Chronic Regional Pain Syndrome

The Facts with a Patient's Perspective©

Mary Sutton, CST, CFA

Introduction

Chronic regional pain syndrome (CRPS), previously referred to as reflex sympathetic dystrophy (RSD), affects millions across the United States. This syndrome is often misunderstood and so most people do not know they have it. Weir Mitchell, MD, first described RSD during the Civil War. He found that certain veterans had "hot pain" after their primary injury healed. This happened most often with a wound involving a nerve injury. He called his syndrome "causalgia."

The Reflex Sympathetic Dystrophy Syndrome Association reports that CRPS is more common in women, which accounts for 75% of its patients. Sixty-five percent of the patients are in their 30s and 40s, although CRPS has been diagnosed in children and adolescents. In 30% of the cases, no precipitating cause is identified. CRPS has been ranked 42 out of 50 on the McGill pain index, making it one of the most painful chronic syndromes that exists. Researchers have found that CRPS occurs following: 1-2% of various fractures, 2-5% of peripheral nerve injuries, and 7-35% of Colles fractures. Unfortunately, for most patients, a diagnosis is not made early. Very mild cases may even resolve without treatment, while others progress until the syndrome becomes chronic and debilitating. Casting or immobilization can worsen the symptoms. CRPS can go into remission then come back after a further injury.

Types of CRPS

There are two types of chronic regional pain syndrome. Type I includes cases where there is no known nerve injury (this type is also referred to as RSD), and cases where the injury does not follow normal healing paths. The sympathetic nervous system (SNS) assumes an abnormal function after the initial injury. Even a minor injury, such as a sliver or an IV placement, can cause CRPS. Type II is also known as causalgia, where there is a distinct, major nerve injury. Again, the SNS assumes an abnormal function.

Symptoms

Symptoms of CRPS usually occur near the site of the injury. They include burning pain, muscle spasms, localized swelling, changes in skin temperature (which can be cold or warm), increased sweating, softening of bones, joint tenderness/stiffness, restricted and/or painful movement, and changes in the nails and/or skin which appear shiny red and later become bluish. The patient may have weakness and skin rashes in the extremity. There are often movement disorders with decreased muscle tone, tremor, difficulty starting a movement, and/or increased reflexes. The patient's pain is out of proportion to the injury and worsens in time. The patient often refers to the pain as burning, aching, or searing. The pain is continuous and increases with emotional distress and/or stress. Movement and being touched become intolerable. The patient's joints become stiff from disuse. The skin, muscle, and bones can atrophy. The patient may have extreme sensitivity to touch, which is called allodynia. There is also an increased perception of the pain called hyperalgesia. Misdiagnosis occurs because the symptoms are so individualized.

Some patients can improve without treatment. The greatest opportunity for recovery is with early recognition and treatment. CRPS is diagnosed primarily by observation of the symptoms.

Diagnosis

The criteria for diagnosis of CRPS Type I includes the presence of initial injury or cause for immobilization of the limb. The patient experiences continuing pain, allodynia, or hyperalgesia with pain disproportionate to the injury, if known. The extremity is edematous, with changes in the skin blood flow that cause both color and temperature variations. There is often abnormal or subnormal motor activity in the region of the pain.

The criteria for diagnosis of CRPS Type II is the presence of continuing pain, allodynia, or hyperalgesia post nerve injury, which is not necessarily limited to the distribution of the nerve.

There are three stages of CRPS (Table 1). Each has definite features, but patients often present with symptoms of different stages at the same time.

Typically, a diagnosis of CRPS is based on four common characteristics: intense prolonged pain, vasomotor disturbances, delayed recovery of function, and various trophic changes. Pain is the clinical feature that is considered the hallmark of CRPS and is the problem that prompts the patient to seek treatment. The pain may cause mobility problems disproportionate to the injury itself. All tactile stimulation to the skin is painful (hyperesthesia). There is diffuse tenderness throughout the affected limb. Myofascial pain is caused by tender spots within the muscle due to the spasms. There can also be spontaneous "jabs" of pain.

As described previously, the skin is significantly affected by CRPS. The skin becomes dry, shiny, and at times scaly. Nails grow faster, then the growth slows, and nails become cracked. Rashes, pustules, and skin ulcers occur as CRPS progresses. The skin can be either warm or cold to touch. Often, the skin temperature reacts to the temperature of the room. The color of the skin can change from white mottled to red to blue.

Swelling is usually diffuse and localized to the painful region. Joints are often swollen and stiff. The extremity itself is difficult and painful to move. Decreased mobilization leads to more muscle atrophy than the syndrome alone causes. Muscle spasms, which can be severe, become debilitating.

As CRPS progresses, pain and symptoms become increasingly diffuse and, in approximately 70% of cases, spread to another limb. There are three types of spread. Continuity spread is to the upper or lower limb on the same side. Mirror image is spread to the limb on the opposite side. Independent spread is progression to a separate, distant region of body, which can be secondary to trauma in that area.

In some cases, there can be wasting of bones and osteoporosis. This is because CRPS can leech calcium from the bone and muscles. The duration of pain and the other symptoms of CRPS vary. A patient can have periods of remission of all symptoms and then exacerbation of those symptoms.

As trauma to the extremity has been discussed as an etiological factor, whether major (surgery/fracture) or minor (sliver, IV) is irrelevant. There are several other factors that should be discussed.

CRPS has occurred in individuals with ischemic heart disease or patients having myocardial infarctions, cervical spine or spinal cord disorders, cerebral lesions, infections to the extremity, and repetitive motion disorders. As discussed, there are also patients with no known etiological event.

In order to understand the physiological cause of CRPS, one must understand the pain reflex. When there is an injury, a pain impulse is sent via sensory nerves to the central nervous system. The impulse triggers a reaction in the sympathetic nervous system (SNS), which travels, back to the site of the injury. At the site of injury, the SNS triggers an inflammatory response, causing vessels to spasm and resulting in swelling and pain. The pain then triggers another impulse to the brain which cycles over and over again. In a normal situation, the SNS stops functioning after a few minutes. In CRPS, the SNS assumes an abnormal function and the cycle continues. The vessels spasm, restricting blood to the muscles, which causes the muscles to spasm. Also, with the vessel spasms, there is the white skin response. Because of the redundant cycle, the long-term effects begin to show.

Unfortunately, there is no proven diagnostic lab test for CRPS; however, there are several studies that may suggest the disorder. Thermograms measure the heat emission of a limb. Usually there is an abnormal change in skin temperature of a CRPS patient, but a normal thermogram cannot rule it out. An abnormal test can be helpful evidence for judicial or insurance inquiries. A radionucleotide (three-phase) bone scan is a nuclear medicine scan, which studies the changes in bone structure. Some physicians use the scan as a tool to diagnosis CRPS. However, studies have found that the results cannot be validated as a definitive diagnostic tool. Doppler studies may also be useful in comparing asymmetrical bilateral blood flow.

TABLE 1 **Stages of CRPS**

Stage I	*Time elapsed from precipitating incident—one week to three months*
	• Severe burning pain at the site of injury • Muscle spasm, joint stiffness • Restricted mobility • Rapid hair and nail growth • Vasospasms that affect the color and temperature of skin
Stage II	*Time elapsed from precipitating incident—three to six months*
	• Pain intensifies and becomes more diffuse • Swelling spreads • Hair growth diminishes • Nails become cracked and grooved • Diffuse osteoporosis • Joints thicken • Muscles begin to atrophy
Stage III	*Time elapsed from precipitating incident—until resolution or indefinitely*
	• Changes in skin and bone become irreversible • Intractable pain • Marked muscle atrophy • Mobility severely limited • Flexor tendon contraction • Occasional limb displacement • Marked bone softening

Treatment

The CRPS patient needs the resources of many modalities for treatment. Pain centers throughout the country involve the services of occupational therapists, physical therapists, psychologists, and pain physicians. The patient needs to feel a part of the team and be involved in all aspects of their care. The most important part of the treatment is patient education. He or she must understand CRPS and the treatment options available.

Most patients need to know that someone understands their pain. The physicians involved must be knowledgeable about CRPS, as it is very individualized. A written record, such as a journal, may be helpful to review the patient's progress. This protocol could include the procedures, medications, physical/occupational therapy, psychosocial issues, and tests, including results.

During this process, the patient should set reasonable goals for outcomes. The patient has to learn to overcome the natural tendency to maintain their disability. A pain psychologist is critical for optimal rehabilitation. The psychologist helps the patient improve his or her coping skills using relaxation techniques. Biofeedback

or self-hypnosis may be used in this process. The pain psychologist must look at the psychosocial issues in the patient's life. They must evaluate the patient's pain-coping skills, as well as their drug-abuse potential. As stress is a known cause for exacerbation of pain, the patient's potential for committing suicide must also be considered.

Maintaining normal mobility of the affected limb is very important. The patient must avoid the automatic reflex to guard the painful extremity and not to use it. There are many modalities used to keep a patient mobile. Oral or transdermal medications may be prescribed. Transcutaneous electrical nerve stimulation (TENS unit) can be used to stimulate the muscles to keep them moving.

Physical therapy is the most important tool used to maintain mobility, and the therapist educates the patient on how to use the extremity. The "no pain, no gain" approach to physical therapy cannot be used, however, as that could cause the CRPS to worsen. The physical therapist must work with the patient to stretch and strengthen the affected limb, as well as work within the patient's pain cycle to avoid further damage. Different modalities—such as ultrasound, hydrotherapy, and massage—can be used to help treat the muscle spasms and weakness.

Occupational therapists are also used to help the patient use the affected limb normally in their work. Modalities such as desensitization are used to help the patient relieve some of the sensation of pain due to light touch. Occupational therapists work on fine motor skills to help the patient deal with everyday life. Just as the physical therapists are important for the continued use of the limb, the occupational therapists are also important in keeping the patient working and using that limb.

Sequential drug trials may be used as treatment. Medications are often "off-labeled," meaning that CRPS is not their primary modality. The dosages are gradually increased to determine the optimal dose for treatment. Different sequences of drugs may be used to create the desired effect. Patients should familiarize themselves with the possible side effects of each of the medications.

Medications are often prescribed according to various symptoms. Nonsteroidal anti-inflammatory drugs (NSAIDs, such as aspirin, ibuprofen, and naproxen) are given for constant pain with inflammation. In fact, certain studies have shown that CRPS may be alleviated within the first six months by NSAIDs alone. Tramadol, a central nervous system agent, may be prescribed for constant pain without inflammation. Constant pain and sleep disturbances are often treated with antidepressants, such as amitriptyline, doxepin, nortriptyline, or oral lidocaine (mexiletine), which is experimental. Paroxysmal pain (spontaneous jabbing pain) is treated with anticonvulsants, such as gabapentin or carbamazepine. For severe pain that is unresponsive to the previous drugs, an oral opioid (eg, morphine, codeine, etc.) may be prescribed.

Some studies suggest that a low dose of methadone to bind to a nonopiate receptor in the spinal cord, thereby reducing pain perception, may help CRPS patients. Any time a narcotic is used, a potential addiction hazard exists. A narcotics contract may be drawn up and signed. For muscle spasms, a variety of muscle relaxants may be used, such as cyclobenzaprine (Flexeril®), clonazepam (Klonopin®), tizanidine (Zanaflex®), or baclofen. A clonidine patch may be useful for SMP treatment. For localized pain due to nerve injury, capsaicin cream may be applied, although its effectiveness for CRPS patients has not been determined.

Sympathetic nerve blocks both diagnose and treat CRPS. A series of nerve blocks treat patients with CRPS that have sympathetic mediated pain (SMP). If the blocks prove effective, it is presumed that the patient was suffering CRPS. Other patients have sympathetic independent pain (SIP), which means their pain is due to something other than the abnormal function of the pain reflex, and the nerve block treatment is ineffective. SIP patients often are harder to treat, as it is more difficult to find the cause of the continued pain. Because of the ability to distinguish between the types of pain, as well as being a treatment protocol for SMP patients, sympathetic blocks are the most credible diagnostic tools for CRPS. With the type of pain established, the treatment process continues, if necessary.

Sympathetic blocks are often considered good treatment options. There are three reasons to

consider a sympathetic block: permanent cure or partial remission of the CRPS, diagnostic information, and prognostic information for further treatments. A good sympathetic block provides increased temperature without increased numbness. The doctor should record the amount of pain relief and any improvement in the range of motion. The goal of sympathetic blocks is to treat but not to over treat. A series of multiple sympathetic blocks separated by brief intervals (eg one week) may be given to determine if these blocks are an effective treatment protocol.

The duration of pain relief and improvement due to a sympathetic block must be closely monitored by the patient and the physician. Patients with sympathetic mediated pain usually experience pain relief that far outlasts the local anesthetic duration (usually just a few hours). The extended pain relief and improved movement of the limb can last from days to months or put the affected limb into remission.

Some patients may not reliably report the effects of the blocks. One of the characteristics of a good sympathetic block is a feeling of warmth. Some patients may mistake this feeling as a relief or even a genuine perceived reduction of pain. Some patients deceitfully report pain relief as a means to get additional treatment and treatment options. Other patients feel that some kind of treatment is better than no treatment at all. The pain physician must be aware of this kind of behavior in order to act accordingly and in the best interests of the patient.

There are several types of sympathetic blocks: Stellate ganglion blocks are used to treat CRPS

THIS IS MY STORY

In February 1997, one year and one week after carpal tunnel surgery, I experienced severe shoulder pain. I also noticed that my hand was cold and swollen. I went to my orthopedic surgeon, who told me that I had a cold in my shoulder and gave me some NSAIDs. I returned several weeks later with slightly different shoulder symptoms, but the same hand symptoms. After seeing him for eight months, he basically told me that he did not know what to do with me. His tone was that of disbelief. I was so discouraged!

On the advice of a friend, I spoke to one of our anesthesiologists who dealt with pain. We went to his office and he placed temperature strips on my hands. My left hand was 85 degrees; my right was 80 degrees. He told me the difference was normal for this syndrome, and gave me my first stellate ganglion block a week later.

I have had numerous stellate ganglion blocks, as well as three stellate ganglion RFLs. In October 1998, I had an endoscopic thoracic sympathectomy and was in remission for a year and six weeks. Since that time, I have had a couple of falls in the operating room. The first resulted in a pulled hamstring. The CRPS spread down that muscle only. The second resulted in a sprained ankle. Several months later, the CRPS spread down the rest of my leg.

My doctor tried to treat both areas by performing a thoracic sympathetic block at T-2 and T-3. I had several of them, as well as a thoracic sympathetic RFL. These were not successful, so I now receive a lumbar sympathetic block for my leg and a stellate block for my arm.

I currently take a number of medications to help control my RSD. Despite blocks and muscle relaxants, muscle spasms occur. I am still in pain, but can handle things better when I am mobile and taking medication. I have been through occupational therapy, as well as years of physical therapy, and will have many more years to come. I find it easier to keep my mobility by going to physical therapy. I am often chasing problems, such as tendonitis or limited movement with my arm, especially due to the muscle atrophy caused by my CRPS.

One thing that helped considerably was seeing a pain psychologist. I learned to focus away from my pain and deal with stress to prevent additional pain. Music also helps me deal with my pain. Listening to music, reading, and working helps me through the day without having a major perception of my pain. I have continued to work a full-time job with call, as well as one to two different part-time jobs. In addition, I have served on the AST Board of Directors. In fact, with my doctor's permission, I went to a Board meeting not even two weeks post sympathectomy.

Where do I go from here I continue to keep doing what I am doing. I try to keep my muscles stretched and as strong possible. I work at a surgicenter, as well as take first assisting call for vitreoretinal surgeons. I keep myself educated on current CRPS trends and do what I can to educate others about this chronic pain syndrome. Hopefully, this education will benefit both surgical technologists and their future patients. Most importantly, I never give in to what I call "my beast." I cannot let it win.

of the upper extremity. The stellate ganglion is located in the chest but the actual block is done at the level of C6-7 along the trachea. The patient is placed in the sitting position after injection to allow the anesthetic to settle on the stellate ganglion. Other possible features of a stellate ganglion block include Horner's syndrome, which is characterized by the flushing of the cheek and drooping of the upper eyelid on the side of the block. Because the block is done along the trachea, the recurrent laryngeal nerve may be affected for a short period of time. The patient experiences hoarseness and trouble eating and drinking. The block may cause numbness along the thumb side of the arm due to the C6 and C7 motor nerves. All these features resolve in a short period of time.

Lumbar sympathetic blocks are given for CRPS in the lower extremity. A C-arm is used to facilitate placement of the block needles. A catheter may be placed for a more specific block than an epidural block. A problem with the catheters is they are easily dislodged.

If a sympathetic block has been successful for a patient, the pain management doctor may try a radio frequency lesioning (RFL) of that area. The goal of RFL is to put the patient into total remission or at least to initiate a longer period of pain relief. The physician must verify, via fluoroscopy with contrast, that the RFL needle is in the correct position to ensure that no structures, other than the sympathetic root, are burned.

Epidural blocks can be used but are not very helpful for diagnosis or treatment of CRPS. Epidural blocks are less specific to the sympathetic nervous system. Most often, they are used to infiltrate steroids. The patient may experience temporary weakness in the legs, making walking difficult following an epidural block. Some doctors try to treat pain by placing a long-term epidural catheter. This has been found to be expensive with a greater risk of life-threatening conditions such as meningitis. Dislodgement of the catheter is also a problem. An epidural block with catheter placement necessitates a hospital stay of at least two days.

Chemical sympathetic blocks can be used as a diagnostic test for SMP. An intravenous sympathetic blocker, phentolamine, may be used to see if the pain is stopped. There is a 43% false negative rate, which makes this procedure rarely used. But it may be a valuable treatment in a situation where a block is not possible or when multiple extremities are involved. Phenol, however, is still used for sympathetic neurolysis in the lumbar region with somewhat good results.

Another technique is the injection of blocking agents into an extremity and limiting spread of the agent by a tourniquet. This procedure relies upon the ability to start an IV in a swollen extremity. There is no evidence that this technique is more effective than the sympathetic block, but it is an option for patients on anticoagulant therapy.

Spinal cord stimulators (SCS) have been used in the treatment of CRPS. They work well in patients with chronic intractable pain due to CRPS. The SCS provides low-intensity electrical impulses to trigger nerve fibers along the dorsal column of the spinal cord. The SCS is believed to stop pain messages to the brain. It replaces the pain with tingling. A temporary trial should be performed before the permanent stimulator placement. The stimulator should focus on the most painful region, which can be difficult for CRPS patients, whose most painful regions constantly change. It is an invasive, costly procedure that may not be covered by insurance. Potential complications of SCS are spinal infection and paralysis, but these are very rare.

CRPS may be treated with morphine pump placement. The pump would produce a single injection of morphine into the spinal fluid. Selective pain blocking occurs. It is also an invasive, costly procedure. There is no evidence of an advantage over oral morphine, though the pump does spare the patient from the side effects of taking oral morphine. Potential complications include tolerance, nausea, constipation, weight gain, decreased libido, edema in legs, and increased sweating. Pumps with baclofen may be used for patients with dystonia.

Another controversial procedure is sympathectomy. Kotzareff described the first open sympathectomy in 1920 for hyperhidrosis

(abnormal sweating of the palms). An endoscopic thoracic sympathectomy was reported by Kux 50 years ago. It became widely accepted in the 1990s. Endoscopic, retroperitoneal, lumbar sympathectomy was first reported in 1993 with only preliminary results recorded at this time.

A sympathectomy disrupts the autonomic nervous system, resulting in interruption of pain pathways. It is used on patients with significant decrease in pain following sympathetic blocks. Only patients with SMP should be considered, as sympathetectomy is a very invasive procedure with risks. The procedure removes a section of sympathetic nerves located near the spinal cord. For CRPS in upper extremities, a sympathetectomy should be performed between T1 to T6 or T7. For lower limbs, it should be performed between L1 to L4. Sympathetectomy should be used for relief of CRPS, restoration of normal blood flow to the arms, and hyperhidrosis.

Complications of sympathectomy include bleeding, infection, and pneumothorax. Some patients experience compensatory sweating or gustatory sweating. Horner's syndrome is a common problem, due to the cutting of the sympathetic chain. In many cases, there has been sympathetic regeneration of the area that was cut.

Prognosis

With such diverse treatment options and individualization of the disease, there is no doubt that the CRPS patient has an uphill battle. There are many problems and effects of CRPS for the patient. Extreme pain, which is often a lifelong situation, is a way of every day life. This causes extreme family disruption and sorrow from family members, who are helpless to provide answers and pain relief. Often the CRPS patient is disabled or unemployed, which causes financial difficulties. Patients go through multiple misdiagnoses and strife from disbelieving health care professionals. Due to the misdiagnoses, the patient undergoes the improper treatment for CRPS. They have multiple surgeries, some of which will be unsuccessful. The quality of life they had is lost. Their health care costs are increased, and treatments may not be covered by insurance.

As the number of pain procedures being performed in the operating room increases, it is up to these professionals to familiarize themselves with chronic pain syndromes and how to take care of patients. Caregivers have to realize these patients are angry, frustrated, and in great pain. Ask the patient where to place EKG pads, what position causes them pain, and how to best touch them so they do not hurt. This allows the patient to relax in a stressful environment. Often these patients will have multiple visits to the operating room. Knowing their caregivers will treat them with respect and have an understanding of their pain syndrome goes a long way in alleviating preoperative stress.

About the author

Mary Sutton, CST, CFA, is currently an instructor at Concorde Career Institute in Jacksonville, FL. When she has time, she is still a surgical first assistant for vitreoretinal physicians. She was certified as a CST in 1984 and as a CFA in 1994. Sutton has been a member of AST since 1984 and served as vice president of the Florida State Assembly from 1999-2003. She served on the AST's national Board of Directors from 1996 to 2000 and currently serves on the LCC-ST Board of Directors.

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