0008-3194/2006/20–26/$2.00/©JCCA 2006

An unusual presentation and outcome of complex regional pain syndrome: a case report

Heather M Shearer, BA(Hon), DC*†
Astrid Trim, BSc, DC, FCCSS(C)**

A 44 year-old woman presented to a chiropractic clinic with swelling and point tenderness over the right metacarpals and right shoulder and elbow pain of insidious onset. Examination revealed right wrist and hand swelling, diminished grip strength, and reduced wrist and cervical ranges of motion. A bone scan, radiographs, and clinical examination led to the diagnosis of complex regional pain syndrome (CRPS). Following chiropractic care, the patient had improved grip strength, functional abilities, and pain reduction. The primary characteristics of CRPS include motor, trophic and sensory changes, usually in a peripheral limb following some form of trauma. Due to the varied symptom presentation, it may be unclear which conservative therapies will be most beneficial in the treatment of CRPS. A multidisciplinary approach to treatment should be pursued with these patients. More investigation of therapies such as chiropractic care as it relates to the pathophysiology of CRPS is needed.
(JCCA 2006; 50(1):20–26)

KEY WORDS: pain, neuropathy, chiropractic.

Une femme, âgée de 44 ans, s'est présentée dans une clinique de chiropratique avec une enflure et une sensibilité, localisée sur le dessus du métarcapien droit et l'épaule droite, ainsi qu'une douleur au coude d'une apparence insidieuse. Un examen a signalé une enflure du poignet et de la main droite, une diminution de la force de préhension, de même qu'une diminution de l'amplitude des mouvements du poignet droit et du cervical. Une scintigraphie osseuse, des radiographies et un examen clinique ont mené au diagnostic du syndrome douloureux régional complexe (SRDC). Suite à des soins en chiropraxie, la patiente a amélioré sa force de préhension, ses capacités fonctionnelles et la douleur a diminué. Les caractéristiques principales du SDRC incluent des changements moteur, trophiques et sensoriels qui apparaissent habituellement sur un membre inférieur, suite à une forme quelconque de traumatisme. Étant donné, la variété de symptômes qui se présentent, il n'est pas toujours évident de déterminer parmi les thérapies conventionnelles, laquelle est la plus favorable pour traiter le SDRC. Une approche multidisciplinaire du traitement devrait être envisagée pour de tels patients. Une analyse plus approfondie des thérapies, telles que, les soins en chiropratique, reliés à la pathophysiologie est nécessaire.
(JACC 2006; 50(1):20–26)

MOTS CLÉS : douleur, neuropathie, chiropratique.

* Clinical Sciences Resident, Graduate Education and Research, Canadian Memorial Chiropractic College, 6100 Leslie Street, Toronto, Ontario M2H 3J1. Phone 416-482-2340 ext: 287.
† Author responsible for negotiations concerning the manuscript.
Correspondence may be sent to Dr. Heather M. Shearer at the above address or by e-mail: hshearer@cmcc.ca
** Faculty, Canadian Memorial Chiropractic College, Private practice, Mississauga, Ontario.
Research procedures conformed to the ethical standards of the Canadian Memorial Chiropractic College Review Board.
The research was approved February 9, 2005.

Introduction

Complex Regional Pain Syndrome (CRPS) represents a revised taxonomic system for two forms of neuropathies. CRPS Type I describes regional sympathetic dystrophy (RSD) and may occur without a definite nerve lesion. Type II represents causalgia, which involves cases in which a definite nerve lesion occurred.[1] The taxonomy for the syndromes was developed in 1994 by the International Association for the Study of Pain (IASP) and was introduced based on patient history, symptoms, and examination findings.[2] The word complex was chosen to represent the varied clinical presentation of the syndromes.

The primary characteristics of the syndrome include motor, trophic and sensory changes, usually in a peripheral limb following some form of trauma to the region. Some features include persistent pain at disproportionate levels to the injury, evidence of edema, and alterations in blood flow and motor activity.[1]

The clinical picture of CRPS Type I has been described as occurring following a range of trauma, from sprains, bruises, soft tissue trauma to fractures or surgery of the affected limb.[3] Additional inciting events may include stroke, myocardial infarction, and shingles.[4] Although CRPS is usually associated with trauma, it has been reported that no precipitant could be identified in ten percent of diagnosed cases.[5] Sensory changes most commonly appear early. Physical examination may reveal a hypersensitivity to light touch and pinprick testing. Autonomic abnormalities of swelling, hyperhydrosis, and skin blood flow are reported as common diagnostic criteria.[6] Temperature differences have often been used to describe the temporal aspect of the syndrome. Acute presentations have been described as having warmer symptomatic limbs, while chronic patients are described as being in a cold end-phase.[4] Others who have not found the temperature changes to be a reliable indicator of disease stage have disputed this temporal classification.[5] Other commonly reported findings include muscle weakness, joint stiffness, impaired accurate movements, and severity of symptoms disproportionate to trauma, and regional osteoporosis in chronic patients.[4]

As noted above, CRPS Type II occurs following a traumatic peripheral nerve lesion. All symptoms are very similar to those of CRPS Type I including extreme hypersensitivity, distal extremity swelling, and symptoms spreading beyond peripheral nerve distribution.[3]

Case Study

History

A 44 year-old woman presented to a chiropractic clinic with swelling over the dorsum of the right hand, point tenderness over the 4th and 5th right metacarpals, and right shoulder and elbow pain. According to the patient, the symptoms had been present for several weeks and were of insidious onset. During the subsequent month, the patient developed progressive weakness of the right hand and wrist and was having difficulty performing daily activities such as brushing her hair and teeth, and writing. She also reported occasional paresthesia in the right hand and arm. The swelling persisted and the patient noted that the affected hand had decreased range of motion and increased temperature in comparison with the asymptomatic hand. At the time of the initial examination, the patient rated her pain as 9/10. Her medical history was unremarkable aside from being post-menopausal since the age of 39.

Findings

At the initial examination, there was visible swelling in the right wrist and hand. All right wrist ranges of motion were reduced by 50%. Grip strength was 4kg and 23kg in the right and left wrists, respectively. An upper limb neurological examination was unremarkable. There was significantly decreased cervical flexion, left rotation, and right lateral flexion. Right shoulder abduction was diminished by 10–20%. Adson's test was positive and restrictions were noted in motion palpation of the right first rib, clavicle, sternoclavicular joint, and cervicothoracic joints.

Right wrist radiographs revealed a generalized osteopenia in the carpals, distal radius, and in all five proximal metacarpals. Patchy osteoporosis was evident in all of the right carpals (See Figures 1–2). Although the lateral aspect of the lunate is not clearly visualized in Figure 1, bright light examination of the original films revealed an intact cortical margin in this area. Left wrist radiographs were unremarkable. The patient was referred to her general practitioner for blood work. Blood tests revealed a mildly elevated platelet count and erythrocyte sedimentation rate and the rheumatoid factor was negative. The patient was subsequently referred to a rheumatologist. A bone scan performed at three months post-onset revealed

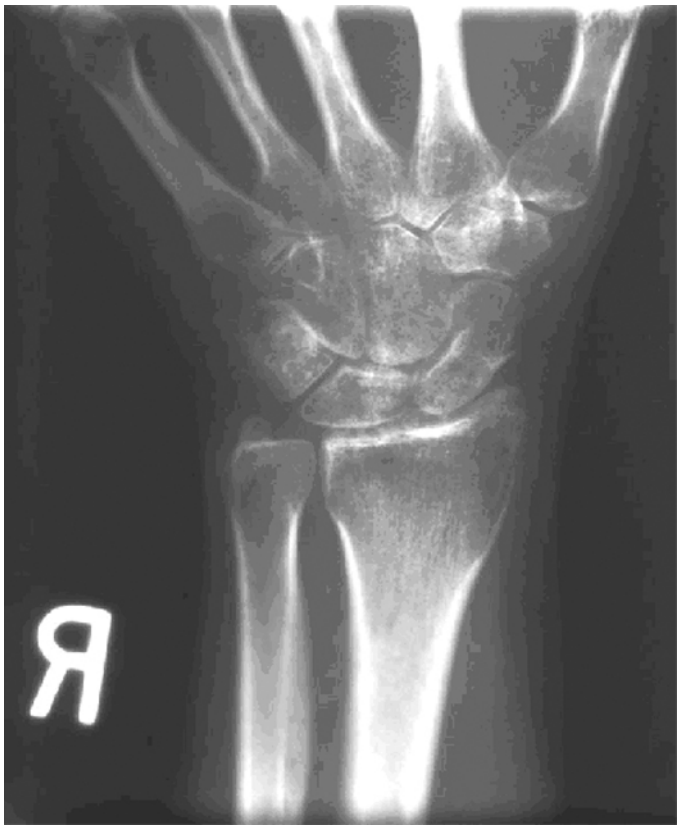

Figure 1 Right wrist with patchy osteoporosis.

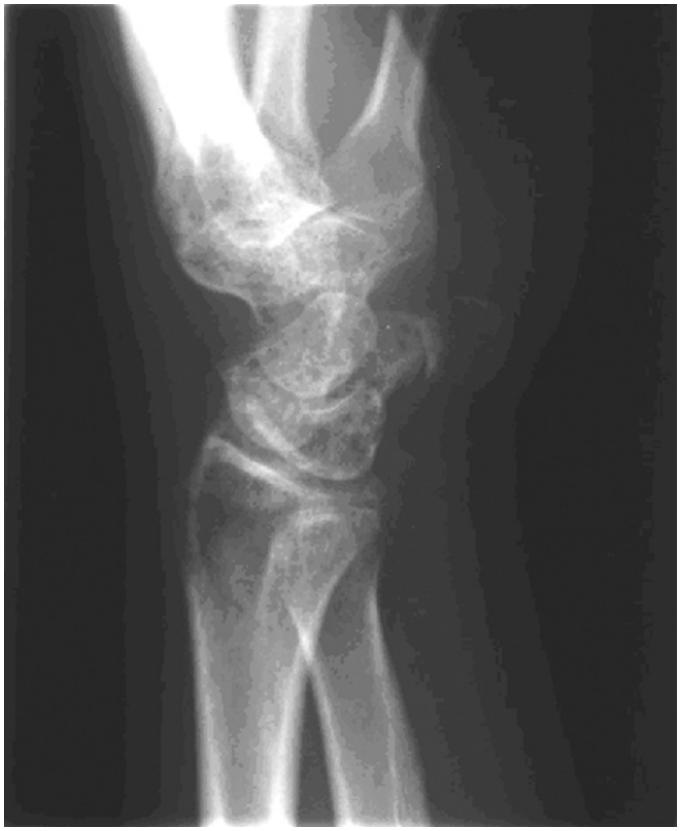

Figure 2 Multiple lucencies in the right carpal bones.

increased activity in either the metacarpals or carpal bones at the base of the right first, second, and third metacarpals (See Figures 3–4). There was milder non-specific diffuse increased uptake in the remaining right wrist region and medial half of the right clavicle. Based on these findings, a medical diagnosis of shoulder-hand syndrome was made by the rheumatologist.

Treatment

The patient was treated by a chiropractor two to three times weekly for the initial three weeks, then one to two times per week for another five weeks. The goal of the treatment was to restore first rib, clavicular, sternoclavicular, and lower cervical joint biomechanics using spinal manipulative therapy. Soft tissue therapy was also performed on the scalenes, trapezius and levator scapulae musculature. Initially, no hand or wrist treatment was provided due to the level of pain. After one week, the patient reported improved right hand dexterity and was able to grip a pencil and utensils. She also noted having improved cervical flexion and increased circulation throughout the right arm. By the completion of three weeks and 10 treatments, she had returned to work full-time. Right hand grip strength increased to 10 kg, and cervical ranges of motion were full with only minor pulling occurring with left and right rotation. Shoulder ranges of motion were full and pain-free. Right clavicle and first rib motion was improved but remained tender. Adson's test was negative. At this point, acupuncture was added to the treatment regime. Acupuncture was performed twice per month while she continued with chiropractic care once to twice per week. Following two months of care, the patient had resumed most of her exercise program, and was able to garden and work full-time. Chiropractic and acupuncture treatments were then alternated weekly for several months to achieve the best, ongoing results.

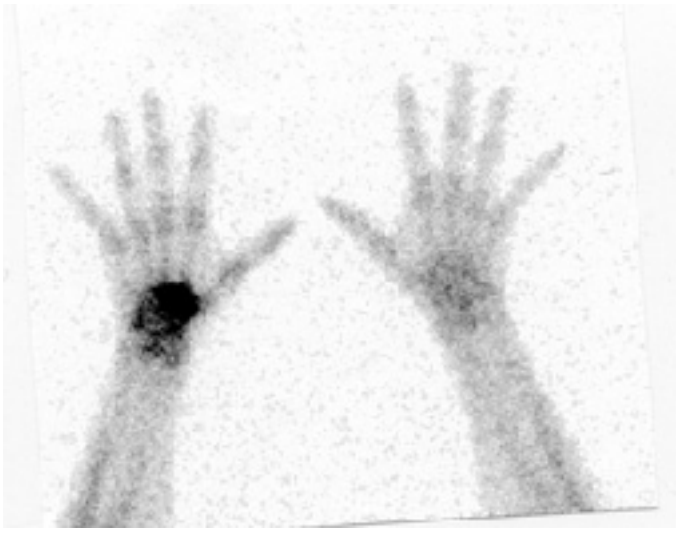

Figure 3 Wrist bone scan with increased uptake in right wrist.

Acupuncture was the chosen modality for pain control. In accordance with Traditional Chinese Medicine patterns, the two common wrist points used were "heart 7" and "triple energizer 5". Both acupuncture and chiropractic care continued for several months.

Results

A follow-up appointment with the rheumatologist occurred at five months. He re-confirmed the diagnosis, although he did remark that it was an unusual presentation of the shoulder-hand syndrome. Due to the improvement in both the hand and shoulder symptoms he recommended the patient continue with her current therapy and exercises. He also suggested the patient take Actonel due to her moderate osteopenia, and noted that it may aid in her CRPS symptoms. The patient chose not to take the medication.

An evaluation performed at nine months post-onset revealed that pain intensity levels had decreased to 1–2/10. The pain was described as a dull ache in the right wrist and shoulder only. Cervical spine and shoulder ranges of motion were unremarkable, as were all orthopaedic tests. Palpation revealed mild hypertonicity of the right scalene, and tenderness of the right clavicle and first rib. At this time, the wrist symptoms had greatly improved since the initial examination. Active and passive wrist flexion was full, while there was 30% decrease in active extension. Tenderness was reported at the base of the right metacarpals and the carpals. Motion palpation of the wrist was tender, with restrictions noted at the radiocarpal joint and midcarpal region. Grip strength was equal bilaterally. Right wrist mobilizations were initiated by the chiropractor at this point.

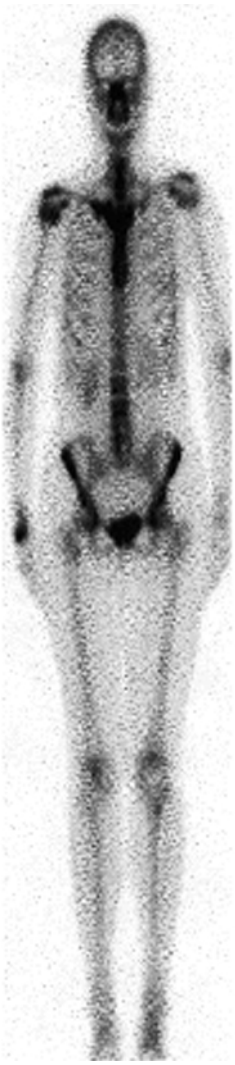

Figure 4 Full-body scan with increase uptake in the right upper limb and SC joint.

At the one-year follow-up, the patient had full cervical and shoulder ranges of motion. Right wrist ranges of motion were full with pain at end-ranges and pain with passive wrist extension. Grip strength was equal bilaterally.

At two-year follow-up, the patient reported some mild residual difficulty with prolonged writing, leading to pain in the right thumb and forearm. Cervical ranges of motion and orthopaedic testing were unremarkable. Mild tenderness was noted over the right first rib and trapezius musculature. Right wrist ranges of motion were full, with mild pain at the end range of extension. Grip strength was equal bilaterally with normal colour and muscle appearance in comparison to the left hand.

Discussion

Although the new taxonomy for CRPS has been in place for several years, it is evident that many clinicians continue to be unfamiliar with the new terminology. A literature search revealed the use of various synonymous terminology including Sudek's atrophy, reflex sympathetic dystrophy, shoulder-hand syndrome, alygodystrophy, and CRPS. In fact, the taxonomy used often varies depending on the country concerned, the specialty treating the patient, or the personally favored terminology. Part of the confusion stems from the variety of applied diagnostic criteria and until further clarity is gained with respect to the clinical presentation of CRPS, it is important that clinicians continue to be very specific and complete when describing the clinical presentation of CRPS.

The pathophysiological mechanism for CRPS remains poorly understood. Early theories focused on hyperactive sympathetic responses causing the symptoms. Proponents of this theory cite the salient response to sympathetic blockades as evidence of the pathogenesis.[6] Others have argued that because blockades are not effective in all patients, other possible pathological mechanisms need to be explored.

Another proposed theory gaining support is the possibility of an exaggerated sensitivity to sympathetic nervous system neurotransmitters. A denervation hypersensitivity of the peripheral B-adrenergic receptor is thought to occur following nerve injury. The lowered adrenergic outflow from sympathetic neurons is compensated for by latent receptor up-regulation. Increased receptor function, synaptic efficacy and exaggerated response to otherwise normal neurotransmitter loads is the result. The end result is symptoms which are attributed to an over-active sympathetic nervous system.[4]

Recent investigations have reported that an exaggerated inflammatory response may also be a possible disease mechanism.[5] Localized neurogenic inflammation may be related to increased vasodilation, acute edema, and sweating. Clinical findings which indicate that there may be an inflammatory process in the pathogenesis of early CRPS include joint effusions measured with MRIs, increased synovial protein concentration, hypervascularity, and neutrophil infiltration.[3]

The lack of a gold standard test for the diagnosis of CRPS may lead to over diagnosis of this condition. As such, CRPS can be a difficult condition for both patients and clinicians. Other neuropathic disorders, undifferentiated arthritis, rheumatism, inflammatory arthropathies, and unilateral vascular occlusive disorders should all be considered as possible differentials. In order to discern between various diagnoses, the clinical and imaging findings must not be examined in isolation. Taken together, the findings will quickly help to narrow down the differential diagnoses. For example, the inflammatory arthropathies which affect the hand and wrist include rheumatoid arthritis, psoriatic arthritis, and gout. Infection should also be considered another differential. Some of the radiographic findings of CRPS include normal joint spaces and margins. This assists in differentiating CRPS from infection or rheumatoid arthritis. In addition, the clinical pictures would be notably different for all three diagnoses. Although the clinical findings between CRPS and gout do have several similarities, the radiographic findings for both this and psoriatic arthritis are not consistent with those of CRPS.

Several authors cite the misdiagnosis of dysfunctional postures as CRPS Type I, and that a multidisciplinary approach for the diagnosis and treatment of both conditions is needed.[8] Currently we must rely on a detailed history and physical examination in order to make the appropriate diagnosis. Certain key information must be ascertained. This includes any history of inciting trauma, and history of sensory, autonomic, and motor changes. The physical examination should reveal any neurological changes, distribution of pain, swelling, sweating, and trophic changes.

In a prospective study, the symptoms and progress of 829 CRPS-diagnosed subjects were examined over an eight-year period. It was concluded that patients could not be subdivided into temporal stages based on the temperature of the affected limb. Instead, it was suggested that patients be categorized by either primarily warm or

cold forms. Those with colder limbs were more inclined to have been suffering from CRPS for a longer period of time.[5] Due to the inconsistencies of presenting symptoms among acute and chronic sufferers, it has also been suggested that the syndrome be classified as either mild, moderate, or severe.[7] The current diagnostic criteria require that the following four criteria be satisfied:[2]

1. The presence of an initiating noxious event or a cause of immobilization.
2. Continuing pain, allodynia, or hyperalgesia with which the pain is disproportionate to any inciting event.
3. Evidence at some time of edema, changes in skin blood flow, or abnormal sudomotor activity in the region of the pain.
4. This diagnosis is excluded by the existence of conditions that otherwise would account for the degree of pain and dysfunction.

In the IASP classification, Merskey and Bogduk noted that only criteria 2 and 4 had to be satisfied.

Paraclinical testing is important to provide information regarding autonomic, sensory, and motor changes. Bone scintigraphy is useful in providing information about vascular bone changes, especially in later disease stages. Plain radiographs need only be taken at the chronic stage, when bone mineralization status can be evaluated. Quantitative sensory testing (QST) and autonomic function testing for these patients has been reviewed in the literature, although it is beyond the scope of this paper to discuss the details of the testing.[3,7]

Not only has the taxonomy of this syndrome changed, the therapeutic approach has also been greatly modified. Sympathetic blockades were a common diagnostic tool and form of therapy. Studies have shown that there is inadequate evidence to unequivocally report the benefit of sympathetic blockades on pain levels and motor function. It was reported that approximately 85% of CRPS patients report a positive acute effect following sympathetic blocks.[3] In contrast, others have reported that sympathetic blockade or sympathectomy had minimal effect with only 7% reporting lasting success.[5]

More recently, there has been a shift towards restored functional abilities instead of pain control as the primary goal of treatment. Exercise therapy is considered one of the key elements of this process.[7] Physical therapy is an important component to treatment although aggressive therapy can be detrimental in early stages when pain levels are high. Although one functional goal is to improve joint motion and strength, passive movements are frequently too painful and should be avoided in acute patients.[4] Passive therapy, followed by active isometric and then active isotonic training can be pursued when pain levels diminish.[3]

A multidisciplinary approach to treatment should be pursued with these patients. Various modes of therapy may be considered. The primary goal of pharmacological intervention is pain relief.[7] In the early onset, under the assumption that neurogenic inflammation is the pathological mechanism, NSAIDs and steroids may be administered as a form of pain control. Even so, Vacariu reported that few clinical trials have shown benefits.[7] There are no long-term studies on the use of opioids in treating CRPS or other neuropathic pain although this is thought to be an effective initial course of therapy in order to manage pain levels. Tri-cyclic antidepressants have been shown to have an analgesic effect and are used in doses smaller than those needed to produce anti-depressant effects (75–150mg/day).[3] The use of TCAs may be related to interrupting the pain cycle by improving sleep, mood, and anxiety. Physical therapy, and to a lesser degree occupational therapy, has been effective in reducing pain and improving active mobility in patients with CRPS I of less than a one year duration.[9]

For evidence-based practitioners, chiropractic practice has progressed to being more than simple joint manipulation. The current case represents the increased awareness that many conditions require some physical therapy component, including specific exercise regimes. The patient had multidisciplinary management and was treated with spinal manipulative therapy, soft tissue therapy, acupuncture, and home exercises by the chiropractor. There have been limited studies published regarding the chiropractic treatment of CRPS and similar conditions. Besides the current case, only one published case report regarding chiropractic management of CRPS was found. This study reported drastic upper limb symptom improvement in one patient following a total of three cervical manipulations and home exercise therapy.[10] The current case had a similar level of improvement in upper limb symptomatology and functional ability following first rib, clavicle, and cervicothoracic manipulation.

These two CRPS cases are encouraging with respect to the results obtained using a non-pharmacological multidisciplinary approach to care. CRPS can be a severe, debilitating condition of which the effects may be long lasting. A key factor in the prognosis and recovery of CRPS patients is early intervention focusing on restoration of functional abilities. Manipulative therapy serves to restore joint function and may be effective in improving somatic dysfunction associated with CRPS and other similar conditions.[4] Home exercises and a return to activities of daily living may limit the overall disability and shorten recovery time.

Although the diagnostic criteria of CRPS were met in the current case, the true pathophysiology of the disease is yet to be completely understood. Due to the varied symptom presentation and extent of affliction, it may be unclear which conservative therapies will be most beneficial in the treatment of CRPS. In order for clinicians to make truly accurate diagnoses and treatment protocols for this group of patients, more investigation of therapies such as chiropractic care, acupuncture, massage, and exercise as they relate to the pathophysiology are needed.

Acknowledgements

The authors would like to thank Dr. Paula Stern for her assistance in reviewing the manuscript.

References

1 Cepeda S, Carr D. Local anaesthetic sympathetic blockade for complex regional pain syndrome. The Cochrane Database of Systematic Reviews 2004, Issue 1. Art. No.:CD004598.
2 Stanton-Hicks M, Janig W, Hassenbusch S, Haddox JD, Boas R, Wilson P. Reflex sympathetic dystrophy: changing concepts and taxonomy. Pain 1995; 63:127–133.
3 Wasner G, Schattschneider J, Binder A, Baron R. Complex regional pain syndrome – diagnostic, mechanisms, CNS involvement and therapy. Spinal Cord 2003; 41:61–75
4 Muir JM, Vernon H. Complex regional pain syndrome and chiropractic. JMPT 2000; 23(7):490–497.
5 Veldman PHJM, Reynan HM, Arntz IE, Goris RJA. Signs and symptoms of reflex sympathetic dystrophy: prospective study of 829 patients. The Lancet 1993; 342:1012–1016.
6 Turner-Stokes L. Reflex sympathetic dystrophy – a complex regional pain syndrome. Disability and Rehabilitation 2002; 24:939–947.
7 Vacariu G. Complex regional pain syndrome. Disability and Rehabilitation 2002; 24:435–442.
8 Stutts JT, Kasdan ML, Hickey SE, Bruner A. Reflex sympathetic dystrophy: misdiagnosis in patients with dysfunctional postures of the upper extremity. The Journal of Hand Surgery 2000; 25A:1152–1156.
9 Oerlemans HM, Oostendorp RAB, de Boo, Goris RJA. Pain and reduced mobility in complex regional pain syndrome I: outcome of a prospective randomized controlled clinical trial of adjuvant physical therapy versus occupational therapy. Pain 1999; 83:77–83.
10 Bortolotto J. Reflex sympathetic dystrophy: an enigmatic improvement with spinal manipulation. JCCA 2000; 44:245–251.