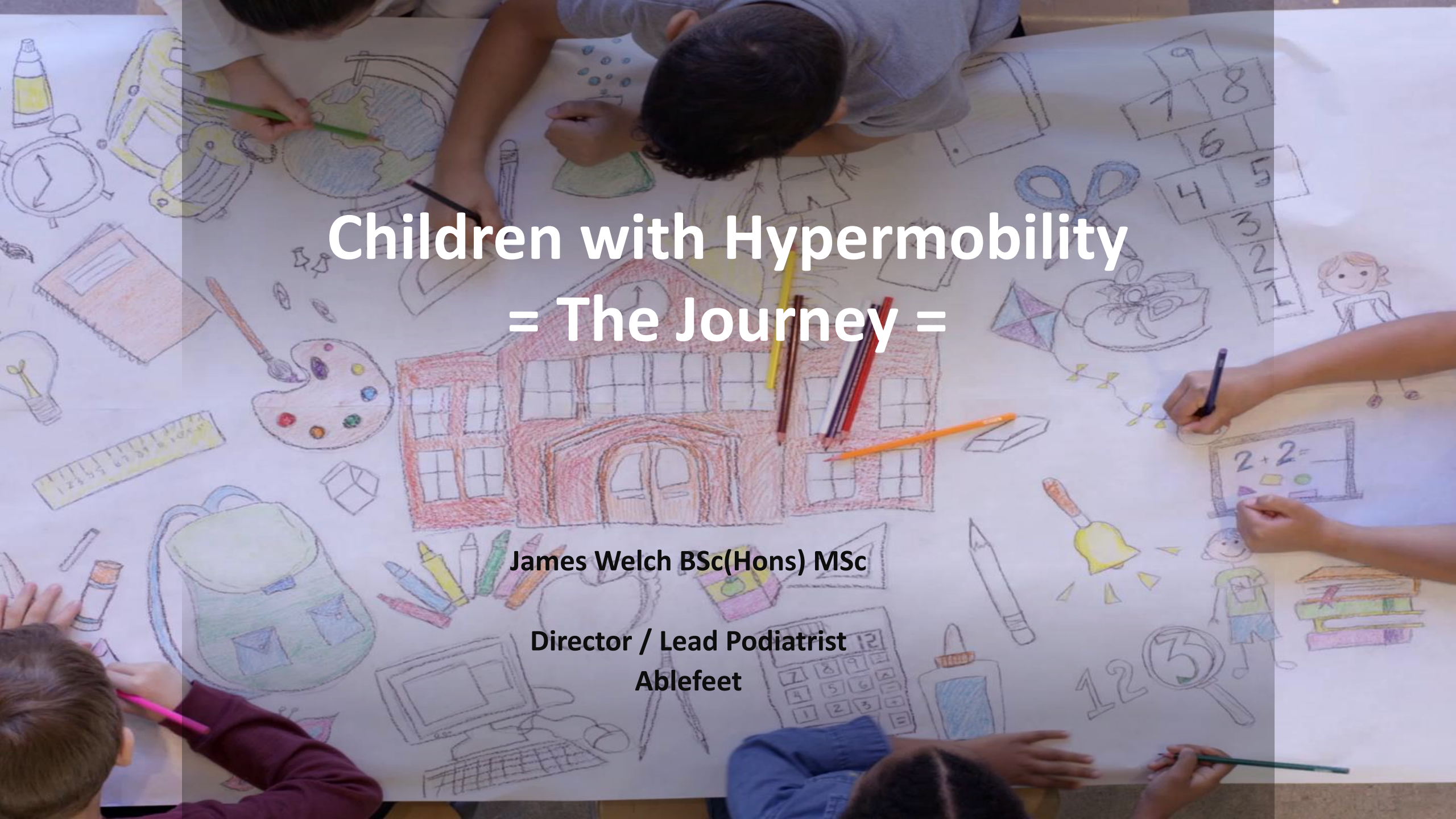


Children with Hypermobility
= The Journey =

James Welch BSc(Hons) MSc

Director / Lead Podiatrist
Ablefeet

The Journey

Podiatry Referral

- Heidi, 7 years old
- Complaining of pain in her feet and ankles, but also knees and hips, from first thing in the morning
- Pain continues throughout the day with no signs abating
- Has dislocated her left knee three times

Podiatry Referral

- She can be rather reserved due to the constant pain, but isn’t the sort of child to complain
- She can feel really tired a lot of the time, sometimes struggling to keep up with peers
- She’s been told she’s hypermobile

Initial assessment

- Full term pregnancy, with no gestational difficulties, born via vaginal delivery, weighing 7lbs
- APGAR Score – 8

Indicator	0 points	1 point	2 points
Appearance	Blue; Pale	Pink body; blue extremities	Pink
Pulse	Absent	Below 100 bpm	Over 100 bpm
Grimace (reflex irritability)	Floppy	Minimal response to stimulation	Prompt response to stimulation
Activity (muscle tone)	Absent	Flexed arms and legs	Active
Respiration	Absent	Slow & irregular	Vigorous cry

Initial assessment

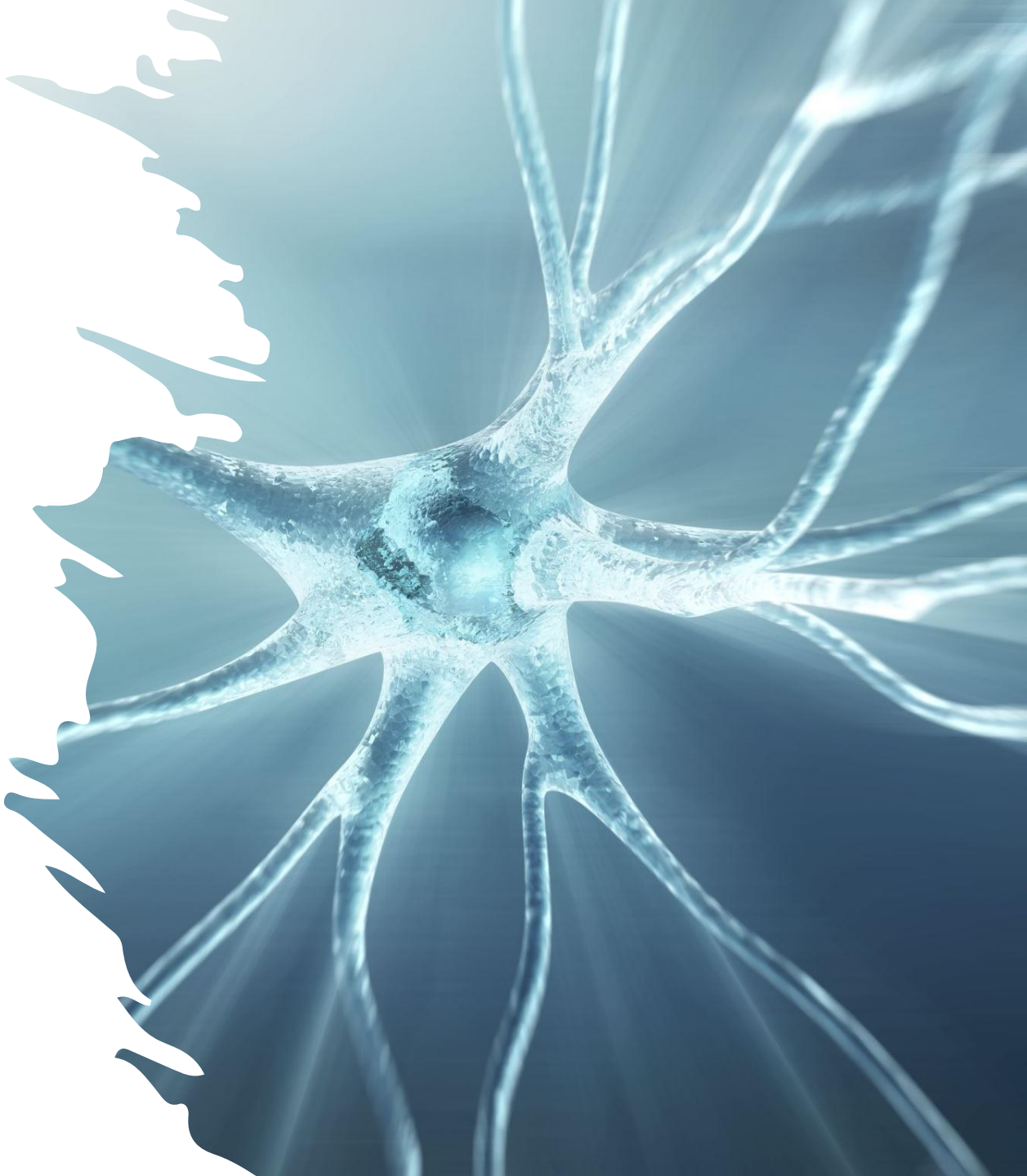
- Not crawling at 12/12
- Not cruising at 18/12
- Only became independently mobile at 2.5 yo
- Difficulty using cutlery, holding a pen, fastening buttons
- History of constipation
- Disrupted sleep pattern
- PMHx: Maternal history of Ehlers-Danlos Syndrome (Hypermobile Type 5)
- Paediatric flat foot

Common phrases

- “He’s struggling to walk”
- “We’ve been told he’s hypermobile, but we don’t know if it’s a problem”
- “She needs orthotics to control her hypermobility”
- “My daughter seems to fall over all of the time, much more than any of her classmates”
- “His knee keeps popping out”
- “She’s got such flat feet which you need to correct”
- “I think my son’s got growing pains”

Pain

- “An unpleasant sensory and emotional experience associated with, or resembling that associated with, actual or potential tissue damage.”
  - Pain is always a personal experience that is influenced to varying degrees by biological, psychological and social factors
  - Pain and nociception are different phenomena. Pain cannot be inferred solely from activity in sensory neurons.
  - Through their life experiences, individuals learn the concept of pain.



Pain

- A person's report of an experience as pain should be respected
- Although pain usually serves an adaptive role, it may have adverse effects on function and social psychological well-being
- Verbal description is only one of several behaviors to express pain; inability to communicate does not negate the possibility that a person experiences pain.

Matryoshka Doll Analogy

Legends of the scene

- Professor Rodney Grahame

- Dr Alan Hakim

Hypermobility

- An excessive range of motion in any given joint over and above what would be deemed a normal range of motion (Grahame, 2008; Simmonds & Kerr, 2007)

- It is most common in childhood and adolescence, in females, and Asian and Afro-Caribbean races

- Hypermobility can be associated with joint and ligament injuries, pain, fatigue and other symptoms.

Hypermobility

- A sign of a more serious underlying condition, which are often passed down through the generations. These conditions are known as Heritable Disorders of Connective Tissue (HDCT).

- Difficulties arise when that patient is “diagnosed” with hypermobility, which occurs all too frequently

- Hakim & Grahame (2003) stated that “*the act of recognition becomes the goal in itself rather than the medium through which effective therapy can be provided”*

Benefits of physical activity (in hypermobile pts)

- Improved mental health
- Improved co-ordination
- Improved balance
- Improves your sleep
- Improves digestion
- Improved bladder and bowel function
- Increased ability to manage pain
- Increased energy levels
- Strengthen bones
- Reduced risk of some cancers
- Increased chances of living longer

Barriers to physical activity (Simmonds et al 2007)

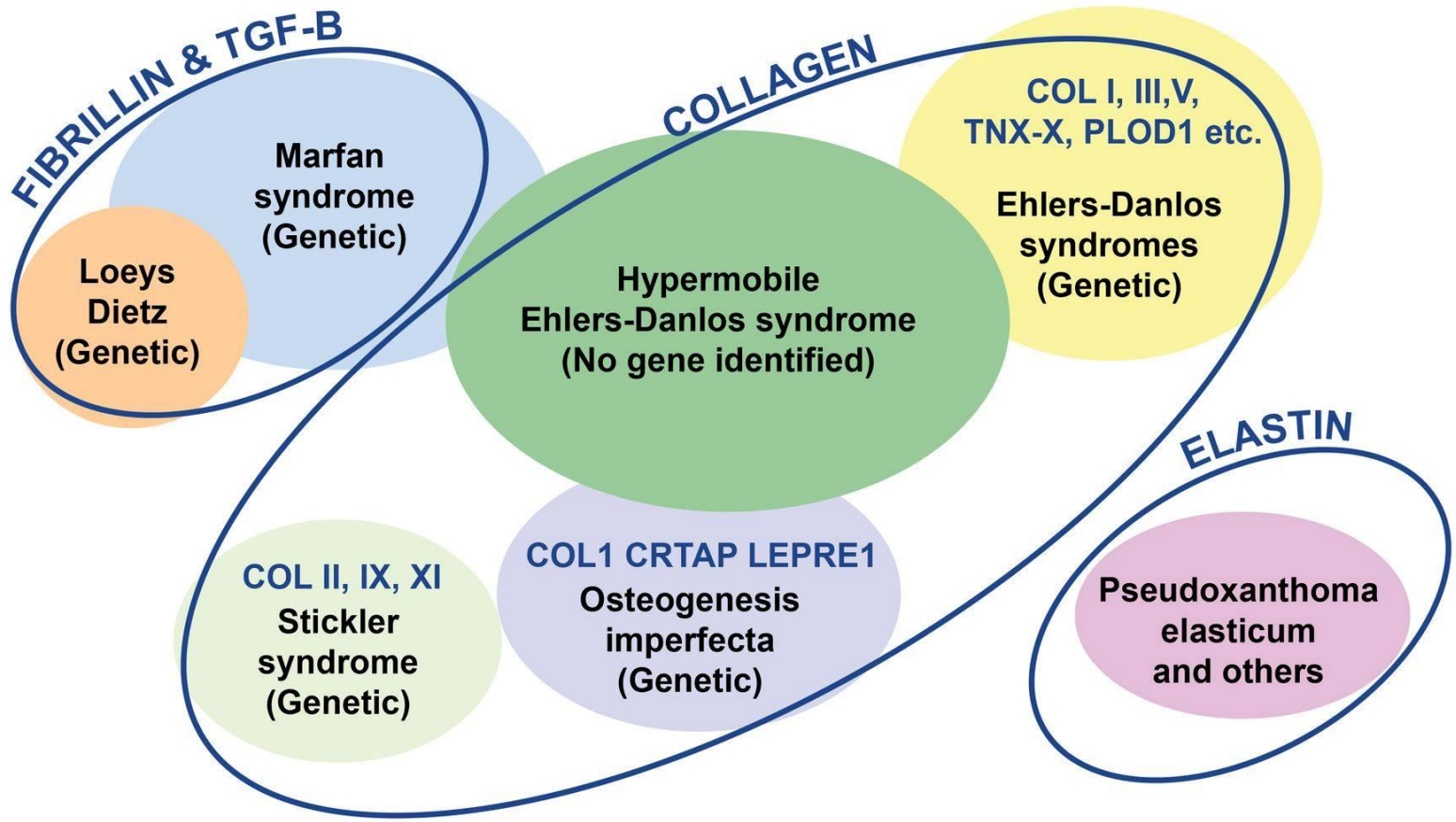
- Misconceptions about physical activity
- Fear of injury and pain
- Fatigue
- Not enough time
- Low motivation
- Boring
- Don’t know what to do

Question

How many hereditable disorders of connective tissue should we be considering?

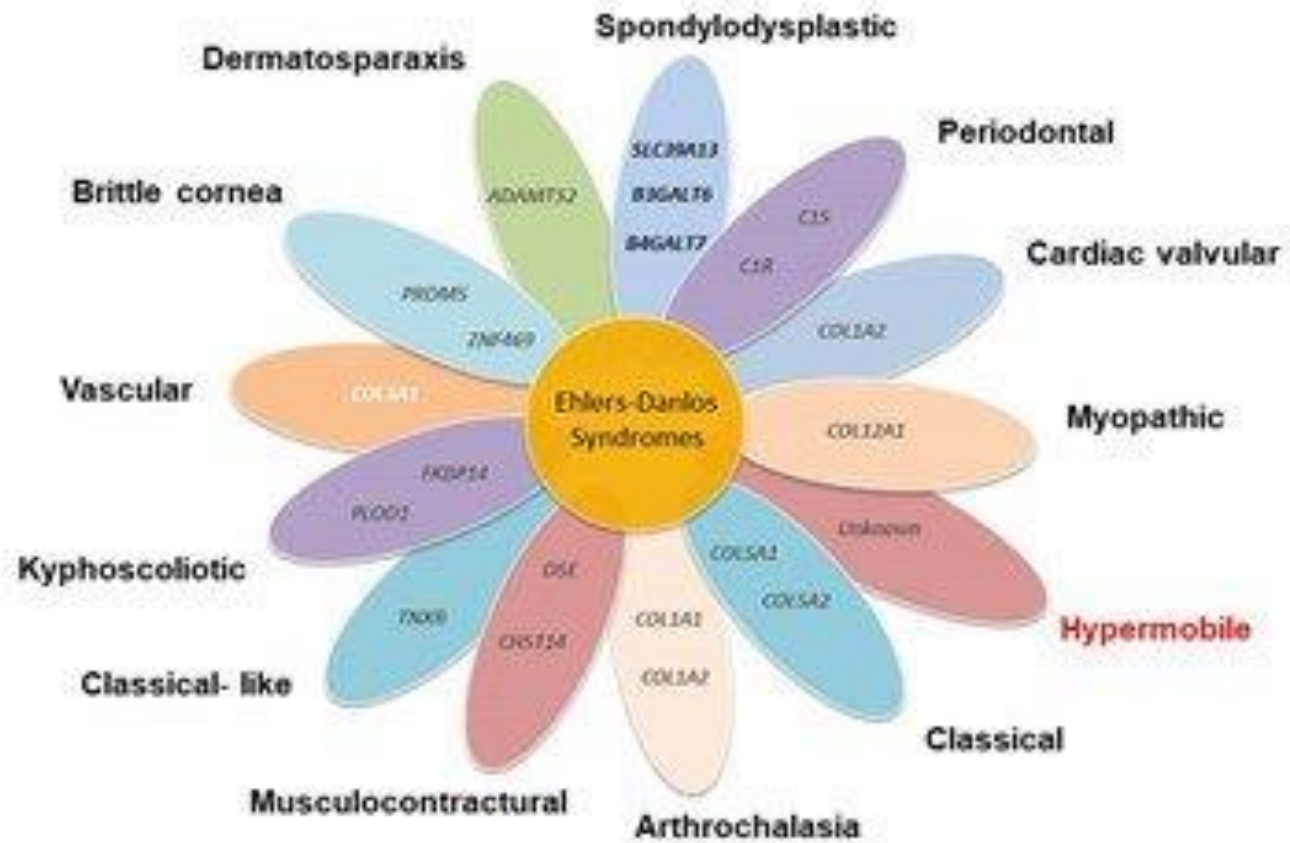
HMSA
Hypermobility Syndromes Association

The More Common Heritable Disorders of Connective Tissue



hypermobility.org

Ehlers-Danlos Syndrome



+ 14th subtype: *AEBP1* gene

J Amberger, C Bochinni, A Hamosh, 2018

“No other disease in the history of modern medicine, has been neglected in such a way as Ehlers-Danlos Syndrome”

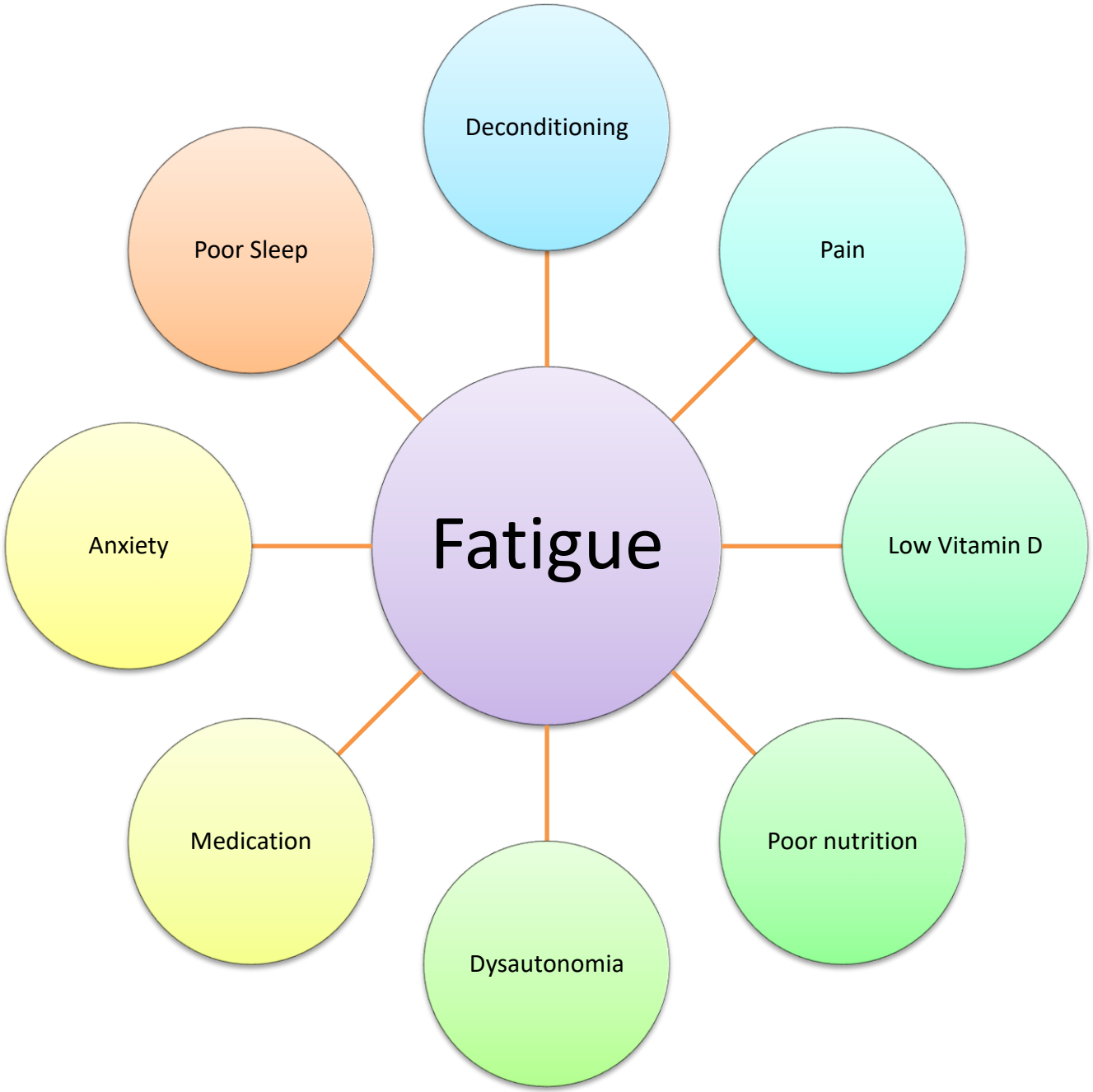
Prof Rodney Grahame

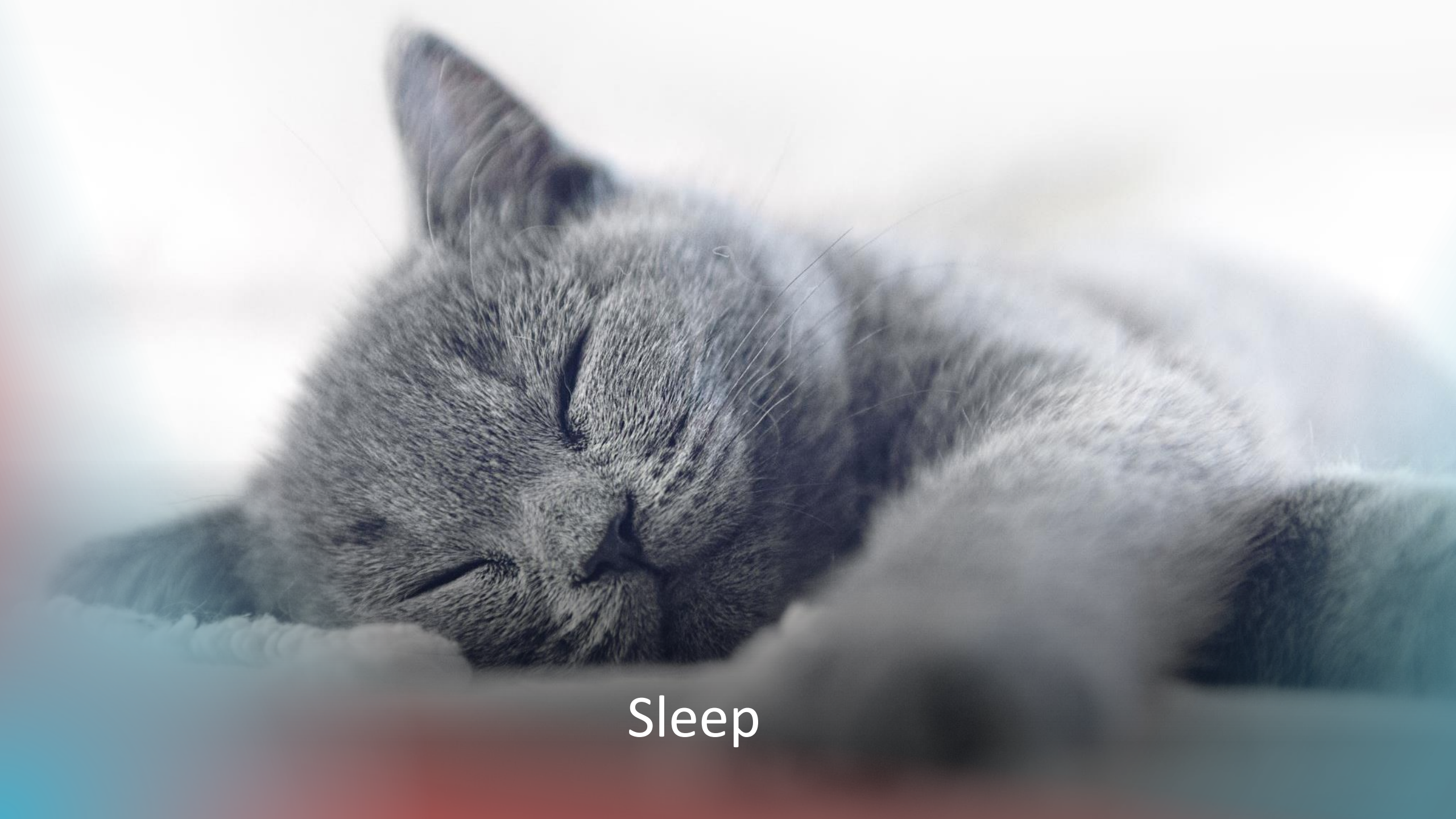
Question

What were the most common symptoms reported by the participants to the EDS Global registry?

EDS

- The EDS Global registry – report of 1334 participants - reporting on symptoms and co-morbidities.
- Highest ranked percentages (95%+)
  - Fatigue
  - MSK pain
  - Mobility concerns
  - Sleep concerns


Deconditioning
Poor Sleep
Pain
Anxiety
Fatigue
Low Vitamin D
Medication
Poor nutrition
Dysautonomia


Sleep

How much sleep is enough?

Age	Recommended amount of sleep
Newborn to 12 months	12-16 hours a day (including naps)
Children 1 -2 years old	11-14 hours a day (including naps)
Children 3-5 years old	10-13 hours a day (including naps)
Children 6-12 years old	9-12 hours a day
Teens 13-18 years old	8-10 hours a day
Adults 18+	7-8 hours a day

Brighton Criteria

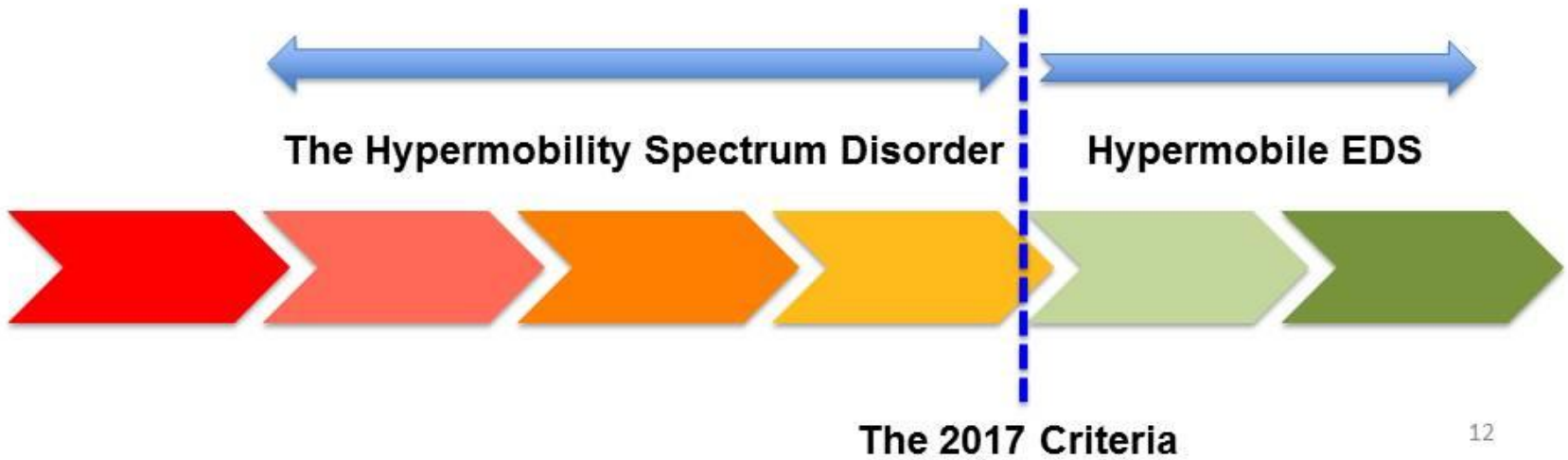
- **Major Criteria**
  - Beighton score of ⩾4 *Figure 4*)
  - Arthralgia for longer than 3 months in 4 or more joints

- **Minor Criteria**
  - Beighton score of 1, 2, or 3 (*Figure\!s>4*)
  - Arthralgia (>3-month duration) in one to three joints or back pain (>3-month duration)or spondylosis, spndylolysis/spondylolisthesis
  - Dislocation or subluxation in more than one joint, or in one joint on more than one occasion
  - Three or more soft tissue lesions (eg, epicondylitis, tenosynovitis, bursitis)
  - Marfanoid habitus (tall, slim, span greater than height (>1.03 ratio), upper segment less than lower segment (<0.89 ratio), arachnodactyly)
  - Skin striae, hyperextensibility, thin skin, or abnormal scarring
  - Ocular signs: drooping eyelids, myopia, antimongoloid slant
  - Varicose veins, hernia, or uterine or rectal prolapse
  - Mitral valve prolapse

- **Requirement for Diagnosis**
  - Any one of the following:
    - two major criteria
    - one major plus two minor criteria
    - four minor criteria
    - two minor criteria and unequivocally affected first-degree relative in family history

The International Consortium with The Ehlers-Danlos Society has sought to answer these questions.

On the far Left the person is hypermobile and well with nothing else to find; on the Right of the dash line are people with hEDS as defined by the new criteria. To the Left of the dashed line i.e. those who do not meet the criteria (or have any other condition) we now use the term Hypermobility Spectrum Disorder (HSD) – people with their own sets of problems due to their hypermobility but who do not have hEDS.



The International Consortium on Ehlers-Danlos Syndromes & Related Disorders
In Association with The Ehlers-Danlos Society

Diagnostic Criteria for Hypermobile Ehlers-Danlos Syndrome (hEDS)

This diagnostic checklist is for doctors across all disciplines to be able to diagnose EDS

Distributed by The Ehlers Danlos Society.

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

The clinical diagnosis of hypermobile EDS needs the simultaneous presence of all criteria, 1 ***and*** 2 ***and*** 3.

CRITERION 1 – Generalized Joint Hypermobility

One of the following selected:

- ☐ ≥6 pre-pubertal children and adolescents
- ☐ ≥5 pubertal men and woman to age 50
- ☐ ≥4 men and women over the age of 50

Beighton Score: 0 /9

If Beighton Score is one point below age- and sex-specific cut off, two or more of the following must also be selected to meet criterion:

- ☐ 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 shoulder or kneecap dislocate on more than one occasion?
- ☐ Do you consider yourself "double jointed"?

CRITERION 2 – Two or more of the following features (A, B, or C) must be present

Feature A (five must be present)

- ☐ Unusually soft or velvety skin
- ☐ Mild skin hyperextensibility
- ☐ Unexplained striae distensae or rubae at the back, groins, thighs, breasts and/or abdomen in adolescents, men or pre-pubertal women without a history of significant gain or loss of body fat or weight
- ☐ Bilateral piezogenic papules of the heel
- ☐ Recurrent or multiple abdominal hernia(s)
- ☐ Atrophic scarring involving at least two sites and without the formation of truly papyraceous and/or hemosideric scars as seen in classical EDS
- ☐ Pelvic floor, rectal, and/or uterine prolapse in children, men or nulliparous women without a history of morbid obesity or other known predisposing medical condition
- ☐ Dental crowding and high or narrow palate
- ☐ Arachnodactyly, as defined in one or more of the following:
  (i) positive wrist sign (Walker sign) on both sides, (ii) positive thumb sign (Steinberg sign) on both sides
- ☐ Arm span-to-height ratio ≥1.05
- ☐ Mitral valve prolapse (MVP) mild or greater based on strict echocardiographic criteria
- ☐ Aortic root dilatation with Z-score >+2

Feature A total: 0 /12

Feature B

- ☐ Positive family history; one or more first-degree relatives independently meeting the current criteria for hEDS

Feature C (must have at least one)

- ☐ Musculoskeletal pain in two or more limbs, recurring daily for at least 3 months
- ☐ Chronic, widespread pain for ≥3 months
- ☐ Recurrent joint dislocations or frank joint instability, in the absence of trauma

CRITERION 3 – All of the following prerequisites MUST be met

1. ☐ Absence of unusual skin fragility, which should prompt consideration of other types of EDS
2. ☐ Exclusion of other heritable and acquired connective tissue disorders, including autoimmune rheumatologic conditions. In patients with an acquired CTD (*e.g.* Lupus, Rheumatoid Arthritis, etc.), additional diagnosis of hEDS requires meeting both Features A and B of Criterion 2. Feature C of Criterion 2 (chronic pain and/or instability) cannot be counted toward a diagnosis of hEDS in this situation.
3. ☐ Exclusion of alternative diagnoses that may also include joint hypermobility by means of hypotonia and/or connective tissue laxity. Alternative diagnoses and diagnostic categories include, but are not limited to, neuromuscular disorders (*e.g.* Bethlem myopathy), other hereditary disorders of the connective tissue (e.g. other types of EDS, Loeys-Dietz syndrome, Marfan syndrome), and skeletal dysplasias (*e.g.* osteogenesis imperfecta). Exclusion of these considerations may be based upon history, physical examination, and/or molecular genetic testing, as indicated.

Diagnosis: _____

Save as... Print ERASE ALL Cannot be undone!

v3

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Pediatric joint hypermobility: a diagnostic framework and narrative review

Louise Jane Tofts, Jane Simmonds, Sarah B. Schwartz, Roberto M. Richheimer, Constance O'Connor, Ellen Elias, Raoul Engelbert, Katie Cleary, Brad T. Tinkle, Antonie D. Kline, Alan J. Hakim, Marion A. J. van Rossum & Verity Pacey

Hypermobility Spectrum Disorder

- Autosomal dominant pattern of inheritance
- Multi-systemic CTD causing systemic tissue laxity, due to collagen mutation
- Poorly diagnosed condition, approx 1 in 20 correctly diagnosed (Grahame, 2008)
- Accounts for 45% of routine general rheumatology referrals

Paediatric Working Group
The International Consortium on Ehlers-Danlos Syndromes & Related Disorders
In Association with The Ehlers-Danlos Society

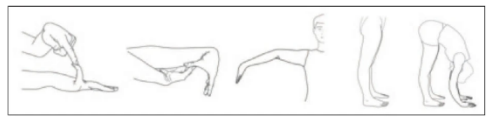
Diagnostic Criteria for Paediatric Joint Hypermobility

This diagnostic checklist is to support doctors to diagnose paediatric joint hypermobility and hypermobility spectrum disorder

Distributed by The Ehlers Danlos Society.

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

Children From 5 Years Of Age Until Biological Maturity



L☐R☐ L☐R☐ L☐R☐ L☐R☐ ☐

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

Skin and Tissue Abnormalities

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

Score: _____/6
Must be a minimum of 3

Musculoskeletal Complications

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

Score: _____/3
Must be a minimum of 2

Co-Morbidities

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

Any number causing distressor disability?
Y / N

Prerequisites:

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

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

GJH & HSD

Table 1 Diagnostic framework for pediatric joint hypermobility in the presence of skin abnormalities, musculoskeletal complications, and/or core comorbid conditions

	Generalized joint hypermobility	Skin and tissue abnormalities	Musculoskeletal complications	Core comorbidities
Asymptomatic				
Pediatric generalized joint hypermobility	Present	Absent	Absent	Absent
Pediatric generalized joint hypermobility with skin involvement	Present	Present	Absent	Absent
Symptomatic conditions				
Pediatric generalized joint hypermobility with core comorbidities	Present	Absent	Absent	Present
Pediatric generalized joint hypermobility with core comorbidities and with skin involvement	Present	Present	Absent	Present
Pediatric hypermobility spectrum disorder, musculoskeletal subtype	Present	Absent	Present	Absent
Pediatric hypermobility spectrum disorder, musculoskeletal subtype with skin involvement	Present	Present	Present	Absent
Pediatric hypermobility spectrum disorder, systemic subtype	Present	Absent	Present	Present
Pediatric hypermobility spectrum disorder, systemic subtype with skin involvement	Present	Present	Present	Present

THE HSD SPECTRUM

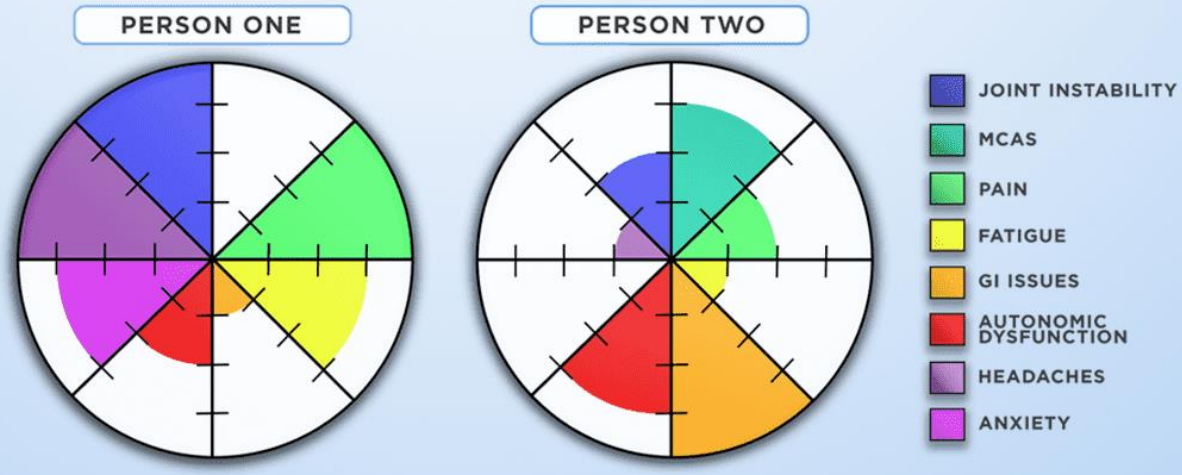
THE HYPERMOBILITY SPECTRUM DISORDERS
DO NOT EXIST ON A LINEAR SPECTRUM LIKE THIS:

EACH PERSON'S EXPERIENCE IS A COMBINATION OF
THE SPECIFIC SYMPTOMS THAT AFFECT THEM.
THE SPECTRUM LOOKS MORE LIKE THIS:

The Ehlers Danlos Society

THE HSD SPECTRUM

TWO DIFFERENT PEOPLE WITH HSD MAY EXPERIENCE VERY DIFFERENT SYMPTOMS



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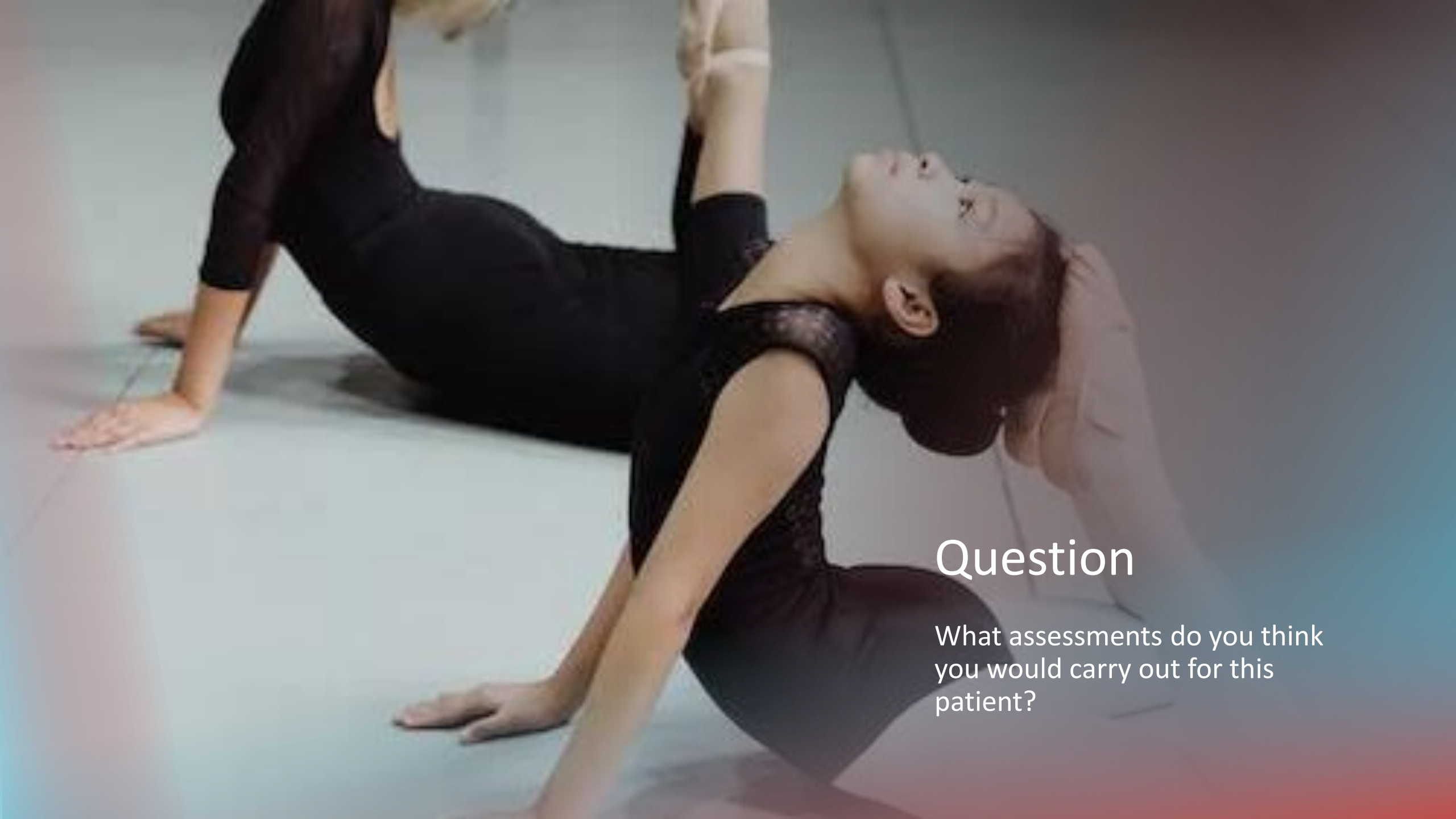


Question

What assessments do you think you would carry out for this patient?

Assessment

- Beighton Scale
- Lower Limb Assessment Score
- Hospital del Mar Criteria

Beighton Scale (1973)

…but

- Simple scale to use & quick
- Only 1 lower limb marker
- ….and for some reason, discounted…
- an excess range of passive dorsiflexion of the ankle and eversion of the foot
- Only tracks a small number of joints, therefore some conditions may be missed
- Why 6/9? Why not 4 or 5?

Lower Limb Assessment Score

- Ferrari et al (2005)
- Included movements of the joints occurring in several planes of motion rather than in just one direction.
- Approx. 15 minutes to complete
- Maximum 12 marks per limb
- 7/12 defined as “hypermobile”













Hospital del Mar Criteria

- Bul
- Ins

Hospital del Mar Criteria for the Clinical Assessment of Joint Hypermobility
Upper Extremities
1. Thumb: Passive apposition of the thumb to the flexor of the forearm at <21 mm.
2. Metacarpophalangeal: With the palm of the hand resting on the table, the passive dorsiflexion of the fifth finger is ≥90°.
3. Elbow hyperextension: The passive extension of the elbow is ≥ 10.
4. External shoulder rotation: With the upper arm touching the body and with the elbow at 90, the forearm is taken in external rotation ≥ 85 of the sagital plane (shoulder-to-shoulder line).
Lower Extremities, Supine Position
5. Hip abduction: The passive hip abduction can be taken to an angle of ≥ 85.
6. Patellar hypermobility: With one hand holding the proximal end of the tibia, the patella can be moved well to the sides with the other hand.
7. Ankle and feet hypermobility: An excess range of passive dorsiflexion of the ankle and eversion of the foot can be produced.
8. Metatarsophalangeal: Dorsal flexion of the toe of the foot over the diaphysis of the first metatarsal is ≥ 90.
Lower Extremities, Prone Position
9. Knee hyperflexion: Knee flexion allows the heel to make contact with the buttock.
Ecchymoses
10. Ecchymoses: Appearance of ecchymoses after hardly noticed, minimal traumatism.


What are your options for Rx?

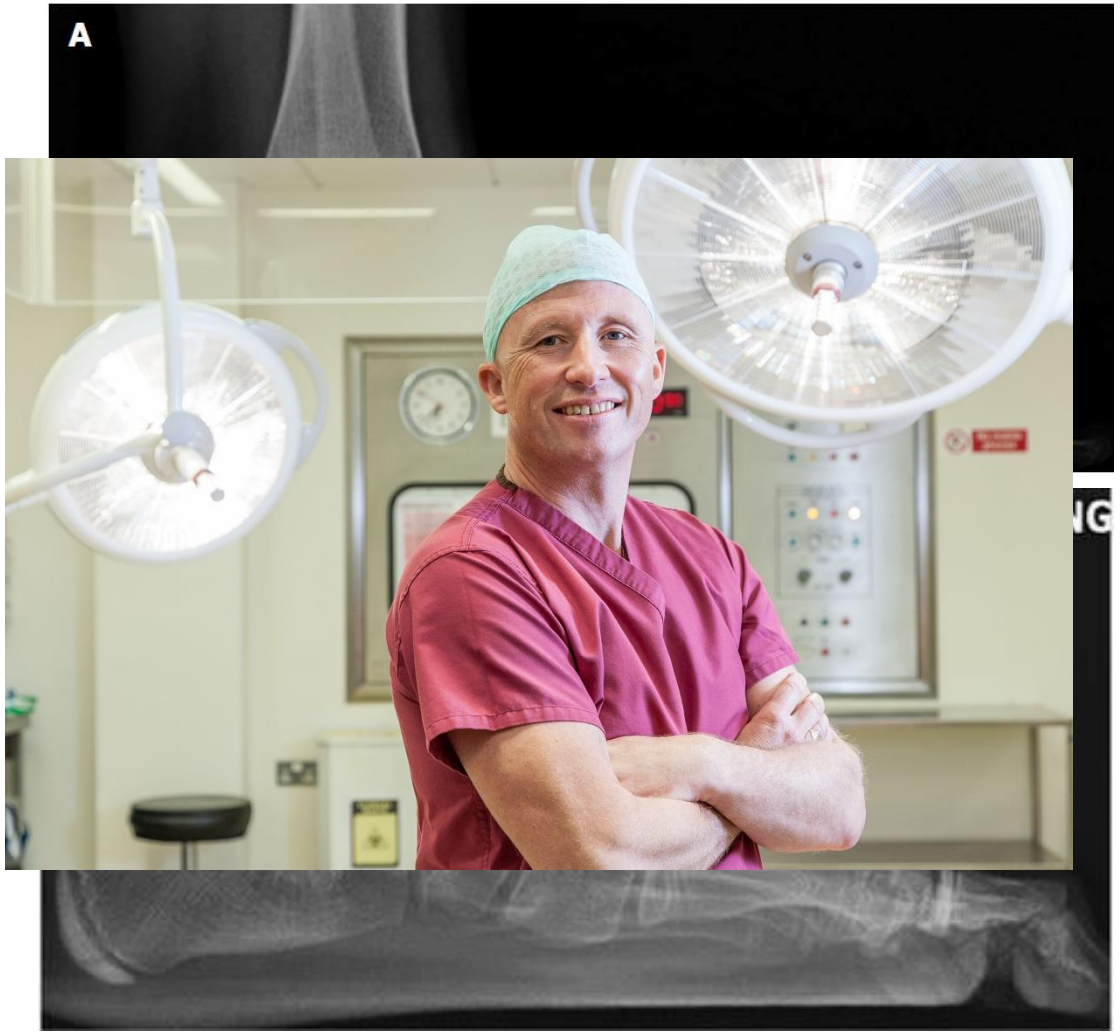
Treatment

- Be mindful of the phrases / terminology you use with the child
- Intensive physiotherapy and hydrotherapy
- Patient / parent education
- Management

Treatment

- Orthoses
- Piedro boots (not always go-to treatment)
- Taping
- Surgical management

Paediatric flat foot – the surgical perspective



Summary

- Make sure you take a thorough history, this area can be very complex
- There’s more to remember than just the Beighton scale
- Is it a normal variant, or something else?

Questions?

- **surgery@ablefeet.com**
- **@Ablefeet**
- **@Ablefeet**

- www.hypermobility.org

- www.rarediseases.org

- www.ehlers-danlos.com

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