

## ICD-11 Development Version Text relating to CRPS

### Description

Complex regional pain syndrome (CRPS) is a chronic pain condition in an extremity with a variable course over time. It is characterized by continuing regional pain (not in a specific nerve territory or dermatome), usually with distal predominance or distal-to-proximal gradient. It typically arises after tissue trauma and is seemingly disproportionate in magnitude or duration to the usual course of pain after such tissue trauma. CRPS is characterized by signs indicating autonomic and neuro-inflammatory changes in the affected body region varying between patients and over time. Often, CRPS is accompanied by significant emotional distress or functional disability. CRPS is multifactorial.

### Additional Information

Complex regional pain syndrome (CRPS) is a chronic pain condition in an extremity with a variable course over time. It is characterized by continuing (spontaneous and/or evoked) regional pain (not in a specific nerve territory or dermatome), usually with distal predominance or distal-to-proximal gradient. It typically arises after tissue trauma and is seemingly disproportionate in magnitude or duration to the usual course of pain after such tissue trauma.

CRPS is characterized by signs indicating autonomic and neuro-inflammatory changes in the affected body region varying between patients and over time.

Often, CRPS is accompanied by significant emotional distress (anxiety, anger/frustration or depressed mood) and/or functional disability (interference in daily life activities and reduced participation in social roles). As any chronic pain condition, CRPS is multifactorial: biological, psychological and social factors contribute to the disease-phenotype. The diagnosis is based on clinical signs and symptoms alone, so the condition can be diagnosed independent of identified biological or psychological contributors and should only be made if there is no other diagnosis which would better account for the presenting signs and symptoms.

Alternative chronic pain diagnoses to be considered include chronic neuropathic pain and chronic musculoskeletal pain including rheumatological disease. Where regional limb pain is associated with discrete peripheral nerve damage, CRPS can only be diagnosed if CRPS signs and symptoms extend beyond the identified injured nerve territory; clinical features of the nerve lesion (numbness, paraesthesias) may be restricted to the nerve territory involved; this situation is also termed 'CRPS II'.

Diagnostic criteria:

Criteria: Given the absence of a diagnostic test, CRPS is diagnosed on the basis of clinical signs and symptoms.

Conditions A to E are fulfilled:

A. Chronic pain in a limb (persistent or recurrent for longer than 3 months) is present

B. The pain is associated with at least one symptom in three of the four following categories\*:

B.1 Sensory: hyperalgesia and/or allodynia

B.2 Vasomotor: temperature asymmetry and/or skin color changes and/or skin color asymmetry

B.3 Sudomotor/edema: edema and/or sweating changes and/or sweating asymmetry

B.4 Motor/trophic: decreased range of motion and/or motor dysfunction (weakness, tremor, dystonia) and/or trophic changes (hair, nails, skin)

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

C.1 Sensory: hyperalgesia (to pinprick) and/or allodynia (for example to light touch, deep somatic pressure, or joint movement)

C.2 Vasomotor: temperature asymmetry and/or skin color changes and/or asymmetry

C.3 Sudomotor/edema: edema and/or sweating changes and/or sweating asymmetry

C.4 Motor/trophic: decreased range of motion and/or motor dysfunction (weakness, tremor, dystonia) and/or trophic changes (hair, nails, skin)

D. The pain is associated with at least one of the following:

D.1 Emotional distress due to pain is present.

D.2 Interference with daily life activities and social participation.

E. The pain is not better explained by another chronic pain condition.

Different types of CRPS can be distinguished:

CRPS Type I: Develops after any type of trauma, especially limb fracture or soft tissue lesion.

CRPS Type I does not involve discrete nerve damage. Diagnostic signs and symptoms of CRPS Type I and CRPS Type II are identical.

CRPS Type II: Occurs after trauma associated with discrete peripheral nerve damage as indicated by neurological examination, electrodiagnostic testing, or other quasi-objective testing. While clinical features of the nerve lesion (numbness, paraesthesias) are restricted to the nerve territory involved, CRPS signs and symptoms must extend beyond the identified nerve territory.

Diagnostic signs and symptoms of CRPS Type I and CRPS Type II are identical.

CRPS with Remission of Some Features: This category is used to refer to patients who were documented to meet full CRPS criteria (either CRPS 1, or CRPS 2) at an earlier point in time but who currently do not display sufficient signs and symptoms to meet full criteria. Patients in this category are not necessarily improved with regards to pain intensity nor are they free of all CRPS-related signs and symptoms.

Clarification about the application of the diagnostic criteria:

- All patients should be asked systematically about all symptoms listed in the criteria at each formal diagnostic evaluation, even if they have not previously reported certain symptoms. CRPS signs and symptoms are clinically observed to fluctuate over time.

- Clarification of the terms “asymmetry” and “changes” as used in the CRPS criteria: For unilateral CRPS, assess asymmetry by comparing the affected side (the side that is most painful) to the unaffected side. For (much rarer) bilateral CRPS, assess changes in the affected limbs relative to an unaffected limb in the patient or to the limbs of a typical healthy individual. Asymmetry is based on clinical judgment only rather than any pre-specified criteria.

- For evaluating possible spreading of CRPS beyond a single limb, the full ICD-11 diagnostic criteria must be applied to each limb individually. True spreading of CRPS is defined as CRPS that meets full ICD-11 diagnostic criteria for multiple limbs – extension of pain to other limbs, which is not unusual, in the absence of other CRPS features is not formally considered to be spreading CRPS.

- SYMPTOMS (as given by or elicited from patient, either currently or historically)

o Hyperalgesia is any minor painful stimulus that is now perceived as intense or prolonged;

o Allodynia is a usually innocuous stimulus now perceived as painful. It is not unusual for a patient to report numbness or hyposensitivity even in the context of clear allodynia in the same body region.

o Temperature asymmetry that is obvious to the patient.

o Color asymmetry that is obvious to the patient.

o Sweating asymmetry that is obvious to the patient.

o Edema that is obvious to the patient (may be described as swelling).

o Dystrophic changes (increased or decreased growth, altered texture) of nails, hair, or skin as described by the patient.

o Motor abnormalities such as weakness, tremor, dystonia (limb locked in unusual position) or jerking of the limb as described by the patient. Decreased range of motion as described by the patient.

- SIGNS (as observed by examiner on the exam date)

o Hyperalgesia is indicated by a mildly noxious stimulus being perceived as intensely painful or the pain lasting longer than the duration of the stimulus. Tested clinically by a single pinprick applied to the affected side (in center of most affected region) and the same site of the contralateral limb.

o Allodynia: Allodynia can be indicated in multiple ways, all based on reporting pain in response to a normally non-painful stimulus. Stimuli used in clinical assessment can include light touch, vibration, cool or warm temperature, tissue pressure in the affected area, or joint movement.

o Suggested clinical assessment procedures follow: allodynia to light touch as tested by light manual touch (or brush); allodynia to tissue pressure as assessed by pressure applied to a joint or other tissue using the evaluator's finger with just enough pressure to make the fingernail bed of the evaluator blanch (turn white), allodynia to vibration as assessed using a graded tuning fork over bony prominence in affected limb; allodynia to temperature.

o Temperature asymmetry in the affected area compared to the comparable area on the contralateral extremity that is obvious to the touch of the dorsum of the hand of the examiner.

o Obvious color asymmetry of a regional nature (i.e. hand, foot, knee or larger region). Please specify the nature of the color changes, for example red, blue, pale, mottled or around a scar.

o Obvious sweating asymmetry.

o Asymmetric edema as observed by the examiner, or may be quantified by objectively measured volumetry (cm<sup>3</sup>) compared side to side, or limb circumference as assessed by tape measure at same location side to side. If a graduated cylinder large enough to hold the extremity or tape measure is unavailable, please just omit the quantification, as long as the categorical "yes/no" response is marked.

o Dystrophic changes of the affected extremity as noted by the examiner in nails, hair (increased or decreased growth) or skin (shiny, thin, thick etc) of the affected side. Please describe the nature of these changes.

o Motor abnormalities as noted by the examiner such as decreased range of motion, weakness, tremor, or dystonia on the affected side.