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

- Mid 16th century
  - Ambroise Paré
  - French surgeon
- 1864
  - Sir Silas Mitchell
  - American doctor
Clinical features: “A CRPS limb”

Sensory
Allodynia
Hyperalgesia
Hyperesthesia
Hyperpathia
Hypoesthesia

Motor
Weakness
Tremor
Dystonia
Myoclonus

Autonomic
Skin temperature ↑ or ↓
Skin colour changes
Sweating ↑ or ↓
Oedema

Psychological
Suffering
Fear
Anxiety
Anger
Depression
Failure to cope
Behavioural illness

Trophic
Hair growth ↑
Nail growth ↑ or ↓
Contraction and fibrosis of joints and fascia,
Glossy skin
Hyperkeratosis
Skin atrophy

Stages

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

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

- **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)\(^a\) (A-D must apply)\(^b\)

<table>
<thead>
<tr>
<th>Category</th>
<th>Signs (you can see or feel a problem)</th>
<th>Symptoms (the patient reports a problem)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 &quot;Sensory&quot;</td>
<td>Allodynia (to light touch and/or temperature sensation and/or deep somatosensory pressure and/or hyperalgesia, to pinprick)</td>
<td></td>
</tr>
<tr>
<td>2 &quot;Vasomotor&quot;</td>
<td>Temperature asymmetry and/or skin colour changes and/or skin colour asymmetry</td>
<td>If you notice temperature asymmetry, must be (&gt;3^\circ)C</td>
</tr>
<tr>
<td>3 &quot;Sudomotor/edema&quot;</td>
<td>Oedema and/or sweating changes and/or sweating asymmetry</td>
<td></td>
</tr>
<tr>
<td>4 &quot;Motor/atrophy&quot;</td>
<td>Decreased range of motion and/or motor dysfunction (weakness, tremor, atrophy) and/or trophic changes (hair, nail, skin)</td>
<td></td>
</tr>
</tbody>
</table>

Pathophysiology

Increased synaptic transmission at somatosensory neurons in the dorsal horn of the spinal cord.

Inflammatory cytokines (e.g., IL-1β, IL-6, TNF-α, and IL-6)
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

Limb dysfunction

Pain intensity

Distress

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)
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%).

References

- Complex regional pain syndrome in adults, RCP UK guidelines for diagnosis, referral and management in primary and secondary care 2018
- Complex Regional Pain Syndrome I (Reflex Sympathetic Dystrophy), Srinivasia N. Raja, M.D., *Theodore S. Gribov, M.D., †Anesthesiology 2002; 96:1254–60
  
  [https://burningnightscrs.org/crpsrsd/](https://burningnightscrs.org/crpsrsd/)
Thank you for listening, any questions?