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# **Complex Regional Pain Syndrome (RSD – Reflex Sympathetic Dystrophy)**

Discussion Paper prepared for

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This medical discussion paper will be useful to those seeking general information about the medical issue involved. It is intended to provide a broad and general overview of a medical topic that is frequently considered in Tribunal appeals.

Each medical discussion paper is written by a recognized expert in the field, who has been recommended by the Tribunal's medical counsellors. Each author is asked to present a balanced view of the current medical knowledge on the topic. Discussion papers are not peer reviewed. They are written to be understood by lay individuals.

Discussion papers do not necessarily represent the views of the Tribunal. A vice-chair or panel may consider and rely on the medical information provided in the discussion paper, but the Tribunal is not bound by an opinion expressed in a discussion paper in any particular case. Every Tribunal decision must be based on the facts of the particular appeal. Tribunal adjudicators recognize that it is always open to the parties to an appeal to rely on or to distinguish a medical discussion paper, and to challenge it with alternative evidence : see *Kamara v. Ontario (Workplace Safety and Insurance Appeals Tribunal)* [2009] O.J. No. 2080 (Ont Div Court).

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## Complex Regional Pain Syndrome – Type 1 (CRPS1)

This is a chronic limb pain that is accompanied by distinct physical features including swelling, abnormal sweating, temperature variation and structural changes to the skin and even the bones of the affected limb. The diagnosis excludes instances where a nerve injury is demonstrable, though the clinical features may overlap.

### Clinical Features:

CRPS(1) typically occurs as a consequence of limb trauma but may also be seen after myocardial infarction, stroke or even without obvious cause. The pain may be described as burning, aching or throbbing and can often be provoked by trivial stimuli such a light touch to the affected limb. The phenomenon is known as *Allodynia*. As the condition evolves the limb may change physically with thickened skin, dystrophic nails, diffuse swelling and alterations in colour and temperature. It may be abnormally dry or conversely excessively sweaty. In severe cases bone demineralization leads to marked osteoporosis; a condition known as *Sudek's Atrophy*.

The evolution of the disorder has been categorized into three stages:

Stage 1: Pain, swelling and vasomotor changes.

Stage 2: Soft tissue edema, skin thickening and muscle wasting.

Stage 3: Limited movement, contractures, nail changes and bony demineralization.

The condition may arrest at any stage<sup>3,4</sup> and early stage disorder may even revert to normal but late stage disease is most often permanent.

### Etiology

The origins<sup>2</sup> of the condition are at present unknown or at the very least disputed. Injury resulting in immobilization of the limb appears to be a common initiator but thereafter the mechanisms of injury are unclear.

role for the autonomic nervous system is suggested by the thermal and sudomotor features of the disorder. This has lead to its previous description as "*Reflex Sympathetic Dystrophy*" and the abnormality attributed to development of an abnormal reflex arc.

Other postulated mechanisms include excess production of Cytokines (regulatory proteins that the immune system releases to act as intercellular mediators in the generation of an immune response) such as substance P, which is demonstrable in some instances. However, it is unclear whether these are cause or consequence.<sup>2</sup> Most recently an immune mechanism has been described as a cause of pain persistence and IV immunoglobulin proposed as a treatment.

## Diagnosis

The diagnosis of CRPS(1)<sup>7</sup> is essentially clinical: though many tests have been devised to measure the degree of abnormality observed, none are sufficiently sensitive or specific to have predictive value. They may be supportive but only if prior probability of CRPS(1) is high.

The clinical diagnostic criteria have been proposed and codified over the past two decades. The International Association for the Study of Pain (IASP)<sup>1</sup> and the American Academy of Disability Evaluation<sup>7</sup> have published similar criteria. Both strive for optimal predictive value

**Table 1: IASP diagnostic criteria for complex regional pain syndrome (CRPS)\***

1. The presence of an initiating noxious event, or a cause of immobilization<sup>†</sup>
2. Continuing pain, allodynia, or hyperalgesia in which the pain is disproportionate to any known inciting event.
3. Evidence at some time of edema, changes in skin blood flow, or abnormal sudomotor activity in the region of pain (can be sign or symptom)
4. This diagnosis is excluded by the existence of other conditions that would otherwise account for the degree of pain and dysfunction.

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\* If seen without "major nerve damage" diagnose CRPS I; if seen in the presence of "major nerve damage" diagnose CRPS II.  
† Not required for diagnosis; 5-10% of patients will not have this.

and are adequately sensitive but, unless strictly applied, not very specific and liable to label non-sufferers inappropriately.

An inclusion criteria set (Table 1) is highly sensitive but relatively non-specific<sup>1</sup>.

A more stringent criteria set has been proposed (Table 2) and includes a useful description of the condition and its principal features<sup>6</sup>.

## Diagnostic Tests

As noted previously, a number of tests have been devised in an attempt to quantify the extent of the physical abnormality, to shed light on the nature of the disorder and even to provide a diagnostic tool. None have proved capable of doing so<sup>4</sup>. Most measure what is clinically obvious. All became increasingly positive as the disease progresses and are invariably normal or equivocal when the clinical examination is similarly uncertain. If they have a role, it may be when diagnosis is disputed because of problems interpreting the clinical data.

**Table 2: Proposed clinical diagnostic criteria for CRPS****General definition of the syndrome:**

CRPS describes an array of painful conditions that are characterized by a continuing (spontaneous and/or evoked) regional pain that is seemingly disproportionate in time or degree to the usual course of any known trauma or other lesion. The pain is regional (not in a specific nerve territory or dermatome) and usually has a distal predominance of abnormal sensory, motor, sudomotor, vasomotor, and /or trophic findings. The syndrome shows variable progression over time.

To make the clinical diagnosis, the following criteria must be met.

1. Continuing pain, which is disproportionate to any inciting event
2. Must report at least one symptom in three of the four following categories:

**Sensory:** Reports of hyperesthesia and/or allodynia

**Vasomotor:** Reports of temperature asymmetry and/or skin color changes and/or skin color asymmetry.

**Sudomotor/Edema:** Reports of edema and/or sweating changes and/or sweating asymmetry.

**Motor/Trophic:** Reports of decreased range of motion and/or motor dysfunction (weakness, tremor, dystonia) and/or trophic changes (hair, nail, skin)

3. Must display at least one sign at **time of evaluation** in two or more of the following 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 (>1oC) and/or skin color changes and/or asymmetry

**Sudomotor/Edema:** Evidence of edema and/or sweating changes and/or sweating asymmetry

**Motor/Trophic:** Evidence of decreased range of motion and/or dysfunction (weakness, tremor, dystonia) and/or trophic changes (hair, nail, skin)

4. There are no other diagnosis that better explains the signs and symptoms

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**For research purposes,** diagnostic decision rule should be at least one symptom in all four symptoms categories and at least one sign (observed at evaluation) in two or more signs categories.

## Management

A variety of treatments have been proposed, none of certain value. Foremost among them have been attempts to block the putative, aberrant, reflex arc either peripherally with drugs such as Guanethidine or centrally with sympathetic ganglion blockade. In all instances the earlier treatments are applied the greater the apparent benefit, but in the absence of randomized studies it is difficult to exclude the effect of natural history i.e. spontaneous improvement or recovery.

## Disability Implications

Currently the most comprehensive review of the disability implications of CRPS(1) is that performed on behalf of the American Academy of Disability Evaluation Physicians (AADEP)<sup>7</sup>. This was an extensive review of the literature to 2002 and reinforced the opinion that the diagnosis is a clinical one. It also concluded that disability should be evaluated according to conventional standards. "Impairment is based on objectively validated limitations in activities of daily living (ADL)".

The authors present a scale of disability to be anticipated and quantified. It provides a decision hierarchy that is as objectively based as current knowledge allows. Causally relating the condition to a workplace injury requires an accurate clinical diagnosis and clear documentation of the injury. Deficiency in either sphere, combined with the possibility of a non-traumatic precipitant, only adds to the complexity of disability evaluation.

## Summary

CRPS(1) is an unusual but well recognized cause of prolonged functional disability that may arise from workplace injury. Since it has many triggers

attribution to the workplace may be problematic. Once suspected, the diagnosis depends on the presence of a defined aggregation of clinical symptoms and physical signs that need to be rigorously sought and documented. No laboratory or imaging tests are definitive but they may support the clinical diagnosis. They are not a substitute for the clinical description.

## References

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