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

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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

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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

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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

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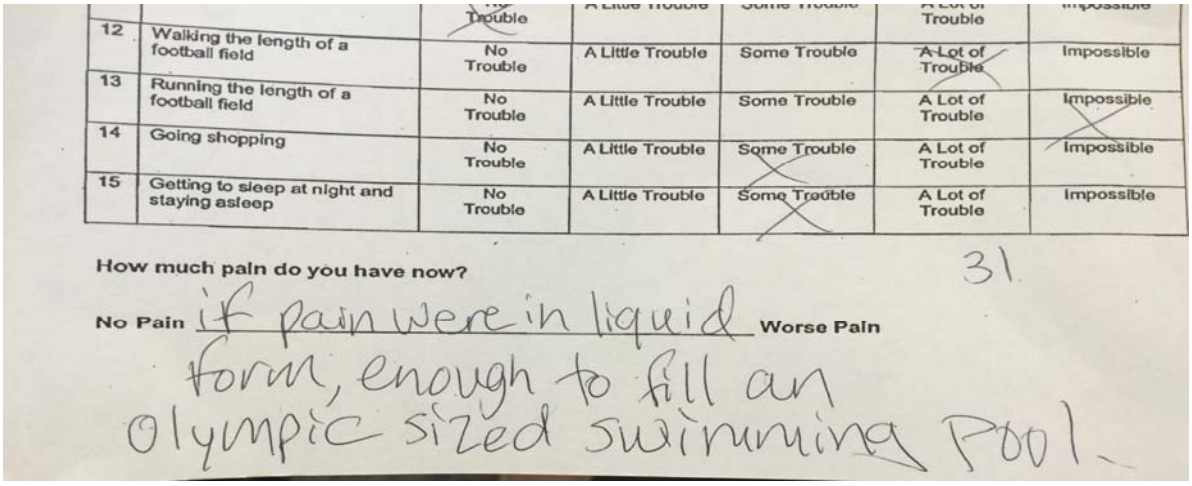

		Trouble			Trouble	
12	Walking the length of a football field	No Trouble	A Little Trouble	Some Trouble	A Lot of Trouble	Impossible
13	Running the length of a football field	No Trouble	A Little Trouble	Some Trouble	A Lot of Trouble	Impossible
14	Going shopping	No Trouble	A Little Trouble	Some Trouble	A Lot of Trouble	Impossible
15	Getting to sleep at night and staying asleep	No Trouble	A Little Trouble	Some Trouble	A Lot of Trouble	Impossible

How much pain do you have now? 31

No Pain if pain were in liquid form, enough to fill an olympic sized swimming pool. Worse Pain

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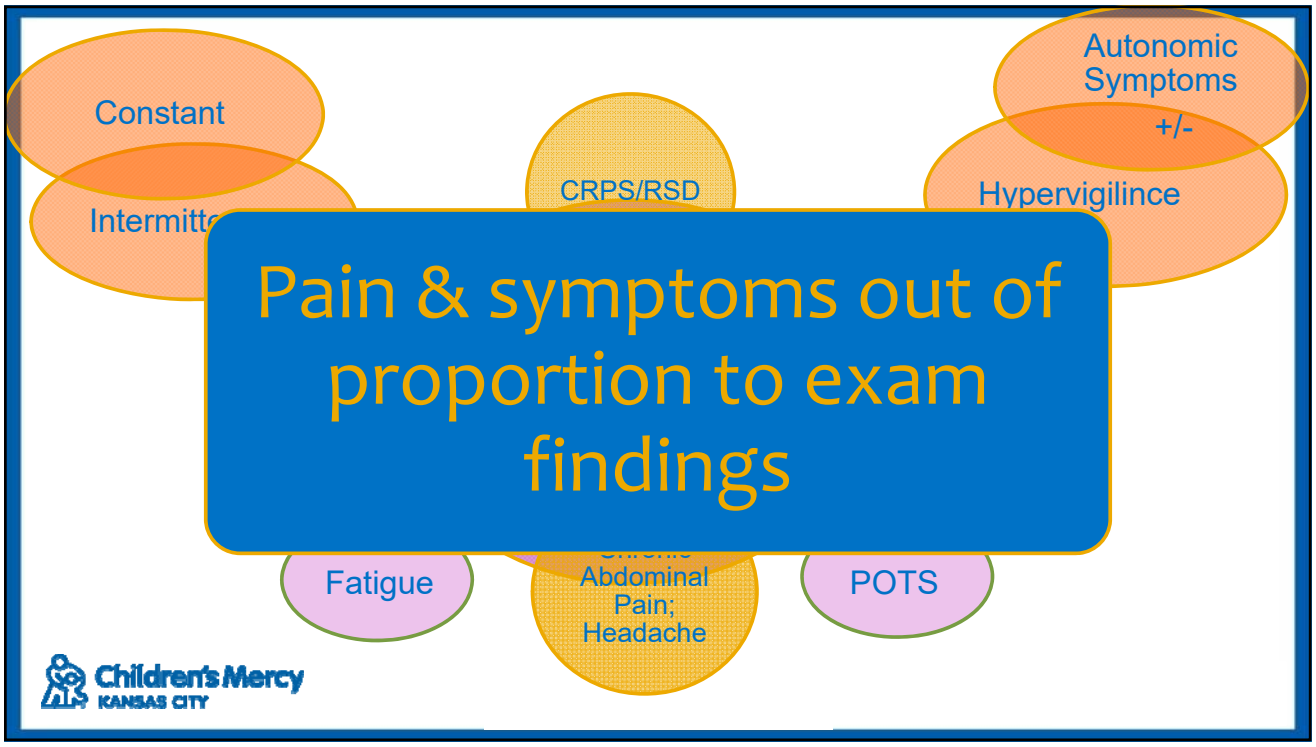

Juvenile Primary Fibromyalgia

-Full body pain

-Positive ROS

-Conversion

-Uses wheelchair at school

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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

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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)

Shoulder, right	Shoulder, left	Upper arm, right	Upper arm, left
Lower arm, right	Lower arm, left	Hip (buttock), right	Hip (buttock), left
Upper leg, right	Upper leg, left	Lower leg, right	Lower leg, left
Jaw, right	Jaw, left	Chest	
Upper back	Lower back	Neck	

WPI Total:_____

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

	No problem	Slight/Mild problem, generally mild or intermittent	Moderate, Considerable problem, often present	Severe, pervasive, continuous, life-disturbing problem
Fatigue	0	1	2	3
Waking Still Feeling Tired	0	1	2	3
Concentration or Memory Problems	0	1	2	3

Total:_____

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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?

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

(see instructions below) C Score: _____

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

SS Total (B + C) : _____

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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

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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)**

Arnold LM, et al. Pediatric Rheumatology (2016) 14:46 DOI 10.1186/s12969-016-0106-4

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Pregabalin Side Effects

Side effects

	Pregabalin	Placebo	
Major depression	2	0	
Choletithiasis	2	0	
Dizziness	30%	13%	
Nausea	22%	9%	
Increased weight	17%	0%	[30% our patients obese]
Somnolence	9%	4%	
Limb Pain	7%	0%	

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Arnold LM, et al. Pediatric Rheumatology (2016) 14:46 DOI 10.1186/s12969-016-0106-4

Complex Regional Pain Syndrome: "Cold Blue Foot"

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Clinical Characteristic	Children	Adults
Affected Extremity	Lower>> Upper (6:1)	Upper>Lower
Sex Ratio	Marked female 7:1	Moderate female 2-4:1
Inciting injury	Less often	More often
Exam findings	Skin cooler; neurologic symptoms less pronounced	More likely to have "stages" of change
Psychological Issues	More common	Less common
Prognosis	Excellent recovery in most cases	Variable – long term disability common

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

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Berde, Anesthesiology 2005

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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

Barkmann, C., et al. Soc Psychiatry (2011) 46:1003-1011

Simons LE, et al. J of Ped Psych 38

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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

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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

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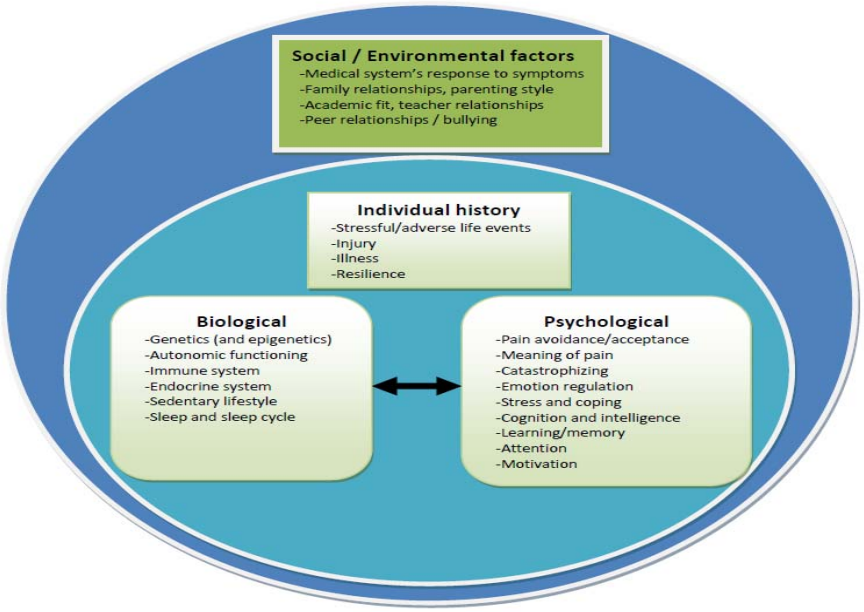
Complex Biopsychosocial Model



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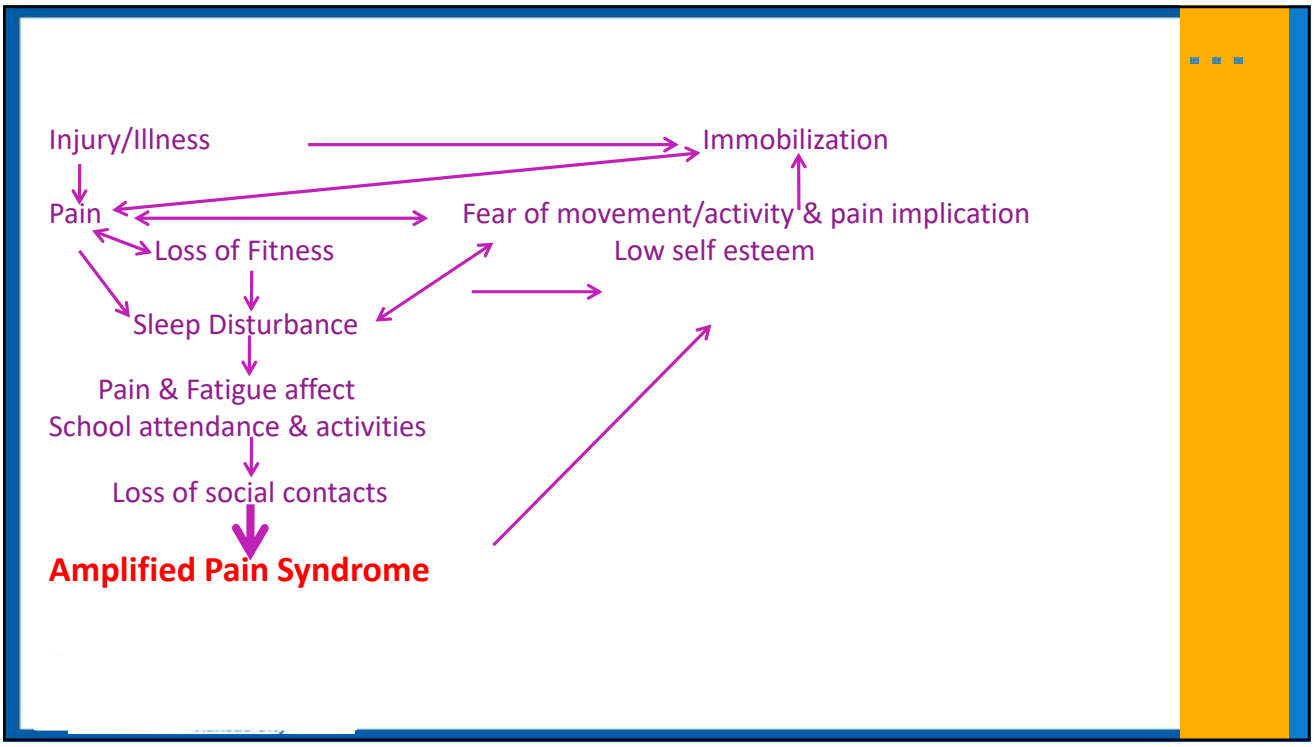

Working Model of Pain

- 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

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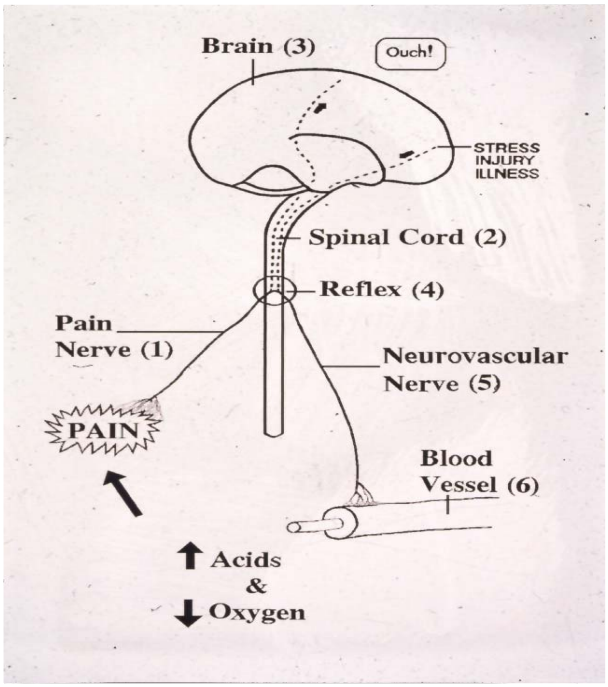

stopchildhoodpain.org

Courtesy David Sherry, MD

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WHO ARE THESE KIDS?

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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

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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

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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

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Social History Clues

- Major life events
- Role model for the same or chronic pain
- Typical personality
  - Mature
  - Excels
  - Pleaser
  - Perfectionist
  - Worrier
  - Sensitive

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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

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Autonomic Signs

- Cold
- Cyanotic
- Clammy
- Decreased pulse
- Dystrophic skin
- Check after exercise

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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

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LABS AND IMAGING

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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

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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!

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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!

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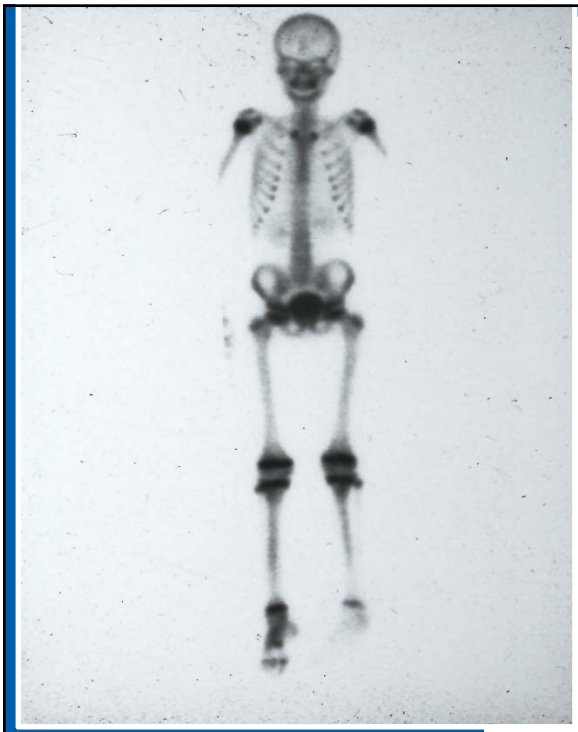

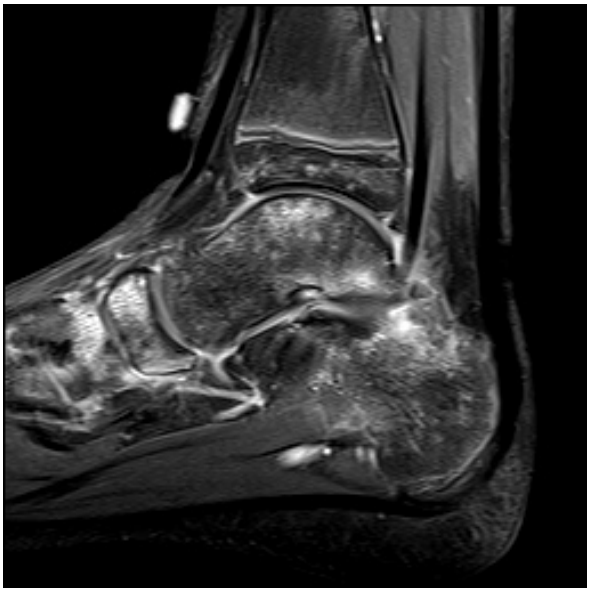

MANAGEMENT

Challenges in Management

- **Complex and Controversial tr**
  - Opioids
  - Anticonvulsants
  - IV sympathetic b
  - Surgical sympa
- Antidepressant trial ren/adolescen cebo respo nitude & small treatment effect
- Anticonvulsants may adverse effects on f cr of suicidal ideation & attempts
- Concern about publicati stry funded trials

Gregoire M, Finley GA. Drugs for c ical practice and the absence of evidence. Pain Res Manag 2013; 18:47–50.

Zernikow B, Dobe M, Hirschfeld G, et al. Please don't hurt me! A plea against invasive procedures in children and adolescents with complex regional pain syndrome (CRPS). Schmerz 2012; 26:389–395.

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Complex Biopsychosocial Model

Requires Multidisciplinary Treatment Approach

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

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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

•Kashikar-Zuck S, Sil S, Lynch-Jordan AM, et al. J Pain 2013

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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

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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"

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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

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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)

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Typical Program

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

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Physical

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

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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.

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Additional Programming

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

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OUTCOMES

After 5 months and 8 blocks…

7 days later with just PT & OT

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RAPS Patients

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

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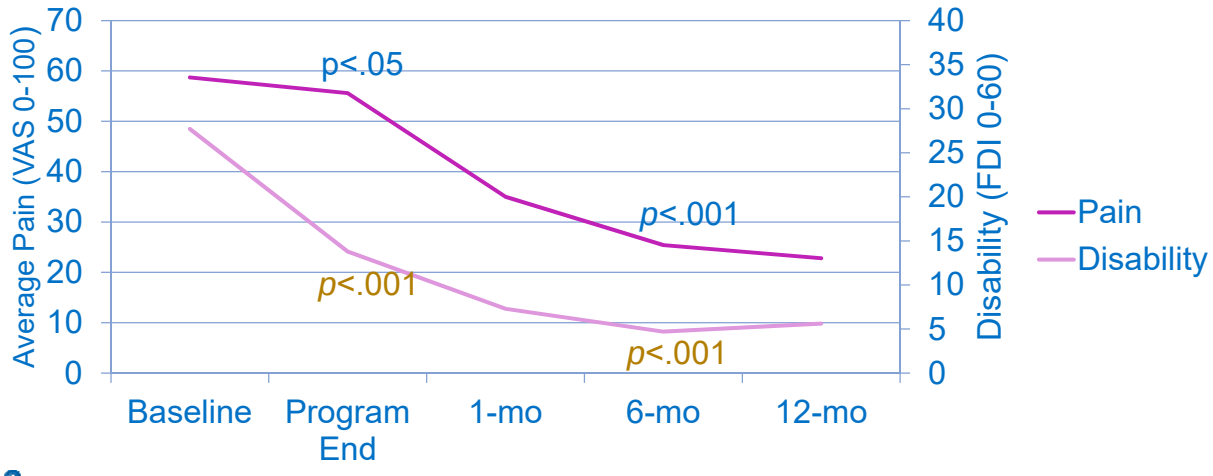
Improvements in Pain and Disability



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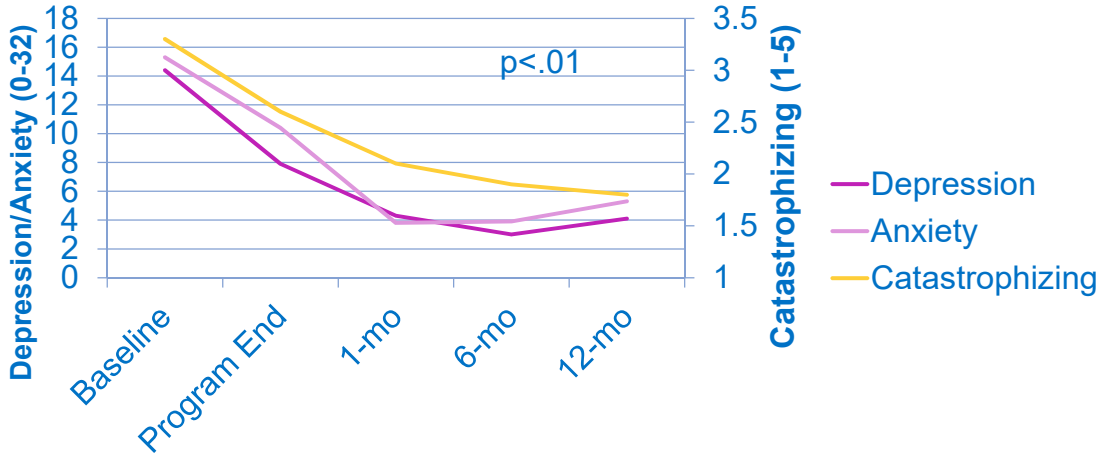
Psychological Improvements



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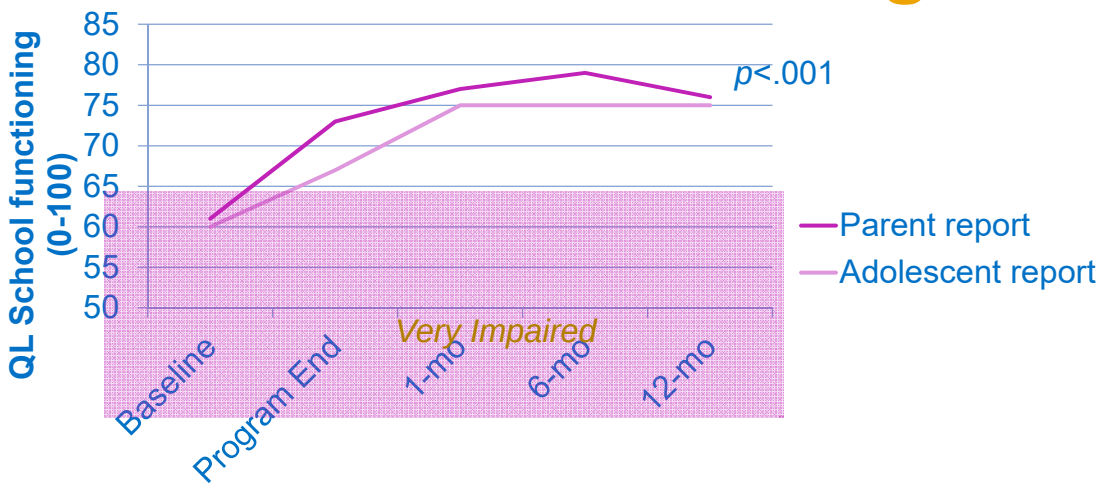
School Functioning



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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

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Additional Resources

stopchildhoodpain.org

www.childrensmercy.org/RAPS/

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Thank You!

cmhoffart@cmh.edu
@CMHPain

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