

Complex Regional Pain Syndrome Formerly Known as Reflex Sympathetic Dystrophy (RSD)

Complex regional pain syndrome (CRPS) is a pain condition that is constant over a long period of time and is believed to be the result of dysfunction in the central or peripheral nervous systems. CRPS is characterized by pain, swelling or stiffness in the affected hand or extremity. The pain may be out of proportion to the injury that triggered it. CRPS is usually associated with an injury, which can sometimes be as minor as a paper cut or small bruise. It causes the nervous system to misfire and send frequent or constant signals to the brain that are interpreted as painful. The nervous system becomes overactive, causing intense burning or aching pain, along with swelling and changes in skin color and moisture.

Causes

The causes of CRPS are not known. CRPS can begin after a minor injury, such as a sprain or small cut, or after major trauma or surgery. Injury to a nerve may also provoke its onset. It is most common among individuals between 25 and 55 years of age, though anyone of any age can be affected. CRPS is three times more likely to occur in women than men. An estimated 60,000 Americans are affected by CRPS.^{1,2}

Signs and Symptoms

The key symptom of CRPS is continuous, intense pain out of proportion to the severity of the injury (if an injury has occurred). CRPS most often affects one of the extremities and is also often associated with the following symptoms:

- "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

The pain may spread 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.

Diagnosis

There is no single test to confirm a diagnosis of CRPS. The diagnosis is primarily through observation of signs and symptoms. Patients must be examined by a qualified physician who does a thorough history and physical examination. X-rays, MRI, EMG/NCV, bone scans, thermography, or pain imaging where available may be helpful. Consultation with other specialists may be needed, and a pain clinic is often recommended.

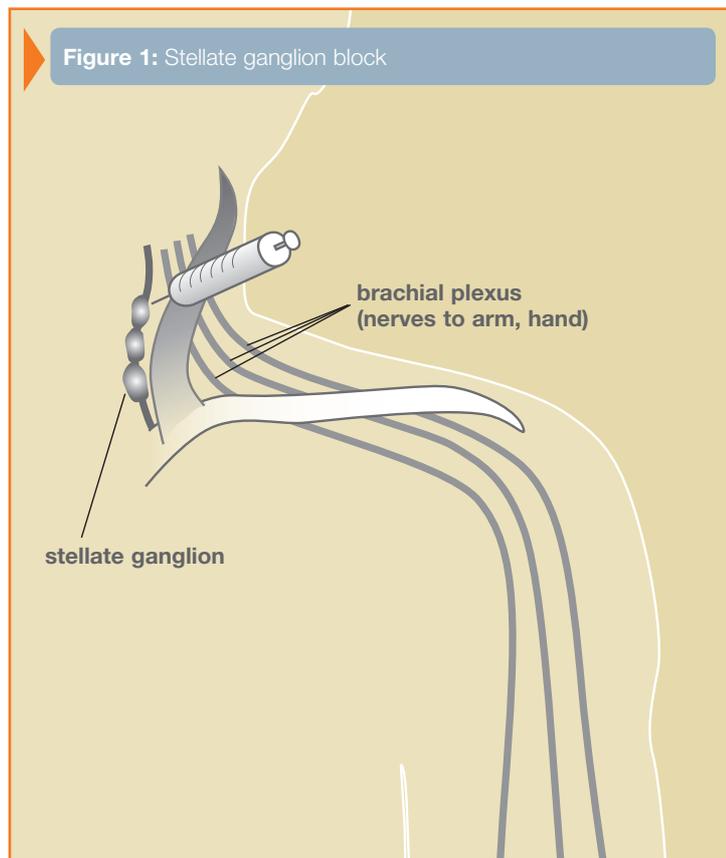
Treatment

The earlier the diagnosis of CRPS is made and treatment started, the better the chance for recovery. Treatment is varied and depends on both the severity of the symptoms and the duration of the problem. Aerobic conditioning, relief of sleep disorders, and treatment of psychological problems can be helpful for treatment. Some patients may have a chronic physical problem, such as a compressed or entrapped nerve, that needs to be addressed. Since there is no simple cure for CRPS, treatment is intended to relieve painful symptoms so that patients can resume their normal lives as well as optimize use of the hand or extremity.

Any of the following may be employed to treat CRPS, often in combination:

- **Occupational/Physical Therapy.** An increasing exercise program to help with motion may help preserve or restore mobility and function to the affected hand. Overall aerobic conditioning is very important to improve coping ability, sleep, and pain control.
- **Psychotherapy.** CRPS can have profound psychological effects on patients and their families. Many people with CRPS have depression, anxiety, or post-traumatic stress disorder. A psychologist or psychiatrist may be able to improve coping ability and motivation as well as detect and address any substance dependency issues.

Figure 1: Stellate ganglion block



- **Nerve Blocks.** Many patients experience significant relief from nerve blocks, in which local anesthetic is injected to numb nerves. By relieving pain, blocks can enable more effective therapy, improve mood, and improve level of activity. Stellate ganglion blocks may be used to numb the stellate ganglion, which is a cluster of sympathetic nerves at the base of the neck, in an effort to reduce the over-activity of the sympathetic nerves seen in CRPS (see Figure 1).
- **Medications.** Many different drugs are used to treat CRPS and associated conditions, such as sleep disorders, depression, and anxiety. Medications may include topical analgesics, anti-seizure drugs, antidepressants, corticosteroids, muscle relaxants, opioids, and sleeping medications.
- **Surgery.** If the CRPS is from a compressed nerve, such as with carpal tunnel syndrome, then surgery to release pressure on the nerve may be needed (e.g., carpal tunnel release). Rarely, an operation known as sympathectomy is used to divide the sympathetic nerves in patients who are helped by nerve blocks, and its use is controversial.

Other options include spinal cord stimulation and intrathecal drug pumps, in which pain medications are injected continuously into the space around the spinal cord. Deep brain stimulation and Electrotherapy (ECT) have also been used, but new therapies continue to emerge.

Prognosis

Each patient with CRPS responds differently to treatment. Spontaneous improvement occurs in some persons. Others may have crippling, irreversible changes in spite of appropriate treatment. Most physicians believe that early treatment is helpful to limit the disability from CRPS. More research is needed to understand the causes, the development of the disease, and how treatment can alter its course.

References

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2. Sandroni P, Benrud-Larson LM, McClelland RL, Low PA. Complex regional pain syndrome type I: incidence and prevalence in Olmsted county: a population-based study. *Pain.* 2003;103:199-207.

Fact Sheet: Chronic Regional Pain Syndrome



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What is Reflex Sympathetic Dystrophy (RSD) or Complex Regional Pain Syndrome (CRPS)

This is a chronic pain syndrome that can occur after an injury, surgery, a stroke or heart attack. It is an abnormally severe and / or prolonged manifestation of a normal post-injury response.

The following features are usually present:

- **PAIN**
This is always present and usually out of proportion to the original injury. This may be burning or shooting or the hand may be sensitive to touch, pressure or light movement.
- **CIRCULATION**
The limb may be pale, appear blue or red or may be hot or cold. There may be abnormal sweating.
- **SWELLING**
May be present or the skin may be shiny.
- **STIFFNESS**
Occurs due to decreased muscle strength or secondary to lack of movement due to pain.

Causes :

There is no correlation between the severity of the injury and RSD developing. The injury / surgery causes nerves to become sensitive to signals which they do not usually respond eg. light touch may cause excruciating pain.

The sympathetic nerves are not the usual nerves for feeling pain or sensation. They control sweating and colour changes in the skin.

In RSD there is often an abnormal firing of these nerves secondary to the injury. RSD can affect a single digit, a single nerve distribution or an entire limb.

RSD is initiated by trauma to a limb and can be made worse by post-traumatic events eg. tight casts or acute carpal tunnel syndrome.

Fractures of the distal radius and ulna are the most common injuries producing RSD. Traumatic or surgical injury to a cutaneous nerve may precipitate RSD or it may occur following surgery for Dupuytren's disease.

The literature does not support a psychological causation. Cigarette smoking is statistically linked to RSD.

Demographics :

The majority of patients are between 30 and 55 years of age (average 45 years). Women are affected three times more commonly than men. Cigarette smoking is statistically linked to RSD.

80% of patients with RSD diagnosed within 1 year of injury will improve significantly. However, 50% of patients with untreated symptoms lasting for more than 1 year will have profound residual impairment.

Investigations

X-rays and / or bone scans may be useful in some cases.

Fact Sheet: Complex Regional Pain Syndrome (CRPS)

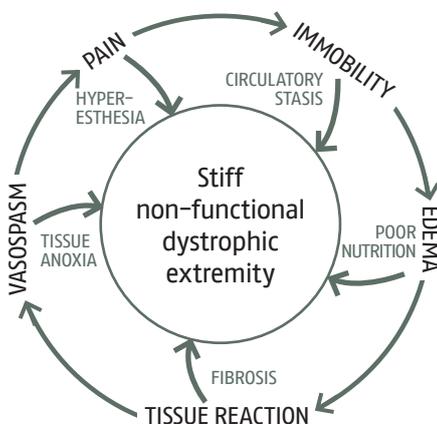


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Treatment

- **Physiotherapy** – this is an essential part of the treatment of RSD. Early active motion of the affected joints is the goal.
“Scrub and carry” programme – stress loading which involves applying weight to the hand while performing an activity eg using a scrubbing brush or carrying a bucket of water or bag in the affected hand.
No passive exercise at all is allowed by anyone except the patient. Heat or ice packs may be helpful but extremes of temperature should be avoided. Deep friction massage may also be helpful.
- **Splinting** – this is an important part of treatment especially in the hand. Splints are used to prevent contracture formation and maintain a balanced hand.
- **(Transcutaneous Electrical Stimulation) or TENS** – electrodes placed on the skin produce cutaneous tingling and can help to block pain transmission.
- **Blocks** – injections around the nerves supplying the hand or leg may temporarily block the pain signals these nerves are sending. Injections in the neck may also block these pain signals. Three or four such blocks may be required.
- **Drugs** – The aim is to break the pain cycle and reduce the inflammation. Pain killers and anti-inflammatories like Voltaren or Celebrex are used. Antidepressant medications such as Tryptanol can be used. These act on the nerves to dampen the signals they are processing.
- **Cognitive Behavioural Therapy (CBT)** – Psychologists are able to help patients by offering ways of coping with the pain and teaching patients relaxation methods. CBT is used to improve skills in the management of stressful situations. A positive outlook and approach can help the patient’s recovery.
- **Surgery** – This is occasionally indicated in the presence of a chronic painful lesion in an extremity eg. a documented carpal tunnel syndrome or a painful neuroma. This will usually be performed under a nerve block.

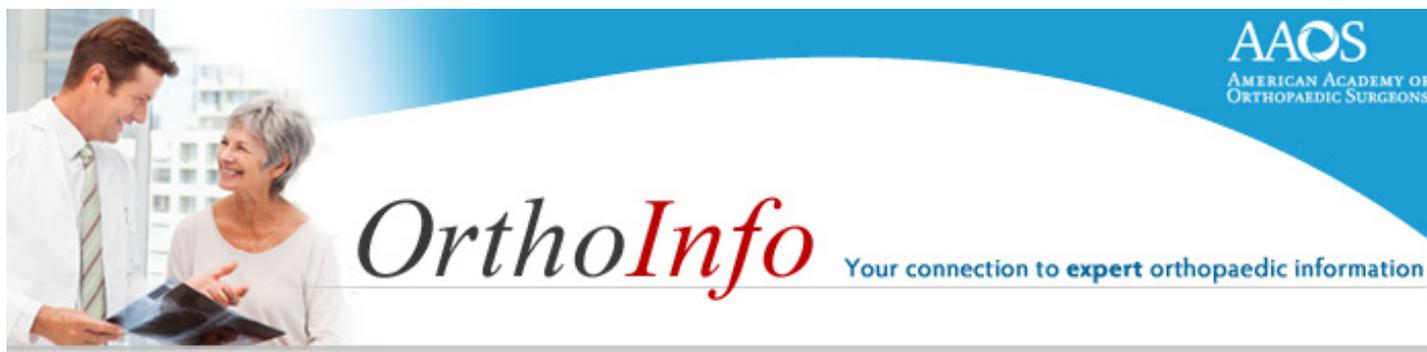
The Vicious Cycle of RSD



Prognosis

The condition may take several months to years to settle down. Approximately 75% will return to normal or near-normal. About 15% have moderate permanent disability and about 10% have severe permanent disability.

50% of patients with untreated symptoms lasting for more than 1 year will have profound residual impairment.



Complex Regional Pain Syndrome (Reflex Sympathetic Dystrophy)

This article is also available in Spanish: *Síndrome de Dolor Regional Complejo (Distrofia Simpática Refleja)* (topic.cfm?topic=A00500).

Complex regional pain syndrome (CRPS) is a condition of intense burning pain, stiffness, swelling, and discoloration that most often affects the hand. Arms, legs, and feet can also be affected by CRPS.

This condition was previously known as reflex sympathetic dystrophy, Sudeck's atrophy, shoulder-hand syndrome, or causalgia.

Description

There are two types of CRPS:

- Type 1 occurs after an illness or injury that did not directly damage a nerve in the affected area
- Type 2 follows a distinct nerve injury

Although the triggers vary, both types of CRPS have the same symptoms and go through the same three stages of disease.

Stage I: Acute

Stage I may last up to 3 months. Burning pain and increased sensitivity to touch are the most common early symptom of CRPS. This pain is different — more constant and longer lasting — than would be expected with a given injury. Swelling and joint stiffness usually follow, along with increased warmth and redness in the affected limb. There may be faster-than-normal nail and hair growth and excessive sweating.



Acute stage CRPS, 2 months after injury

Stage II: Dystrophic

Stage II can last 3 to 12 months. Swelling is more constant and skin wrinkles disappear. Skin temperature becomes cooler. Fingernails become brittle. Pain is more widespread, stiffness increases, and the affected area becomes more sensitive to touch.

Stage III: Atrophic

Stage III occurs after 1 year. The skin of the affected area becomes pale, dry, tightly stretched, and shiny. The area is stiff and there is less hope of getting motion back. Pain may decrease and the condition may spread to other areas of the body.

Cause

Although the two types of CRPS can be tied to injury or illness, the exact cause of CRPS is unknown. One theory is that a "short circuit" in the nervous system is responsible. This "short circuit" causes overactivity of the sympathetic (unconscious) nervous system which affects blood flow and sweat glands in the affected area.

Symptoms most commonly occur after injury or surgery. Other causes include pressure on a nerve, infection, cancer, neck problems, stroke, or heart attack.

Doctor Examination

After discussing your medical history and symptoms, your doctor will carefully examine your hand or affected limb.

People with CRPS are unusually protective of the involved limb. Even a light touch may evoke expressions of severe pain.

Tests

There is no single test that can make the diagnosis of CRPS. Some imaging studies, such as x-rays, bone scans, and magnetic resonance imaging (MRI) scans can help your doctor make a firm diagnosis.

Treatment

Early diagnosis and treatment are important in order to prevent CRPS from developing into the later stages.

It is also important that these patients not be told that the pain is "in their heads." CRPS is a physiological condition. Even though it is not fully understood, CRPS is treatable.



After 6 months of treatment, this patient's hands have regained normal color and are no longer swollen.

Nonsurgical Treatment

Medications. Non-steroidal anti-inflammatory drugs (NSAIDs), oral corticosteroids, anti-depressants, blood pressure medications, anti-convulsants, and opioid analgesics are medications recommended to relieve symptoms.

Injection therapy. Injecting an anesthetic (numbing medicine) near the affected sympathetic nerves can reduce symptoms. This is usually recommended early in the course of CRPS in order to avoid progression to the later stages.

Biofeedback. Increased body awareness and relaxation techniques may help with pain relief.

Therapy. Active exercise that emphasizes normal use of the affected limb is essential to permanent relief of this condition. Physical and/or occupational therapy are important in helping patients regain normal use patterns. Medications and other treatment options can reduce pain, allowing the patient to engage in active exercise.

Surgical Treatment

If nonsurgical treatment fails, there are surgical procedures that may help reduce symptoms.

Spinal cord stimulator. Tiny electrodes are implanted along your spine and deliver mild electric impulses to the affected nerves.

Pain pump implantation. A small device that delivers pain medication to the spinal cord is implanted near the abdomen.

Results from surgical procedures may be disappointing. Many patients with chronic CRPS symptoms benefit from psychological evaluation and counseling.

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Contributed and/or Updated by: Robert Brent Blake, MD

Peer-Reviewed by: Charles D. Jennings, MD; Colin F. Moseley, MD

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The American Academy of Orthopaedic Surgeons
9400 West Higgins Road
Rosemont, IL 60018
Phone: 847.823.7186
Email: orthoinfo@aaos.org