PERIODIC PARALYSIS: OTHER SYMPTOMS

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Disclaimers

- I have no financial conflicts of interest
Disclaimers

- Discussion is predicated on an unproven observation
Disclaimers

- I am not a neurologist
Objectives

1) Describe possible association between Periodic Paralysis and Joint Hypermobility

2) Discuss symptoms associated with Joint Hypermobility

3) Discuss general management of two common Hypermobility problems
Periodic Paralysis

“Some signs and symptoms noted at the PPA Conference that may also be a part of familial hypokalemic periodic paralysis (but have not been formally validated) are:”

www.periodicparalysis.org/site/primary-hypokalemic-periodic-paralysis
PP Other Symptoms

Periodic Paralysis Association Website

Migraines
Heart rhythm abnormalities
ADD/ADHD
Insensitivity to local anesthetics
Severe PMS
Pain
Cramps
PP Other Symptoms

Review of the Medical Literature
PP Other Symptoms

Personal Observations

Headache
Palpitations
Joint Pain
Insomnia
Diarrhea / Constipation
Abdominal pain
Anxiety / Irritability
Painful menstrual cycles
Joint hypermobility
Difficulty thinking clearly
TT Hypermobility Clinic Symptoms

Headache
Palpitations
Joint/Muscle Pain
Insomnia
Tinnitus
Diarrhea / Constipation
Abdominal pain
Anxiety / Irritability
Painful menstrual cycles
Increased likelihood of endocrine problem (thyroid/sugar)
Abnormal bleeding / Scarring
Joint dislocations/ injury
Difficulty thinking clearly ("Brain Fog")
Syncope
Insensitivity to local anesthetics
PP Other Symptoms

- Migraines
- Heart rhythm abnormalities
- ADD/ADHD
- Insensitivity to local anesthetics
- Severe PMS
- Pain
- Cramps

Hypermobility

- Headache
- Palpitations
- Joint/Muscle Pain
- Insomnia
- Tinnitus
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### Hypermobility

- Headache
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- Diarrhea /Constipation
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<tr>
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Hypermobility overlap with PP?

PP

Hypermobility

“Other Symptoms?”
Joint Hypermobility

- Up to 10% of women
- Up to 20% of men
- Just biology
Hypermobility overlap with other syndromes

- Fibromyalgia
- CFS
- Hypermobility?
Hypermobility associations noted in the literature (partial list)

- Chronic Fatigue Syndrome (myalgic encephalomyelitis)
- Fibromyalgia
- Chiari malformation
- Postural Orthostatic Tachycardia Syndrome (POTS)
- Eosinophilia
- Job Syndrome
- Slow-transit constipation
- Vesico-ureteral reflux
- Recurrent UTI
- Idiopathic growth hormone deficiency
- SLE
- Vocal cord dysfunction
- Mast Cell dysregulation
Joint Hypermobility - Defined
Beighton Score
Joint Hypermobility

• Beighton Score 4 (or 5)/9
• 10% Adult men
• 20% Adult women
• Up to 40% of Children depending on age
Joint Hypermobility Syndromes

- Benign Joint Hypermobility Syndrome (BJHS)
- Ehlers-Danlos Syndrome- hEDS
- Marfan Syndrome
- Loeys-Dietz Syndrome
- Stickler Syndrome
How Common is Hypermobility in PP?
How is hypermobility associated with symptoms?

- **Direct Effect**
  - Joint pain
  - Dislocations
  - Easy bleeding (altered collagen affecting platelet activation?)
  - Headache?
    - Vascular spasm
    - Altered stability of cervical spine
      - Occipital nerve irritation
      - Excessive muscle contraction to stabilize
    - Dynamic alterations in CSF flow
How is hypermobility associated with symptoms?

- Indirect Effect
  - Dysautonomia
Dysautonomia - Defined

- Central Nervous System
  - Conscious control
  - Unconscious (Automatic) control
    - Sympathetic – “Fight or Flight”
    - Parasympathetic – “Veg and Chill”
Dysautonomia - Defined

- Abnormal regulation of the Autonomic Nervous System

This Photo by Unknown Author is licensed under CC BY-NC-ND
ANS Effects Examples

Sympathetic
- Heart rate Increase
- Blood vessel constriction/dilation
- Depress stomach motility
- Increase sweating
- Pupil size increases
- Release of adrenalin

Parasympathetic
- Heart rate decreases
- Blood vessel constriction/dilation
- Increase stomach motility
- Decrease sweating
- Pupil size decreases
- Suppress adrenalin
Sympathetic Nervous System Triggers

- Fear / startle
- Exercise
- Cold exposure
- Glucose changes
- Positional changes
- Infection
- Anything that causes “stress”
Typical sympathetic dysautonomia “flare”

- Anxious
- Feel short of breath
- Palpitations
- Dizziness with standing
- Pale or Flushed
- Abdominal pain
- Worsening headaches
- Fatigue
- Syncope
Possible PP - Dysautanomia Connections?

Speculation Alert!!!
Possible PP - Dysautanomia Connections?

• Hypothesis 1
  • Altered depolarization process of autonomic nerves (similar to somatic nerves)
  • “Periodic Paralysis of the autonomic nerves”
Possible PP Dysautonomia Connections?

- Hypothesis 2
  - Some PP patients have unrelated hypermobility
  - Hypermobility associated with dysautonomia
  - Sympathetic activation causes serum potassium drop due to increased catecholamine levels
  - Dysregulated ANS results in more frequent / severe paralytic attacks
Possible PP Dysautonomia Connections?

• Hypothesis 3
  • Some patients with "gene negative" PP have direct nerve damage rather than channelopathy
Hypothesis 3
Hypothesis 3: pB-C2 measurement
Hypothesis 3: pB-C2 measurement

Fig. 3b: The axial view through C2 shows high stress (45-60 N/cm²) in the posterior and lateral columns, correlating with the widespread sensory changes, hyper-reflexia and Babinski sign. Even higher stress (70 N/cm²) is seen in the anterior gray matter, possibly underlying the tongue thrusting on presentation of the patient.

FC Henderson, et al 2010
Practical Stuff
Management of “Other Symptoms”

• Joint pain / dislocations
• Dysautonomia / Postural Orthostatic Tachycardia Syndrome
Joint Pain / Dislocations - Microtrauma

- Excess joint laxity results in "micro-trauma" to articular surface
- Body adjusts gait / motion to relieve stress on damaged joints
- Abnormal gait / motion increases stress on "compensatory" joints
Joint Dislocations - Macrotrauma

- Each excessive stretch causes long lasting effect on ligaments
  - Dislocate shoulder once, and more likely to happen again
- Repetitive macrotrauma results in joint failure
Joint Pain - Management

• Avoid high impact activities (eg rugby, football, pole vaulting)
• Habitual muscle toning exercises (bicycle riding, reasonable weight lifting)
• If joint sprained, give time to heal
  • No “walk it off”
• Physical Therapy for “trigger joints”
  • Joints with consistent pain
  • Joints with dislocation
Joint Pain - Management

• Orthotics – Good and Bad (Personal Opinion)
  • Stabilize ligaments, allowing healing
  • Cause muscle atrophy

• If “muscular joint” (hips, shoulders, knees)
  • Short immobilization if necessary
  • Physical therapy to strengthen joint build muscle bulk
  • Surgery if fail conservative therapy

• If “ligamentous joints” (ankles, wrists, fingers)
  • Immobilization if necessary
  • Trial of physical therapy
  • Consider orthotics
  • Surgery if fail conservative therapy
Dysautonomia Management

- **Postural Orthostatic Tachycardia Syndrome (POTS)**
  - Over-activation of sympathetic nervous system
  - Defined by abnormal rise in heart rate with tilt-table testing or orthostatic vital signs
  - DYNAMIC, not static (this is not your grandfather’s dysautonomia)
  - Treatment is focused on capping the sympathetic response
POTS Management

- Catecholamine blockade
  - Beta blockers (non-cardioselective)
    - Eg. Propranolol, Labetolol
    - There seems to be diminishing returns when “standard” dosing used.
    - Typically low dose (Start Propranolol 10-15mg/day, no more than 45mg/day)
    - Advantages – maybe benefit for headaches? Benefit with anxiety?
    - Disadvantages – may worsen insomnia? Classically risk of exacerbation of depression when used at standard blood pressure doses
POTS Management

- Catecholamine blockade
  - Central alpha agonists
    - Eg. Clonidine
    - Longer acting variants are available
    - Stimulate negative feedback loop to sympathetic system
  - Advantages – Benefit if insomnia?
  - Disadvantages - Sleepiness
POTS Management

- Catecholamine stimulation
  - Peripheral alpha agonists
    - Midodrine
    - Direct vasoconstriction of peripheral blood vessels
    - Increase blood flow return to heart
  - Advantages – Great med if isolated cardiovascular symptoms
  - Disadvantages – Does not work on any other symptoms
POTS Management

• Other
  • Funny channel blockade
    • Ivabradine
    • Slows the rate of cardiac pace setting
    • Two retrospective studies showed equivalence with other therapies
    • Advantages – Benefit in POTS with severe fatigue?
    • Disadvantages - Cost
POTS Management

- Non-pharmacologic
  - Fluid resuscitation
    - Sports drinks rather than just H2O
    - Hydrate before anticipated vigorous activity
  - Avoid sugar load
POTS Management

- Non-pharmacologic
  - Cooling Vest
POTS Management

- Non-pharmacologic
  - Biofeedback
    - Psychologist
    - Computer based
    - Transcutaneous vagal stimulation?
Summary

• Relationship between hypermobility and PP?

• Lots of symptoms potentially associated with hypermobility

• Management of joints and dysautonomia in hypermobility
TT HM Clinic Experience

**Average Score (0-7) for targeted symptoms**

<table>
<thead>
<tr>
<th>Symptom</th>
<th>Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>Headache</td>
<td>5.0</td>
</tr>
<tr>
<td>Neck Pain</td>
<td>4.5</td>
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<tr>
<td>Joint Pain</td>
<td>4.0</td>
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<tr>
<td>Dizziness</td>
<td>3.5</td>
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<tr>
<td>Syncope</td>
<td>3.0</td>
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<tr>
<td>Fatigue</td>
<td>2.5</td>
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<tr>
<td>Blurred vision</td>
<td>2.0</td>
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<tr>
<td>Tinnitus</td>
<td>1.5</td>
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<tr>
<td>Thinking probl.</td>
<td>1.0</td>
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<td>Anxiety</td>
<td>0.5</td>
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<td>Angry</td>
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<tr>
<td>Sad</td>
<td>0.0</td>
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<tr>
<td>Constipation</td>
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<tr>
<td>Diarrhea</td>
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<tr>
<td>Vomiting</td>
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<td>Abd Pain</td>
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<tr>
<td>Ulcers Palpitation</td>
<td>0.0</td>
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</tbody>
</table>
TT Hypermobility Clinic Experience

It’s a Fan!

It’s a Spear!

It’s a Wall!

It’s a Rope!

It’s a Snake!

It’s a Tree!
TT HM Clinic Experience

Age versus Height percentile

Height Percentile

Age
TT HM Clinic Experience

Age versus Height percentile

- Height Percentile
  - 0 to 120
- Age
  - 0 to 70
TT HM Clinic Experience

Height percentile versus normative

- Percentage of patients
- 5th, 5-25th, 25-50th, 50-75th, 75-95th, 95th percentiles

- Blue line: Hypermobility
- Red line: Expected
TT HM Clinic Experience

**BMI percentile versus normative**

- **Percentage of patients**
- **5th percentile**
- **5-25th percentile**
- **25-50th percentile**
- **50-75th percentile**
- **75-95th percentile**
- **95th percentile**

- **Hypermobility**
- **Expected**
### TT HM Medical Questionnaire

**Study ID Number**

**Date:**

---

**MEDICAL QUESTIONNAIRE – X-ray Study**
Department of Pediatrics, TTUHSC – Amarillo

Please answer the following questions about symptoms you may have been having over the past 2 months. The answers range from 0” which is no problem at all, to 7” which is a problem that keeps you from being able to do your daily activities. Circle the answer that most closely matches how you feel.

<table>
<thead>
<tr>
<th>Symptom</th>
<th>0</th>
<th>1</th>
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<tbody>
<tr>
<td>Headaches</td>
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<td>Neck pain</td>
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<td>Joint pain or muscle pain</td>
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<td>Dizziness with standing</td>
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<td>Passing out or “blacking out”</td>
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<td>Fatigue or feeling tired</td>
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<td>Blurred vision</td>
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<td>Ringing or buzzing sound in your ears</td>
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<td>Feeling sad</td>
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<td>Pain in your belly</td>
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<td>Mouth or nose ulcers</td>
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<td>Polpitations or racing heart</td>
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**Gender**

- [ ] Male
- [ ] Female
HIT-6 Scale

1. When you have headaches, how often is the pain severe?
   - Never
   - Rarely
   - Sometimes
   - Very Often
   - Always

2. How often do headaches limit your ability to do usual daily activities including household work, work, school, or social activities?
   - Never
   - Rarely
   - Sometimes
   - Very Often
   - Always

3. When you have a headache, how often do you wish you could lie down?
   - Never
   - Rarely
   - Sometimes
   - Very Often
   - Always

4. In the past 4 weeks, how often have you felt too tired to do work or daily activities because of your headaches?
   - Never
   - Rarely
   - Sometimes
   - Very Often
   - Always

5. In the past 4 weeks, how often have you felt fed up or irritated because of your headaches?
   - Never
   - Rarely
   - Sometimes
   - Very Often
   - Always

6. In the past 4 weeks, how often did headaches limit your ability to concentrate on work or daily activities?
   - Never
   - Rarely
   - Sometimes
   - Very Often
   - Always

To score, add points for answers in each column.

Total Score

Higher scores indicate greater impact on your life.

Score range is 30-78.
Correlation = 0.84
TT HM Clinic Experience

- Headache
- Neck Pain
- Joint Pain
- Dizziness
- Syncope
- Fatigue
- Blurred vision
- Tinnitus
- Thinking probl
- Anxiety
- Angry
- Sad
- Constipation
- Diarrhea
- Vomiting
- Abd Pain
- Ulcers
- Palpitation

Female | Male
--- | ---

Percentages of symptoms by gender.
Fibromyalgia

• Canadian Consensus Statement
  • History of Pain
    • Bilateral
    • Above and below waist
    • Includes axial skeleton
  • 11 of 18 tender point sites on physical exam
  • Present for at least 3 months

Van de Sande, et al
Journal of Musculoskeletal Pain 2003
CFS/SEID/ME diagnostic criteria

• IOM 2015
  • Substantial reduction in activity for more than 6 months that is not substantially alleviated by rest.
  • Post-exertional malaise
  • Unrefreshing sleep
  • Plus either
    • Cognitive impairment
    • Orthostatic intolerance

Overlap with “defined” syndromes

Pathophysiology?

- Fibromyalgia
- CFS
- Hypermobility?
Hypermobility associations

• Mast Cell Dysregulation
Hypermobility associations

- Mast Cell Dysregulation
- Chiari Malformations
Overlap with “defined” syndromes

- Fibromyalgia
- CFS
- Hypermobility?
- Chronic Migraines
Overlap with “defined” syndromes

Pathophysiology?

- Fibromyalgia
- CFS
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- Chronic Migraines
Overlap with “defined” syndromes

- Fibromyalgia
- CFS
- Hypermobility?
- JIA?
- Chronic Migraines
Overlap with “defined” syndromes

- Fibromyalgia
- CFS
- JIA?
- Hypermobility?
- Normal Pressure Hydrocephalus
- Chronic Migraines
Overlap with “defined” syndromes

- Fibromyalgia
- CFS
- Hypermobility?
- JIA?
- Normal Pressure Hydrocephalus
- Multiple Sclerosis
- Chronic Migraines
Overlap with “defined” syndromes

- Fibromyalgia
- CFS
- Hypermobility?
- Chronic Migraines
- JIA?
- Normal Pressure Hydrocephalus
- Pseudotumor cerebri
- Non-TB Mycobacterium
- Multiple Sclerosis
- Chronic Migraines
Unquantified Hypermobility associations noted at TT
Unquantified Hypermobility associations noted at TT

- Insomnia
- Hypersomnolence
- Tuberculosis
- Non-TB Mycobacterium?
- Gastroparesis
- Osteochondritis dissicans
- Livedo reticularis
- Generalized anxiety disorder
- Menstrual irregularities
- Hematuria
Thoughts/Hypothesis/Meanderings

• Association with Chiari?
• Association with Cranio-cervical instability?
• Changes in Intracranial Pressure?
• Relationship to Mast Cell regulation?
• Histamine dysregulation?
Chiari Malformation – Type 1

• Increased prevalence in EDS-HT population
• 6 mm ptosis cerebellar vermis below the foramen magna
• Little correlation between symptoms and radiographic findings
Chiari Malformation
Chiari Malformation – “Type 0”

- Stringent definition – Chiari Symptoms without radiographic evidence of Chiari, but presence of syringomelia
- Common definition – Presence of a headache and an internet connection.
Chiari Malformation
Chiari Malformation – Type 1

- Arnatouvic et al December 2014
- Literature Review 1965-2013
  - 145 operative series of CM-1
  - 3000+ patients?
  - 65% of patients with Syringomelia
  - 99% of series used posterior decompression
- Overall
  - 75% “Improved”
  - 17% “No Change”
  - 9% “Worsened”
Chiari Malformation – Type 1

- Arnatouvic et al December 2014
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  - 145 operative series of CM-1
  - 3000+ patients?
  - 65% of patients had Syringomelia
  - Peak age for Children 9 yrs, Adult 43 years
  - 99% of series used posterior decompression
- Overall
  - 75% “Improved”
  - 17% “No Change”
  - 9% “Worsened”
- Best case scenario?
Arguments against CM as cause of SYMPTOMS

• Why 10% worsened?
• Why 17% unchanged?
• Why symptoms without Chiari and no symptoms with Chiari?
• What else?
Grabb et al 1999

- 40 Adolescents and young adults
- All with Chiari I
- Attempted to quantify the degree of brainstem distortion by measuring the degree to which odontoid retroflexes
Grabb-Oakes Measurement (pB-C2)

• “in a sagittal image is the distance to a perpendicular line traced from the most posterior region of the dura mater covering the dens to the line that goes from the inferior surface of the basion to the posterior inferior aspect of the C-2 vertebral body”

• Batista et al. Neurosurgery Focus 2015
FIGURE 2

Ventral Brain Stem Compression in Pediatric and Young Adult Patients with Chiari I Malformations.
Grabb, Paul; Mapstone, Timothy; Oakes, W


FIGURE 2. Sketch of a sagittal view of the craniocervical junction showing the B-C2 line, drawn from the basion to the inferoposterior aspect of the C2 body, and a line perpendicular to this line, pB-C2, drawn through the odontoid tip to the ventral cervicomedullary dura. The distance of pB-C2 is then measured in millimeters between the thick and the thin arrows.
Ventral Brain Stem Compression in Pediatric and Young Adult Patients with Chiari I Malformations.
Grabb, Paul; Mapstone, Timothy; Oakes, W

FIGURE 1. Examples of the subjective grades of VBSC in association with Chiari I malformations as revealed by sagittal MRI. A, absent; B, mild (note ventral flattening of the cervicomедullary junction); C, moderate (note frank distortion by a ventral vector of the cervicomедullary junction).
<table>
<thead>
<tr>
<th>Ventral Brain Stem Compression in Pediatric and Young Adult Patients with Chiari I Malformations.</th>
<th>TABLE 3. Cohorts of pB-C2 Measurements Grouped as Low, Medium, and High (with the means and standard deviations in parentheses) of the Accompanying Factors</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>TABLE 3</strong></td>
<td><strong>Ventral Brain Stem Compression in Pediatric and Young Adult Patients with Chiari I Malformations.</strong> Grabb, Paul; Mapstone, Timothy; Oakes, W</td>
</tr>
<tr>
<td></td>
<td>Low pB-C2 Group (&lt;6 mm)</td>
</tr>
<tr>
<td>------------------</td>
<td>--------------------------</td>
</tr>
<tr>
<td>Age (yr)</td>
<td>4.3 (1.0)</td>
</tr>
<tr>
<td>pB-C2 (mm)</td>
<td></td>
</tr>
<tr>
<td>Tonsillar descent (mm)</td>
<td>8.3 (4.7)</td>
</tr>
<tr>
<td>Odontoid’s relation to Chamberlain’s line (mm)</td>
<td>-1.4 (2.6)</td>
</tr>
<tr>
<td>Odontoid’s relation to McRae’s line (mm)</td>
<td>-5.4 (2.4)</td>
</tr>
<tr>
<td>Length of clivus (mm)</td>
<td>32.1 (3.1)</td>
</tr>
<tr>
<td>Basal angle (degrees)</td>
<td>119.6 (6.5)</td>
</tr>
<tr>
<td>Foramen magnum anteroposterior diameter (mm)</td>
<td>34.3 (7.5)</td>
</tr>
</tbody>
</table>

* Significantly different when compared with the other two groups (P < 0.01).
### TABLE 1

| Stratum | All Patients  
<table>
<thead>
<tr>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>n = 40</td>
</tr>
<tr>
<td></td>
<td>Subjective VBSC, Absent n = 10</td>
</tr>
<tr>
<td></td>
<td>Subjective VBSC, Mild n = 19</td>
</tr>
<tr>
<td></td>
<td>Subjective VBSC, Moderate n = 11</td>
</tr>
<tr>
<td>Control Participants n = 12</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Parameter</th>
<th>All Patients</th>
<th>Subjective VBSC, Absent</th>
<th>Subjective VBSC, Mild</th>
<th>Subjective VBSC, Moderate</th>
<th>Control Participants</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tonsillar descent (mm)</td>
<td>13 (6.6)</td>
<td>7.1 (3.8)</td>
<td>12.9 (4.3)</td>
<td>18.5 (7.4)</td>
<td>NM</td>
</tr>
<tr>
<td>Odontoid's relation to Chamberlain's line (mm)</td>
<td>-0.1 (3.8)</td>
<td>-1.6 (2.8)</td>
<td>-0.5 (2.6)</td>
<td>1.5 (5.6)</td>
<td>NM</td>
</tr>
<tr>
<td>Odontoid's relation to McRae's line (mm)</td>
<td>-4.8 (2.1)</td>
<td>-5.7 (2.4)</td>
<td>-4.9 (1.7)</td>
<td>-3.7 (2.3)</td>
<td>NM</td>
</tr>
<tr>
<td>Length of clivus (mm)</td>
<td>33.0 (3.5)</td>
<td>31.6 (2.5)</td>
<td>34.2 (4.0)</td>
<td>31.9 (2.3)</td>
<td>NM</td>
</tr>
<tr>
<td>Foramen magnum anteroposterior diameter (mm)</td>
<td>33.5 (5.9)</td>
<td>34.6 (8.3)</td>
<td>34.2 (4.6)</td>
<td>31.5 (5.2)</td>
<td>NM</td>
</tr>
<tr>
<td>Basal angle (degrees)</td>
<td>121.1 (9.6)</td>
<td>118.6 (7.2)</td>
<td>118.5 (8.4)</td>
<td>127.5 (10.8)</td>
<td>NM</td>
</tr>
<tr>
<td>Age (yr)</td>
<td>8.6 (5.9)</td>
<td>6.3 (4.2)</td>
<td>8.9 (5.6)</td>
<td>10.3 (7.5)</td>
<td>9.0 (3.2)</td>
</tr>
<tr>
<td>pB-C2 (mm)</td>
<td>7.0 (2.3)</td>
<td>4.1 (0.9)</td>
<td>7.2 (1.4)</td>
<td>9.1 (1.6)</td>
<td>3.7 (0.7)</td>
</tr>
</tbody>
</table>

* a VBSC, ventral brain stem compression; NM, not measured.

b P < 0.05 when subjective grade groups are compared with one another.

c Negative and positive values correspond to caudal and cranial to the reference line, respectively.

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**Ventral Brain Stem Compression in Pediatric and Young Adult Patients with Chiari I Malformations.**

Grabb, Paul; Mapstone, Timothy; Oakes, W

Grabb et al 1999

- 40 Adolescents and young adults
- All with Chiari I
- 12 Had pB-C2 >9mm
- 11/12 underwent posterior decompression
- 4/11 underwent cervical spine fusion/odontoidectomy
- Remaining 7 had resolution of primary neurologic sign
- Conclusion – If pB-C2 <9mm, posterior decompression adequate
pB-C2

- Asymptomatic adult average about 6mm
- Asymptomatic pre-adolescent average about 3mm
- TT HM Clinic average 9mm
- ?
pB-C2 Measurement

Headache Severity vs Grabb-Oakes

- HIT-6 score vs Grabb-Oakes (mm)
• Neither Chiari malformations nor Grabb-Oakes measurements correlate well with symptoms in literature
• Both are “static” measurements
Atlantoaxial Instability?

- Goel – 2015
- Retrospective review of 65 patients with Chiari 1
- 84% with Syringomelia
- All treated with Atlantoaxial fixation
Atlantoaxial Instability?

- Goel – 2015
- Retrospective review of 65 patients with Chiari 1
- 84% with Syringomelia
- All treated with Atlantoaxial fixation
- 1 Died
- 1 Worsened
- 63 Improved
- Post-op MRI only available for 11
Neurosurgeons always give 110%!
Atlantoaxial Instability?

- Goel – 2015
- Different ages?
- Different pathophysiology?
- Different Disease?
Gaol 2015

Patient age - Gaol 2015

- 11-20: 28%
- 21-30: 42%
- 31-40: 20%
- 41-50: 14%
- 51-60: 6%

Neurosurgeons always give 110%!
Chiari Malformation – Type 1

- Arnatouvic et al December 2014
- Literature Review 1965-2013
  - 145 operative series of CM-1
  - 3000+ patients
  - 65% of patients had Syringomelia
- **Peak age for Children 9 yrs, Adult 43 years**
  - 99% of series used posterior decompression
- Overall
  - 75% “Improved”
  - 17% “No Change”
  - 9% “Worsened”
- Best case scenario?
“Sub-clinical” Cranio-cervical instability

- Same collagen in vertebral ligaments as in ligaments of appendicular joints
- Back pain common complaint among joint hypermobility patients
- Intervertebral disc disease common
- “Dynamic” pathology rather than static pathology
OM study

- 20 female patients
  - Joint hypermobility plus diagnosis of dysautonomia or headaches
- Medical Questionnaire, HIT-6 Scale
- Orthostatic vital signs
- Optic nerve ultrasound
- Dynamic open mouth odontoid X-rays

- Still enrolling!
OM study
OM study
OM study
OM study
OM study

$r = .74$, $r^2 = .55$, $p < 0.05$
OM study

$r = .81$, $r^2 = .66$, $p < 0.05$
OM study

Correlations between C1/C2 shift and symptoms

- Headache
- Neck Pain
- Joint Pain
- Dizziness
- Syncope
- Fatigue
- Blurred vision
- Tinnitus
- Thinking probl
- Anxiety
- Angry
- Sad
- Constipation
- Diarrhea
- Vomiting
- Abd Pain
- Ulcers
- Palpitation
- Total Q Score
- HIT-6
OM study

Significant Correlations

Correlation Coefficient

Headache, Neck Pain, Joint Pain, Dizziness, Syncope, Fatigue, Tinnitus, Constipation, Diarrhea, Abd Pain, Palpitation, HIT-6

= $p<0.05$, HIT-6 $p=0.052$
OM Study

• First evidence of atlantoaxial instability as potential etiology of headache and dysautonomia in patients with joint hypermobility?
P3-PT Study

- 8 female patients
  - Joint hypermobility plus diagnosis of dysautonomia or headaches recruited from OM Study
- Medical Questionnaire, HIT-6 Scale
- Orthostatic vital signs
- Optic nerve ultrasound
- 6 weeks of PT focused on strengthening C1/C2

- 3 enrolled to date, one completed
P3-PT Study

- Baseline
- Syncope 20x per month
- Chronic Daily headache
- Dropped out of school
- 12 week assessment
- Syncope 1x per month
- Headaches “cut in half”
- Getting married (my intended outcome was to go back to school).
P3-PT Study

• $N=1$ to date
Hunches

- Dysautonomia symptoms in adolescents with HM is a combination of anatomic craniocervical compression (Chiari or pB-C2) plus atlantoaxial instability
- Mast cell regulation plays a role in symptom variability
- Intracranial pressure due to impaired *venous* egress (due to above) determines headache significance
Acknowledgements

Nancy Lewis, PT
Rosie Gellman, MS-3
Kassi Ackerman, M.D.
Saneea Almas, M.D.
Laszlo Nagy, M.D.
Mandy Griffin, M.D.
Johnnie Faircloth, M.D.
Rodney Young, M.D.
Karen Saunders
Questions?
Chiari Malformation