

CRPS

(Complex Regional Pain Syndrome)

TOWARDS THE DEVELOPMENT OF DIAGNOSTIC CRITERIA AND TREATMENT GUIDELINES

By

WCB Evidence Based Practice Group

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Compensation and Rehabilitation Services Division

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Introduction

Complex Regional Pain Syndrome (CRPS) is a diagnosis seen with increasing frequency in injured workers. It is a condition that can be very disabling, but for which the pathophysiology remains unclear. Currently, there are few, if any, universally effective treatments for this syndrome^{1,2,5,17,18,19}. At the Workers' Compensation Board of British Columbia Pain Programs, injured workers have been referred with this diagnosis at a rate that appears significantly higher than what is 'expected' in a typical chronic pain population.

Because of the increase in this diagnosis, there have been concerns raised that this 'label' may be used inappropriately, resulting in an extraordinary array of subsequent clinical, social, psychological and administrative costs^{2,3,4}. Injured workers, employers, clinicians and administrative bodies have all raised such issues in recent years.

The following paper is based on a review of the referenced medical literature concerning this condition. The objective of this review is to initiate a discussion on this subject and to develop reasonable and practical science based clinical criteria for the diagnosis and treatment of CRPS.

It is important to understand that CRPS is a specific diagnostic entity that would be considered in the differential diagnosis of what might be characterized as "chronic regional pain"⁵. These are often conditions that defy categorization with a specific clinical diagnosis based on known pathophysiology. Such conditions include:

- Complex Regional Pain Syndromes Type I and II
- Myofascial Pain Syndrome
- Cumulative Trauma Disorder
- Peripheral Pain of Central Origin

The above list is not exhaustive. It does, however, appear to illustrate the difficulty clinicians have had and continue to experience when attempting to assess and treat this small group of patients with their real yet ill-defined problem(s).

Introduction (cont'd)

It would be very useful to consider a consistent diagnostic label within the context of both clinical practice and the WCB for this regional pain of unknown etiology and pathophysiology. Guidelines for the management of such pain can then be developed and refined as necessary. Multiple compensation jurisdictions have medical advisory guidelines that present reasonable models for an approach to CRPS – these jurisdictions include the Alberta Workers' Compensation Board, The Department of Labor and Industry of Washington State², American Academy of Disability Evaluating Physicians (AADEP)⁶, and the Centre for Pain Studies, Colorado⁴.

The following document on diagnosis and treatment of CRPS is based on a non-systematic review of the present literature, a compilation of expert opinion and in depth discussion with numerous practitioners who frequently see patients with CRPS. As such, and within the field of Evidence-based Medicine, the level of evidence of this document on its own could never be graded higher than level 4 (see Appendix 1. Quality of Evidence). However, because of the nebulous nature of the disease process, the extraordinarily variable level of 'research' on this subject (both in diagnosis and treatment) and the wide array of treatment offered, it is felt this document may represent an initial attempt to put locally produced guidelines into practice.

History of Complex Regional Pain Syndrome as a Diagnosis

There have been a variety of diagnostic terms applied over the last one hundred and fifty years to the condition now thought of as CRPS^{7,8,9}. Specifically, these terms referred to the syndrome of pain, hyperesthesia, skin and vasomotor changes in an extremity usually coming on after an injury. This was first identified by Mitchell et al¹⁰ in survivors of gunshot wounds after the American civil war. Terms such as algodystrophy, Sudeck's atrophy and post-traumatic vasomotor syndrome have been used¹³. More recently the terms Reflex Sympathetic Dystrophy and Causalgia were and continue to be used.

A workshop of international experts for the International Association for the Study of Pain in Orlando in 1993 reached a consensus to rename Reflex Sympathetic Dystrophy and Causalgia as Complex Regional pain Syndrome Type I and Type II⁷, respectively. This conference also produced a set of diagnostic criteria for these conditions. These criteria were criticized as being mainly subjective and too inclusive of other conditions.⁶ Subsequent efforts have been made to design and validate stricter diagnostic criteria, by Wilson et al in 1996¹¹ and Bruehl et al of the Center for Pain Studies, Rehabilitation Institute of Chicago in 1999⁷. There are also diagnostic criteria in use by a number of Workers' Compensation systems including the Washington State Department of Labor and Industries in 1999² and the State of Colorado Department of Labor and Employment, Division of Workers' Compensation in 1998⁴. Finally, there are diagnostic criteria for CRPS, type 1 in development by The American Academy of Disability Evaluating Physicians (AADEP)⁶. The latter sets of criteria require factors of history AND subjective symptoms AND objective signs in order to establish a diagnosis.

This document outlines what the WCB of BC feels are reasonable diagnostic criteria. They are an amalgamation of expert opinion from BC based physicians and surgeons and existing criteria from other institutions including Washington State, Center for Pain Studies², State of Colorado⁴ and AADEP Draft Paper⁶.

Proposed Diagnostic Criteria for CRPS

The terms CRPS Type I and Type II are meant to describe certain chronic pain syndromes. They do not embody any assumptions about cause or pathophysiology. For the most part, the clinical phenomena characteristic of CRPS type I are the same as are seen in CRPS Type II. The central difference between Type I and Type II is that, by definition, Type II occurs following a known peripheral nerve injury with damage to nerve function, whereas Type I occurs in the absence of any known nerve injury.

Incidence of CRPS varies enormously from 0.05% to 35%^{1,13,14,23}, depending on the population surveyed or studied. It is estimated that 20%¹ - 35%²⁴ of these cases will remain incapacitated and only 20% - 30% return to their previous full-time employment²⁵. Most patients with widespread pain in an extremity that does not fit an obvious anatomic pattern do not have CRPS. It is often more appropriate to describe a patient as having “**regional pain of unknown origin**” than to diagnose CRPS.

Proposed Diagnostic Criteria:

1. The patient must have continuing pain that is disproportionate to any inciting event.
2. The patient must report at least one **symptom** in at least three out of the following four categories in the affected extremity:
 - Sensory: reports of hyperesthesia;
 - Vasomotor: reports of temperature asymmetry and/or skin colour changes and/or skin colour asymmetry;
 - Sudomotor/edema: reports of edema (with or without joint stiffness) and/or sweating changes and/or sweating asymmetry; or
 - Motor/trophic: reports of decreased range of motion and/or motor dysfunction (weakness, tremor, dystonia) and/or trophic changes (nails, hair, skin).
3. The patient must display at least one **sign** in two or more of the following categories in the affected extremity:
 - Sensory: evidence of hyperalgesia (to pinprick) or allodynia (to light touch);
 - Vasomotor: evidence of temperature asymmetry and/or skin color changes and/or asymmetry;
 - Sudomotor/edema: objective evidence of edema (with or without joint stiffness) and/or sweating changes and/or sweating asymmetry; or
 - Motor/trophic: evidence of decreased range of motion (including joint stiffness) and/or motor dysfunction and/or trophic changes.
4. It has been noted by a number of authors^{5,9,12,15}, that at this time there are no definitive diagnostic tests of CRPS including imaging modalities such as plain X-

ray or three-phase scintillography, EMG, thermography or sympathetic nerve blocks.

Given the above diagnostic criteria of patients having at least 3 symptoms and ≥ 2 signs, we expect to be able to 'accurately' diagnose $\pm 80\%$ of potential CRPS patients. With even more restrictive criteria, Bruehl et al⁷ showed that employing 4 symptoms and 2 signs as diagnostic criteria for CRPS is the most effective criteria to rule in and rule out CRPS across different populations.

A number of expert clinicians in this field suggest that criteria such as we have presented above may well be helpful in the later stages of the CRPS process and that perhaps less rigid criteria should be employed in an effort to 'pick up' the earlier cases. It is argued that only by doing this could one expect to positively affect outcomes on a larger population basis. While this is an attractive argument many, including the WCB would, at present prefer the more rigid criteria.

The literature, clinicians and administrative bodies will frequently debate what criteria are truly 'objective' vs. 'subjective' when considering such diagnostic criteria. The WCB feels there is no 'right' answer to this, but would like to point out that, in the face of solely subjective criteria in an insurance setting, careful consideration of other entities to explain complaints needs to be considered. It is recognized that many adjudicative decisions will be based on appropriate, science based diagnoses – hence, the need to ensure that the vast majority of clinicians dealing with CRPS patients feel comfortable with these criteria. Ultimately, it would be of benefit to have access to a clinical study or studies looking at clinician's diagnostic inter rater reliability, but this information on CRPS is not available. Hence the need for reasonable criteria that allows consistency in clinical diagnosis. The proposed diagnostic criteria is presented again, in tabular form in Table 1 at the end of this paper.

Sympathetic Blockade in the Diagnosis of CRPS

A patient's response to a diagnostic sympathetic block provides information about whether his/her pain is sympathetically maintained, but neither establishes nor refutes a diagnosis of CRPS. Therefore, a sympathetic block is not considered to be a definitive diagnostic test for CRPS.

In the patient with CRPS, the purpose of a sympathetic block is to guide treatment²⁰. If a CRPS patient responds positively to a sympathetic block (indicating that his/her pain is sympathetically maintained) repeat blocks might be useful in the overall treatment plan.

An Overview of Treatment

Experts in CRPS believe the probability of a patient developing this condition can be reduced by early mobilization/activation following injury or surgery. Conversely, unnecessarily prolonged immobilization following injury or surgery may set the stage of iatrogenic CRPS^{16,20,21}. With this in mind, it is clear that prevention of the development of this condition post injury is likely medicine's most important 'tool' in the overall management of CRPS. All therapy for CRPS should be directed toward the goals of physical restoration and pain control²¹. Details regarding treatment are presented in Tables 2 and 3 located at the end of this paper.

A. Physical Restoration

Experts agree that CRPS patients usually become trapped in a vicious cycle in which guarding and activity restrictions perpetuate the pain of CRPS. Therapy for CRPS should be directed toward breaking the pain cycle by having patients participate in a progressive activation program for the affected limb.

1. Because patients usually resist using the affected extremity, the physical restoration program generally requires direct supervision by a physical therapist or occupational therapist.
2. Involvement of a physical or occupational therapist is important so that repeated measurements of a patient's functional capacity can be made.
3. The frequency with which a patient receives physical or occupational therapy must be individualized by the attending physician. Generally, this would be daily treatment within a CRPS program.
4. Physical or occupational therapy occasionally continues beyond the time period during which pain control interventions such as sympathetic blocks are administered. Prolonged therapy, as long as there is evidence of ongoing improvement of function of the limb, is felt to increase the potential for better outcomes.
5. Patients need to understand they must use their symptomatic limb in the course of their usual daily activities as well as during physical or occupational therapy sessions. Patients must commit themselves to physical restoration on a 24-hour per day basis.

B. Pain Control

1. Interventions to reduce pain are typically needed so that patients can get enough relief to participate in an activation program.
2. It is crucial that pain control interventions be linked closely with physical/occupational therapy. Physical or occupational therapy sessions should be scheduled as soon as possible after a sympathetic block. The interval between block and therapy should always be less than 24-hours. In general, physical/occupational therapy should be directed toward activation and desensitization in the affected limb. Details are given in [Table 3](#).
3. Clinicians use a variety of medications to control pain in patients with CRPS. These include NSAIDs, α -adrenergic blockers, corticosteroids, calcitonin, antidepressants, antiseizure medications, mexiletine, opiates, capsaicin and sympathetic nerve blocks. The Workers' Compensation Board of BC has no formal guideline regarding a specific medication regimen for CRPS.
4. Opiates are not recommended as first line treatment in CRPS patients²⁷. Opiates should only be considered when non-opiates are unable to adequately control pain. Opiates should only be used as part of integrated pain management program in CRPS patients.
5. To this date there is insufficient evidence on the effectiveness of Spinal Cord Stimulation in treating CRPS patients²⁸.

C. Sympathetic Blocks

1. In a patient who meets the criteria for CRPS, **up to three sympathetic blocks** will be authorized to allow the attending physician **to determine whether the patient has sympathetically mediated pain**.
2. Additional blocks should be undertaken ONLY if there is evidence from the first three that the patient has sympathetically mediated pain.
3. The physician who performs each sympathetic block should document:
 - a. Measurable evidence that a sympathetic blockade in the target limb was achieved – e.g., hand/foot temperature before and after the block, observed color changes and/or venodilation.
 - b. The extent and duration of the patient's pain relief, based on a pain diary.
4. A patient should be seen by a physical or occupational therapist during the time interval when a sympathetic block would be expected to have an effect – that is, within a few hours of the block. The therapist should document the functional status of the patient's symptomatic limb during the therapy session.
5. The attending physician or the physician performing sympathetic blocks should correlate the information previously described in #3 and #4 to determine whether a block has produced the intended effects on pain, function and observable manifestations of CRPS.

D. Psychological Treatment

The clinical course of many patients with chronic pain, such as those with CRPS, may be complicated by pre-existing or concurrent psychological or psychosocial issues. A one time psychological/psychiatric consultation may be requested to assist in the evaluation of such patients.

E. Treatment Phases

Treatment is divided into six-week phases. A maximum of three phases may be authorized. The second phase will be authorized only if the first phase has led to demonstrable functional improvement. The third phase may be authorized only if the first and second phases have led to demonstrable functional improvement.

1. In the first six-week phase, up to five sympathetic blocks will be authorized (along with other accepted conservative measures such as medication management).
2. During the second six-week phase, a total of three sympathetic blocks will be authorized.
3. Up to three more sympathetic blocks may be authorized for patients who go on to the third phase of treatment.

F. Hospitalization

Hospitalization is rarely appropriate in the treatment of CRPS. The only exception to this would be when CRPS is diagnosed in an extremity, and there is a coexisting orthopedic condition amenable to surgery in that limb. In such cases, CRPS patients are at high risk for post-operative exacerbation of their condition. Therefore, it is reasonable and appropriate for such patients to be admitted to hospital prior to surgery so that aggressive pain control measures may be undertaken pre-operatively.

Proposed Outcome Evaluation

Each patient undergoing a diagnostic or treatment evaluation should also have other important outcomes objectively measured. With this in mind, the WCB would suggest:

- i) Functional assessment – MD, PT or OT musculoskeletal assessment of the affected extremity;
- ii) Visual analogue pain score assessment; and
- iii) Quality of life scoring (SF12, 36 or similar)

In any treatment program, all assessments/scorings would be appropriate and should be completed on admission and at the end of each 'session' or when the patient has plateaued or recovered.

Table 1. CRPS: Proposed Diagnostic Criteria (adapted from 7,22)

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1. Continuing pain, which is disproportionate to any inciting events.

 2. Report of at least one **symptom** in three of the four following categories:
 - Sensory: reports of hyperesthesia;
 - Vasomotor: reports of temperature asymmetry and/or skin colour changes and/or skin colour asymmetry;
 - Sudomotor/edema: reports of edema (with or without joint stiffness) and/or sweating changes and/or sweating asymmetry; or
 - Motor/trophic: reports of decreased range of motion and/or motor dysfunction (weakness, tremor, dystonia) and/or trophic changes (nails, hair, skin).

 3. At least one **sign** in two or more of the following categories:
 - Sensory: evidence of hyperalgesia (to pinprick) or allodynia (to light touch);
 - Vasomotor: evidence of temperature asymmetry and/or skin color changes and/or asymmetry;
 - Sudomotor/edema: objective evidence of edema (with or without joint stiffness) and/or sweating changes and/or sweating asymmetry; or
 - Motor/trophic: evidence of decreased range of motion (including joint stiffness) and/or motor dysfunction and/or trophic changes.
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Table 2. CRPS: Proposed Treatment Guidelines (adapted from 2,5)

1. **Conservative care:**
 - The goal of the treatment is physical restoration and pain control;
 - Early, aggressive care is encouraged; and
 - Emphasis should be on improved functioning of the symptomatic limb.

 2. **First 6 weeks of care:**
 - Physical/occupational therapy should be focused on increasing functional level;
 - Sympathetic or somatic blocks, maximum of **five**. Each block should be followed by physical/occupational therapy; and
 - Other medications are at MD's discretion as long as it promotes improved function.

 3. **After the 1st six weeks of care:**
 - Strongly consider psychiatric or psychological consultation if disability has extended beyond 3 months;
 - Continued physical/occupational therapy based on documented progress toward goals established during the first 6 weeks (Item 2 above); and
 - Sympathetic or somatic blocks only if response to previous blocks has been positive, maximum of 3 every six weeks for a maximum of 12 weeks (a maximum of 11 blocks can be delivered over the total 18 week period).

 4. **Sympathectomy is not the treatment of choice for CRPS :**
 - Sympathectomy should only be contemplated in highly unusual and extraordinary cases.
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Table 3. CRPS: Protocol for Physical/Occupational Therapy ^(adapted from 2,5).

1. **Evaluation should:**
 - Include a date of onset of original injury (to determine disease stage) and a date of onset of CRPS symptoms;
 - Establish a baseline for strength and motion;
 - Establish a baseline for weight bearing for lower extremity;
 - If lower extremity, evaluate distance able to walk and need for assistive device;
 - If upper extremity, establish a baseline for grip and pinch strength and shoulder range of motion;
 - If possible, measure the edema (e.g. using volume displacements); and
 - Define functional limitations.

 2. **Set specific functional goals for treatment related to affected extremity.**

 3. **All treatment programs should include a core of:**
 - A progressive active exercise program, including a monitored home exercise program;
 - Progressive weight bearing for the lower extremity (if involved);
 - Progressive improvement of grip strength, pinch strength and shoulder range of motion of the upper extremity (if involved); and
 - A desensitization program.

 4. **For specific cases, additional treatment options may be indicated to enhance effectiveness of the above core elements. Documentation should reflect reasons for these additional treatment options.**

 5. **Documentation should include:**
 - At least every two weeks, assessment of progress towards goals;
 - Response to treatment used in addition to core elements (item no.3 above); and
 - Evidence of motivation and participation in home exercise program such as using diary or quota system.
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Other Literature of Interest

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Appendix A

Workers' Compensation Board of BC - Evidence-based Practice group.

Quality of evidence (adapted from 29, 30, 31)

Table 1. Quality of Published Evidence

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| 1 | Evidence from at least 1 properly randomized controlled trial (RCT). |
| 2 | Evidence from well-designed controlled trials without randomization. |
| 3 | Evidence from well-designed cohort or case-control analytic studies, preferably from more than 1 centre or research group. |
| 4 | Evidence from comparisons between times or places with or without the intervention. Dramatic results in uncontrolled experiments could also be included here. |
| 5 | Opinions of respected authorities, based on clinical experience, descriptive studies or reports of expert committees. |