

# **COMPLEX REGIONAL PAIN SYNDROME**

## **What is complex regional pain syndrome?**

Complex regional pain syndrome (CRPS) is a chronic pain condition that is believed to be the result of dysfunction in the central or peripheral nervous systems. Typical features include dramatic changes in the color and temperature of the skin over the affected limb or body part, accompanied by intense burning pain, skin sensitivity, sweating, and swelling. CRPS I is frequently triggered by tissue injury; the term describes all patients with the above symptoms but with no underlying nerve injury. Patients with CRPS II experience the same symptoms but their cases are clearly associated with a nerve injury.

Older terms used to describe CRPS are "reflex sympathetic dystrophy syndrome" and "causalgia," a term first used during the Civil War to describe the intense, hot pain felt by some veterans long after their wounds had healed.

CRPS can strike at any age and affects both men and women, although most experts agree that it is more common in young women.

## **What are the symptoms of CRPS?**

The key symptom of CRPS is continuous, intense pain out of proportion to the severity of the injury (if an injury has occurred), which gets worse rather than better over time. CRPS most often affects one of the extremities (arms, legs, hands, or feet) and is also often accompanied by:

- "burning" pain
- increased skin sensitivity
- changes in skin temperature: warmer or cooler compared to the opposite extremity
- changes in skin color: often blotchy, purple, pale, or red
- changes in skin texture: shiny and thin, and sometimes excessively sweaty
- changes in nail and hair growth patterns
- swelling and stiffness in affected joints
- motor disability, with decreased ability to move the affected body part

Often the pain spreads to include the entire arm or leg, even though the initiating injury might have been only to a finger or toe. Pain can sometimes even travel to the opposite extremity. It may be heightened by emotional stress.

The symptoms of CRPS vary in severity and length. Some experts believe there are three stages associated with CRPS, marked by progressive changes in the skin, muscles, joints, ligaments, and bones of the affected area, although this progression has not yet been validated by clinical research studies.

***Stage one*** is thought to last from 1 to 3 months and is characterized by severe, burning pain, along with muscle spasm, joint stiffness, rapid hair growth, and alterations in the blood vessels that cause the skin to change color and temperature.

***Stage two*** lasts from 3 to 6 months and is characterized by intensifying pain, swelling, decreased hair growth, cracked, brittle, grooved, or spotty nails, softened bones, stiff joints, and weak muscle tone.

In ***stage three*** the syndrome progresses to the point where changes in the skin and bone are no longer reversible. Pain becomes unyielding and may involve the entire limb or affected area. There may be marked muscle loss (atrophy), severely limited mobility, and involuntary contractions of the muscles and tendons that flex the joints. Limbs may become contorted.

### **What causes CRPS?**

Doctors aren't sure what causes CRPS. In some cases the sympathetic nervous system plays an important role in sustaining the pain. The most recent theories suggest that pain receptors in the affected part of the body become responsive to a family of nervous system messengers known as catecholamines. Animal studies indicate that norepinephrine, a catecholamine released from sympathetic nerves, acquires the capacity to activate pain pathways after tissue or nerve injury. The incidence of sympathetically maintained pain in CRPS is not known. Some experts believe that the importance of the sympathetic nervous system depends on the stage of the disease.

Another theory is that post-injury CRPS (CRPS II) is caused by a triggering of the immune response, which leads to the characteristic inflammatory symptoms of redness, warmth, and swelling in the affected area. CRPS may therefore represent a disruption of the healing process. In all likelihood, CRPS does not have a single cause, but is rather the result of multiple causes that produce similar symptoms.

### **How is CRPS diagnosed?**

CRPS is diagnosed primarily through observation of the signs and symptoms. But because many other conditions have similar symptoms, it can be difficult for doctors to make a firm diagnosis of CRPS early in the course of the disorder when symptoms are few or mild. Or, for example, a simple nerve entrapment can sometimes cause pain severe enough to resemble CRPS. Diagnosis is further complicated by the fact that some people will improve gradually over time without treatment.

Since there is no specific diagnostic test for CRPS, the most important role for testing is to help rule out other conditions. Some clinicians apply a stimulus (such as touch, pinprick, heat, or cold) to the area to see if it causes pain. Doctors may also use triple-phase bone scans to identify changes in the bone and in blood circulation.

### **What is the prognosis?**

The prognosis for CRPS varies from person to person. Spontaneous remission from symptoms occurs in certain people. Others can have unremitting pain and crippling, irreversible changes in spite of treatment. Some doctors believe that early treatment is helpful in limiting the disorder, but this belief has not yet been supported by evidence from clinical studies. More research is needed to understand the causes of CRPS, how it progresses, and the role of early treatment.

### **How is CRPS treated?**

Because there is no cure for CRPS, treatment is aimed at relieving painful symptoms so that people can resume their normal lives. The following therapies are often used:

***Physical therapy:*** A gradually increasing exercise program to keep the painful limb or body part moving may help restore some range of motion and function.

***Psychotherapy:*** CRPS often has profound psychological effects on people and their families. Those with CRPS may suffer from depression, anxiety, or post-traumatic stress disorder, all of which heighten the perception of pain and make rehabilitation efforts more difficult.

***Sympathetic nerve block:*** Some patients will get significant pain relief from sympathetic nerve blocks. Sympathetic blocks can be done in a variety of ways. One technique involves intravenous administration of phentolamine, a drug that blocks sympathetic receptors. Another technique involves placement of an anesthetic next to the spine to directly block the sympathetic nerves.

***Medications:*** Many different classes of medication are used to treat CRPS, including topical analgesic drugs that act locally on painful nerves, skin, and muscles; antiseizure drugs; antidepressants, corticosteroids, and opioids. However, no single drug or combination of drugs has produced consistent long-lasting improvement in symptoms.

***Surgical sympathectomy:*** The use of surgical sympathectomy, a technique that destroys the nerves involved in CRPS, is controversial. Some experts think it is unwarranted and makes CRPS worse; others report a favorable outcome. Sympathectomy should be used only in patients whose pain is dramatically relieved (although temporarily) by selective sympathetic blocks.

***Spinal cord stimulation:*** The placement of stimulating electrodes next to the spinal cord provides a pleasant tingling sensation in the painful area. This technique appears to help many patients with their pain.

**Intrathecal drug pumps:** These devices administer drugs directly to the spinal fluid, so that opioids and local anesthetic agents can be delivered to pain-signaling targets in the spinal cord at doses far lower than those required for oral administration. This technique decreases side effects and increases drug effectiveness.

### **Additional Information**

- StopPain.org - <http://www.stoppain.org>
- National Pain Foundation - <http://www.nationalpainfoundation.org/>
- International Research Foundation for CRPS -  
<http://www.rsdfoundation.org/>

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