Complex Regional Pain Syndrome

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Objectives

• Define the epidemiology of CRPS
• Evaluate the pathophysiology of CRPS
• Review the clinical symptoms of CRPS
• Discuss the differential diagnosis of CRPS
• Explain the treatment of CRPS
Epidemiology

• More common in females
• Mean age at diagnosis is 12yo
• Lower extremity more often than upper extremity
• CRPS Type I predominant form
• ?Genetic component
• Family history
Normal Pain Processing

3. A signal is sent via the _ascending_ tract to the brain, and perceived as pain.

4. The _descending_ tract carries modulating impulses back to the dorsal horn.

2. Impulses from afferents depolarize dorsal horn neurons, then, extracellular Ca^{2+} diffuse into neurons causing the release of Pain Associated Neurotransmitters:
   - Glutamate
   - Substance P

1. Stimulus sensed by the peripheral nerve (e.g., skin)

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Gate Control Theory

Pain signals sent along acute and chronic pain fibres

Gate in spinal cord is open, so pain messages get through to the brain

Pain signals are still sent via acute and chronic pain fibres

Rubbing the injury sends messages along touch and pressure fibres

Messages from touch and pressure fibres activate the gating nerve cell in the spinal cord

The gate in the spinal cord is closed, so no pain messages get through to the brain
# Gate Control Theory

<table>
<thead>
<tr>
<th>Fiber Type</th>
<th>What they Transmit</th>
<th>Characteristics</th>
<th>Effect on Gate</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Small Fibers</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Aδ</td>
<td>Sharp, Prickly Pain</td>
<td>Thin, myelinated, slow</td>
<td>Opens</td>
</tr>
<tr>
<td>C-Fibre</td>
<td>Dull, Aching Pain</td>
<td>Thin, unmyelinated, slow</td>
<td>Opens</td>
</tr>
<tr>
<td><strong>Large Fibers</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Aβ</td>
<td>Non-painful Stimuli</td>
<td>Thick, myelinated, fast</td>
<td>Closes</td>
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</table>

1. C fibres transmit pain from the periphery.
2. The projection neurone transmits pain signals to the sensory cortex.
4. Mechanical stimulation activates the inhibitory neurone which is able to inhibit the projection neurone to limit pain.
Central Sensitization

brain: malfunctioning of central pain inhibitory pathways arising from periaqueductal gray matter and rostral ventral medulla in brainstem

brain: cognitive-emotional sensitisation

brain: sensory-motor conflict

brain: malfunctioning of descending pain-modulating pathways

dorsal horn: hyperexcitability of NMDA-receptor sites of second-order neurons & progressive increase in discharges from second-order neurons

injury / trauma / arthritis

peripheral tissues: enhanced responsiveness of nociceptive endings

overactive ascending pain facilitatory pathways
Central Sensitization

• Pain modifies the way the central nervous system works
• Sensitized patients are more sensitive
• Pain echoes
• Not the cause of pain but the cause of the chronicity
• Disease of over reaction to threats
• Pain Hallucinations
Experimental Evidence

- Abnormalities in Quantitative Sensory Testing
- Neurochemical abnormalities
- Changes in hypothalamic-pituitary-adrenal axis
- Neuroimaging
- Epigenetic and pathophysiologic changes
- Decreased opioid binding potential
Clinical Diagnosis

Table. Budapest Criteria for CRPS

All of the following statements must be met:
- The patient has continuing pain that is disproportionate to any inciting event
- The patient has at least 1 sign in 2 or more of the categories below
- The patient reports at least 1 symptom in 3 or more of the categories below.
- No other diagnosis can better explain the signs and symptoms.

<table>
<thead>
<tr>
<th>No.</th>
<th>Category</th>
<th>Signs/Symptom</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Sensory</td>
<td>Allodynia (pain to light touch and/or temperature sensation and/or deep somatic pressure and/or joint movement) and/or hyperalgesia (to pinprick)</td>
</tr>
<tr>
<td>2</td>
<td>Vasomotor</td>
<td>Temperature asymmetry and/or skin color changes and/or skin color asymmetry</td>
</tr>
<tr>
<td>3</td>
<td>Sudomotor/edema</td>
<td>Edema and/or sweating changes and/or sweating asymmetry</td>
</tr>
<tr>
<td>4</td>
<td>Motor/trophic</td>
<td>Decreased range of motion and/or motor dysfunction (weakness, tremor, dystonia) and/or trophic changes (hair/nail/skin)</td>
</tr>
</tbody>
</table>
Psychological Factors

• Stress related disorder
• High levels of anxiety
• Tendency toward over achieving or to learning difficulties
• School absenteeism
• Two distinct types of families:
  • high levels of cohesion, expression and organization and with low to average levels of conflict
  • high overt conflict levels and low levels of family cohesion, expressiveness and organization
• Parental enmeshment with their child
Differential Diagnosis

• Fibromyalgia
• Hypermobility
• Myofascial pain
• Fracture/strain/sprain
• Arthritis
• Spondyloarthropathy
• Leukemia
• Progressive diaphyseal dysplasia
• Idiopathic juvenile osteoporosis
• Thyroid disease

• Spinal cord tumors
• Chronic recurrent multifocal osteomyelitis
• Raynaud’s disease
• Fabry’s disease
• Erythromelalgia
• Chronic compartment syndrome
• Peripheral mononeuropathy
• Vitamin D deficiency
How do we treat pain?

• Conventional Analgesics
• Nonconventional Analgesics
• Interventional Procedures
• Physical Therapy/Occupational Therapy
• Cognitive Behavioral Therapy/Psychotherapy/Biofeedback
• Complementary Medicine
• Integrative therapies
“Neuro-Education”

• Pain is real
• Problem is not with the tissues themselves but in the pain system itself
• Disordered pain processing system interconnects with other components of the nervous system
• “Real” disease or disorder
• Treatment is available
Inpatient Rehab Program

• Fail outpatient program/Limited resources
• Psychological readiness
• Understand the goals of the program
• Accept the structure
• Prepare for discharge
Program Overview

- 2 week program
  - Scheduled admission under pain service
  - Changed from one week to two in 2013
- Admit conference, mid-stay conference, and discharge conference
- Parents present at all conferences, in the evenings 5-7pm, half day on Saturdays, and full day on Sundays
- Limit to one patient with chronic pain at a time
Program Overview

• 3 hours of therapy (PT and OT), 6 days per week
• CBT with clinical therapist, Neuropsychological testing, Biofeedback
• School time
• Complementary therapies - massage, acupuncture, pet therapy, art, music, child life
• Therapeutic Outings
  • Continue to work on therapy goals
  • Community re-entry
  • School outing in some cases
**Prognosis**

• More favorable than for adults
• Relapses are common
• Younger age at time of injury correlates with less pain, better function, fewer remaining signs of autonomic dysfunction on follow-up and a shorter total duration of symptoms. Younger patients more likely to return to sports
It Takes Team Work

Physician
Dr. Gandhi, Dr. Brooks, Dr. Borg
Dr. Chang, Dr. Yamout

Neuropsychologist
Marsha Gabriel, Beth Coluca

Clinical Therapist
Matthew Reed

Midlevels
Fred Ortiz, Wendy Wood

Nurses
Barbara Deleon, Chansey Pullen

Therapy Dogs

Art Therapist

Music Therapist

Psychophysiologist
Laura Barnes

Physical Therapist
Bruce Morgan, Jenny Arey, Melissa Bro, Erin, Nikki, Kendra, Lindsey

Occupational Therapist
Leanna, Jamie, Hannah

Billing
Phyllis Bomberger

Rehab Techs

School Teacher
Jan Sordini

Massage Therapist
Joni Carroll
Parental Involvement

Conquering Your Child’s Chronic Pain
A Pediatrician’s Guide for Reclaiming a Normal Childhood
Lonnie K. Zeltzer, M.D.,
Director of the Pediatric Pain Program, JCA Shriners Children’s Hospital
and Christina Blackett Schlank

Relieve Your Child’s Chronic Pain
Elliot J. Krane, M.D.,
with Deborah Mitchell

When Your Child Hurts
Effective Strategies to Increase Comfort, Reduce Stress, and Break the Cycle of Chronic Pain
Rachael Coakley, Ph.D.
References