Q. What do a blood draw, a rear-end collision, a gunshot wound, carpal tunnel syndrome, and a sprained ankle have in common?
A. They are ways of developing complex regional pain syndrome.

INTRODUCTION
Complex regional pain syndrome (CRPS), a painful condition previously known as reflex sympathetic dystrophy (RSD), has been described by the court as an uncommon, chronic condition that usually affects the arm or leg. The sufferer may experience intense burning or aching pain along with skin discoloration, swelling, abnormal sweating, altered temperature, and hypersensitivity in the affected portions of the body. The nature of the condition is most puzzling, and the cause is still not clearly understood. In fact, the condition remains clinically challenging both in terms of accurate diagnosis and effective treatment. This led one speaker at a conference on pain sponsored by Harvard Medical School in June 2009 to opine that CRPS is the “most controversial and least understood of all chronic pain problems.”

Although CRPS usually affects an arm or leg, there are reports of this condition occurring in other parts of the body such the face and penis. Seen more often in female patients than male patients in a ratio of 2:1 to 4:1, CRPS occurs in approximately 26.2 cases per 100,000 persons each year. This disorder does occur in children but is rare before 6 years of age, usually starting in the 10- to 12-year-old range and continuing into adolescence. The legs are more likely to be involved than arms in a ratio of 6 to 8:1, and girls are affected six times more often than boys. Few physicians question the legitimacy of this diagnosis, although some physicians who are not pain specialists reportedly still consider CRPS a “psychosomatic illness.” Understanding this disorder and the criteria used for its diagnosis has changed over the past few years, creating much confusion and controversy in medical and legal circles.

The basic diagnostic problem of this condition – severe, unrelenting pain out of proportion to the inciting injury – is significantly complicated by the subjective nature of the pain and the lack of a clear objective basis of the problem. Added to this mix is the fact that there is no diagnostic test specific for CRPS. In a medical setting, these issues create considerable debate over the accuracy of the diagnosis and appropriate treatment. In a compensation setting, subjective pain that is out of proportion to the injury is a recipe for unrelenting controversy and protracted litigation.

This chapter will examine CRPS from two different perspectives. The first part will provide a medical overview of CRPS with a brief historical review, a detailed examination of the diagnostic criteria, an explanation of the current theory of causation, and a discussion of available treatments. The second section will analyze how the courts
The issue of severe, unrelenting pain caused by trauma was first noted in 1872 by Silas Weir Mitchell, M.D. A Union Army physician during the American Civil War, Dr. Mitchell reported the occurrence of disabling pain in the limbs of soldiers due to bullet or shrapnel wounds. Most of these cases resulted from large, low velocity projectiles, also known as minie balls, utilized by the Confederate Army; Mitchell termed this disorder *causalgia*. Over the subsequent years, a number of other phrases such as Sudeck’s atrophy, posttraumatic dystrophy, chronic traumatic edema, reflex neurovascular dystrophy, algodystrophy, peripheral trophoneurosis, and idiopathic neurodystrophic disorder have been applied to identify this malady.

In 1946, James A. Evans coined the phrase reflex sympathetic dystrophy (RSD) in an effort to describe the problem based on a theory of its pathophysiology, concluding that the sympathetic nervous system is the key to this condition. The term RSD stuck as a mechanistic description, implying that the malady resulted from a reflex arc of abnormal firings by the sympathetic nervous system causing disabling pain and loss of function to an extremity.

With further research and experience with RSD, however, a growing discomfort with this proposed mechanism developed among clinicians and research physicians, resulting in a reevaluation of the terminology. In 1994 a consensus workshop of investigators and other experts in the field coined the term complex regional pain syndrome or CRPS. The reason for this change was descriptive and not to imply any pathophysiologic basis for the disorder. Complex regional pain syndrome was further subdivided into two –types: CRPS I and CRPS II. The former term was to replace RSD; CRPS II was to replace causalgia. In turn, this new terminology was codified by the International Association for the Study of Pain (IASP).

The medical community, however, has been slow to accept this new nomenclature, and subsequent validation studies using the new CRPS diagnostic criteria reveal that the disorder is being overdiagnosed. In addition, pressures from nonmedical groups, such as personal injury lawyers, are calling for a new reevaluation: ... certain influential groups have resisted the change (e.g., personal injury lawyers, who may benefit by a “looser” criteria, and some ill informed patient advocacy organizations that fear a “tighter” criteria may cause many previously diagnosed patients to be thrown into diagnostic limbo...). As a consequence, the full benefits of the common consensus-defined IASP criteria have not been completely realized.

In response to these concerns, a workshop was held in Budapest in 2003 to reexamine this disorder. The attendees, medical experts in the field of CRPS, published their consensus findings in 2005 and submitted them to the IASP for approval. As of this writing, however, the IASP has not published the new criteria and they are being revalidated. These proposed diagnostic criteria for CRPS, listed in Table 2, are to replace...
the previously used ones from Table 1. The main difference between the two sets of diagnostic criteria is the additional requirement that at the time of evaluation, there must be objective evidence of abnormalities in the painful area such as swelling, sweating and/or temperature changes, motor dysfunction, and exquisite hypersensitivity.

**CRPS Diagnosis: Clinical Criteria**

Physicians can detail the symptoms associated with patients with CRPS and make some comments as to the likelihood that the condition exists based on the symptoms a patient exhibits. However, the scientific community does not know precisely the pathologic process at work.24

The correct diagnosis of a medical problem is the cornerstone for successful care and treatment of any patient. This point is especially important when the patient is seeking compensation based on that diagnosis. Further, researchers investigating the disorder need assurance that their subjects have the same medical problem as those patients being seen in doctors’ offices. To understand the development and importance of accurate diagnostic criteria for CRPS, one needs to consider the concepts of sensitivity and specificity. These parameters are useful for determining the accuracy of clinical criteria as well as diagnostic testing for CRPS.25

*Sensitivity* refers to the ability to detect a disorder when it is present, thereby eliminating the possibility of false-negatives.26 *Specificity* refers to identifying the normal population, eliminating false-positives.27 These parameters are determined by statistical data analyses and are scored based on mathematical formulae.28 Ideally, the sensitivity and specificity values should both be 1.0 (or 100%), such that every patient with the disorder is identified (sensitivity) and no one without the condition is diagnosed (specificity).29 For practical purposes, as the sensitivity approaches 1.0, the probability of underdiagnosis is reduced.30 Likewise, as the specificity value approaches 1.0, the likelihood of overdiagnosis is reduced.31

This statistical analysis has been applied to the diagnostic criteria for CRPS as developed by both consensus workshops. Following the first workshop in 1994, Bruehl et al. evaluated 117 patients meeting those IASP criteria (Table 1) and compared them with 43 patients with neuropathic pain but not CRPS.32 They found the criteria to be very sensitive (0.98) in identifying CRPS patients, but the specificity or elimination of false-positives was quite low (0.36).33 Accordingly, these authors determined that the diagnosis of CRPS is correct in only 40% of the cases.34

**TABLE 1**

IASP Diagnostic Criteria for Complex Regional Pain Syndrome (CRPS): 1994 Consensus Workshop35

1. The presence of an initiating noxious event, or a cause of immobilization;
2. Continuing pain, allodynia, or hyperalgesia in which the pain is disproportionate to any known inciting event;
3. Evidence at some time of edema, changes in skin blood flow, or abnormal sudomotor activity in the region of pain (can be sign or symptom); 
4. This diagnosis is excluded by the existence of other conditions that would otherwise account for the degree of pain and dysfunction; and 
5. If seen without “major nerve damage,” diagnose CRPS I; if seen in the presence of “major nerve damage,” diagnose CRPS II.

A similar analysis of the more restrictive 2003 Budapest criteria was discussed by Harden et al. These requirements, based on symptoms (what the patient reports) and signs (what is observed at the time of examination), are listed in Table 2. The inclusion of signs (i.e., objective findings) at the time of examination is an important addition to the criteria, providing objective measures of pathology rather than relying solely on subjective history and complaints of pain by the patient. By requiring two of the four sign categories and three of the four symptoms, the sensitivity rate dropped somewhat (0.85) but the specificity improved (0.69). When the criteria are modified to require that two of the four signs and all four of the symptom categories be positive, the specificity significantly increased to 0.94 but the sensitivity dropped to 0.70. At the time of this writing, the more recent Budapest consensus group has proposed both of these two sets of criteria for CRPS: one for clinical application and the other (with the enhanced specificity) for research work. In an accompanying editorial, Rollin Gallagher makes the point that the one set of criteria for clinical use will “support our patients’ claim to a legitimate disease or obtain appropriate treatment.” In addition, “we can use the new research criteria to refine our samples in clinical research to establish the efficacy of these and other new and promising treatments.”

TABLE 2
Proposed Clinical Diagnostic Criteria for CRPS: 2003 Consensus Workshop

General definition of the syndrome – CRPS describes an array of painful conditions that are characterized by a continuing (spontaneous and/or evoked) regional pain that is seemingly disproportionate in time or degree to the usual course of any known trauma or other lesion. The pain is regional (not in a specific nerve territory or dermatome) and usually has a distal predominance of abnormal sensory, motor, sudomotor, vasomotor, and/or trophic findings. The syndrome shows variable progression over time.

To make the clinical diagnosis, the following criteria must be met-
1. Continuing pain, which is disproportionate to any inciting event. 
2. Must report at least one symptom in three of the four following categories: 
   • Sensory – reports of hyperesthesia and/or allodynia 
   • Vasomotor – reports of temperature asymmetry and/or skin color changes and/or skin color asymmetry 
   • Sudomotor/edema – reports of edema and/or sweating changes and or sweating asymmetry 
   • Motor/trophic – reports of decreased range of motion and/or motor dysfunction (weakness, tremor, dystonia) and/or trophic changes (hair, nail, skin)
3. Must display at least one sign at the time of evaluation in two or more of the following categories:
   • Sensory – evidence of hyperalgesia (to pinprick) and/or allodynia (to light touch and/or temperature sensation and/or deep somatic pressure and/or joint movement)
   • Vasomotor – evidence of temperature asymmetry (>1 degree centigrade) and/or skin color changes and/or asymmetry
   • Sudomotor/edema – evidence of edema and/or sweating changes and/or sweating asymmetry
   • Motor/trophic – evidence of decreased range of motion and/or motor dysfunction (weakness, tremor, dystonia) and/or trophic changes (hair, nail, skin)

4. There is no other diagnosis that better explains the signs and symptoms.

For research purposes, the diagnostic decisions rule should be at least one symptom in all four symptom categories and at least one sign (observed at evaluation) in two or more sign categories.

However, it is possible that if these dual sets of criteria are validated and accepted by the IASP, a much more confusing situation will exist than what is current, as physicians struggle to identify which patient does or does not have CRPS. From a law-related standpoint, this ambiguity as to which criteria to use to make the diagnosis of CRPS will further complicate resolution of the issues in a compensation setting.

The consensus workshop approach, as a method of formulating the CRPS diagnostic criteria, has been criticized as a valid method as opposed to evidence-based medicine, which relies on application of the scientific method to arrive at diagnostic and treatment recommendations. Although the participants of the consensus workshops are respected experts in the field, each contributes from their own experiences and biases. As stated by Harden et al., “consensus-derived criteria that are not subsequently validated may lead to over- or under-diagnosis, and will reduce the ability to provide timely and optimal treatment.” Because the recent concepts of CRPS are newly defined, “there has been an almost complete absence of evidence-based information about this condition.”

It is hoped that future research will provide the needed evidence-based information to provide more definitive diagnostic and treatment parameters for CRPS.

The older articles on RSD specified a series of three progressive stages that each patient went through leading to the terminal state of a useless extremity. Each of the stages was detailed and defined as to increasing severity of symptoms. Further, each stage had a specified duration. The recent CRPS consensus panels, however, have recommended elimination of the concept of stages, preferring not to characterize the disorder in such restrictive, artificial constraints, but rather as a continuum of signs and symptoms or possibly a series of subtypes of CRPS.

In addition to pain and the associated changes in limb appearance, abnormalities in motor function have been described. As reported by Schwartzman and Kerrigan in 43 patients with CRPS, movement disorders include tremor of the affected limb as well