NCPD 2.0 ANCC Contact Hours

Complex Regional Pain Syndrome
The Importance of Early Diagnosis

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Complex regional pain syndrome (CRPS) is a potentially life-altering, neuropathic pain condition that often affects an extremity in response to surgery or even minor trauma. The pain is often disproportionate to the injury sustained or the surgical procedure performed—and it has been described as excruciating and unbearable. CRPS can result in irreversible, disabling changes in the affected limb. In some cases it has resulted in amputation or severe psychological issues. However, CRPS outcomes are generally more successful with early detection and treatment. This article presents an overview of CRPS and discusses the critical role nurses can play in recognizing the condition early, assisting with treatment, and providing psychological support.

Many chronic pain conditions affect a patient's extremities, and among the most agonizing is complex regional pain syndrome (CRPS), a neurologic disorder that often follows a traumatic event. CRPS is characterized by autonomic and inflammatory features, with the patient experiencing pain that is worse in magnitude and/or duration than would be typically expected from the inciting incident. This incident is most commonly a fracture, though it can also be a surgical procedure or a trivial injury, such as a contusion, ankle sprain, an injection, or intravenous infusion. Or its cause may be unknown (de Mos et al., 2007; Goh et al., 2017; Shim et al., 2019; Weber & Mackinnon, 2010).

Nurses who are charged with the care of patients, including children, with musculoskeletal extremity issues, such as trauma or such issues after surgery, should be familiar with CRPS. These medical professionals may need to assist in the treatment of patients diagnosed with CRPS, or more importantly, they may be the first to identify a patient who may have this affliction. The prompt recognition of CRPS is essential, as early initiation of treatment has been shown to improve the patient's outcome (Breivik & Stubhaug, 2016; Grieve et al., 2019). Delays in diagnosing CRPS can lead to intractable pain, disuse of the limb, the spread of CRPS to other limbs, and psychological consequences. Known as "the suicide disease," CRPS can be so debilitating that some sufferers choose to take their own life. Other patients with resistant CRPS have opted to have the painful limb amputated, though outcomes for these people are still unclear (Ayyaswamy et al., 2019; Goebel et al., 2018; Henderson, 2019; Lee et al., 2014; van Rijn et al., 2011).

Definition and Pathophysiology

Although the first documented instance of CRPS occurred in the 16th century when King Charles IX developed CRPS after a bloodletting procedure, a comprehensive description of the clinical signs and symptoms was not published until the American Civil War. Silas Weir Mitchell, MD, was a young U.S. Army contract physician who treated soldiers with gunshot wounds. He observed that after the wounds of some injured soldiers had healed, they continued to experience severe pain. The disorder was termed "causalgia" from the Greek word for fever (*kausos*) and pain (*algia*) (Iolascon et al., 2015; Mansano & Trescot, 2016).

Since Mitchell's discovery in the late 1860s, CRPS has also been referred to as Sudeck's dystrophy, minor causalgia, algodystrophy, and reflex sympathetic dystrophy (Ratti et al., 2015). In 1994, the definitive nomenclature—complex regional pain syndrome—was ratified by the International Association for the Study of Pain. The term "CRPS" includes two subclassifications: CRPS type 1, where nerve damage is not identified; and type 2, where nerve damage is identified.

Although the pathophysiology of CRPS is not fully understood, the available literature suggests that even with CRPS 1, an initial nerve injury in an extremity—even one of minimal severity—can be a trigger that results in the development of CRPS (Bruehl & Warner, 2010; Mansano & Trescot, 2016; Oaklander et al., 2006). At the time of the injury, a combination of abnormal mechanisms occur simultaneously involving the peripheral and the central nervous system, causing distinct aberrant neuroplasticity and an exaggerated immune and inflammatory response, combined with autonomic

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Table 1. Possible Differential Diagnoses of Complex Regional Pain Syndrome

Neurological	*Musculoskeletal*
Neuropathy	Strains/sprain
Nerve compression	Osteoarthritis
Radiculopathy	Fracture
Spinal tumor	Compartment syndrome
Inflammatory	*Vascular*
Infection	Raynaud's disease
Seronegative arthritis	Thrombosis
Rheumatologic	Erythromelalgia
Erysipelas	Traumatic vasospasm
Connective tissue disorder	*Psychiatric*
Lupus	Somatoform pain disorder
Scleroderma	Munchhausen syndrome

dysfunction (Baronio et al., 2020; Cutts et al., 2020; Shim et al., 2019).

Diagnosis

No test exists to determine the onset of CRPS; as a result, the diagnosis is clinical and is made by exclusion. As such, it is important to rule out problems that may appear to be CRPS (see Table 1) (Goebel et al., 2019; van Eijis et al., 2011). The criteria for diagnosing CRPS have evolved, and currently the diagnosis is based on the Budapest criteria (see Table 2), which have a high sensitivity for CRPS (Harden et al., 2010· Shim et al., 2019· Urits et al., 2018). Significantly, one must consider the fact that the signs and symptoms of CRPS vary over time (Goh et al., 2017).

History

Of those patients who have been diagnosed with CRPS, their medical history usually includes extremity-sustained trauma. This may be in the form of surgery or perhaps a seemingly trivial injury. The pain of these patients is frequently disproportionally greater than would be expected. Moreover, this pain extends beyond the area of the injury or surgery. Research suggests that patients suffering from posttraumatic stress disorder have an increased incidence of CRPS. Further, there may be an association of CRPS with pain catastrophizing, anxiety, and depression (Im et al., 2021; Lohnberg & Altmaier, 2013; Shim et al., 2019).

Nurses and other healthcare personnel should be astute when a patient complains of the symptoms listed in the Budapest criteria (see Table 2). Allodynia is pain from a stimulus that is not normally painful, such as the patient's inability to wear a shoe or even a sock. Hyperalgesia is an exaggerated pain response, such as the patient complaining of severe pain after a minor procedure. In addition, allodynia and hyperalgesia can both be present (He & Kim, 2020; Shim et al., 2019; van Eijs et al., 2011). A patient suffering from this condition often states that the pain is constant and not alleviated by any treatment (e.g., pain medication, ice, and other palliative protocols). These patients often are afraid to move the extremity, relate that the skin feels warm or cold, and that the pain extends beyond the area of the injury. For example, the patient may relate his entire foot hurts, though he only injured one toe.

Table 2. Budapest Criteria to Diagnose Complex Regional Pain Syndrome

The patient must meet all of the following criteria (A to D):

A. Pain that is disproportionate to the inciting incident

B. At least one symptom in three of the following four categories:
   1. Sensory: hyperalgesia and/or allodynia
   2. Vasomotor: temperature asymmetry and/or skin color changes
   3. Sudomotor: edema and/or changes in sweating
   4. Motor/trophic: decreased range of motion, motor abnormality (weakness, dystonia, tremor), and/or trophic changes (skin, hair, nails)

C. At least one sign present during the examination in two of the following categories:
   1. Sensory: hyperalgesia (to pinprick) and/or allodynia (to light touch, deep somatic pressure, or joint movement)
   2. Vasomotor: temperature asymmetry and/or skin color changes or skin color asymmetry
   3. Sudomotor: edema and/or changes in sweating
   4. Motor/trophic: decreased range of motion, motor abnormality (weakness, dystonia, tremor), and/or trophic changes (skin, hair, nails)

D. There is no other diagnosis that can better explain the patient's signs and symptoms

Physical Examination

When nurses and other healthcare personnel suspect CRPS, the Budapest criterion is the recommended guide for further diagnosis and confirmation of the malady. The presentation of CRPS varies, and different phases of the disorder have been described and observed. Initially, the extremity often shows the common inflammatory signs: warmth, pain, redness, and swelling. After 6 months, the extremity becomes cold, and there is more likely to be atrophy of the muscle, subcutaneous tissues, and the skin. In addition, the skin may appear dusky or shiny, and it may be beneficial to compare the skin with that of the contralateral extremity (see Figure 1). Allodynia can be evaluated by lightly brushing a cotton swab against the patient's skin, which typically should not elicit pain. Hyperalgesia can be tested for by a gentle pinprick (He & Kim, 2020; Kabani & Brassard, 2014; Misidou & Papagoras, 2019; Shim et al., 2019; van Eijs et al., 2011).

Testing

To reiterate, no specific test confirms a CRPS diagnosis; however, testing can be beneficial in excluding other problems. Laboratory tests, including C-reactive protein and the erythrocyte sedimentation rate, are normal in CRPS, but help in differentiating other diseases. Radiographs and MRI can help rule out other problems, and late stages of CRPS may show reduced bone density in the affected extremity. Nonetheless, such a finding is not conclusive for CRPS (Cutts et al., 2020; van Eijs et al., 2011). A positive finding on bone scintigraphy cannot be relied upon to diagnose CRPS type 1; however, a negative bone scintigraphy test reduces the likelihood of the presence of CRPS 1 (Wertli et al., 2017).

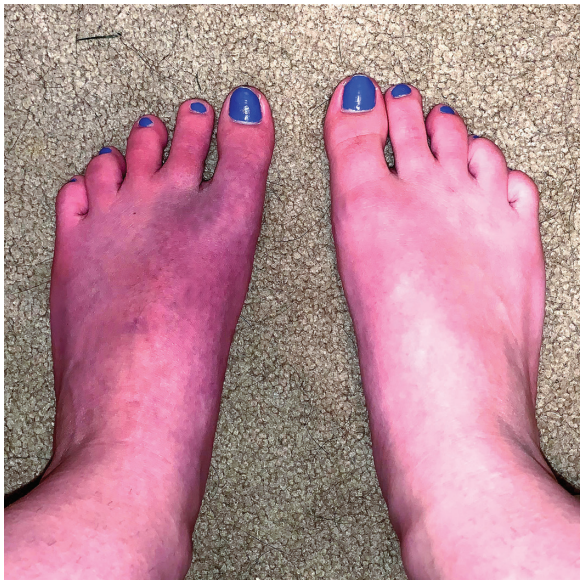

FIGURE 1. A comparison of the feet of a 17-year-old who has complex regional pain syndrome in the left lower extremity and no problems on the right. The color version of this figure is available in the online issue at https://journals.lww.com/orthopaedicnursing.

Epidemiology

A population-based study in the United States found that CRPS occurred in 5.46 persons per 100,000, with a predilection to affect women four times more than men. The median age of onset was 46 years (Sandroni et al., 2003). Population-based research in the Netherlands found CRPS occurred in 26 persons per 100,000, with women affected 3.4 times more than men. The median age in that study was 52.7 years (de Mos et al., 2007). Research in Scotland found the risk of CRPS in children ages 5–15 years was 1.2 per 100,000 (Abu-Arafeh & Abu-Arafeh, 2016).

Both the U.S. and Netherlands adult population studies found that the inciting incident was most often a fracture, accounting for 46% of the U.S. cases and 44% of the Netherlands cases. The U.S. researchers found that CRPS resolved—often spontaneously—74% of the time (de Mos et al., 2007; Sandroni et al., 2003). A Danish study revealed similar findings as those mentioned, and also noted that CRPS occurs three times more often after surgery than after nonsurgical treatment (Petersen et al., 2018). Postsurgical CRPS has been shown to have a risk of 2%–5% after carpal tunnel surgery, a 22%–39% risk after distal radius repair surgery, and a 4.36% risk after foot or ankle surgery (Li et al., 2010; Rewhorn et al., 2014). CRPS risk also increases dramatically when an extremity is immobilized, such as when a limb is placed in a cast (Pepper et al., 2013).

Treatment

Most important in the treatment of the CRPS patient is the urgent, early initiation of active range-of-motion exercises of the affected limb often through physical therapy. Pharmacological treatment generally also starts early, often with gabapentin, an anticonvulsant that has shown to be mildly effective (Goh et al., 2017; Van de Vusse et al., 2004).

Typically, management of CRPS is done in conjunction with (or direct referral to) a pain management specialist and sometimes with the concomitant help of a psychological expert. Other treatments include behavior therapy, the use of bisphosphonates, antioxidants (typically vitamin C), corticosteroids, and nonsteroidal anti-inflammatories, benzodiazepines, along with botulinum toxin and/or opioids. Interventional treatments include transcutaneous electrical nerve stimulation, intravenous regional blockade, peripheral sympathetic blocks, spinal cord stimulation, neurectomy, and nerve decompression (Cutts et al., 2020; Duong et al., 2018; Goebel et al., 2019; Poppler & Mackinnon, 2019). In severe cases, amputation has been done, though some authors consider this controversial as research on outcomes is limited (Ayyaswamy et al., 2019; Schrier et al., 2018, 2019).

Nursing Implications

Nurses, especially orthopaedic nurses, have the potential to recognize CRPS in their patients. Given the importance of early diagnosis of CRPS, nurses—especially orthopaedic nurses—are in a position to recognize CRPS in their patients even before other healthcare practitioners and in time to accord the patient a successful outcome. As examples, a routine bandage change, removal of sutures, or injury triage on a patient is a valuable opportunity to recognize the presence of CRPS. As such, it is essential for nurses to keep the possibility of CRPS in mind, even though they may not be directly charged with diagnosing CRPS. Nurses should not feel compelled to determine whether the patient has CRPS type 1 or 2, as this can be done in conjunction with other experts after the initial diagnosis is made.

Nurses who care for patients with CRPS should have a thorough understanding of the disorder to assist with their treatment and to appreciate the fact that these patients may be dealing with some of the most unbearable pain imaginable. Importantly, these patients should not be inadvertently mistaken for malingers or "drug seekers," though such patients are possible. With that said, nurses should empathize with the patient and focus on helping him or her accept their situation and continue to work with all concerned healthcare providers to ensure the best outcome.

Summary

Nurses should be alert to identify the possibility of CRPS in their patients, and in such situations partner with healthcare providers, including pain management experts, to confirm the diagnosis and initiate treatment. When treating patients who suffer from this rare neurologic disorder, nurses should focus on assisting with treatment and providing the patient with education and emotional support—two critical components of care and living with this disease, which currently has no cure.

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