EDS: A STRENGTH TRAINING APPROACH

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DISCLOSURES

- No financial disclosures.

BACKGROUND

- 2nd year SPT at Ohio State University
- B.S in Kinesiology Michigan State University

THE OHIO STATE UNIVERSITY

OBJECTIVES

After this presentation the learner will be able to…

- Demonstrate a basic understanding of Ehlers-Danlos syndrome and clinical signs/symptoms
- Understand how muscular physiology is altered in patients with EDS type III and its relation to hypertrophy
- Demonstrate an understanding of how to implement an individualized strength program for effective gains in patients with EDS type III/generalized hypermobility.
- Develop awareness of other interventions that may be appropriate for patients with EDS type III.
- Apply learned knowledge regarding diagnosis and strength programming to a clinical case.

SO WHAT IS EDS?

AND WHAT CAN I EXPECT?

GENERAL CHARACTERISTICS

- Heritable connective tissue disorder
- Collagen mutation
- Females > Males
- Hypermobility with cutaneous involvement, cardiovascular, gastrointestinal, and/or urogynecological symptoms

Bathen et al. 2013

COLLAGEN 101

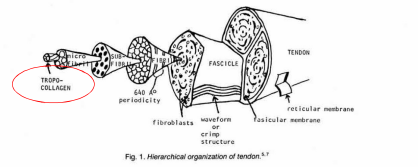


Fig. 1. *Hierarchical organization of tendon.*[5-7]

- 70% of dry weight of both ligaments and tendons
- Major structural unit of the body's connective tissues
- EDS= defect in synthesis and assembly

Hinton, 1986

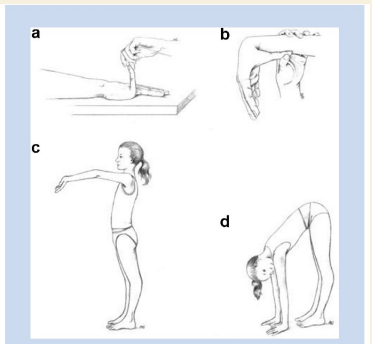
Type	Main Characteristics
Classical (Type I/II)	**Skin hyperextensibility, *widened atrophic scars*, joint hypermobility, subcutaneous spheroids, molluscoid pseudotumors**
Hypermobility (III)	**Skin hyperextensibility, smooth/velvety skin, generalized hypermobility, chronic limb/joint pain (>3 months)**
Vascular (IV)	**Thin skin, arterial/intestine/uterine fragility/rupture, extensive bruising, characteristic facial appearance (decreased adipose tissue), hypermobility at small joints, acute abdominal/flank pain**
Kyphoscoliosis (VI)	*Most severe*, joint laxity, severe muscle hypotonia at birth, scoliosis at birth and progressive, scleral fragility, bruising, tissue fragility, possible loss of ambulation
Athrochalasia (VIIA/VIIB)	Severe generalized hypermobility (recurrent dislocations), congenital bilateral hip dislocations (seen in all), tissue fragility, easy bruising, kyphoscoliosis
Dermatoparaxis (VIIC)	*Severe skin fragility*, skin that is soft, doughy, sagging, and redundant, easy bruising, large hernia

Beighton, et al. 1997

VILLEFRANCHE DIAGNOSTIC CRITERIA

- Generalized joint hypermobility (Beighton score ≥5 joints)
- Skin involvement (hyperextensibility, smooth, soft, and velvety)
- Recurring joint dislocations
- Chronic limb and joint pain (≥3 months)
- Positive family history

Beighton, et al. 1997



Bathen, et al. 2013

BEIGHTON SCORE

a) Passive DF of 5th digit joint ≥ 90˚

b) Passive apposition of thumb to flexor aspect of forearm ≥ 90˚

c) Elbow and knee hyperextension ≥ 10˚

d) Forward flex trunk with palms flat on the ground

One point for each limb at each position

Total points: 9

COMMON SYMPTOMS YOU WILL SEE

- Excessive ROM
- Pain
- Fatigue
- Decreased endurance
- Muscle atrophy
- Reports of falls/clumsiness
- Fear of movement
- Depression
- Scapular winging
- Easily bruised skin
- Reports of subluxations/dislocations
- Abnormal autonomic responses
- Other systemic involvement

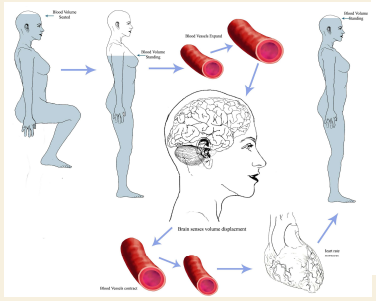
Most appear 100% healthy

Table 2. Multisystemic Nature of EDS-HT.

System	Manifestations
Cardiovascular	Aortic regurgitation, aortic root dilatation, mitral valve prolapse, mitral regurgitation, tricuspid regurgitation, Reynaud phenomenon
Autonomic Nervous System	Palpitations, dizziness, pre-syncope, syncope
Gastrointestinal	Gastroesophageal reflux, dyspepsia, gastritis, delayed gastric emptying, irritable bowel syndrome
Hematologic	Easy bruising, bleeding tendency, prolonged bleeding time, oral mucosal bruises, menometrorrhagia
Ocular	Myopia, strabismus
Gynecologic	Dysmenorrhea, menorrhagia, dyspareunia, uterine prolapse
Urologic	Constipation, fecal soiling, urinary tract infections, urinary incontinence, bladder prolapse, rectal prolapse,
Obstetric	Short labor and delivery, premature rupture of membranes, pelvic pain, varicose veins, worsening of dysautonomia during pregnancy, postpartum hemorrhage, complicated perineal wounds
Neurologic	Headache, local anesthesia failure, postural instability, increased frequency of falls, impaired proprioceptive acuity, Chiari 1 type 1
Psychiatric	Kinesiophobia, anxiety, depression

Gazit et al.2016

LETS TALK ABOUT POTS



Box 1 Diagnostic criteria for postural tachycardia syndrome (POTS)

- An increase in heart rate by ≥30 bpm on standing in adults (or ≥40 bpm in children) with no orthostatic hypotension (fall in systolic blood pressure by ≥20 mm Hg or diastolic blood pressure by ≥10 mm Hg)
- Associated symptoms that are worse with standing (light-headedness, fatigue, palpitations, syncope) and better with recumbence
- Additional causes of tachycardia excluded, including prolonged bed rest, hyperthyroidism, medications, acute blood loss
- Chronicity implies symptoms for longer than 6 months

Jones et al, 2016

THE LOWDOWN ON EDS-HT

- Most common type (HT and classic account for 90% of cases)- 1% population
- Genetic mutation encoding collagen
- 273 patients questioned, (162 HT, 45 classic, 13 other, 53 unknown)
  - 237 had one or more surgeries
  - HT reported highest VAS score for current pain, least severe pain, and most severe pain
  - 92% reported pain lasting longer than 1 year; continuous in 85%
  - 89% regularly use one or more analgesics
  - 78% had previous dislocations (severe pain correlated)
  - **214 of subjects reported pain impairing them in ADLs (SF-36)**
  - Pain mainly in neck, shoulders, hips, legs

Voermans et al. 2010

HOW SERIOUS IS THE LIMITATION?

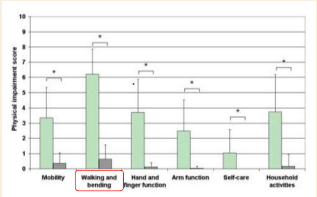


Figure 2. Physical impairment. Descriptive statistics are shown as the mean ± SD. Green bars = Ehlers-Danlos syndrome hypermobility type group; grey bars = control group; * = $P < 0.05$.

Table 4. Functional muscle performance*

Chair rise test variable	EDS-HT group	Control group	*P*
Average time per test, seconds	2.3 ± 0.96	1.3 ± 0.24	< 0.001†
Total time, seconds	8.3 ± 5.21	4.2 ± 1.48	< 0.001†

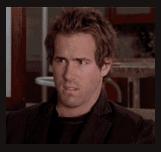
* Values are the mean ± SD. EDS-HT = Ehlers-Danlos syndrome hypermobility type.
† $P < 0.05$.

Rombaut et al. 2012

Physical therapy is considered one of the most successful, mainstay treatments for EDS-HT

Castori et al. 2012

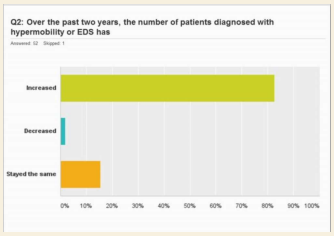
SO WHY AM I HERE?



BUT CAN PATIENTS WITH EDS STRENGTH TRAIN?! CAN THEY RUN?!

- Case example

CINCINNATI CHILDREN'S GRAND ROUNDS



12/31/16
Chris Peltier, MD, FAAP & Derek Neilson, MD

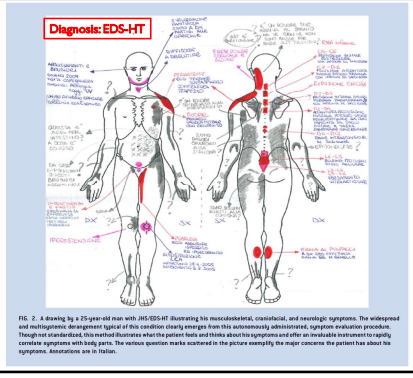


FIG. 2. A drawing by a 25-year-old man with JHS/EDS-HT illustrating his musculoskeletal, craniofacial, and neurologic symptoms. The widespread and multisystemic derangement typical of this condition clearly emerges from this autonomously administrated, symptom evaluation procedure. Though not standardized, this method illustrates what the patient feels and thinks about his symptoms and offer an invaluable instrument to rapidly correlate symptoms with body parts. The various question marks scattered in the picture exemplify the major concerns the patient has about his symptoms. Annotations are in Italian.

Castori et al. 2012

SENDING OUT AN SOS

325

96%

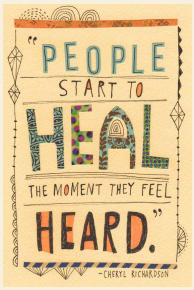
42/70

34%

Rombaut et al. 2015

PATIENT EXPERIENCE

"In general I was being told repeatedly that my troubles were 'all in my head'. Even when the doctor was relatively kind about it, it was still hard to take. When they were rude it was even worse"



Berglund et al. 2010

GOOD NEWS, WE CAN HELP!

http://ehlers-danlos.com/loose-connections/LooseConnections_2016_Spring_S.pdf

does. But I still had the fatigue and the pain was getting worse. The drug list was getting longer. I often spent days at a time in bed, avoiding any movement that would trigger my joint pain. I felt like I was an unsolvable medical mystery.

The pain got so bad that one of my doctors sent me to a physical therapist to ease my painful joints and muscles. The diagnosis was fibromyalgia syndrome and pelvic floor dysfunction. During the intake visit when the physical therapist was filling out her standard questionnaire, she looked me over and said, "You don't look like you have fibromyalgia. Has a doctor ever mentioned Ehlers-Danlos syndrome to you?" She had worked with Dr. Howard Levy's EDS patients from Johns Hopkins. I came to her as a horse; she saw a zebra.

That was when everything changed.

I began my journey to diagnosis at age fifty-seven.

WHERE WE NEED HELP

- Lack of treatment consensus
- Lack of clinical evidence for interventions
- Mixed messages between patient, physician, and PT

PHYSIOLOGY BEHIND IT ALL

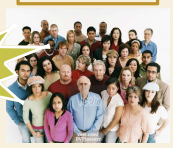
IDENTIFY IMPAIRMENTS AND WHY THEY ARE HAPPENING

JOINT HYPERMOBILITY

2-57% of population

WHY?

3.3% females 0.6% males



Scheper et al. 2015

THEORIES

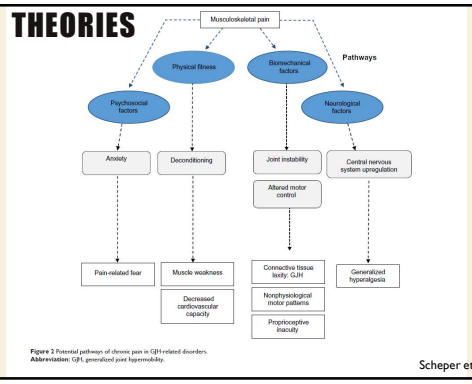


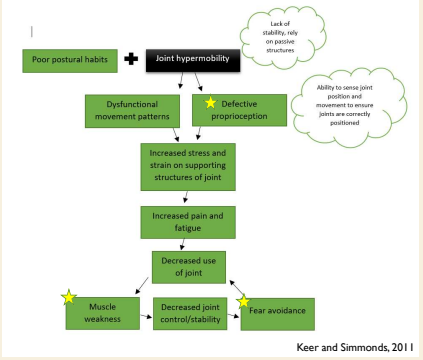
Figure 2 Potential pathways of chronic pain in GJH-related disorders.
Abbreviation: GJH, generalized joint hypermobility.

Scheper et al, 2015

LETS LOOK AT WHAT WE KNOW:

1. JOINTS ARE LOOSE
2. PATIENT IS IN PAIN
3. SOMETHING ELSE IS GOING ON

OKAY, SO LETS BREAK IT DOWN



Keer and Simmonds, 2011

DEFECTIVE PROPRIOCEPTION

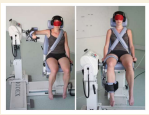


Table 2 Mean, standard deviation (SD) of the active and passive absolute errors for joint reposition test at the knee and shoulder joint of the EDS type III group and the control group

	Active		Passive	
	Mean ± SD (°) EDS type III	Mean ± SD (°) Control	Mean ± SD (°) EDS type III	Mean ± SD (°) Control
JPS 30°FL knee	**6.7±5.47**	4.0±2.67	**6.9±5.53**	4.6±4.48
JPS 60°FL knee	**5.4±4.53**	4.3±3.54	**7.4±5.70**	5.8±4.12
JPS 45° ER shoulder	6.2±6.19	5.2±3.74	6.9±4.81	6.5±3.51
JPS 75° ER shoulder	7.9±5.12	6.1±4.02	7.8±5.02	7.1±4.99

EDS type III Ehlers–Danlos syndrome type III, *JPS* joint position sense, *FL* flexion, *ER* external rotation, ° degrees of absolute error. Bold: significant between-group difference (EDS type III group versus control group) considering SI as covariate ($P<0.05$)

Commonly also seen in proximal interphalangeal joints (Ferell et al. 2004)

Rombaut et al. 2009

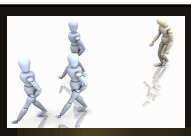
BUT WHY?

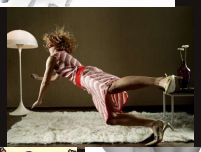
" Proprioception is defined as the ability to sense joint position and movement in order to ensure joints are correctly positioned and have suitable muscle tone for activity"

- Muscle atrophy → decreased proprioceptive sensors
- Can't generate enough mechanical force from lax joint capsule → increased activation threshold, decreased input
- Inhibited/reduced knee reflex
- Damage to joint receptors due to excessive joint mobility
- More concentration required to stabilize

Keer and Simmonds, 2011; Scheper et al. 2015

IMPLICATIONS





☆MUSCLE WEAKNESS

Table 2. Muscle strength*

	EDS-HT group	Control group	Difference, %	P
Test at 60°/second				
PT, Nm				
Knee extensors	84 ± 38.2	120 ± 26.1	−30.0	< 0.001†
Knee flexors	44 ± 20.5	69 ± 12.8	−36.2	< 0.001†
PT/BW, Nm/kg				
Knee extensors	1.2 ± 0.52	1.8 ± 0.39	−33.3	< 0.001†
Knee flexors	0.6 ± 0.26	1.0 ± 0.20	−40.0	< 0.001†
H:Q, %	53 ± 14.1	58 ± 9.2		0.115
Test at 180°/second				
PT, Nm				
Knee extensors	56 ± 26.9	79 ± 18.5	−29.2	< 0.001†
Knee flexors	31 ± 16.2	46 ± 11.7	−32.7	< 0.001†
PT/BW, Nm/kg				
Knee extensors	0.8 ± 0.36	1.2 ± 0.26	−33.3	< 0.001†
Knee flexors	0.4 ± 0.22	0.7 ± 0.18	−42.9	< 0.001†
H:Q, %	54 ± 16.3	59 ± 10.8		0.199

* Values are the mean ± SD unless otherwise indicated. EDS-HT = Ehlers-Danlos syndrome hypermobility type; difference = 100 × ([EDS-HT − control]/control); PT = peak torque; PT/BW = PT normalized for body weight; H:Q = PT hamstrings:PT quadriceps ratio.
† $P < 0.05$.

Also seen clinically…
- Deep core musculature
- Glute max/med
- Scapular stabilizers

Don't forget about muscular endurance too!

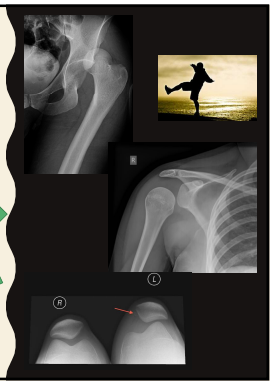
Rombaut et al. 2012

BUT WHY?

- Deconditioning (…what about athletes?)
- Altered structural integrity of connective tissue
  - Decreased force transfer
- Postural misuse
- Atrophy due to pain

Keer and Simmonds, 2011; Scheper et al. 2015

IMPLICATIONS

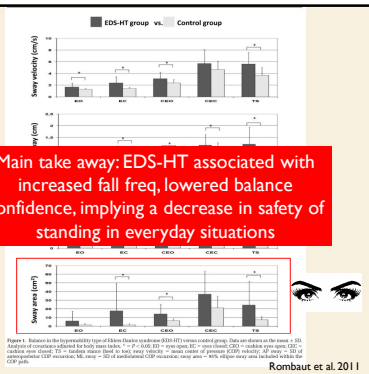


DECREASED ENDURANCE

- Rombaut et al. (2012) used the Borg scale to analyze fatigue after various muscular tests
- EDS patients showed…
  - Fatigue even before the test
  - Greater level of fatigue at every moment than control
  - Decreased ability to recover

IMPAIRED BALANCE

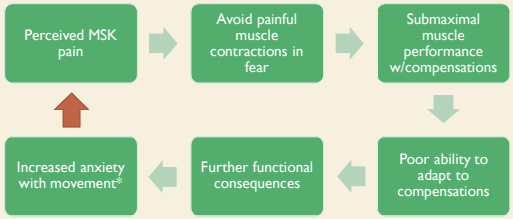
- 21 out of 22 female subjects with EDS-HT reported at least one fall in the past year
- 4= not fearful of falling
- **18= little to moderate**
- 0= very fearful



Main take away: EDS-HT associated with increased fall freq, lowered balance confidence, implying a decrease in safety of standing in everyday situations

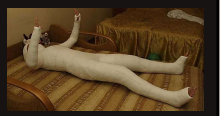
Rombaut et al. 2011

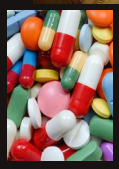
FEAR AVOIDANCE



Scheper et al. 2015

IMPLICATIONS



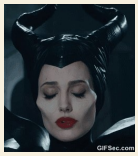


SUMMARY OF IMPAIRMENTS

Pain and fatigue
Decreased proprioception
Muscle weakness/decreased endurance
Impaired balance
Gait deviations
Fear avoidance

THERE ARE BENEFITS!

- Increased skill at specific sports/hobbies
- Shorter duration of labor
- Possible decreased risk of coronary artery disease and stroke



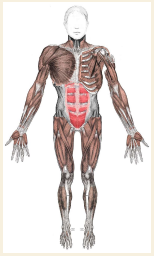
SAM'S SUBJECTIVE REPORT

NOW WHAT WE ALL CAME FOR.

HOW DO I TREAT THESE PATIENTS?!?!

CAN DEFECTIVE COLLAGEN HYPERTROPHY?

- Stiffness of patellar tendon average increase from 1795 N/mm → 2519 N/mm
- 3x/week → 48 sessions
- Connective tissue in these patients is capable of adapting to physical training



Moller et al, 2014

BUT IT DOESN'T STOP THERE

- 15 → 18 on chair rise test (average)
- Sway area decreased from 0.144m2 → 0.108m2
- CIS20 of fatigue: 68 and 33 subscale → 56, 25
- Increases in training loads LE: 31%; UE: 34% in 5-RM tests

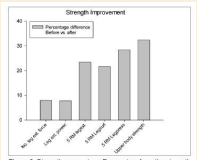


Figure 3. Strength parameters: Parameters from the strength tests are shown. The percentage differences are calculated as averages of the individual changes for the three participants. Iso. leg ext. force, Isometric leg extension force; ext., extension; 5 RM, 5 Repetition Maximum

Moller et al, 2014

WHY STRENGTH TRAIN

- 12 females: EDS-HT after 3 months of exercise showed improvements in performance of ADLs, increased muscle strength and endurance, decreased kinesiophobia, increased overall satisfaction.
- 18 subjects; JHS- 8 week physical therapy regimen: CKC and static HS exercises.

Proprioception

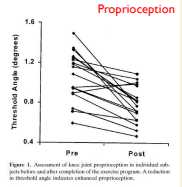


Figure 1. Assessment of knee joint proprioception in individual subjects before and after completion of the exercise program. A reduction in threshold angle indicates enhanced proprioception.

Balance

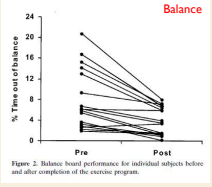


Figure 2. Balance board performance for individual subjects before and after completion of the exercise program.

Bathen, 2013; Ferrell 2004

WHY STRENGTH TRAIN

After intensive PT…

- No pain/apprehension about shoulder instability
- No further patella or hip subluxation
- 2 months later returned to swimming, throwing sports, no further complaints
- Shoulder dislocations d/t ↓HEP but no further dislocation afterwards
- Steady ↑ in strength in all muscle groups- maintained with HEP
- Nocturnal shoulder and thigh cramping disappeared
- School attendance and peer interaction ↑

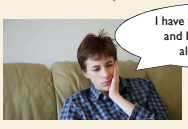
"The attending physician felt that the spontaneous remission of the pt's symptoms was remote and that the exercise program was chiefly responsible for increases in functional joint stability"

Hinton, 1984

SPORT-SPECIFIC RETURN

16 year old, JHS

17 weeks of magic



Simmonds and Keer, 2008

HOW DID THEY DO THAT?

Initial
- Posture
- Lumbar control
- PNF, taping, mirror cueing
- SIMPLE

Middle
- CKC circuit
- Pain-free range
- Gradually increase reps
- Endurance

Late
- Sport specific
- Running, cutting
- LE plyo
- Racquet activities on minitramp

Simmonds and Keer, 2008

Exercise programs are not associated with the side effects common to some pharmacologic interventions and they also empower patients to manage their own condition

Ferrell, 2004

COMPLIANCE

Patient 10: "I think that every time that we are supported or guided, like in physical therapy, like the osteopath…We can do things better. Because when you're alone you're scared! You're scared of getting hurt; you don't know what needs to be done. In the end, that's what made me quit doing the moves."

TOO COMPLEX

Patient 6: "We almost have 10 exercises. It's too much. There should be a limit: 3 or 4 max."

TOO MUCH

Patient 11: "I have my rehabilitation sheets that I really struggle to follow every day, because it's so boring!"

BORING

Patient 3: "It's like being fed up. I can't spend my whole life doing this all the time…When am I getting better?" This feeling could be worse for patients who experienced a resurgence of symptoms despite good adherence.

UNEDUCATED/UNMOTIVATED

Palazzo et al. 2016

SOLUTIONS TO THINK ABOUT

Palazzo et al. 2016

Patient 21: "Renewing the exercises, for me it's a good thing, because if you put a little bit of change, that makes it more enjoyable…From the moment you start a new exercise, it will stimulate you."

UNIQUE

Patient 2: "A video, that would be good really…that would be perfect…it's a simulation straight from the rehabilitation department." "A real person practicing exercises! Imitate and follow!…it's better with images because you mimic."

VIDEO DEMOS

More detailed explanations on the disease, the objectives of exercises, and the choice of exercises included in the program were cited as needed; the need for individualized advice to integrate exercises into daily life was also noted.

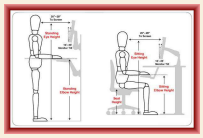
EDUCATION

Patient 27 "To send a spreadsheet at the end of the week saying what I've done, by email or some stuff like that, that's something that could motivate me."

ACCOUNTABILITY

EDUCATION

- Activity modification
- Importance of joint neutral and avoidance of end ranges
- Watch your language used- be careful not to discourage
- Low impact activities **until increase in strength/stability**
- Use of ADs and other resources
- Importance of posture and optimal positioning
- Pain management



DrScholl's

EDUCATION

TABLE VIII. Lifestyle Recommendations Concerning Pain and Fatigue in JHS/EDS-HT

Recommendation
Promote regular, aerobic fitness
Promote fitness support with strengthening, gentle stretching and proprioception exercises
Promote postural and ergonomic hygiene especially during sleep, at school and workplace
Promote weight control (BMI <25)
Promote daily relaxation activities
Promote lubrication during sexual intercourse (women)
Promote assumption of generous isotonic liquid intake (2–2.5 L/day)
Promote assumption of high salt intake (avoided in case of arterial hypertension)
Promote early treatment of malocclusion
Avoid high impact sports/activities
Avoid low environmental temperatures
Avoid prolonged sitting positions and prolonged recumbency
Avoid sudden head-up postural change
Avoid excessive weight lifting/carrying
Avoid large meals (especially of refined carbohydrates)
Avoid hard foods intake and excessive jaw movements (ice, gums, etc.)
Avoid bladder irritant foods (e.g., coffee and citrus products) intake
Avoid nicotine and alcohol intake

Note: These recommendations are based on authors' experience as well as basic pathophysiologic principles related to laxity of the connective tissue.

Castori, 2012

KT TAPING

- Trial and error but patients typically have a **positive** response
- Be careful with skin reactions (milk of magnesia?)
- Postural cues
- Knee stability
  - Case study showed improvements in gait biomechanics and reduction in knee pain (Camerota et al, 2015)
- Enhances sensory input to skin to help with proprioception (Keer and Simmonds, 2011)
- Not much luck for fallen arches clinically
- Can supplement strength training with cueing of optimal positioning

Our goal is to give the patient knowledge and tools to empower them to manage their impairments and pain LONG TERM through ACTIVE treatments.

CREATING AN INDIVIDUALIZED PROGRAM

SPECIFIC INTERVENTIONS

THINGS TO KEEP IN MIND

- Start at low/moderate intensity
- Supervision is key before becoming HEP
- Important to explain reasoning behind exercises
- Train patterns not just isolated movements early on
- Focus on stability and control
- Educate the slower progression
- Motivation is key
- Avoid end-ranges, stay in neutral
- GET CREATIVE
- Choose appropriate outcome measure and goals
- Start with full-body/multi-joint 2-3x/wk
- Stretching conservatively

STRENGTH TRAINING DOESN'T JUST MEAN DUMBBELLS!

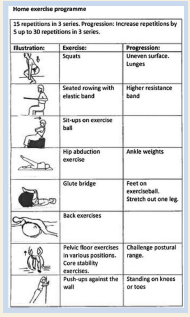
REP SCHEMES AND FREQUENCY

Table 2. Targeted progression in the lower body protocol.

Session	1-12	13-16	17-24	25-32	33-39	40-45	46-48
Nr. of sets	3	3	4	4	5	5	3*
Repetitions	10,10,10	10,9,8	10,9,8,7	9,8,7,6	10,9,8,7,6	9,8,7,6,5	6,6,6

* In the last sessions a tapering period were carried out with the same weight but lower number of sets and reps. The tapering period in the last 3 sessions were introduced to allow for the anabolic response.

- Bodyweight exercises may need more reps.
- Don't push the envelope too early; buy-in (50% of 10-RM)
- Healthy adults: low-load training to failure can effectively induce hypertrophy in untrained
  - Low rep training (1-5RM) enhances neuromuscular adaptations
  - High repetitions (15+) can help delay fatigue and increase time under load



Home exercise programme

15 repetitions in 3 series. Progression: Increase repetitions by 5 up to 30 repetitions in 3 series.

Illustration:	Exercise:	Progression:
	Squats	Uneven surface. Lunges
	Seated rowing with elastic band	Higher resistance band
	Sit-ups on exercise ball	
	Hip abduction exercise	Ankle weights
	Glute bridge	Feet on exerciseball. Stretch out one leg.
	Back exercises	
	Pelvic floor exercises in various positions. Core stability exercises.	Challenge postural range.
	Push-ups against the wall	Standing on knees or toes

Bathen et al. 2013; Moeller et al. 2014; Schoenfeld 2013

OVERALL STRENGTHENING

OKC vs. CKC

- ★Need to add ext resistance
- ★Stressful to joints
- ★Doesn't facilitate proprioception as well

- ★Less strain on knee ligaments
- ★Increases stability and co-contraction
- ★Increases proprioception through WB
- ★Body weight resistance

Ferrell et al. 2004 and Keer and Simmonds, 2011

GENERAL OR TARGETED PROGRAM?

- First RCT conducted in 2010 on children with BJHS and PT

Generalized Exercise n=17
- Shuttle runs
- Bunny hops
- Squat thrusts
- Sit to stands
- Step ups
- Star jumps
- 6 weekly apts x 30min + HEP

Targeted Exercise n=15
- Control joint neutral (optimal alignment)
- Re-train dynamic control (specific muscles)
- Motion control through entire ROM
- Specific tissue lengthening
- 6 weekly apts x 30min + HEP

The results:
- No sig difference between groups, child's assessment of pain ↓ in both (-30.64 and -21.23)
- Sig difference in parent's global assessment of pain in favor of targeted (3.7 and -17.59)
- CHAQ both decreased; shuttle run both increased- not sig
- Conclusion: both are effective; combine them

Kemp et al. 2010

IDEA OF METABOLIC STRESS

- Induced mechanical stress is critical stimulus for hypertrophy
- Review article: Exercise induced metabolic stress can maximize muscle development (Schoenfeld, 2013)
- Goal is to ↑ training volume without ↑ stress to the joint

MAY NOT BE APPROPRIATE FOR EARLY PATIENTS WITH EDS, assess response to resistance training before initiating gradual inclusion.

CREATING METABOLIC STRESS

Drop sets:
↑ volume without ↑ load

the weight by 20% for 3-4x OR change the angle without changing weight

Resistance bands/mid set isometrics:

What is it? 5-8s isometric holds with rowing, pressing, and pulling movements

Giant sets:
Stress same muscle group

as you can for all three movements

Use clinical judgment about gradual inclusion in very late stages with EDS patients!

https://drjohnrusin.com/maximizing-metabolic-stress-with-intensity-techniques-for-hypertrophy/

EXAMPLES OF GIANT SETS:

- **Quads:** Heel Elevated Front Squats, Unilaterally Loaded Split Squats, Wall sits
- **Hamstrings:** Dumbbell RDLs, Glute Bridges, Unilateral Lying Leg Curls
- **Biceps:** EZ Bar Curls, Reverse Barbell Curls, Cross Body Hammer Curls
- **Triceps:** Dips, DB Pullovers, Skullcrushers
- **Chest:** Banded Incline Hammer Press or Banded Incline Dumbbell Press, Incline Cable Flies, Push-Ups
- **Back:** Pull-Up Negatives, Straight Arm Pulldowns, T-Bar Rows

https://drjohnrusin.com/maximizing-metabolic-stress-with-intensity-techniques-for-hypertrophy/

PROGRAMMING

1. Warm-up (~3 exercises/5)
2. Accessory
3. Foundational Movement
4. Accessory
5. Accessory (*optional early on*)
6. Aerobic (*optional*)

Squat, hip hinge, lunge, upper body push, pull

WARM UP IDEAS

https://www.youtube.com/watch?v=xOtxpkdujsM – Justin Ocha

- Bridging → activate posterior chain
- Banded rowing → horizontal pulling before vertical to activate scapular stabilizers
- Slo-mo air squats → hold at the bottom, show control
- Bird dogs → coordinate core control with dynamic movement

- Mobility things depending on patient deficits:
  - Lateral lunge for adductors
  - Front lunge with palms on ground for hip flexors

PATIENT CASE

TIME TO TREAT SAM

SAM'S SUBJECTIVE REPORT

REMINDER

Add-in known POTS dx

"Even thinking about running causes a little bit of pain"

Has given up most physical activity, used to enjoy running, 6 mo since last run; usually ran ~5miles

Muscle atrophy and decreased endurance

Wearing of braces, frequent stretching

Finger dislocations, generalized joint pain

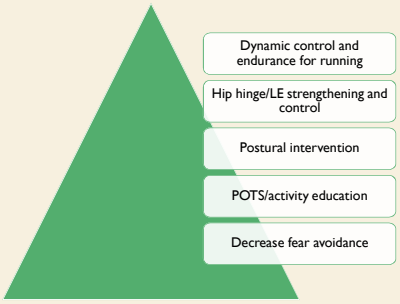
L knee pain w/walking and stairs, better with rest

EDUCATION

Activities	Hips	Knees	Ankles	Shoulders	Hands
Walking	•	•	•		
Swimming				•	•
Running	••	•••	•••		
Rowing	•	••	•	••	••
Climbing stairs	•••	•••	•		
Cycling (sta)	•	••	•		
Tennis/ Racquet	•••	•••	•••	•••	•••
Low impact aerobics	••	••	••	•	

The Christ Hospital Health Network

WHAT WE NEED TO ADDRESS



DECREASING FEAR AVOIDANCE

Responses highly dependent on the patient

Figure out their interests, motivation

Show them pain-free exercise

Use success stories, research, support groups, etc.

Gain their trust as early as you can

Educate the patient on importance of muscular support

POTS EDUCATION

- No cure- but no reported deaths.
- Non-pharm treatments should PRECEDE meds
  - Increase daily fluid (3L) and dietary salt (8-10g) intake
  - Waist high compression stockings
  - **Exercise has repeatedly been shown to improve symptoms in POTS**
    - **3 mo, decreased orthostatic tachycardia, reduced symptom burden, and increased QoL**
    - **Slow, graded program; primarily aerobic and some leg resistance (rowing, swimming)**
    - **30 min/every other day (5d/wk)**
    - **May initially feel worse**

Jones et al. 2016

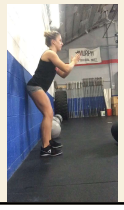
POSTURAL INTERVENTION

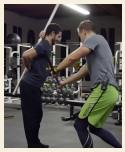
- Chat about: lumbar roll, studying at a desk, KT tape, frequent breaks
- Take pictures or use mirrors due to decreased proprioception
- T-spine mobility: foam roller, quadruped
- TELL THEM WHY

TEACHING THE HIP HINGE

- **Learn the pattern, then add resistance**
- "In order to gain access to high quality frontal plane power [agility] and stability, hip extension is a key potentiator" – Craig Liebenson, D.C

FOCUS ON CONTROL





WHAT IF SHE ASKS WHEN SHE CAN RETURN TO RUNNING?

OSU RETURN TO RUN PROTOCOL

- Ideal for athletes with non-surgical injuries post-surgical patients following Alter G or Deep Water Running Progression and runners that average 20-40 miles per week
- This guideline is intended for end stage rehabilitation return to running and the clinician should use their own clinical judgement when it is safe to return the athlete to plyometrics and running
- Supplement with Alter G and Deep Water Running Progression for post-surgical patients

Phase I: Walking and Plyometrics

Criteria to Start Phase I	• Ability to walk 30 minutes pain-free • Full joint range of motion • At least 80% strength compared to the unaffected limb (specifically post-surgical injuries) • Trace to no edema present
Goals	• Tolerate single leg impact activities • Demonstrate proper lower extremity biomechanics • Walking without limitations • Demonstrate equal quality and power bilaterally
Guidelines	• Double limb jumps progressed to single limb hops • Unilateral to multi-directional plane hops

Sample Functional Hop Progression	
Double Leg	**Single Leg**
Hop in Place	Hop in Place
Forward Hop	Forward Hop
Backward Hop	Backward Hop
Triple Hop	Triple Hop
Side-to-Side Hop	Side-to-Side Hop
Crossover	Cross-over
Scissor Hops	Dot Drills
Dot Drills	Lateral Bounds/Skaters
180 Degree Hops	90 Degree Hops

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https://wexnermedical.osu.edu/~/media/Files/WexnerMedical/Patient-Care/Healthcare-Services/Sports-Medicine/Education/Medical-Professionals/Return-to-Running/IntermediateReturnToRunning.pdf?la=en

STAGE 1- MIDRANGE GENERAL STRENGTHENING

Exercise	Reps	Sets
90-90 breathing	5	2
Thoracic rotation quadruped	8	4
Goblet box squats	8	2
Banded pull-through	10	2
Bear position static hold	15s	3

Encourage rowing and cycling for aerobic exercise

- Use the 30s chair rise test once you feel that she can properly due a hip hinge with limited cueing.
  - Take subjective report of pain into account
  - Focus on quality of movement
- Progress exercises to stage 2 if there is no increase in pain with these exercises AND improved quality
  - Look at ability to complete 10-15 reps **pain free**

ASSESS/ REASSESS

STAGE 2-FLIRT WITH END RANGE, STABILITY

Exercise	Reps	Sets
Monster walk with throw	15	4
Forward-reverse lunge	6	4 (2 each side)
KB bottoms up deadbug	8	3
Side to side step downs	12	3

Alternative:

Exercise	Reps	Sets
Lateral SL throws	10	4 (2 each side)
KB deadlift	10	3
Banded unstable row	15	3
Bird Dog	12	3

- Core stability
- Bottoms up KB rhythmic stabilization for time
- Single limb squat progress
  - Take subjective report of pain into account
  - Focus on quality
- Progress exercises to stage 3 if there is no increase in pain with these exercises and she shows adequate CONTROL

ASSESS/ REASSESS

OSU RETURN TO RUN PROTOCOL

Most likely too early

Phase II: Walk to Run Progression

Criteria to Start Phase II	• Athlete is able to tolerate 200-250 foot contacts ○ Athlete is able to tolerate number of foot contacts for ~1/3rd of a mile of running • No symptoms reported by the patient and demonstrates adequate plyometric form with minimal to no knee valgus, toe to heel landing, no trunk lean, and demonstrates soft landing • Ability to perform 15 heel taps with proper LE mechanics
Goals	• Progression back to continuous running without aggravation of symptoms and antalgia
Guidelines	• Prior to walk to run progression complete 5 minute dynamic warm-up (example at wexnermedical.osu.edu/sports-medicine/treatments/endurance-medicine) • Athlete must take at least one running off day in between each return to running workout, non-impact cross training during off days • Take at least one complete rest day a week • If athlete develops pain, return of other symptoms, or cannot complete the phase they remain at that phase until they are able to complete it without symptoms • Complete only one phase per day

Intermediate Walk to Run Program	Warm-up	Run:Walk	Repetitions	Cool down	Total	Days
Phase 1	5-10 min	2 min:1-2 min	2-4	5-10 min	20-30 min	2
Phase 2	5-10 min	4 min:1-2 min	2-4	5-10 min	25-35 min	2
Phase 3	5-10 min	6 min:1-2 min	2-4	5-10 min	30-40 min	2
Phase 4	5-8 min	8 min:1-2 min	2-4	5-8 min	35-45 min	2

https://wexnermedical.osu.edu/~/media/Files/WexnerMedical/Patient-Care/Healthcare-Services/Sports-Medicine/Education/Medical-Professionals/Return-to-Running/IntermediateReturnToRunning.pdf?la=en

STAGE 3- ISOLATED STRENGTH, ENDURANCE, POWER

	Exercise	Reps	Sets
Superset	Banded hip march	45s	4
Superset	Pallof press	12	4
	SL KB deadlift (stop)	10	4
	Max wall ball toss for time (lbs is clinical decision)	1 min	2

Alternative:

	Exercise	Reps	Sets
	Bear crawl for distance	Max	2
	Banded hip thrusts for power	15	3
Superset	Landmine lunge march	5-8	4
Superset	Box double/single leg	10	4

IS SHE READY?

What else do we need to consider?

Intermediate Walk to Run Program	Warm-up	Run:Walk	Repetitions	Cool down	Total	Days
Phase 1	5-10 min	2 min:1-2 min	2-4	5-10 min	20-30 min	2
Phase 2	5-10 min	4 min:1-2 min	2-4	5-10 min	25-35 min	2
Phase 3	5-10 min	6 min:1-2 min	2-4	5-10 min	30-40 min	2
Phase 4	5-8 min	8 min:1-2 min	2-4	5-8 min	35-45 min	2

What else do we need to assess?

RUNNING ASSESSMENT

Foot contact

Knee flexion angle at IC

Vertical displacement

Stride length

Cadence

Torso angle

- **Check progress towards goals**
  - Look at quality and quantity of SLS
  - Landing mechanics with hop-downs
  - Re-assess segmental rolling and KB stabilization

Education on long-term maintenance and collaborative planning

- How is she going to keep moving?
- Tools she needs
- Understanding of proper exercise

ASSESS/ REASSESS

OSU RETURN TO RUN PROTOCOL

This progression is not a cookie cutter for every patient!!

Phase III: Running Progression

Criteria to Start Phase III	• Able to complete Phase II without pain or symptoms • At least 90% strength and Limb Symmetry Index compared to the unaffected limb (specifically post-surgical injuries) • Ability to perform 12 inch hop downs from box with proper LE mechanics
Goals	• Increase daily and weekly mileage gradually • Return to normal running routine within 5 weeks • No return of pain or symptoms
Guidelines	• Athlete can cross train or rest on off days, but must take at least one rest day a week • Prior to run progression complete 5 minute dynamic warm-up and 5-10 min walking warm-up • After run complete 5-10 min walking cool down and post-run stretch

Intermediate Running Progression	Day 1	Day 2	Day 3	Day 4	Day 5	Day 6	Day 7	Total Miles
Week 1	-	1	-	2	-	2	-	5
Week 2	-	2	-	3	-	4	-	9
Week 3	-	4	-	3	-	4	-	11
Week 4	5	-	4	3	-	5	-	17
Week 5	5	-	6	4	-	5	-	20

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Wexner Medical Center

LONG-TERM

- Its all about empowerment and education
- Maintenance of this strength is CRUCIAL
- CROSS TRAIN, CROSS TRAIN, CROSS TRAIN
- Dumbbells over barbells → stability
- CAUTION w/dynamic movements-
  - NONE with weights- use med balls or body weight
  - No push press, OH squats, oly lifting, kipping movements, handstands unless your clinical judgement allows SAFETY with these movements
- Build your own program resource

WHAT IF I DON'T HAVE TIME TO WRITE PROGRAMS FOR EACH PATIENT?

You don't have to!

Day one	Day two	Day three	Day four	Day five	Day six
• 1 major • 2-3 acc • 1 core					

HOME EXERCISE PROGRAM

EXAMPLE

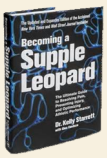
Push	Pull	UE accessory
• DB bench press • Push up • Banded sled push • Lunge to cable chest press • DB Shoulder press	• Bent over row • Single arm DB row • Inverted row • Face pull downs • Banded shoulder extension • Prone row	• Rear delt raise • Bicep curls • Tricep pull downs (NO DIPS) • Horizontal band pull aparts • Bottoms up KB

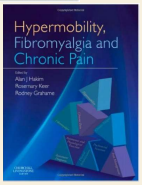
Hip Hinge	Single leg	LE accessory
• Deadlift • Goblet squat • KB sumo squat • Hip thrust • Goblet pause squats • DB Box squats	• Single leg deadlift • Front lunge • Reverse lunge • Step ups • Heel taps • Lateral lunge • Front to reverse lunge	• Lateral side to side • Banded lateral walks/Side steps • Single leg bridges • Nordic HS curls • HS rollouts

Core stability	Aerobic
• Side plank • Pallof press • Single leg press • bear crawl • deadbug • single leg raise with tension	• Rowing • Cycling • Elliptical • Walking (incline?)

OTHERS

- http://ehlers-danlos.com/
  Support groups, resources for professionals/patients, events, research
- http://hypermobility.org
- Postural aids: intelliskin.net, str8-n-up
- http://www.otpbooks.com/
  Free articles, discounted products
- http://www.thebarbellphysio.com/
- https://drjohnrusin.com/
- https://mikereinold.com/





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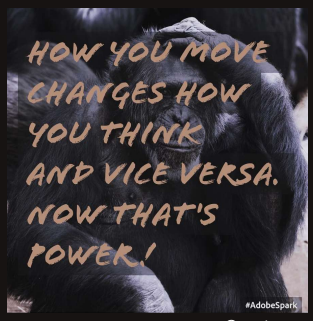

@stopchasingpain

REFERENCES

- Scheper MC, de Vries JE,Verbunt J, Engelbert RHH. Chronic pain in hypermobility syndrome and Ehlers-Danlos syndrome (hypermobility type): it is a challenge. *J Pain Res* 2015;8:591-601.
- Scheper M, Rombaut L, de Vries J, De Wandele I, van der Esch M,Visser B, Malfait F, Calders P, Englebert R. The association between muscle strength and activity limitations in patients with the hypermobility type of ehlers-danlos syndrome: the impact of proprioception. *Disabil Rehabil* 2016: 1-7.
- Palazzo C, et al. Barriers to home-based exercise program adherence with chronic low back pain: Patient expectations regarding new technologies. *Ann Phys Rehabil Med.* 2016;59(2):107-113.
- Hinton RY. Case study: Rehabilitation of multiple joint instability associated with ehlers-danlos syndrome. *JOSPT.* 1986;8(4): 193-198.
- Gazit Y, Jacob G, Grahame R. Ehlers-Danlos syndrome-hypermobility type: a much neglected multisystemic disorder. *Rambam Maimonides Med J.* 2016;7(4).
- Jones PK, Shaw BH, Raj SR. Clinical challenges in the diagnosis and management of postural tachycardia syndrome. *Pract Neurol.* 2016;16:431-438.
- Long Z, Spencer J.The Best Kettlebell Rehab Exercises. The Barbell Physio. http://www.thebarbellphysio.com/kettlebell-rehab-exercises. Accessed November 26, 2016.
- Moller MB, Kjaer M, Svensson RB, Andersen JL, Magnusson SP, Nielsen RH. Functional adaption of tendon and skeletal muscle to resistance training in three patients with genetically verified classic Ehlers Danlos Syndrome. Muscles Ligaments Tendons J. 2014;4(3): 315-323.
- Voermans NC, Knoop H, Bleijenberg G, van Engelen BG. Fatigue associated with muscle weakness in Ehlers-Danlos syndrome: an explorative study. Physiotherapy 2011;97(2):170-174.
- Kemp S, et al. A randomized comparative trial of generalized vs targeted physiotherapy in the management of childhood hypermobility. Rheumatology. 2010;49(2):315-325.
- Camerota F, et al. The effects of neuromuscular taping on gait walking strategy in a patient with joint hypermobility syndrome/Ehlers-Danlos syndrome hypermobility type. Ther Adv Musculoskeletal Dis 2015;7(11): 3-10.
- Ferrell WR, et al. Amelioration of symptoms by enhancement of proprioception in patients with joint hypermobility syndrome. Arthritis Rheum. 2004;50(10):3323-3328.
- Shoenfeld, BJ. Potential mechanisms for a role of metabolic stress in hypertrophic adaptations to resistance training. Sports Med. 2013;43:179-194.
- Morales-Artacho AJ, Lacourpaille L, and Guilhem G. Effects of warm-up on hamstring muscles stiffness: Cycling vs foam rolling. *Scand J Med Sci Sports.* 2017;00:1–11

REFERENCES

- Simmonds JV, Keer RJ. Hypermobnilty and hypermobility syndrome, part 2: assessment and management of hypermobility syndrome: illustrated via case studies. *Man Ther.* 2008;13(2):e1-11.