Hearing Hoofbeats and Identifying Zebras

An interdisciplinary approach for the treatment of Ehlers Danlos Syndrome

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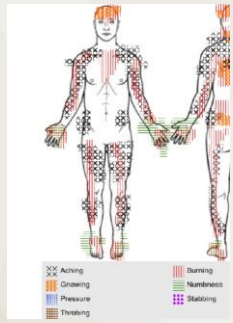
1

Vanderbilt Osher Center for Integrative Health

- High volume of clients with central sensitization diagnosis
- Health psychology
- Massage therapy
- Acupuncture
- Nursing for guidance, nutrition, bioimpedance
- Yoga therapy
- Thai Chi
- Physician trained in western and Ayurvedic medicine



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Objectives

- **Examine** characteristics of Hypermobile Ehlers Danlos Syndrome (hEDS) to promote early detection, safer execution of rehabilitation and to identify other necessary referrals.
- **Classify** treatment considerations unique to hEDS to promote successful intervention plans.
- **Judge** appropriate joint supports, adaptive equipment, and orthoses for joint protection and promote functional independence for clients with hEDS.
- **Appraise** therapeutic interventions that promote joint stability and proprioceptive neuromuscular control to assist with improving functional limitations.
- **Recommend** therapeutic and behavioral approaches to encourage empowerment and to limit fear-based behavior.

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What is EDS?

- According to The Ehlers-Danlos Society, **E**hlers-**D**anlos **s**yndromes (EDS) are a group of 13 heritable connective tissue disorders
- The conditions are caused by genetic changes that affect connective tissue
- Each type of EDS has its own set of features with distinct diagnostic criteria
- Some features are seen across all types of EDS, including joint hypermobility, skin hyperextensibility, and tissue fragility

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Type of EDS	Distinguishing Features
Hypermobile	Generalized joint hypermobility, joint instability, chronic pain
Classical	Skin fragility with extensive atrophic scarring, very stretchy skin with velvety or doughy texture
Vascular	Arterial fragility with aneurysm/dissection/rupture, organ fragility and rupture, extensive bruising, pneumothorax
Periodontal	Severe, early-onset gum disease with tooth loss pretibial plaques (discoloration of shins)
Kyphoscoliotic	Congenital/early-onset kyphoscoliosis, congenital hypotonia
Spondylodysplastic	Short stature, muscle weakness, limb bowing, craniofacial features
Brittle Cornea Syndrome	Severe problems with the cornea of the eye, hearing loss

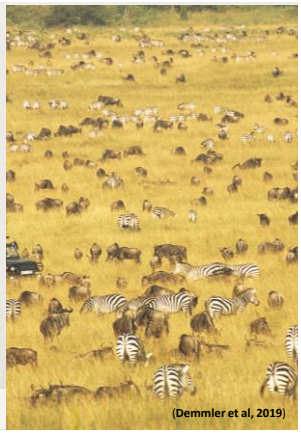
5

EDS type continued	Symptoms
Arthrochalasia	Severe joint hypermobility, congenital bilateral hip dislocation
Musculocontractural	Congenital multiple contractures, craniofacial features
Classical-like	Stretchy, velvety skin without atrophic scarring, foot deformities, leg swelling
Dermatosparaxis	Extreme skin fragility, craniofacial features, loose excessive skin, severe bruising, short limbs
Myopathic	Congenital hypotonia, proximal joint contractures
Cardiac-vulvular	Severe heart valve insufficiency

6

Incidence of hEDS

- 1 in 3100
  - Underestimate
- Men: 30%, Women: 70%
- Women more likely to be diagnosed later in life than men
- Minimal statistics regarding incidence in different ethnicities



(Demmler et al, 2019)

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Commonly Referred Diagnosis

Fibromyalgia	Postural orthostatic tachycardia syndrome (POTS)	Pelvic floor dysfunction	Chronic fatigue
Scoliosis	Delayed motor development	Joint dislocation/subluxation	Headaches/TMD
	Joint pains	Trigger points/muscle spasms	Tendon ruptures or "itis"

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(Russek 2019)

8

Associated Symptoms may include

GI dysfunction/organ prolapse	Hernias	Frequent Falls
CRPS	Frequent bruising	Raynauds
Poor tolerance to medication	Anxiety/Depression	Poor wound healing
Memory/concentration	Poor immune system	Sleep disturbance

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(Russek 2019)

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Subjective reporting

- Easily exhausted
- Clumsy
- Contortionist/party tricks
- Frequently pop joints to relieve pain
- Worsening of symptoms following a period of inactivity or stress

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Treatment response

- Nightmare disorder
- Odd unexplainable symptoms
- Standard protocols are painful
- Forget to perform HEP
- Easily confused with cues
- Frequently cancel due to GI issues, worsening pain, or fatigue (Russek 2023)

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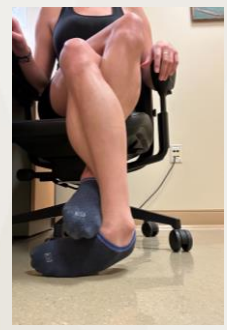
Objective/Observations

- Decreased quality of movement
- Genu recurvatum and sway back
- Hyperextend arms in quadruped
- Flat feet with medially aligned talus and everted calcaneus
- Sit with feet in chair or excessively crossed
- Point to pain with hyperextended fingers
- Visible nodules in the heel during weight bearing



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12

13

Screening tools

- Beighton Scale
- 5 Part Questionnaire
- Lower Limb Assessment Score
- Upper Limb Hypermobility Assessment Tool

14

Beighton score

With the palm of the hand and forearm resting on a flat surface with the elbow flexed at 90°, if the metacarpal-phalangeal joint of the fifth finger can be hyperextended more than 90° with respect to the dorsum of the hand, it is considered positive, scoring 1 point.

With arms outstretched forward but hand pronated, if the thumb can be passively moved to touch the ipsilateral forearm it is considered positive scoring 1 point.

With the arms outstretched to the side and hand supine, if the elbow extends more than 10°, it is considered positive scoring 1 point.

While standing, with knees locked in genu recurvatum, if the knee extends more than 10°, it is considered positive scoring 1 point.

With knees locked straight and feet together, if the patient can bend forward to place the total palm of both hands flat on the floor just in front of the feet, it is considered positive scoring 1 point.

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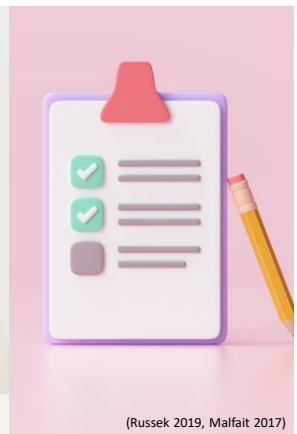
15

Beighton Score

- 6/9 for prepubertal
- 5/9 for adolescents to 50 y.o
- 4/9 for >50 y.o
- Limited to younger clients with hypermobility in the upper body more than lower body
- Beighton score alone can lead to over diagnoses in children and under diagnoses in elders
- No matter the score, the scale alone does not diagnose EDS



(Russek 2019, Malfait 2017)

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16

5 Part Questionnaire

- Can you now *(or could you ever)* place your hands flat on the floor without bending your knees?
- Can you now *(or could you ever)* bend your thumb to touch your forearm?
- As a child, did you amuse your friends by contorting your body into strange shapes or could you do the splits?
- As a child or teenager, did your kneecap or shoulder dislocate on more than one occasion?
- Do you consider yourself "double-jointed"?

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5 Part Questionnaire

- According to the EDS Society website, a yes to 2 or more questions suggests hypermobility
  - sensitivity 91%
  - specificity 75%

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18

Treatment considerations

All clients with hEDS have, or have a history of, joint hypermobility.

However; additional challenges with pain, fatigue, dysautonomia, coordination, cognitive deficits, etc. can vary significantly .

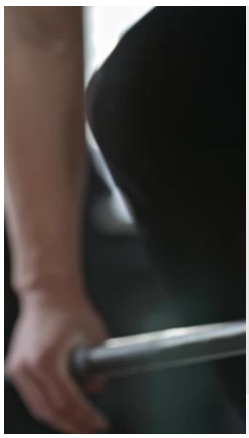
(Russek 2019)

19

Manual Therapy

- Soft tissue treatments targeting trigger points can be useful
- Joint manipulations are not recommended
- Joint mobilizations should be used with caution

20



Strengthening

- Acute pains can interfere with graded exercise progression
- Working towards muscle strength to support painful joints can help avoid fear of movement
- Strengthening progression should be slow and protect the integrity of the tendons/joints (Russek 2019)

21

Stretching

- Stretching may be appropriate to address a muscle imbalance
- No overpressure in open packed positions
- Protect the joints

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Russek 2019

22

Coordination/ Proprioception

- Clients may demonstrate decreased proprioception/balance which can contribute to frequent falls, bumping into objects such as doorways, or missing the doorknob
- Improved coordination can help decrease pain and injury

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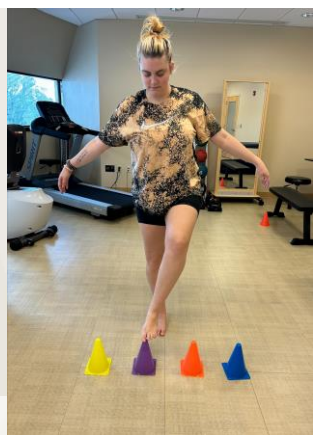

23

Barriers

- May have cognitive deficits for memory and tasks
- Few/no supporting diagnostic images correlate to symptoms
- Mild upper cervical spine instability symptoms may be common and could be muscular or neurological
- Major upper cervical spine instability is less common
- Those with hEDS may be slower to heal from surgery

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(Russek 2019/2023)

24

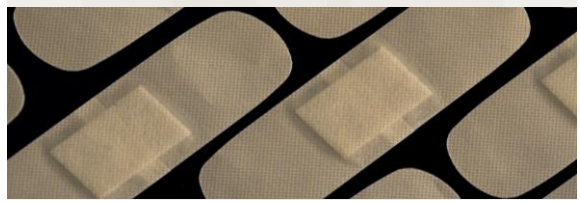
Interventions

- Hip/low back pain may be referred from pelvic floor hypertonicity
- Be sure to assess the quality of functional tasks such as rolling in bed, ascending/descending stairs, moving from the floor to standing, gait mechanics
- Caution with adhesives (such as E-stem electrodes and KT Tape) if the client suffers from mast cell activation syndrome
- Elastic taping with Thrive tape tends to be better tolerated

25

Bracing/ Assistive Devices

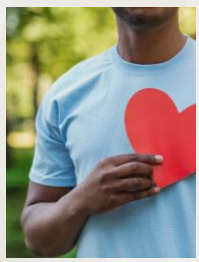
- May be helpful in short time intervals (hours to days)
- May help to limit deconditioning by empowering client to continue to move
- For severe upper cervical spine instability, a firm collar that does not irritate the TMJ may be critical for function (Russek 2023)



26

Postural Orthostatic Tachycardia Syndrome (POTS)

- A type of dysautonomia that can result in dizziness and fatigue from poorly controlled heart rate
- Incidence reported to be as high as 50% in those with hEDS (Celletti 2017)
- CHOPS or Levine protocols are prescribed and are started in a horizontal position
- Slow 8-month progression towards upright exercise
- Positional pain may interfere with performance of standard protocol
- Deep squat can be a recovery position
- Increased stress (physical or psychological) can result in POTS flare or onset



27

Pacing cycle

- Poor pacing results in a bang and bust behavior
- The behavior can result in deconditioning
- The behavior supports the chronic pain cycle

28

Pacing guidance

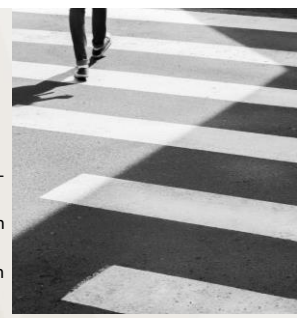
- What is the activity do you want to do more of?
- How much time in minutes can you do the activity on a good day?
- How much time can you do the activity on a difficult day?
- Add the two together
- Divide the total by 2
- Subtract a few more minutes to create a buffer
- This is how much activity that is recommended on a difficult day AND on a good day
- Once you can perform the activity regularly for 1 week, slowly begin to add minutes to the total time

(Zoffness 2020)

29

Kinesiophobia

- Excessive, irrational, debilitating fear of movement due to concern for injury or re-injury
- Effects 51-72% of patients with chronic pain
- Frequently clients will move in unusual patterns to protect from **expectation of** pain



(Perrot 2018)

30

Pain Science Education

Pain does not indicate tissue damage

Pain is a protector!

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Pain Science Education- The House Alarm Analogy

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32

Kinesiophobia treatment

- Support pain free exercise to feed a sense of safety and improve compliance
- Utilize paraphrasing to demonstrate active listening
- Teach or encourage mindfulness practice
- Encourage safe descriptions of sensations

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(Gordon 2021)

33

Our wording is important

Instead of this

- You are double jointed
- You are very weak

Try this

- You have extra motion
- Strengthening your muscles will help support your joints

34

Patient wording is also important

Instead of this

- My body is broken
- My arm is on fire

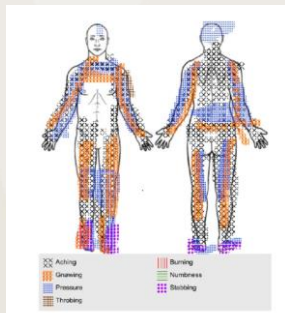
Try this

- I currently struggle with
- A sensation of heat

35

Case Study- Sarah

- 24 y/o F referred to PT for bilateral foot, hip, wrist, and hand pain
- Suspected hEDS without formal diagnoses yet
- Has POTS- takes Propanolol



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36

Sarah- Subjective

- Reports frequent knee, hip, wrist, and finger popping
- Difficulty sitting or standing for too long
- Uses wheelchair for long distances due to POTS and pain
- Works at a foster care facility in safety and security
- Completing psychology degree online
- Trouble sleeping, likes to sleep on her stomach
- Former collegiate swimmer- would like to swim again
- Enjoys bowling but has not been able to participate recently due to pain

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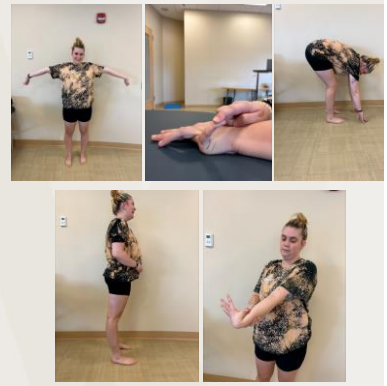
37

Sarah- Beighton Score



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38

Sarah- Objective

- Strength: Decreased posterior chain strength and poor core control
- Flexibility: decreased B hamstring, hip flexor, and upper trapezius flexibility
- Palpation: hypersensitivity to touch, significant guarding with tenderness to palpation in B upper trapezius, suboccipitals, and hip flexors
- Single leg Balance: <10 seconds with increased ankle strategy and pain
- Posture: Sway back with B knee hyperextension and anteriorly displaced pelvis, and forefoot pronation, and valgus position of calcaneus
- Squat: bilateral in-toing with dynamic genu valgus, excessive trunk forward flexion, and pain

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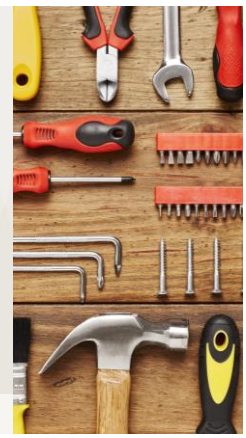
Sarah-Education

- Sleeping - switched to side lying with body pillow
- Working- talked through ergonomics and standing posture
- Exercising- new tennis shoes, pacing, fan for heat intolerance
- Self manual release- use of tennis balls



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40

With great power comes great responsibility

With great flexibility comes great responsibility

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41

Interventions- Safe Stretching

- It's OK to stretch with hypermobility, but you must pay attention to form and joint protection!
- Hamstring Stretch: seated to prevent knee hyperextension
- Hip Flexor Stretch: modified Thomas test position, making sure to monitor for lumbar hyper-lordosis
- Breathing, breathing, breathing!

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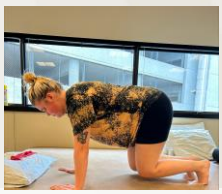
42



Interventions- Core Activation

- Starting laying down with attention to POTS symptoms as you progress
- External cues from ball press helpful
- Quadruped positioning for increased joint proprioception
  - Watch for elbow positioning!

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43

Interventions- External Cuing

- Single leg deadlift for posterior chain strengthening
- Using bench and cone for external cuing
- Mirror for visual feedback
- Shoes off to increase foot strengthening

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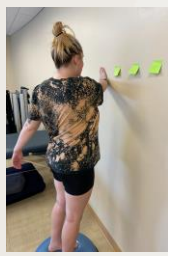
44

Interventions- Dual Tasking and Fun

- Final level of proprioceptive training
- Find fun games to divide attention between technique and tasks
- Exercise should be fun!



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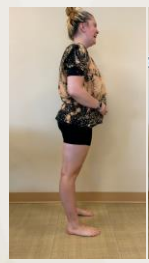
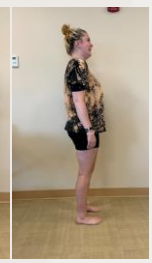
45

Where is Sarah now?

- Has done 15 visits over 8 months
- Significantly increased strength of posterior chain
- Improved postural awareness and control
- Can now balance on each leg for >20 second
- Using rollator for prolonged ambulation for POTS control
- Been swimming twice with focus on pacing
- Has re-joined a bowling league and now bowls weekly
- Plans to transition into OT next!





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46

EDS Inter-disciplinary Collaborations: Occupational Therapy Considerations

Jamie Bergner, OTD, OTR/L, CHT, COMT

47

Disclosures & Affiliations

- Vanderbilt University Medical Center
- MSOT & OTD Programs Cox College
- EndoEvolve
- MedBridge
- International Academy of Orthopedic Manual Therapy – US, Hand & Upper Extremity Track

48

Occupational Therapy

- Focus on the things you *want and need* to do in your daily life.
- OT interventions use everyday life activities (occupations) to promote health, well-being, and your ability to participate in the important activities in your life.
- Looks different for everyone!
  - taking care of yourself and your family
  - working, volunteering, going to school, among many others.

49

OT: The Evaluation

Exhibit 1. Aspects of the Occupational Therapy Domain
All aspects of the occupational therapy domain transact to support engagement, participation, and health. This exhibit does not imply a hierarchy.

Occupations	Contexts	Performance Patterns	Performance Skills	Client Factors
Activities of daily living (ADLs) Instrumental activities of daily living (IADLs) Health management Rest and sleep Education Work Play Leisure Social participation	Environmental factors Personal factors	Habits Routines Roles Rituals	Motor skills Process skills Social interaction skills	Values, beliefs, and spirituality Body functions Body structures

American Occupational Therapy Association. (2020). Occupational therapy practice framework: Domain and process (4th ed.) American Journal of Occupational Therapy, 74(Suppl.2). 7412410010. https://doi.org/10.5014/ajot.2020.7452001

50

Hand & Upper Extremity Specialty

- Specialized skills and training in considerations for the upper extremity
- Apply biomechanical principles to analyzing movement patterns with integrated knowledge of anatomy
- Adaptive experts, Integrate occupation
  - Environmental supports
  - Body supports
  - Psychological supports

Che Daud, A.Z., Yau, M.K., Barnett, F., Judd, J., Jones, R.E., & Nawawi, R.F.M. (2016) Integration of occupation-based intervention in hand injury rehabilitation: A randomized controlled trial, Journal of Hand Therapy, 29(1). 30-40. https://doi.org/10.1016/j.jht.2015.09.004.

51

Let's Look at an example!

- Who: Casey
  - 20-year-old female
  - Working fulltime in a customer service/desk job
  - Planning to go back to school in the next year
  - Roles: daughter, worker, girlfriend, friend
  - Quiet, hard worker, rarely complains, pushes through

52

Who: Systems, Comorbidities

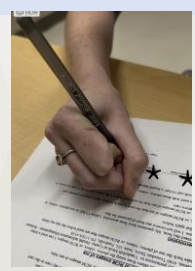
- Hypermobile, as long as she can remember
  - Hip dysplasia
  - TMJ arthritis
  - Generalized weakness
- Gastroesophageal reflux disease
- Fatigue, difficulty sleeping
- Recurrent major depressive disorder
- Generalized anxiety disorder
- Childhood trauma history

Bulbena-Cabré, A., Baeza-Velasco, C., Rosado-Figuerola, S., & Bulbena, A. Updates on the psychological and psychiatric aspects of Ehlers-Danlos syndromes and hypermobility spectrum disorders. American Journal of Medical Genetics Parc C: Seminars in Medical Genetics. 187C:482–490. https://doi.org/10.1002/ajmg.c.31955

53

What: Current Concerns

- Hands hurt after a full day of writing and typing (work)
- Hands hurt after origami (leisure)



Keer, R., & Simmonds, J. (2011). Joint protection and physical rehabilitation of the adult with hypermobility syndrome. *Curr Opin Rheumatol, 23*. DOI: 10.1097/BOR.0b013e3328342d3af

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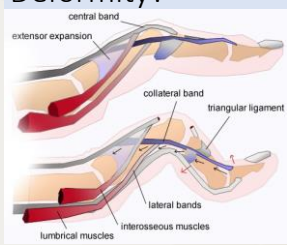
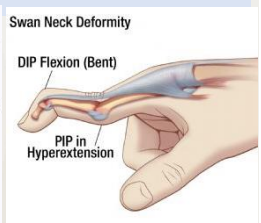
54

Boutonniere or Swan Neck Deformity?





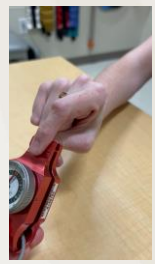
Boutonniere deformity, October 7, 2018. Kevin Lu, https://wikem.org/wiki/File:Boutonniere_deformity.jpg#file

Swan neck deformity, March 25, 2010. Allie Christian, https://www.physio-pedia.com/File:Swan-neck_deformity.jpg#filelinks

55

Or can it be both? Watch thumb MCP and IPJ under load

Tip pinch (MCP flexion, IPJ hyperextension pattern - Boutonniere)



VIDEO

Lateral pinch (MCP hyperextension, IPJ f ck)

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VIDEO

56

Let's analyze the activity - Typing

VIDEO

Photo and video credit/released to Jamie Bergner

57

Typing: This hand needs support!

- Thumbs are okay (with this task anyway)!
- DIPs hyperextend
- PIPs flex (boutonniere pattern)
- Tried immobilizing the hyperextending DIPs of digits
  - Made the PIP hyperextend into swan neck
  - Made the DIP hyperflex
- Fluctuations of extremes!
- We can't immobilize all of her!

58

Better....and Pain-free by end of workday!

PIPs:
Oval 8 splints to block hyperextension

DIPs:
Silipose gel caps with tips cut to support but not immobilize the DIPs to minimize hyperextension

VIDEO

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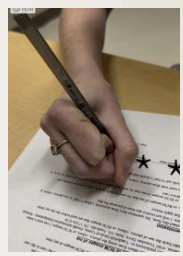
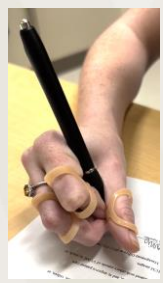
59

Repeated with writing....

Painful



Better



Best – Pain-free

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60

Leisure Participation: Origami

Painful Thumbs

VIDEO

Better

VIDEO

Pain-Free

VIDEO

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61

Are Supports....*Supported with EDS Clients?*

- Retrospective review (n=98)
- Pulled data about the following interventions:
  - Complimentary/Alternative Treatments
  - Opioids
  - NSAIDs
  - PT
  - OT
  - Muscle Relaxants
  - Neuropathic modulators
  - Steroids
  - Surgery/Procedures
  - Acetaminophen
- Separated into 3 categories: Improvement, No Effect, Worsening

Song, B.O., Yeh, P., Nguyen, D., Ikpeama, U., Epstein, M., & Harrell, J. (2020). Ehlers-Danlos syndrome: An analysis of the current treatment options. Pain Physician (23), 1533-3159. PMID: 32709178

62

Are Supports....*Supported with EDS Clients?*

- Results
  - Occupational therapy and Bracing/Splints/Orthotics were the <u>most effective</u> interventions in their entire sample
  - 70% reported improvement
  - Attributed to role in improving proprioception & joint stability
- Comparison
  - PT 43.4% but authors note this is lower in this study compared to others (63%)
  - Attributed to variability in their PT sample, recommending referral to PT knowledgeable in EDS

Song, B.O., Yeh, P., Nguyen, D., Ikpeama, U., Epstein, M., & Harrell, J. (2020). Ehlers-Danlos syndrome: An analysis of the current treatment options. Pain Physician (23), 1533-3159. PMID: 32709178

63

What about <u>*REHABILITATION*</u> in OT?

- Significantly compromised mechanoreception by attenuated ligaments
  - Reduces mechanical stability
  - Reduces protective responses to joints from diminished **afferent** functions of the ligaments
- Slow, non-reflexive **efferent** pathways
  - Goal is to facilitate/inhibit muscle activation patterns
  - Protective off-loading of joints does not happen quick enough to protect against painful subluxations/dislocations under load

64

We need to 'soup-up' our systems!

Let's Look at Some Examples.....

Balon Greyjoy, CC0, via Wikimedia Commons

65

Afferent Pathways

Mirror Box

EARLY stage:
Reduce Pain, Reframe thoughts around pain
Encourage "noticing" instead of "focusing"

Fluidotherapy

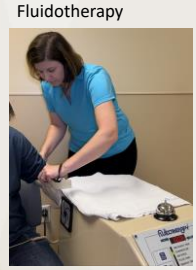

Photo and video credit/released to Jamie Bergner

Hincapie, O. L., Elkins, J. S., & Vasquez-Welsh, L. (2016). Proprioception retraining for a patient with chronic wrist pain secondary to ligament injury with no structural instability. Journal of Hand Therapy, 29(2), 183–190. https://doi.org/10.1016/j.jht.2016.03.008

66

Efferent Pathways:

Early/Mid Stage: Isometrics, Stable positions

Remember this?

NO PUTTY!

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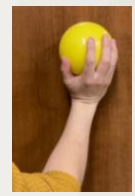
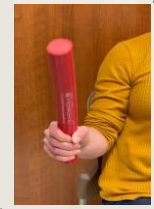
67

Afferent & Efferent Pathways: Neuromuscular Control

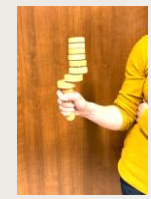
Proprioceptive Activities



Dynamic Co-contraction in functional planes (dart thrower's motion)



True Balance



Grading: Eyes Open, Eyes Closed

VIDEO

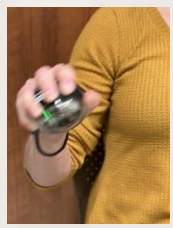
Esplugas, M., Garcia-Elias, M., Lluch, A., & Llusa Perez, M. (2016). Role of muscles in the stabilization of ligament-deficient wrists. J Hand Ther, 29(2), 166–174. https://doi.org/10.1016/j.jht.2016.03.009
Salva-Coll, Guillem, Garcia-Elias, M., & Hagert, E. (2013). Scapholunate Instability: Proprioception and Neuromuscular Control. Journal of Wrist Surgery, 02(02), 136–140. https://doi.org/10.1055/s-0033-1341960

68

Reflexive: Rapid Muscle Activation

LATE STAGE:
Powerball / Gyroscopes

Flexbar perturbations
-Self Eyes Open
-Self Eyes Closed
-Other person: Eyes Open
-Other person: Eyes Closed



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69

The rest of Casey's story....

- Casey discharged from OT to home program
- Last check in- intermittently using finger supports for task specific items like origami
- No pain at the end of the workday in the hands with intermittent use of supports based on 'noticing'

Outcomes: 6 OT visits
0/10 Pain in B UE
16% Improvement on Upper Limb Functional Index Score (ULFI)
20# increase R grip strength
12# increase in L grip strength

- Knee subluxation
- Referred to PT for dynamic strengthening
  - MD wanted to try immobilization period first
- Encouraged genetic counseling
- Connected with the EDS Society for resources
- Open door – she can get a new referral if other challenges 'pop up' or activities or demands change

70

Occupational Therapy Care

- Connect the client back to DOING their occupations (address contexts, demands of the activities, needed supports) - ***THEY*** prioritize needs
- Establish a rehab 'maintenance plan' focused on combo afferent & efferent
- Teach Self-Advocacy (support groups, resources, counseling)
- Listen first, retrain faulty thought patterns as you go
- Keep the intervention short in frequency-duration
- Be their OT for life – open door policy
- Refer as needed but guard over-medical-izing their life

71

Potential referrals

Immunology
Rheumatology
Cardiology
Gastroenterology
Genetics
Nuerology
Sleep study
Health psychology
PM&R/pain management
Psychiatry
PT/OT
Speech Therapy

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72

In Summary....

Inter-disciplinary EDS Care

- Establish goals important to the client
- Identify the barriers to achieving goals
- hEDS affects MORE than joints, refer accordingly
- Educate safe movement practice for long term management
- Reconnect client with meaningful, purposeful movement
- Listen to the patient

73

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74

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75

Helpful Online Resources

- https://www.ehlers-danlos.com/assessing-joint-hypermobility/
- https://thrivetape.com/
- https://www.zoffness.com/
- https://www.muldowneypt.com/living-life-to-the-fullest-with-ehlers-danlos-syndrome/
- https://jeanniedibon.com/
- https://www.tamethebeast.org/
- https://www.youtube.com/watch?v=YrbXg7crshs (The Pelvic Piston - How You Breathe Affects Your Pelvic Floor)

76

Helpful Books

- Disjointed: Navigating the Diagnosis and Management of hypermobile Ehlers-Danlos Syndrome and Hypermobility Spectrum Disorders edited by Diana Jovin
- Hypermobility without Tears: Moving Pain Free with Hypermobility and EDS by Jeannie Di Bon
- Living Life to the Fullest with Ehler's Danlos Syndrome by Kevin Muldowney PT

77

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78