COMPLEX REGIONAL PAIN SYNDROME

PART 1: ESSENTIALS OF ASSESSMENT AND DIAGNOSIS

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DISEASE/DISORDER

DEFINITION

Complex Regional Pain Syndrome (CRPS) is a disorder characterized by continued debilitating regional pain that is inordinate to the provoking event, with or without a specific nerve injury and does not need to follow a dermatomal pattern. It usually develops after trauma; however there is no relation between severity of injury and development of this disorder and in some cases there is no precipitating event.

The pain is often associated with a combination of sensory and motor deficits, vasomotor disturbances, sudomotor disturbances, edema and/or trophic changes, all of which tend to present at distinct times throughout the course of the syndrome.

It was originally classified by the International Association for the Study of Pain (IASP) in 1994 and formally updated to the Budapest Criteria (2003), and now to the Valencia consensus (2019) due to its higher sensitivity and specificity; however CRPS remains a diagnosis of exclusion.

CRPS is further classified into: a) CRPS type I (previously Reflex Sympathetic Dystrophy) which typically develops after trauma or injury and does not involve a specific peripheral nerve; and b) CRPS type II (previously Causalgia) which develops after peripheral nerve injury. It can be further subdivided into “warm” (sympathetically maintained) or “cold” (sympathetically independent).

CRPS nomenclature has evolved through the years. It was previously referred to in the literature as Reflex Sympathetic Dystrophy, Algodystrophy, Causalgia, Sudeck’s atrophy among others.

ETIOLOGY

CRPS type I has a higher propensity to develop after an injury and can be due to several precipitating events ranging from sprains, fractures, visceral disease or even immobilization, while CRPS type II is known to occur after obvious peripheral nerve damage and can be related to a multitude of initial events. CRPS most often develops after acute tissue trauma including fractures, crushing injuries, sprains and surgeries. Patients may also develop CRPS after burns, stroke, frostbite, injections or local infections. However, it has been reported in cases without any identifiable precipitating event. There is currently no evidence that any particular injury regularly manifests as CRPS or that the severity of injury is related to the development of this syndrome.

The focused literature research revealed that immune reactions, alterations in receptor populations (e.g., upregulation of adrenoceptors and reduced cutaneous nerve fiber density) and central changes in autonomic drive seem to contribute to regional and systemic disturbances in sympathetic activity and to sympathetically maintained pain in CRPS. Evidence for dysfunction of both the innate and adaptive immune systems in CRPS is presented, through measured cytokines and other inflammatory mediators in the skin of affected limbs. Additional results from studies of mediator levels in animal models are evaluated in this context. Similarly, the production of autoantibodies and the potential targets of those antibodies from human, animal, and translational studies is reviewed.

Compelling evidence of autoinflammation in skin and muscle of the affected limb has been collected from CRPS patients and laboratory animals. Cytokines including IL-1β, IL-6, TNFα, and others are reliably identified during the acute phases of the syndrome.

More recently, autoimmune contributions have been suggested by the discovery of self-directed pain-promoting IgG and IgM antibodies in CRPS patients and model animals. Both the autoimmune and the autoinflammatory components of CRPS appear to be regulated by neuropeptide-containing peripheral nerve fibers and the sympathetic nervous system. Earlier age at CRPS onset and more severe symptoms have been found in patients with some human leukocyte antigen and tumor necrosis factor-alpha (TNF-α) polymorphisms.

A recent study identified four single nucleotide polymorphisms in genes expressed in peripheral nervous system neurons and immune cells that were associated with CRPS type 1 development, and more common in men. Several case reports and retrospective studies suggest a familial inheritance; however, no clear inheritance pattern has been established.

EPIDEMIOLOGY INCLUDING RISK FACTORS AND PRIMARY PREVENTION

Literature reports a frequency of 26.2 per 100,000 persons per year for CRPS type I and 5.5 per 100,000 persons per year for CRPS type II. Females have a higher propensity compared to males at a 4:1 ratio, and there is an increased incidence in the fourth decade and from 61 to 70 years old. No racial differences have been noted.

Fractures are the most common precipitating event, particularly in the upper extremity. The development of CRPS following surgery is a major cause of concern as this complicates post- operative management and has significant clinical ramifications. Operative procedures of the shoulder (incidence 0.9-11%), distal radius (22-39%), carpal tunnel (2-5%) Dupuytren’s contracture (4.5-40%), foot and ankle surgery (4.36%) have been associated with the manifestation of CRPS.

In a prospective study of patients with tibial fractures, the incidence of CRPS following surgical repair was documented at 31%; 33.3% of patients treated with intramedullary nailing, 28.6% of patients treated with nails and screws and 28.6% of patients treated with external fixation.

Other risk factors include asthma, history of migraine, angiotensin-converting enzyme inhibitor use, menopause, osteoporosis, cigarette smoking. Evidence exists that vitamin C supplementation may reduce the risk of CRPS type I for wrist fractures, in foot and ankle trauma, and following total knee replacement or lumbar spine surgery at a 12 month follow up.

The oral vitamin C dosage varied from 200 to 1,500 mg in a meta-analysis; however generally considered optimal. Duration of supplementation is typically 45-50 days. The most common side effects of supplementation are fatigue and lethargy, along with increased risk for nephrolithiasis for higher doses. In addition, intravenous supplementation or inclusion of vitamin C in regional blocks may reduce the risk of CRPS. The exact mechanism of action for the effects of vitamin C on CRPS are unknown, but are believed to be due to its anti-oxidant capabilities and improvement of bone density, and physical performance. In the pediatric population, CRPS is more common among early adolescent females with CRPS type 1 affecting the lower extremity.

PATHO-ANATOMY/PHYSIOLOGY

The physiological process of CRPS is not completely delineated. Several theoretical mechanisms have been proposed, which include sympathetically maintained pain, sensory and motor dysfunction, peripheral and central sensitization, and protective disuse. A large majority believe that continued noxious stimuli related to damaged areas may lead to sensitization of the peripheral and central nervous system. Peripheral injured nociceptors lead to an overexpression of neurotransmitters and eventual hypersensitivity due to repetitive stimulation.

The overstimulation of the peripheral nervous system leads to increased nociceptive awareness in the central nervous system. A distorted perception of nociceptive stimuli causes a continuous torrent of downstream ramifications and an ongoing exaggerated response to the initial stimuli. The involvement of the sympathetic nervous system is not fully understood and has lost some of its significance over time.

There may be a relation to increased catecholamine concentrations and subsequent interactions among nociceptive neurons. Additional mechanisms revolve around the body's active inflammatory response in an attempt to heal injured areas. Inflammatory markers released into the bloodstream and injured tissue lead to vasodilation and eventual engorgement, presenting as edema, warmth, or signs of inflammation.

Whilst many observations have been made of physiological abnormalities, how these explain the condition and who does and doesn't develop CRPS remains unclear. A new overarching hypothesis that may explain the condition invokes four dynamically changing and interacting components of tissue trauma, pathological pain processing, autonomic dysfunction (both peripheral and central) and immune dysfunction, primarily involving excessive and pathological activation of dendritic cells following trauma or atrophy.

Other proposed mechanisms include classic and neurogenic inflammation. It has been found that patients with CRPS present with increased pro-inflammatory cytokines locally, in the bloodstream and in CSF. These substances may produce plasma extravasation and vasodilation, thereby producing localized edema, warmth and erythema characteristic of CRPS.

Neuroimaging studies suggest a reorganization of somatotopic maps in the cortex of patients with CRPS. The degree of somatotopic reorganization correlates significantly with pain intensity and degree of hyperalgesia. The degree to which individual mechanisms contribute to CRPS may differ between patients and even within one patient over time.

DISEASE PROGRESSION INCLUDING NATURAL HISTORY, DISEASE PHASES OR STAGES, DISEASE TRAJECTORY (CLINICAL FEATURES AND PRESENTATION OVER TIME)

Initial symptoms of CRPS typically develop within 6 weeks of the precipitating event. Early symptoms tend to include edema, sensory changes, distal vasomotor changes, and pain at the area of insult. Over time, symptoms become less localized.
CRPS was historically divided into three stages, with initial stages lasting up to 6 months.

Stage 1 (acute): edema, vasomotor dysregulation, severe tenderness, and allodynia.
Stage 2 (dystrophic): skin and muscle atrophy, brittle nails and atrophy, intense proximal pain, mottled skin, and brawny edema.
Stage 3 (atrophic): skin becomes pale, smooth, shiny, and cyanotic, contractures and flexion deformities, pain decreases, and vasomotor changes stop.

Initial symptoms of CRPS typically develop within 6 weeks of the precipitating event. Early symptoms tend to include edema, sensory changes, distal vasomotor changes, and pain at the area of insult. Over time, symptoms become less localized. CRPS was historically divided into three stages, with initial stages lasting up to 6 months. However, evidence to date suggests that the evolution of CRPS is unlikely to be sequential.

Another CRPS classification is to "warm" and "cold", with individuals sometimes exhibiting early warm symptoms but transitioning to cold later on.

Warm: warm, red, dry and edematous extremity, shorter duration (4.7 months)
Cold: cold, blue, sweaty and less edematous extremity, longer duration (20 months)

Pain in itself has a multitude of different features or presentations, from burning, stabbing, or a dragging character, to throbbing and constricting. A large proportion of pain is located in deeper structures (muscles, tendons, bones) as compared to the more superficial locations. Autonomic and trophic changes have a propensity for hyperemic or livid features and a larger majority of individuals show no alteration in the growth of hair and nails. Edema or areas of swelling present most of the time and changing in sweating patterns are inconsistent among individuals. Reduced strength is more prevalent than normal strength and almost half have some form of movement disorder. Sensory disorders tend to be the most consistent deficit and could be increased or decreased at the time of exam.

SPECIFIC SECONDARY OR ASSOCIATED CONDITIONS AND COMPLICATIONS

Secondary complications from CRPS include sequelae of disuse including contractures as well as altered sensation of the affected limb. Infection, edema, dystonia, and myoclonus can also occur. Dystrophic changes of nails, skin, and hair, osteopenia (Sudeck's atrophy), and small nerve fiber dropout may occur. Impaired sympathetic vasoconstriction may also occur and lead to presyncope or syncope.

ESSENTIALS OF ASSESSMENT

HISTORY

A detailed history of present illness is essential. The most common complaints are hyperesthesia and/or allodynia. Pain is often described as hot, burning, and ache-like, and can either follow a dermatomal or a no dermatomal distribution. It is relieved with medication and rest, and is exacerbated by temperature changes, limb movement, stress and physical activity. Signs and symptoms are highly variable between patients, and often change over time. History of recent surgery or trauma, including sprains, crush injuries and fractures should be thoroughly assessed as this may be a trigger for CRPS.

To make a clinical diagnosis of CRPS, the following criteria (the Budapest criteria) must be met. The patient must have continuing pain, which is disproportionate to any inciting event. The patient must have at least one symptom in 3 of the 4 following categories:

Sensory- Sensory reports of hyperesthesia, hyperalgesia and/or allodynia
Vasomotor - Report of temperature asymmetry and/or skin color changes
Sudomotor/edema - Report of edema and/or sweating changes and/or sweating asymmetry
Motor/Trophic - Report of decreased range of motion and/or motor dysfunction (weakness, tremor, or dystonia) and/or trophic changes (hair, nail, or skin)

The patient must display at least one sign during the physical exam in 2 of the 4 categories:

Sensory - Evidence of hyperalgesia (to pinprick) and/or allodynia (to light touch and/or deep somatic pressure and/or joint movement)
Vasomotor - Evidence of temperature asymmetry between limbs and/or skin color changes/asymmetry
Sudomotor/Edema - Evidence of edema and/or sweating changes/asymmetry
Motor/trophic - Evidence of decreased range of motion and/or motor dysfunction (manual muscle testing weakness, tremor, or dystonia) and/or trophic changes (hair, nail, skin)

There is no other diagnosis that better explains the symptoms and signs. A diagnostic subtype called CRPS-not otherwise specified (NOS) was created that would capture those patients who did not meet the new clinical criteria but whose signs and symptoms could not be better elucidated by any other diagnosis.

The Budapest criteria has been reported to have sensitivity of 0.99 and specificity of 0.79.21. Specific diagnostic criteria for the pediatric population are currently in development. Diagnostic accuracy of the Budapest criteria in children is somewhat limited as 37% of pediatric CRPS patients do not qualify as having CRPS.

PHYSICAL EXAMINATION

The patient must display quasi-objective findings on the exam in at least 2 of the 4 categories. Examination tools should include pin, temperature tape or infrared thermometer, von Frey testing or weighted pin, algometer, and goniometer.

Sensory signs are elicited through light touch and pinprick. Light touch can be performed with a cotton-tipped swab or soft brush. Motor changes can present as reduced range of motion and tremors. Trophic changes may manifest as growth or loss of hair and nail changes. Vasomotor signs can be detected by assessing skin color and measuring skin temperature in affected and contralateral limbs.

CRPS may spread to other extremities, and it may be important to measure skin temperature in arms and legs. The examiner must also assess for sudomotor changes (sweating changes/asymmetry) and edema. Trophic changes manifest as shiny, edematous, and erythematous skin findings with loss of hair. There are isolated findings in CRPS, which are more suggestive of other conditions on the differential. Limited and painful joint range of motion can be found with adhesive capsulitis. Skin warmth and swelling around the joints may be the first finding for inflammatory arthritis. Red, hot, and painful extremities can be found in erythromelalgia, although this condition is frequently symmetric, while CRPS is at least initially unilateral. Cold limbs might indicate vascular insufficiency. Neuropathic pain can be exquisite in postherpetic neuralgia and shingles. In each of these cases, other physical examination findings may assist in organizing the differential when considering CRPS.

FUNCTIONAL ASSESSMENT

An impaired limb can be disabling. Inquiry into how patients minimize discomfort while pursuing activities of daily living, including bathing and dressing, can guide functional restoration efforts. Because upper extremities are more frequently affected by CRPS, dominant hand function may be affected. Physical impairment may exceed those found with limb amputations and patients may demonstrate distress similar to that found with other chronic pain conditions. Additionally, neuropsychological changes such as asomatognosia, or “neglect-like” changes, in the absence of any brain lesion have been reported.

LABORATORY STUDIES

At this time, no laboratory studies are used for the assessment of CRPS. Testing to exclude other diagnoses on the differential, including vascular studies to evaluate compromised limb perfusion, electrodiagnostics for specific neuropathies, and rheumatologic laboratory studies to evaluate for arthritis, may be helpful. While multiple biomarkers have been studied with varied results, currently none are accepted for routine clinical use.

IMAGING

Various imaging modalities have been used for CRPS and could potentially be used to help diagnosis and monitoring; however at this time there are no radiologic diagnostic criteria established and the diagnosis is primarily clinical.

Triple phase bone scans are generally considered the most useful; however a meta- analysis demonstrated that it has no added value to CRPS diagnosis and should not be used for confirmation. However, a negative bone scan may help to exclude the disease or rule out other etiologies. Plain films may reveal patchy osteoporosis at periarticular regions in the affected limb. Magnetic resonance imaging (MRI) is occasionally performed to evaluate for other muscle, joint, or soft tissue etiologies of pain.

Some data regarding use of functional MRI, CT, PET-CT and diffusion tensor imaging exist; however their use tends to be more experimental.

SUPPLEMENTAL ASSESSMENT TOOLS

Historically, the diagnosis of CRPS required abrupt and transient relief from pain and dysesthesia with a systemic chemical sympatholytic (Bier Block) and/or a diagnostic sympathetic block with a stellate ganglion block or a lumbar paravertebral block. However, as the role of the sympathetic nervous system in the pathogenesis of CRPS remains unclear and contradictory, it is now widely accepted that a positive response to sympathetic block is not diagnostic of CRPS.

Furthermore, there is low evidence to support or recommend against use of local anesthetic sympathetic blockade is effective for reducing pain in CRPS. Autonomic tests such as resting sweat output (RSO), the resting skin temperature (RST), and the quantitative sudomotor axon reflex test (QSART) have been used to evaluate CRPS. Quantitative sweat tests and sudomotor axon reflex are sometimes used to assess sudomotor dysfunction but are rarely used and have limited availability.

Electrodiagnostic testing may be useful in the diagnosis of CRPS type II to demonstrate nerve injury, although tolerance to this examination may be limited.

EARLY PREDICTIONS OF OUTCOMES

Early diagnosis and intervention in CRPS is associated with improved outcome and function. Although pain may decrease over time, detrimental changes arise from neuroplasticity. Additionally, long-term deficits in muscles, bones, and nerves are seen if the syndrome advances or goes undiagnosed and untreated. In a study with a mean follow up of 5.8 years, 30% of patients considered themselves fully recovered, 54% were stable and 16% reported progressive disease. Cold CRPS and CRPS involving the upper extremity are associated with worse outcomes. Female sex, greater baseline pain, anxiety and disability were associated with worse CRPS severity long term. Among older adults with distal radius fractures, higher BMI, immobilization time, and lower physical activity were associated with lower functional outcomes at 6 week and 1 year follow-up. Recurrence of CRPS is not uncommon; estimates of recurrence range from about 10 to 30%, with the higher rates occurring in younger patients, including children.

ENVIRONMENTAL

Weather may exacerbate CRPS symptoms. Tobacco and alcohol use is reported in 56% and 78% of patients with CRPS, respectively.

SOCIAL ROLE AND SOCIAL SUPPORT SYSTEM

The most pragmatic assessment of pain must include biological, psychological, and sociological aspects. Secondary psychological responses and dysfunction are ubiquitous. A 2020 cross-sectional study showed patients with CRPS with more negative disease perceptions experienced greater pain, disability, and kinesiophobia.

Psychological stress may influence disease progression as patients with higher levels of anxiety, perception of disability, and pain-related fear have been shown to have a worsened disease course. Plasma levels of epinephrine and norepinephrine were elevated in a sample of CRPS patients compared with healthy control subjects. While theoretically, stress and emotional distress could elevate these catecholamines, a causal relationship between social and environmental influences and the development of CRPS is far from proven.

Further, catastrophizing, commonly seen in CRPS, has been shown to increase pro-inflammatory cytokines which may also be linked to disease progression. This psychosocial distress among CRPS patients may result in higher pain intensity compared to similar distress in a non-CRPS chronic pain patient. Addressing psychologic and social factors may improve outcomes; however, neither have been proven as the cause of CRPS. The role of psychologic and social factors must be viewed with caution. It is critical to identify and aggressively treat all spheres of the pain experience.

COMPLEX REGIONAL PAIN SYNDROME

PART TWO: MANAGEMENT AND TREATMENT

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Originally published: July 25, 2012 Last updated: October 31, 2024

REHABILITATION MANAGEMENT AND TREATMENTS

AVAILABLE OR CURRENT TREATMENT GUIDELINES

Awareness of complex regional pain syndrome (CRPS) by general practicing physicians is poor, which often leads to delays in treatment. Aggressive treatment should not be delayed as progressive worsening of symptoms is associated with poor prognosis. Rehabilitative therapies coupled with pharmacotherapy are the mainstays of early treatment. Interventional treatments are considered if conservative strategies fail. There are no well-accepted treatment guidelines for pharmacotherapy. Best evidence supports a multidisciplinary approach.

TRADITIONAL TREATMENTS

PHYSICAL THERAPY AND OCCUPATIONAL THERAPY

Physical therapy (PT) and occupational therapy (OT) can improve outcomes in CRPS, when started early (symptoms for less than 1 year). Objectives of PT and OT in CRPS are to improve range of motion, desensitization, minimize swelling, promote normal positioning, decrease muscle guarding, and increase functional use of the extremity.

MIRROR BOX THERAPY

Mirror box therapy may improve affected limb range of motion (ROM) by cortical reorganization of pain and motor neural networks. It is an adjuvant treatment of post-stroke upper limb CRPS. Besides optimizing pain control and function, it is also associated with shorter hospital stays.

GRADED MOTOR IMAGERY

Graded motor imagery (GMI) treatment is focused on training the brain to re-connect to the body part affected by pain. When you have CRPS in one part of your body, your brain recognizes the extremity affected as a painful threat. There are 4 steps to GMI, and these include laterality, imagery, sensory discrimination, and mirror therapy. GMI has been shown to improve altered central processing in CRPS, which may improve symptoms. Trials have demonstrated improvement in pain and functional disability at 6 months in patients with CRPS I.

TACTILE (OR SENSORY) DISCRIMNATION TRAINING

Tactile (or sensory) discrimination training has been shown to help pain and function in CRPS. By teaching the body part and the associated area of the brain how to differentiate between various sensations, it helps clear the picture in the brain.

TRANSCUTANEOUS NERVE STIMULATION (TENS)

Results in decreased pain and edema and provides minimal functional benefits in combination with PT.

COGNITIVE BEHAVIORAL THERAPIES

Regardless of the duration of the condition, all CRPS patients and their families should receive education about the negative effects of disuse, the pathophysiology of the syndrome, and possible interactions with psychological/behavioral factors. All patients with chronic CRPS should receive a thorough psychological evaluation, followed by cognitive-behavioral pain management treatment, including relaxation training with biofeedback, reframing, hypnosis, and behavioral modifications.

NON-TRADITIONAL TREATMENTS

COMPLEMENTARY THERAPIES

Isolated case studies show a potential role for acupressure and acupuncture. Herbal medicines, anti-inflammatory type diets, and natural supplementation have also found roles in treatment.

HYPERBARIC OXYGEN

Hyperbaric oxygen therapy has been shown to induce analgesic effects in nociceptive, inflammatory and neuropathic pain in animal models. In patients with post-traumatic CRPS of the wrist, it may lead to potential improvement in range of motion, pain control, and edema management. It is not clear these results are generalizable to other populations.

ELECTROCONVULSIVE THERAPY

There are case reports of secondary improvement in CRPS symptoms when electroconvulsive therapy is used for depression.

PHARMACOLOGIC THERAPIES

NONSTEROIDAL ANTI-INFLAMMATORY AGENTS

Clinical trials have shown mixed results, questioning their benefit in CRPS.

ANTIEPILEPTIC DRUGS

Gabapentin has moderate evidence in improving pain symptoms such as hyperesthesia and allodynia. Gabapentin and amitriptyline were compared in the pediatric population and showed both drugs were effective in reducing pain intensity and improving sleep, but no difference between them. Other options include pregabalin and topiramate. Also, carbamazepine was studied in a small trial with 600mg per day for 8 days and showed pain reduction.

BISPHOSPHONATES

Bisphosphonates may be beneficial through several different mechanisms. They can reduce osteoclastic activity and modify inflammatory cytokines, although the exact mechanism remains unclear. Studies show consistent statistically significant effects in pain relief, functional improvement and overall improvement. A review from 2022 found high-quality evidence supporting bisphosphonates (and ketamine) as a first-line treatment for upper extremity CRPS.

CALCITONIN

Analgesic properties in the central nervous system through release of β-endorphins and bone resorption inhibition. Conflicting evidence, though treatment is relatively simple, safe and better on early CRPS.

PHENOXYBENZAMINE

Oral phenoxybenzamine is an alpha-1 antagonist that has shown benefit in CRPS. The dose is slowly increased up to a maximum daily dose in the range of 40 to 120 mg, with treatment duration of 6 to 8 weeks. Orthostatic hypotension and ejaculatory problems can be expected at the higher dose range.

CORTICOSTEROIDS

Frequently used in clinical practice despite relatively weak evidence for effectiveness. 22Oral prednisolone has been found to be effective in alleviating CRPS symptoms, with doses ranging from 30 to 100 mg/day.34 The limited evidence available suggests that a short course of steroids may be indicated in early CRPS, as per the fifth edition of the Diagnostic and Treatment Guidelines for Complex Regional Pain Syndrome.

NIFEDIPINE

Limited data may indicate that the calcium channel blocker, nifedipine, may be helpful at daily doses of up to 60 mg.

OPIOIDS

There is a lack of evidence to support long term (>6 months) opioids in CRPS. While tramadol may be of benefit in neuropathic pain, there is, again, little scientific support in CRPS. Similarly, there is insufficient evidence to support or refute fentanyl use for any neuropathic pain condition.

KETAMINE

NMDA receptor and hyperpolarization activated cyclic nucleotide gated potassium channel 1 receptor antagonist that also has dopaminergic effects which may produce improvement in pain and can decrease opioid requirements. The largest study randomized subjects to a 4 day infusion of IV ketamine vs normal saline titrated according to pain relief and side effects. However, the initial improvements in pain lasted until week 11th of the study. Offers temporary relief from severe, debilitating pain and does not improve affected limb functionality. Ketamine has recently been suggested as a first-line treatment for upper extremity CRPS.

ANTIDEPRESSANTS

While commonly used in neuropathic and chronic pain conditions, there is little specific evidence of their benefits in CRPS.

DISEASE MODIFYING ANTIRHEUMATIC DRUGS

TNF-α signaling has been reported to contribute to the development of nociceptive sensitization in CRPS and tissue necrosis factor-alpha inhibitors has shown effectiveness in case studies. However, a randomized-controlled trial failed to demonstrate the benefit of this medication in CRPS.

INTRAVENOUS IMMUNOGLOBULIN (IVIG)

IVIG can interfere with proinflammatory markers and cytokines. Small studies of low dose IVIG have shown some benefit in chronic pain syndromes and CRPS. However, the LIPS trial, a randomized placebo-controlled study, concluded that IVIG therapy is not an effective analgesic regimen for long-standing CRPS.

THERAPEUTIC PLASMA EXCHANGE

Considering the evidence of immune system involvement in CRPS, plasma exchange showed to be effective in a subset of patients with long standing CRPS. Plasma exchange is hypothesized to reduce a number of factors that contribute to neuropathic pain such as inflammatory cytokines and fibrinogen, and it can also increase serum anti-inflammatory cytokines.

FREE RADICAL SCAVENGERS

There is moderate evidence for topical 50% dimethyl sulfoxide (DMSO) and oral N-acetylcysteine in early and chronic CRPS effectiveness respectively.

TOPICAL AGENTS:

- Capsaicin Long-term topical capsaicin application can reduce epidermal C fiber density with resultant decreased substance P production. There are clinical strengths ranging from 0.025% to 0.15%, but this has been poorly tolerated in CRPS because of inherent burning sensation when applied despite concentrations administered.
- Transdermal lidocaine Compared with capsaicin, transdermal lidocaine may be better tolerated. Few cases in the literature demonstrate long term improvement of pain.
- Isosorbide dinitrate Vasodilator, for which there has been only a small study in the CRPS1 population, but not CRPS2. 24 patients with "cold" CRPS of the hand were randomized for topical ointments applied 4 times daily for 10 weeks vs placebo. No difference was observed in skin temperature, pain, activity level or levels of NO and endothelin 1.20 Another study showed some improvement in mean skin temperature in "cold" CRPS1.
- Topical clonidine Clonidine is an alpha-2 adrenergic agonist, and topical administration may help local CRPS induced allodynia and hyperalgesia.
- Topical diclofenac Diclofenac is a non-steroidal anti-inflammatory drug, and it may serve as an effective treatment option for patients with neuropathic pain from CRPS.

INTERVENTIONAL THERAPIES

SYMPATHETIC BLOCK

Considering autonomic dysregulation and exaggerated response to catecholamines is thought to contribute to the pathophysiology of CRPS, sympathetic blocks have been used for both diagnostic and therapeutic purposes in CRPS. Sympathetic block is generally considered the first choice when interventional treatments are considered. Stellate ganglion blocks are indicated for upper-extremity CRPS, and lumbar sympathetic blocks are indicated for lower-extremity CRPS. Sympathetic blocks are generally more likely to help if skin discoloration and temperature changes are present. A series of injections is usually prescribed; however, there is no convincing evidence to conclude that a series of sympathetic blocks is indicated unless there is progressive improvement of symptoms with each injection. Relatively weak evidence exists supporting effectiveness, mostly used in chronic lower limb CRPS.

INTRAVENOUS REGIONAL BLOCKS

Administration of Intravenous (IV) medication after exsanguination of a limb followed by tourniquet placement. This method was formerly known as a Bier block. Guanethidine, reserpine, droperidol, and atropine have not been shown to be effective. However, regional blockades with bretylium or ketanserin can result in significant pain reduction. Relatively weak evidence for effectiveness and if used, confined to patients that respond to phentolamine tests. No significant differences on pain relief, and functional improvement.

DORSAL ROOT GANGLION STIMULATION

The dorsal root ganglion (DRG) is considered an important target for neuropathic pain management since it transmits input from peripheral to central nervous system. Has the potential of achieving pain relief in focal neuropathic pain syndromes, including those difficult to maintain or target with spinal cord stimulation (SCS). DRGS demonstrated greater improvement in quality of life and psychological disposition with less postural interference from stimulation or unwanted paresthesia compared to SCS in refractory lower limb CRPS. Also, it was preferred over dorsal column stimulation in another study evaluating patients with knee CRPS. DRGS is a promising method since it results in significantly improved analgesia, function and mood at one year compared to SCS with high evidence levels.

INTRATHECAL BACLOFEN THERAPY

A few studies have shown that intrathecal baclofen therapy may be of benefit in CRPS1 patients, particularly those with dystonia. Also, a combination of intrathecal baclofen with spinal cord stimulation may decrease pain and improve dystonia in patients with CRPS refractory to conservative treatment.

IMPLANTABLE INTRATHECAL CONTINUOUS INFUSION PUMP

Opioid Infusion: Studies specific to CRPS are lacking with implantable continuous infusion pumps with opioid therapy and use is generally not recommended in CRPS. It has been considered; however, only in specific patients with very poor pain control, hypersensitivity, and markedly decreased range of motion. In rare cases, it should be combined with aggressive physical therapy to improve mobilization Clonidine and Adenosine Infusion: Preclinical data suggest that intrathecal clonidine and adenosine reduce hypersensitivity. The prevalence of sensory gain and loss on testing in patients with neuropathic pain varies as a function of presumed etiology, with hyperalgesia being most common in CRPS. Both intrathecal clonidine and adenosine acutely inhibit experimentally induced and clinical hypersensitivity in patients with chronic regional pain syndrome.

SPINAL CORD STIMULATION

Spinal cord stimulation (SCS) directly stimulates the dorsal columns to modulate neuropathic pain. Good evidence to support spinal cord stimulation durability for long term pain, satisfaction and quality of life improvement. Limited evidence supporting functional improvement. Most recently, SCS has been found to improve sensory, vasomotor, and sudomotor symptoms, and reduce use of opioids in patients with chronic/refractory CRPS. There is moderate evidence for reduction of pain symptoms. Still, studies have not been able to demonstrate an improvement in functionality after treatment with SCS. Risks associated with invasiveness of procedure must be balanced against potential benefit.

SURGICAL INTERVENTIONS

AMPUTATION

Considered in "end-stage" CRPS and may be appropriate for pain relief and improving quality of life in therapy resistant disease, intractable, debilitating pain, totally dysfunctional limb, severe recurrent infections and chronic trophic ulcers. Several retrospective studies of CRPS1 (but not CRPS2) patients indicate that approximately half had pain improvement after amputation. In patients with intractable CRPS, when compared to nonamputees, those who undergo amputation exhibit better pain scores, less disability, improved quality of life, and less depression. However, there is the risk of CRPS recurrence stump or phantom limb pain.

SURGICAL SYMPATHECTOMY

If there is excellent but temporary improvement from sympathetic blockade on repeated occasions, then surgical sympathectomy may be of benefit. Its chance of success is best if performed within the first 3 months after the initial trauma. Relief of pain may decline with time

MOTOR CORTEX STIMULATION

Recent case reports illustrate the use of motor cortex stimulation in CRPS. A craniotomy is performed and placement of an extradural grid is optimized using somatic evoked responses to cover the areas of pain. The mechanism of action probably involves spinal cord structures including spinal sympathetic nucleus and ventral roots.

DEEP BRAIN STIMULATION

DBS is the most invasive form of neuromodulation. Specifically, it involves targeting a deep structure in the brain. DBS targets CNS structures; if such structures are injured/maladaptive, suboptimal inhibition may be evident. There are no recent studies addressing this treatment option specifically for CRPS, and given the very low quality of evidence, the recommendation is inconclusive, with studies favoring non- invasive strategies research.

PREVENTION OF CRPS

Vitamin C has been studied as a prophylactic treatment for CRPS that acts by inhibiting pro-inflammatory pathways mediated through antioxidant mechanisms. There have been mixed results in its effectiveness. A 2022 review found that vitamin C administration reduced the rate of CRPS-1 after distal radius, wrist, foot, and ankle surgeries, with an odds ratio of 0.33 compared to placebo.32 However, because it is a low risk intervention physicians may consider it as a prevention strategy.1 Daily supplementation with 500 mg of vitamin C per day for 50 days may be beneficial to decrease risk of developing CRPS-I after distal radial fracture, foot and ankle surgery or trauma.

TREATMENT CONCLUSIONS

Treatment for CRPS can be difficult and frustrating. Each patient will be different, and an individualized approach is essential. Aggressive early treatment should be emphasized through an interdisciplinary approach. Most treatments are not well-documented in the evidence-based literature. Early physical and occupational therapy is important. Sympathetic blockade can be considered as the first interventional technique. Medication management to include corticosteroids and bisphosphonates are best supported in the literature. Other adjuvant medication can be considered, as previously described. Different pharmaceutic interventions can then be attempted to try to improve symptoms

EMERGING/UNIQUE INTERVENTIONS

Prognosis is best with early diagnosis and treatment. Once delayed, CRPS can spread proximally in the affected limb and to other areas of the body. Significant loss of function, atrophy, and contractures can result. Non-organic factors may worsen CRPS. As such, psychological therapy can be an important component and may include cognitive behavioral therapy.

CUTTING EDGE/EMERGING AND UNIQUE CONCEPTS AND PRACTICE

In a 2022 randomized, double-blind, controlled trial, lumbar sympathetic ganglion block was performed using botulinum toxin type A. The proposed mechanism for botulinum toxin is the blocking of cholinergic transmission, thus blocking sympathetic outflow when administered near sympathetic ganglia. This study included 48 participants. The results showed improvement in limb temperature at 1 month, with this effect lasting for 3 months. There was also improvement in pain at 1 and 3 months compared to blockade with levobupivacaine. There were no adverse events observed.

The transient receptor potential ankyrin 1 (TRPA1) is a non-selective cation channel found in calcitonin gene-related peptide (CGRP)-positive nociceptors. TRPA1 has been suggested to have a role in CRPS, particularly in post-ischemia pain, by modulating bone resorption through nociceptor activation. The role of TRPA1 may thus be understood through the proposed concept of senso-immunology, which is the study of the interaction between the immune system and the sensory system. The proposed association in the case of CRPS is that nociceptors, by releasing neuropeptides, may regulate the activation of osteoclasts, osteoblasts, and hematopoietic stem cells. At the same time, immune cells, epithelial cells, and osteoclasts can stimulate nociceptors and generate pain.

Dorsal root ganglion stimulation (DRGS) has shown similar success rates in treatment of CRPS when compared to spinal cord stimulation (SCS). In fact, some studies have found better results with DRGS than with SCS. Nonetheless, further research would be required to understand the mechanism of DRGS.

An inflammatory immune response can be activated in CRPS. IVIG can affect proinflammatory markers and cytokines and is an encouraging treatment for CRPS. Randomized controlled studies in refractory CRPS have shown benefit.5 Even in a variety of chronic pain syndromes, open label studies using low-dose IVIG have been effective in pain reduction. However, these studies are small, and there are valid concerns about the cost and availability of such treatments.

In regard to the inflammatory response of CRPS, dexmedetomidine is a selective α2-adrenoceptor agonist, and it might alleviate allodynia through GRK2 upregulation in sympathetic postganglionic neurons.

Memantine is a drug with the ability to block NMDA receptors in the brain and a potent inhibitor of central and peripheral sensitization. Some studies suggest that it may be a promising option for the treatment of CRPS.

Microvascular dysfunction and ischemia in muscle play a role in the development of cutaneous tactile allodynia in chronic post-ischemia pain. Pentoxifylline, a vasodilator and hemorheologic agent may be beneficial if used early in treating CRPS related to chronic post-ischemia pain.

Transcranial magnetic stimulation (TMS) is thought to help describe important neurophysiological and pathophysiological aspects of brain involvement in CRPS. In addition, repetitive TMS can modulate cortical excitability and induce long-lasting neuroplastic changes. Non-invasive procedures based on rTMS are now emerging as an alternative treatment of drug resistant pain. However, well-designed studies are needed to corroborate initial findings. Currently there is a clinical trial running of TMS for CRPS that hypothesizes TMS will improve CRPS-related pain and other symptoms such as cognitive, emotional and physical, when compared to baseline.

GAPS IN THE EVIDENCE-BASED KNOWLEDGE

There is controversy over the accepted pathophysiology of the disorder. Initially, CRPS was thought to be predominantly mediated through the sympathetic nervous system; however, autonomic symptoms are often not seen in clinical presentation. Moreover, some CRPS patients do not get relief from sympathetic blockade and plasma catecholamine levels are generally lower in the affected limb.

Other mechanisms thought to be involved include cortical reorganization, exaggerated inflammatory response, and neurogenic inflammation primarily through neuropeptide mediators including bradykinin, calcitonin gene-related peptide, and substance P.

A major gap in the evidence is the paucity of double-blinded placebo-controlled clinical trials. For a variety of reasons, CRPS patients are commonly excluded from pharmaceutical studies. There are no absolute specific or generalized guidelines for management of CRPS, nonetheless the treatment must be tailored according to each type of patient based on chronicity and presenting symptomatology. There is not enough evidence to fully support any of the proposed treatment approaches. Some of the factors mentioned include the limitation to find an adequate sample for randomized controlled trials, leaving us with a lack of powerful enough studies to establish standardized management.

SOURCE

HTTPS://NOW.AAPMR.ORG/COMPLEX-REGIONAL-PAIN-SYNDROME-PART-1-ESSENTIALS-OF-ASSESSMENT-AND-DIAGNOSIS

ORIGINAL VERSION OF THE TOPIC SUNJAY MATHUR, MD.
COMPLEX REGIONAL PAIN SYNDROME PART 1: ESSENTIALS OF ASSESSMENT AND DIAGNOSIS. 8/9/2012.

PREVIOUS REVISION(S) OF THE TOPIC
LUIS BAERGA-VARELA, MD, RAFAEL ARIAS-BERRIOS, MD, SHIRLEY GRIGG, MD, NATALIA M. BETANCES RAMÍREZ, MD, JUAN GALLOZA OTERO, MD.
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LAURENTIU DINESCU, MD, JUSTIN FAYE, MD, ALEC GUERZON, MD, MICHAEL MOSIER, MD.
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HTTPS://NOW.AAPMR.ORG/COMPLEX-REGIONAL-PAIN-SYNDROME-PART-1-ESSENTIALS-OF-ASSESSMENT-AND-DIAGNOSIS

ORIGINAL VERSION OF THE TOPIC STEPHEN KISHNER, MD.
COMPLEX REGIONAL PAIN SYNDROME PART 2: MANAGEMENT AND TREATMENT. 7/25/2012.

PREVIOUS REVISION(S) OF THE TOPIC
STEPHEN KISHNER, MD, BRANDON L HICKS, MD.
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COMPLEX REGIONAL PAIN SYNDROME PART 2: MANAGEMENT AND TREATMENT. 5/7/2021

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MCGILL PAIN INDEX

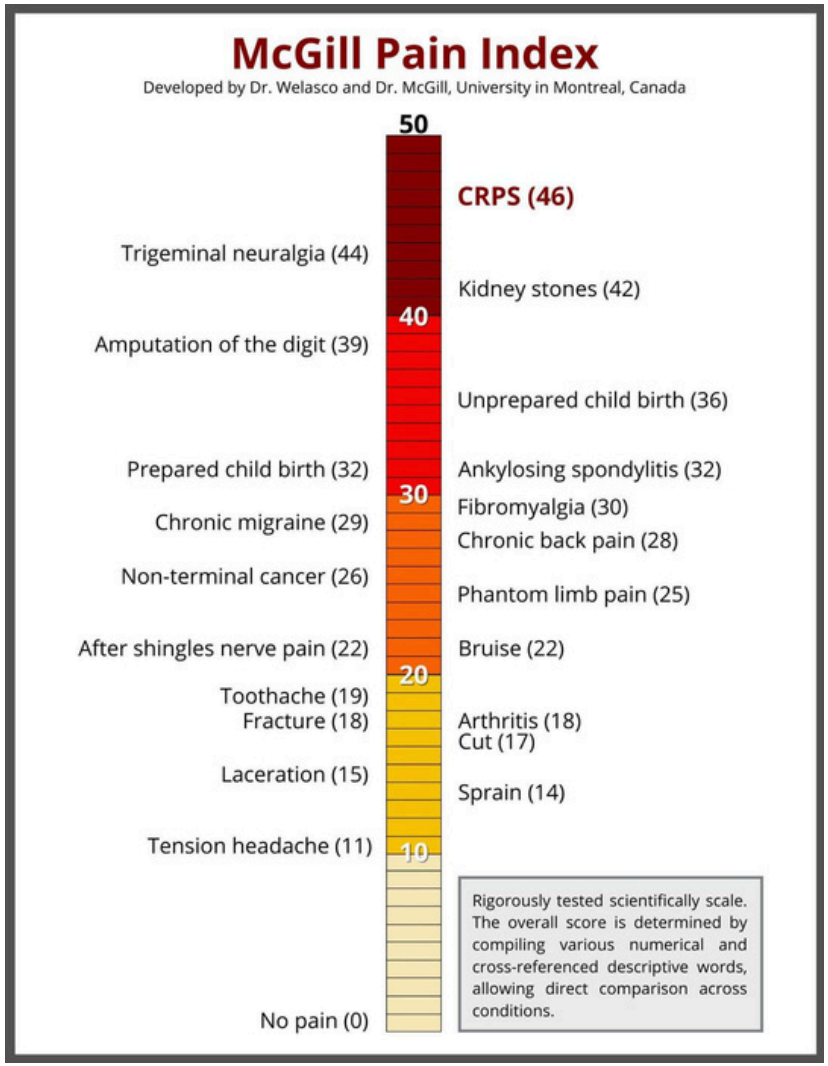
CRPS IS RANKED AT 46 OUT OF 50 ON THE MCGILL PAIN INDEX

Developed in 1971 by Ronald Melzack and Warren Togerson at McGill University in Montreal, Canada, the McGill Pain Index revolutionized pain research by challenging the idea that pain is strictly cause and effect. Melzack described how the brain perceives pain, which is influenced by past experience and other brain inputs.

The McGill Pain Index measures pain from multiple dimensions, including sensory, affective, cognitive, and behavioral. It was designed to provide quantitative measures of clinical pain that can be treated statistically.

It uses 3 major classes of word descriptors - sensory, affective and evaluative - that are used by patients to specify subjective pain experience. It also contains an intensity scale and other items to determine the properties of pain experience.

The 3 major measures are: (1) the pain rating index, based on two types of numerical values that can be assigned to each word descriptor, (2) the number of words chosen; and (3) the present pain intensity based on a 1-5 intensity scale.



McGill - Melzack Pain Questionnaire

Objective:
Self-reporting measure of pain used for patients with a number of diagnoses. It assesses both quality and intensity of subjective pain. The MPQ is a multi-dimesional tool for pain assessment and it has three components, which are the sensory intensity, the cognitive evaluation of pain and the emotional impact of pain.

Method of Use:
The MPQ is composed of 78 words, of which respondents choose those that best describe their experience of pain. Seven words are selected from the following categories: dimension 1 to 10 (pain descriptors), three words; dimensions 11 to 15 (affective components of pain), dimension 16 (evaluation of pain) one word, and dimension 17 to 20 (miscellaneous) one word. Scores are tabulated by summing values associated with each word; scores range from 0 (no pain) to 78 (severe pain). Qualitative differences in pain may be reflected in respondent's word choice

PAIN DESCRIPTORS

- ☐ Flickering
- ☐ Quivering
- ☐ Pulsing
- ☐ Throbbing
- ☐ Beating
- ☐ Pounding
- ☐ Jumping
- ☐ Flashing
- ☐ Shooting
- ☐ Pricking
- ☐ Boring
- ☐ Drilling
- ☐ Stabbing
- ☐ Lacerating
- ☐ Sharp
- ☐ Cutting
- ☐ Lacerating
- ☐ Pinching
- ☐ Pressing
- ☐ Gnawing
- ☐ Cramping
- ☐ Crushing
- ☐ Tugging
- ☐ Pulling
- ☐ Wrenching
- ☐ Hot
- ☐ Burning
- ☐ Scalding
- ☐ Searing
- ☐ Tingling
- ☐ Itchy
- ☐ Smarting
- ☐ Stinging
- ☐ Dull
- ☐ Sore
- ☐ Hurting
- ☐ Aching
- ☐ Heavy
- ☐ Tender
- ☐ Taut
- ☐ Rasping
- ☐ Splitting

AEFFECTIVE COMPONENTS:

- ☐ Tiring
- ☐ Exhausting
- ☐ Sickening
- ☐ Suffocating
- ☐ Fearful
- ☐ Frightful
- ☐ Terrifying
- ☐ Punishing
- ☐ Grueling
- ☐ Cruel
- ☐ Viscous
- ☐ Killing
- ☐ Wretched
- ☐ Blinding

EVALUATION OF PAIN:

- ☐ Annoying
- ☐ Troublesome
- ☐ Miserable
- ☐ Intense
- ☐ Unbearable

MISCELLANEOUS:

- ☐ Spreading
- ☐ Radiating
- ☐ Penetrating
- ☐ Piercing
- ☐ Tight
- ☐ Numb
- ☐ Drawing
- ☐ Squeezing
- ☐ Tearing
- ☐ Cool
- ☐ Cold
- ☐ Freezing
- ☐ Nagging
- ☐ Nauseating
- ☐ Agonizing
- ☐ Dreadful
- ☐ Torturing

ACCOMPANYING SYMPTOMS:

- ☐ Nausea
- ☐ Headache
- ☐ Dizziness
- ☐ Drowsiness
- ☐ Constipation
- ☐ Diarrhea

PPI:

- ☐ 0 No Pain
- ☐ 1 Mild
- ☐ 2 Discomforting
- ☐ 3 Distressing
- ☐ 4 Horrible
- ☐ 5 Excruciating

SLEEP:

- ☐ Good
- ☐ Fitful
- ☐ Can't Sleep

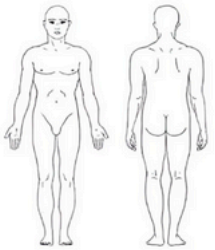
FOOD INTAKE:

- ☐ Good
- ☐ Some
- ☐ Little
- ☐ None

ACTIVITY:

- ☐ Good
- ☐ Some
- ☐ Little
- ☐ None

BRIEF ○	RHYTHMIC ○	CONTINUOUS ○
MOMENTARY ○	PERIODIC ○	STEADY ○
TRANSIENT ○	INTERMITTENT ○	CONSTANT ○



○ E = EXTERNAL
○ I = INTERNAL

CIRCLE THE BODY PART BEING TREATED TODAY

COMMENTS:

SOURCE:

REPRINTED WITH PERMISSION FROM MAPI RESEARCH TRUST
MELZACK R. THE MCGILL PAIN QUESTIONNAIRE: MAJOR PROPERTIES AND SCORING METHODS. PAIN 1975; 1:277-99

FOR ALL MEMBERS
OF MY CARE TEAM

HANDLE WITH CARE!

I HAVE BEEN DIAGNOSED WITH
COMPLEX REGIONAL PAIN SYNDROME
ALSO KNOWN AS CRPS OR RSD

PLEASE ALWAYS ASK BEFORE TOUCHING ME!

CRPS AFFECTS EVERY PATIENT DIFFERENTLY

- Persistent pain described as burning, throbbing, "pins and needles" sensation, or as if the affected limb was being squeezed.
- Allodynia: Sensitivity to touch or cold (light touch or normal physical contact)
- Hyperalgesia: severe/long-term pain after a mildly painful stimulus such as a pin prick.
- Changes in skin temperature, skin color, or swelling of the affected limb: affected limb may feel warmer or cooler than the opposite limb, skin on the affected limb may change color, becoming blotchy, blue, purple, gray, pale, or red.
- Changes in skin texture: shiny and thin or thick and scaly.
- Sweating, nail, and hair growth: On the affected limb, hair and nails may grow very quickly or not at all, and they may notice patches of profuse sweating or no sweating.
- Thinning of the bone or excess bone growth.
- Stiffness in affected joints.
- Impaired muscle strength and movement disorders.
- Atrophy to the affected limb.

IMPORTANT GUIDELINES FOR MY CARE

- Do not use ice or heat on me without asking first. This may make everything more painful.
- Pre-op shave to be done after anesthesia (this is very painful, like shaving a sunburn).
- Phlebotomy should be done by someone experienced and on unaffected limb only using pediatric needles (do not puncture the skin more times than absolutely necessary).
- Adjust rate and temperature of iv fluids if possible, some respond better to slower and warmer infusions.
- Whenever possible, place patients in a quiet part of the hospital and in the second bed to eliminate unnecessary traffic & being bumped when moving the other patient).
- Frequent linen changes are appreciated due to asymmetric sweating.
- Extra linens being available to regulate temperature is appreciated.

FROM CAREGIVER/PATIENT

- CRPS is nicknamed "The Suicide Disease". I am hopeless about my condition. Please don't be like the medical professionals that have mistreated me and/or didn't know what crps is.
- I am hypersensitive to many things that most people aren't. I have brain fog. I forget things (sometimes instantly) & lose time. Please keep this in mind!
- I am in more pain that you can fathom, so I may be snippy or short tempered. I apologize for this.
- With the exception of surgery, I am not seeking and do NOT want pain medication. I already take more medications than I'd like to, I am not looking for more.
- All secondary illnesses are amplified by CRPS and need to be accessed with this in mind and taken seriously. Please don't dismiss them as not bad enough.
- My symptoms are not exaggerated.

Budapest Criteria: Diagnostic Criteria
Distinct Factors of Complex Regional Pain Syndrome

Pain and at least one symptom in 3 of these categories:

Sensory: hyperalgesia and/or allodynia
Vasomotor: temperature asymmetry and/or skin color changes and/or skin color asymmetry
Sudomotor/Edema: edema and/or sweating changes and/or sweating asymmetry
Motor/Trophic: decreased range of motion and/or motor dysfunction (weakness, tremor, dystonia) and/or trophic changes (hair, nail, skin)

Must display at least one sign at the time of evaluation in two or more of the following categories:

Sensory: hyperalgesia (to pinprick) and/or allodynia (to light touch and/or deep somatic pressure and/or joint movement)
Vasomotor: temperature asymmetry and/or skin color changes and/or asymmetry
Sudomotor/Edema: edema and/or sweating changes and/or sweating asymmetry
Motor/Trophic: decreased range of motion and/or motor dysfunction (weakness, tremor, dystonia) and/or trophic changes (hair, nail, skin)

There is no other diagnosis that better explains the signs and symptoms

Symptoms	Comments
Continuing regional pain	Disproportionate to any inciting event
Sensory	Hyperesthesia Allodynia
Vasomotor	Temperature asymmetry Skin color changes Skin color asymmetry
Sudomotor/Edema	Swelling due to fluid in the body's tissues Sweating changes Sweating asymmetry
Motor/Trophic	Decreased range of motion Motor dysfunction Changes in hair, skin, and nails

SOURCE:

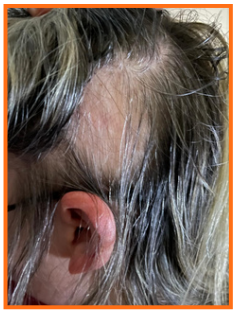
PAIN MED. 2007; 8(4): 326-331.
R. NORMAN HARDEN, STEPHAN BRUEHL, MICHAEL STANTON-HICKS, PETER R. WILSON

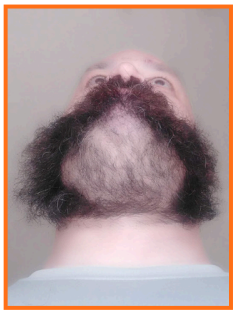
WHAT SYMPTOMS LOOK LIKE

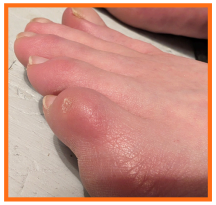
PHOTOS DONATED BY PATIENTS WITH CONFIRMED CRPS DIAGNOSIS

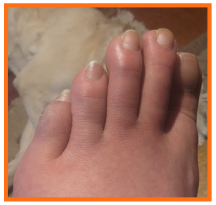
MOTOR/TROPHIC

HAIR, SKIN, NAIL CHANGES --- SKIN: TENDER, THIN, SHINY --- HAIR/NAILS: STRENGTH, GROWTH/LOSS



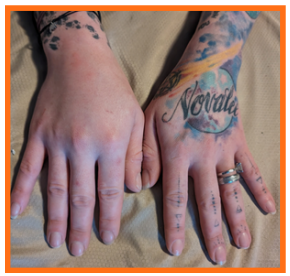


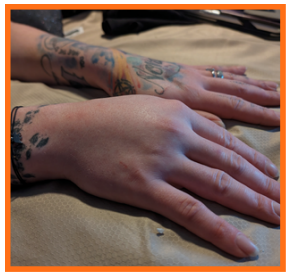


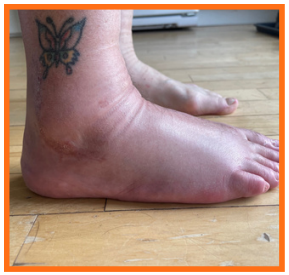


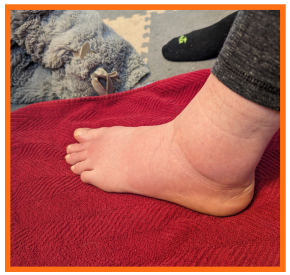
SUDOMOTOR/EDEMA

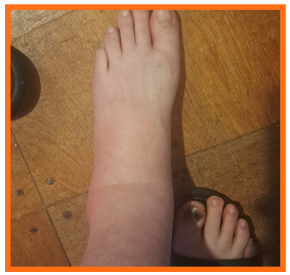
SWELLING/EDEMA --- SWEATING ASYMMETRY

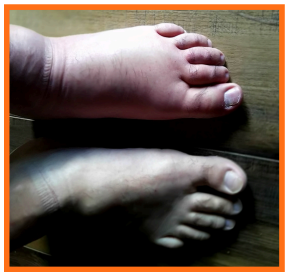


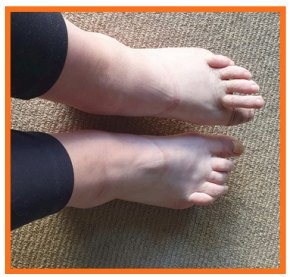


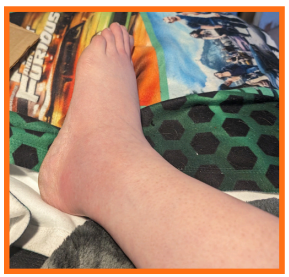






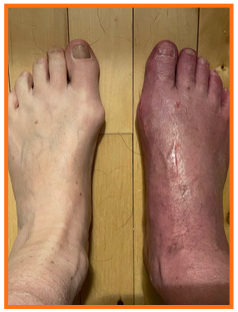


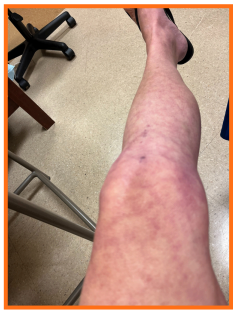


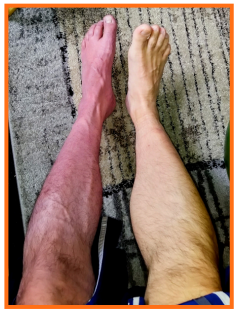


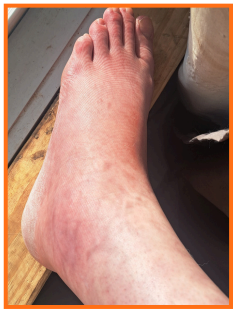
VASOMOTOR

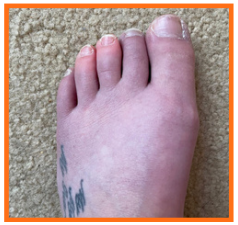
TEMPERATURE ASYMMETRY --- SKIN COLOR CHANGES

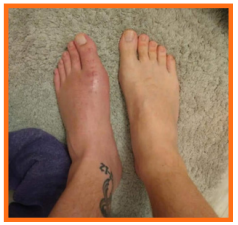


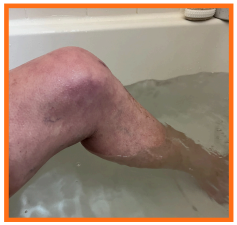


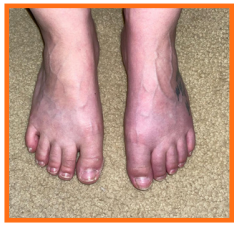












Real Words from Real Patients

Complex Regional Pain Syndrome
The Suicide Disease

Complex Regional Pain Syndrome (CRPS) patients go through various stages of grief and acceptance as they cope with their condition. It is important to understand how fragile these individuals can be due to the physical and emotional toll that CRPS takes on their bodies. Compassion and patience are crucial when interacting with CRPS patients as they may be experiencing a range of emotions and physical challenges. It is essential to provide support and understanding to help them navigate the complexities of their condition and find ways to improve their quality of life.

Hope

Peter Conti

"The main nerve in my left leg was crushed during a reduction procedure while I was in the hospital for surgery to put my broken hip back together with 14 pins and 4 plates. Aa you might imagine, the pain was overwhelming. This was the beginning of over sensitizing my central nervous system to the point where two years later I'd been to 23 different doctors and tried everything under the sun to try and get my life back. My brain had been sent into a loop where my foot and leg hurt like crazy even though there was nothing wrong with it.

I knew that I didn't want to live my life like that, so I made a decision to try to hike the Appalachian Trail. I figured that if I hiked over 2,000 miles then my leg would HAVE to get better. The result was quite an adventure, as I've shared in my book, "Only When I Step On It - One Man's Inspiring Journey to Hike the Appalachian Trail Alone." Unfortunately, while my hike helped immensely, afterwards I was still dealing with pain, fortunately at lower levels than before.

Another two full years later a doctor I was seeing about implanting a spinal cord stimulator said "I think you have CRPS - Complex Regional Pain Syndrome." And here I am, still on my healing journey today, one step at a time."

Holly Watson

I have a whole new respect for this monster. I developed CRPS after a meniscus repair gone bad and 3 surgeries later believe it has spread. I'm on a mission to get better and feel like I'm truly fighting for my life and limbs.

Learning to mourn the old life I was able to live. The online support groups have helped me the most and I cling to others for support and knowledge of the steps they took to get into remission.

On days I wanna give up I rest. Other days I wanna give up and get up and put up more of a fight and then rest. Exercise and movement is my friend. The PT where I run a cotton ball and work up to a loofa is very slowly getting a bit better.

I make gratitude lists and hold tight for better days. It's not easy but I know it's in me and I'm gonna fight! 🧡🔥🧡

Brian Hatt

I've been living with Complex Regional Pain Syndrome since 2011, the same year my life changed forever the day before my 33rd birthday. When I left for work that day, I had no idea the man that I had been all my life, the man I had worked my hands to the bone to become, would never walk through that door again.

It took me 3 years and countless medical professionals that either dismissed me or treated me like their guinea pig to get diagnosed. During this time, I watched everything fade away. I watched pieces of myself disappear, only to be replaced with my illness. I watched my friends fall off one by one. I watched my ex-wife fall out of love with me as I became nothing but a burden she did not want understand. I watched my children become farther and farther away from me when I could no longer do the things I used to for and with them. My ex-wife sided with doctors against me, my sons still do not know what CRPS is, my mother would act sympathetic while the nature of my illness went in one ear and out the other, and friends stopped calling. I suddenly I realized how alone I was and how wrong I had been about the people I called friends, and that I called family. People that claimed loyalty, disappeared.

The darkest corners of my mind were my home, they were the only place I felt I belonged. I became consumed by the dark abyss I had created to protect me and I had given up. I again reached the point of no return and this world would not have to suffer my existence much longer. At this darkest hour, of my life someone reached out. Someone cared. That's the only reason I'm still breathing. Most days I still want to quit. Things don't get better. I don't feel better. I'm still invisible. It's indescribable to walk through life while losing time and not knowing who you are.

Kimberly Cegelka

"This is grief. This is accepting the person I was before, a person I loved and worked hard to become, I can no longer be. However, as long as their is breathe in my lungs there will be hope in my heart.

It took 7 months for me to find a Dr who diagnosed me with CRPS. It is a hard pill to swallow, acceptance of an "incurable" and painful disease is a daily struggle. I am blessed to have a supportive family and to have found a Dr who is determined to help reduce my pain and truly listens, validates, and is empathetic. Please know what it means to CRPS patients when you truly hear them and help fight with them.

There may not be a cure yet, but there is still hope that the pain can lessen to a manageable level. CRPS patients need hope and support. It is hard for people to understand the pain levels we live with and I pray someday the awareness of CRPS means patients are diagnosed and given treatments early and don't also have to fight to be believed. "

Brittany Summerville

I started as a caregiver for my boyfriend who suffers from CRPS that has spread throughout his body. A year and 3 months in, I went from advocate, supporter, and caregiver to diagnosed CRPS patient.

Living with neurodiversity and severe mental illness has been a lifelong journey for me. A day without pain is not something I have ever known. Having multiple illnesses that involve having extremely painful hypersensitivities, both mentally and physically, to things people can't understand or even fathom costs you relatability. When people can no longer relate to you, they no longer what to be a part of your life. The barrier has always been there for me. Isolation forces you to sit in a dark place where feelings of not being enough and never doing anything right become the narrative you believe and tell yourself. A place so dark no one can see you and you feel unworthy of love.

The day I got diagnosed with CRPS was the day I realized I would always exhausted, I would always be on fire, and I would always reach a point where I would want it all to just end. My level of self-control has to be at max power 24/7. I am constantly navigating my pre-existing 3rd degree emotional burns in the form of ultra rapid cycling, disassociation, and fear of the unknown. My physical pain is masked by my intolerable emotional pain. The isolating nature of the beast are the extreme symptoms. You come off as "too much" or "not enough" - there's no gray of understanding. When others can't comprehend the level of pain you feel, it's hard to have compassion. Choosing quality over quantity can make us look selfish and full of excuses - but the simple reality is - having limited tolerance, functionality, and high pain means I have to wisely choose where I put what little energy I do have.

While this has been very defeating at times, I try to stay positive. My goal is to let others know that they aren't alone in what they are facing. Feeling alone while in a scary situation can be a traumatic experience, I aim to give more people a place to turn when they feel that have no where to go.

Charlie Mandril

Throughout my 40-plus years, I've faced overwhelming odds, traumatic situations, and life-altering events that required me to rebuild myself time and time again. So, in December 2020 when I woke up with a disease I had never heard of—given to me by a poorly located nerve block injection, during what was supposed to be a minor surgery—I arrogantly assumed it was just another challenge I would overcome. I was so very wrong! This is the only battle of my life that I will not be able to call myself the victor. It has stolen everything from me, and that's not just an expression. I've lost two careers, the ability to run, and eventually, even the ability to walk more than 200 feet in a single attempt. I no longer sleep soundly, I cannot climb stairs everyday, take a shower while standing, swim, lift weights, have a normal relationship with my wife or child, I cannot even pick up my 6-year-old daughter when she needs consoling. But not everything was taken from me, right? I've gained things too like: unrelenting and unimaginable pain; 100 pounds in the first year of dealing with CRPS; insomnia; severe depression; PTSD with vivid nightmares; the spread of CRPS to other limbs; emotional instability; arguments and lies from doctors; medication cocktails that make my hair fall out; anger; frustration; failed attempts at pain management; treatments that caused the CRPS to spread; and other medical failures that have darkened my outlook on life. But, who really listens to the fucking guy in the wheelchair anyways, he's just mad about his life and the fact he has to breathe everybody else's exhaust.

Lori Wilson

I never understood the importance of Awareness until I was diagnosed with something that very few are "aware" of. Now I understand all too well. The physical symptoms of CRPS suck, and I think anyone who witnesses them understands that. It's the mental toll that it takes that most people could never comprehend. And there is really no way to put it into words.

It starts with having to mentally digest the whole "No Cure" thing and all the other horrible fun facts that are found on Google. It then moves into having to re-convince almost EVERY doctor of your diagnosis, being treated like a lying, faking drug seeker (even without requesting drugs) by doctors, lawyers, and even people who you thought were friends.

If "you don't look sick", most people automatically believe that you're fine, and the same person that you were before. But you're not. No matter how hard you try, you can't be the same person. Your life will never be the same, and very few people will ever truly understand that.

And then you realize that if no one knows that this awful Twilight-Zone-meets-Stephen-King condition exists, then no one is trying to find the cure. Which makes you feel hopeless. And more alone than I ever knew was possible, regardless of having an amazing support system. A cure is the goal, and I know understand that Awareness makes up a large part of the path to that goal.

April Davenport

I was diagnosed with CRPS at the age of 11 after I injured my arm, resulting in doctors coming to the conclusion that it was broken. After two weeks in a cast, the doctors realized that my arm had not been broken after all, and I went home feeling relieved. That quickly changed as I realized my arm was in the most pain I had ever experienced. I lost the use of my left arm for a year and half, but after I finally regained its full range of motion, the CRPS in my arm went into remission.

Unfortunately, around the time my CRPS in my arm subsided, I injured my leg, causing CRPS to develop in my knee. This time, instead of going into remission on its own, the symptoms persisted and even spread to my other knee. To this day I have CRPS in both my legs which has come with its own additional medical diagnosis.

My journey with CPRS has not been linear, but I do feel I am on the final stage of grieving the life I once thought I would have as an adult. First, I experienced denial, that caused me to push myself to the breaking point over and over again to participate in activities that I was no longer able to take part in in a healthy way. I then became angry, with doctors, the medical system who I felt like failed me, and with myself. I fought to bargain with myself and those around me to let me hold on to any bit of normalcy that I would have gotten to experience living a life without pain. When I found myself deep into my teen years grappling with loss of sleep and normal social interaction, I battled deep sadness. But finally, I have found myself in a state acceptance that this is my new normal. I still find myself in dark places when my pain flares, and loss the use of my legs temporarily. But I find strength in the fact that I have made it this far, and that I am not doing this alone.

Cody Johnson

I wish I could convey the magnitude of the mental toll this takes on a person. I wish doctors had prepared me for it when I got sick. Instead, they couldn't even look me in the face to give me the diagnosis.

This disease is unfathomable, inhumane, and downright unsustainable. Your brain begs you for it to be over more than your own thoughts can exist. It's robbed me of my body, my family, my mind, and my physical and mental functionalities. You become a shattered, fragmented shell of what you once were.

After my bunionectomy surgery that got me sick, I went home and withered away in front of my kids for three and half months while I waited for my pain management appointment, where I was finally told. Waiting all that time with no diagnosis and no surgeons seeming to be legally able to tell me what was wrong with me, even though I knew they knew what they'd done to me. I lost 45 pounds in that time.

We have to find a way to test, such as an MR Nuerography, and treat immediately. This disease is the ugliest monster I could ever imagine. We need help to fight it. I feel utterly betrayed and abandoned by the medical field. What I've learned from my community of warriors and my own journey is that we're on our own. That can't be. That just can't be. I promise you I'm combating as hard as humanly possible, but I can't keep up with the unspeakable force threatening my life every second of every minute. It's been 1,219 days, and I haven't had a single break.

Please find a way. Help us.

Chele Sykora

I am a 40 y/o CRPS Warrior, wife, and mama of two amazingly wild 8 y/o twin boys. I am sharing the following, first, to give a brief description on my medical training and knowledge in the medical field and healthcare system prior to my CRPS diagnosis.

I started taking healthcare courses from 1998 to 2001. I became a CNA at 17. Later, I attained my Asoociates Degree in Science of Nursing - RN in 2009. I worked in all settings; specializing in ER, Pediatrics, & Pediatric ER medicine. Fast forward from 2020 to present day. A dimensional shift is pretty much the only way I can describe the Healthcare system today. A very sad, dysfunctional, broken, and indoctrinated medical system that has let down, mistreated, ignored, and gaslit patients. This is due to medicine and (Big Pharma) not treating patients the way they should be treated, and our tried and trusted insurance system that is in bed with the docs, and Big P. We are no longer taken seriously. Doctors no longer try to get to the root of the problem. They no longer care, as they just prescribe medications, get perks from big pharma, and then keep patients sick from the medications they prescribe. Which causes the need for more medications to treat all of the side effects these toxic drugs cause.

I was diagnosed with CRPS 8 months after an "outpatient, routine, exploratory abdominal surgery" in Jan 2020, for possible endometriosis. My life, my world blew up as I knew it. If the young, surgery hungry doc that performed my surgery would have looked at my MRI scans, he would have seen that I had adenomyosis and could have concluded that my symptoms were due to that vs causing CRPS for an exploratory surgery for endometriosis.

There is more back story to having CRPS, years prior from other surgeries, but it went undiagnosed. I can leave out those details because, the point is, I was diagnosed with CRPS. The way I was diagnosed, was a heartless, uncompassionate doctors visit. I was trying to figure out all the 'whys'. Why I was in excruciating pain, why I could not walk and function as I once did, why I could not care for my kids and family, why I was exhausted beyond recognition, why I could not eat, sleep, think. Why I could no longer be the active, vivacious women I once was. A gym rat, a mom that could pick up and hold her babies. Why I was having to explain to my almost 2 year old boys (when CRPS actually started but kept getting dismissed by the doctors). Why mommy couldn't pick them up anymore, or pick them up to nurse them and daddy had to help. Why was this happening? Why does each doctor I see that is clueless - just keep referring me from one doctor to the next? (Big P and getting insurance to pay for all of these "I dunno, go here" visits. My doctor, a neurologist that I worked with, handed me a business card with CRPS written on the back of it, and he told me to Google it, maybe follow up with pain medicine, and gave me a horrible prescription for nerve pain medications along with steroids that causes more crippling & debilitating side effects than are worth taking.

The ironic thing is, we, healthcare personnel, are all too familiar with patients that Google symptoms. We tell them to never Google their symptoms. Yet, this doctor could not take a moment to sit with me and explain this horrific, life altering disease to me. He handed me a business card and walked out. I was bawling in pain, fear, confusion and devastation. I got to a very dark place. As most warriors are in, or get to. I will never forget that first year. Being bed/couch bound. Screaming in pain into the couch cushions/pillows when I was at my whits end. I remember blacking out at multiple times. This one time in particular, my boys ran up to the couch, my husband was asleep in the recliner behind me. I was literally screaming into the back of the couch cushions. My, then, 4 y/o sons were so concerned. During my screams, I remember thinking, what the hell am I putting these poor boys through. I screamed one last time after contemplating that they would be better off without me. I had to make a change, on my own, without the doctors. I pushed myself up off the couch and said, no more. I WIĹL be here for my family, regardless of whether I need to take a lot of breaks, or be in a wheelchair, or limit/cancel activities. The old me is gone and I had to let go of her. It didn't mean that the new, altered me couldn't have a life as well.

Even though I have a medical education, CRPS was never taught in any of my schooling. After being diagnosed, I learned very quickly that it is not taught in medical school either. It is almost impossible to find a doctor that has taken a course, or specialized training in CRPS. Which makes this condition one of the worst, most dangerous diseases out there, as patients go for far too long without a diagnosis, without proper early treatment, with dismissed symptoms, medical gaslighting, inappropriate medication prescriptions just to 'appease' the steps they have to take to say they 'addressed the patients pain/symptoms' to the medical system, government or higher ups that control all of that. It is insane, barbaric, and inhumane how the medical system has failed CRPS Warriors! And, a lot more patients with other diagnosis' as well.

When I say it is dangerous, I will clarify by saying, CRPS is nicknamed the "suicide disease". By medical professionals lacking the education to properly diagnose patients early on, or at anytime; it is extremely dangerous. It can cause the patients to not get early access to treatment that may help put them in remission, only if caught and treated early. It can cause a patient to get so frustrated, and despaired with the doctors referring them from one doctor to the next, waiting for pre-authoritions, or approvals to see one doctor to the next, or wait for testing, or scans. Waiting for insurance pre-authorizations for the next doctor. Only to try and schedule an appt to find that they are months booked out. It is an ongoing, vicious cycle that haunts warriors. Warriors are already in the battle of their life with this complex condition, let alone trying to navigate the doctors and Healthcare system game. We often get so discouraged with no signs of help, relief, or end in sight. We break. We physically and mentally cannot handle all of this. And god forbid a warrior is on their own with no support system. Many warriors, end their own life as it is just too much. Too much pain, too much to handle with all that the doctors, insurance, and broken medical system puts them through. This is never ok or acceptable.

Final words of advise from a healthcare worker and CRPS warrior: please educated yourselves, and other doctors. Take courses in CRPS, treatment options and pain management. Have compassion with patients presenting with unrelenting pain that is out of proportion from the original injury. Help patients with treatment options that are out of the usual prescribing medications and referring out. Be the doctor that helps us, not hurts us.

Kari Dabrowski Nainggolan

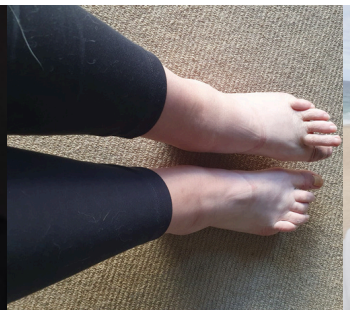
In march 2018, 4 months post surgery my surgeon diagnosed me with CRPS. He handed me a prescription for Cymbalta, mentioned side effects briefly and said to see your pain doc and good luck. I had no idea what was about to happen to me over the next 7 years.

The first 2 years I spent in bed, barely able to move from bed to bath without assistance. And the "golden standard" of medicine I was prescribed had so many awful side effects and weight gain that I had to stop taking it. My CRPS started in my left ankle and has recently spread to my left arm and possibly an organ. My left side of my body is also an inch smaller due to muscle wasting.

Like all Warriors, I struggle just to move every day and I seem to find a new version of a pain level 10 every few months. This disease has taken my career of 16 years, my independence, my bank account and left me isolated. I fight every day, but it is so much harder with little outside support. I am grateful for the Warriors Foundation.

Over the last 7 years I have tried every therapy I could find including alternative medicine and plant based medicine. Ironically, the therapies I have found to work were all self researched or suggested by my pharmasist/naturalpath.

My words to doctor's treating Warriors would be the following:
- don't dismiss us when we have small complaints as they are usually the beginning of the end when CRPS starts to spread
- don't prescribe medicines that cause weight gain and then question the patient why they wont lose weight
- work with all doctors to create the full picture so nothing gets missed
- just because we actually made it to our appointment and look presentable had no reflection on the agony we are hiding behind
- above all, simply listen. If you do, you will hear what you need



Yesina Cantu

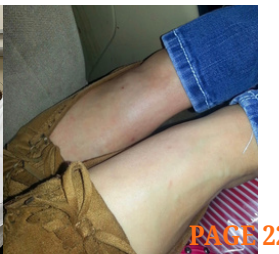
When I was 14 years old, I got rushed to the hospital with a frozen cold leg and shooting pain rushing down my leg. My leg started turning purple due to no circulation. The doctors were puzzled not able to come up with an explanation as to what was going on with my body. They had me stay in the hospital for a month, continuously doing constant tests and exams.

After being stuck in a wheelchair and losing my ability to walk I had to go to rehabilitation to learn how to walk again. During one of these sessions my leg flared up having temperature and color changes. The doctors rushed in with a handful of books and started searching to find my diagnosis, complex regional pain syndrome.

After finally getting a diagnosis, I got sent home because they knew they couldn't do anything for me at the hospital. I continued to go to my sessions to relearn to walk and was sent to a doctor out in Dallas, Texas. When I went, I had a low pain day and when she saw me, she assumed I was fine and sent me back home.

After being dismissed for the second time I decided to take matters into my own hands, and I did the research on my own disorder and found ways to take care of myself. With the right diet, light exercise/movement, and supplements I learned how to safely live with my disease. I searched to find the right doctors for myself and medication that was best suited for my body. I knew that the only person that knows what's best for me is me and I will continue to be the best advocate for myself.

I know that there are still days that I will have bad flare ups and there are still some things that I can not do, but I am capable of some amazing things, and I will continue to strive and achieve all I possibly can. With this mindset I have achieved getting my associates and bachelor's degree in business, became a licensed nail technician and started doing pageantry representing my platform of CRPS. I am committed to raising awareness, providing support, and advocating for those affected by this condition.





In loving honor and memory of

Jake Filut

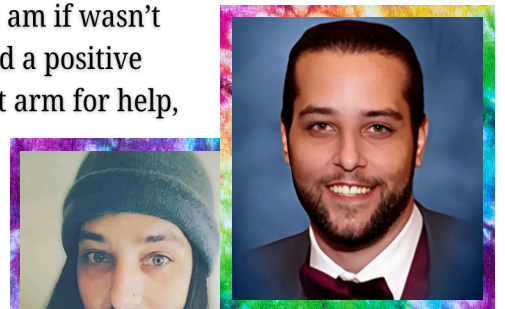
Dec 17, 1998 – Oct 12, 2023

Jake was an outgoing, creative with a passion music. He wrote and recorded his own rap songs and played multiple instruments. Jake was curious and fearless, always wanting to try new things. He loved the outdoors and taking long walks at the park

In January 2021, Jake injured his right arm while weight-lifting and lost complete it, while suffering from intense pain. Jake saw many doctors and was finally diagnosed with CRPS a year and nine months later. Jake was a true warrior who never complained and always did the best he could to remain active even though his physical abilities became extremely limited from pain.

A best friend describes him as "the kind of friend that everyone needs but not everyone finds...from making you laugh until you can't breathe or encouraging you to step out of your comfort zone and do something amazing...or helping you grow into the person you were born to be." Another friend said, "I don't think I'd be the person I am if wasn't blessed with Jake's life." Others describe Jake as kind, funny, open-minded, special, and a positive person. His 91 year-old grandmother said, "When I could no longer hold on to his right arm for help, I held onto his left arm...he was my rock!"

Jake brought joy to all he touched.



In loving honor and memory of

Matthew Manning

June 18, 1988 – July 11, 2023

Matt was naturally curious and a genuine epistemophile, making him wildly knowledgeable on a myriad of subjects. His mental acuteness allowed him to draw connections and bridge gaps. He was always up for a deep and thoughtful conversation. A true creative, Matt appreciated the beauty in everything.

Many people don't know that Matt suffered with Complex Regional Pain Syndrome (CRPS), which greatly limited his physical capabilities. Undeterred by this dehibilitating prognosis, he fought for his autonamy and continued to seek out ways to improve his quality of life. Despite the physical pain he felt, nothing stopped him from being Uncle Matt. He would absolutely light up when spending time with them.

Matt made a lasting impression on everyone who had the honor of knowing him. He was one of a kind, a truly unique individual in a small town, and will be deeply missed by his family and friends.

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