

# Complex Regional Pain Syndrome

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## Outline

- Definition
- Epidemiology
- Historical perspective
- Clinical features
- Diagnostic criteria
- Pathophysiology
- Investigations
- Treatment
- Prognosis



## Definition

- A **debilitating painful chronic condition** in a limb or extremity.
- characterised by **limb pain, and dysfunction** within the **motor, sensory and autonomic nervous systems**.
- **Pain is the leading symptom** and is usually **disproportionate (in time or degree) to the original/inciting event**.
- The pain is not restricted to a specific nerve territory or dermatome.

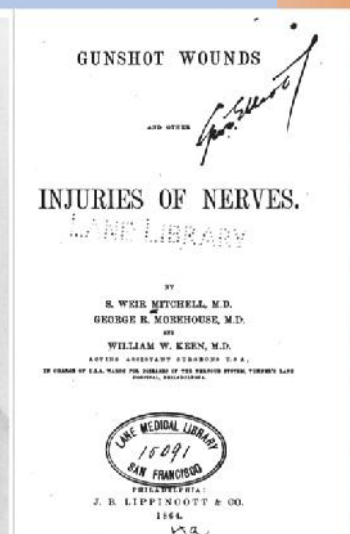
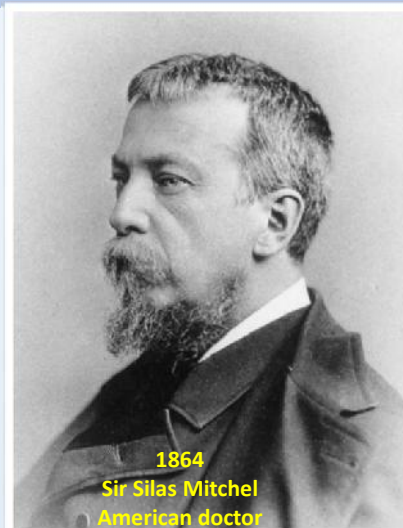
## Earlier names for CRPS

- Algodystrophy
- Causalgia
- Algoneurodystrophy
- Reflex sympathetic dystrophy
- Sudeck's atrophy
- Shoulder–hand syndrome
- Reflex neurovascular dystrophy
- Fracture disease

## Epidemiology and impact

- The European incidence rate of CRPS is 20–26/100,000 person-years (like MS)
- More common in women, with a female-to-male ratio of 2:1 to 4:1
- Incidence appears to be highest in postmenopausal women
- Usually post-traumatic (e.g. following radial fracture), although 10% cases have no obvious causal event.
- Usually unilateral although in approximately 7% of cases there is later involvement of additional limbs.
- 80% improve or resolve within 18 months.
- 50% continue with long-term functional problems
- 50% do not return to work due to chronic functional disability and residual pain

## Historical perspective



## Clinical features: "A CRPS limb"



### **Sensory**

Allodynia  
Hyperalgesia  
Hyperesthesia  
Hyperpathia  
Hypoesthesia

### **Autonomic**

Skin temperature  $\uparrow$  or  $\downarrow$   
Skin colour changes  
Sweating  $\uparrow$  or  $\downarrow$   
Oedema



### **Psychological**

Suffering  
Fear  
Anxiety  
Anger  
Depression  
Failure to cope  
Behavioural illness



### **Motor**

Weakness  
Tremor  
Dystonia  
Myoclonus



### **Trophic**

Hair growth  $\uparrow$   
Nail growth  $\uparrow$  or  $\downarrow$   
Contraction and fibrosis of joints and fascia,  
Glossy skin  
Hyperkeratosis  
Skin atrophy



## Stages

Stages	Pain	Extremity	Skin	X-ray	Timeframe
1 (Acute)	Localised, severe and burning	Warm	Dry and red	Normal	Within weeks of injury
2 (Dystrophic)	More diffuse and throbbing	Cold/cyanotic/ oedematous Muscle wasting	Sweaty and brawny	Osteoporosis	Within months of injury
3 (Atrophic)	Less severe and can involve other extremities	Severe muscle atrophy and contractures Limitation of movement	Glossy and atrophic	severe demineralisation	Within years after injury

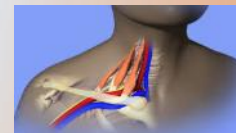
## Types

Signs and symptoms	CRPS Type 1 (formerly termed reflex sympathetic dystrophy/RSD)	CRPS Type 2 (formerly termed "causalgia")
Precipitating event	Sometimes	Yes
Peripheral nerve injury	<b>No</b>	<b>Yes</b>
Physiological change in affected limb	Yes	No
Progressive	Yes	Sometimes
Bone atrophy	Yes	No
	90% of clinical presentations	10% of clinical presentations

- **Warm:** increased skin temperature at the onset of symptoms
- **Cold:** decreased skin temperature at the onset of symptoms

## Differential Diagnosis

- Infection (bone, soft tissue, joint or skin)
- Orthopaedic mal-fixation
- Joint instability
- Arthritis or arthrosis
- Bone or soft tissue injury
- Compartment syndrome
- Neural injury / neuropathy (DM, EToH)
- Thoracic outlet syndrome (due to nerve or vascular compression)
- Arterial insufficiency
- Raynaud's disease
- Lymphatic or venous obstruction
- Gardner–Diamond syndrome
- Brachial neuritis or plexitis
- Erythromelalgia (may include all limbs)
- Self-harm



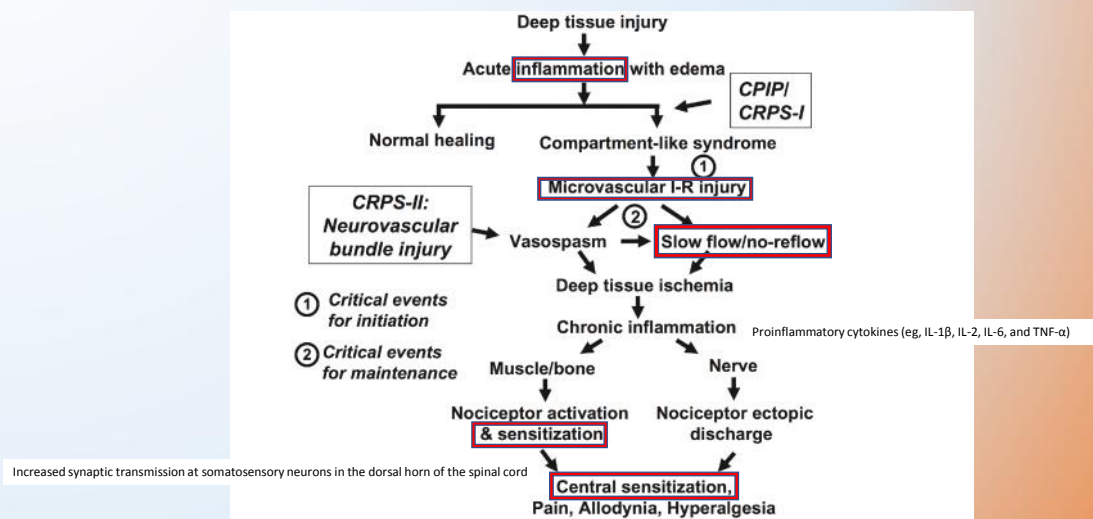
## Diagnostic criteria (Budapest Criteria)

Table 1 Diagnostic criteria for CRPS (Budapest criteria)<sup>17</sup> (A–D must apply)<sup>1</sup>

A) The patient has continuing pain which is disproportionate to any inciting event	<input type="checkbox"/>	
B) The patient has at least one sign in two or more of the categories	<input type="checkbox"/>	
C) The patient reports at least one symptom in three or more of the categories	<input type="checkbox"/>	
D) No other diagnosis can better explain the signs and symptoms	<input type="checkbox"/>	
Category	Sign (you can see or feel a problem)	Symptom (the patient reports a problem)
1 'Sensory'	Allodynia (to light touch and/or temperature sensation and/or deep somatic pressure and/or hyperalgesia (to pinprick))	Hyperesthesia does also qualify as a symptom
2 'Vasomotor'	Temperature asymmetry and/or skin colour changes and/or skin colour asymmetry	
3 'Sudomotor/oedema'	Oedema and/or sweating changes and/or sweating asymmetry	
4 'Motor/trophic'	Decreased range of motion and/or motor dysfunction (weakness, tremor, dystonia) and/or trophic changes (hair/nail/skin)	



## Pathophysiology



## Investigations

- Clinical diagnosis (no "gold-standard" test)

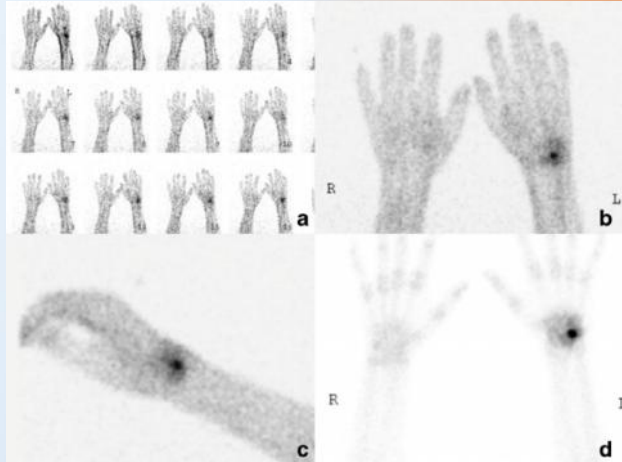
Some useful tests:

Side-by-side radiographs: spotty bone decalcification

Triple phase bone scintigraphy: increased radiotracer uptake (during the mineralization) in joints distant from the trauma site.

Autonomic tests: resting sweat output (RSO), the resting skin temperature (RST)

- MRIs/NCSs to exclude other diagnoses



## Range of services used by patients with CRPS

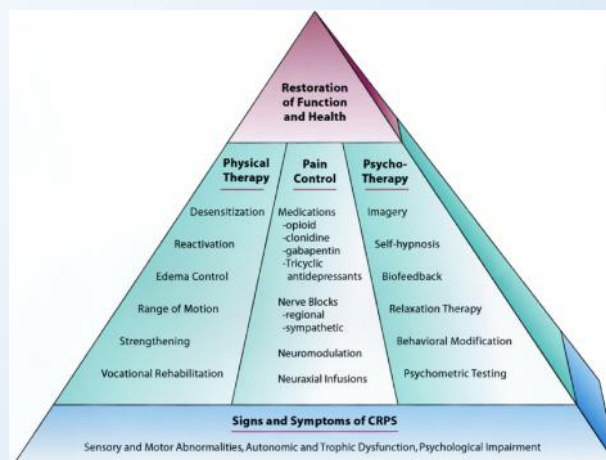




## Four pillars of treatment for CRPS – an integrated interdisciplinary approach



## Therapeutic goals and strategies





## Occupational/Physiotherapy

- Gentle limb movement (unless contraindicated for surgical reasons)
- Frequent attention to the affected limb
- Normalising the sensation of the affected limb, 'desensitisation', following appropriate guidance
- Progressing to more active use (e.g. weight bearing and stretching) when tolerated.
- Pain management programmes (PMPs)

## Therapeutic approaches

- Patient education and support
- Self-administered tactile and thermal desensitisation with the aim of normalising touch perception
- General exercises and strengthening
- Functional activities
- Mirror visual feedback
- Gait re-education
- Transcutaneous electrical nerve stimulation (TENS)
- Postural control
- pacing, prioritising and planning activities
- Goal setting
- Relaxation techniques
- Coping skills
- Hydrotherapy
- Sleep hygiene
- Oedema control strategies
- Vocational support
- Facilitating self-management of condition
- Splinting (generally short term, in acute CRPS)‡



Mirror visual feedback

## Yellow flags

- Iatrogenic factors, i.e. previous negative experiences with HCPs
- Poor coping strategies, e.g. ongoing 'guarding' of the limb despite education
- Involved in litigation/securing benefits (note that this may affect progress with treatment in some patients, but there must be no assumption that this applies in every patient)
- Overuse of appliances
- Distress/anxiety/depression
- Lack of willingness to set goals
- Passive in treatment sessions
- Inaccurate beliefs despite education
- Fear avoidance
- Negative family influences

## Management of pain

In early CRPS

NSAIDs: (Ibuprofen/Naproxen)

An anticonvulsant, such as gabapentin or pregabalin.

A TCA drug that is effective for neuropathic pain. (amitriptyline or nortriptyline)

Topical: lidocaine cream (2 to 5 %) or capsaicin cream (0.025 to 0.075 %)

Oral glucocorticoids

Pamidronate (single 60 mg IV)

### **Refer to pain team asap**

Trigger point/tender point injections, regional sympathetic nerve block, spinal cord stimulation, or epidural clonidine, IV Baclofen, Botox.

## Experimental treatments

- Anaesthetic blockade with specific agents (lidocaine, clonidine, labetalol)
- Analgesic cream (Ketamine, Pentoxifylline, Clonidine, DMSO)
- Brachial plexus analgesia (low dose morphine)
- Combined spinal cord stimulation and intrathecal therapy
- Dry needling
- Electroconvulsive therapy (ECT)
- Hyperbaric oxygen therapy
- Ketamine PO/IV (NMDA receptor antagonists)
- Topical capsaicin
- Surgical sympathectomy
- Plasma exchange
- Topical lidocaine – patches

## Psychological intervention

- Psychologic assessment and therapy in high risk patients: (CBT)
  - CRPS of more than two months duration at presentation
  - Insufficient response to treatment
  - Suspected comorbid psychologic or psychiatric disorder

## Prognosis

- Prognosis is uncertain!
- A substantial proportion of pts have some degree of prolonged disability.
- Recurrence of CRPS is not uncommon (10-30%).

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**Thank you for listening, any  
questions?**

