Excessive limb guarding may increase complex regional pain syndrome risk

The skin of his left hand was reddish and shiny, with the tight appearance of drawn leather. His fingernails were long and seemed brittle with notable longitudinal ridges. "How would I describe my pain?" he asked. "My own words would never be adequate. Dante described my pain in The Inferno, when he wrote '... ever burning, but never consumed.'"

This patient suffers from complex regional pain syndrome, or CRPS. Unfortunately, most individuals who carry this diagnosis would find Dante's description an agonizingly accurate way to characterize the unrelenting searing pain they are constantly subjected to in their affected limbs. CRPS is a painful condition typically affecting the extremities, where long-standing pain persists at a magnitude of severity and duration out of proportion to the apparent precipitating level of tissue damage.\(^1\) Although the specific limb symptoms presented by the patient described above are not always present, or required for a CRPS diagnosis, individually varying patterns of associated symptoms such as edema, alterations in blood flow, local trophic changes, and motor impairments do accompany the disorder.\(^2\)

The actual prevalence of CRPS is unknown. It has been established, however, that it can appear in people of any age (the average age of onset between 36 and 50 years), about 70 - 75% of cases occur in females, the number of upper versus lower extremity cases is roughly equal, and 11 - 16% of patients have more than one limb affected.\(^2\)

A new name for "RSD"

Many names have historically been attached to the condition we now call CRPS. These have included post-traumatic neuralgia, traumatic dystrophy, algodystrophy, sympathalgia, shoulder-hand syndrome, Sudek's atrophy, and causalgia. Until recently, the disorder was most commonly referred to as reflex sympathetic dystrophy (RSD), because John Bonica reported in the 1950s that a number of patients experienced pain relief following administration of anesthetic blocks to sympathetic ganglia supplying the limbs.\(^3\) This clinical finding implicated a dysfunctioning sympathetic nervous system as the pain generating culprit, hence RSD seemed to be an appropriate name for the problem.\(^4\)

Current research suggests that this syndrome may have multiple causes.\(^2,5\) It is now felt that patients whose pain is apparently maintained by the sympathetic nervous system actually represent the minority causes.\(^2\) Most CRPS patients manifest what can
be termed sympathetically independent pain. Therefore, in 1994 the International Association for the Study of Pain officially renamed the condition complex regional pain syndrome. The new label is more than merely a name change, it now encourages researchers and clinicians to look beyond the sympathetic nervous system for explanations and treatment options.

**Diagnostic foundation**

Since CRPS may have multiple pathophysiological etiologies, diagnosis is made on the basis of clinical presentation. Results from sympathetic nerve blocks and three-phase bone scans are no longer required for the diagnosis and, in the case of the latter, are believed to have limited clinical utility. Essential to the diagnosis is the presence of some initiating traumatic event or cause of immobilization; continuing pain disproportionate to the inciting event; evidence at some time of either edema, skin blood flow changes, or abnormal perspiration at the painful site; and the absence of other existing conditions which might otherwise account for the abnormal degree of pain and dysfunction. CRPS is further categorized as either Type I or Type II. The Type II diagnosis is made if there is evidence of known injury to a peripheral nerve and Type I identifies cases where there is an absence of a known nerve injury. A hallmark of both types is allodynia, where the skin becomes so exquisitely sensitive to touch or temperature, that normal light contact on the affected limb produces severe pain.

Authorities are in agreement that CRPS is not an independent disease process. Rather, it is best viewed as a reaction pattern to injury or the excessive restriction of activity. It may be a complication of injury or iatrogenically induced. The terms CRPS Type I and CRPS Type II are meant as descriptors of certain chronic pain syndromes. They do not embody any assumptions about pathophysiology. Both central and peripheral nervous system mechanisms have been suggested as contributing to the development of the syndrome. Although too numerous to completely discuss here, some peripheral theories offered include abnormally increased resting discharge of sympathetic activity, peripheral nociceptor sprouting secondary to local demyelination, nociceptive “C” fibers increasing substance P release at the site of original injury, and adrenergic supersensitivity of vasomotor alpha receptors as a result of damaged sympathetic control. Hypothesized central nervous system possibilities range from abnormal modulation of second-order wide dynamic range tract cells to neuroplastic alterations in the somatosensory cortex. However, as Galer states in the current edition of the encyclopedic text, *Bonica's Management of Pain*, "the pathophysiology of CRPS is not known."

**Risk factors**

For health care practitioners and patients alike, the burning question is - why? Why is it that hundreds, or more likely thousands, of uncomplicated surgical procedures and relatively simple orthopedic injuries can heal as expected without residual pain, while one case develops into CRPS? Can we identify any salient, and hopefully preventable, risk factors which steer healing off course and into the realm of unrelenting
chronic pain? Severity of trauma is not the issue. Even very minor inciting incidents can ultimately result in a very severe long-term CRPS response. One patient was walking barefoot through her living room and tripped on her toddler's tennis shoe, resulting in mild subluxation of the first toe. By the time she sought help from a neurologist with expertise in CRPS, her entire lower extremity was affected to the point where she could not tolerate the touch of clothing on her leg and could ambulate only with the aid of bilateral axillary crutches.

Although psychological issues show frequent comorbidity with CRPS and chronic pain in general (approximately 30% of CRPS patients meet criteria for major depression, attempts to identify a premorbid psychological pattern have not yielded any solid predictive factor or personality profile which may predispose an individual to develop the syndrome posttraumatically.

Deactivation pain

An emerging factor which may help explain CRPS risk for many patients is based on altered behavioral patterns of limb usage and biomechanical changes in response to pain. This theory refers to deactivation pain and is based on the idea that when one excessively limits the movement and functional usage of a limb, a series of pathophysiological changes take place which establish multiple new pain generation sites. This means that even after the original lesion has completely healed, pain may be perpetuated by other tissue changes which resulted from prolonged disuse of the limb.

Imagine yourself for a moment suffering from CRPS affecting your right forearm and hand. The ever-present burning pain flares in intensity with each movement. When you attempt to use the hand - type a letter, push a mower, cut fabric with scissors, or just drive your manual transmission car through traffic - you pay for it the next day as a delayed surge in pain intensity tests your limits of tolerance. You can't ignore pain . . . you obey it. As pain continues and builds, using the hand for anything becomes unthinkable. You are also a rational and fast learner. The medical advice of Dr. Groucho Marx rings true in your mind "Well, if it hurts when you do that, then . . ." You take that hand out of the game. Any daily living activity, job related task, or leisure pursuit which requires the participation of your right hand will now come to an end. Not only will functional loss limit use and mobility, but thanks to allodynia you must protect the limb from tactile contact. Imagine walking through a crowd with the constant searing pain in your hand instantly launched beyond tolerance by the slightest brush from another person. So, like many CRPS patients, you adopt a protective guarding posture - the "pledge of allegiance guard." Hand clutched protectively to your chest, you don't use it, it doesn't move.

Traditional learning models tell us the obvious, that patients rapidly develop kinesiophobia. Movement and usage increases pain, so patients stop moving and using their affected limbs, hoping to minimize the suffering. Additionally, if it hurts to move, they assume movement must be producing further damage. Pain is a signal that tissue damage is being done . . . right? In chronic pain conditions, this is not necessarily true.
Another relatively recent pain/behavior theory suggests that in an effort to cope with severe chronic pain, the patient may cognitively dissociate the painful body part. Mentally walling it off may facilitate some level of detachment from the pain, but it also contributes to a "neglect-like" condition whereby the patient subconsciously ceases to treat the limb as his/her own, further reducing the involvement of the limb in movement or any functional activity.

Understandably, pain behaviorally restricts movement and functional limb use. However, it may be the case that prolonged and excessive restriction of limb use may maladaptively prolong pain. The sequella of pathophysiological changes developing from excessive immobility and disuse is not difficult to envision. The development of deactivation pain begins with postural guarding. The individual fixes the limb in an immobile, protective position. The rhythmic pumping action of muscles moving joints of the extremity ceases. Venous return from the limb is reduced. Edema remains unresolved. Increased venous pressure begins limiting arterial perfusion and oxygen delivery. Oxidative metabolism is reduced and cells of the region begin shifting into glycolytic activity. The region becomes acidotic, with reduced circulatory flow to flush out anaerobic metabolites. Regional damage to the tissues over time leads to fibrosis.

Over time, immobility and postural guarding result in additional changes which can exacerbate limb pain as secondary pain generation sources are created through disuse adaptation. Soft connective tissue adaptively shortens, resulting in contracture and adhesions. Lack of full range arthrokinematic motion leads to peripheral breakdown of articular cartilage. Attempts to move into normal joint ranges now pulls on contracted connective tissue and puts compressive force on desiccated joint surfaces. This is painful. The patient further restricts movement so as not to move into the new painful range. A downward immobility/pain spiral is created. As unused muscles weaken, they no longer contribute their share of effort to stabilizing joints and noncontractile ligamentous structures must now take on a greater burden of resisting joint destabilizing forces, increasing the probability of joint instability, ligamentous tears, and further pain.

By eliminating skin contact in an effort to avoid painful allodynia, the patient essentially eliminates the normal somatosensory mechanism of exogenous analgesia. In the absence of normal tactile input along large diameter fibers, an important component of our natural pain inhibition mechanism is lost. Without the option to rub a sore area, or scratch an itch, pain messages are free to ascend unchallenged through the dorsal horn of the spine and on to multiple destinations within the brain. Finally, diminished overall physical activity reduces general aerobic capacity, lowering anaerobic threshold, thereby making even mild activity an anaerobic exercise, perhaps again contributing to increased pain.

Restricted movement of proximal joints to brace the limb in protective guarding often leads to painful myofacial adhesions; particularly in the periscapular region during cases of upper extremity involvement. The condition is sometimes perceived by patients to "spread." The spread of CRPS to adjacent proximal joints and contralateral limbs is not due to the migration of a pathogen, but rather may be the result of transferring
biomechanical stresses to other areas away from affected sites and/or proximal muscular bracing.

A spectrum of specific evidence linking immobility to CRPS symptoms is mounting. A prospective study demonstrated CRPS symptoms developing in normal individuals after an extended period of casting following orthopedic procedures. The symptoms then resolved following a course of active physical therapy.\textsuperscript{17} Similar results have been found by immobilizing noninjured healthy volunteers.\textsuperscript{15} Animal investigations have demonstrated allodynia and neuroplastic changes in pain processing in rats following limb immobilization.\textsuperscript{18,19} In one study, just seven days of hind paw immobilization produced several weeks of both tactile and thermal allodynia in rats.\textsuperscript{19} Gellman, et al, reported brain-injured patients developing RSD in limbs affected by disuse secondary to neglect.\textsuperscript{20}

Health care practitioners and exercise physiologists are quite familiar with the deconditioning effects of inactivity and reduced mobility. In the case of CRPS, and perhaps other chronic pain conditions, inactivity also creates new pain sources. "Experts agree that CRPS patients usually become trapped in a vicious cycle in which guarding and activity restrictions perpetuate the pain of CRPS."\textsuperscript{6}

**Treatment approaches**

CRPS treatment is multifaceted. "The key to successful treatment of CRPS is a trained, coordinated, and experienced interdisciplinary team employing a functional restoration approach."\textsuperscript{13} The physician manages medications and coordinates the treatment team, a cognitive/behavioral psychologist frequently participates to help the patient cope with stress related to functional deficits and reevaluate kinesiophobia, and physical and occupational therapists guide the patient through carefully dosed and appropriately paced physical and functional reactivation. While a variety of treatment modalities are brought into play, "normalized movement is critical in the recovery of CRPS patients."\textsuperscript{13}

Although clearly a disorder with multifaceted etiology, a key component to the prevention and successful treatment of CRPS may lie in appropriately paced normalization of movement and functional limb use. The basic treatment philosophy follows Robinson's 1997 guideline; "Therapy for CRPS should be directed toward breaking the pain cycle by having patients participate in a progressive activation program for the affected limb."\textsuperscript{6} Treatment objectives include the following:

- Attenuate edema, pain, and allodynia.
- Arrest and/or reverse the biomechanical spread of CRPS.
- Restore bilateral symmetry of movement and weightbearing.
- Restore normal quality of movement to the affected limb.
- Return the affected limb to normal participation in functional movement and daily activity.\textsuperscript{21}
Successful treatment components, used in combination, include progressive physical reactivation of motion, strength, endurance, and functional activity via a quota-based exercise/activity dosing system;\(^{2,21,22}\) weightbearing and axial loading,\(^{21,23,24}\) along with tactile\(^{25}\) and thermal\(^{10,26,27}\) desensitization for the management of allodynia.\(^{2,21,28}\)

While the factors placing a given patient at risk for developing CRPS have yet to be fully established, patients who display excessive limb guarding and behavioral overprotection of an affected limb early in the recovery process are waving a serious warning flag which should be heeded and addressed by their physicians and therapists.

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**References**


