

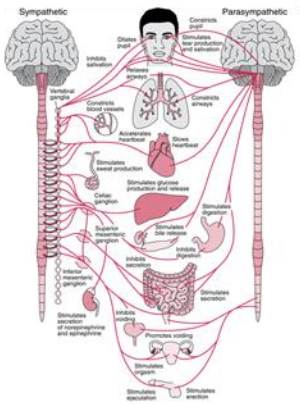


MANAGEMENT OF HYPERMOBILITY SPECTRUM DISORDERS (HSD) HYPERMOBILE EHLERS DANLOS SYNDROME (hEDS) IN CHILDREN AND ADOLESCENTS

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Ehlers-Danlos Support UK
MAKING OUR INVISIBLE VISIBLE

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

HMSA
Hypermobility Syndromes Association

UCL
GREAT ORMOND STREET INSTITUTE OF CHILD HEALTH

Plan

Spectrum of hypermobility

How big is the problem

Improving services

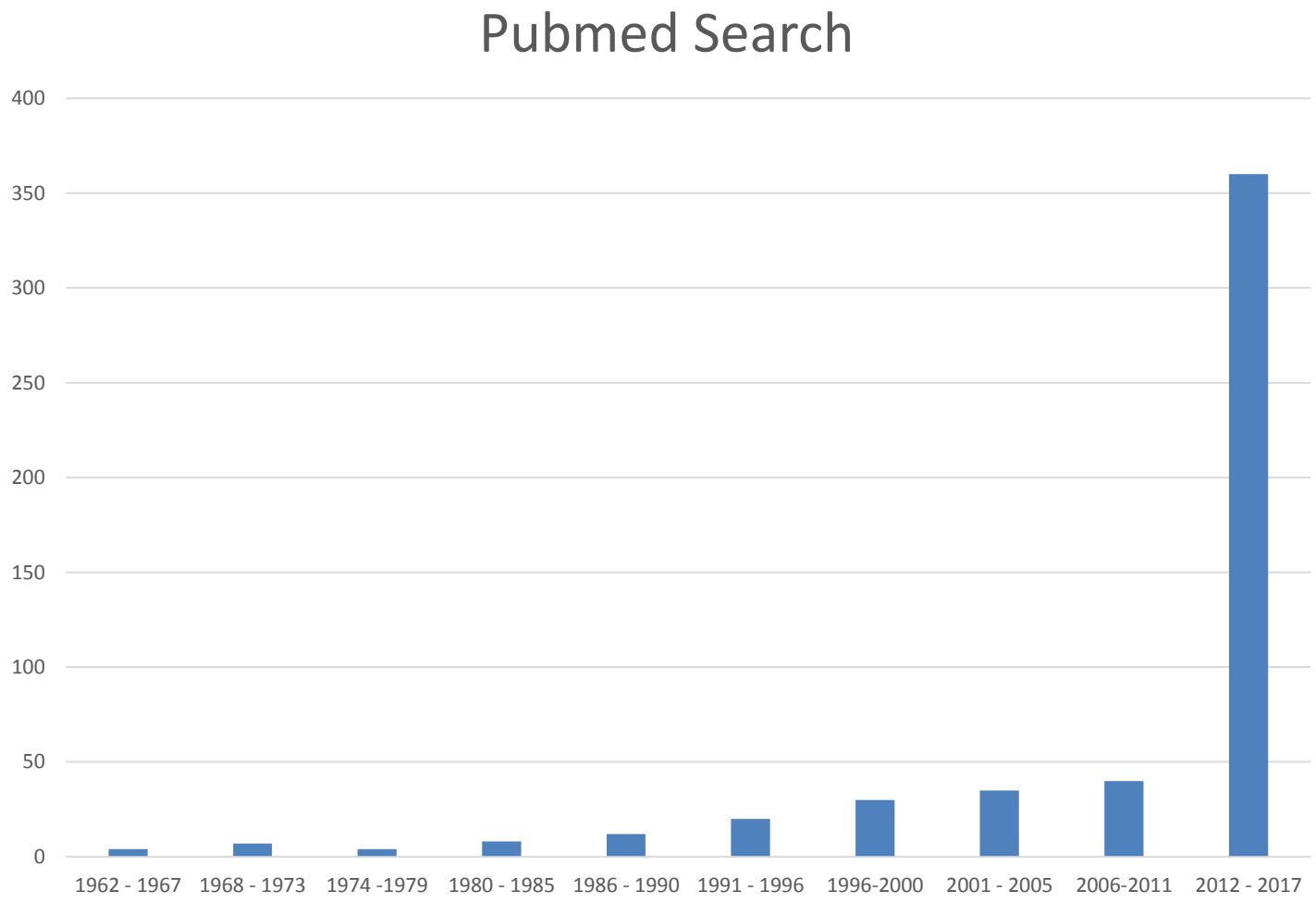
Assessment

Management

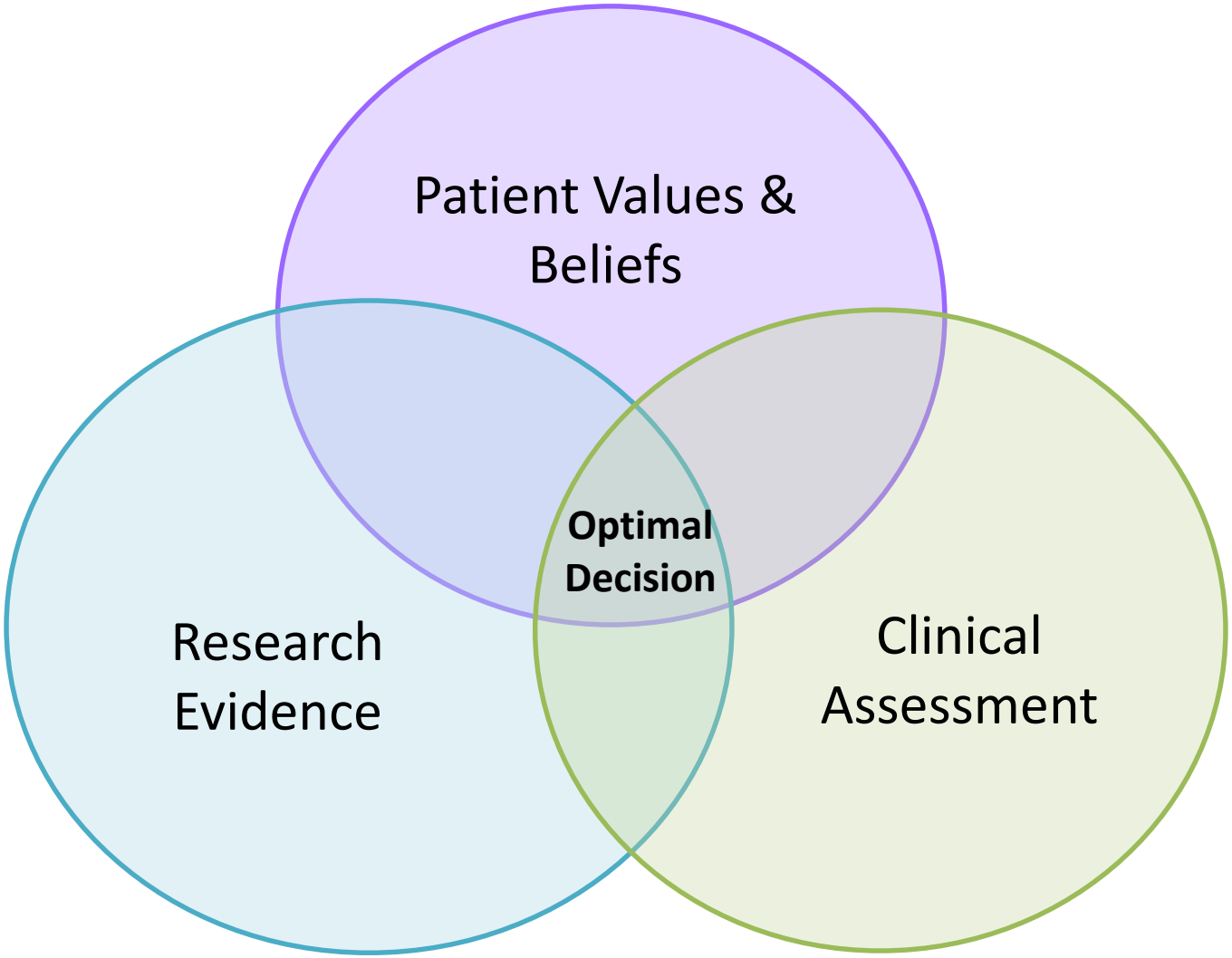
Cases

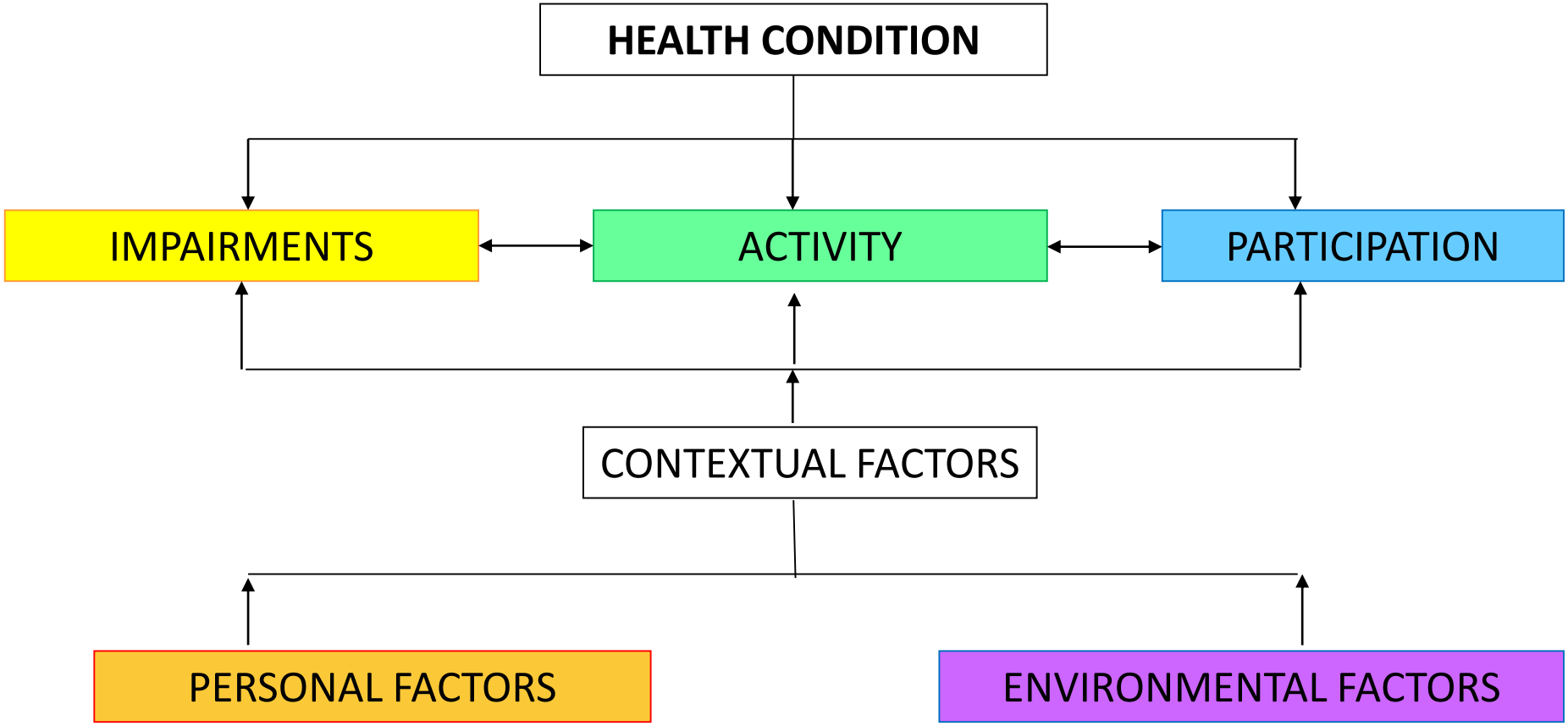
Learning outcomes

- Appreciate the hypermobility spectrum in children and adolescents
- Select objective measures and outcome measures
- Apply clinical reasoning and the evidence base to develop a management plan
- Consider preventative strategies
- What do you want to learn?

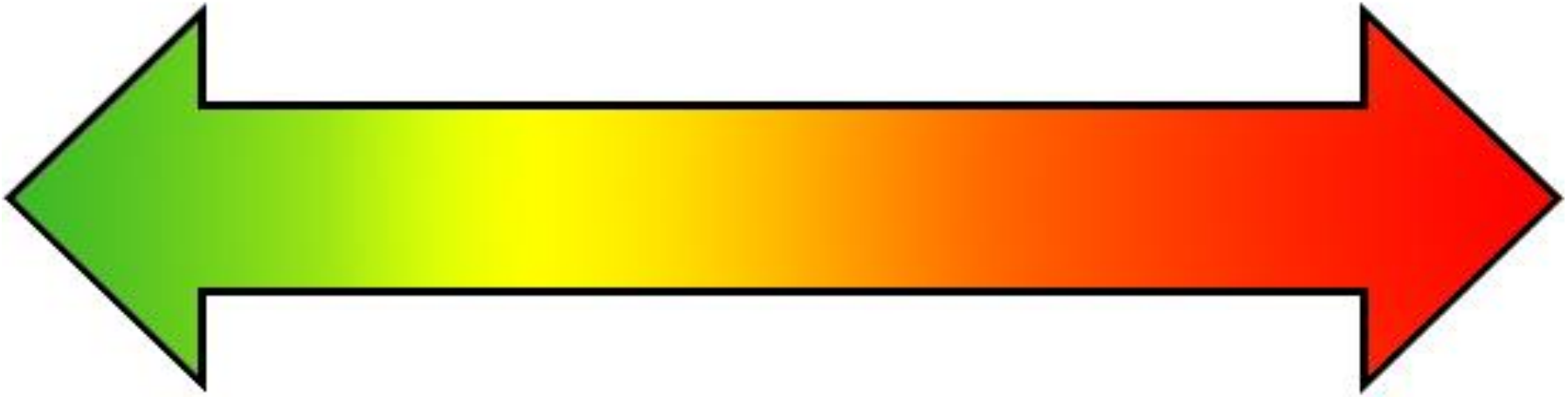


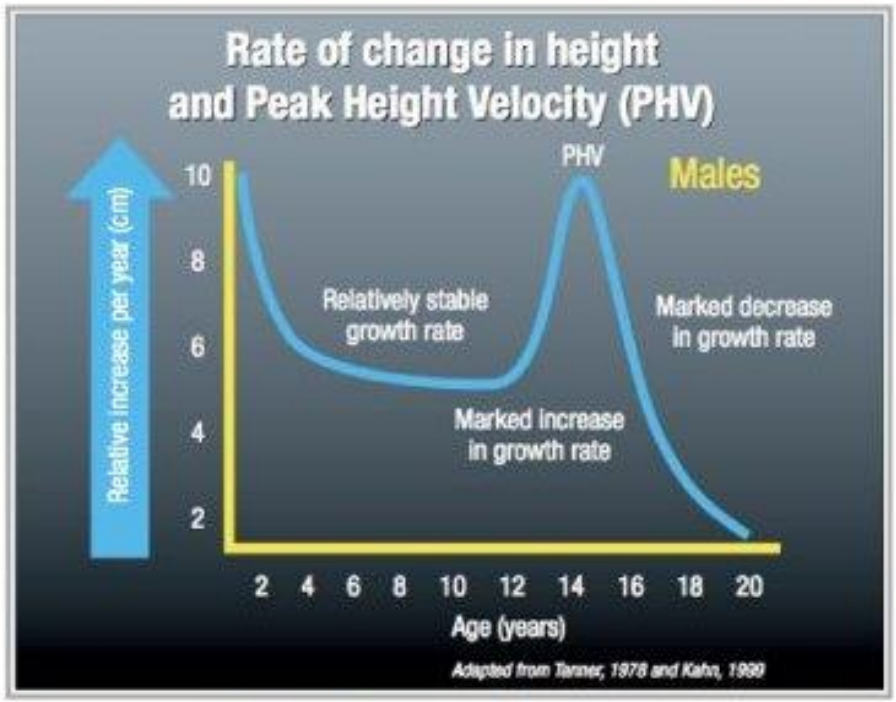
Search terms: Joint Hypermobility; Hypermobility Syndrome; Ehlers Danlos Syndrome Hypermobility Type

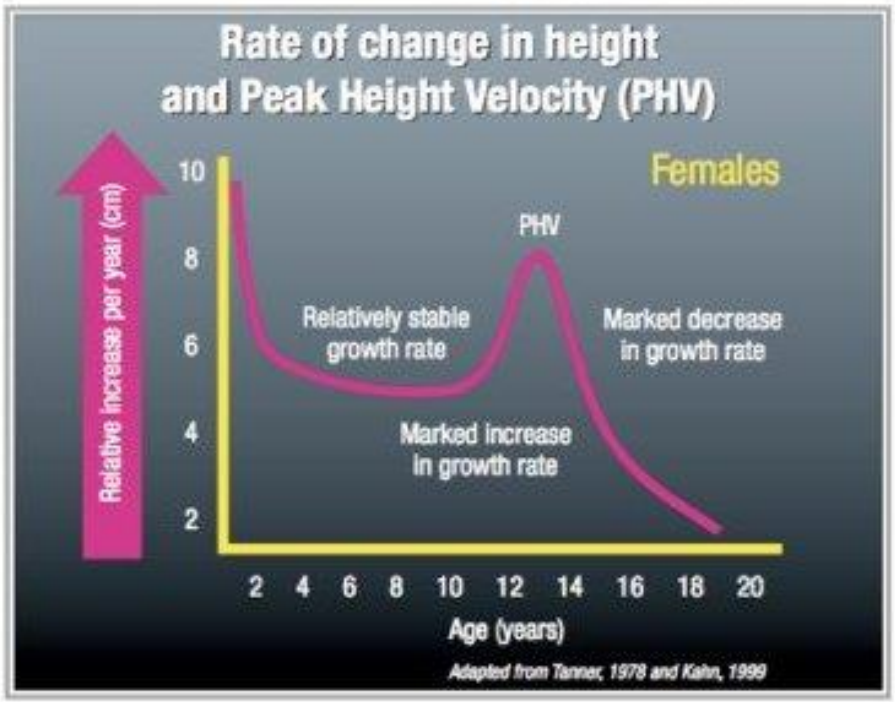

Patient Values & Beliefs
Optimal Decision
Research Evidence
Clinical Assessment



International Classification of Functioning Disability and Health (WHO)







Reflect on the key physiological and psychological changes during childhood

1. Limb development/ gait
2. Hormonal changes
3. Growth – PHV

Seminal study – Kirk et al., (1967) described three quarters of patients developed symptoms prior to age 15 years

Hypermobile children are at greater risk of developing musculoskeletal problems than non hypermobile counterpart (Tobias et al 2013)

Meet Sam
5 years

Hx: late walker 19 months
Talked late

I wanna play with the other kids

IMPAIRMENTS

Joint hypermobility 9/9 + hips & shoulders

Everted ankles/over pronated, flat feet

Coordination problems (gross and fine motor)

Poor balance – unable to hop

Low muscle tone

Muscle weakness

Generalised leg pain

Tired

Slow gut motility

ACTIVITY

Not keeping up with peers
Unable to walk for > 10 mins
Struggling to run
Unable to hop
Struggling to throw and catch
Unable to ride a bike/ tricycle
Struggling with dressing
Struggling with pencil skills

PARTICIPATION

Struggling at nursery
Parents nervous about Sam starting school

PERSONAL FACTORS

Male
Low confidence/ self esteem

ENVIRONMENT

Supportive family
Very sport father and sister

Meet Helen
14 years

Hx: Hypermobility detected in early school years
Recurrent subluxations
Physiotherapists treated single areas
Never got on top of the problem

I want to be a dancer/ actor

IMPAIRMENTS

Widespread hypermobility ++
Recurrent shoulder and knee subluxations, fingers collapse,

Persistent pain – knees, shoulders and hips

Fatigue

Anxious

Low mood

Early satiety when eating/ bloating, nausea, vomiting and slow transit constipation

ACTIVITY

Unable to dance and struggling to act
Struggling with eating
Unable to travel on public transport

PARTICIPATION

Reduced attendance at & having to modify college
Reduced social activity with friends

PERSONAL FACTORS

Female
High achiever – A student
Depressed
Low confidence/ self esteem

ENVIRONMENT

Protective family
2 siblings
Mother with EDS

Meet Beth
16 years

Hx: Motor vehicle accident 18 months ago – whiplash. Previously fit and well.

I want to go to university, wants to go to concerts, wants to be a photo be a journalist

IMPAIRMENTS

Joint hypermobility 6/9 +
Widespread pain

Fatigue

Head aches

Brain fog

Severely dizzy, fainting (started 2 years ago)

Deconditioned ++

Muscle weakness

Slow transit constipated

Fatigue and sleeps poorly

ACTIVITY

Unable to walk for > 5 mins
Uses wheel chair

PARTICIPATION

Unable to go to school
Socialising only on social media only

PERSONAL FACTORS

Female
Depressed
Loves writing/ reading/ photography
Tends to “boom & bust”

ENVIRONMENT

Supportive family

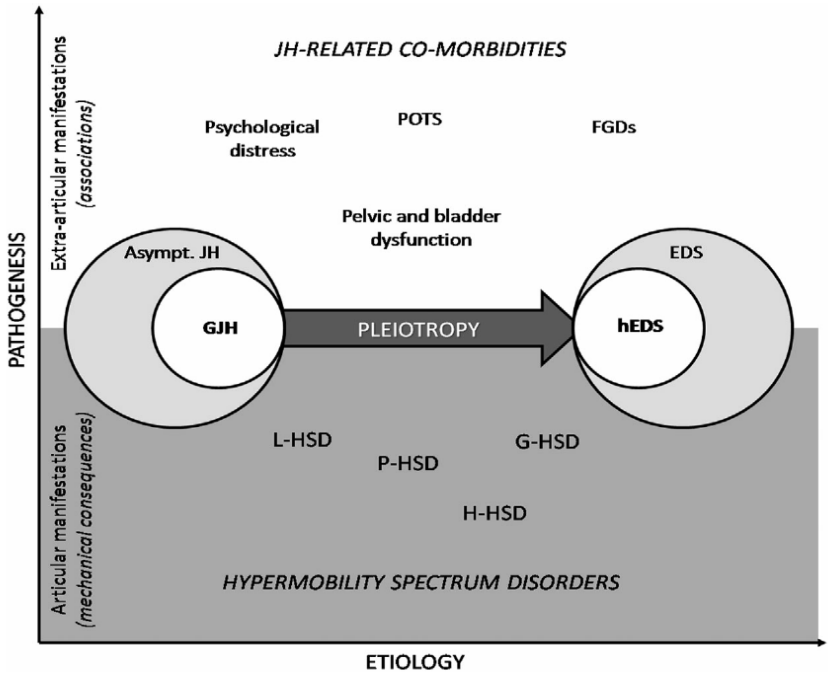
New Nosology – Criteria

New

Broad term ……..Hypermobility Spectrum Disorders (HSD)

Local, peripheral, generalised HSD……

JHS/ EDS-HT = Hypermobility Ehlers Danlos Syndrome (hEDS)



** Castori et al 2017. Am J Med Genetics. Seminars in Medial Genetics. Part C

** Malfait et al 2017. Am J Med Genetics. Seminars in Medial Genetics. Part C (out today)

** Juul Kristensen et al 2017. Am J Med Genetics. Seminars in Medical Genetics. Part C

New Classification

New

The clinical diagnosis of hEDS needs the simultaneous presence of criteria 1 & 2 & 3

Criterion 1: generalized joint hypermobility (GJH)

Pre pubertal and adolescents ≥ 6 /9 or more joints

≥5 for pubertal men and women up to the age of 50

≥4 for those >50 years of age for hEDS

…… where there has been trauma, amputation etc 5 part questionnaire (has only been validated in adults)

Criterion 2: extra articular signs and symptoms

5 signs and symptoms (eg mild skin hyperextensibility, striae, heal papules, prolapse, mitral valve prolapse, aortic root dilatation z>+2)

Criterion 3: exclusion

Marked skin fragility (consider other HDCT)

Hypermobility related to causes eg dysplasia, hypotonia, other HDCTs

How big is the problem?

Adults

Prevalence of JHS/EDS- HT in the general population is estimated at 0.75-2%
(Hakim & Sahota, 2006)

58% women 29% in men attending a London rheumatology clinic had JHS/ EDS-HT
(Hakim & Grahame, 2004)

55% of female patients presenting in the OPD in Oman had JHS/EDS-HT (60% returners)
(Clark & Simmonds, 2011)

30% of men and women presenting to a London Neuromusculoskeletal Physiotherapy Triage clinic had JHS/EDS-HT (Connelly et al, 2015)

39% presenting in a pain clinic, 10% Orthopaedics in London
(To et al., 2016)

Children and adolescents

Unknown

Cause of concern in rheumatology and physiotherapy clinics (School and PE participation)
(Adib et al 2005; Murray, 2006; Pacey et al., 2015)

Optimising Physiotherapy

Health professionals' and patients' descriptions of an 'ideal' physiotherapy Intervention for JHS/EDS-HT "notably similar"

Patients perceptions – negative experiences of PT

- exacerbation of symptoms
- lack of understanding or recognition of condition
- focus on single joint
- lack of diagnosis and flexibility

Health professional perceptions – frustration

- correcting incorrect information & unravelling mismanagement
- picking up diagnosis & looking globally, but only allowed 6 sessions
- delay in diagnosis leading to chronicity and lots of psychological baggage
- not fitting with model of acute injury and recovery

Palmer et al 2016 Physiotherapy management of JHS – a focus study of patient and health professional perspectives. Physiotherapy 102, 93-102

Important to have:

Continuity of therapist, trained in JHS/EDS-HT, who provided reassurance

Treatment flexibly delivered, patient-led, meeting and managing goals & expectations

Holistic, long term approach with ongoing maintenance

Education

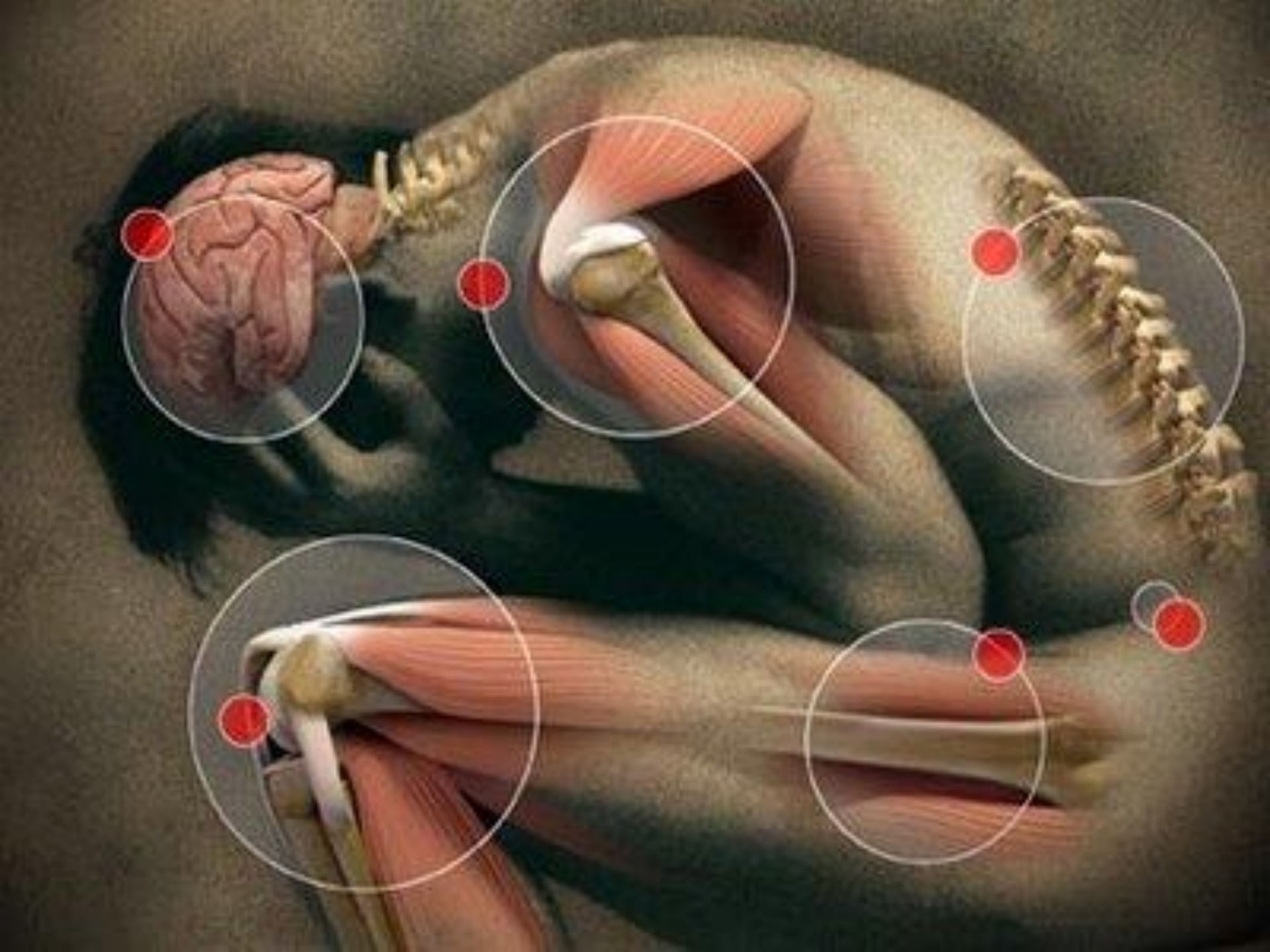
Timely intervention to prevent chronicity and equip patient with skills to manage JHS over their life course to enable them to maximise their function

Palmer et al 2016 Physiotherapy management of JHS – a focus study of patient and health professional perspectives. Physiotherapy 102, 93-102


LIFE
CHANGING
EVENT


SCHOOL


University



Hypermobility Scoring Systems

Beighton Scale

Beighton et al (1973) Annals of the Rheumatic Diseases.32:413 -7

Rotés-Querol

Rotés-Querol J, Argany A. (1957) Rev Rhum Mal Osteoartic :24:535-539.

Contompasis

McNerney JE & Johnston WB (1979) J Am Pediatr Assoc. 69: 69 - 72

Lower Limb Assessment Scale (prac)

Ferrari et al (2005) Clinical and Experimental Rheumatology. 23: 413 – 420

5 Part Hypermobility Questionnaire

Hakim AJ, Grahame R. (2003) Int J Clin Pract. 57: 163-166.

The Beighton 9-Point Hypermobility Score

	Right	Left
5th MCP	1	1
Thumb	1	1
Elbow	1	1
Knees	1	1
Lumbar Spine	1	
	9	

Cut-off 4/9, 5/9 6/9?

Beyond the Beighton Score

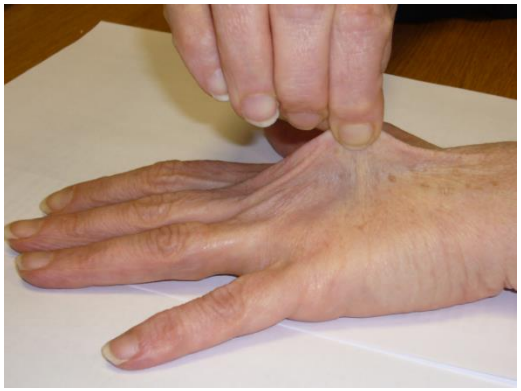
Beyond the Beighton Score

Beyond the Beighton Score

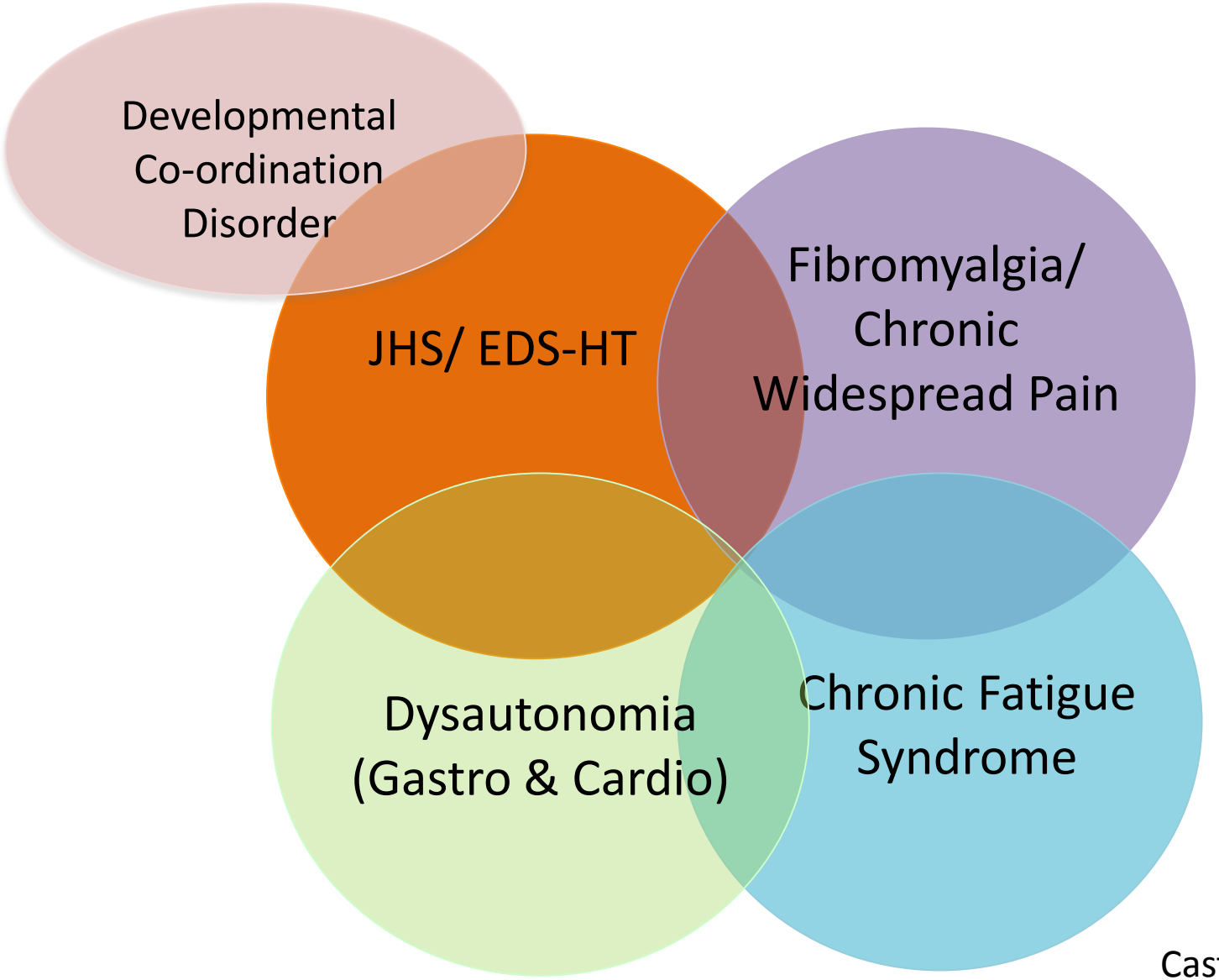

1 year
3 years
6 years
10 years

Beyond the Beighton Score

Skin signs



Overlapping Conditions and Symptomology



Castori et al., 2015

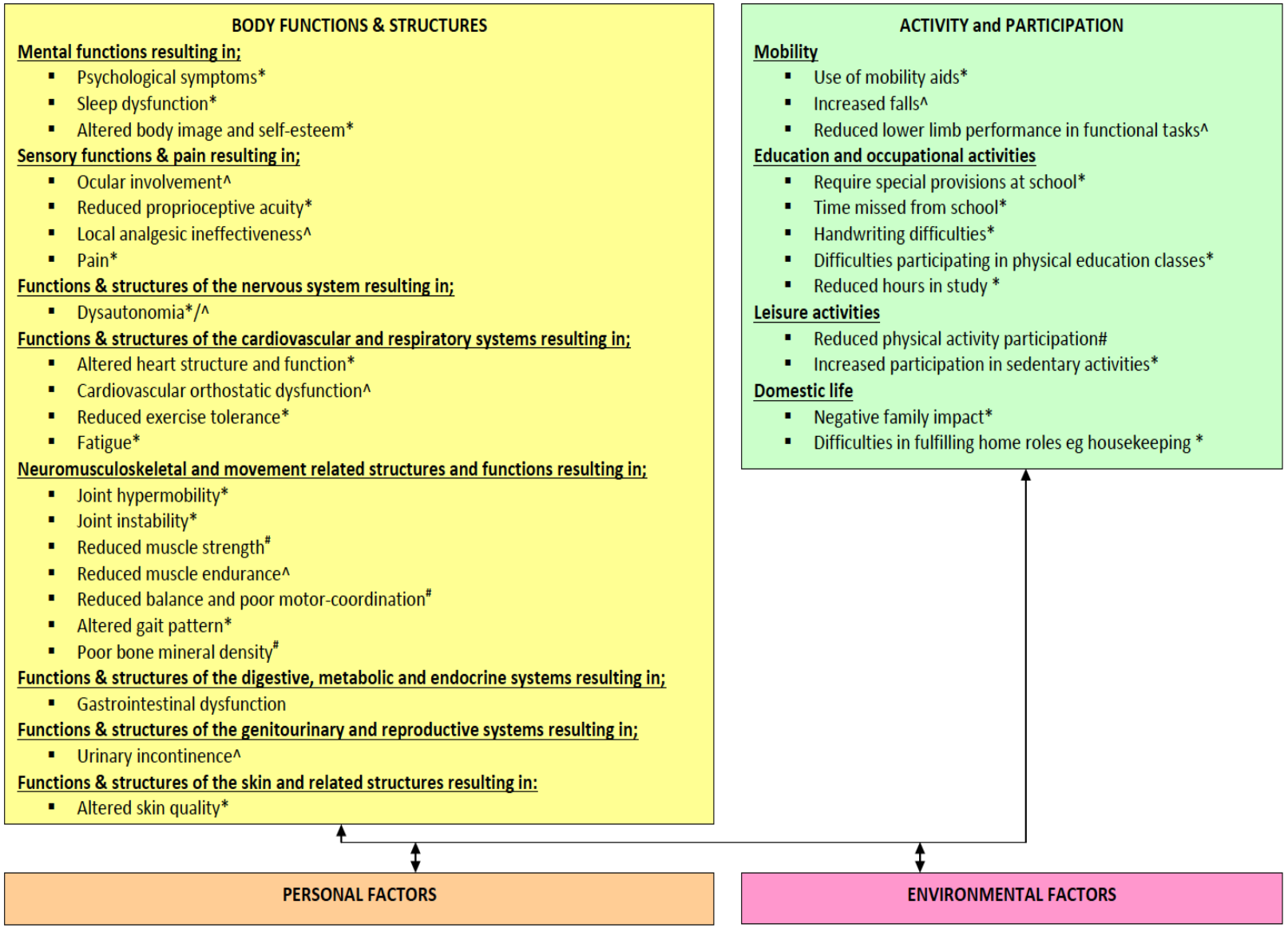


Figure 2.6 Proposed ICF model of Joint Hypermobility Syndrome in children

* Consistent evidence in children with JHS

Inconsistent evidence in children with JHS

^ Evidence in adults with JHS and theoretical basis assumes it could be possible in children, and no contradicting studies in children with JHS

NB. Current studies suggest varicose veins and asthma are not more prevalent in children with JHS than their non-affected peers, and are therefore not included in the model.

Pacey, 2014 PhD thesis

CHILDREN

BODY FUNCTIONS AND STRUCTURES / IMPAIRMENTS

- Pain (Adib et al., 2005; Pacey et al., 2015)
  - Weight bearing joints, especially the knee joints
  - Chronic widespread
- Fatigue (Pacey et al., 2015)
- Joint instability, dislocations and subluxations (Pacey et al., 2015)
- Muscle tone, strength and endurance
  (Mitz-Itzen et al., 2009; Celetti et al., 2012; Pacey et al., 2015)
- Proprioception, balance, coordination and gait
  (Ferrell et al., 2004,7; Kirby et al., 2007; Hanewinkel et al., 2009; Fatoye et al., 2009;11; Celetti et al., 2012; Schubert – Hajlmarsson et al., 2012)
- Bone density (Engelbert et al, 2003)

CHILDREN

BODY FUNCTIONS AND STRUCTURES / IMPAIRMENTS

- Cardiovascular fitness (Engelbert et al., 2006)
- Gastrointestinal dysmotility (Abonia et al., 2013)
- Bladder dysfunction (Pacey et al., 2015)
- Psychological – mood, self esteem and body image, sleep disturbance (Pacey et al., 2013)

Dysautonomia Symptoms

Near syncope on standing

Venous pooling with colour changes

Tachycardia – when standing and changing position

Can result in massive anxiety

Excessive heart rate on exercise

Heat intolerance

Reflux – IBS/ Slow transit

Mathias et al (2011)

Cardiovascular Sympathetic Dysautonomia

- Postural Orthostatic Tachycardia (POTS)

> 30 bpm rise in pulse adults

> 40 bpm children

Patchy dysautonomia, pooling of blood in peripheral circulation

Activation / hypersensitivity of cardiac sympathetic system

Mechanisms
Deconditioning?
Laxity?
Hormonal?
Sympathetically driven

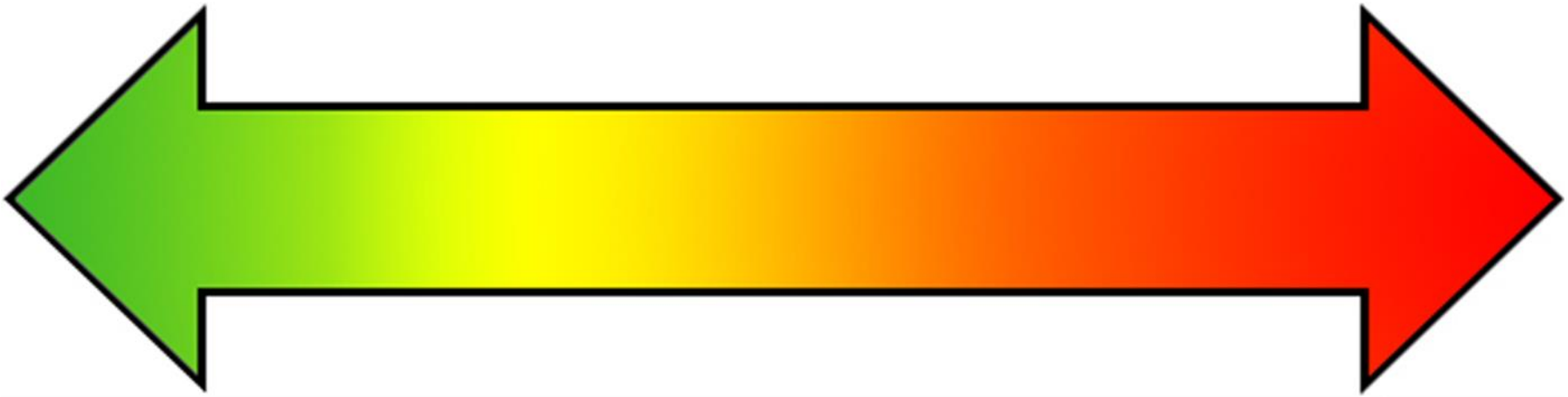
Mathias et al (2011)

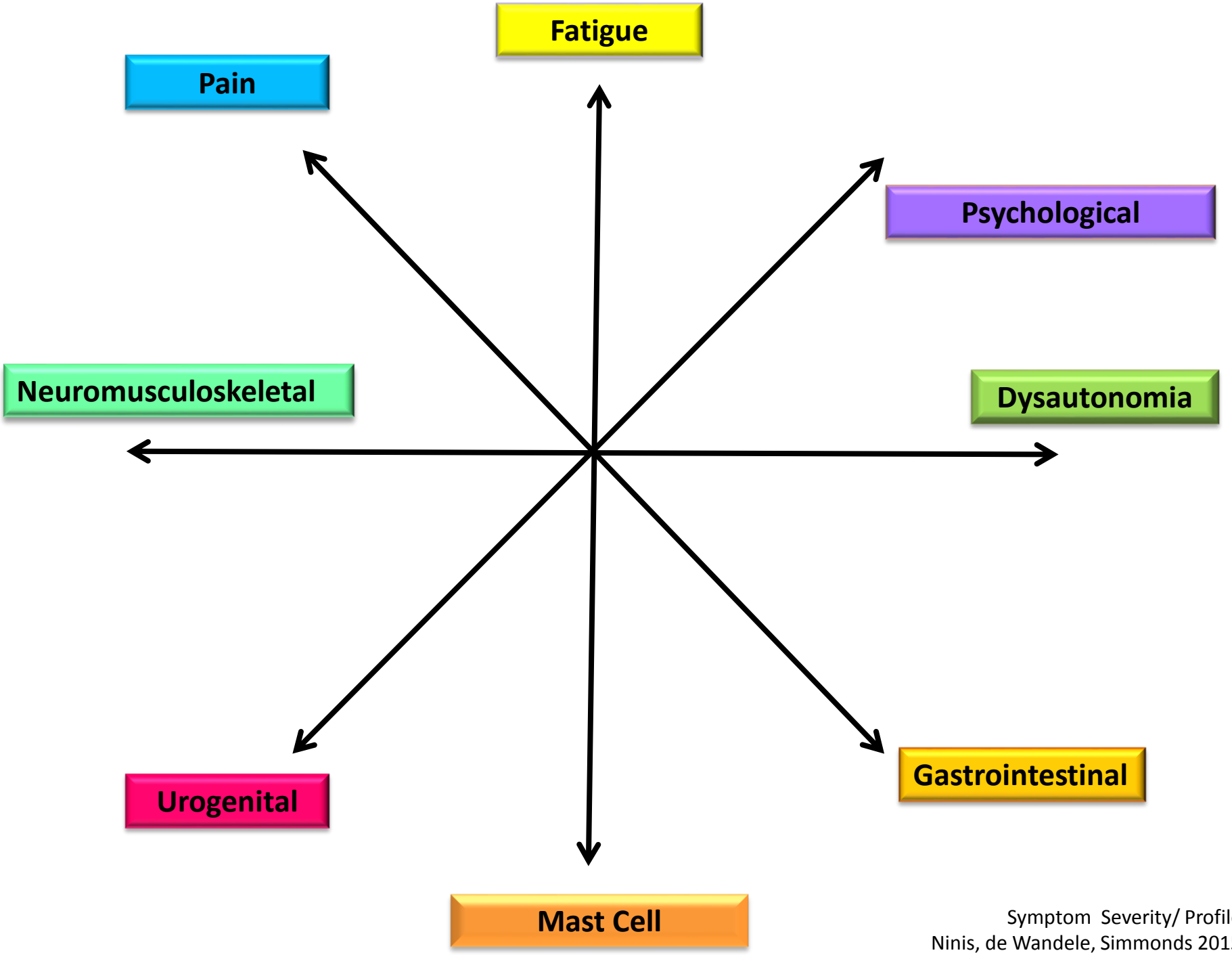
JHSD/EDS-HT - The Spectrum

SIMPLE/ACUTE

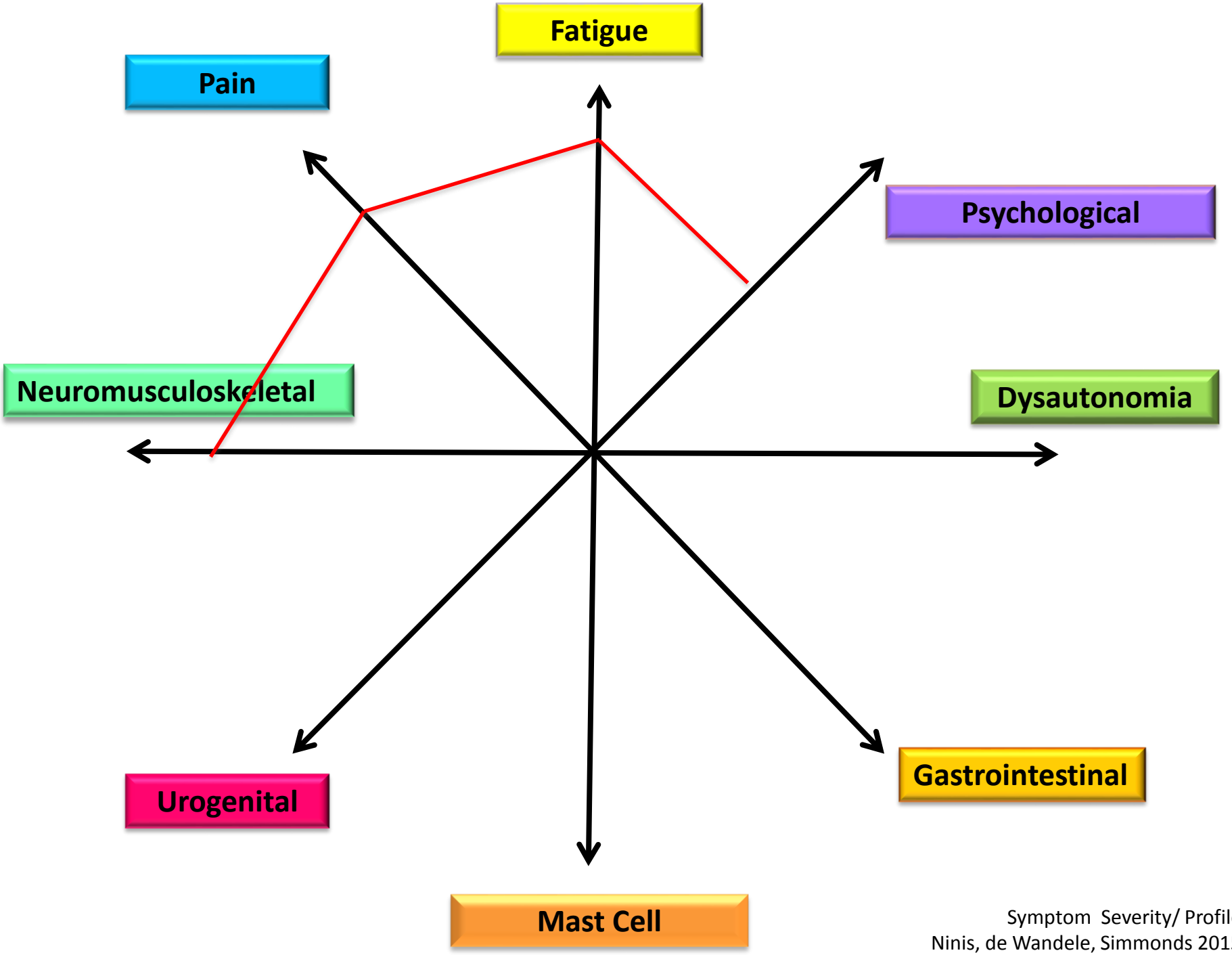
INTERMEDIATE

COMPLEX/CHRONIC





Symptom Severity/ Profile
Ninis, de Wandele, Simmonds 2015



Symptom Severity/ Profile
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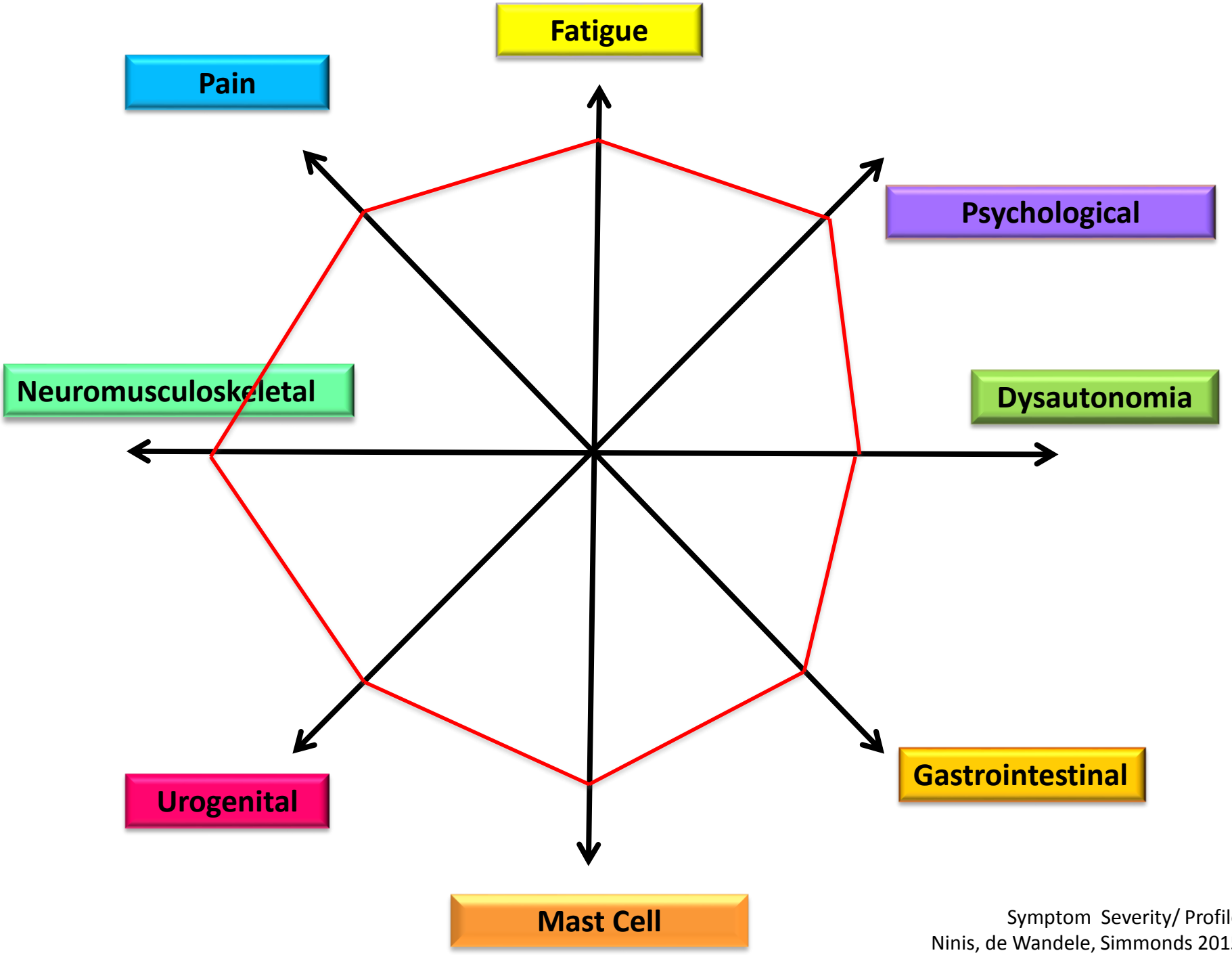


Symptom Severity/ Profile
Ninis, de Wandele, Simmonds 2015

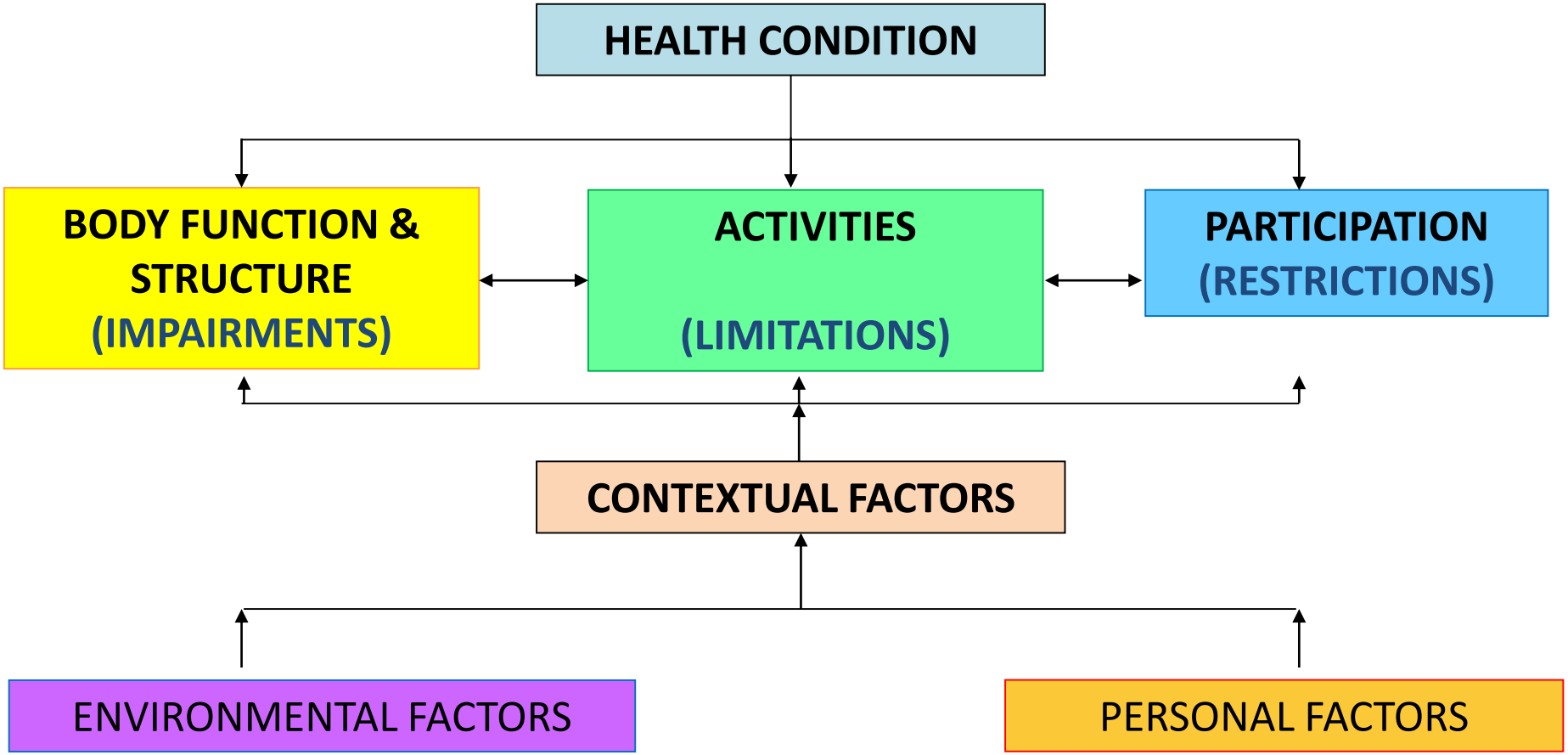
CHILDREN

ACTIVITY AND PARTICIPATION

- Mobility (Adib et al., 2005)
  - Mobility aids
- School attendance and performance (Jansonn et al., 2004; Birt et al. 2014)
  - Handwriting
- Physical education and sport participation (Jansonn et al., 2004; Birt et al., 2014)
- Increased sedentary activities (Schubert – Hajlmarsson et al., 2012)
- Impact on domestic life (Schubert – Hajlmarsson et al., 2012)
- Quality of life (Pacey et al., 2015)
  - Pain, fatigue and stress incontinence can have the biggest impact on quality of life

OUTCOME MEASURES

International Classification of Functioning Disability and Health ICF (WHO, 2015)



Which Outcome Measures?

Impairment level

- Pain (Included in PedsQL)
  - Visual analogue Scale - Local & global/ Baker Wong faces
- Fatigue VAS - (PROM)
  - PedsQL Multidimensional Fatigue Scale
    (Varni et al 2004. J Rheumatology. 31.12, 2494 – 500)
- Balance - proprioception
  - Single leg stance – eyes open/ eyes closed
  - Injury prediction Star (Y) Excursion Balance Test
    Issues with standardisation, time consuming – practice
    (Plisky et al 2006 JOSPT, 30,12, 912 – 920, Pacey et al 2013)
- Fitness/ functional capacity
  - CPET – gold standard (Expensive and time consuming)
  - Incremental shuttle walk test (need space)
  - 6 minute walk test
  - iStep (Rand et al 2015)

Which Outcome Measures?

Functional / Activity level

- Physical Activity Questionnaire (PAQ – A) (PROM)
- 9 – Item questionnaire
- Good validity and moderate reliability

- Handwriting – Detailed Ass Speed Handwriting (DASH)
- Personal Factors – Self Efficacy **(Hot off the press)**
  - Adolescent measure of confidence and musculoskeletal Performance (AMCAMP)
    - Very good psychometric properties
    - Reliability of scores on each of three clusters of items identified by factor analysis was assessed with coefficient alpha (range = 0.82 to 0.94), Standard Error of Measurement (1.38 to 2.74), and Minimum Detectable Change (3.83 to 7.6).

Kowalski, K. et al (1997) Convergent validity of the Physical Activity Questionnaire for Adolescents (PAQ-A). Pediatric Exercise Science. 9, 342 - 352

May KM et al (2016) Adolescent measure of confidence and musculoskeletal performance. IJSPT. 11(5):698-707

Which Outcome Measure shall I use?

Functional /Participatory level

PedsQL (Paediatric Quality of Life Inventory)

- Brief (23 items) and Practical (Less than 4 minutes to complete)
- Developmentally Appropriate (Ages 2-18; Child Self-Report Ages 5-7, 8-12, 13-18; Parent Proxy-Report Ages 2-4, 5-7, 8-12, 13-18).
- Multidimensional (**Physical, Emotional, Social, School Functioning**).
- Reliable (Total Scale Score: 0.88 Child Self-Report; 0.90 Parent Proxy-Report).
- Valid (Distinguishes between healthy children and children with acute and chronic health conditions; distinguishes disease severity within a chronic health condition).
- Responsive to clinical change over time
- Translated into multiple languages
- Cost implication – need a licence

Palmer 2016. Physiotherapy. In press available on online
Varni et al., 2002. Arth & Rheum. 46,714 - 725

Alternatives: Health Related Quality of Life

CHQ (Child Health Questionnaire)

- 3 domains – emotional, physical, social
- Parent version short (28) and long (56)
- Child version (87 items)
- Response to change

CHAQ (Child Health Assessment Questionnaire)

- 2 Domains Disability and Discomfort
- 8 functional tasks
- Pain scale
- Reliable and validity – Inflammatory / Autoimmune populations Juvenile Arthritis

(McErlane et al., 2013 Rheumatology, 52,11, 1941- 1951)

(Landgraf et al., 2013 Instruments for Children 3rd Edn. Laurence Erlbaum. London)

MANAGEMENT APPROACH

- Holistic, empowering, evidence based approach
- Education and reassurance
  - Sign post to patient support groups and information
- Specialist referral - multi-systemic or associated conditions

Clinically reasoned, goal directed functional restoration programme

- May or may not be alongside multidisciplinary team
- Exercise interventions **carefully implemented** based on American College of Sports Medicine Guidelines (ACPSM) recommendations and motor control theory (Faigenbaum 2009;10; Garber et al., 2011; Smidt, 2013)

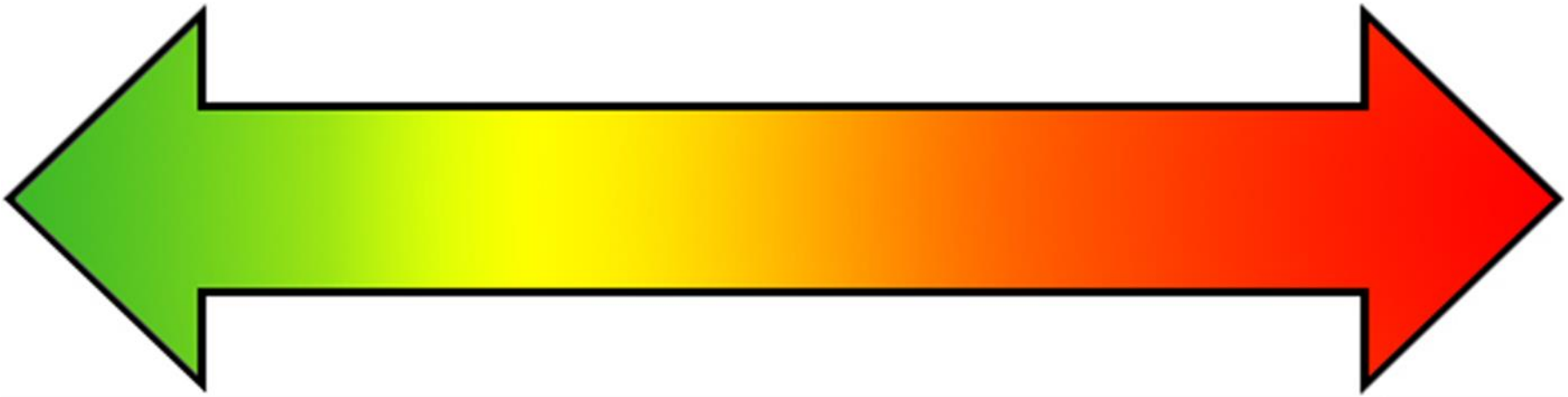
Englelbert R, Bjuul-Kristensen, Pacey V, de Wandelle I, Smeek S, Scheper M, Russek L, **Simmonds JV** (2017) The evidence-based rationale for physical therapy of children, adolescents and adults with Joint Hypermobility Syndrome/ Ehlers Danlos Syndrome - Hypermobility type. American Journal of Medical Genetics. Part C Medical Seminars

The Spectrum

SIMPLE/ACUTE

INTERMEDIATE

COMPLEX/CHRONIC



STRATIFIED MANAGEMENT

SIMPLE/ EARLY
Episode of acute musculoskeletal injury, enthesopathies, sprains, dislocation and subluxations

SIMPLE/ EARLY
Electrotherapy, support, ice, manual therapy, movement, advice, **education - prevention**

Rose 1985, Russek 2000, Macgregor et al 2005
Callaghan et al 2002, 2008, Aminaka & Gribble 2008
Christou 2004, Keer & Simmonds 2011

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Electrotherapy, support, ice, manual therapy, movement, advice, **education - prevention**

INTERMEDIATE

Recurrent episodes, series of episodes at different sites, with some deconditioning, some central/ peripheral sensitization, mild systemic conditions

Subjective - History

- Late developmental milestones
- Clumsy
- Fatigue
- Party/Cortortionist tricks
- Coordination a problems (gross and fine)
- Growing pains/restless leg
- Easy bruising
- Subluxations and dislocations
- Adolescence – biomechanical and hormonal changes
- ‘Clicking’
- Family history – wider family
- Slow tissue healing and unresolved injury ***
- Poor response to analgesics – dentist/ epidural
- *Gastro intestinal dysmotility*
- *Dizziness/fainting and syncope/reflux*
- *Allergies, rashes*

Subjective Examination

Problems – prioritise

- Pain
- Joint instability – subluxations, dislocations, clicking
- Fatigue
- Coordination problems **
- Anxiety
- Low mood/ depression
- Gastrointestinal dysmotility and pain
- Dysautonomia - PoTS
- Bladder problems
- Allergies

Explore

Physical activity/ Physical Education

Social, School, University, Work

General health

Blood pressure, dizziness, palpitations, drop attacks, chest pain, abdominal symptoms bowels, changes in skin, pooling of blood, profuse sweating, bladder, bladder, allergies, rashes

- Family history and thorough developmental history **

Objective Assessment - Functional

Joint range and quality

Soft tissue laxity and tone

- Posture – static and dynamic
  - compensatory patterns
- Gait
- Heal raise
- Sit to stand/ squat
- Balance
- Repositioning
- Strength/ activation
- Cardiovascular

Principles of functional rehabilitation programme

Condition specific education, advice & support

www.HMSA.org www.ehlers-danlos.org www.potsuk.org

Time to listen reassurance, promote self efficacy - Readiness for change; Motivational interviewing (Simmonds 2003; Prochaska & Di clementi 1993)

Pain management & pacing (Harding 2003)

Manual therapy - muscle spasm, neurodynamics, joint mobilisation (Sahrmann 2002)
Acupuncture
TENS

Stretching (Harding 2003)

Postural control (Kendall et al 1993, Booshanam et al 2010)

Motor control & graded exercise (Rousell et al 2009, Boudreau et al 2010, Palmer et al 2013;Pacey et al 2013)

- specific motor-skill training, pain-free, goal-orientated/cognitive, quality>quantity

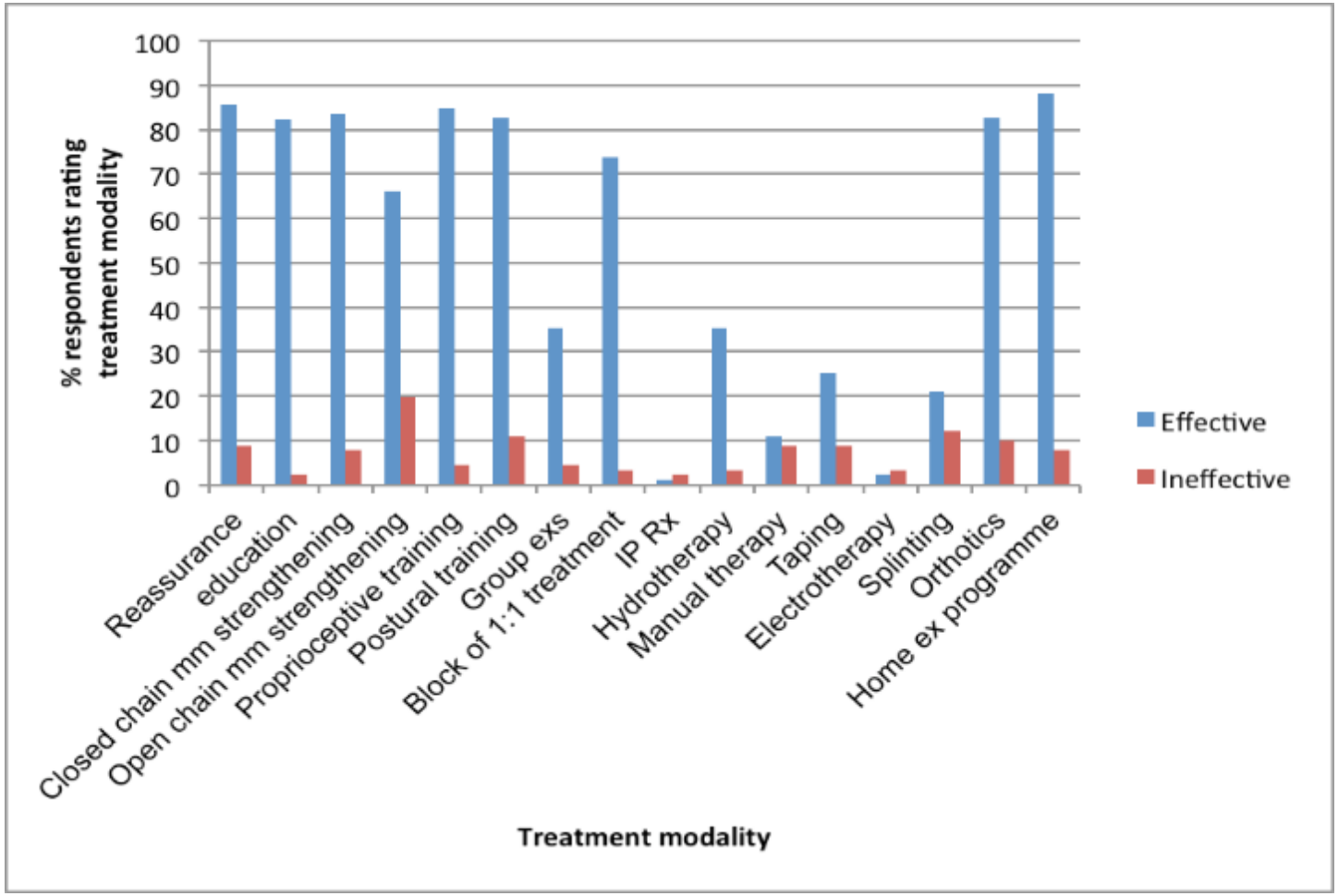
Proprioception and balance – closed chain into hypermobile range (Ferrell et al 1004, 2007, Sahin et al 2008,)

General fitness/ sports/performance specific – strength - endurance - cardiovascular

Simmonds & Keer 2007, Keer & Simmonds 2011

National survey

Treatment modalities considered effective by **paediatric physiotherapists**



Billings, Deane & Simmonds (2015), Physiotherapy Practice and Research

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SIMPLE/ EARLY

Electrotherapy, support, ice, manual therapy, movement, advice, **education - prevention**

INTERMEDIATE

Recurrent episodes, series of episodes at different sites, with some deconditioning, some central/ peripheral sensitization, mild systemic conditions

INTERMEDIATE

Physiotherapy modalities have temporary effect, no effect or exacerbates. Modalities modified/adapted

Functional Restoration

Opinion/ anecdotal**

MANAGEMENT - CHILDREN

RHEUMATOLOGY

Rheumatology 2010;49:315–325
doi:10.1093/rheumatology/kep362
Advance Access publication 30 November 2009

Original article

A randomized comparative trial of generalized *vs* targeted physiotherapy in the management of childhood hypermobility

Sue Kemp[1,*], Ian Roberts[1,*], Carrol Gamble[2], Stuart Wilkinson[1], Joyce E. Davidson[1], Eileen M. Baildam[1], Andrew Gavin Cleary[1], Liza J. McCann[1] and Michael W. Beresford[3]

6 week graduated exercise intervention

* Improvements in pain – child and parent perspectives
- Parental global assessment reported better outcomes with a targeted motion control approach

MANAGEMENT - CHILDREN

Exercise in children with joint hypermobility syndrome and knee pain: a randomised controlled trial comparing exercise into hypermobile versus neutral knee extension

Pacey *et al.*

BioMed Central

Pacey *et al. Pediatric Rheumatology* 2013, **11**:30
http://www.ped-rheum.com/content/11/1/30

8 week graduated exercise intervention

* Improvements in knee strength and pain in both groups
* Parent reported - psychological health, self esteem , mental health and behaviour was significantly different in favour of exercising into the hypermobile range

MANAGEMENT > 18(!)

Rheumatol Int (2008) 28:995–1000
DOI 10.1007/s00296-008-0566-z

ORIGINAL ARTICLE

Evaluation of knee proprioception and effects of proprioception exercise in patients with benign joint hypermobility syndrome

Nilay Sahin · Akin Baskent · Aysegul Cakmak · Ali Salli · Hatice Ugurlu · Ender Berker




8 week graduated proprioception, balance and plyometric training

* Reduced knee pain and improved proprioception

MANAGEMENT - CHILDREN

Orthotics and footwear?

Assessment of gait characteristics and orthotic management in children with Developmental Coordination Disorder: Preliminary findings to inform multidisciplinary care

Stewart C. Morrison [a,*], Jill Ferrari [a], Sally Smillie [b]

[a] School of Health, Sport and Bioscience, University of East London, United Kingdom
[b] Medway Community Healthcare, United Kingdom

Research in Developmental Disabilities 34 (2013) 3197–3201

* Improved gait efficiency

Evans & Rome 2011 Cochrane Review of evidence for non surgical intervention for flexible flat feet. Eur J Phys Rehab Med. 47 (1): 69 - 89

* Judicious use of orthotics or sensible footwear

MANAGEMENT - CHILDREN

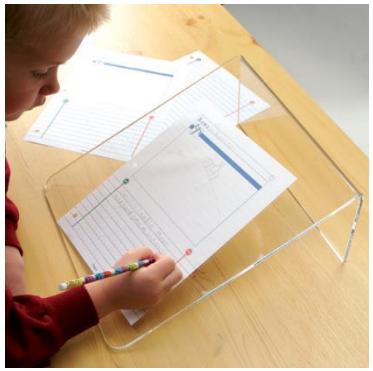
Splinting?

Frolich et al., 2011 Physical & Occupational Therapy in Paediatrics 32(3):243–255

* Splints not effective for hand pain or writing speed

Expert opinion - Judicious use **


0.5kg
0.5kg
5LB
2LB
2LB



MANAGEMENT of PoTS

Dysautonomia – Postural Tachycardia Syndrome (PoTS)

- Reassurance
- Advice – fluids, electrolyte, compression tights
  - Positioning, anti syncope manoeuvres
- Monitoring of medications when prescribed (Midodrine, Fludrocortizone, Beta blockade)
- Respiratory physiotherapy – hyperventilation
- Anxiety management – psychological support
- Graded cardiovascular exercise and resistance training – focus on lower limbs
- Recumbent to upright

* **Incorporating** exercise to manage joint instability

Mathias et al., 2011
Fu et al., 2011
Jarjour 2013

Clinical expert opinion **

20 Point Borg Scale

RPE
Rate of Perceived Exertion

POINT	EFFORT	DESCRIPTION	% OF MAXIMUM HEART RATE	√
6	No Exertion	Little to no movement, very relaxed	20%	
7	Extremely Light	Able to maintain pace	30%	
8			40%	
9	Very Light	Comfortable and breathing harder	50%	
10			55%	
11	Light	Minimal sweating, can talk easily	60%	
12			65%	
13	Somewhat Hard	Slight breathlessness, can talk	70%	
14		Increased sweating, still able to hold conversation but with difficulty	75%	
15	Hard	Sweating, able to push and still maintain proper form	80%	
16			85%	
17	Very Hard	Can keep a fast pace for a short time period	90%	
18			95%	
19	Extremely Hard	Difficulty breathing, near muscle exhaustion	100%	
20	Maximally Hard	STOP exercising, total exhaustion		

Source: Gunnar Borg, PhD, M.D.

STRATIFIED MANAGEMENT

SIMPLE/ EARLY
Episode of acute musculoskeletal injury, enthesopathies, sprains, dislocation and subluxations

SIMPLE/ EARLY
Electrotherapy, support, ice, manual therapy, movement, advice, **education - prevention**

INTERMEDIATE
Recurrent episodes, series of episodes at different sites, with some deconditioning, some central/ peripheral sensitization, mild systemic conditions

INTERMEDIATE
Physiotherapy modalities have temporary effect, no effect or exacerbates.
Modalities modified/adapted
Functional Restoration

COMPLEX LONG TERM
Chronic, longstanding, severe, unremitting pain with profound deconditioning/ comorbidities, disability (Rhombaut 2011; Scheper, 2016)

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COMPLEX/ LONG TERM
Multi disciplinary management programme using cognitive behavioural approaches. (Finsterbush & Pogrund 1982; Bathen et al, 2014)

Examples Cases

Meet Sam
5 years

Hx: late walker 19 months
Talked late

I wanna play with the other kids

IMPAIRMENTS

Joint hypermobility 9/9 + hips & shoulders

Everted ankles/over pronated, flat feet

Coordination problems (gross and fine motor)

Poor balance – unable to hop

Low muscle tone

Muscle weakness

Generalised leg pain

Tired

Slow gut motility

ACTIVITY

Not keeping up with peers
Unable to walk for > 10 mins
Struggling to run
Unable to hop
Struggling to throw and catch
Unable to ride a bike/ tricycle
Struggling with dressing
Struggling with pencil skills

PARTICIPATION

Struggling at nursery
Parents nervous about Sam starting school

PERSONAL FACTORS

Male
Low confidence/ self esteem

ENVIRONMENT

Supportive family
Very sport father and sister

Meet Sam
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Hx: late walker 19 months
Talked late

Which Outcome Measures?

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Therapeutic Approach

- Consider Developmental Coordination Disorder/ Hypermobility Spectrum Disorder
  - Education – patient, family & school
- Rehab in the form of play
- Top down – functional approach
  - Practice
- Bottoms up – strengthening/stability/ fitness

Agree and set goals

1. Learn to throw and catch
2. Learn to ride a bicycle
3. Improve pencil skills
4. Learn to tie laces independently

Reduce anxiety
Improve quality of life
Improve self efficacy/ confidence

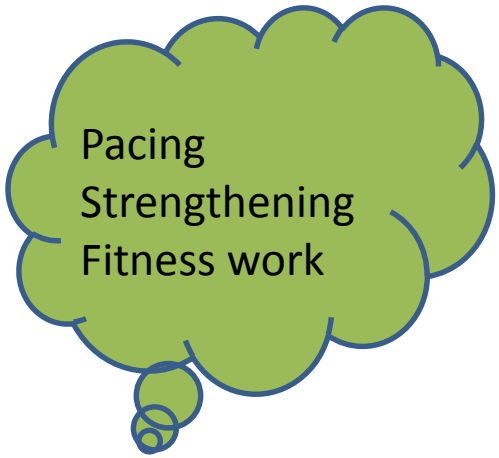
Mum concerned also about pain and fatigue

Task

Physical requirements

Deconstruct the task into mini tasks/goals

- Throwing and catching
- Riding a bike
- Tying shoe laces
- Pencil skills



Consider MATCH approach

- M: Modify the task
- A: Alter expectations
- T: Teach strategies
- C: Change the environment
- H: Help understanding

Advocated by canchild

https://canchild.ca/en/resources/123-m-a-t-c-h-flyers-a-resource-for-educators

Top Tips

Break down task

Take care with communication, make sure the child understands

Repetition/ practice helps

Achievable goals – feedback/feedforward

Circuits and practice

Strengthening….fitness

Make it fun!

American Psychiatric Association (2000). Diagnostic and Statistical Manual of Mental Disorders. (4th ed.) Washington, DC: Author.

https://canchild.ca/en/resources/126-children-with-coordination-difficulties-a-flyer-for-physical-educators

Meet Helen
14 years

I want to be a dancer/ actor

IMPAIRMENTS	ACTIVITY	PARTICIPATION
Widespread hypermobility ++ Recurrent shoulder subluxations, fingers collapse, low back pain Widespread persistent pain and fatigue Anxious Low mood Early satiety when eating/ bloating, nausea, vomiting and slow transit constipation	Unable to dance and struggling to act Struggling with eating Unable to travel on public transport	Reduced attendance at & having to modify college Reduced social activity with friends

PERSONAL FACTORS	ENVIRONMENT
Female High achiever – A student Depressed Low confidence/ self esteem	Protective family 2 siblings Mother with EDS

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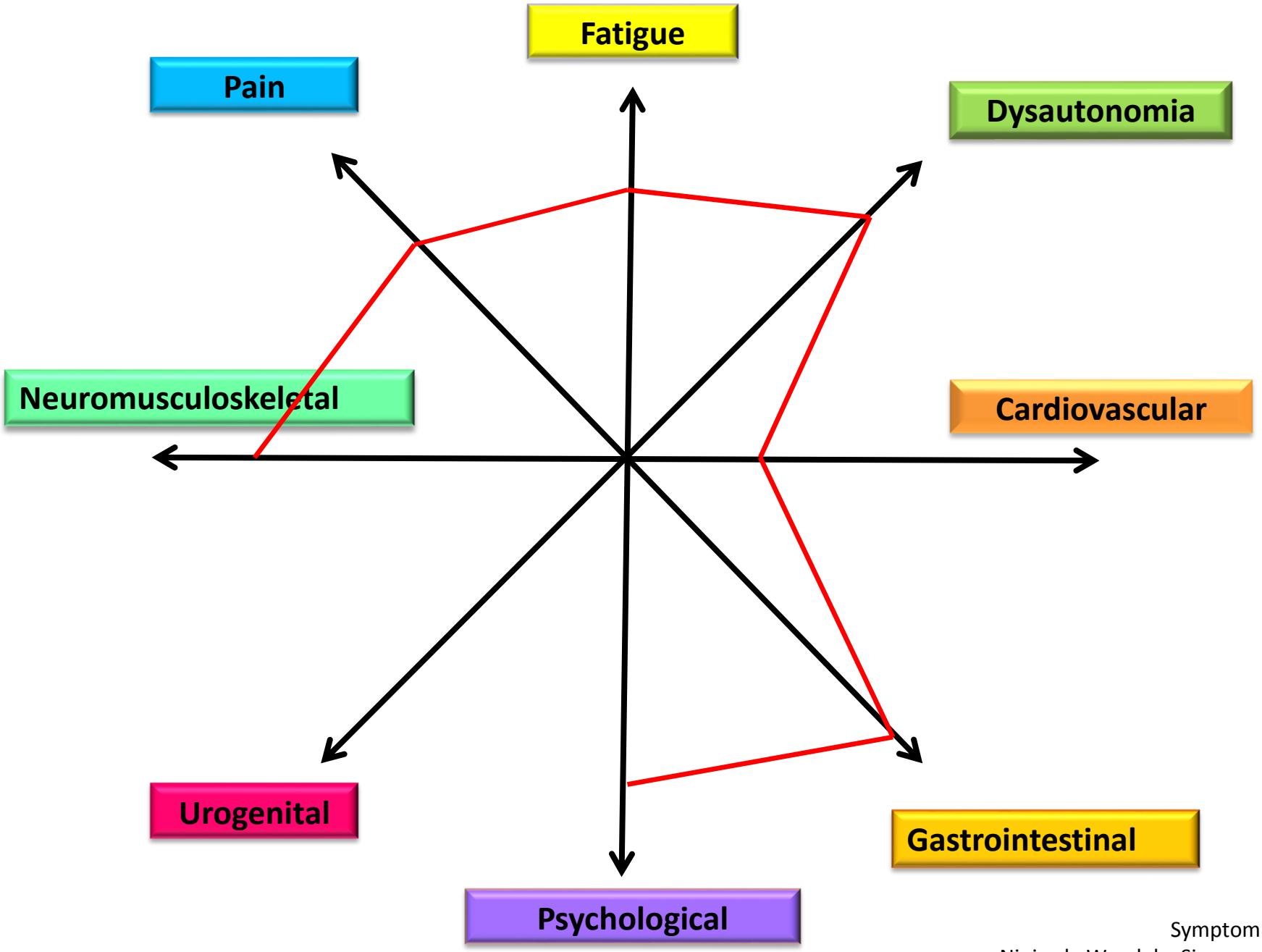
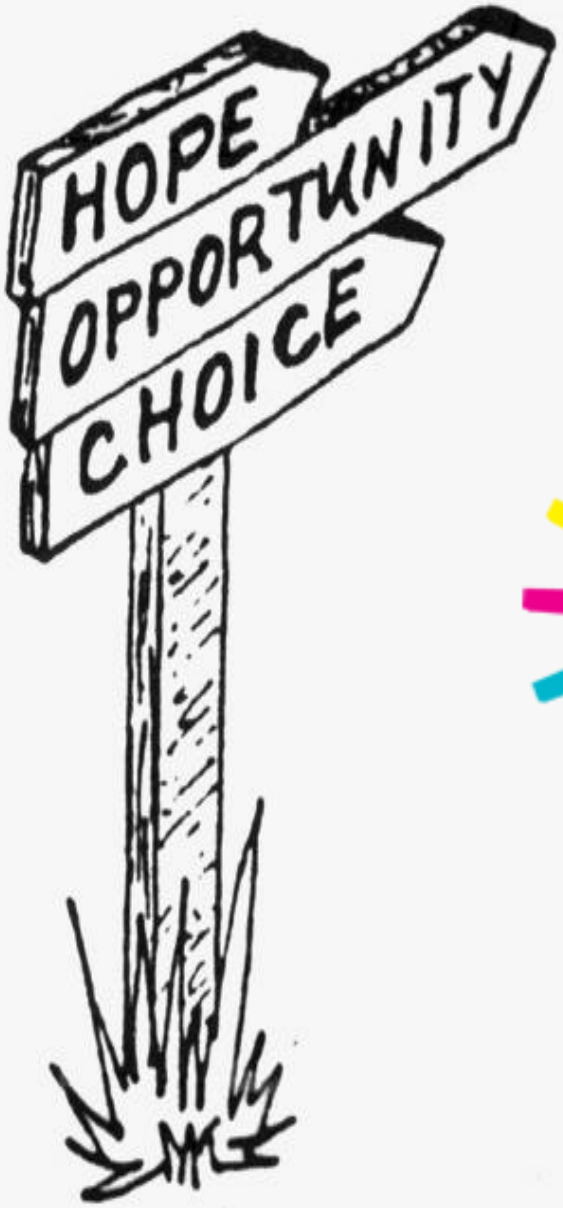


Symptom Profile
Ninis, de Wandele, Simmonds 2015

Therapeutic Approach

- Education
- Tertiary referral
- GI distress
- Mood - psychology
- Manage pain – visualisation, psychology
- Pacing
- Address musculoskeletal issues



CAMHS

Child & Adolescent Mental Health Services

I want to be an actor/dancer

I want to go to drama school

Meet Beth

16 years

Hx: Motor vehicle accident 18 months ago – whiplash. Previously well.

I want to go to university, wants to go to concerts, wants to be a photo be a journalist

IMPAIRMENTS

Joint hypermobility 6/9 +
Widespread pain

Fatigue

Head aches

Brain fog

Deconditioned ++

Muscle weakness

Slow transit constipated

Fatigue and sleeps poorly

ACTIVITY

Unable to walk for > 5 mins
Uses wheel chair

PARTICIPATION

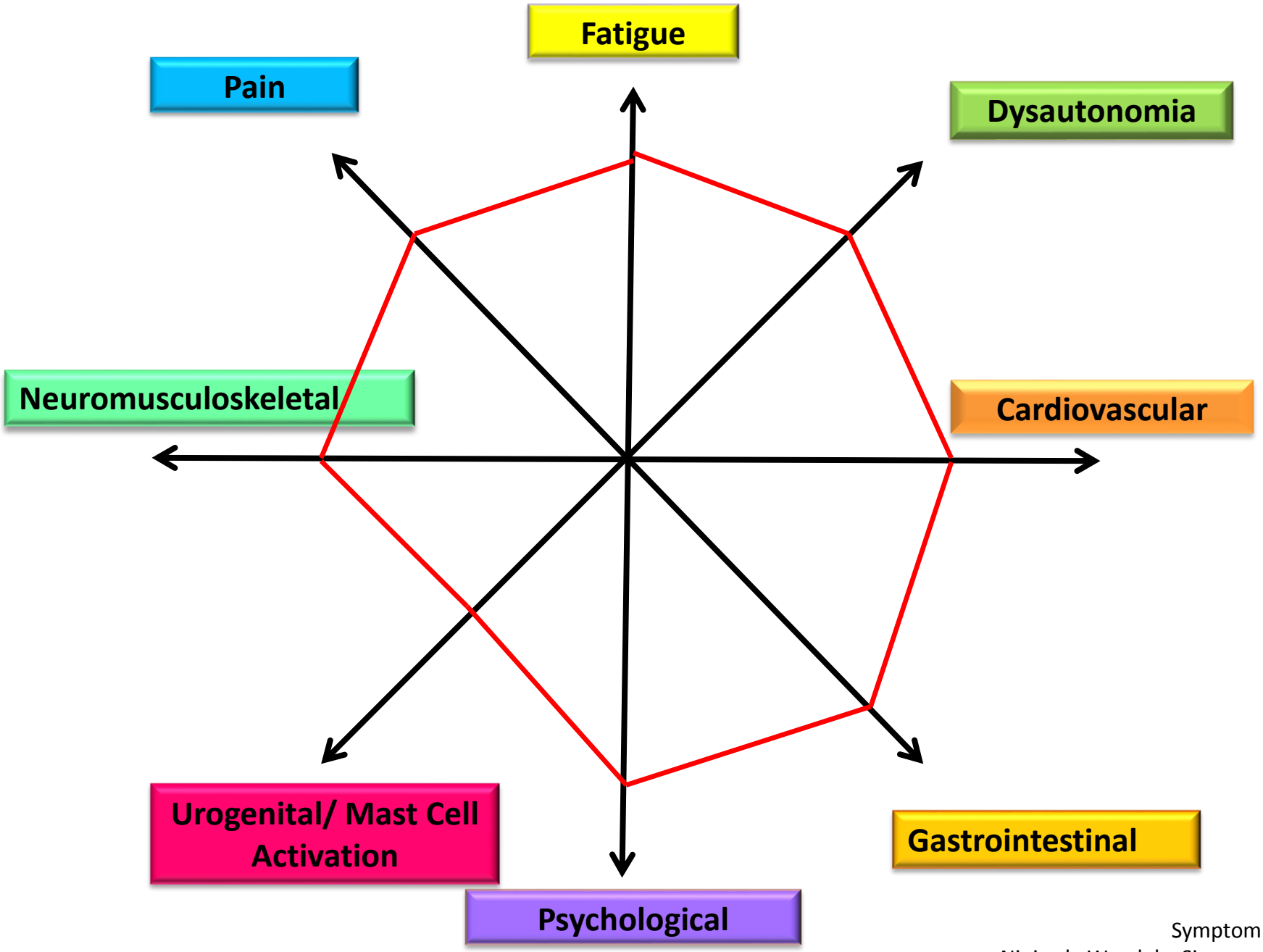
Unable to go to school
Socialising only on social media only

PERSONAL FACTORS

Female
Depressed
Loves writing/ reading/ photography/ animals
Tends to “boom & bust”

ENVIRONMENT

Supportive family



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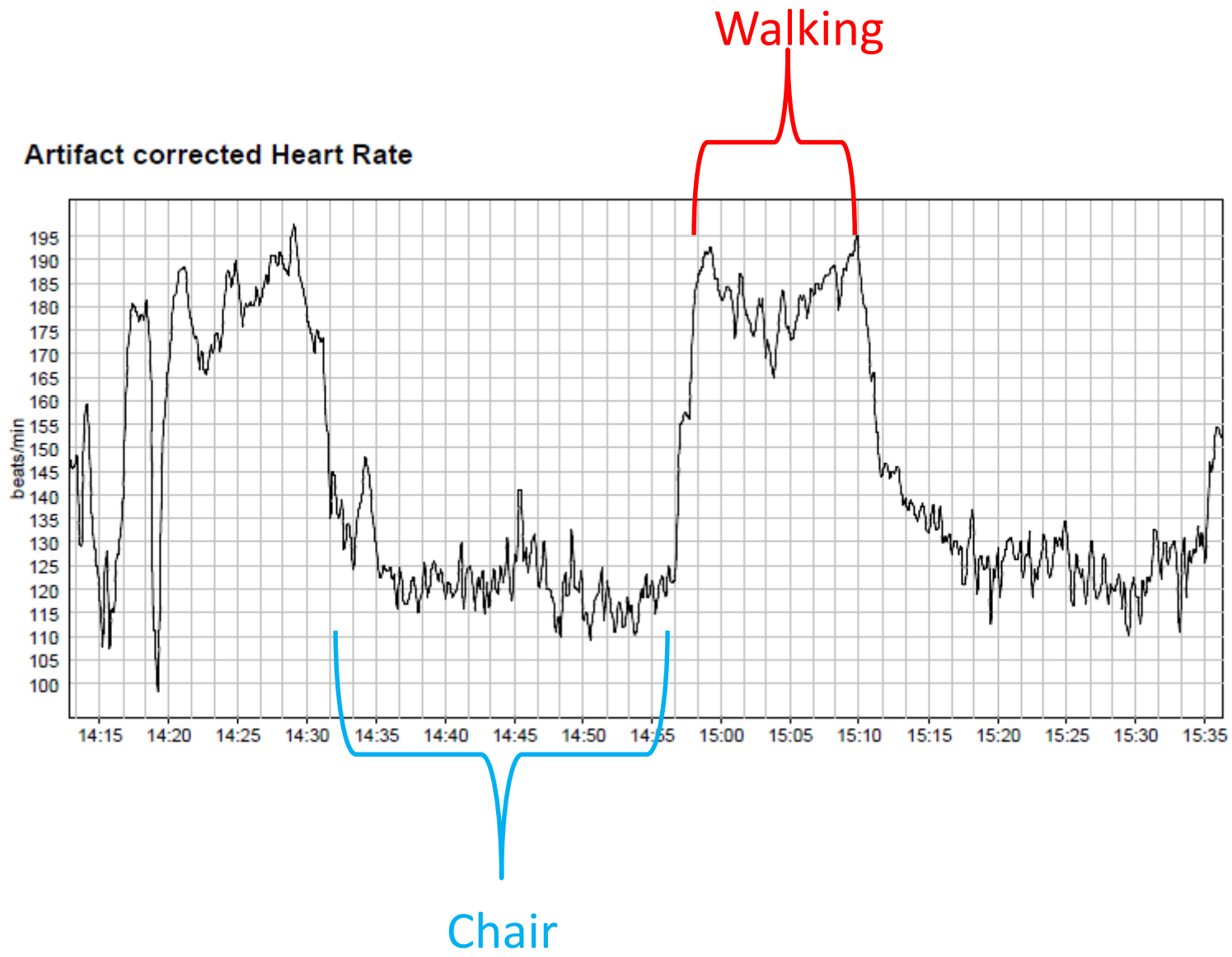
Walking
Artifact corrected Heart Rate
beats/min
195
190
185
180
175
170
165
160
155
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145
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135
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125
120
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15:00
15:05
15:10
15:15
15:20
15:25
15:30
15:35
Chair

PoTS / EDS Functional Rehabilitation

- Medical management
  - Fludrocortizone, Midodrine, B Blockers
- Non pharmacological management
  - Fluids, salt, syncope strategies
  - Pacing/ sleep hygeine
  - Breathing exercises
- Graded exercise
  - Morning regime
  - Cardiovascular (5 per week)
    - Working towards 30 minutes
    - 40-50% MHR ⟶ 60-70% MHR (meaningful)
    - Recumbent to vertical
    - Fun and relevant
  - Muscular strength/endurance (3 times per week)
  - Joint stability/ motor control programme (daily)
- Manual therapy

MAYO Patient information. 2009
Ferrell et al. Arthritis Rheumatism. 2004, 50, 3323-3328
Hauser et al. Arthritis Research & Therapy. 2010, 12, R79
Fu et al. Hypertension. 2011. 58, 167-175
Pacey et al. Paediatric Rheumatology. 2013, 11, 1-9
Mathias et al 2012

Beth’s Video

Top Tips

- Make a plan – **give hope**
- Decide what needs to change…family and individual
- Goals (realistic)
- Routine – nutrition, hydration, exercise, fun, pacing, daily relaxation, stay optimistic
- Strategy for set backs
- Share positive experiences
- Celebrate achievements
- Realise life isn’t perfect …..but can be fun

Educational resources

Www.hypermobility.org

Www.ehlers-danlos.org

Mayo Clinic leaflet

POTS UK
POSTURAL TACHYCARDIA SYNDROME

www.potsup.org

Thank you
Questions

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