

Pain, distress and hypermobility

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Outline

- Case presentation
- Definition of hypermobility
- Rehabilitation intervention for musculoskeletal pain
- Exercise advice/prescription for hypermobility
- Short review of other associated problems with hypermobility

Case presentation

- F/25
- Pain in elbows, wrists, knees, ankles and toes since 2003, at age of 20
- Also neck and back pain
- Each attack lasts for few days and subsided spontaneously
- Symmetrical pattern
- No joint swelling or redness
- No morning stiffness
- Pain is aggravated after she has exercise

Pain history

- No trigger point or tender spots
- Pain is widespread
- Pain sites are around joints and muscle insertions
- Big muscles, quad, biceps, deltoids are also painful
- Could be quite distressing, affecting her sleep and work
- Pretty normal between attacks

- She is seeing pain specialist for her diffuse pain
- Both CBT and pharmacological treatment have been prescribed

Drug history

1. Voltaren gel
2. Mohrus (ketoprofen) Patch 2%
3. Lyrica 25mg nocte

Past health

1. Bilateral hallux valgus with left osteotomy done in New York in 3/07
2. Mild PID C3-6 (by MRI cervical spine) without clinical neurology
3. Mild S-shaped scoliosis
4. Amenorrhoea for many years

No history of spontaneous pneumothorax or hernia

Social history

- American Chinese, lives and studies in US before her 20s
- Come back to HK to work, now lives alone
- She is a writer, flexible and yet long working hours
- Significant work stress
- Non-smoker
- Non-drinker
- Non-drug abuser

P/E

- She is thin, short stature, 1.56m
- Scoliosis+
- Hallux valgus on right side, left side has osteotomy scar
- Bilateral pes planus
- Beighton score 9/9, Arm span to body ratio <1
- No clinical synovitis
- Spine movement is normal
- Systems are NAD. No inguinal hernia

- No blue sclera
- No high arched palate
- No pectus carinatum/excavatum

Physical exam





Investigations

- CBP, ESR normal
- ANA, ENA negative
- XR: S scoliosis, shoulder, elbow, hip, knee showed normal anatomy
- XR foot: L osteotomy wires, R hallux valgus

Diagnosis?

- Hypermobility
- BJHS (Benign joint hypermobility syndrome)

Case presentation 2

- F/45
- On and off polyarthralgia for 5 years
- Symmetrical polyarthralgia large and small joints, hips, knees, Shoulders, MCPs, PIPs and MTPs
- Pain is more obvious after exercise but most of time is unpredictable
- Joint pain usually subsided spontaneously after few days

- No obvious swelling or redness
- No significant EMS

- Back pain for >10 years
 - Comes and goes
 - at thoracic and lumbar region
 - Never last more than few hours
 - No nocturnal pain or EMS

Likely to be mechanical than inflammatory

- No enthesitis, heel pain or buttock pain
- No eye symptoms
- No dry eyes or dry mouth
- No skin rash
- No features of lupus
- No bowel or urinary symptoms

Past health

- Polyarthralgia since teenager, mainly both knees and hips

Drug history/treatment received for her musculoskeletal pain

- NSAIDs & Coxibs
- Acupuncture
- Massage/Tui Na from TCM

Family Hx

- No connective tissue disease
- Father has similar problems
- Younger sister has joint laxity but no clinical symptoms

Social Hx

- Mother of 2 daughters
- Head swimming coach of an international school
- Elite swimmer, member of HK swimming team at teenage, participating Commonwealth Game
- Master swimming competition record holders

Training history

- Large training volume each day
 - Swim 3-4K daily in an hour
 - 6 days a week
- And dry land exercise
 - At least one hour
 - 6 days a week

P/E

- Height: 169cm
- Weight: 53kg
- BMI: 18.3
- Arm span 164cm
- No deformity
- No synovitis at hands and feet
- Bilateral pes planus
- Mild hallus valgus

- Hip flexion is limited by trunk only(0-140°)
- Hip extension 15-20° (0-10°)
- Hip internal rotation normal (0-50°)
- Hip ext rotation 60° (0-40°)

- Hyperextended knee
- Anterior and posterior draw tests are neg
- Patellar apprehension test are neg
- Shoulder ROM: normal
- Neer's test and Hawkin's sign positive on both sides
- Empty can test positive, supraspinatus muscle weakness
- Beighton score 5/9

Diagnosis

- BJHM + supraspinatus tendinitis and impingement syndrome (swimmer shoulders)

Benign joint hypermobility syndrome (BJHS)

- Defect in collagen, leading to fragility in ligaments, tendons, cartilage, and blood vessels
- AD inheritance
- Hypermobility is relative common 5-15% in population
- Musculoskeletal pain is the major complaints, arthralgia, myalgia and bone pain
- Relative benign condition, no potential life threatening complications
- The difference between EDS and Marfan is there is no widespread tissue fragility, skin, eyes or heart valves

REVISED DIAGNOSTIC (Beighton 1998) CRITERIA FOR THE BENIGN JOINT HYPERMOBILITY SYNDROME (BJHS)

Journal of Rheumatology 2000; 27: 1777-1779

- **Major Criteria**
- A Beighton score of 4/9 or greater (either currently or historically)
- Arthralgia for longer than 3 months in 4 or more joints
- **Minor Criteria**
- A Beighton score of 1, 2 or 3/9 (0, 1, 2 or 3 if aged 50+)
- Arthralgia (> 3 months) in one to three joints or back pain (> 3 months), spondylosis, spondylolysis/spondylolisthesis.
- Dislocation/subluxation in more than one joint, or in one joint on more than one occasion.
- Soft tissue rheumatism. > 3 lesions (e.g. epicondylitis, tenosynovitis, bursitis).
- Marfanoid habitus (tall, slim, span/height ratio >1.03, upper: lower segment ratio less than 0.89, arachnodactily [positive Steinberg/wrist signs]).
- Abnormal skin: striae, hyperextensibility, thin skin, papyraceous scarring.
- Eye signs: drooping eyelids or myopia or antimongoloid slant.
- Varicose veins or hernia or uterine/rectal prolapse.

Grahame *et al* proposed a validated set of diagnostic criteria for BJHS

- The BJHS is diagnosed in the presence 2 major criteria, or 1 major and 2 minor criteria, or 4 minor criteria.
- BJHS is excluded by presence of Marfan or Ehlers-Danlos syndromes.

Why Beighton Criteria is needed?

- Prof Beighton pointed out that Beighton scoring system is developed for epidemiological study, not for clinical diagnosis
- Hypermobility $\neq$ disease
- Hypermobilit Syndrome means hypermobility + symptoms

Beighton score system

- Score one point if you can bend and place your hands flat on the floor without bending your knees



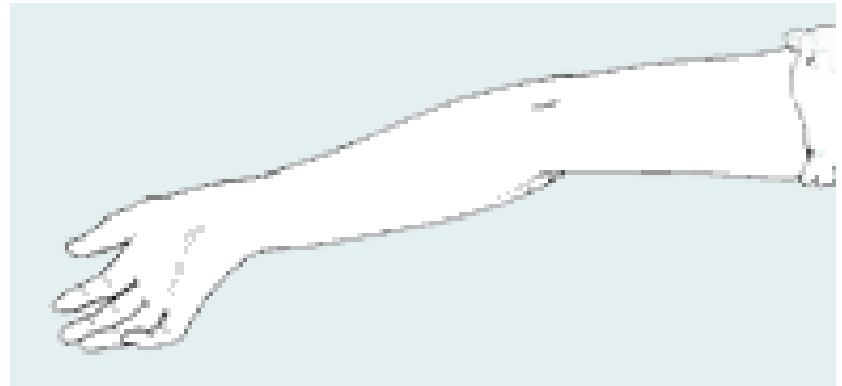
Beighton score system

- Score one point for each knee that will bend backwards (hyperextension $\geq 10^\circ$).



Beighton score system

- Score one point for each elbow that will bend backwards (hyperextension $>0^\circ$).



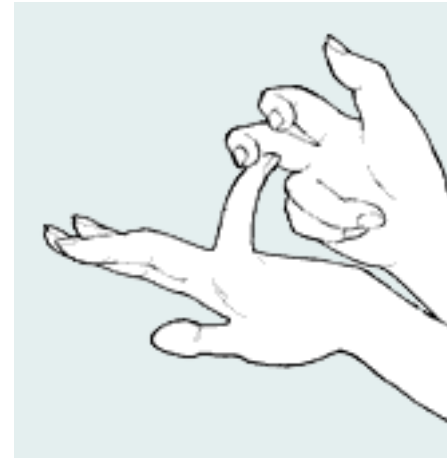
Beighton score system

- Score one point for each thumb that will bend backwards to touch the forearm.



Beighton score system

- Score one point for each hand when you can bend the little finger back beyond 90°.



Problems for both cases

- Post exercise musculoskeletal pain
 - Joint
 - Tendons/ligaments
 - Muscle
- Joint deformity secondary to hypermobility
 - Pes planus
 - Hallux valgus
 - Scoliosis

Reason for referral

- Widespread musculoskeletal pain affects their daily activities, work and social/sports life
- Excludes underlying autoimmune disease

Their wishes

They both want us to help:

1. Ease their pain
2. Go back to their exercise and training

MANAGEMENT

What can be offered to ease their
widespread musculoskeletal pain?

Physical therapies for musculoskeletal pain

- 1. Thermal modalities e.g. heat, cold
- 2. Exercise
- 3. Electrotherapy e.g. TENS, IFT
- 4. Acupuncture
- 5. Manual manipulation e.g. massage

1. Thermal therapy

- A. Heat
 - - Superficial heat
 - - Deep tissue heating
- B. Cryotherapy

1. Thermal therapy

- Prescription for heat or cold:
 - Indication or diagnosis
 - Modality
 - Location
 - Intensity
 - Duration
 - Frequency

A. Heat therapy

- Postulated mechanisms for analgesia:
 - Cutaneous counterirritant effect
 - Vasodilatation -> decreased ischaemic pain, washout of pain mediators
 - Reduce muscle spasm leading to ischaemia
 - Elevation of pain threshold

A. Heat therapy

- **(i) Superficial heat**

- Moist hot packs, immersion in hot water baths, paraffin wax application etc
- Duration ~ 20-30 min
- Only heat superficial tissues

A. Heat therapy

- (ii) Deep heat
- Ultrasound
 - Use of high-frequency acoustic energy to produce thermal effects in tissue
 - Heats deeper tissues, predominantly at tissue interfaces

Ultrasound therapy for musculoskeletal disorders

- A systematic review in 1999 included 38 studies, evaluating the effects of ultrasound therapy on pain, disability or range of motion for lateral epicondylitis, shoulder pain, degenerative rheumatic disorders, ankle distortions, temporomandibular pain or myofascial pain and a variety of other disorders.

Ultrasound therapy for musculoskeletal disorders: a systematic review. Pain. 1999 Jun; 81(3): 257-71

- There seems to be little evidence to support the use of ultrasound therapy in the treatment of musculoskeletal disorders

USG therapy

- Several other systematic reviews also showed little evidence for pain relief in different musculoskeletal disorders.
 - Therapeutic ultrasound for the treatment of rheumatoid arthritis. *Cochrane Database Syst Rev.* 2002; (3)
 - Ultrasound therapy for acute ankle sprains. *Cochrane Database Syst Rev.* 2002; (1)
 - Therapeutic ultrasound for treating patellofemoral pain syndrome. *Cochrane Database Syst Rev.* 2001; (4)

B. Cryotherapy

- Ice/cold packs, cold water immersion, vapocoolant spray
- Acute injury / inflammatory pain
- *Mechanisms for pain relief*
 - : slows conduction velocity of peripheral nerves -> decrease nociceptive information transmission / neuronal activity of dorsal horn neurons ; cutaneous counterirritant effect; vasoconstriction and reduce swelling

Physical therapies for musculoskeletal pain

- 1. Thermal modalities e.g. heat, cold
- 2. *Exercise*
- 3. Electrotherapy e.g. TENS, IFT
- 4. Acupuncture
- 5. Manual manipulation e.g. massage

2. Exercises

- Different forms of exercise are used in the treatment of painful musculoskeletal disorders, e.g. aerobic exercise, strengthening exercises, aquatic exercise

Meta-analysis: Exercise therapy for treatment of non-specific low back pain

Ann Intern Med May 2005 3; 142(a)765-75

- 61 randomized controlled trials (6390 participants) met inclusion criteria: acute (11), subacute (6) and chronic (43) low-back pain (1 unclear).
- Evidence suggests that exercise therapy is effective in chronic back pain relative to comparisons at all follow-up periods; pooled mean improvement was 7.3 points (95% CI, 3.7 to 10.9) for pain (out of 100), 2.5 points (CI, 1.0 to 3.9) for function (out of 100) at earliest follow-up.

Meta-analysis: Exercise therapy for treatment of non-specific low back pain

Ann Intern Med May 2005 3; 142(a)765-75

- **Conclusions:**
 - Exercise therapy appears to be slightly effective at decreasing pain and improving function in adults with chronic low-back pain

Aquatic exercise

- *Pain relieving mechanisms:*
 - Warmth and buoyancy of water may block nociception by acting on thermal and mechanoreceptors; enhance blood flow; facilitate muscle relaxation
 - Ease of movement may activate supraspinal pathways resulting in a reduction in pain intensity

Does Aquatic Exercise Relieve Pain in Adults With Neurologic or Musculoskeletal Disease? A Systematic Review and Meta-Analysis of Randomized Controlled Trials

Archives of Physical Medicine and Rehabilitation Volume 89, Issue 5, May 2008, Pages 873-883

- Randomized controlled trials (RCTs) that included adults with neurologic or musculoskeletal disease, pain as an outcome measure, and exercise in water were included.
- 5 RCTs included in meta-analyses
 - aquatic exercise vs no treatment n=3
 - aquatic exercise vs land treatment n=2

Does Aquatic Exercise Relieve Pain in Adults With Neurologic or Musculoskeletal Disease? A Systematic Review and Meta-Analysis of Randomized Controlled Trials

Archives of Physical Medicine and Rehabilitation Volume 89, Issue 5, May 2008, Pages 873-883

- Results:
- Aquatic exercise has a small post-treatment effect in relieving pain compared with no treatment ($P=.04$; standardized mean difference [SMD], $-.17$; 95% confidence interval [CI], $-.33$ to $-.01$), but it is not possible to draw a firm conclusion because of the lack of consistency of evidence across studies.
- Comparable pain-relieving effects were found between aquatic and land-based exercise ($P=.56$; SMD=.11; 95% CI, $-.27$ to $.50$).

Does Aquatic Exercise Relieve Pain in Adults With Neurologic or Musculoskeletal Disease? A Systematic Review and Meta-Analysis of Randomized Controlled Trials

Archives of Physical Medicine and Rehabilitation Volume 89, Issue 5, May 2008, Pages 873-883

- **Conclusions:**
 - There is sound evidence that there are no differences in pain-relieving effects between aquatic and land exercise.
 - Compared with no treatment, aquatic exercise has a small pain-relieving effect; however, the small number of good-quality studies and inconsistency of results means that insufficient evidence limits firm conclusions.

Physical therapies for musculoskeletal pain

- 1. Thermal modalities e.g. heat, cold
- 2. Exercise
- 3. *Electrotherapy e.g. TENS, IFT*
- 4. Acupuncture
- 5. Manual manipulation e.g. massage

3. Electrotherapy

- A. Transcutaneous electrical nerve stimulation (TENS)
 - To provide continuous electrical impulses via surface electrodes

Transcutaneous electrical nerve stimulation (TENS)

- Administered at varying frequencies of stimulation
 - *High frequency* (>50Hz)
 - *Low frequency* (<10 Hz)
- Intensity – determined by the response of the patient as being sensory level or motor level TENS
 - *Low intensity*: voltage increased until patient feels a comfortable tingling or tapping sensation without motor contraction
 - *High intensity*: motor contraction

Transcutaneous electrical nerve stimulation (TENS)

- *Conventional TENS*
 - high frequency TENS administered at low intensities
- *Strong, low-rate, or acupuncturelike TENS*
 - low frequency TENS administered at high intensities so that a motor contraction is produced

Transcutaneous electrical nerve stimulation (TENS)

- *Proposed mechanisms for pain relief:*
 - Gate control theory of pain
 - Release of endogenous opioids
 - Reduce dorsal horn neuron responses to noxious stimuli in normal and arthritic animals

Transcutaneous electrical nerve stimulation (TENS)

- **Transcutaneous electrical nerve stimulation (TENS) versus placebo for chronic low-back pain.** *Cochrane Database Syst Rev. 2008 Oct 8;(4)*
 - Conflicting evidence about whether TENS was beneficial in reducing back pain intensity
 - Consistent evidence in two trials (410 patients) that it did not improve back-specific functional status.
- **Transcutaneous electrical nerve stimulation (TENS) for the treatment of rheumatoid arthritis in the hand.** *Cochrane Database Syst Rev. 2003; (3)*
 - Conflicting effects of TENS on pain outcomes in patients with RA.
- **Transcutaneous electrical nerve stimulation for knee osteoarthritis.** *Cochrane Database Syst Rev. 2000; (4)*
 - Pain relief and knee stiffness improved significantly in active treatment group
 - TENS is effective in pain control over placebo for OA knee

3. Electrotherapy

- B. Interferential therapy (IFT)
 - Two medium-frequency(KHz) currents are applied to the skin, a low-frequency current (difference in frequency between the two medium-frequency currents) will be induced in the deep tissues

Interferential therapy (IFT)

- Physiologic and therapeutic effects similar to those of TENS
- Allows effective stimulation of deep tissues, whereas TENS is predominantly a cutaneous or superficial stimulus
- Larger pain-relieving areas

Physical therapies for musculoskeletal pain

- 1. Thermal modalities e.g. heat, cold
- 2. Exercise
- 3. Electrotherapy e.g. TENS, IFT
- 4. *Acupuncture*
- 5. Manual manipulation e.g. massage

4. Acupuncture

- Insertion of needles into designated acupuncture points; needles manually manipulated or electrical current administered via the needles (electroacupuncture)
- *Pain relief mechanisms:*
 - Gate Control Pain Theory
 - Stimulates endogenous opioids and other neurotransmitters in the modulation of pain

Scientific Evidence of Acupuncture

- Almost 500 randomized controlled trials conducted on the efficacy of acupuncture since 1970s
- Often, these reviews report equivocal efficacy with exception of RCTs of acupuncture for emesis and dental pain showing overwhelmingly positive result

- Acupuncture: Theory, Efficacy, and Practice. *Ann Intern Med.* 2002 Mar 5;136(5):374-83. Review.

Acupuncture for peripheral joint osteoarthritis: a systematic review and meta-analysis.

- Rheumatology (Oxford). 2006 Nov;45(11):1331-7.

- 18 RCTs: 10 trials tested manual acupuncture; 8 trials tested electro-acupuncture.
- Overall, ten studies demonstrated greater pain reduction in acupuncture groups compared with controls.
- The meta-analysis of homogeneous data showed a significant effect of manual acupuncture compared with sham acupuncture (standardized mean difference 0.24, 95% confidence interval 0.01-0.47, $P = 0.04$, $n = 329$), which is supported by data for knee OA.
- The extent of heterogeneity in trials of electro-acupuncture prevented a meaningful meta-analysis.

Physical therapies for musculoskeletal pain

- 1. Thermal modalities e.g. heat, cold
- 2. Exercise
- 3. Electrotherapy e.g. TENS, IFT
- 4. Acupuncture
- 5. *Manual manipulation e.g. massage*

5. Manual manipulation

- Massage
 - Soft-tissue manipulation using hands or a mechanical device on any body part
 - To produce relaxation, relieve muscle tension, reduce pain, increase mobility of soft tissue and improve circulation

Massage for low-back pain.

- Cochrane Database Syst Rev. 2008 Oct 8;(4)

- Thirteen randomized trials (1596 participants) assessing effects of massage therapy for non-specific low-back pain were included
- Conclusions: Massage might be beneficial for patients with subacute and chronic non-specific low-back pain, especially when combined with exercises and education.

Summary

- Ice and Heat useful for post-exercise pain, arthralgia
- USG for acute injuries
- TENS/IFT/massage should be useful for Case 2's back pain
- Acupuncture may or may not be helpful
- *?Exercise*

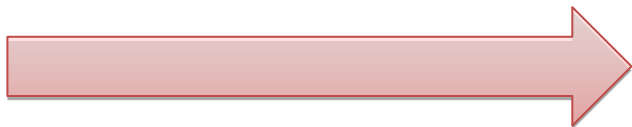
Exercise advice/prescription for hypermobility

The benefits of regular exercise

- More energy to sustain the events of the day
- Improved circulation
- Stabilization of blood pressure
- Improved kidney function
- Improved posture
- Increased muscle strength
- Improved sleep patterns
- Stabilization of blood sugar levels
- Improved concentration and improved mental function
- Increased ability to cope with stress
- Faster recovery from illness and injury
- Improved heart and lung function
- Better digestion
- It makes you feel good

Potential risks of exercise in hypermobility

- Lack of ligamentous strength to hold joints
- Muscle imbalance
- Poor proprioception
 - Joint dislocation
 - Soft tissue injuries
 - Muscle or ligamentous tears



High injury risks

Principles of Joint protection technique

- Respect Pain
 - Fear of pain results in inactivity and loss of motion and strength. Disregard for pain results in joint damage and pain.
- Wear splints and/or braces
 - Protect and provide rest to very weak or unstable joints. During activity, provide external stability to an unstable joint. Work on large joints
- Avoid positions of deformity
- Avoid pressure against the radial side of each finger (thumb side)
 - Don't rest your chin on the side of your fingers. Hold handles straight across the palm.
- Avoid a tight grasp
 - Don't carry heavy handbags, pails, and bags by the handle. Hold everything no tighter than necessary. Release tight grasp frequently if you have to use it.

Postural exercise

- Poor postures are common in hypermobility
 - Swayback, flatback, rounded shoulders
- Good posture reduces the stress placed on joints, muscles and ligaments, thus reducing wear and tear
- It helps maintaining normal joint range and muscle length
 - Vopr Kurortol Fizioter Lech Fiz Kult. 2000 (1):8-11

Co-ordination/proprioception exercise

- Poor proprioception is found in BJHS
 - Phys Ther (1999)79:591-599
 - Br J Rheumatol (1995)34:121-125
- Walking backward, heel walking, walking with eye closed, single leg standing
- wobble board, half roll and Swiss ball exercises.
- They help in synchronizing limb and trunk movements, reducing the risk of dislocation, subluxation and “tripping over” injuries
- Proprioceptive ex enhance proprioception and reduce symptoms
 - Arthritis Rheuma 2004,50(10):3323-8
 - Rheumatol Int (2008)28:995-1000

Stretching/Flexibility

- Emphasize on stretching muscles not ligaments
- Do not overstretch, hyper-movement is not encouraged
- Watch alignment when stretch
- Always warm up a little bit before stretching
- It is the muscle we want to stretch, not the tendons or ligaments.

Muscle strengthening

- To maintain good muscle balance
- Rotator cuff muscles to avoid shoulder injuries or dislocation
- Advice: less repetitions

Aerobic exercise

- Low impact exercise vs high impact
 - Swimming, walking, cycling, golfing, double tennis,
 - Running, football, Basketball, rugby

Exercise plan for patient 1

- Exercise log book
- Heart rate to gauge intensity (polar watch)

	Mon	Tue	Wed	Thur	Friday
Proprioceptive exercise					
Back walking	30s		30s		30s
Heel walk					
Toe walk					
Side walk					
Rocker bottom wood					
Exercise ball					
Running dist	7K				8K
HR (average)	120/min				122
Swimming	500m			600m	
Record events during exercise	Pain in knees				
					84

Exercise plan for patient 2

- Exercise log
- Postural and proprioception exercise
- Rotator cuff/scapular stabilizer muscles strengthening exercise
- Stop usual swimming program which consists mainly endurance/interval of 4 styles
- New program consists of kicks/drills/breaststroke only

Swimming program (sample)

warm up:

- 100m freestyle
- 200m side kick

Main set:

3x 200m free kick/200 fly kick/200 back kick/200
BS kick

3x 25m BS between each style of kick

Total milage: 3k

Other problems associated with BJHS

- Urinary or anal incontinence
 - Higher prevalence of urinary and anal incontinence
 - UI 18/30 (HMS) vs 9/30 (control)
 - AI 7/30 (HMS) vs 0/30 (control)

Other problems associated with BJHS (2)

- Relationship between BJHS, MVP and elastic properties of aortic wall
 - High incidence of MVP in BJHS and proportional to Beighton score
 - (45.6%, (21/46) vs 12% (3/25), $P < 0.0001$)
 - BJHS patients have high elastic property of aortic wall
 - lower index of aortic wall stiffness (IAOS), higher aortic wall distensibility (AOD)

Other problems associated with BJHS (3)

- Bone mineral density in females of BJHS
 - Cross-sectional study of 25 Caucasian female 19-57 y.o
 - Lower BMD at spine and femoral head

Calcif Tissue Int. 2000 Jul; 67(1):37-44

Take home messages

1. BJHS is not a simple disorder, it has impact on patient's life
2. Management of pain and distress in BJHS requires knowledge, patience, compassion and understanding
3. Innovative approaches are required
4. Among evidence, postural and proprioception exercise is safe and useful