

# Complex Regional Pain Syndrome (CRPS)

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## Financial Disclosures

- I have no relevant financial disclosures.

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- BSEE Ole Miss 2005
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## Complex Regional Pain Syndrome

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- Basics
- Disease Progression
- Diagnosis
- Outcomes
- Treatment

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## Complex Regional Pain Syndrome



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## CRPS Basics

- Disorder characterized by continued debilitating pain that is inordinate to the provoking event.
- Damage to:
  - Peripheral Nerve, or
  - Region without direct nerve involvement
- Multitude of Disturbances
  - Sensory and Motor deficits
  - Vasomotor disturbances: dilation/constriction of blood vessels
  - Sudomotor disturbances: change in sweat
  - Edema
  - Trophic changes: changes in skin texture, atrophy of the finger pulps, changes in the nail beds, changes in hair growth, and slower skin healing.

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## CRPS Basics

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- Classification (first done by International Association for the Study of Pain in 1994)
- Updated in 2003 to include Budapest Criteria
  - currently considered the most accepted diagnostic approach
- Old names in literature include:
  - Reflex Sympathetic Dystrophy, Algodystrophy, Causalgia, Sudeck's atrophy among others.

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## CRPS Basics

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- 4:1 Female to Male
- Highest frequency in fourth decade of life
- No differences in race
- Fracture most common cause and more often in upper extremity.
- Estimated Incidence following Surgery:
  - Shoulder: 0.9-11%
  - Distal radius: 22-39%
  - Carpal tunnel: 2-5%
  - Dupuytren's contracture surgery: 4.5-40%
  - Tibial fracture repair: 28.6-33.3%

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## CRPS Mechanism- A Theory

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- A large majority of researchers believe:
- Continued noxious stimuli related to damaged areas may lead to sensitization of the peripheral and central nervous system.
- Peripheral injured nociceptors lead to an overexpression of neurotransmitters and eventual hypersensitivity due to repetitive stimulation.
- The overstimulation of the peripheral nervous system leads to increased nociceptive awareness in the central nervous system.
- A distorted perception of nociceptive stimuli causes a continuous torrent of downstream ramifications and an ongoing exaggerated response to the initial stimuli.

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## 2 Types

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- Type I
  - Previously Reflex Sympathetic Dystrophy
  - Develops after trauma or injury
    - sprains
    - fractures
    - visceral disease
    - even immobilization
  - Does NOT involve a specific peripheral nerve
- Type II
  - Previously Causalgia
  - Develops after peripheral nerve injury

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## Disease Progression

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## Disease Progression

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- Initial Symptoms typically within 6 weeks
- Early Symptoms include:
  - edema
  - sensory changes
  - distal vasomotor changes
  - pain at the area of insult
- Over time, symptoms become less localized.

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## 3 Stages?

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- Stage 1 (acute): edema, vasomotor dysregulation, severe tenderness, and allodynia.
- Stage 2 (dystrophic): skin and muscle atrophy, brittle nails and atrophy, intense proximal pain, mottled skin, and brawny edema.
- Stage 3 (atrophic): skin becomes pale, smooth, shiny, and cyanotic, contractures and flexion deformities, pain decreases, and vasomotor changes stop.
- Currently, there is little support for these sequential stages

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## 3 Subtypes?

- New evidence supporting CRPS described as three different subtypes:
- Subtype 1
  - vasomotor changes
- Subtype 2
  - neuropathic pain
  - sensory disturbances
- Subtype 3
  - motor limitation and
  - trophic skin changes

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## Pain?

- Multiple Presentations:
  - Burning, stabbing, or a dragging character, to throbbing and constricting.
- A large proportion of pain is located in deeper structures (muscles, tendons, bones)

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## Other Findings

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- Edema/swelling present most of time
- Change in sweat inconsistent
- Reduced strength more prevalent than normal strength
- Almost half have some form of movement disorder
- Sensory disorders tend to be the most consistent deficit and could be increased or decreased at the time of exam

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## Secondary/Associated Conditions

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- Sequelae of disuse including contractures
- Altered sensation of the affected limb
- Infection
- Edema
- Dystonia
- Myoclonus
- Dystrophic changes of nails, skin, and hair
- Osteopenia (Sudeck's atrophy)
- Impaired sympathetic vasoconstriction may also occur and lead to presyncope or syncope.

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## Diagnosis

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## Budapest Criteria

- The patient must have continuing pain, which is disproportionate to any inciting event.
- At least 1 symptom in 3 of 4 categories
- At least 1 exam finding in 2 of 4 categories
- There is no other diagnosis that better explains the symptoms and signs
- A diagnostic subtype: CRPS-not otherwise specified (NOS)  
Created to capture those patients who did not meet the new clinical criteria but whose signs and symptoms could not be better elucidated by any other diagnosis .

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## Budapest Symptoms

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|                        |   |
|------------------------|---|
| <b>SENSORY</b>         | SENSORY REPORTS OF HYPERESTHESIA AND/OR ALLODYNIA   |
| <b>VASOMOTOR</b>       | REPORT OF TEMPERATURE ASYMMETRY AND/OR SKIN COLOR CHANGES   |
| <b>SUDOMOTOR/EDEMA</b> | REPORT OF EDEMA AND/OR SWEATING CHANGES AND/OR SWEATING ASYMMETRY   |
| <b>MOTOR/TROPHIC</b>   | REPORT OF DECREASED RANGE OF MOTION AND/OR MOTOR DYSFUNCTION (WEAKNESS, TREMOR, OR DYSTONIA) AND/OR TROPHIC CHANGES (HAIR, NAIL, OR SKIN) |

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## Budapest Exam Findings

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|                        |  |
|------------------------|--|
| <b>SENSORY</b>         | EVIDENCE OF HYPERALGESIA (TO PINPRICK) AND/OR ALLODYNIA (TO LIGHT TOUCH AND/OR DEEP SOMATIC PRESSURE AND/OR JOINT MOVEMENT)                                    |
| <b>VASOMOTOR</b>       | EVIDENCE OF TEMPERATURE ASYMMETRY BETWEEN LIMBS AND/OR SKIN COLOR CHANGES/ASYMMETRY  |
| <b>SUDOMOTOR/EDEMA</b> | EVIDENCE OF EDEMA AND/OR SWEATING CHANGES/ASYMMETRY  |
| <b>MOTOR/TROPHIC</b>   | EVIDENCE OF DECREASED RANGE OF MOTION AND/OR MOTOR DYSFUNCTION (MANUAL MUSCLE TESTING WEAKNESS, TREMOR, OR DYSTONIA) AND/OR TROPHIC CHANGES (HAIR, NAIL, SKIN) |

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## Diagnostic Testing

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## Diagnostic Testing

- Laboratory Studies
  - Used only to exclude other diagnoses
  - Nerve studies for neuropathies or nerve injury(Type II)
  - Rheumatologic labs, etc
- Imaging
  - Low sensitivity
  - Look for other source of pain
  - Triple phase bone scan: negative helps rule out
- Sympathetic Nerve Block
  - Positive response not diagnostic for CRPS but is an indicator of sympathetically maintained pain.

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## Outcomes?

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## Predictions of outcomes

- EARLY diagnosis and intervention KEY
  - Long term damage to nerves, bones, muscles, etc.
- Recurrence rate 10-30%
  - Higher in children

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## Mind Destroys Body

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- Negative disease perceptions experienced greater pain, disability, and kinesiophobia.
- Higher levels of anxiety, perception of disability, and pain-related fear have been shown to have a worsened disease course.
- Catastrophizing, commonly seen in CRPS, has been shown to increase pro-inflammatory cytokines which may also be linked to disease progression.

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## Treatment

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## Where to start

- Awareness of complex regional pain syndrome (CRPS) by general practicing physicians is poor, which often leads to delays in treatment.
- Aggressive Treatment should not be delayed
- Rehab + Meds mainstay of early treatment

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## Traditional Treatments

- PT/OT: Range of motion, desensitization, promote normal positioning, decrease muscle guarding, increase functional use of the extremity.
- Mirror box therapy
- Graded motor imagery
  - Teaching your brain that the affected limb is not a painful threat.
- Tactile (or sensory) discrimination training
  - Differentiate between various sensations
- Transcutaneous nerve stimulation (TENS)
  - Decreases pain and edema
- Cognitive Behavioral Therapy

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## Non-Traditional Treatments

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- Complementary Therapies
  - Isolated studies show benefit
    - Acupressure, acupuncture, herbal medicines, anti-inflammatory type diets, and natural supplementation
- Hyperbaric oxygen
- Electroconvulsive therapy
  - Treat depression may help CRPS-case reports

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## Pharmacologic Treatments

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- NSAIDS: mixed results in trials
- Antiepileptic drugs
  - Gabapentin has moderate evidence for pain
  - Other: amitriptyline, pregabalin(Lyrica)
- Bisphosphonates
  - Studies show consistent statistically significant effects in pain relief, functional improvement and overall improvement
- Calcitonin
- Corticosteroids: frequently used, weak evidence
  - Better for post stroke than post trauma
- Opioids: lack of evidence to support long term use
- Ketamine: temporary relief, does not affect function
- Topicals

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## Interventional Treatments

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- Sympathetic block – generally 1<sup>st</sup> choice
  - Stellate ganglion blocks for upper extremity
  - Lumbar sympathetic blocks for lower
  - Best if skin discoloration or temp changes
  - Do series if progressive improvement
- Radiofrequency sympathectomy
- Intravenous regional blocks
  - Tourniquet then IV med in to affected limb
  - Some meds help pain

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## Interventional Treatments

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- Spinal Cord Stimulation
  - Good evidence for long term pain, satisfaction and quality of life
  - Limited evidence for functional improvement
- Dorsal Root Ganglion Stimulation
  - Better than SCS for lower in some studies
- Intrathecal infusion pump:
  - Opioid(no), Clonidine(maybe) or Baclofen(maybe)

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## Surgical Options

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- Amputation
  - End Stage Option
  - Studies of Type I not II
  - 50% had improvement after amputation
  - Risk of recurrence in stump or phantom pain
- Surgical Sympathectomy
  - If multiple blocks help
  - Best if within 3 months of injury
- Motor Cortex Stimulation or Deep Brain stimulation

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## CONCLUSION

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## Conclusion

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- CRPS Sucks
- Recognize and Treat as fast as possible!
- Send to Pain Management as soon as ANY concern for CRPS.

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## Questions?

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