Benign Joint Hypermobility Syndrome
Implications for Clinical Practice

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No disclosures

Background: JHS

• Benign implies “not harmful” or “harmless”
• This condition is not necessarily “benign”...... evidence suggests that it is in fact the source of significant pain, activity limitations and participation restrictions.
• DIFFERENT from “joint hypermobility (JH)” or “laxity” where the patient is asymptomatic.
• Along the spectrum of EDS-HT (Ehlers-Danlos Syndrome, Hypermobility Type)

Voerman et al, Dis & Rehab, 2011
Background: JHS

- Inherited connective disorder (also in first degree relatives in 50% of cases)
- Type I collagen (most abundant in the body) is hyperelastic.
  - Type I is mostly found in tendons, ligaments, joint capsules, skin and bone.
- Manifests as hypermobility in multiple joints.
- JHS is reserved for the syndrome in which joint laxity is associated with pain in multiple joints.

Russek et al., Physiother Res Int, 2016
Albayrak et al., Cilt Rheumotol, 2015

Background: JHS

- The persistent nociceptive input due to joint abnormalities may trigger a central sensitization phenomenon in the dorsal horn and cause widespread pain.

Di Stefano et al., Eur J Pain, 2016

Prevalence of JHS

- Varies, but:
  - Greater among children, women and people of Asian or African descent
  - In the US, the range is from 5% to 24% (higher among adolescents, children and women)
  - 64.8% of patients with fibromyalgia, 13.2% of patients in a rheumatology clinic
  - Adult patients with JHS have complaints related to pain; children present with developmental delay, poor coordination or ‘clumsiness’

Russek et al., Physiother Res Int, 2016
Prevalence of JHS in college population

- Prevalence of generalized JH in females: 36.7%
- Prevalence of JHS in females: 24.5%
- Individuals with JHS were significantly more likely to have had sprains, back pain, and stress fractures, report clumsiness, easy bruising, and balance problems than those who did not have JHS.

Yet, under recognized and poorly managed

- Many rheumatologists did not believe JHS had a pathophysiological basis and underestimated its prevalence and the impact it had on patients' lives.

US PT knowledge of JHS

- Web-based survey through APTA members about FMS, JRA, ARA and JHS.
- Most knowledge of JHS was
  - Self-taught (31%)
  - PT school (24%)
- Have not learned about the condition: 39%
- Not at all confident in managing JHS: 21%
- Did not know the Brighton Criteria: 73%
Testing for JHS

- Beighton Scale
- Brighton Criteria (Beighton scale is part of Brighton criteria)

Beighton Scale

As passive hyperextension of fifth finger, so they are parallel to the extensor surface of the forearm: E) ability to hyperextend the knee beyond 10°; C) flexion, so that the individual can place their palms on the floor without bending their knees; D) passive opposition of the thumb to the flexor surface of the forearm; E) ability to hyperextend the elbow beyond 10°

Beighton Scale

- Scoring: ≥4/9 or ≥5/9 was considered diagnostic of hypermobility
Brighton Criteria

- Developed to emphasize the fact that BJHS is a systemic and symptomatic condition

Brighton Criteria

<table>
<thead>
<tr>
<th>Major Criteria</th>
<th>Minor Criteria</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Beighton Scale of ≥4/9</td>
<td>1. Beighton score of 1, 2 or 3 (0-3 if age &gt;50)</td>
</tr>
<tr>
<td>2. Arthralgia for &gt;3 months in 4+ joints</td>
<td>2. Arthralgia (≥3 months, in 1-3 joints, or back pain (≥3 months) with spondylosis, sashitis)</td>
</tr>
</tbody>
</table>

BJHS is diagnosed by:
1. Presence of two major criteria
2. One major and two minor criteria
3. Four minor criteria

*Major and minor criteria two are also mutually exclusive

5-Point Hypermobility Questionnaire

1. Can you now (or ever) place hands flat on floor with knees straight?
2. Can you now (or ever) bend your thumb to touch your forearm?
3. As a child, did you amuse friends by contorting your body into strange shapes or could you do the splits?
4. As a child or teenager, did your kneecap or shoulder dislocate on more than one occasion?
5. Do you consider yourself "double-jointed?"

Yes to 2+ suggests hypermobility
Sens: 85% Spec: 90%

Hakim et al., Int J Clin Pract, 2003
Pain is the chief complaint of patients with JHS

- Due to decrease in joint position sense, making more susceptible to injury
- Static stabilizers are ineffective, dynamic stabilizers (muscles) must work overtime
- Arthralgia and osteoarthritis may occur in the long term due to excessive movement of the joints
- Most commonly affected joints: Knee, Ankle, Lumbar Spine

Albayrak, Clin Rehumatol, 2015

Other signs and symptoms associated with JHS

- Clumsiness or motor delay
- Chronic fatigue
- Unrefreshing sleep
- Tendinitis
- Proprioception and balance deficits
- Orthostatic hypotension
- Headaches
- Paresthesias
- Memory and concentration problems
- Muscle cramps and pain
- Strains and sprains
- Dislocations
- Chronic gastritis
- Dizziness
- Tachycardia
- Varicose veins
- Anxiety
- Panic

Russak et al., Physiother Res Int, 2016

Sleep, QoL, Depression in JHS

- 115 patients with JHS and 115 controls were assessed for level of pain, depression level, fatigue, sleep, and quality of life.
- Persons with JHS showed higher scores for:
  - Depression (Beck Depression Inventory)
  - Sleep (Pittsburgh Sleep Quality Index)
  - QoL (Short Form-36)

Albayrak, Clin Rehumatol, 2015
JHS and joint proprioception; pain, fatigue depression and disability

- People with JHS demonstrated statistically significantly poorer lower limb joint position sense (JPS) (p < 0.001) and threshold detection to movement (p < 0.001) than those without JHS; UE was not statistically significant
- Fatigue and symptoms of anxiety and depression showed strong associations with disability.
- Pain can be reduced by a combination of physical and cognitive approaches

From the patient’s perspective ...

Patients report that the biggest issues are:
- Pain
- Fatigue
- Proprioception difficulties
- Repeated cycles of injury
- Lack of awareness of JHS from health professionals and society
- Diagnosis and access to appropriate health-care services was often slow

Research supporting PT Management

- 1 RCT in children
- 1 RCT in adults
- 2 cohort studies in adults

- Persons with JHS who undertake exercise improve over time regarding pain, global assessment of the impact of hypermobility, and QoL without adverse effects
PT Management of JHS: Big Picture

• Acutely: taping, bracing, splinting; NSAIDs
• Exercise is the mainstay of treatment for JHS
• NM education of the stabilizing muscles
• Closed chain exercises may reduce strain and increased proprioception
• Coordination and balance exercises
• Cardiovascular activity is also very important

Prevent acute joint and muscle pain

• Regular physical activity
• Avoid smoking
• Maintain healthy weight
• Stabilize joints with soft tape or brace
• Improve ergonomic at home and workplace
• Prevent osteopenia with Vitamin D supplementation

RX of acute/recurrent pain

• Active rest
• Cold/heat
• Joint stabilization (via immobilization)
• PT for symptom relief
Prevent chronicization of pain

- Personalized, long-term exercise program geared toward improving strength and proprioception of joint stabilizers and balance
- Physical activity via appropriate pacing
- Stress management
- Improve sleep quality
- CBT

Putting it all together: Case

- Female OW crew athlete, 20 years old
- Grade I Spondylolisthesis

- Height: 71.5 in
- Weight: 162 lbs
- 23% BF
- PMH: MDI in right shoulder (which still limits activity)
- HPI: Gradual insidious onset of LBP, increase with rowing, was painfree at rest, now has pain at rest.
- Pain with extension and extension based movement

Beighton Scale
Other factors (Brighton Criteria)

<table>
<thead>
<tr>
<th>YES</th>
<th>NO</th>
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<tbody>
<tr>
<td>Tall, slim</td>
<td>Span/height ratio 1.03</td>
</tr>
<tr>
<td></td>
<td>72.75&quot; / 71.5&quot; = 1.017</td>
</tr>
<tr>
<td>Myopia</td>
<td>Arachnodactyly</td>
</tr>
<tr>
<td>Subluxation in one joint &gt;1 time (MDI)</td>
<td>Varicose veins, thin skin, or prolapse</td>
</tr>
<tr>
<td>Arthralgia with spondy</td>
<td>Soft tissue rheumatism</td>
</tr>
</tbody>
</table>

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Brighton et al. / Rheum. 2000

Management

Acutely:
- Soft-sided back brace for PRN use
- Short levers for ADLs
- DC rowing and riding in or driving the launch
- Stim/ice/modalities that relieved pain
- OTC medications
- Deferred injection
- Core stabilization (stability stage of motor control)
  - US for biofeedback with TA, ED and multifidi
Management

Subacute:
- Out of brace
- CV: biking with intermittent erging (SS, rate cap) with minimizing the finish (working catch drive)
- Rowing: tanking (working on the finish with minimal resistance, mostly for technique)
- Controlled mobility exercises, starting with short lever

Management

- Successful RTS after ~3 months.
- Rowed for ~12 months with minimal difficulty
- Huge exacerbation after winter training (weight room?)
- Difficulty with ADLs, sitting in class; chose to red shirt and shut down from all activity other than ADLs
- “Fought” conservative care for 3-4 months before agreeing to an injection
Management

- Post-injection: better controlled mobility, but difficulty with rowing persisted; eventually RTS ~ 5 months post injection
- Rows for the team (went from 1st boat to 3rd boat)
- Has a significant leadership role on the team
- Received the “Unsung Hero: Female Athlete” Award in April 2016 (voted upon by all UW Athletes)
- Manages condition independently

Summary

- Benign Joint Hypermobility Syndrome is anything but benign!
- Beighton Scale and Brighton Criteria
- Case involving a female rower who barely fit the criteria (making it more difficult to treat)
  - Findings were very subtle
- Look for criteria in those females who are really having difficulty with improving condition

THANK YOU!