunction, chronic and acute pain, deconditioning, psychological issues, and nutritional deficiencies
- **Gastrointestinal disorders**
 - Structural (hiatal hernia, rectal prolapse)
 - Functional (irritable bowel syndrome, gastroparesis)
 - Treatment is based on symptoms

EDS - Management

Psychological Symptoms

- Association between EDS and anxiety, depression, and neurodevelopmental disorders (ADHD, ASD)
- Treatment can include medication, psychotherapy, and treatment of physical symptoms

Orthopedic Management

- EDS can cause early damage to joints and joint instability
- Professionals involved include physical therapists and orthopedic surgeons with EDS experience

<https://www.ehlers-danlos.com/ehlers-danlos-info/>

EDS & HSD – Management

Physical Therapy

- Key is individualized treatment for each client
- Useful to address: pain, proprioception, balance, & muscle tone

Autonomic Dysfunction

- Includes: postural orthostatic tachycardia syndrome, orthostatic intolerance, and naturally mediated hypotension
- Treatment: increased fluids and salt, compression wear, and increased physical activity

Pain Management

- Pain can be related to the joints, GI system, TMJ, and headaches/migraines
- Management: treating the cause, physical therapy, medications, braces or cushion, and compression wear

Questions or Comments

What is the role of OT in EDS & HSD Management?

Implications

What does this mean for therapists and our clients & families?



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Health Conditions

1. Arthritis (Rheumatology)
2. Attention Deficit Disorder(see also Learning Disabilities)
3. Autism
4. Burns
5. Brain Injury
6. Cerebral Palsy
7. Developmental Coordination Disorder
8. Diabetes, Digestive and Kidney Disease
9. Down Syndrome
10. Dyslexia
11. Epilepsy
12. Feeding/Nutrition Issues
13. Fetal Alcohol Spectrum Disorders
14. Fibrodysplasia Ossifications Progressiva (FOP)
15. Genetic Databases
16. Hearing Impairment
17. HIV/AIDS
18. Intellectual Disabilities
19. Joint Hypermobility and Related Disorders
20. Learning Disabilities (see also Attention Deficit Disorder)
21. Mental Health
22. Movement Disorders
23. Multiple Sclerosis
24. Muscle Disease
25. Neurological Disorders
26. Pain
27. Rett Syndrome
28. Sleep Disturbances
29. Spinal Cord Lesions
30. Stroke
31. Turner Syndrome

CDR Website Poll

Did you find this website useful?

☐ Yes☐ No[Vote!](#) [Results](#)

Child Development & Rehabilitation

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Joint Hypermobility and Related Disorders

1. Ehlers-Danlos Syndromes (EDS) & Hypermobility Spectrum Disorders (HSD)
2. Identification and Management of Pediatric Joint Hypermobility (Cincinnati Children's Hospital Medical Center)
3. Hypermobility Syndromes Association
4. Open access articles from the International Consortium on EDS
5. Ehlers-Danlos Syndrome at Medline Plus

Last Updated: May 24, 2018

CDR Website Poll

Did you find this website useful?

School-Based OT Management of Hypermobility (HSD) in Children

WITH A FOCUS ON MANAGEMENT OF FATIGUE,
POSTURE AND FINE MOTOR

Prevalence – Across the Age Span

- Hypermobility is very common, especially in children and women (Castori and Hakim, 2017)
- The estimated prevalence of HSD in children and adults varies from 2% to 57% (Remvig et al. 2007)



Prevalence - Pediatrics



- The reported prevalence of joint hypermobility in children varies widely - between 2.3% and 39% depending on the criteria used and the population studied [\(Coles et al., 2018\)](#)
- In a sample of 285 English school children excessive mobility in 4 or more joint pairs was noted in 7% [\(Coles et al., 2018\)](#)
- Asymptomatic in some children, however in many it is associated with significant pain and disability with consequent impact to the child and family [\(Coles et al., 2018\)](#)

Co-Morbidity with ASD



SPEAKER
Carolina Baeza-Velasco

PRESENTATION

Autism, hypermobility and EDS



- Evidence of the association between autism and HSD/EDS
- Clinical illustrations of the association autism and HSD/EDS

Co-Morbidity with ASD

- Scarce but growing body of research suggesting that ASD and HSD co-occur more often than expected by chance (Baeza-Velasco et al. 2018)



Autistic manifestations expression in a hypermobile body

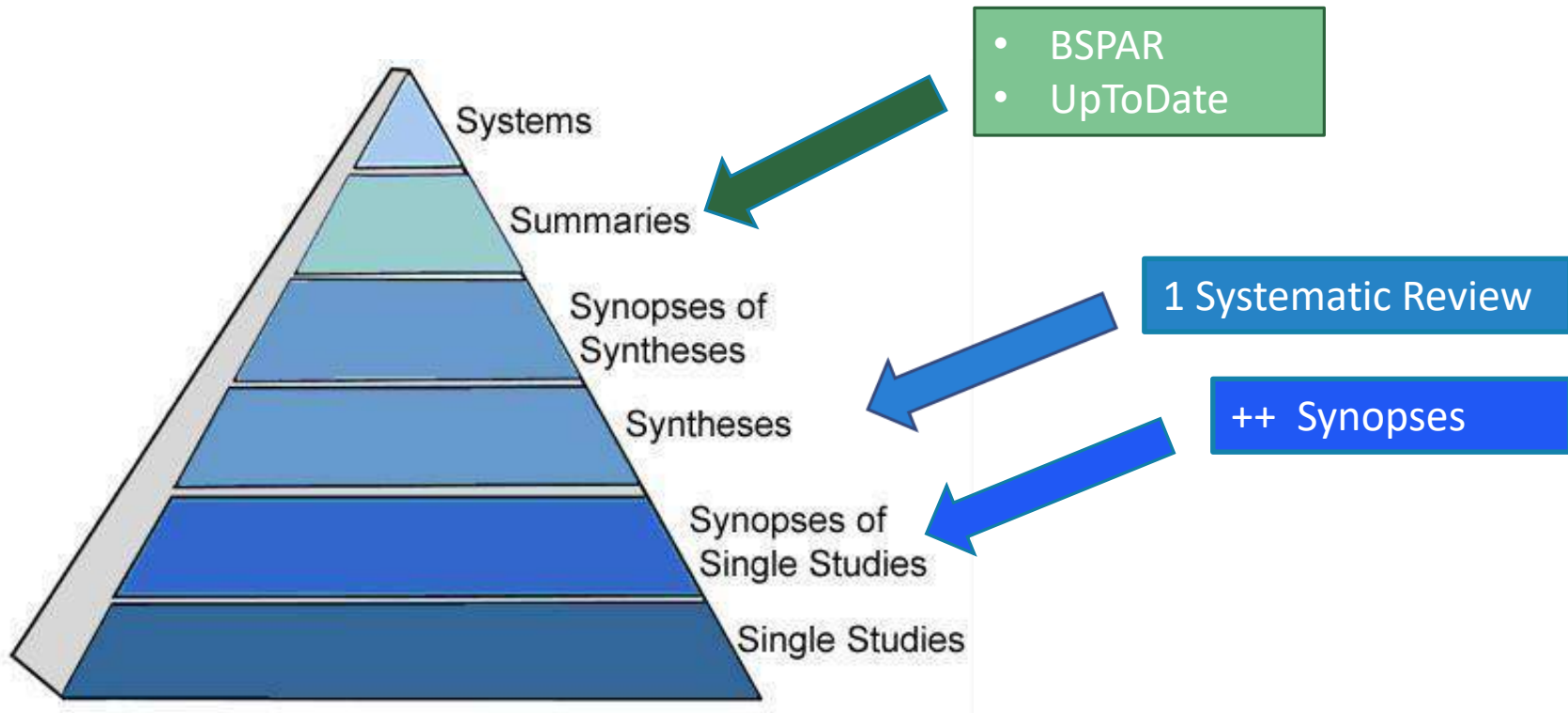


Co-Morbidity with DCD

- Hypermobility has been noted to co-occur with DCD; prevalence is noted to be significantly higher in children with DCD than children who do not have DCD
(Baudinette et al., 2010; Jelsma et al., 2013; Kirby & Davies, 2007).
- More mobility of the joints may be a disadvantage when motor coordination is poorly developed



Evidence



6s Hierarchy of Pre-Appraised Evidence

PLUS: Expert Opinion

2017 Consortium with +++ supporting literature

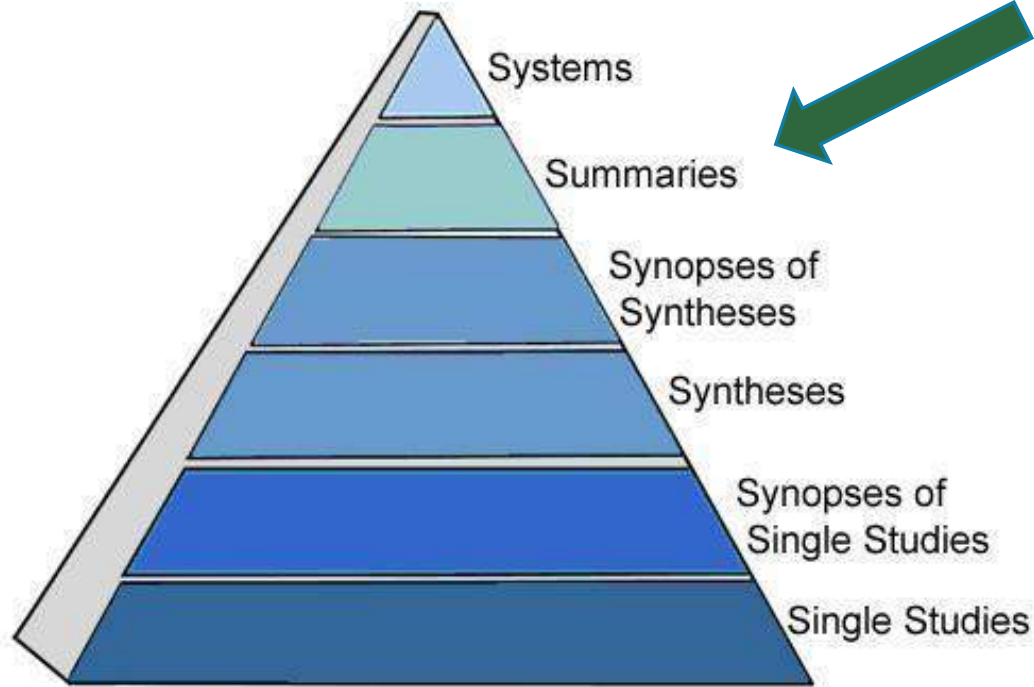
<https://www.ehlers-danlos.com/2017-eds-international-classification/>



The Ehlers-Danlos Society

Ehlers-Danlos Syndromes and
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Evidence – Guidelines (Summaries)



- BSPAR
- UpToDate

6s Hierarchy of Pre-Appraised Evidence

S2 Level: Summary - BSPAR Guideline



2013

The British Society for Paediatric and Adolescent Rheumatology
Registered Charity # 1132967

Guidelines for Management of Joint Hypermobility Syndrome in Children and Young People.

A Guide for professionals managing young people with this condition

These guidelines have been compiled by the Allied Health Professionals Group of the British Society for Paediatric and Adolescent Rheumatology (BSPAR, 2012).

In order to cover the complexities of this condition some aspects of management have been divided into different professions, however there will be significant overlap as to who provides the intervention depending upon local teams.

Introduction

These guidelines are designed to help and support therapists working with children and young people with Joint Hypermobility and musculoskeletal pain.

AGREE II Appraisal Instrument¹ for BSPAR 2013 article

Rating Questions	Reviewer
1. The overall objective(s) of the guideline is (are) specifically described.	5
2. The health question(s) covered by the guideline is(are) specifically described.	3
3. The population (patients, public, etc.) to whom the guideline is meant to apply are specifically described.	2
Scope and Purpose Domain Total Score	
4. The guideline development group includes individuals from all the relevant professional groups.	2
5. The views and preferences of the target population (patients, public, etc.) have been sought.	1
6. The target users of the guideline are clearly defined.	3
Stakeholder Involvement Domain Total Score	
7. Systematic methods were used to search for evidence.	1
8. The criteria for selecting the evidence are clearly described.	1
9. The strengths and limitations of the body of evidence are clearly described.	1
10. The methods for formulating the recommendations are clearly described.	1
11. The health benefits, side effects and risks have been considered in formulating the recommendations.	2
12. There is an explicit link between the recommendations and the supporting evidence.	1
13. The guideline has been externally reviewed by experts prior to its publication.	1
14. A procedure for updating the guideline is provided.	4
Rigour of Development Domain Total Score	
15. The recommendations are specific and unambiguous.	3
16. The different options for management of the condition or health issue are clearly presented.	3
17. Key recommendations are easily identifiable.	2
Clarity and Presentation Domain Total Score	
18. The guideline describes facilitators and barriers to its application.	1
19. The guideline provides advice and/or tools on how the recommendations can be put into practice.	2
20. The potential resource implications of applying the recommendations have been considered.	1
21. The guideline presents monitoring and/or auditing criteria.	1
Applicability Domain Total Score	
22. The views of the funding body have not influenced the content of the guideline.	1
23. Competing interests of guideline development group members have been recorded and addressed.	1
Editorial Independence Total Score^a	
Overall Guideline Assessment Quality^b	2
Overall Guideline Assessment Recommendation for Use^c	Yes with modifications

S2 Level: Summary - Up to Date (2017)

UP TO DATE

Joint hypermobility syndrome

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Contributor disclosures

Rodney Grahame, MD, FRCP, FACP Nothing to disclose. **Alan J Hakim, BA MB BChir** Nothing to disclose. **Peter H Schur, MD** Nothing to disclose. **Paul L Romain, MD** Nothing to disclose.

Contributor disclosures are reviewed for conflicts of interest by the editorial group. When found, these are addressed by vetting through a multi-level review process, and through requirements for references to be provided to support the content. Appropriately referenced content is required of all authors and must conform to UpToDate standards of evidence.

AGREE II Appraisal Instrument¹ for UpToDate 2017

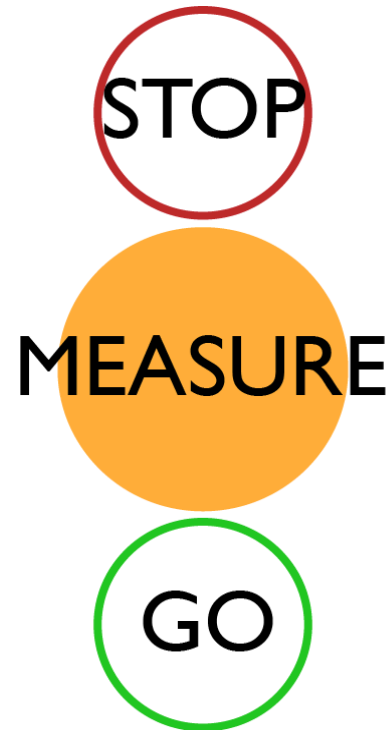
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BSPAR (2013) Guideline & Up To Date (2017) Summary

- Both did not follow traditional guideline development criteria. Therefore both scored low on the AGREE II
- AGREE II INSTRUMENT – Both scored only a 2/7
- However, these guidelines match typical clinical presentation, concerns, management principles and strategies used
- Need to be updated and expanded to better adhere to standard guideline development considerations

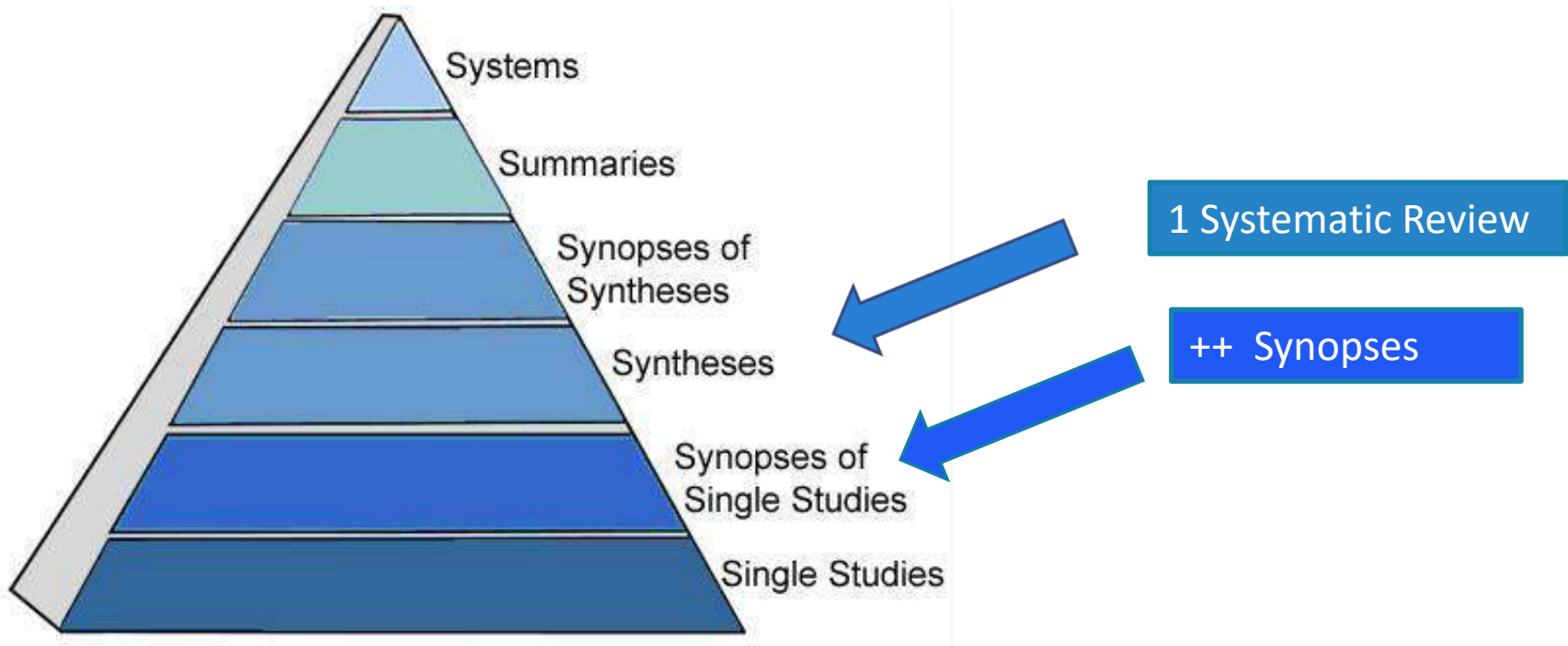
TRAFFIC LIGHT : Highest Level of Evidence (2 Guidelines)

- Group design Level I or II evidence of poor quality, regardless of outcome
- Insufficient Evidence



Evidence: S4 & S5

Systematic Review & Synopses



6s Hierarchy of Pre-Appraised Evidence

S4 Level: Syntheses – Systematic Review

Only 3 studies:

- 2/3 pediatric
- Of those 2 studies:
 - 1 OT (Hand & wrist splints)
 - 1 PT
- OT study had a high risk of bias

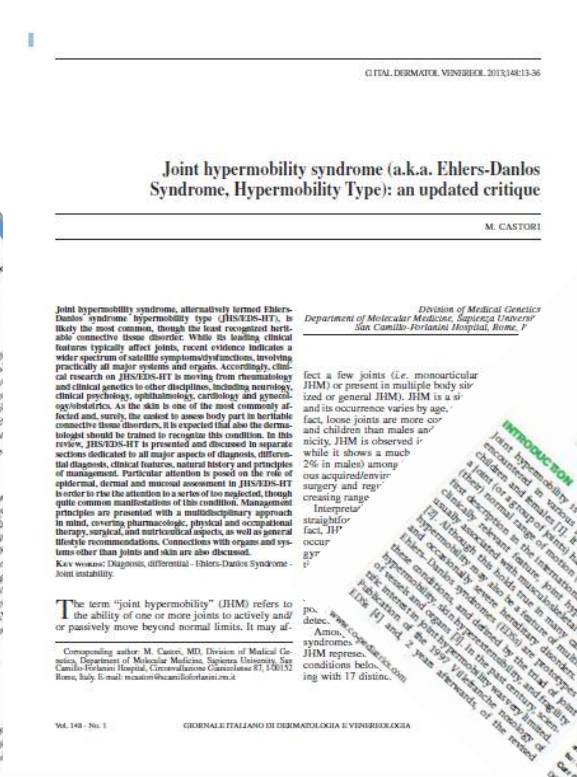


OT Specific Conclusions:

- Insufficient evidence
- Limited size and quality of research

([Smith et al., 2014](#))

S5 Level: Synopses – Review Articles



Plus 2 Existing Handouts



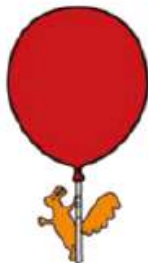
2017

AN EDUCATOR'S GUIDE

Meeting the Needs of
the Ehlers-Danlos Child

A PARENT'S GUIDE

Helping Your Child
Succeed at School



Occupational Therapy Department

Hypermobility

Information for parents, carers and schools

Introduction

All joints have varying ranges of movement. Ligaments and muscles provide a joint with stability and movement and receptors in the joints inform us of our position in the environment. Children who are hypermobile have too much movement around their joints. The looseness of the supporting structures lead to joint instability. Excess movement may cause brief discomfort, pain and swelling. A growth spurt, lack of exercise or an accident can increase symptoms.

If there is an underlying reason for joint hypermobility, this will be investigated by the doctors. Therapy advice will remain the same, regardless of the reason for the hypermobility.

Possible symptoms

- **Fatigue.** Children may experience two types of fatigue:
 - specific joint or muscle fatigue because joints and muscles are working extra hard
 - general overall fatigue due to increased effort to perform activities
- **Pain.** Increased range of movement can lead to pain around joints.
- **Co-ordination.** The receptors that tell us the position of our bodies when we can't see are in our joints. When there is too much movement around the joints the receptors don't get the same messages as when there is less movement. These symptoms may lead to poor co-ordination and difficulties with daily activities such as handwriting, holding cutlery and getting dressed.



As a child develops and becomes more active, hypermobile joints often / generally become stronger and pain subsides. Co-ordination may improve and your child should be less tired.

Management strategies for parents and carers

Good body mechanics:

The way that your child carries their body largely affects how much strain they put on their joints. Considering posture and movement allows your child to use their body more efficiently and conserve energy.

- When sitting at a desk your child's feet should be flat on the floor. Their thighs and forearms should be horizontal and the desk just below elbow height.
- When your child is using a keyboard for long periods consider using a chair with arms or wrist or forearm supports.

2019

KEY MESSAGES:

These evidence summaries, synopses along with the consortium resources were used to address our main concerns in school-aged children:

- Fatigue
- Decreased endurance
- Decreased posture
- Fine motor challenges
- Decreased “emotional regulation” and need for strategies to manage subsequent to fatigue

Pain and Fatigue (Castori 2013)

- Chronic/recurrent pain and fatigue are, by far, the most common neurologic complaints, reported in, perhaps, all adults
- Pain manifestations are widespread and involve the musculoskeletal system, as well as the nervous system and internal organs
- Multiple studies demonstrated that chronic fatigue is a major contributor to disability
- Associated complaints include muscle weakness, sleep disturbance and other features of chronic fatigue syndrome

**I'm still tired from
yesterday's tired.
Today isn't looking
so good, and I've already
used up tomorrow's tired.**



Lifestyle recommendations (Castori 2013)

Promote:

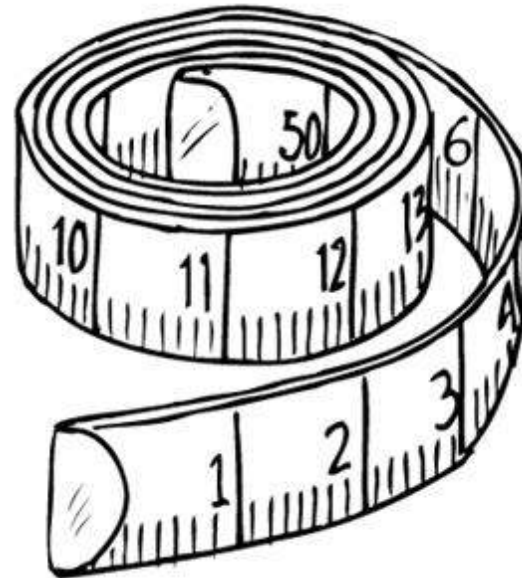
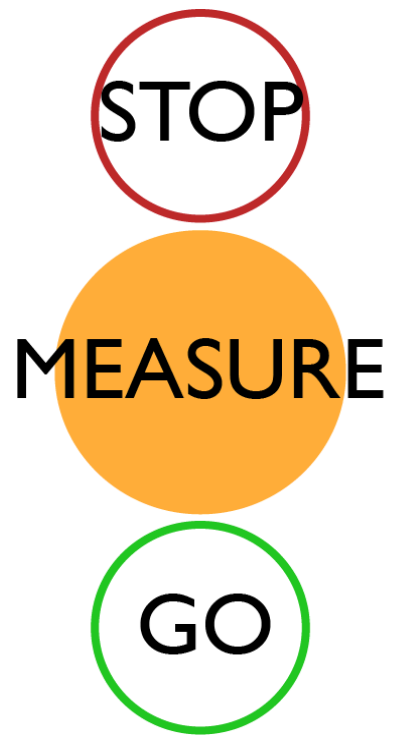
- Regular, aerobic fitness
- Strengthening and proprioceptive exercises
- Postural and ergonomic hygiene
- Daily relaxation activities

Lifestyle recommendations (Castori 2013)

Avoid:

- High impact sports/activities
- Low environmental temperatures
- Prolonged sitting positions and prolonged recumbency
- Sudden head-up postural change
- Excessive weight lifting/carrying

Measurement



Measures of Fatigue (Hewlett et al., 2011)

Arthritis Care & Research
Vol. 63, No. S11, November 2011, pp S263–S286
DOI 10.1002/acr.20579
© 2011, American College of Rheumatology

MEASURES OF PATHOLOGY AND SYMPTOMS

Measures of Fatigue

Bristol Rheumatoid Arthritis Fatigue Multi-Dimensional Questionnaire (BRAFMQ), Bristol Rheumatoid Arthritis Fatigue Numerical Rating Scales (BRAFNRS) for Severity, Effect, and Coping, Chalder Fatigue Questionnaire (CFQ), Checklist Individual Strength (CIS20R and CIS8R), Fatigue Severity Scale (FSS), Functional Assessment Chronic Illness Therapy (Fatigue) (FACIT-F), Multi-Dimensional Assessment of Fatigue (MAF), Multi-Dimensional Fatigue Inventory (MFI), Pediatric Quality Of Life (PedsQL), Multi-Dimensional Fatigue Scale, Profile of Fatigue (ProF), Short Form 36 Vitality Subscale (SF-36 VT), and Visual Analog Scales (VAS).

SARAH HEWLETT, EMMA DURES, AND CELIA ALMEIDA

INTRODUCTION

Fatigue is common to all the rheumatic conditions, in varying degrees, and is a frequent, often severe problem that has major consequences on patients' lives (1–4). In response to these concerns, a body of research subsequently led to international consensus that fatigue must be evaluated in all clinical trials of rheumatoid arthritis and potentially all fibromyalgia syndrome trials (5,6). The 12 fatigue patient-reported outcome measures (PROMs) reviewed in this section have been selected because they are currently or have recently been used in rheumatology

alphabetical order. Three additional scales with fatigue components are reviewed elsewhere in this edition: the Bath Ankylosing Spondylitis Disease Activity Index in the Measures of Ankylosing Spondylitis article, the Fibromyalgia Impact Questionnaire in the Measures of Fibromyalgia article, and the Nottingham Health Profile in the Adult Measures of General Health and Health-Related Quality of Life article.

When selecting a fatigue PROM, researchers and clinicians should consider whether their needs are best served by a single-item PROM as a screening tool, by multi-item PROMs that explore broader fatigue issues to create a

Pediatric Quality of Life (PedsQL): Multi-Dimensional Fatigue Scale (Varni, Burwinkle, & Szer, 2004)

Pediatric Quality of Life Inventory™ (PedsQL™)
Varni JW

PROQOLID™
Distributed by Mapi Research Trust

> Basic description

> Access this questionnaire

> Contact and conditions of use

BASIC DESCRIPTION

DISEASE-SPECIFIC MODULES:
All the available PedsQL™ modules are listed on the "RELATED COA INSTRUMENTS" section in the left column.

Authors
Varni JW

Copyright
Copyright © 1998 JW Varni, Ph.D. All rights reserved

- A free tool!

https://eprovide.mapi-trust.org/instruments/pediatric-quality-of-life-inventory#basic_description

Objective : To assess health-related quality of life in children in various diseases areas. Applicable for healthy school and community populations, as well as pediatric populations with acute and chronic health conditions

PedsQL Purpose

- The PedsQL was developed:
 - To measure child and parent perceptions of fatigue
 - For use in patients with cancer but is intended as a generic measure for pediatric patients
- Versions available:
 - Young adults (ages 18–25),
 - Teenagers (ages 13–18)
 - Children (ages 8–12)
- Developmentally appropriate language, with mirror versions for their parents (“smiley-face” response version is available for children, a written version for parents, and a parent version for toddlers)

PedsQL Content

Covers domains of general fatigue (e.g., I feel tired), fatigue related to sleep/rest (e.g., I feel tired when I wake up in the morning), and cognitive fatigue (e.g., it is hard for me to keep my attention on things)

- Number of items: 18 items, giving a total fatigue score and including 3 subscales, each of 6 items (general fatigue, sleep/rest fatigue, and cognitive fatigue)
- Response options: 5 response options from “Never a problem” to “Almost always a problem.”
- Recall period for items: Acute version 7 days, standard version 1 month

PedsQL Examples of Use

The PedsQL Multi-Dimensional Fatigue Scale is a module from PedsQL Measurement model, a modular approach to measuring pediatric health-related quality of life

It has been used in studies of mixed rheumatologic disorders, fibromyalgia syndrome (FMS), and juvenile idiopathic arthritis (JIA) as well as patients with cancer, cerebral palsy, obesity, cerebral tumours, chronic pain, and multiple sclerosis

Psychometric Information

Reliability

Internal consistency: In rheumatology, Cronbach's alpha ranged from 0.88–0.95 for the total scale and 3 subscales for all age-appropriate versions (n 163); in fibromyalgia syndrome (FMS), Cronbach's alpha ranged from 0.76–0.94 (n 29)

Test–retest: No data could be found for the PedsQL.

Interrater reliability: Child and parent (proxy) fatigue scores correlated in a rheumatology population (n 163)

Validity

Content validity: Items were generated through literature review, and patient and parent focus groups and individual interviews in cancer populations

Construct validity: In 175 children with a range of rheumatologic conditions, total fatigue and the 3 subscales *correlated strongly with quality of life, pain, physical and psychosocial health, and emotional, social, and school functioning* at 0.53–0.91, while *all scales had slightly lower, but still positive, associations with daily activities* (0.48–0.58)

Ability to detect change: No sensitivity to intervention data found for any population







Plus: Mu et al. 2018 used PedsQL for HSD

DOI: 10.1002/ajmg.a.61055

WILEY AMERICAN JOURNAL OF PART
medical genetics **A**

ORIGINAL ARTICLE

Factors affecting quality of life in children and adolescents with hypermobile Ehlers-Danlos syndrome/hypermobility spectrum disorders

Weiyi Mu¹  | Michael Muriello¹  | Julia L. Clemens¹ | You Wang² |
Christy H. Smith¹  | Phuong T. Tran^{3,4} | Peter C. Rowe¹ | Clair A. Francomano⁵  |
Antonie D. Kline⁵  | Joann Bodurtha¹ 

Method

- Survey responses were completed for 47 children and adolescents with hEDS/hypermobility spectrum disorder (81% female, mean age 16 years), some by the affected individual, some by their parent, and some by both.
- Clinical data derived from chart review were compared statistically to survey responses

Findings

- All outcomes correlated moderately to strongly with each other
- Using multiple regression, **general fatigue and pain scores** were the **best predictors of the PedsQL total score**
- Additionally, presence of any psychiatric diagnosis was correlated with a lower PedsQL score
- Current management guidelines recommend **early intervention to prevent disability from de-conditioning**; these results may help identify target interventions in this vulnerable population

PedsQLTM

Pediatric Quality of Life Inventory

Version 4.0

CHILD REPORT (ages 8-12)

DIRECTIONS

On the following page is a list of things that might be a problem for you. Please tell us **how much of a problem** each one has been for you during the **past ONE month** by circling:

- 0 if it is **never** a problem
- 1 if it is **almost never** a problem
- 2 if it is **sometimes** a problem
- 3 if it is **often** a problem
- 4 if it is **almost always** a problem

There are no right or wrong answers.
If you do not understand a question, please ask for help.

In the past **ONE month**, how much of a **problem** has this been for you ...

ABOUT MY HEALTH AND ACTIVITIES (problems with...)	Never	Almost Never	Some- times	Often	Almost Always
1. It is hard for me to walk more than one block	0	1	2	3	4
2. It is hard for me to run	0	1	2	3	4
3. It is hard for me to do sports activity or exercise	0	1	2	3	4
4. It is hard for me to lift something heavy	0	1	2	3	4
5. It is hard for me to take a bath or shower by myself	0	1	2	3	4
6. It is hard for me to do chores around the house	0	1	2	3	4
7. I hurt or ache	0	1	2	3	4
8. I have low energy	0	1	2	3	4

ABOUT MY FEELINGS (problems with...)	Never	Almost Never	Some- times	Often	Almost Always
1. I feel afraid or scared	0	1	2	3	4
2. I feel sad or blue	0	1	2	3	4
3. I feel angry	0	1	2	3	4
4. I have trouble sleeping	0	1	2	3	4
5. I worry about what will happen to me	0	1	2	3	4

HOW I GET ALONG WITH OTHERS (problems with...)	Never	Almost Never	Some- times	Often	Almost Always
1. I have trouble getting along with other kids	0	1	2	3	4
2. Other kids do not want to be my friend	0	1	2	3	4
3. Other kids tease me	0	1	2	3	4
4. I cannot do things that other kids my age can do	0	1	2	3	4
5. It is hard to keep up when I play with other kids	0	1	2	3	4

ABOUT SCHOOL (problems with...)	Never	Almost Never	Some- times	Often	Almost Always
1. It is hard to pay attention in class	0	1	2	3	4
2. I forget things	0	1	2	3	4
3. I have trouble keeping up with my schoolwork	0	1	2	3	4
4. I miss school because of not feeling well	0	1	2	3	4
5. I miss school to go to the doctor or hospital	0	1	2	3	4

Questions or Comments

Our Knowledge Product: A User Friendly Handout

MANAGEMENT OF
HYPERMOBILITY IN SCHOOL
AGED-CHILDREN



FINE MOTOR MANAGEMENT OF HYPERMOBILITY

The following suggestions are additional strategies might be needed for children with Joint Hypermobility Disorder and Ehlers-Danlos Syndrome. Children who are hypermobile are extra flexible. In the past, they exist by itself or be a part of a condition. If there is an underlying reason for the hypermobility, your doctor. Therapy and management of the hypermobility can be wide-spread (five or more joints). The impacts of hypermobility from minor to moderate dislocations, trouble with muscles and effort to perform. For many of the hypermobility cravens, easy management.

- Scheduling and Pacing:**
- Plan daily activities to allow for a balance and variety of different types of activity and rest.
 - Spread out difficult or demanding tasks over the day or week.
 - Split activities into small achievable tasks.
 - Use two sets of textbooks (one for home and one for school) to avoid transport.
 - A small snack and a brisk walk or other type of exercise will often re-energize the body in order to continue with the day's activities.
 - Alternatively a rest may be needed.
- Rest Periods:** As part of pacing, rest periods may be needed and can be the daily school routine. Rest advice might include:
- Be proactive in giving consistent energy throughout the day.
 - Afternoon may result in lack of productivity and need for rest.
 - Even a short rest period may have a large benefit.
 - Offer a quiet space for resting, ideally away from noise.
 - Provide comfortable supportive furniture for resting.
 - chair, or children's arm chair. Encourage children to sit on the floor.
 - Provide comfortable supportive furniture for resting.
 - chair, or children's arm chair. Encourage children to sit on the floor.
 - Provide comfortable supportive furniture for resting.
 - chair, or children's arm chair. Encourage children to sit on the floor.



Developed 2010 by Ivonne Montgomery, Lynda Swain and Samantha Jenkins, Occupational Therapists
<http://www.childdevelopment.ca/FineMotorResourcesGroup/ClassroomResources.aspx>

2) Classroom Seating and Positioning:

- Supportive seating is very important and allows rather than focusing on sitting.
- Fit the chair first. Ensure that the child should be at 90 degrees. Lower back chair. Providing a smaller chair, footrests cannot be met.
 - Once the chair has been fitted, ensure the child's arms are well supported, weight through 2 inches above the elbows. Encourage the child to sit "tummy to desk" to sit close to the desk.
 - Use of a slant board may be helpful for wrist positioning for printing. A slant board can be used for keyboarding.
 - Ensure forearms are well supported.
 - Avoid sitting for extended periods.
 - When standing, the work should be comfortably without stooping.
 - During carpet time try a beanbag chair or book case rather than a chair.



Images retrieved from:
<http://www.childdevelopment.ca/FineMotorResourcesGroup/ClassroomResources.aspx>



3) Fine Motor and Other Accommodations:

- Trial use of a "pencil seat belt" to provide extra support and stability. (Use of a seatbelt is often accepted when a pencil grip is not.)



Images retrieved from:
<http://www.childdevelopment.ca/FineMotorResourcesGroup/ClassroomResources.aspx>

Health Centre for Children

Our handout focuses on 4 areas:

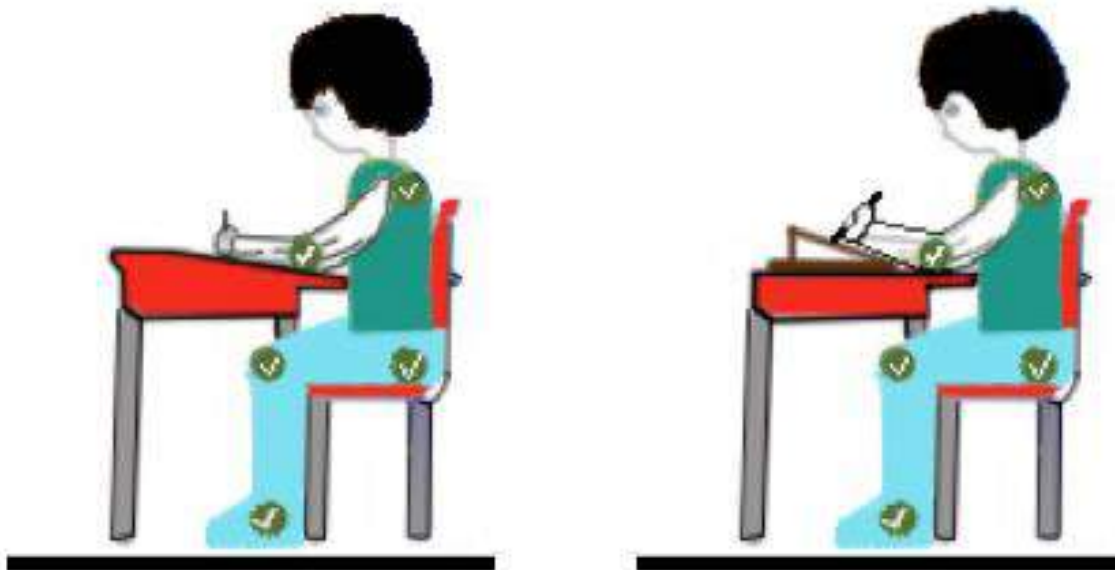
- 1) Classroom Seating and Positioning:
- 2) Energy Management/Pacing:
- 3) Fine Motor and Other Accommodations
- 4) Tips for parents:
 - Sleep
 - Physiotherapy
 - Relaxation techniques

1) Classroom Seating & Positioning

Supportive seating allows the student to best concentrate on learning rather than on posture

- Fit the chair first with hips and knees at 90 degrees and feet flat on the floor
- Table top should be adjusted so that the forearm is well supported with the elbow at 90 degrees ; ideally 2" above bent elbow height so they can bear weight
- Avoid sitting for extended periods of time

Classroom Seating & Positioning



Classroom Seating & Positioning



Image credit: <https://www.amazon.com/Adapt-Ease-Ergonomic-Writing-Slant-Board/dp/B06XY7VMF8>



Image credit: https://store.schoolspecialty.com/OA_HTML/ibeCCtpItnDspRte.jsp?minisite=10224&item=1580571

Classroom Seating & Positioning



Classroom Seating & Positioning



Image credit: CAP Furniture

<https://capfurniture.com.au/product/650-650-cutout-table/>



Image credit: School Outfitters

https://www.schooloutfitters.com/catalog/product_info/pfam_id/PFA_M7252/products_id/PRO18342

Classroom Seating & Positioning



Image credit: <https://www.ikea.com/in/en/p/poaeng-childrens-armchair-birch-veneer-almas-beige-s49337939/>

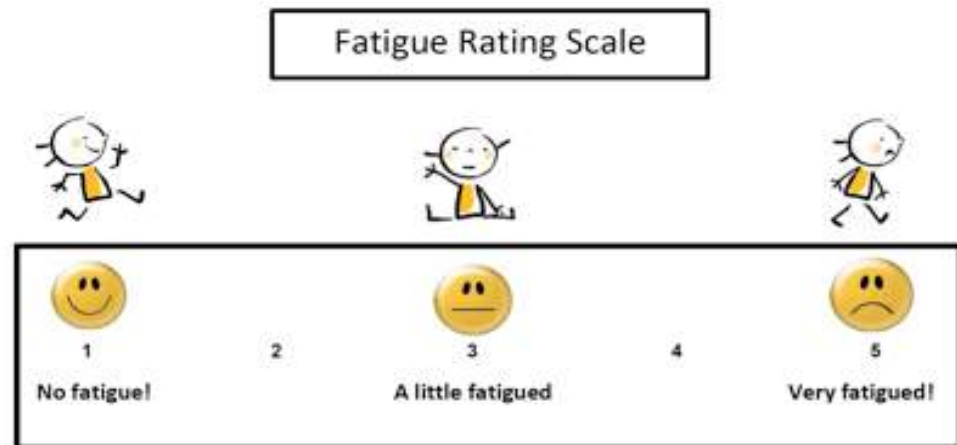


Image credit: <https://www.wintergreen.ca/flex-space-comfy-floor-seat-green-lc405gr>

2) Energy Management / Pacing

Students with hypermobility often suffer from fatigue and once exhausted can take a long time to recover

- Plan activities to allow for balance, variety and rest
- Break activities into smaller bits
- Be proactive in providing rests
- Teach student to self-monitor their fatigue and self-advocate for rests



Pacing with use of Rest Breaks

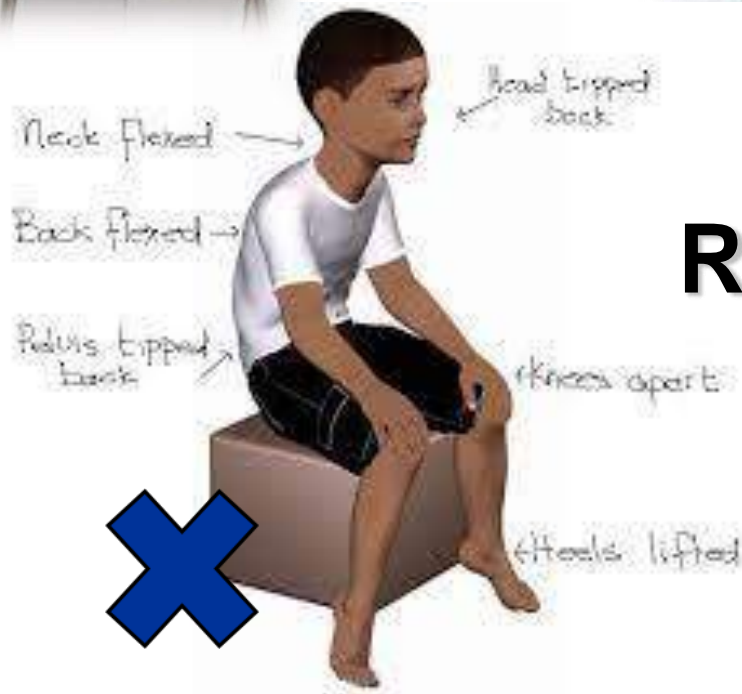
Image credit: <https://tinyurl.com/3acnf9jj>



Image credit: rb.gy/tl0fgi



Image credit: <https://rb.gy/z5ywx>



Rest Breaks



Image credit: <https://bit.ly/3vAtwiY>

Image credit: <https://bit.ly/3nCc87y>

Classroom Seating & Positioning Furniture for Rest Breaks

Key Ingredients



Image credit: <https://bit.ly/3aUwYxf>



Head Supported



Reclined



Feet Elevated



Image credit: <https://bit.ly/3u9roi4>

Classroom Seating & Positioning Furniture for Rest Breaks



Head
Supported



Reclined



Feet
Elevated

Self/Co-Monitoring of Fatigue

Tracking with use of a Fatigue Rating Scale

Fatigue Rating Scale



1

No fatigue!

2



3

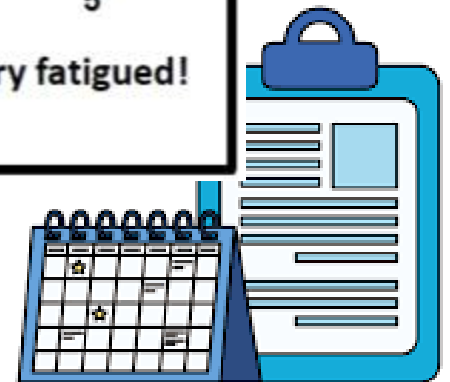
A little fatigued

4



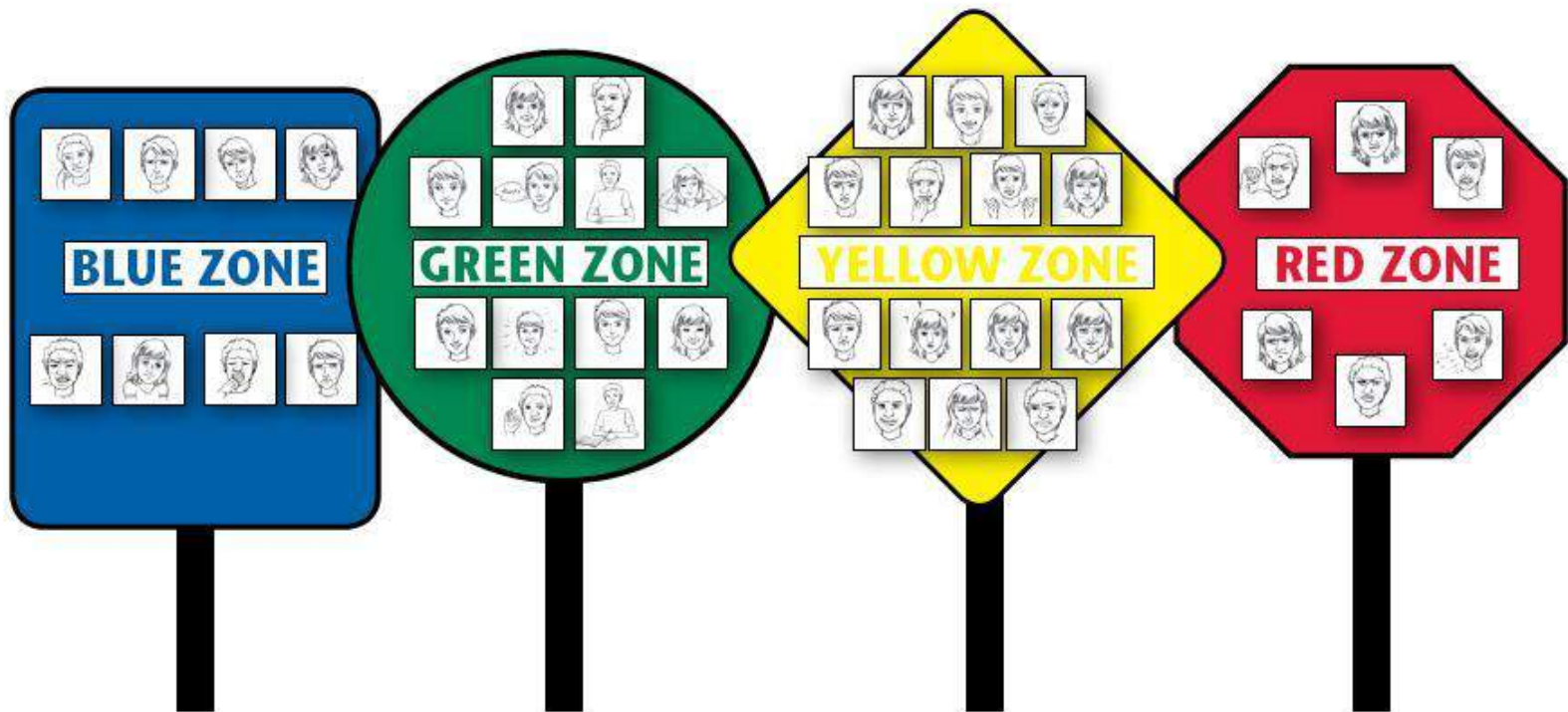
5

Very fatigued!



Tracking with use of a Social- Emotional Framework

The **ZONES** of Regulation[®]



Measurement & Monitoring

Monday	Tuesday	Wednesday	Thursday	Friday

Baseline

Measurement & Monitoring

Pro-active rest breaks:

- Collaboratively planned
- Morning and afternoon
- Twice per day
- Quiet activity

Monday	Tuesday	Wednesday	Thursday	Friday
REST	REST	REST	REST	REST
REST	REST	REST	REST	REST

Measurement & Monitoring

Before



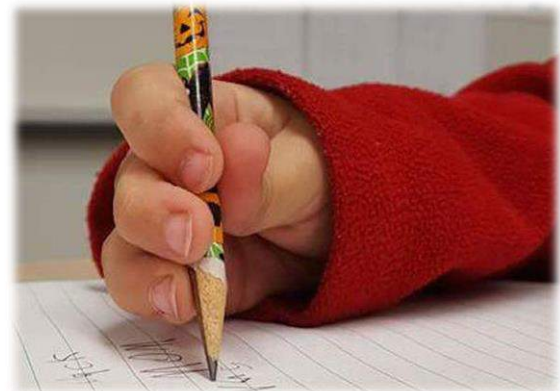
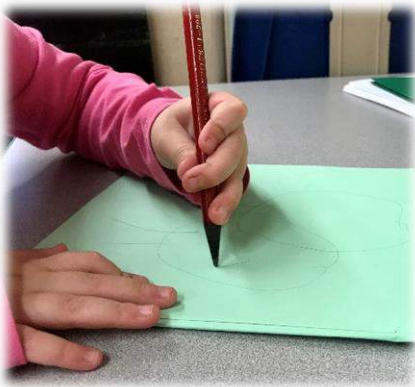
Monday	Tuesday	Wednesday	Thursday	Friday

After



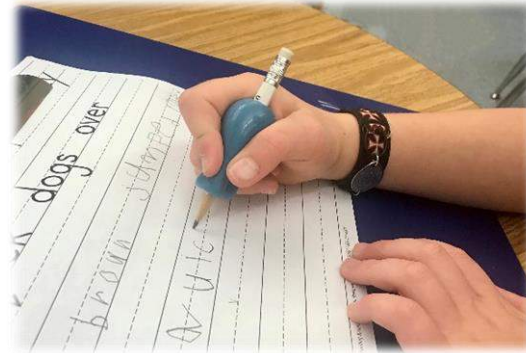
Monday	Tuesday	Wednesday	Thursday	Friday
REST	REST	REST	REST	REST
REST	REST	REST	REST	REST

3) Fine Motor: Unusual Pencil Grasps



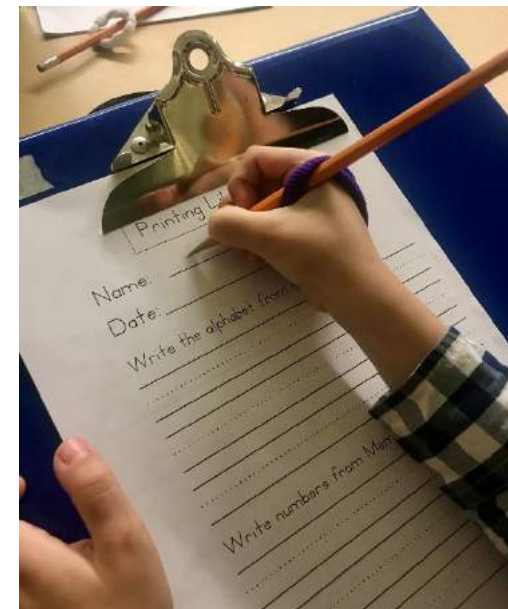
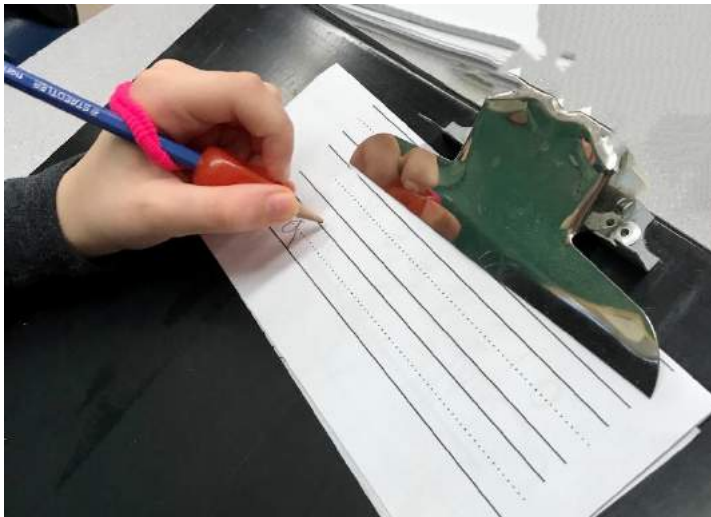
3) Fine Motor Accommodations

Pencil grips



3) Fine Motor Accommodations

- Trial use of a “pencil seat belt”
- Alone or with a pencil grips with slant board



3) Fine Motor Accommodations

- Trial using softer lead (6B or 8 B)
- Older students
- Consider use of technology



- Utilize adapted dressing/ feeding equipment (Velcro, built up grips)

3) Fine Motor Accommodations

- Use the strongest joint for the job
- Carry objects close to the body
- Carry school books in a backpack, worn using both straps to symmetrically distribute weight
- Accommodations such as added time or alternate forms of assessment may be necessary in higher grades



4) Tips for Parents

Sleep:

- Ensure enough sleep to maximize energy/endurance during the day
- Some children with hypermobility expend more energy than their peers and may require more sleep
- Tips on good sleep hygiene:
<https://www.choc.org/wp/wp-content/uploads/2016/04/Sleep-Hygiene-Children-Handout.pdf>

Tips for Parents



Physiotherapy: Consult a physiotherapist to ensure the right amount of exercise and activity

Physiotherapy Role and Rx:

- Striking a balance between getting the right amount of exercise to promote good sleep/fitness while not contributing to day time fatigue
- Exercises and activities for strength, posture, stability and endurance
- Restricting, as appropriate, contact sports or weight bearing activities on arms or wrists such as handstands and cartwheels



Leslie Russek, PT, DPT, PhD, OCS
SPEAKER

PRESENTATION

Physical Therapy for EDS



https://www.ehlers-danlos.com/wp-content/uploads/2020vsc_leslie_russek.pdf

Tips for Parents

Relaxation techniques may help in the management of pain, sleep problems, and comorbid stress or anxiety.



Additional Resources

Hypermobility information and advice

Children with **hypermobile joints** have too much movement in their joints. This can occur with just a couple of joints or all joints. A joint is the place on the body where two bones meet. Often even normal activities that put stress on loose joints will irritate them.

For some children hypermobility can cause the symptoms described below. A growth spurt, lack of exercise, illness or an accident can sometimes increase these symptoms:

Fatigue

Children may complain of a general tiredness and fatigue, because they are working very hard to maintain positions and move due to laxity in the joints. Children may also experience joint or muscle fatigue.

Pain

Children often experience joint pain, again because their joints and muscles are working harder to stabilise the joint and move throughout the day. Repetitive activities may cause pain due to muscle fatigue and should be paced and regular rest breaks scheduled.

Difficulty with activities

Children may have difficulties with pencil grip, managing clothes fastenings or manipulating objects. They may be slower to complete activities than their peers.

Poor coordination

Children may appear less coordinated and have more accidents than their peers. Injuries to joints as a result of a fall, for example, may take longer to heal.

Knowing where joints are in space

Children may have difficulties feeling where their bodies are without looking as the receptors which send this information to the brain are located in our joints.

What can I do to help?

Encourage joint protection by demonstrating adapting and practicing the following:

- Try to use larger handled objects rather than narrow ones, which will increase the tension through the hands and stress through the joints. For instance chunky pencils, fatter handled cutlery and an electric rather than standard toothbrush.
- Hold a book, plate or mug in the palms of the hands. If the child is reading for long periods use a book holder. Place iPads on a stand/holder too and use a table or desk rather than holding the tablet in the hands.
- If a child is carrying items, encourage them to make several small trips rather than carrying one very heavy item. When in secondary school it would be beneficial for young people to use a locker rather than carry all of their belongings.

Avoid keeping joints in the same position for a prolonged period of time:

- Don't give joints a chance to become stiff, this can cause more discomfort, keep them moving.
- Gentle exercise and movement throughout the day, even if in pain will help.
- When writing or doing hand work release the grip every 5-10 minutes or more for younger children.
- On long car journeys get out of the car, stretch and move around at least every hour.
- Whilst watching TV get up and move around every half hour.



Children's Occupational Therapy

Hypermobility information and advice



Balance periods of rest and activity during the day:

- Effectively managing the workload throughout the day can help avoid overworking joints.
- Encourage the child to work at a steady moderate pace and avoid rushing.
- Allow rest periods before the child becomes fatigued or sore.
- Alternate light and heavier work throughout the day.
- Take regular stretch breaks.

Strengthen:

- Encourage regular low impact strengthening activities such as swimming, children's yoga or walking. These are all helpful to improve joint strength.
- Avoid inactivity.
- Being overweight can add extra stress on joints.



Pain management:

- Heat or ice: warm baths, hot water bottles or heat packs can help relax muscles. Ice can reduce swelling.
- Distraction techniques: focusing on pain will make it feel worse so help the child to keep their mind busy with activities to distract them.
- Visit the GP if the pain isn't manageable

How can school help and support?

- When the child is at their desk ensure that feet are flat on the floor, thighs and forearms are horizontal and the desk is just below elbow height.
- Using a desk slope can further improve wrist position for writing. If children appear to be slumping in their chair a foam wedge cushion may improve this. Encourage children to sit straighter as although it is hard work, slumping will cause pain.
- If sitting on the carpet or with crossed legs is hard work allow children to sit on a chair or bench. Discourage children sitting in a 'w sit' position on their knees with their bottom between on the floor.
- Children may need to move around a lot and rather than sitting still may fidget. This will reduce the risk of stiffening up and should be allowed.
- Writing may be hard work for children with hypermobile joints. Using chunky pens may help with grip and regular rest breaks are useful to reduce pain. Completing hand warm ups before handwriting may also help.
- Have a plan to allow children to rest if they get over tired at school or in pain.



PE:

- Unless told otherwise children should participate as much as they can in PE (the curriculum may need to be differentiated).
- This will help keep joints strong to reduce pain
- Make sure they always wear supportive sports shoes with cushioned soles rather than bare feet, if ankles and knees are equally mobile.
- Avoid unnecessary high impact sports if a child is experiencing discomfort.



The **Ehlers-Danlos** Society™



Who We Are

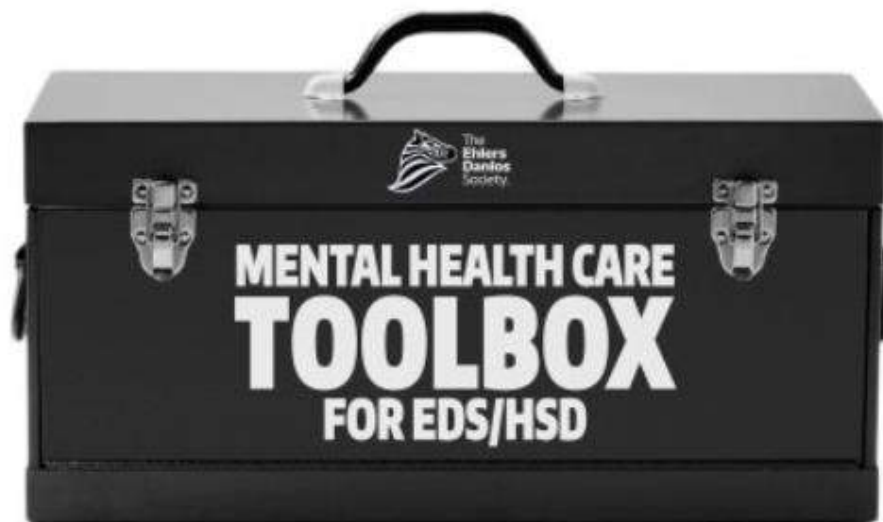
EDS & HSD Info

Community Resources

Get Involved

Professionals

MENTAL HEALTH CARE TOOLBOX FOR EDS AND HSD



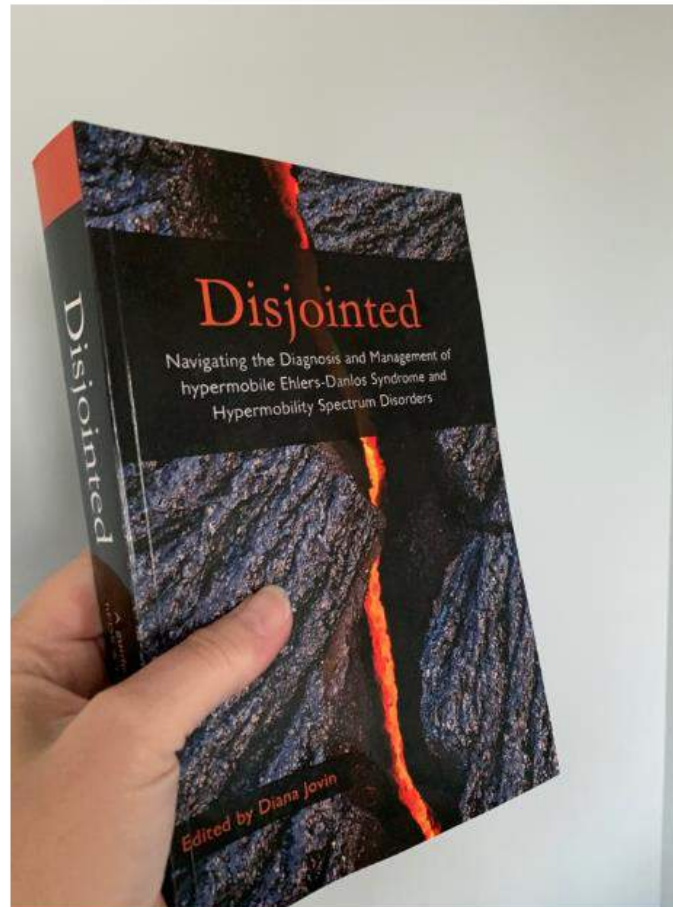
2020 VIRTUAL SUMMER CONFERENCE – DAY 3



[Watch the presentations from Day 3 of the Virtual Summer Conference 2020](#)

Resources/Scholarship

“Disjointed” is a new book for patients with hEDS/HSD and their physicians




The book is 38 chapters:

The first 5 are more about setting the context – history of EDS, patient experiences, dealing with specialists, and so forth.

Ch 6 – 31 were each written by physicians or healthcare providers. They cover, in order: genetics (2 chapters), physical therapy, TMJ dysfunction, dysautonomia, MCAS, neurology, gastrointestinal, autoimmunity, imaging, neurosurgery, anesthesiology and preoperative concerns for surgery, pain management (3 chapters), fatigue, sleep, diet, urogynecology, obstetrics, sensory processing, OT, assistive devices, psychiatry (3 chapters).

Then there are six resource chapters that address different things like parenting, education, service dogs, disability and so forth. The book includes some brief patient stories as illustrations of points being made, but it is meant to be a resource book, not a “this was my experience” book.

Contributors: With specialist chapters by: Paldeep Atwal, M.D., Richard Barnum, M.D., Linda Bluestein, M.D., Pradeep Chopra, M.D., Tania Dempsey, M.D., Shanda Dorff, M.D., Matthew Hamilton, M.D., Kristin Herman, M.D., Myles Koby, M.D., Petra Klinge, M.D. & Abby McElroy D.V.M, M.S., Anne Maitland, M.D., Andrew J. Maxwell, M.D., John Mitakides, D.D.S., Alan G. Pocinki, M.D., Lila Rosenthal, M.D., David Saperstein, M.D., Jill Schofield, M.D., Jordan Tishler, M.D., Sarah Seward Ph.D., Jennifer Varga M.P.H., R.D., Emily Block OTR/L, Nancy Block P.T, A.T.Ret., and more...



Knowledge Translation

- Advocacy and education including medical community
- Resource: <https://ehlers-danlos.com/wp-content/uploads/doctor-handout.pdf>



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The 2017 Ehlers-Danlos Syndromes International Classification

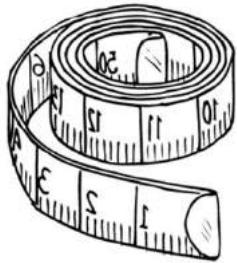
American Journal of Medical Genetics
Part C: Seminars in Medical Genetics (March 17, 2017)

Articles available free at
ehlers-danlos.com/2017-eds-international-classification

Please check out the newly released classification and management and care guidelines on the Ehlers-Danlos syndromes and related disorders, released March 17, 2017. The *American Journal of Medical Genetics* has released a supplement issue containing 18 new articles on the Ehlers-Danlos syndromes and related disorders. These papers were compiled by an International Consortium of over 90 researchers and clinicians from around the world who specialize in the Ehlers-Danlos syndromes. Topics include:

- Expanded classifications to include 13 distinct types of Ehlers-Danlos syndromes as well as the newly defined hypermobility spectrum disorders.
- New diagnostic criteria for all types with management and care guidelines.
- Information on co-morbid conditions and symptoms, including management and care guidelines.
- Guidelines for specialists in Orthopedics, Physical Therapy, Cardiology, Gastroenterology, Neurology, Immunology, Psychiatry and Psychology, Dentistry, and Pain Management.

Knowledge Translation - Research & Scholarship



Therapy/OT Intervention



POLL – KNOWLEDGE CHECK

*How knowledgeable are you **now** about OT management considerations (both broader and targeted) for joint hypermobility?*

1. Not that knowledge
2. Knowledgeable but not sure how to go about it
3. Familiar
4. Have experience with some children and youth
5. Very familiar with this population and OT management

POLL

*How important is occupational therapy for this population?
(both broader and targeted management and intervention)*

1. No opinion
2. Not that important
3. Slightly important
4. Important
5. Fairly important
6. Very important

POLL

How satisfied are you with your current knowledge and skills in occupational therapy management and intervention for this population? (both broader and targeted management and intervention)

1. No opinion
2. Not that satisfied
3. Slightly satisfied
4. Satisfied
5. Fairly satisfied
6. Very satisfied

Feedback

- ❖ One thing that surprised you
- ❖ One thing that you did not know
- ❖ One way this might change your practice



Questions or Comments

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References

An Educator's Guide (2016) Retrieved from: <https://ehlers-danlos.com/wp-content/uploads/Educator-Parent-Guide-2016.pdf>

Baeza-Velasco, C., Cohen, D., Hamonet, C., Vlamynck, E., Diaz, L., Cravero, C., ... & Guinchat, V. (2018). Autism, joint hypermobility-related disorders and pain. *Frontiers in psychiatry*, 9, 656.

Baudinette, K., Sparks, J., & Kirby, A. (2010). Survey of paediatric occupational therapists' understanding of developmental coordination disorder, joint hypermobility syndrome and attention deficit hyperactivity disorder. *British Journal of Occupational Therapy*, 73(8), 366-372.

References

Castori, M. (2013). Joint hypermobility syndrome (aka Ehlers-Danlos Syndrome, Hypermobility Type): an updated critique. *Giornale Italiano Di Dermatologia Venereologia* 148(1), 13-36.

Castori, M., & Hakim, A. (2017). Contemporary approach to joint hypermobility and related disorders. *Current Opinion in Pediatrics*, 29(6), 640-649.

Castori, M., Morlino, S., Celletti, C., Celli, M., Morrone, A., Colombi, M., ... & Grammatico, P. (2012). Management of pain and fatigue in the joint hypermobility syndrome (aka Ehlers-Danlos syndrome, hypermobility type): principles and proposal for a multidisciplinary approach. *American Journal of Medical Genetics Part A*, 158(8), 2055-2070.

Castori, M., Tinkle, B., Levy, H., Grahame, R., Malfait, F., & Hakim, A. (2017, March). A framework for the classification of joint hypermobility and related conditions. In *American Journal of Medical Genetics Part C: Seminars in Medical Genetics* (Vol. 175, No. 1, pp. 148-157).

Grahame, R., Hakim, A., Schur, P., & Romain, P. (2017). UP TO DATE Joint hypermobility syndrome. Retrieved from <https://www.uptodate.com/contents/joint-hypermobility-syndrome>

Coles, W., Copeman, A., & Davies, K. (2018). Hypermobility in children. *Paediatrics and child health*, 28(2), 50-56.

Hewlett, S., Dures, E., & Almeida, C. (2011). Measures of fatigue: Bristol Rheumatoid Arthritis Fatigue Multi-Dimensional Questionnaire (BRAf MDQ), Bristol Rheumatoid Arthritis Fatigue Numerical Rating Scales (BRAf NRS) for Severity, Effect, and Coping, Chalder Fatigue Questionnaire (CFQ), Checklist Individual Strength (CIS20R and CIS8R), Fatigue Severity Scale (FSS), Functional Assessment Chronic Illness Therapy (Fatigue)(FACIT-F), Multi-Dimensional Assessment of Fatigue (MAF), Multi-Dimensional Fatigue Inventory (MFI), Pediatric Quality Of Life *Arthritis care & research*, 63(S11), S263-S286.

Jelsma, L. D., Geuze, R. H., Klerks, M. H., Niemeijer, A. S., & Smits-Engelsman, B. C. (2013). The relationship between joint mobility and motor performance in children with and without the diagnosis of developmental coordination disorder. *BMC pediatrics*, 13(1), 1-8.

Juul-Kristensen, B., Schmedling, K., Rombaut, L., Lund, H., & Engelbert, R. H. (2017, March). Measurement properties of clinical assessment methods for classifying generalized joint hypermobility—a systematic review. In *American Journal of Medical Genetics Part C: Seminars in Medical Genetics* (Vol. 175, No. 1, pp. 116-147).

Kirby, A., & Davies, R. (2007). Developmental coordination disorder and joint hypermobility syndrome—overlapping disorders? Implications for research and clinical practice. *Child: care, health and development*, 33(5), 513-519.

Maillard, S. Stott, C., & Kemp, S. (2013). Guidelines for management of joint hypermobility syndrome in children and young people. British Society for Paediatric and Adolescent Rheumatology, 1-16.

Mu, W., Muriello, M., Clemens, J. L., Wang, Y., Smith, C. H., Tran, P. T., ... & Bodurtha, J. (2019). Factors affecting quality of life in children and adolescents with hypermobile Ehlers-Danlos syndrome/hypermobility spectrum disorders. American Journal of Medical Genetics Part A, 179(4), 561-569.

Occupational Therapy Department Hypermobility Information for parents, carers and schools. (2019, May 25). Retrieved from <https://alderhey.nhs.uk/parents-and-patients/patient-information-leaflets>

Remvig L, Jensen DV, Ward RC. Epidemiology of general joint hypermobility and basis for the proposed criteria for benign joint hypermobility syndrome: review of the literature. J Rheumatol. 2007;34:804–809.

Smith, T. O., Bacon, H., Jerman, E., Easton, V., Armon, K., Poland, F., & Macgregor, A. J. (2014). Physiotherapy and occupational therapy interventions for people with benign joint hypermobility syndrome: a systematic review of clinical trials. Disability and rehabilitation, 36(10), 797-803.

Scheper, M. C., Juul-Kristensen, B., Rombaut, L., Rameckers, E. A., Verbunt, J., & Engelbert, R. H. (2016). Disability in adolescents and adults diagnosed with hypermobility-related disorders: a meta-analysis. *Archives of physical medicine and rehabilitation*, 97(12), 2174-2187.

Varni, J. W., Burwinkle, T. M., & Szer, I. S. (2004). The PedsQL Multidimensional Fatigue Scale in pediatric rheumatology: reliability and validity. *The Journal of rheumatology*, 31(12), 2494-2500.

What is HSD? (n.d.). Retrieved from: <https://www.ehlers-danlos.com/what-is-hsd/>