Chronic Pain Syndromes in Children

Cara Hoffart, DO
Associate Professor of Pediatrics
Medical Director Rehabilitation for Amplified Pain Syndromes (RAPS) Program
Rheumatology and Pain Management
cmhoffart@cmh.edu

Disclosure

• I have no relevant financial relationships with the manufacturers(s) of any commercial products(s) and/or provider of commercial services discussed in this CME activity

• I do not intend to discuss an unapproved/investigative use of a commercial product/device in my presentation.
Changes you may wish to make in practice

• Recognize patients who “just don’t make sense”
• Effectively discuss the pathophysiology and treatment strategy for amplified pain with patients
• Prescribe a treatment approach that does not require drug therapy for pain or sleep aide

PAIN

• An unpleasant sensory and emotional experience associated with actual or potential tissue damage, or described in terms of such damage

• Pain is always subjective
Chronic Pain Etiology

- Associated with underlying medical conditions:
  - Arthritis
  - Inflammatory bowel disease
  - Duchene's muscular dystrophy
  - Cerebral palsy
  - Cystic fibrosis
  - Osteogenesis imperfecta
  - Epidermolysis bullosa
  - Post cancer treatment
  - Sickle Cell
Intermittent Constant Amplified Pain Syndromes

CRPS/RSD Widespread Pain/Juvenile Fibromyalgia

Chronic Abdominal Pain; Headache

Fatigue Autonomic Symptoms +/- Hypervigilence

Pain & symptoms out of proportion to exam findings

Juvenile Primary Fibromyalgia

- Full body pain
- Positive ROS
- Conversion
- Uses wheelchair at school
The MOST common look of an adolescent in pain:

- Longstanding pain with no known cause
- Try to appear normal & “keep it together”
- Missing school but making good grades
- Considering disability
- Too hurt and tired to spend time with friends
- Anxiety

PAIN AND SYMPTOM ASSESSMENT QUESTIONNAIRE

A. Have you had pain (every day or almost every day) in the following location(s) in the last three months? (Widespread Pain Index)

<table>
<thead>
<tr>
<th>Location</th>
<th>Shoulder, right</th>
<th>Shoulder, left</th>
<th>Upper arm, right</th>
<th>Upper arm, left</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lower arm, right</td>
<td>Lower arm, left</td>
<td>Hip (buttock), right</td>
<td>Hip (buttock), left</td>
<td></td>
</tr>
<tr>
<td>Upper leg, right</td>
<td>Upper leg, left</td>
<td>Lower leg, right</td>
<td>Lower leg, left</td>
<td></td>
</tr>
<tr>
<td>Jaw, right</td>
<td>Jaw, left</td>
<td>Chest</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Upper back</td>
<td>Lower back</td>
<td>Neck</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

WPI Total:_____

B. How much of a problem have the following been for you during the past three months?

<table>
<thead>
<tr>
<th>Problem</th>
<th>No problem</th>
<th>Slight/Mild problem, generally mild or intermittent</th>
<th>Moderate, Considerable problem, often present</th>
<th>Severe, pervasive, continuous, life-disturbing problem</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fatigue</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>Feeling Tired</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>Concentration or Memory Problems</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
</tbody>
</table>

Total:_____
PAIN AND SYMPTOM ASSESSMENT QUESTIONNAIRE

C. Have you had problems (every day or almost every day) with any of the following during the past three months?

<table>
<thead>
<tr>
<th>Muscle weakness</th>
<th>Nervousness</th>
<th>Dry eyes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Numbness/tingling</td>
<td>Depression</td>
<td>Itching</td>
</tr>
<tr>
<td>Headache/migraine</td>
<td>Frequent urination</td>
<td>Dizziness/light headedness/balance problems</td>
</tr>
<tr>
<td>Abdominal pain/cramps</td>
<td>Loss of appetite</td>
<td>Shortness of breath</td>
</tr>
<tr>
<td>Constipation</td>
<td>Blurred vision</td>
<td>Thinking problem</td>
</tr>
<tr>
<td>Heartburn</td>
<td>Ringing in ears</td>
<td>Dry mouth</td>
</tr>
<tr>
<td>Nausea</td>
<td>Easy bruising</td>
<td>Tenderness to touch</td>
</tr>
<tr>
<td>Irritable Bowel Syndrome (diarrhea, bloating, nervous stomach)</td>
<td>Insomnia (problems falling or staying asleep)</td>
<td>Sensitivity to loud sounds, bright lights or strong smells</td>
</tr>
</tbody>
</table>

For clinician use:
For Section C score:
0 = no symptoms (0), 1=few symptoms (≤5), 2=moderate symptoms (6-9), 3=great deal of symptoms (>10)
Screening cut-off - WPI ≥ 7 and SS ≥ 5 OR WPI 3-6 and SS ≥ 9
WPI = Widespread Pain Index
SS = Symptom Severity

(see instructions below) C Score: _____

SS Total ( B + C) : _______

Medication use in JPFM

- (NSAIDs, TCA, SSRI, Muscle relaxants, pregabalin - Lyrica)
  - Almost no studies of medication efficacy in pediatrics
  - Adult studies mixed
    - Combination of pharmacologic methods with CBT & exercise showed larger effect than pharmacological interventions alone
- Pregabalin (Lyrica)
  - Treat neuropathic pain & fibromyalgia
  - Anticonvulsant – works by decreasing number of pain signals sent out by damaged nerves in the body.
  - Adult study: Greater proportion of patients on Lyrica (48%) had clinically meaningful reduction in pain vs placebo (27%).
    - Clinically meaningful pain reduction defined as 30% or > improvement
- Risk Benefit Ratio: Medications to Adverse Effects
Pregabalin

107 adolescents randomized
- 54 pregabalin 44 completed
- 53 placebo 36 completed

Primary outcome: change in mean pain score
- Not significant P=0.121 (CI -1.51,0.18)


Pregabalin Side Effects

<table>
<thead>
<tr>
<th>Side effects</th>
<th>Pregabalin</th>
<th>Placebo</th>
</tr>
</thead>
<tbody>
<tr>
<td>Major depression</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>Choletithiasis</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>Dizziness</td>
<td>30%</td>
<td>13%</td>
</tr>
<tr>
<td>Nausea</td>
<td>22%</td>
<td>9%</td>
</tr>
<tr>
<td>Increased weight</td>
<td>17%</td>
<td>0% [30% our patients obese]</td>
</tr>
<tr>
<td>Somnolence</td>
<td>9%</td>
<td>4%</td>
</tr>
<tr>
<td>Limb Pain</td>
<td>7%</td>
<td>0%</td>
</tr>
</tbody>
</table>

Complex Regional Pain Syndrome: “Cold Blue Foot”
<table>
<thead>
<tr>
<th>Clinical Characteristic</th>
<th>Children</th>
<th>Adults</th>
</tr>
</thead>
<tbody>
<tr>
<td>Affected Extremity</td>
<td>Lower_BUILD UP (6:1)</td>
<td>Upper_BUILD DOWN</td>
</tr>
<tr>
<td>Sex Ratio</td>
<td>Marked female 7:1</td>
<td>Moderate female 2-4:1</td>
</tr>
<tr>
<td>Inciting injury</td>
<td>Less often</td>
<td>More often</td>
</tr>
<tr>
<td>Exam findings</td>
<td>Skin cooler; neurologic symptoms less pronounced</td>
<td>More likely to have “stages” of change</td>
</tr>
<tr>
<td>Psychological Issues</td>
<td>More common</td>
<td>Less common</td>
</tr>
<tr>
<td>Prognosis</td>
<td>Excellent recovery in most cases</td>
<td>Variable – long term disability common</td>
</tr>
</tbody>
</table>

**Duration of CRPS does not predict outcome in children**
**May have more than one limb affected**

Berde, Anesthesiology 2005
Effects of Chronic Pain

- ~15-25% of children experience chronic or recurrent pain
- Impaired psychosocial function
- Health services utilization
  – Intensive diagnostic and therapeutic efforts
- Increased social and mental health problems
- School absences

Simons LE, et al. J of Ped Psych 38
Effects of Chronic Pain

Strong and specific association between childhood recurrent abdominal pain & anxiety in young adulthood.

Children with frequent headache have increased risk in adulthood of

- headache (OR 2.22, 95% CI 1.62-3.06)
- multiple physical symptoms (1.75, 1.46-2.10)
- psychiatric morbidity (1.41, 1.20 – 1.66)

Fearon P. BMJ 2001; 322:1145

PATHOPHYSIOLOGY & THE BIOPSYCHOSOCIAL MODEL
Tremendous Phenotypic Variability....

- 12 y.o. girl with cold, blue foot, no sock, on crutches
- 13 y.o. girl cannot bend her knee or walk, legs lock
- 8 y.o. boy cannot go to school after playing soccer – crawls home
- 12 y.o. girl who arrives to clinic in a wheelchair
- 14 y.o. girl with widespread pain, +ROS and painful points
Injury/Illness Immobilization

Pain Fear of movement/activity & pain implication

Loss of Fitness Low self esteem

Sleep Disturbance

Pain & Fatigue affect School attendance & activities

Loss of social contacts

Amplified Pain Syndrome

Working Model of Pain

- Makes the pain understandable and real
- They are NOT faking it!
- Validate
- This is not “all in your head”
- Pain can arise from:
  - Injury
  - Illness
  - Psychological stress
  - Idiopathic
WHO ARE THESE KIDS?
Pain Amplification Syndromes: Putting it all together

- Who are these kids really??
  - Generally healthy (primary vs secondary pain)
  - Females ~80%
  - Caucasian ~80%
  - Mean age: 12 years
  - Mean Duration is > 1 year
  - Most have constant pain
  - Pain may be in multiple sites

Historical Clues

- Increasing pain over time
- Minor trauma or illness common (? not recalled)
- Allodynia
- Worse with rest, cast, splint
- Failed all prior therapy
- May just have history of autonomic signs
Past Medical History Clues

• Slow healer
• + Review of Symptoms
  – (fatigue, sleep disturbance, headache, dizziness, chest pain, blurry vision, palpitations, abdominal pain, diarrhea, nausea)
• High level athlete, may have history of multiple injuries

Social History Clues

• Major life events
• Role model for the same or chronic pain
• Typical personality
  – Mature
  – Excels
  – Pleaser
  – Perfectionist
  – Worrier
  – Sensitive
Physical Exam Clues

- Spokes-parent
- Pseudo-mature
- Incongruent affect
- La belle indifference
  - Unconcern toward physical symptoms by patients
  - Physical symptoms may provide positive reinforcement or relieve anxiety

Autonomic Signs

- Cold
- Cyanotic
- Clammy
- Decreased pulse
- Dystrophic skin
- Check after exercise
Allodynia & Hyperalgesia

- Light touch or light pressure
- Gentle pinch of a fold of skin

- CHECK BORDERS REPEATEDLY
  - may vary 4 to 12 cm within seconds
Complete Appropriate Evaluation

• Reasonable evaluation should be complete
• Evaluation should essentially be normal
  – Caveat: secondary pain syndrome
• Patients can have more than one thing
• Fine line between complete evaluation and over-medicalizing

Laboratory and Imaging

• APS is a diagnosis of exclusion!
• Every patient deserves:
  – Baseline labs
  – Possible x-rays, ultrasound or other appropriate imaging
  – Thoughtful consideration of a differential diagnosis!!
• Don’t miss thrombus, ESR of 70, etc!
Laboratory & Imaging Studies

• Blood studies
  – Normal

• Radiographs
  – Normal or osteoporosis

• Bone Scan
  – Decreased
  – Can be normal or spotty (adult-like)

• MRI
  – Edema – soft tissue & bone → sometimes leads to a boot!
Challenges in Management

- Complex and Controversial treatment regimens
  - Opioids
  - Local anesthetics
  - Anticonvulsants
  - Intrathecal baclofen
  - IV sympathetic blockade
  - Spinal cord stimulator
  - Surgical sympathetic denervation
- Antidepressant trials in children/adolescents
  - Large placebo response magnitude & small treatment effect
- Anticonvulsants may lead to adverse effects on mood, increase risk of suicidal ideation & attempts
- Concern about publication bias in industry funded trials


Complex Biopsychosocial Model

Requires Multidisciplinary Treatment Approach

This is where patients often have VERY mixed feelings about me!

Treatment Overview

• **Definitive** diagnosis and plan
• **STOP** all medical investigations and pain medications
• Do not immobilize to treat pain
• More **exercise** than you can imagine!
• **Desensitization**
• Counseling and **stress** management
• Decreased **attention** to pain
• Required school attendance & other **functional** activities

Outpatient Treatment

- Most get better outpatient
- Permission to get better & work through pain
- Work closely between services to provide exercise that is appropriate considering injury history.
- More complex or disabled patients may need multidisciplinary pain management referral
  - In the meantime, can get treatment started

Outpatient Treatment Plan

“No Magic Pill”

If it hurts to do something, that’s what you should do!
Function comes back before pain goes away
No pain medications or sleep aides
Pain may worsen before it gets better
You want me to do what?

• Anyone can give the pain talk... or at least a version of it
• Focus on nerve and blood vessel dysfunction – make it about physiology
• Carefully weave in the stress piece
• Time consuming
  – Benefits of established patient-provider relationship
• Start the discussion early
  – “We will rule out.... But if all is normal, this could be....”
• Once treated for Amplified Pain... future injury/illness “heal like a normal person”

What if Outpatient Treatment Fails?

Some patients need more intensive interdisciplinary treatment:

Rehabilitation for Amplified Pain Syndromes (RAPS) Program at
Children’s Mercy Hospital – Kansas City

Other pain rehab programs can be found on the American Pain Society Website:
http://americanpainsociety.org/get-involved/shared-interest-groups/pediatric-adolescent-pain
Pain Rehab Candidates

- Adolescents with disability, severe pain, and failure of outpatient interventions
- All pain medications are discontinued prior to program entry!
  - (Not true of all programs)

Typical Program

- Intensive Day Hospital or Inpatient Program
- Average 3-4 week duration
- Goals:
  - Return of physical function
  - Reduce pain
  - Improve associated symptoms
Physical

• 4-5 hours daily PT/OT
  – Timed activities
  – Desensitization
  – ADLs
  – Therapeutic Outings
  – Focus on Function

Self-regulation & Coping

• Talk Time (Cognitive Therapy)
  – Individual & Group

• Music Therapy
  – Individual & Group

• Therapeutic Art
  – Individual & Group

• Yoga – Three 1.5 hour sessions/wk

• Self-regulation – Two 30-minute sessions/wk
  – Guided imagery, relaxation breathing, progressive muscle relaxation, etc.
Additional Programming

• Psycho-educational Testing
• Parent Group 2-3 hours/wk
• Individual parent sessions as needed
• School return coordination
• Daily Team Huddle

OUTCOMES
After 5 months and 8 blocks...
7 days later with just PT & OT

RAPS Patients

<table>
<thead>
<tr>
<th>Baseline demographics</th>
<th>N=125</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age in years, mean (SD, range)</td>
<td>15.4  (1.9, 10-19)</td>
</tr>
<tr>
<td>Female, n (%)</td>
<td>105   (84)</td>
</tr>
<tr>
<td>Race, n (%)</td>
<td></td>
</tr>
<tr>
<td>White</td>
<td>107   (85.6)</td>
</tr>
<tr>
<td>Black</td>
<td>11    (7.2)</td>
</tr>
<tr>
<td>Other</td>
<td>7     (7.2)</td>
</tr>
<tr>
<td>Pain Characteristics, n (%)</td>
<td></td>
</tr>
<tr>
<td>Widespread pain</td>
<td>103   (82.4)</td>
</tr>
<tr>
<td>CRPS or localized pain</td>
<td>22    (17.6)</td>
</tr>
<tr>
<td>Duration of Pain in years, mean (SD, range)</td>
<td>3.7   (3.7, 0.1-15.6)</td>
</tr>
<tr>
<td>Conversion Symptoms, n (%)</td>
<td>37    (30)</td>
</tr>
<tr>
<td>Program Duration in weeks, mean (SD, range)</td>
<td>3.9   (1.1, 2-11)</td>
</tr>
</tbody>
</table>
Improvements in Pain and Disability

- Average Pain (VAS 0-100)
- Disability (FDI 0-60)

- Baseline
- Program End
- 1-mo
- 6-mo
- 12-mo

P<.05
P<.001

Psychological Improvements

- Depression/Anxiety (0-32)
- Catastrophizing (1-5)

- Baseline
- Program End
- 1-mo
- 6-mo
- 12-mo

P<.01
School Functioning

Final Analysis

- Great kids who are in real need
- We can significantly not only address their symptoms (short term benefit), but also address psychological dysfunction (long term benefit)
- Very time consuming
- Very rewarding
Additional Resources

stopchildhoodpain.org
www.childrensmercy.org/RAPS/

References


References


Thank You!

cmhoffart@cmh.edu
@CMHPain