COMPLEX REGIONAL PAIN SYNDROME

Overview

Complex regional pain syndrome (CRPS), formerly known as Reflex Sympathetic Dystrophy (RSD), is an uncommon but often very disabling neuropathic condition that affects the extremities and less often other bodily structures. The key symptom of CRPS is continuous, intense pain out of proportion to the severity of the injury, which gets worse rather than better over time. CRPS most often affects an extremity (arm, leg, hand, foot). This condition was first described after the U.S. Civil War, when soldiers reported severe pain even after their wounds had healed. The condition was often referred to as "hot pain" during that time period.

Women are more likely to be affected than are men. This condition can affect persons of all ages, but the peak age range is between 20 to 35 years. The condition is characterized by aching or burning as well as swelling, skin discoloration, altered temperature, sweating, and hypersensitivity in the affected areas.

Although there is a tremendous amount of research ongoing, there is currently no cure for this condition.

Cause

The cause of CRPS is not truly known. Scientists throughout the world are actively working to identify factors which trigger the disease state. Many experts feel that the condition involves abnormal activity of the sympathetic nervous system, and others believe that there is a triggering of the immune response. Many cases of CRPS occur after a traumatic injury to an arm or a leg, such as a gunshot wound or shrapnel blast. However, the condition can also occur after a seemingly minor injury like an ankle sprain or localized infection.

Complex regional pain syndrome occurs in two types. Both have similar clinical appearance but different causes:

- <u>Type I</u> Previously known as Reflex Sympathetic Dystrophy, this condition occurs after an illness or injury that did not directly damage the nerves in the affected limb. Approximately 90% of people with CRPS are classified as Type I patients.
- <u>Type II</u> Previously referred to as "Causalgia", this condition usually follows a distinct nerve injury.

Signs & Symptoms

The main symptom of CRPS is intense pain, often described as "burning." Additional signs and symptoms can include:

- Skin hypersensitivity Simply touching the skin of an affected limb can produce significant pain.
- Changes in skin temperature, color and texture. At times the skin of an affected area can be sweaty and at other times it may be cold. Skin color can range from white and mottled to red or blue. Skin may become tender, thin or shiny in the affected area.
- Changes in hair and nail growth.
- Joint stiffness, swelling and damage.
- Muscle spasms, muscle weakness and loss of muscle mass or atrophy
- Decreased ability to move the affected body part.

CRPS typically has three stages. Not every patient experiences these phases at the same pace:

- <u>Stage 1</u> Severe pain develops in the affected limb. Swelling, sensitivity to touch or to cold, and skin changes, such as drying or thinning, begin to appear. This stage usually lasts one to three months.
- <u>Stage 2</u> Changes to the color and texture of skin become increasingly obvious, and the swelling spreads. One may experience muscle and joint stiffness. This stage may last three to six months.
- <u>Stage 3</u> Severe damage in the affected area is evident, including profound limitation in movement of the affected limb, irreversible skin damage, muscle atrophy and contractures in nearby digits.

Diagnosis

- **Medical History** The onset of CRPS symptoms can often be traced back to an accident, illness, or injury.
- **Physical Examination** An examination of the skin, muscles, and joints in the affected extremity may reveal information about the source of tenderness and pain. There may be changes in the normal texture and color of the skin, and one may have problems with limited range of motion in the joints of the affected extremity.
- **Bone Scan.** A radioactive substance injected into the circulation permits viewing of the bones with a special camera. This procedure may show increased circulation to the joints in the affected area.
- Sympathetic Nervous System Tests These tests look for disturbances in the sympathetic nervous system. <u>Thermography</u> measures the skin temperature and blood flow in an affected extremity. Other tests can measure the amount of sweat produced by both limbs. Different amounts of sweat produced by the affected and non-affected extremity can suggest the diagnosis of CRPS. However neither of these tests is a reliable diagnostic tool.

- **X-Rays** Loss of minerals from the bones in the affected area may show up on an X-ray in later stages of CRPS.
- **MRI** Magnetic Resonance Imaging may show a number of tissue changes, including skin thinning and muscle atrophy.
- Response To Sympathetic Nerve Blocks These injection procedures are specifically directed at numbing or blocking certain (sympathetic) nerves that may be involved in the cause of this condition. The diagnosis of CRPS is suggested in patients who note improvement or remission in their symptoms following a series of sympathetic nerve blocks. However, lack of response to these nerve blocks does not entirely rule out the diagnosis of CRPS.

Treatment

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. Numerous support groups, both locally and nationally, are available for patients with CRPS and can be found through a simple Google search for "CRPS Support or RSD Support."
- **Sympathetic Nerve Blocks:** These procedures are often effective in reducing pain and facilitating other treat measures, including physical and occupational therapy and medications. The type of nerve block is determined by the location of the pain. For upper extremity CRPS symptoms, a *Stellate Ganglion Block* is performed. For lower extremity CRPS symptoms, a *Lumbar Sympathetic Block* is performed. For more details please refer to the "Treatments" section of this website (coming soon).
- **Medications:** Many different classes of medication are used to treat CRPS, including topical analysis 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.
- **Hyperbaric Oxygen Therapy:** There are some recent studies indicating that this treatment is effective in decreasing pain and swelling and improving range of motion for some CRPS patients. More studies are necessary.
- **Surgical Sympathectomy:** The use of surgical sympathectomy, a technique that destroys the sympathetic 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 electrodes into the epidural space of the spine stimulates a portion of the spinal cord (dorsal column) which changes the sharp or burning pain into more of a pleasant tingling sensation in the painful area. This technique is often very helpful in reducing pain as well as disease progression and restoring function. For more details please refer to the "Treatments" section of this website.
- Intrathecal Drug Delivery: These devices are programmable computerized pumps that are implanted into a person's body to administer medications like morphine, local anesthetics, clonidine, and other medications directly into the spinal fluid. The medications bind to special receptors on the spinal cord to exert their pain relieving effects. Intrathecally delivered medications are infused in doses far lower than those used for oral or intravenous use. Therefore side effects are limited and the drugs are much more effective. This treatment is often reserved for patients who have failed to respond to other more conservative therapies, including all those listed above.

<u>Links For More Information</u> - For more information on CRPS, please refer to some of the following websites:

- eMedicine:
- National Institute for Neurologic Disorders and Stroke:
- National Pain Foundation
- WebMD: