


eMedRef “Single Disease Card Template”

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[Rel. Topics]  (Pepid will create)

COMPLEX REGIONAL PAIN SYNDROME

Background

1. ICD-9 Code(s) (Pepid will do this)
2. Definition: chronic pain condition, not necessarily confined to specific nerve distribution, usually affecting one limb or portion of one limb, often characterized by allodynia (pain in response to non-noxious stimuli)
 - Complex regional pain syndrome I – no demonstrable nerve damage (previously known as reflex sympathetic dystrophy) About 90% of cases.
 - Complex regional pain syndrome II – demonstrable nerve damage to specific nerve (previously known as causalgia)
3. General Information:
 - Stage I: occurs shortly after an injury or with no apparent trigger. Burning/throbbing pain, vasomotor instability, hypesthesia, sensitivity to heat or cold, possibly edema
 - Stage II: progression of soft tissue edema, skin thickening & color changes (brawny), muscle wasting, early bone demineralization
 - Stage III: contractures, significant osteopenia, waxy skin, brittle nails

Pathophysiology

1. Pathology of Disease:
 - Not completely known
 - A disease of the sympathetic nervous system.
 - Multiple theories including sympathetic nervous dysfunction related to nociception, peripheral pain receptors hypersensitive to circulating catecholamines, changes in pain sensing in dorsal horn of spinal cord.
2. Incidence, Prevalence:
 - 1-5% of patients who have sustained peripheral nerve damage
 - 30% in patients post-Colles fracture
 - Many cases resolve spontaneously over 9-12 weeks).¹
3. Risk Factors: precipitating event often unknown. Inactivity can increase risk, especially after trauma
 - Trauma: sprain, fracture, nerve injury
 - Iatrogenic: after carpal tunnel release or arthroscopic surgery, venipuncture, IM injection
 - Medical conditions: diabetic neuropathy, malignant invasion of nerve plexus, etc.
 - Psychosocial stress is NOT a risk factor, but can develop as a result of the disease.
4. Morbidity / Mortality:
 - Morbidity: osteopenia (from inactivity), muscle atrophy, substantial psychosocial stress related to pain management

Diagnostics

1. Diagnosis based on history & physical exam **[SOR B]**²

2. History:
 - a. Chief complaint includes:
 - i. severe dysesthesia (spontaneous occurrence of pain in absence of painful stimuli)
 - ii. allodynia (painful response to thermal or mechanical stimuli that would not normally cause pain)
 - iii. hyperesthesia (prolonged/exaggerated response to painful stimuli)
 - b. HPI:
 - i. pain can be throbbing, burning, constant, or aching
 - ii. pain often begins days or weeks after triggering incident, and lasts much longer than anticipate for normal healing
 - iii. pain often triggered by change in temperature, light touch, movement, or psychosocial stress/excitement
 - iv. pain can progress proximally to involve entire limb
3. Physical Examination:
 - a. Compare affected & unaffected limb for: color, warmth, sensitivity to touch, edema, atrophy
 - b. progressive changes may occur
 - i. initially affected area warm, erythematous and dry, with accelerated hair and nail growth
 - ii. hair then becomes sparse, nails grooved and brittle
 - iii. skin becomes cool, cyanotic and moist
 - iv. swelling and vasomotor changes can lead to skin dystrophy
 - v. soft puffy edema changes to tight, shiny swelling and loss of skin creases
 - vi. atrophic limb develops: decrease in fat pads, digits thin & pointed, muscle spasms & wasting, joint thickening
 - vii. eventually marked bone and muscle atrophy, weakness and flexor tendon contractures
4. Diagnostic Testing: no definitive testing available
 - Laboratory evaluation
 - Diagnostic imaging
 - X-ray: may show osteopenia (69%)³
 - Bone scan may be helpful if x-ray non-diagnostic
 - Other studies: immersion in ice water triggers substantial pain similar to pain patient describes as characteristic
 - Diagnostic Criteria: Listed below are the International Association for the Study of Pain diagnostic criteria^{4,5} (note: other acceptable criteria include Bruehl's & Veldman's)
 - for complex regional pain syndrome type I
 - presence of initiating noxious event or cause of immobilization (this criterion not necessary for diagnosis)
 - continuing pain, allodynia or hyperalgesia with which pain is disproportionate to inciting event
 - evidence at some time of edema, changes in skin blood flow or abnormal sudomotor activity in painful region
 - diagnosis excluded by existence of conditions that would otherwise account for degree of pain + dysfunction
 - for complex regional pain syndrome type II
 - continuing pain, allodynia or hyperalgesia after nerve injury, not necessarily limited to distribution of injured nerve

- evidence at some time of edema, changes in skin blood flow or abnormal sudomotor activity in painful region
- diagnosis excluded by existence of conditions that would otherwise account for degree of pain + dysfunction

Differential Diagnosis

Key Differential Diagnoses:

- Musculoskeletal: sprain, fracture, bursitis, myofascial pain syndrome, rotator cuff tear
- Neurologic: postherpetic neuropathy, diabetic neuropathy, radiculopathy
- Infectious: cellulitis
- Psychosomatic: Munchhausen's, malingering (esp when secondary gain issues exist, e.g., workmen's compensation)

Extensive Differential Diagnoses

- Vascular: Reynaud's disease, vasculitis
- Rheumatic: systemic lupus erythematosus, rheumatoid arthritis

Therapeutics

- 1) Acute Treatment: diagnosis of CRPS made over weeks to months, acute treatment is not relevant
- 2) Further Management (24 hrs): NA
- 3) Long-Term Care: develop clear treatment plan, develop mutual goals and expectations for therapy.
 - a) Spontaneous resolution is frequent
 - b) Watchful waiting and psychological support appropriate early in course
 - c) Initiate medical and physical therapy simultaneously for optimal effect.¹ Expect some effect over 2-3 weeks, with gradual resolution of symptoms over months
 - d) Educate patient that pain does not represent ongoing injury, that mobilization & desensitization of affected area are keys to successful recovery.
 - e) Medical: Listed therapies may have some effect, and are unlikely to cause harm.
 - i) Topical DMSO: particularly helpful for analgesia during therapy **[SOR B]¹**
 - ii) IV bisphosphonates: requires 4-8 weeks of therapy, good long term effect **[SOR A]¹**
 - iii) Calcitonin **[SOR B]¹**
 - iv) Oral Corticosteroids (limited course) **[SOR B]¹**
 - f) Physical Therapy: mobilization of affected limb important, but painful

Follow-Up

- 1) Return to Office: follow-up by phone and/or in person to provide psychological support and assess efficacy of watchful waiting and/or interventions such as medical and physical therapy.
- 2) Refer to Specialist: not indicated unless diagnostic uncertainty
- 3) Admit to Hospital: not indicated unless unable to manage pain as outpatient (rare)

Prognosis

- 1) Not well documented.
- 2) Most patients do experience lessening of symptoms with combined medical, physical, and psychological therapy.
- 3) Many eventually reach level of remission that allows for normal daily activities.

Prevention

- 1) Early mobilization after trauma [SOR C]^{1,2}
- 2) Vitamin C 500 mg by mouth daily for 60-90 days reduces the risk of CRPS in elderly patients with wrist fractures [SOR A]²

Patient Information

- handout from [American Academy of Family Physicians](#)
- handout including organizations for more information from [National Institute of Neurological Disorders and Stroke \(NINDS\)](#)

Date Edited/Initials:

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