

# Reflex Sympathetic Dystrophy in Adolescents: Lessons for Adults

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Persistent pain without clear explanation frequently involves the musculoskeletal system, with the core clinical features being the presence of pain and abnormal tenderness in the same area. Such conditions may be widespread or localized, that is, to an upper or lower quadrant or, less commonly, to a more restricted distal limb region. The causes, pathophysiologic mechanisms, clinical presentations, and management strategies of these chronic pain syndromes remain controversial (1). Any insight into any aspect of these problems is welcomed.

It is often difficult for a clinician to know where to begin when assessing such patients. There may be a number of possible causes, differing severity of clinical features, and marked variation in response to treatments. Outcomes also differ greatly. It is no wonder that with such multidimensional disorders any commentator on the topic is inevitably partly right and partly wrong! Psychological and social factors provide input into the mechanisms of disordered pain biology but are also major consequences of this process. As such, the biopsychosocial model of disease is usefully applied to these disorders (2,3). Both the common fibromyalgia syndrome and less common complex regional pain syndrome (CRPS), also known as reflex sympathetic dystrophy (RSD), are better managed with this approach.

As in other pain syndromes, the study of CRPS/RSD has been associated with refinement of terminology and classification criteria (4). For many years, terms such as algodystrophy and Sudeck's atrophy have emphasized the uncommon outcome of severe tissue damage, whereas the popular term reflex sympathetic dystrophy incorporates the putative role of the sympathetic nervous system in this disorder. More recently, the descriptive term complex regional pain syndrome has gained favor to emphasize the

clinical features without implication of causative mechanisms (5), somewhat akin to the reasons for use of the term fibromyalgia syndrome (6). Parallel to taxonomic changes, criteria to classify and diagnose CRPS continue to evolve (7,8). In classic CRPS, the clinical features are usually obvious and often spectacular. Indeed, orthopedic surgeons may make this clinical diagnosis! Seeing is believing—swelling, vascular change, and marked tenderness all may be prominent. However, CRPS exists on a spectrum with milder CRPS being far more common than the severe types that characterize textbook descriptions. In many cases, clinical features may be more subtle, particularly initially.

Many of these issues are highlighted in the article by Maillard et al (8) in this issue of *Arthritis Care & Research*, in which 23 adolescent patients with CRPS, who attended a single pediatric unit at a London hospital, are reviewed. Despite the limitations of the methodology involved in this retrospective review, the described clinical approach to CRPS is very sensible and contributes further understanding to an important and often underreported condition. The observations in these adolescents carry important lessons for those managing CRPS in adult patients.

First, the review highlights the difficulty of diagnosing CRPS. This difficulty arises because there is often a low level of awareness of the condition among the various health professionals who are confronted by persons with persisting regional pain problems. However, it also relates to the overlap between early clinical features of CRPS and those that associate with the normal and expected temporary process of postinjury pain sensitization. In such settings, CRPS may simply “creep up on you.” What, then, might alert a health care professional to a person being a candidate to develop CRPS? One important clue is the presence of a specific trigger, such as tissue injury, that occurs in a highly charged emotional context. Examples range from myocardial infarction to arthroscopy. Here, psychosocial distress may associate with either pain out of proportion to the underlying tissue damage or persisting beyond the normal time of tissue healing. In such situations, the astute clinician will usually anticipate a possible pain syndrome well before any specific criteria are met. In this setting preventive strategies, such as explanation, mo-

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Submitted for publication November 5, 2003; accepted November 10, 2003.

bilization, counseling, and support, are all part of normal care.

Even so, it may be necessary to perform additional investigations to clarify the diagnosis. These need to be planned cautiously and accompanied by careful explanation to avoid excessive concern and further pain amplification. The diagnosis of these pain syndromes, CRPS and fibromyalgia alike, are not diagnoses of exclusion nor, importantly, are they exclusive diagnoses. Many individuals have an associated pain-generating disorder that needs treatment in its own right, independent of the CRPS pain sensitization process. This could include bone fracture, soft-tissue injury, inflammatory arthritis, or, uncommonly, nerve injury, to name but a few.

Early diagnosis allows for early intervention. The first intervention, as outlined in the Maillard review, is appropriate education as to the nature of the CRPS problem. The package of information given to an individual will vary according to the level of understanding, the surrounding clinical circumstances, and the severity of the problem (2,3). The concept that CRPS is a disorder of pain neurophysiology, rather than being due to tissue damage in the painful region, is critical. Notions of pain modulation must offset the common fear of aggravation of perceived tissue damage that so often significantly interferes with the crucial need for mobilization and activity.

Physical therapy strategies may be simple or complex. In adolescents, hydrotherapy is extremely helpful and this may also be so in adults. Pursuit of activity requires appropriate analgesia, which is best kept as simple as possible. Desensitization of the involved area may require techniques ranging from touching, gentle massage, and mobilization to relaxation and psychological therapy, which will often involve skills from a wide range of personnel. This multidisciplinary team has dual primary goals: improved function and pain relief. A key goal in adolescents is return to school and in adults return to work, in addition to routine recreational and household activity. It is essential that the treating team speak with one voice, providing the same message about management strategies and goals. The psychologist in the team is often invaluable in defining entry into a variety of coping and pain (life) management strategies. However, every therapeutic input has associated psychology and sometimes a one-on-one physical therapist provides this input.

Thus, early diagnosis and early intervention are clearly usefully applied to adults. The next lesson from adolescent CRPS is in regard to psychosocial inputs into the disorder (9). Many persons who developed this syndrome had family histories of chronic pain of different types. Many had previous episodes of persistent pain. In adolescents, subtle and sometimes not so subtle psychological factors are often prominent and occur in the background of the CRPS presentation. In adults, CRPS is often triggered in the context of work or injury. Depending on the society, with differing attendant safety-net legislations, an expected self-limited injury to the musculoskeletal system (particularly in a distal limb area) may translate into a persistent pain syndrome. The inevitable focus on biologic factors as the primary cause for the ongoing pain, coupled

with lack of resolution of this "biologic" pain, confuses both the health care professional and the patient. Issues of fear, frustration, mood and sleep disturbance, and persistent and aberrant stress response compound and feed back into an activated pain system. Perturbations of medicolegal dispute and loss of important life roles create powerful ongoing psychosocial forces that do not favor resolution of the pain problem (10). This occurs despite the use of powerful analgesic and psychotropic medication or an ever-increasing variety of interventions aimed at blocking components of the disordered pain system. An early appreciation of the psychosocial factors involved in the presentation of individuals with injury as a potential trigger for any persistent pain may obviate the postinjury pain problem, of which CRPS is often a spectacular example.

Finally, perhaps the most important lesson is that of reversibility of this problem. In the current study, >80% of subjects were doing very well at 6 months, and this is in a population from the more severe end of the CRPS spectrum, all of whom required management through a tertiary care unit. The finding that a severe chronic pain syndrome can potentially reverse provides us with extremely important and powerful knowledge. Even though it has traditionally been stated that younger people with CRPS have a good prognosis (11), this same situation also pertains to adults. Although the literature and previous terminology of this disorder emphasize the severe end of the CRPS spectrum with poor outcome, i.e., dystrophy, the more usual outcome is actually resolution of the problem. This knowledge is required to help the individual heal, to help the management team persist with strong purpose, and to allow the law to understand that this condition does not persist in the majority. When it does persist, complex psychosocial factors are often persistent, and these include the medicolegal, disability, and other safety-net systems that all too often become part of the problem, significantly contributing to a futile cycle of suffering. Earlier diagnosis and earlier intervention shift the CRPS spectrum to better outcomes.

There are of course differences between clinical features of CRPS in adolescents and adults; for instance, lower limbs are more affected than upper limbs, blood flow based on nuclear scan evaluation is diminished, and psychosocial factors are usually more obvious. However, the plasticity that characterizes the spectacular up and down of the activated adolescent pain system does not fade as one ages. Attention to both psychosocial and biologic factors together is essential to down-regulate the pain system and optimize outcomes in these conditions at all ages.

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