Complex Regional Pain Syndrome
Work-Related Complex Regional Pain Syndrome (CRPS): Diagnosis and Treatment

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Acknowledgements
I. INTRODUCTION

Complex regional pain syndrome (CRPS), sometimes referred to as reflex sympathetic dystrophy or causalgia, is an uncommon chronic condition with clinical features that include pain, sensory, sudomotor disturbances, trophic changes, and impaired motor function. This condition may involve the upper or lower extremities and can affect men or women of any age, race, or ethnicity. The majority of people with onset of CRPS are females and adults. Females are affected at least three times more than males. The pathophysiology of CRPS is not fully understood. When CRPS occurs it typically follows an injury, such as a fracture, sprain, crush injury, or surgery. Immobilization, particularly post-fracture or post-surgery, is a well-described risk factor.

Two types of CRPS have been described: CRPS I and CRPS II. For the most part, the clinical characteristics of both types are the same. The difference is based on the presence or absence of nerve damage: CRPS I (also known as reflex sympathetic dystrophy) is not associated with nerve damage, whereas CRPS II (also known as causalgia) is associated with objective evidence of nerve damage. Treatment for either form of CRPS should follow the recommendations in this guideline, although if there is objective evidence for CRPS II, other references and treatment guidelines for the particular nerve injury may also apply.

II. ESTABLISHING WORK-RELATEDNESS

CRPS may occur as a delayed complication of a work-related condition or its treatment. Usually, CRPS occurs following an injury. In rare situations, CRPS may occur following an occupational disease.

An injury is defined as “a sudden and tangible happening of a traumatic nature producing an immediate or prompt result and occurring from without, and such physical conditions as result there from”. The only requirement for establishing work-relatedness for an injury is that it occurs in the “course of employment”.

For an occupational disease, establishing work-relatedness requires a more critical analysis that demonstrates more than a simple association between the disease and workplace activities. Establishing work-relatedness for an occupational disease requires all of the following:

1. Exposure: Workplace activities that contribute to or cause the condition, and
2. Outcome: A medical condition that meets certain diagnostic criteria, and
3. Relationship: Generally accepted scientific evidence, which establishes on a more probable than not basis (greater than 50%) that the workplace activity (exposure) in an individual case was a proximate cause of the development or worsening of the condition (outcome).

Establishing CRPS as a work-related condition requires documentation of all of the following:
1. Another work-related condition has been previously accepted, and
2. A diagnosis of CRPS that meets the criteria in Section IV, and
3. CRPS involves the same body part as the accepted, work-related condition.

### III. PREVENTION

CRPS is believed to be incited by trauma or immobilization following trauma. It is most likely to occur in the setting of bone fracture, especially of the distal extremity. The greatest risk for CRPS appears to be certain types of fractures such as distal radial, tibial, and ankle as well as limited movement of the affected limb.⁶⁻⁹

CRPS may be preventable if the alert clinician is on the lookout for CRPS. Therefore, in addition to the usual protocols for a particular injury, close surveillance of patients at risk for CRPS is recommended. For such patients, extra office visits may be appropriate, especially if the clinician suspects a patient may not follow the expected course of recovery within the expected length of time.

The use of Vitamin C (500mg by mouth every day for 50 days) has been shown to reduce the incidence of CRPS following radial, foot, and ankle fractures.⁶⁻⁹*

CRPS may be prevented or arrested by early identification of risk factors and taking prompt action when they are present. The emphasis should be on pain control, mobilization, and monitoring from onset of acute injury through the normally expected treatment time, typically a few weeks to a few months. Following these few precautions can help prevent CRPS:

**A. KNOW THE RISK FACTORS**
1. Prolonged immobilization (e.g. due to bone fractures or soft tissue injury, especially in upper or lower distal extremities)
2. Longer than normal healing times
3. Delays in reactivation after immobility (e.g. due to inadequate control of acute pain)
4. Lack of weight-bearing on lower extremities
5. Tobacco use which can delay fracture healing
6. Reluctance to move or reactivate due to fear of pain or injury (fear avoidance)
7. Nerve damage

**B. IDENTIFY CASES EARLY AND TAKE ACTION**
1. Intentionally solicit symptoms and watch for signs
2. Educate the patient to immediately report any CRPS symptoms
3. Give clear and specific instructions to patients about mobilization and use of the injured part
4. Manage patients’ expectations about pain relief
5. Use vitamin C at recommended doses in cases of fracture

**C. ENCOURAGE ACTIVE PARTICIPATION IN REHABILITATION**
1. Have patient keep a recovery diary, logging pain level, symptoms, and activities
2. Provide or facilitate activity coaching
3. Set recovery goals with specified time frames (e.g. next office or PT visit)

* Based on Level I and Level II Evidence
4. Use medications or interventional procedures in concert with rehabilitative strategies

IV. MAKING THE DIAGNOSIS

Most patients with pain in an extremity do NOT have CRPS. Avoid the mistake of diagnosing CRPS primarily because a patient has widespread extremity pain that does not fit an obvious anatomic pattern. In many instances, there is no diagnostic label that adequately describes the patient’s symptoms. It is often more appropriate to describe the condition as “regional pain of undetermined origin” than to diagnose CRPS. However, it is equally important to identify CRPS when it does occur, so that appropriate treatment can be instituted.

A. SYMPTOMS AND SIGNS

CRPS is an uncommon syndrome based on a particular pattern of symptoms and signs in addition to pain. Symptoms and signs may be present at rest or elicited by exercise or activity involving the affected limb. The primary symptom associated with CRPS is continuous pain that is disproportionate to the inciting event. Pain is often described as “burning” or “sharp” and may be associated with changes in skin sensation such as hyperalgesia (increased sensitivity) or allodynia (pain perception to stimuli that are normally not painful). Other symptoms and signs in the affected area may include:

1. Skin temperature dysregulation
2. Skin color variability
3. Sweat dysregulation
4. Swelling or edema
5. Changes to the texture or growth pattern of hair, nails, or skin
6. Motor weakness, decreased range of motion (ROM), tremors, dystonia

B. THREE-PHASE BONE SCINTIGRAPHY

Three-phase bone scintigraphy can be a useful supplement to making the clinical diagnosis of CRPS. Abnormalities related to CRPS that may be seen in a three-phase bone scan include increased blood flow and increased blood pool uptake to the region of interest, with delayed images showing increased uptake in a periarticular pattern. Including the bone scan as a criterion is intended to increase diagnostic sensitivity. A normal bone scan neither increases nor decreases the likelihood of the diagnosis of CRPS. An abnormal bone scan is not required for a CRPS diagnosis.

C. DIAGNOSTIC CRITERIA

Diagnostic criteria for CRPS known as the “Budapest criteria” were adopted by the subcommittee, with slight modification, after careful consideration of existing criteria and available scientific evidence. Information about the sensitivity and specificity of the diagnostic signs and symptoms can be found in the literature.

†Based on Level II and Level IV Evidence
### Diagnostic Criteria for Complex Regional Pain Syndrome (CRPS)

To make a clinical diagnosis, the patient must meet *all* four of the following criteria:

1) Continuing pain, which is disproportionate to any inciting event

2) At least one symptom in *three of the four* following categories must be reported:
   - Sensory: Reports of hyperalgesia and/or allodynia (to pinprick, light touch, deep somatic pressure, and/or joint movement)
   - Vasomotor: Reports of instability and/or asymmetry of skin temperature and/or color
   - Sudomotor/Edema: Reports of instability and/or asymmetry of sweating and/or edema
   - Motor/Trophic: Reports of decreased range of motion and/or motor dysfunction (e.g. weakness, tremor, dystonia) and/or trophic changes (e.g. hair, nails, skin)

3) At least one sign in *two or more* of the following categories must be identified by objective clinical findings documented in the medical record over the course of one or more examinations:
   - Sensory: Evidence of hyperalgesia and/or allodynia (to pinprick, light touch, deep somatic pressure, and/or joint movement)
   - Vasomotor: Evidence of instability and/or asymmetry of skin temperature and/or color
   - Sudomotor/Edema: Evidence of instability and/or asymmetry of sweating and/or edema
   - Motor/Trophic: Evidence of decreased range of motion and/or motor dysfunction (e.g. weakness, tremor, dystonia) and/or trophic changes (e.g. hair, nails, skin)

   *A three-phase bone scan that is abnormal in a pattern characteristic of CRPS can be substituted for one of the signs in this section. (This is the committee’s modification of the Budapest criteria.)*

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

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### V. TREATMENT

#### A. HAVE A TREATMENT PLAN

Treatment for CRPS should be initiated early and aggressively. An interdisciplinary approach is often useful. A treatment plan should encourage patients to take an active role in their rehabilitation plan. This can include having the patient keep a journal, to record symptoms, activity tolerance, and pain and function levels. Emphasis should be on improving functional activity in the *symptomatic* limb and should include elements of the following:

- Physical therapy (PT) or occupational therapy (OT)
- Medication for pain control
- Psychological or psychiatric consultation and therapy
- Sympathetic blocks
- Multidisciplinary Program for Pain Management

1. **Physical and Occupational Therapy**

   A physical or occupational therapy treatment plan specific to CRPS should be developed by a therapist who is experienced in the treatment of CRPS. Therapy should be active, focused on desensitization, normalizing movement patterns, improving strength and range of motion and improving functional activities. A CRPS- focused physical or occupational therapy plan should include the following elements:
A. An Evaluation to include:
   1. Date of onset of original injury (helpful in determining if early or late stage) and a date of onset of the CRPS symptoms and signs
   2. Baseline objective measurements including ROM of all involved joints, strength, sensory loss, hypersensitivity, appearance, temperature, function (e.g. weight bearing and gait for lower extremity; fine motor tasks, pinch and grip for upper extremity), and use of assistive devices, braces and orthotics. If possible, include objective measurements of swelling

B. Specific, measurable functional goals which will allow assessment of progress and the effectiveness of treatment for the affected area

C. All treatment programs should include a core of:
   1. Desensitization
   2. Neuromuscular re-education, which might include graded motor imagery\textsuperscript{16,17}, mirror box therapy\textsuperscript{18} or other techniques to promote normalization of neuromuscular function\textsuperscript{1}
   3. A progressive, active exercise program designed to promote improvement in ROM, strength and endurance
   4. Activities targeted to attain the functional goals, e.g. weight bearing and gait training for the lower extremity and fine motor tasks for the upper extremity
   5. A monitored home exercise program to promote the patient’s participation in rehabilitation activities on a daily basis

D. Documentation should be done at least every two weeks to include:
   1. Reassessment of relevant baseline measurements described in A2 above. This provides objective evidence of response or non-response to treatment
   2. Assessment of progress toward functional goals (e.g. how the condition interferes with daily activities or activities related to employment)
   3. Level of patient motivation
   4. Participation in a home exercise program

2. Medication for Pain Control
Pain inhibits movement, and inadequate pain control may be an obstacle to activity, so judicious use of medications for pain control can be a useful adjunct to therapy. There is no drug with high-quality evidence to support use in either pain reduction or facilitation of function in CRPS. However, the committee recognizes that various medications are commonly used in clinical practice to manage pain or associated symptoms in CRPS.\textsuperscript{19} The categories of medications often used include non-steroidal anti-inflammatory drugs (NSAIDs), anticonvulsants\textsuperscript{20}, antidepressants, opioids, N-methyl-D-aspartate receptor antagonists (NMDA), antihypertensives, alpha-adrenergic agents, calcitonin\textsuperscript{21} and bisphosphonates\textsuperscript{22}. Selection of a particular agent may be influenced by the specific symptom or associated co-morbidities. These medications may be useful in helping a patient engage in therapy and regain function — the keys to successful management of CRPS.

The benefits of pain control should be weighed against the risks associated with adverse side effects. This is a particular challenge when using opioid medications for chronic pain. The Guideline on Opioid Dosing for Chronic Non-Cancer Pain, developed by the Washington State Agency Medical Directors Group (AMDG) can help:\textsuperscript{23}

\texttt{http://www.agencymeddirectors.wa.gov/Files/OpioidGdline.pdf}

3. Psychological or Psychiatric Consultation and Therapy
It is not uncommon for a fear-avoidance behavior pattern to emerge with a CRPS diagnosis. Patients are frequently fearful that pain indicates danger. They are sometimes concerned that ongoing pain means their condition has been misdiagnosed. Consequently, education and frequent reassurance are essential. This may be addressed using cognitive-behavioral therapy. In many cases, there is a more substantial

\textsuperscript{1} Based on Level II Evidence
psychological barrier to using the limb that warrants direct attention. If a co-morbid mental illness is identified that warrants formal psychiatric evaluation and treatment, screening or referral to the appropriate specialist may be needed.

4. Sympathetic Blocks
Sympathetic blocks have long been a standard treatment for CRPS and can be useful for a subset of cases. Stellate ganglion blocks (cervical sympathetic blocks) and lumbar sympathetic blocks are widely used in the management of upper and lower extremity CRPS. There is limited evidence to confirm effectiveness. An initial trial of up to three sympathetic blocks should be considered when the condition fails to improve with conservative treatment, including analgesia and physical therapy.

The most common way to administer sympathetic blocks is single local anesthetic injections. Selection of sympathetic block technique depends on each case, reflecting in part the patient’s needs and the interventional pain specialist’s preference and expertise. The current standard of practice is to use image guided approaches, such as with fluoroscopy and ultrasound, since complications of blind injections may include airway hematomas, inadvertent intravascular or central neuraxial injections, and esophageal puncture. Sympathetic blocks done without imaging guidance will not be authorized by the Department.

When sympathetic blocks are helpful, the benefit is evident within the first days following the nerve block. The optimal timing, number, or frequency of blocks, have not been specified. Patients who have a shorter duration of symptoms seem to have a greater response to treatment. Documentation of a physiologic response (e.g. change in skin temperature of the affected limb or Horner’s syndrome) is required to demonstrate that the block was successful. For sympathetic blocks to support lasting improvement, they should be combined with physical and behavioral therapies. Therapy should occur within 24 hours of the block or, if possible, on the same day of the block. An effective block is expected to produce at least 50% improvement in pain and a concomitant increase in function. Sympathetic blocks may be repeated, only when there is objective evidence of progressive improvement in pain and function.

5. Multidisciplinary Treatment
A multidisciplinary program for pain management will provide coordinated and closely monitored care using physical and/or occupational therapy, medication management, psychological screening and counseling, patient education, and other pain management techniques. The goal is to coordinate therapeutic interventions that ensure adequate pain control so reactivation of the affected body part can occur.

It is recommended that the attending provider and the pain management team communicate regularly about the patient’s treatment plan and progress towards treatment goals. Therapists and pain management staff should routinely report objective and quantifiable measures of functional improvement and pain tolerance and alert the attending provider if progress is not occurring. The objective is to act quickly so that the treatment team may take actions to quickly get the patient back on the expected course of recovery.

B. TREATMENT IN PHASES
Treatment can be thought of in phases. Although each phase has a general time frame, the time needed for an individual case is difficult to predict. Each phase can be shortened or lengthened as needed, allowing patients to move from one phase to another depending on their individual progress.

1. Phase One – Prevention and Mitigation of CRPS Risk Factors
The duration of Phase One will depend on the expected healing time for the specific injury, commonly spanning the first few weeks following the injury. The emphasis during Phase One is on pain control, appropriate mobilization, and monitoring of pain and function. After an initial injury, the patient should
be encouraged to move as much as is safe for whatever injury he or she has. PT/OT will be directed at what is appropriate for the specific injury and may be limited during this phase.

While there are no fixed rules as to the time of immobilization for a given injury, 6-8 weeks for the upper extremity and 8-12 weeks for the lower extremity are typical durations. It may be worth noting that mobility can continue in spite of casting. For example a patient in a long arm cast can still move his fingers, and a patient in an ankle cast can still move his toes. With appropriate immobilization, pain should generally decrease progressively with time. If pain is not decreasing over time, the provider must reassess the plan of treatment. If at any point the patient demonstrates unusual distress, pain complaints that appear to be out of proportion to the injury, or unexpectedly slow progress, the frequency of clinic visits should be increased. In this situation, it is important to consider the possibility of a missed diagnosis or an unrecognized comorbidity such as a behavioral or substance abuse disorder.

2. Phase Two – Recovery is Not Normal
The sooner treatment for suspected CRPS is initiated, the more likely it is that the long term outcome will be good. When recovery is delayed, and if no specific cause for the delay is identified, CRPS may be the diagnosis. Referral to a pain management or rehabilitation medicine specialist is strongly recommended.

3. Phase Three – CRPS Initial Treatment
Following a CRPS diagnosis, treatment should be initiated early and aggressively in the patient’s community whenever possible. Care should be coordinated and include physical or occupational therapy, psychological or psychiatric therapy, and medication management. An initial sympathetic block trial may be considered in cases that do not demonstrate functional gains during initial treatment.

4. Phase Four – CRPS Intensive Treatment
When the patient is unlikely to benefit from Phase Three treatment, an immediate referral to a multidisciplinary treatment program may be made. If the patient’s condition has not substantially improved within 6 weeks of Phase Three treatment, referral to an approved multidisciplinary treatment program is recommended.

5. Treatment Not Authorized for CRPS
The Department will not authorize the following interventions for CRPS:
   • Sympathectomy (no effect/no improvement in function\textsuperscript{26})
   • Spinal cord stimulation (non-covered benefit; see Health Technology Assessment decision 2010: \texttt{http://www.hta.hca.wa.gov/documents/adopted_findings_decision_scs_102510.pdf})
   • Ketamine infusions (no effect/no improvement in function, serious adverse events\textsuperscript{27,28})\textsuperscript{§}

\textsuperscript{§} Based on Level II Evidence
References


This course was developed from the public domain document: “Work-Related Complex Regional Pain Syndrome (CRPS): Diagnosis and Treatment” by the Industrial Insurance Medical Advisory Committee (IIMAC) and the Washington State Department of Labor & Industries.