Part 5
Case 59
Contributions from Johns Hopkins Medical Institutions under Deborah A. Schwengel

All I had was a knee bursectomy; now do I have RSD (CRPS)?

Adam J. Carinci and Paul J. Christo

The case

Marcus is a 40-year-old dialysis technician who presents with severe, bilateral lower extremity pain following a right knee bursectomy in January 2006. His past medical history includes gastroesophageal reflux disease, coronary artery disease treated with a stent, hypertension, and a right knee bursectomy. He is married with no children. He has no history of substance or alcohol abuse; likewise, there is no family history of substance or alcohol abuse.

The patient's present pain began in January 2006, following a right knee bursectomy. The pain initiated in the right lower extremity and subsequently spread to the left lower extremity (contiguous and mirror image spread, respectively). He describes the pain as constant burning, aching, throbbing, shocking, stabbing, lacerating, wrenching, cruel, tearing, vicious, torturing, and unbearable. He is unable to wear pants due to allodynia and is unable to walk due to severe pain -- he is wheelchair bound. His numeric rating pain score is 7 out of 10 at rest and 10 out of 10 with activity. Aggravating factors include cold, touch, walking, and standing, and alleviating factors include rest and sitting. The pain is associated with allodynia, vasomotor changes, sweating, swelling, and weakness, discoloration, and ulcers in lower extremities.

Marcus is angry and depressed secondary to pain. The pain has affected his relationship with his wife in the form of a decreased libido. Marcus is no longer able to socialize with friends or take annual vacations to the local state park. Previous treatments included physical therapy (water-based) and interventional therapy with lumbar sympathetic blocks. Previous medication trials included oxycodone/APAP, hydrocodone/APAP, gabapentin, morphine sulfate, pregabalin, methadone, duloxetine, and cyclobenzaprine.

The patient was eventually diagnosed with complex regional pain syndrome type I (CRPS type I). He subsequently underwent spinal cord stimulator (SCS) implantation, which has produced 70% relief of bilateral lower extremity pain, and combined medical therapy (e.g., cyclobenzaprine, gabapentin, duloxetine, oxycodone/acetaminophen) relieves the remaining 30% of his pain. SCS therapy has permitted discontinuation of methadone (opioid sparing), increased mobility (out of wheelchair), elevated mood, 6-pound weight loss, and ulcer healing.

Patient care

Residents must be able to provide patient care that is compassionate, appropriate, and effective for the treatment of health problems and the promotion of health.

- Communicate effectively and demonstrate caring and respectful behaviors when interacting with patients and their families.

- Patients in chronic pain are desperately seeking relief. A compassionate, thorough history is indispensable in assessing the patient's complaints and crucial to establishing a diagnosis. Moreover, chronic pain patients may also have the additional burden of convincing the health care provider that their pain is, in fact, real because no objective signs or tests can confirm the diagnosis of pain. Caring and respect for patients are imperative.

- Gather essential and accurate information about their patients.

- Complex pain problems necessitate a thorough history and physical exam.

- Make informed decisions about diagnostic and therapeutic interventions based on patient information and preferences, up-to-date scientific evidence, and clinical judgment.

- CRPS is a debilitating neurologic syndrome characterized by pain and hypersensitivity, vasomotor skin changes, functional impairment, and various degrees of trophic change. No one treatment modality is the
panacea; rather, a multimodal, combined pharmacologic and interventional approach is often necessary.

Develop and carry out patient management plans.

The goal of treatment in patients with CRPS is to improve function, relieve pain, and enhance quality of life. Current guidelines recommend interdisciplinary management, emphasizing three core treatment elements: pain management, rehabilitation, and psychological therapy.

Multimodal therapy is key to effective treatment of CRPS. A thorough algorithm for the treatment of CRPS can be found in the literature [1].

Counsel and educate patients and their families.

Psychosocial counseling in addition to medical and interventional treatments is important in patients with CRPS.

Use information technology to support patient care decisions and patient education.

Vascular studies, electromyogram/nerve conduction testing, magnetic resonance imaging, X-rays, and blood testing are warranted. These rule out possible causes of the patient’s symptoms other than CRPS. Thermography, a three-phase bone scan, sudomotor testing, sympathetic blockade, and phenolamine infusion can help support the diagnosis of CRPS.

Perform competently all medical and invasive procedures considered essential for the area of practice.

Typical treatment incorporates medications (opioids, tricyclic antidepressants, antiepileptics, topical agents, bisphosphonates), interventions (sympathetic blocks, SCS, implantable drug delivery systems such as intrathecal pumps), and psychological counseling.

No two patients will respond exactly alike, and oftentimes, a trial of therapy is necessary, and different combinations of interventions can be trialed to arrive at an acceptable regimen. All therapies assist in achieving the primary objective of functional restoration.

Provide health care services aimed at preventing health problems or maintaining health.

Ongoing patient education and follow-up are often needed to help patients deal with the chronic pain of CRPS. Once the patient is on a stable regimen and pain is well controlled, follow-up appointments can be made once every several months. Acute flares of CRPS will necessitate more frequent follow-up to reassess the patient’s overall clinical presentation and any new changes that may have produced the acute exacerbation. CRPS is an extremely debilitating and disabling syndrome. Patients may experience months of adequate pain control, only to suffer repeated flares and setbacks.

Work with health care professionals, including those from other disciplines, to provide patient-focused care.

Referrals to pain psychologists and/or support groups often benefit patients dealing with pain and disability secondary to CRPS.

Medical knowledge

Residents must demonstrate knowledge about established and evolving biomedical, clinical, and cognitive (e.g., epidemiological and social-behavioral) sciences and the application of this knowledge to patient care.

Demonstrate an investigatory and analytic thinking approach to clinical situations.

The diagnosis of CRPS can be challenging. Again, a thorough physical exam and history of the patient’s complaints are essential to aid in diagnosis. Patients should report at least one symptom in each of the four categories and display one sign in two or more categories, according to the 1999 modified diagnostic criteria:

- sensory: report hyperesthesia as increased sensitivity to a sensory stimulation; evidence of hyperesthesia or allodynia
- vasomotor: temperature asymmetry or skin color changes
- sudomotor/edema: edema or sweating changes
- motor/trophic: decreased range of motion or weakness, tremor, dystonia or trophic changes (hair, nail, skin changes)

Once a presumptive diagnosis of CRPS is made based on physical exam and history, sympathetic blocks can then be utilized both to confirm the diagnosis of sympathetically maintained pain associated with CRPS and to treat the painful symptoms. Because
the pain in CRPS may be caused by the sympathetic nervous system, a sympathetic block (stellate ganglion block for upper extremities and ipsilateral face and lumbar sympathetic block for lower extremities) can interrupt the aberrant signaling and ameliorate the pain. Furthermore, the use of neuromodulation (spinal cord stimulation or intrathecal medications) may be required to facilitate treatment goals in patients who achieve limited benefit from more standard therapies.

Early recognition and diagnosis of CRPS is associated with better outcomes. It is essential for patients to continue using the affected limb to prevent atrophy and maintain function.

Know and apply the basic and clinically supportive sciences that are appropriate to their discipline.

Practitioners should be familiar with the typical presentation and physical exam findings as well as treatment modalities when caring for patients with CRPS. Refer to previous discussion for further details.

**Practice-based learning and improvement**

Residents must be able to investigate and evaluate their patient care practices, appraise and assimilate scientific evidence, and improve their patient care practices.

Analyze practice experience and perform practice-based improvement activities using a systematic methodology.

Proposed diagnostic and treatment algorithms for CRPS are available. Practitioners should avail themselves of such aids to help guide diagnostic and treatment decisions. PubMed is an excellent source for recent peer-reviewed research and investigations. In addition, secondary sources, such as UpToDate and MD Consult, provide review articles that synthesize the latest thinking and treatment approaches.

Locate, appraise, and assimilate evidence from scientific studies related to their patients’ health problems.

Chronic pain literature [e.g., 2–7] is replete with case reports, case series, and investigational uses of medications and interventions that have shown benefit in treating patients with CRPS.

Obtain and use information about their own population of patients and the larger population from which their patients are drawn.

What benefits one patient may or may not benefit another. A broad exposure to a variety of patients will help expand the practitioner’s knowledge base. Furthermore, seeking the opinion of more seasoned colleagues can be especially helpful in diagnosing and treating CRPS.

Apply knowledge of study designs and statistical methods to the appraisal of clinical studies and other information on diagnostic and therapeutic effectiveness.

References in the chronic pain literature are useful in diagnosing and treating CRPS [see 2–7].

**Professionalism**

Residents must demonstrate a commitment to carrying out professional responsibilities, adherence to ethical principles, and sensitivity to a diverse patient population.

Demonstrate respect, compassion, and integrity; a responsiveness to the needs of patients and society that supersedes self-interest; accountability to patients, society, and the profession; and a commitment to excellence and ongoing professional development.

Patients with CRPS have diverse pain needs. A compassionate, patient-focused, and comprehensive history and physical coupled with a multimodal treatment algorithm is essential in providing maximum benefit to patients.

Demonstrate a commitment to ethical principles pertaining to provision or withholding of clinical care, confidentiality of patient information, informed consent, and business practice.

Observe all HIPAA regulations (don’t discuss the case where others can overhear the conversation; don’t reveal any confidential patient information; provide the most relevant complications associated with specific nerve blocks, implantations, or pharmacotherapies).
Demonstrate sensitivity and responsiveness to patients' culture, age, gender, and disabilities.

A respect for culture, age, gender, and so on is important when diagnosing and treating patients with CRPS. No two patients are identical in their clinical presentation or psychosocial background; therefore practitioners must treat every patient as an individual with unique needs, requirements, and expectations.

Interpersonal and communication skills
Residents must be able to demonstrate interpersonal and communication skills that result in effective information exchange and teaming with patients, their patients' families, and professional associates.

Create and sustain a therapeutic and ethically sound relationship with patients.

Often patients with CRPS require intense support. This is an opportunity for practitioners to develop a firm physician-patient relationship with clear boundaries, expectations, and requirements. Patients with CRPS may often feel desperate or helpless, and this is a wonderful opportunity for physicians to establish compassionate avenues for communication and encouragement.

Practitioners should realize that CRPS is a syndrome that often waxes and wanes because patients may experience acute exacerbations that worsen their pain even after several months on a stable regimen. Patients may appear angry, exasperated, and dejected over these setbacks, and this may affect their personalities and ability to communicate effectively with their providers. Residents need to be patient and kind with CRPS patients and maintain empathy.

Use effective listening skills and elicit and provide information using effective nonverbal, explanatory, questioning, and writing skills.

Thorough documentation of treatment successes and failures is ultimately necessary to ensure that failed treatments are not repeated and that patients are provided with procedural interventions and medications appropriate to their specific needs.

Work effectively with others as a member or leader of a health care team or other professional group.

Any treatment plan for CRPS must be multimodal. Interdisciplinary treatment is the mainstay of effective management of CRPS. Treatment plans will often involve physical therapists, pain medicine specialists, psychiatrists and/or psychologists, nurses, recreational therapists, and occupational therapists. Respect for each member of the team will ultimately improve patient care and patient outcomes.

System-based practice
Residents must demonstrate an awareness of and responsiveness to the larger context and system of health care and the ability to effectively call on system resources to provide care that is of optimal value.

CRPS is a challenging medical problem. Effective treatment will involve practitioners from multiple specialties over the course of several years. An understanding of a team approach to treating patients with CRPS within the greater context of the health care system will help ensure that patients receive appropriate treatment, follow-up, and monitoring.

Effective multidisciplinary teams may include a pain physician, psychiatrist, psychologist, physical therapist, nurse, physician assistants, and social workers. A treatment approach that encompasses physical and psychosocial needs is ideal.

Practice cost-effective health care and resource allocation that does not compromise quality of care.

An understanding of both effective and less successful medical and interventional treatments will prevent practitioners from repeating costly tests or therapies and will avoid patient disappointment from duplicating ineffective treatments.

Advocate for quality patient care and assist patients in dealing with system complexities.
In addition to the patient with CRPS, the patient's family members and social networks are also significantly affected. Engaging the family or social supports and educating them about the course of CRPS will help each group cope with the often protracted nature of the syndrome. It will further assist them with the substantial psychosocial impact of the disease.

Know how to partner with health care managers and health care providers to assess, coordinate, and improve health care and know how these activities can affect system performance.

The pain specialist should communicate regularly with the patient's primary care physician, physical therapist, and psychologist. Integrating available inputs will better help craft treatment and tailor interventions to the unique needs of the patient. Moreover, this allows for closer follow-up and greater patient satisfaction from knowing that the entire team is collaborating with the treatment plan.
References


