



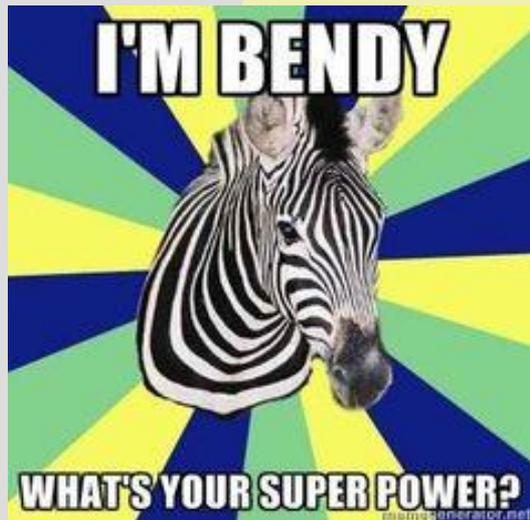
Wits University  
Donald Gordon  
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MEDICLINIC 



# Hypermobility Spectrum Disorder: treatment considerations



Dr Gail Faller  
Donald Gordon Medical Centre

# Conflicts of interest



- NONE

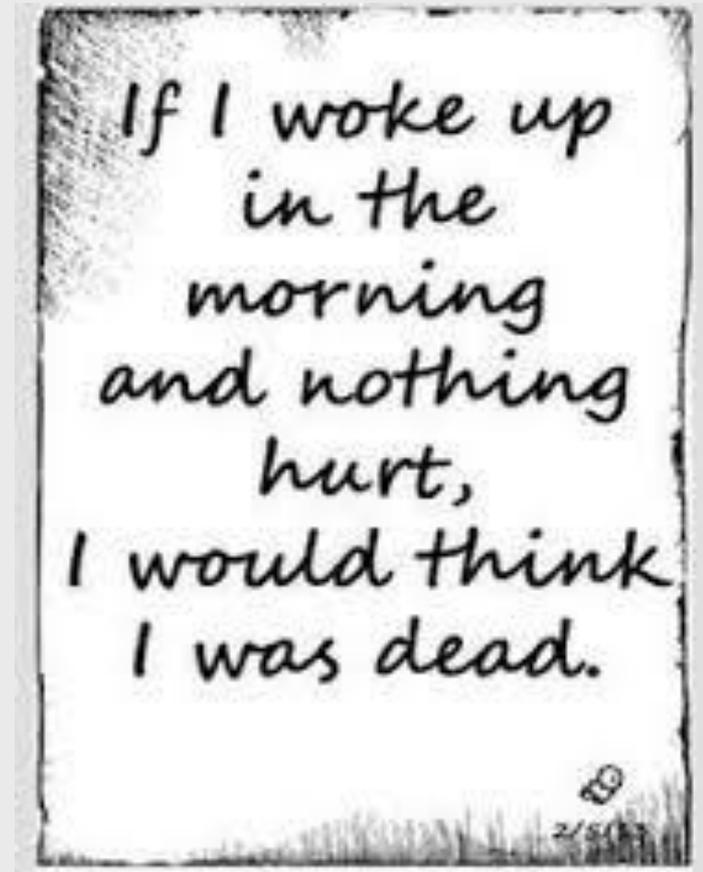
# Joint Hypermobility Syndrome

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Rodney Grahame, CBE, MD, FRCP<sup>b,\*</sup>

It is not a trivial articular problem occurring in healthy individuals; it is now recognized as a multisystemic disorder and a major source of chronic widespread pain, dysautonomias, and gastrointestinal dysmotility. It is a neglected area within rheumatology

# Definition: hypermobility spectrum disorder (HSD)

- Flexibility beyond normal for age and activity
- Associated clinical problems including pain
- Impacts on daily life



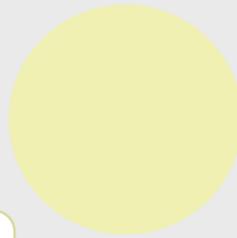
# Major problems needing treatment

- PAIN, PAIN, **PAIN**
- Autonomic dysfunction
- GIT dysmotility

# Other problems



- Tendonitis and recurrent sprains
- Fatigue
- Anxiety
- Migraines
- Infections
- Poor healing and scars
- Proprioception problems



Physio/  
Bio

school

OT



family

podiatry

Gastro

Psych

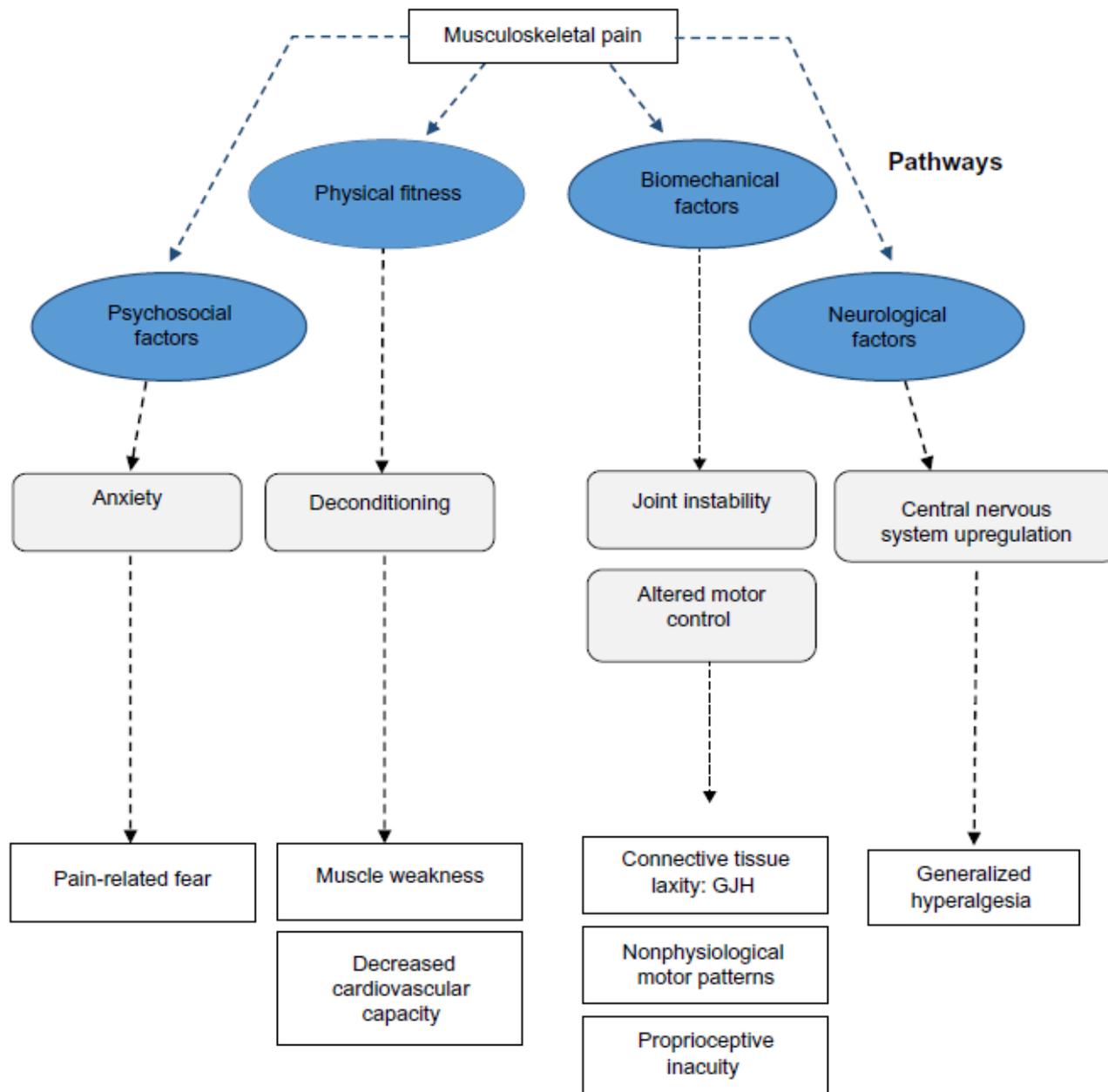


**Function  
Despite  
Pain**



**Fitness  
is  
your  
friend**

*The Rheumatologist, May 2010*



ARTICLE

# The Evidence-Based Rationale for Physical Therapy Treatment of Children, Adolescents, and Adults Diagnosed With Joint Hypermobility Syndrome/Hypermobile Ehlers Danlos Syndrome

RAOUL H.H. ENGELBERT \*, BIRGIT JUUL-KRISTENSEN , VERITY PACEY,  
INGE DE WANDELE, SANDY SMEENK, NICOLETA WOINAROSKY, STEPHANIE SABO,  
MARK C. SCHEPER, LESLIE RUSSEK, AND JANE V. SIMMONDS

*Beyond the Beighton scale,  
other assessment measures  
should be utilized within each  
domain of the ICF and  
clinical reasoning should  
underpin where appropriate,*

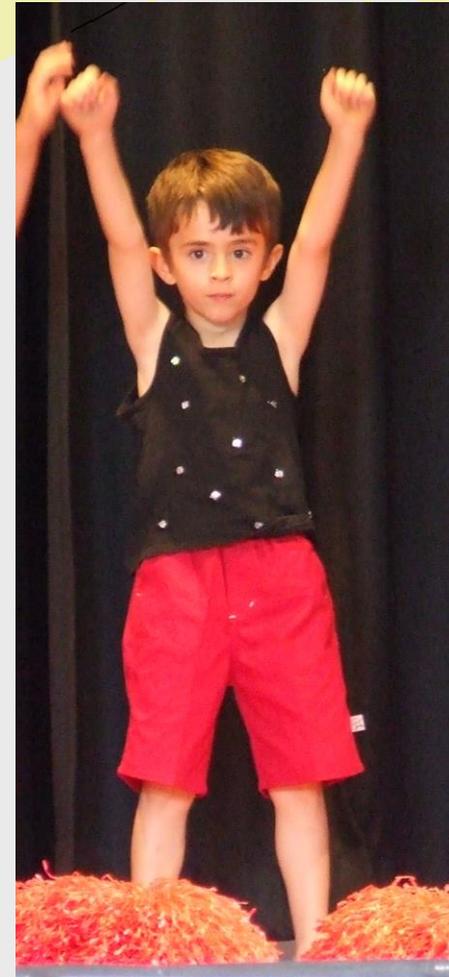
# Treating pain



- Improve the physical condition
- Avoid opioids: use NSAID, Panado, muscle relaxants
- Use ice, strapping, massage, rehab injuries promptly
- DO NOT Immobilise for long periods
- ? TENS machine

*Kemp et al. performed the first prospective randomized controlled trial (RCT) in children comparing a 6-week generalized program, improving muscular strength and fitness, with a targeted program aimed at correcting motion control of symptomatic joints. It was demonstrated in 57 children that significant improvements in both the children's and parental pain scores were reached in both groups.*

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# Fatigue: Multiple factors

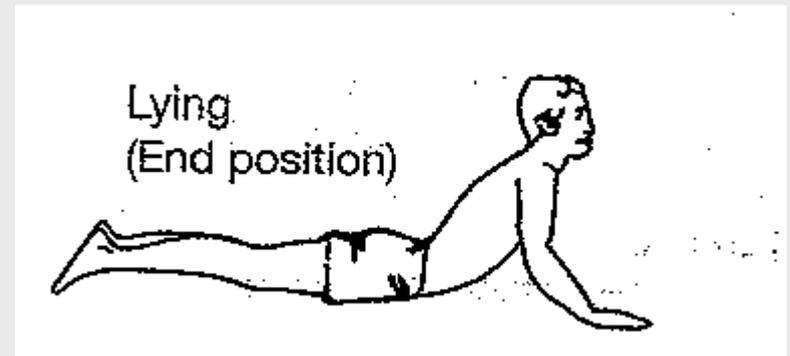
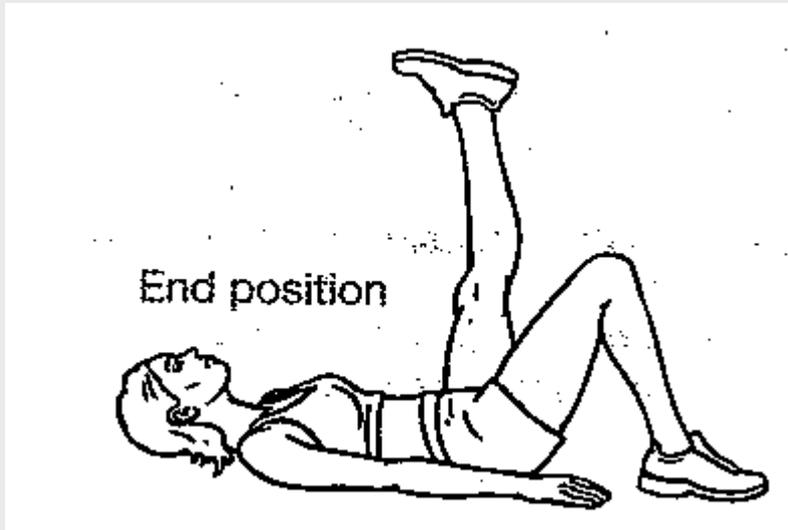
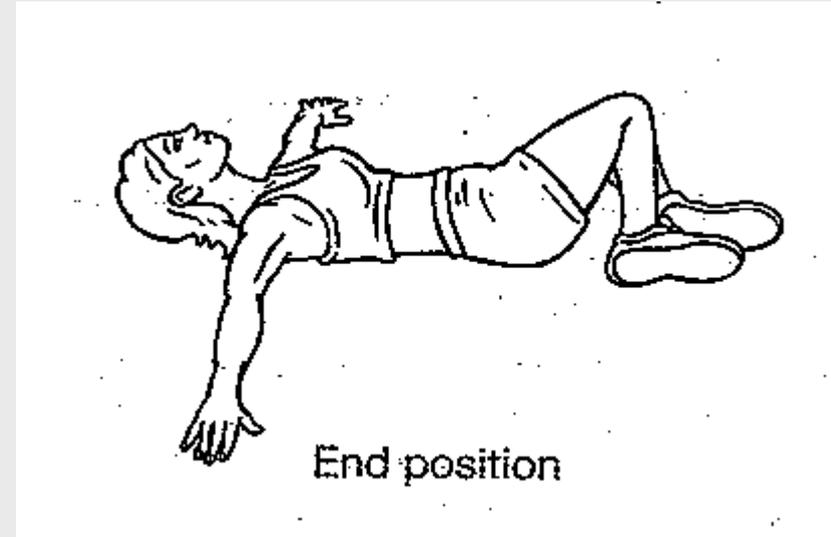


- Muscle weakness
- Respiratory insufficiency
- Unrefreshing sleep
- Dysautonomia
- Intestinal malabsorption
- Depression/anxiety
- Excessive use of analgesics

*Fatigue symptoms are heterogeneous in nature and can vary from mild to severe. However, patients often report fatigue symptoms as the most disabling complaint.*

# Other considerations

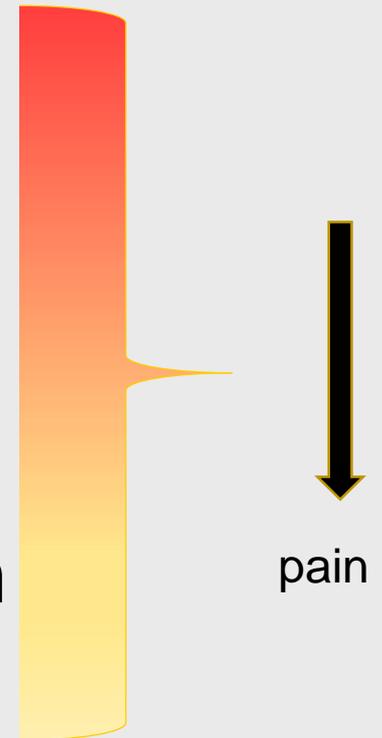
Daily stretches are very important importance.  
The thoracic spine is the first place to stiffen in adolescents



# Physical therapy



- Addresses weakness
- Core most important
- Improves tone
- Improves fitness
- Better balance and proprioception
- Improved sleep



# Sports

- Swimming
- Dancing
- Martial arts



- Rackets sports for upper limb strength
- Avoid: rugby, judo, high impact gymnastics

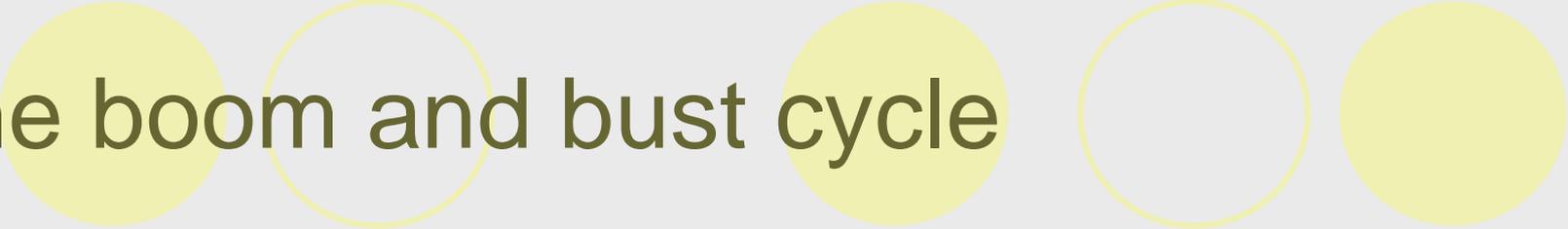
# Occupational therapy

- Hand function
- Hand strength
- Proprioception
- Accelerated fine motor and balance
- School skills

*Decreased muscle strength is associated with activity limitations in JHS/hEDS patients. Joint proprioception has been found to influence this association and should be considered in the development of new treatment strategies for patients with JHS/hEDS.*

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# The boom and bust cycle



- Especially prevalent in children
- Short cycles      long cycles
- Periods of hyperactivity followed by periods of being unable to function
- Pacing is key
- During the bust: stretching

# GIT problems



- Treat the constipation even when it is “not too Bad”
- Untreated constipation leads of bowel dysfunction
- Take food sensitivity seriously: test for coeliac disease
- Encourage pelvic floor exercises for improved urinary function.

# The Gastroenterologist



- Scope for rule out ulcers
- Low grade colonic inflammation is common
- Test for helicobacter
- Treatments for IBS are often helpful
- Reflux treatment

# Treating neuropsychiatric disease

- Limit medication for sleep
- Melatonin
- Trepiline
- Short acting benzodiazepines
- Anti-anxiety medication
- Anti-depressants



# Dysautonomia

- Improved by physical conditioning
- Avoid drugs and triggers—heat, dehydration
- Use pressure garments to avoid venous pooling

*Symptoms can be highly debilitating in hEDS. There is recognition of an association of POTS with fatigue, reduced quality of life, and a greater incidence of migraine and syncope with POTS in patients with joint hypermobility syndrome compared to those without.*

## **Cardiovascular Autonomic Dysfunction in Ehlers–Danlos Syndrome—Hypermobile Type**

ALAN HAKIM,\* CHRIS O'CALLAGHAN, INGE DE WANDELE, LAUREN STILES, ALAN POCINKI, AND PETER ROWE

# Drug therapy



- Fludrocortisone for orthostatic Hypotension
- B-blockers may help recurrent syncope/tachycardia
- Hormonal medications may help young women who worsen during menstruation
- Desmopressin may help those with nocturia who dehydrate overnight



Cont..

- Pyridostigmine may help postural tachycardia (POTS)
- Clonidine for anxiety
- Methylphenidate (Ritalin/Concerta) for attention, also vasoconstricting
- SSRI
- Reflux meds



Cont..

- Midodrine increases BP, not available in South Africa
- Low dose naltrexone
  - Controversial in medical circles
  - But established in EDS literature for pain

# Archives of diseases in Childhood

## January 2015

- “Research into childhood joint hypermobility is singularly lacking, yet the problem seems to be increasing and poses difficulties for specialists and generalists alike”

Armon K. Arch Dis Child 2015;100:2–3

# Conclusion



- HSD can be a cause of major morbidity
- More research is needed to determine the size of the problem and outcomes.
- Evidence based approaches to management lacking
- Continuous support, physical therapy are most important

Thank-you

