Unraveling the Myth Mysteries of Complex Regional Pain Syndrome

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History

- Been around for about 2000
  - Earliest description in the 5th century
    During times of war surgeons would describe patients complaining of severe burning pain which would last longer that the time normal healing

Psychological VS Real Pain

- Early physicians termed this pain “nervous” pain and tended to interpret this as a psychological problem or as a imaginary illness rather than as real pain.

War Time

- During the time of the Civil War by Dr. S. Weir Mitchell the father of modern neurology, defined the term causalgia as the burning pain.
- Later during WWI he further defined causalgia and we see descriptions of glossy skin and hypertrichosis.

World War II

WWII- new definitions develop

Reflex sympathetic dystropy – no nerve injury

Believed to be driven by the sympathetic nerves symptoms

Name Changes

- Complex Regional Pain Syndrome
- CRPS-I
  - RSD
  - No nerve injury
- CRPS-II
  - Causalgia
  - Nerve injury

Epidemiology

- Poorly understood due to diversity of clinical presentation
  - Mean age ranges from 36-46 with women predominating (60-81%)
  - Upper extremity is involved in 44-61% of cases
  - Lower extremity is involved in 16-46%
  - In some cases the etiology remains undetermined.

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Diagnostic Terms

- Neurological
  - Hyperesthesia: Increased sensibility to sensory stimuli
  - Allodynia: Perception of pain from a non-painful stimulus
  - Hyperalgesia: Excessive sensibility to a painful stimulus
  - Tremor: A quivering, esp. continuous of a convulsive nature
  - Dystrophic: Progressive weakening of muscle

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Diagnostic Terms

- Autonomic dysregulation
  - Vasomotor: Pertaining to the nerves having muscular control of the blood vessel walls
  - Sudomotor: Pertaining to nerves that control the secretion of sweat

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Diagnostic Terms

- Trophic changes: Concerned with nourishment, particularly with a type of efferent nerves believed to control the growth and nourishment of the parts they innervate (i.e. skin, nails, muscle and bone).

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Clinical Presentation

- Sensory signs and symptoms
  - Excruciating pain and hyperaesthesia
  - Decreased temperature
  - Decreased sensation to pinprick
    - Rommel and colleagues

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Clinical Presentation

- Autonomic signs and symptoms
  - Vasomotor or sudomotor changes
    - Swelling, color, and temperature changes and sweating abnormalities.

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IASP Diagnostic Criteria 1994

- The presence of an initiating **noxious event** / immobilization **without** (CRPS 1) or **with** (CRPS 2) evidence of **nerve injury**
- **Pain + allodynia, or hyperalgesia** that is **disproportionate** in severity to any inciting event
- Evidence at some point in time of edema, changes in skin blood flow (temp. difference of 1.1 deg. Celsius), or abnormal sudomotor activity in the region of pain (**trophic change**)
- Exclusion of conditions that would otherwise account for the degree of pain and dysfunction (**diagnosis of exclusion**)

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Clinical Presentation

- Motor and dystrophic signs and symptoms
  - Weakness
  - decreased ROM
  - tremor
  - dystonia
  - myoclonus

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Clinical Presentation

- Myofascial dysfunction
  - More common when upper extremity is affected
  - Related to the duration of disease

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CPRS I (RSD) Etiology

- Noxious Event
- UE > LE
- Inciting Event can include:
  - Minor trauma
  - Sprains
  - Bone fractures
  - Surgery
  - MI
  - Stroke

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The Clinical Course of CRPS I (RSD)

- Pain expands along the limb or migrates to other body parts in nearly 70% of patients
- The pain becomes bilateral in up 50% of cases.

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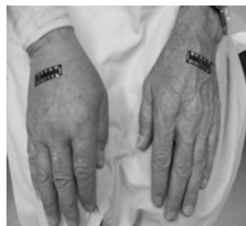
Three Stages of CRPS I (RSD) Stage 1

- Defined by Bonica
  - Stage 1 or acute stage features pain, edema, warm skin, and increased swelling.



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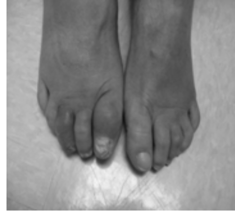
Stage 2

- Stage 2 or dystrophic stage is marked by cold, dry skin, and trophic changes.



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Stage 3

- Stage 3 or atrophic stage shows atrophied skeletal muscles and bones, joint contractures, progressive loss of function, and persistent pain.

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CRPS I (RSD) Clinical Features

- Allodynia
- Altered sweating (absent, excessive, or reduced)

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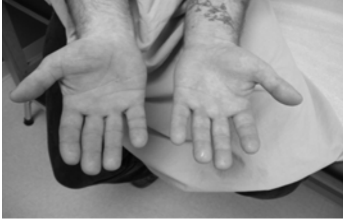
Sweat beading up on finger tips of CRPS patient



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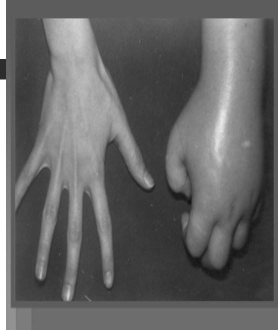
Skin Changes

- Atrophy of skin with loss of wrinkles (glossiness of skin)



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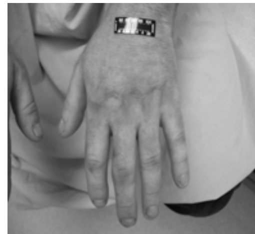
Color Changes

- Color changes of skin (cyanotic, erythematous, pale, blotchy)



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CRPS I (RSD) Clinical Features

- Involuntary movements: tremor, dystonia, spasms
- Joint stiffness (acute or chronic arthritic changes)
- Inappropriate warmth or coldness

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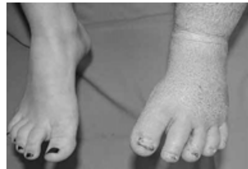
Skin Changes

- Dupuytren's and other contractures
- Hair changes
- Skin changes



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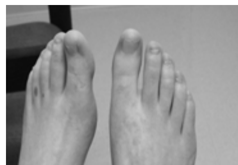
Nail Changes

- Muscle wasting and/or weakness
- Nails (brittle or clubbed; curved, thin, ridged)



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Other Signs / Symptoms

- Osteoporosis: spotty, localized, or widespread
- Pigmentation changes
- Subcutaneous atrophy or thickening
- Swelling

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CRPS II (Causalgia)

- Noxious event
- Disproportionate to inciting event
- Distribution over several nerve paths
- Evidence of edema
- Skin blood flow changes
- Abnormal sudomotor activity
- Allodynia
- Hyperalgesia

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CRPS II (Causalgia)

- Is defined as a partial injury to a peripheral nerve or one of its major branches.
- Cardinal symptoms are:
  - spontaneous burning pain
  - hyperalgesia
  - mechanical dysfunction
  - cold allodynia.

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Pathophysiology

- Not completely understood
- Multiple mechanisms
  - Neuorgenic inflammation
  - Immunological mechanisms
  - Plastic changes in the sympathetic and CNS

Pathophysiology

- Evidence to support the hypothesis that sympathetic nervous system plays a role
- Clinically patient with type I CRPS
  - Impairment of sympathetic nervous system
    - Decreased sympathetic outflow
    - Increased adrenergic responsiveness

Pathophysiology

- Central mechanism
- Rommel et al
- Moihofner et al

Goals of Treatment

- Perform a comprehensive diagnostic evaluation
- Be prompt and aggressive in treatment interventions
- Assess and reassess the patient's clinical and psychological status
- Be consistently supportive
- Strive for maximal amount of pain relief and functional improvement

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Guidelines

- CRPS clinical pathway three domains
  - Rehabilitation
  - Pain management
  - Psychological treatment

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Rehabilitation

- Rehabilitation is the mainstay of CRPS treatment
  - Occupation and physical therapies are crucial to a patient progress through specific areas of the clinical pathway

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Rehabilitation

- Early stages of CRPS treatment
  - Adequate analgesia
  - Encouragement
  - Education about the disease process

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Rehabilitation

- Step two
  - Increase flexibility
  - Stretching
  - Strengthening
  - Postural correction
  - Electrical stimulation
  - Edema control

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Rehabilitation

- Final stage
  - Normalization of use,
  - Assessment of ergonomics and posture
  - Implementation of required modifications at home and at work

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Psychological Therapy

- Pain less than 2 months does not require formal psychological intervention
- After 2 months should have evaluation including treatment for
  - Anxiety
  - Depression
  - Personality disorders

Management

- Difficult to treat mechanisms of pain not well understood
- Close collaboration important
  - Psychologist
  - Physical and occupational therapist
  - Neurologist
  - Pain management

Management

- Treatment goals
  - Pain relief
  - Functional recovery
  - Psychological improvement

Pain management

- Pain is the major symptom of CRPS and treatment of pain varies from patient to patient.

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Pharmacologic Management of CRPS

- Few randomized controlled trials
  - Disability or liability claims lead to exclusion into clinical trials
- 2006 few trials with experimental drugs
  - Failed to show efficacy

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Pharmacologic Management of CRPS

- Anti-inflammatory drugs
  - Acute phase of the disease shows inflammatory process
    - Swelling
    - Erythema
    - warmth

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Pharmacologic Management of CRPS

- Antiseizure medications tricyclic antidepressants, and opioids mainstays of therapy
- Proven to be effective in treatment of other types of neuropathic pain
  - Post herpatic neuralgia
  - Diabetic neuropathy

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Pharmacologic Management of CRPS

- Bisphosphonates
  - Clodronate, pamidronate and alendronate
  - Selectively inhibit bone reabsorption
  - CRPS do manifest some degree osteoporosis in the involved extremity so may be helpful.

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Pharmacologic Management of CRPS

- Opioids
  - Used but do not work well for neuropathic pain
  - If using would recommend longer acting meds vs shorting acting meds.

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Interventional Therapeutic Techniques

- Intravenous regional sympathetic blocks
- Local anesthetic sympathetic blockade
- Stellate Ganglion block
- Lumbar sympathetic blocks

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Blocks

- Intravenous regional sympathetic blocks
  - Introduced in 1908 by Bier
  - Results showing long term advantages are questionable
  - Studies have shown some favorable results
    - Tountas and Noguchi (1991)
    - Zyluk (1998)

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Blocks

- Local anesthetic sympathetic blockade
  - Done by placing a needle tip the proximity of the sympathetic structures
    - Stellate ganglion
    - Lumbar sympathetic chain

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Stellate ganglion block

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Lumber sympathic block

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Benefits of blocks

- Sharma, et al
  - Benefits of doing blocks
    - Pain relief
    - Reduction in vasomotor symptoms
    - Improved mobility
    - Range of motion
    - Motor strength

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Spinal cord stimulation

- Newest therapeutic modality
- Mechanisms not widely know
  - Believed of effect neuromodulation and restore normal gamma y-aminobutyric acid levels to reduce neuropathic pain

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Spinal cord stimulation

- Taylor et al
  - 2 point mean reduction in Visual Analogue Pain Scale in patients with CRPS
  - Reports a lifetime cost savings of approximately $60,000
  - They were also able to show that almost 2/3 of both type I and II patients reported at least 50% improvement in pain scores.

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Spinal Cord Stimulation

- Harke, et al
  - Studied 29 patients
    - Showed SCS reduced deep pain and allodynia
    - Improvement in ADL's
    - 77% showed significant improvement in motor strength

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Guideline for implantation

- Screen carefully
  - Pyschological evaulation
  - Physical therapy evulation
  - Understand of goals of stimulation
  -

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Spinal Cord Stimlator leads at implantation

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Conclusions about SCS

- Symptom management
- Improving function
- Improving physical therapy goals
- Cost effective in the long term

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Peripheral Nerve Stimulation

- Limited literature
- Does show some promise due to localization of pain to the extremity
- Should be considered in patient resent to other forms of treatment

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Intrathecal Pain Pumps

- Considered only when patients have failed all other treatment options
- Medications vary but include
  - Morphine
  - Baclofen
  - Prialt

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