# Complex Regional Pain Syndrome of the Upper Extremity

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The diagnosis and management of complex regional pain syndrome is often challenging. Early diagnosis and intervention improve outcomes in most patients; however, some patients will progress regardless of intervention. Multidisciplinary management facilitates care in complex cases. The onset of signs and symptoms may be obvious or insidious; temporal delay is a frequent occurrence. Difficulty sleeping, pain unresponsive to narcotics, swelling, stiffness, and hypersensitivity are harbingers of onset. Multimodal treatment with hand therapy, sympatholytic drugs, and stress loading may be augmented with anesthesia blocks. If the dystrophic symptoms are controllable by medications and a nociceptive focus or nerve derangement is correctable, surgery is an appropriate alternative. Chronic sequelae of contracture may also be addressed surgically in patients with controllable sympathetically maintained pain. *(J Hand Surg 2011;36A:1553–1562. Copyright © 2011 by the American Society for Surgery of the Hand. All rights reserved.)*

**Key words** Chronic pain, complex regional pain syndrome, management, reflex sympathetic dystrophy, upper extremity.

After trauma or surgery, physiologic events and adaptations that contribute to pain (allodynia, hyperpathia, and cold sensitivity), dystrophic events, or impaired function are common. The abnormal prolongation or persistence of these events with persistent pain, cold sensitivity, autonomic dysfunction, microvascular abnormality, localized atrophy, and functional impairment constitutes complex regional pain syndrome (CRPS, also known as reflex sympathetic dystrophy and algodystrophy). Complex regional pain syndrome increases injury or surgical symptoms, delays recovery, and may compound morbidity by secondary complicating events such as arthrofibrosis and delayed healing. Complex regional pain syndrome can increase permanent impairment and strain the physician–patient relationship.

The purposes of this review were to define the subjective and objective manifestations of CRPS, to outline diagnostic criteria, discuss nonoperative and operative treatment options, elucidate common myths and misconceptions, and delineate standard-of-care issues.

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## DEFINITIONS

Complex regional pain syndrome is a clinical entity without a single pathognomic test or marker. Although multiple synonyms exist, the most common are reflex sympathetic dystrophy, causalgia, and algodystrophy (Table 1).

Based on the recommendation from the International Association for the Study of Pain, complex regional pain type 1 entails pain syndrome, autonomic dysfunction, trophic changes, and functional impairment without an identifiable peripheral nerve component. This entity corresponds to traditional reflex sympathetic dystrophy. Complex regional pain syndrome type 2 includes the above and identifiable nerve involvement (classic causalgia). The lessening of symptoms and clinical improvement after sympatholytic intervention



 

**TABLE 1. Definitions**

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| Pain: An unpleasant emotional response associated with actual or potential cellular damage |
| Analgesia: Absence of pain in response to an insult that should produce pain |
| Nociception: Response to an unpleasant (noxious) stimulus that produces pain in human subjects under normal circumstances |
| Allodynia: Pain in a specific dermatomal or autonomous distribution associated with light touch to the skin; a stimulus that is not normally painful |
| Hyperalgesia: Increased sensitivity to stimulation (includes allodynia and hyperesthesia) |
| Hyperesthesia: Increased sensitivity to stimulation (pain on response to a mild nonnoxious stimulus) |
| Sympathetic pain: Pain in the presence of or associated with overaction of the sympathetic pain fibers; by definition, the pain is relieved by sympatholytic interventions |
| Hypoesthesia: Decreased sensitivity to stimulation |
| Hyperpathia: Abnormally painful reaction to a stimulus (especially repetitive); often includes extended duration of pain, frequently with a delay |
| Dysesthesia: An unpleasant abnormal sensation |
| Paresthesia: An abnormal sensation |

is defined as sympathetically maintained pain (SMP). Failure of improvement after sympatholytic intervention including pharmacologic intervention or sympathetic blocks is termed *sympathetically independent pain*. Pain may be nociceptive or neuropathic. The former denotes a mechanical or inflammatory process that serves as a constant or intermittent source of pain initiation. "Neuropathic pain" refers to pain related to peripheral nerve irritation or excitability from compression, neuroma, neuroma-in-continuity, or inflammation. If this pain spreads beyond the normal distribution of the involved nerve and is associated with autonomic changes, trophic events, and functional impairment, CRPS type 2 is defined.

## WHY YOU SHOULD CARE

Postinjury and postoperative outcomes are negatively affected by CRPS. The recovery course is prolonged, complications are frequent, rehabilitation is impaired, stiffness is common, functional results are inferior, time to return to work is increased, litigation is increased, and patient satisfaction is compromised. Patients with poor outcomes and CRPS may be litigious. Furthermore, plaintiffs with CRPS who prevail in court have increased monetary rewards. An understanding of the clinical subtleties and manifestations of CRPS are crucial in improving patient outcomes and decreasing liability. In patients with persistent pain and refractory symptoms—especially stiffness—CRPS should be in the differential diagnosis, and that deliberation, positive or negative, should be recorded.

### Incidence/prevalence

The incidence and prevalence of CRPS are unknown; however, its incidence is higher in smokers versus nonsmokers[1] and it occurs 3 to 4 times more often in women than in men.[2] In Olmsted County, Minnesota, the incidence was reported as 5.5 per 100,000 and the prevalence as 20.7 per 100,000 in 2003.[3] The incidence after fracture of the distal radius varies from 4% to 39% in prospective series and may occur after carpal tunnel release with or without iatrogenic damage.[4–6]

### Anatomy and physiology of pain

Pain with cellular damage initiated in the periphery may be from mechanical, thermal, chemical, and ischemic events. Pain signals, potentiated by local reflexes and humeral factors, are relayed via peripheral nerves to the dorsal horn of the spinal cord (wide dynamic range neurons), where they may be amplified and modified and then transmitted to cortical centers. The magnitude of pain depends on the mechanisms of initiating events, afferent information transmitted, efferent modulation, and central nervous system interpretation (Fig. 1). Painful (nociceptive) information is activated peripherally by mechanical, thermal, chemical, and ischemic events and transmitted by small myelinated (A-Δ) and small unmyelinated C-afferent peripheral nerve fibers to the spinal cord. The perception and physiologic consequences are related to a complex interplay of physiologic events and psychological factors. Complex regional pain syndrome is conceptually an exaggeration or abnormal prolongation of the expected pathophysiologic events after injury or surgery.

Complex regional pain syndrome may result in irreversible end-organ dysfunction, including loss of the normal arteriovenous shunt mechanisms and permanent alterations in central neurologic responses. Ongoing segmental ischemic and cell death may have a substantial role in the process. Swelling and stiffness, and atrophy and contracture may occur and persist. Pain is often associated with arteriovenous shunting and a relative ischemia of the extremity, mechanical events, or nerve irritation.

### Natural history

Complex regional pain syndrome types 1 and 2 occur, in part, as a departure from the orderly and predictable