Plasticity of Complex Regional Pain Syndrome (CRPS) in Children

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Abstract

Complex regional pain syndrome I (CRPS I) is defined by the International Association for the Study of Pain (IASP) criteria to include pain that is disproportionate to the inciting event, sensory disturbances such as allodynia/ hyperalgesia, autonomic dysfunction, and motor dysfunction that usually occurs after trauma that is frequently trivial and generally expressed in an extremity. These symptoms are well described in the adult population, but there are relatively few data or reports of its prevalence in the pediatric population. Recent studies have demonstrated that unlike the adult population, about 90% of the cases reported are females in a range of 8 to 16 years, the youngest being 3 years old. There tends to be delay in recognizing the diagnosis, which may be as long as 4 months. In contrast to adults, the response to treatment, particularly exercise therapy with behavioral management will achieve almost 97% remission. While the pathophysiology is poorly understood, many features, particularly the neurologic abnormalities, suggest both peripheral and central nervous system involvement. Peripheral small fiber neuropathy as an etiology and inflammation involving small nerve fibers (neurogenic inflammatory pain) has been suggested. A tissue inflammatory etiology has been investigated over the past 25 years. However, these inflammatory aspects differ from those seen in other conditions involving tissue inflammation. The suggestion that CRPS in children is a different clinical entity than that seen in the adult, is probably incorrect, as recent evidence would suggest that the pathophysiology is most likely identical involving endocrine, behavioral, developmental, and environmental factors that distinguish clinical presentation in children from the adult. Behavioral management is a mandatory accompaniment of any program of exercise therapy and the sometimes extreme sensory disturbances and parental enmeshment do distinguish the clinical presentation from that in the adult. Interventional procedures may be required in the face of extreme allodynia preventing exercise therapy, and in occasional cases interruption of the sympathetic nerves may reverse this symptom in a few children. Occasionally, continuous analgesia techniques such as that which can be delivered by tunneled epidural catheter or an externalized neurostimulator (spinal cord stimulation) for short periods of time are effective.

Key Words. Complex Regional Pain Syndrome (CRPS); Therapy; Reflex Sympathetic Dystrophy (RSD); Occupational Therapy; Physical Therapy; Behavioral Therapy

Introduction

While complex regional pain syndrome (CRPS) is particularly incapacitating and generally occurs after minor injury or abrasion, in approximately 10% of the patients there is no history of any trauma [1–16]. All tissues in the affected region are involved to a greater or lesser extent and all function in the region may be temporarily lost. While a pathophysiological mechanism for CRPS remains unknown, number etiologies have been proposed: a peripheral small fiber neuropathy, an exaggerated regional inflammation (neurogenic inflammatory pain involving small nerve fibers), and autonomic (sympathetic) dysfunction [7,8,17,18]. Extreme variation of the presenting characteristics includes sensory, motor, neurovascular, and sudomotor dysfunction. These characteristics not only vary from patient to patient, but can undergo temporal change in the same patient. In fact, disagreement concerning a potential mechanism is reflected in the lack of consensus concerning which diagnostic criteria are most important [14,19]. While several attempts have been made to validate and provide uniform diagnostic criteria, these have all involved the evaluation of symptoms in an adult population and understandably have been compared with those in established neuropathic pain syndromes [20,21]. Furthermore, inter-observer agreement of the sensitivity and specificity in relation to quantitative sensory tests for clinical examination in comparison with neuropathic pain syndromes has only a tenuous correlation [14].
During the past 30 years, an increasing number of reports have demonstrated an incidence of CRPS in children that is distinguished by both clinical differences and the ratio of males to females [3,4,13]. Until recently most clinical trials and case series reporting these sensory abnormalities as a component of the diagnostic criteria for CRPS have not analyzed patterns of cutaneous sensitivity in children and adults. In fact there has been no standardized physical inventory of these features in children and adolescents [22,23]. Only one research group has made a point of comparing clinical features with those in a large series of adults. These results and those of other observers are discussed [4,24].

Materials and Methods

This review will look at the clinical experience gained through the management of young children and adolescent’s with CRPS. 69 children (55 female), with an average age of 14.58 years, were enrolled in a 3-week comprehensive rehabilitation program that is focused on physical and behavioral measures, but which in a few cases utilizes interventional methods. The salutary clinical response in these children will be discussed in the light of recent literature, which demonstrates increased awareness that CRPS does occur with regular frequency in children, but is associated with slightly different characteristics than are commonly seen in the adult. Before looking at the recent literature regarding CRPS in children, it is worth describing the pediatric pain rehabilitation program at the Cleveland Clinic. This is a two-part program that consists of in-patient and day hospital components. The program’s length was chosen to conform to what was considered to be an adequate length of time and intensity to achieve resolution in most cases, namely 2 weeks as in-patient and 1 week as a day hospital patient. The interdisciplinary nature of the program incorporates specialists from pediatric medicine, psychiatry, behavioral medicine, physical and occupational therapy, nutrition, and social work. Other specialists from anesthesiology, gastroenterology, neurology, psychiatry, psychology, rheumatology, pediatric spine, upper, and lower extremity surgery are also involved in the overall management of these children. Of particular importance are the behavioral aspects that include training or self management skills such as relaxation, diaphragmatic breathing, and mental imagery, combined with stress management and problem solving aspects. Children attend music therapy and schooling is provided concurrently at their particular grade. Parents are also provided with guidance as to how their children should be managed. To avoid the all-work-and-no-play, aspects of treatment, recreational therapy is an integral aspect of the program. Throughout the 3-week period an effort is taken to foster one-on-one small group sessions. These are used to instill new interests and help the child to return to their previous activities. The Social worker not only prepares the family for their child’s hospitalization, but also following their structured rehabilitation to help reintegrate the child back in their home surroundings. Preliminary evaluation of these patients suggests that the program is accomplishing its goals, particularly in the area of improved functioning. Initial results show a 51.4% improvement in pain severity, 62.5% improvement in physical functioning, and improvements in pain-specific anxiety (53.2%) and social functioning (37.8%) (Figures 1–3). These preliminary data suggest that our interdisciplinary rehabilitation approach can be effective in helping children return to normal activity despite pain. Longer term, more comprehensive assessment of a larger group of these patients is ongoing and will be important for better evaluating program effectiveness.

The recent report by Wilder underscores the demography of CRPS in children and emphasizes the importance of early clinical diagnosis and early rehabilitation using physiotherapeutic measures. The diagnosis of CRPS is based on the IASP diagnostic criteria: [1]

1. The presence of an initiating noxious event.
2. Continuance of pain, allodynia or hyperalgesia with which the pain is disproportionate to an inciting event.
3. Evidence of edema, changes in blood flow, or abnormal sudomotor activity.
4. The diagnosis is excluded by the existence of conditions that would otherwise account for the degree of pain and dysfunction.

Children most often describe their pain as burning and use adjectives that describe sensations of dysesthesia. The most prominent pediatric symptom in an extremity is the change of temperature and color. The change in color suggests that the sympathetic nervous system is most likely involved and this association may be demonstrated by the response to a sympathetic block that may relieve both the clinical signs and much of the burning and dysesthetic pain [13]. This has been described as sympathetically maintained pain (SMP) [25,26]. That nociceptors can be sensitized by nor-adrenaline has been demonstrated by Drummond and co-workers [17]. Most children that are seen with the clinical signs of CRPS are active in sports and gymnastics. Not uncommon is the level of enmeshment of the child and parent [4,13]. This is particularly true in those cases in which marital discord has an adverse impact on driving the severity of the syndrome often after minor trauma [11]. Unlike the adult, the tremor and other motor signs are a relatively rare component [14,27]. Because excessive pain is the hallmark of CRPS that is a sequel of trauma, it is essential to exclude any physical, e.g., orthopaedic process or latent pathology that can undergo treatment. The literature regarding scintigraphy as an aid to diagnosis is both confusing and nonspecific.

Both treatment and the “plasticity” of pediatric CRPS distinguishes its management and response from that in the adult. Although the use of physical modalities as exemplified by two authors, Bernstein and Sherry, almost 30 years apart, the recent randomized prospective trial by Lee et al. who used a once weekly or three times weekly outpatient physical therapy and weekly cognitive behavioral therapy, did not show any statistically significant differences in outcomes in each group [12,28,29]. What seems to be clear from our own results is that the majority of children do well without any interventions beyond those used to facilitate physical activity in the rehabilitation program described earlier. Approximately 6–7% of children and adolescents do require additional analgesia, not feasible orally, that can be implemented in the form of epidural infusion, continuous regional analgesia (block) or SCS [5,16]. In such cases where the severity of alldynia precludes any effective participation in physical therapeutic maneuvers, it is imperative to proceed with one of the foregoing modalities. In fact the impact of pain relief on the child not only encourages total investment in their well being, but also improves participation in their concurrent behavioral management (Figures 1–3). The truth is, continuous epidural analgesia or an extended SCS trial were used in a limited manner for the duration of the program, and in two (2/68) of the children were continued for 6 weeks and 2 months, respectively, before their removal. We did experience a similar rate of recidivism, 30%, described in the literature [3–5,12]. All children, however, responded positively after a second course of treatment—up to 4 weeks in the rehabilitation program.

For years, CRPS has been associated with sympathetic dysfunction to the extent it was considered primary pathophysiology. As a consequence, blocks of the sympathetic nervous system were considered the main treatment with little thought to other possible mechanisms. During at least the initial stages of the syndrome this lead to a monotherapy with repeated blocks of the sympathetic nervous system even if the response was frequently only the duration of local anesthetic effect. While a single sympathetic block may prove useful to determine the presence of SMP and therefore a predictor of their response to alpha-1-adrenoceptor blockade, a few children may have a dramatic and prolonged duration of effect allowing full participation in exercise and behavioral therapy. The use of indwelling tunneled epidural catheters provides a means for adequate analgesia without incurring motor inhibition sufficient to interfere with therapy. In the few cases that require this modality, it has been used for a period of 4 to 6 weeks.

**Literature Review**

1. A recent article from France describes the use of regional analgesic techniques for managing children who have been refractory to conservative measures [5]. Thirteen children with both upper and lower extremity CRPS were studied in a prospective manner. The study evaluated the effect of placing either a popliteal catheter for the lower extremity or an axillary catheter for the upper extremity followed by a Bier Block with a mixture of Lidocaine, hydroxyethyl starch and bupivacaine for 20 minutes.

In a study of 21 families, the authors reported significant enmeshment with one or both parents. Most children were high achievers and were also very compliant. Almost half of these families were associated with marital discord. Fifty percent had difficulties in school. No major psychopathology was revealed and one child demonstrated a high score for somatization. All children who had not responded to conventional treatment of CRPS were included in the study protocol. It was also noted that a...
Comparative control group was not permitted and was considered to be unethical. It was also noted that the excellent control of pain allowed both physical therapy and psychotherapy to proceed without hindrance. Of special mention was the completeness of pain relief, which enabled both intensive occupational and physical therapeutic measures to be used.

Supervised therapy by a physical therapist began 1 hour after the institution of the continuous nerve block and again at 6–12–24 hours at the hospital. In addition exercises were programmed to be continued twice daily at home. The total duration of therapy occupied 4 days. A total of 13 children, one with upper extremity CRPS were treated. All lower extremity CRPS were walking after 24 hours. No side effects occurred and a complete remission in all children was achieved at 2 months.

2. Spinal cord stimulation for children is highlighted in a recent article in which severe refractory CRPS in seven girls aged 11–14 years was instituted [16]. All children were incapacitated and had been resistant to both regional analgesic blocks and conservative therapy. The authors make the point that in spite of the invasive nature of spinal cord stimulation and the possibility of a strong placebo effect, the symptoms in these patients were so severe, devastating and chronic that there was little else that could be offered to facilitate physical and behavioral measures. In four cases, because of complete resolution of their symptoms, the spinal cord stimulator was removed [30]. The author’s also point to one recent study in which a randomized controlled format was used. Most of the recent reviews or meta-analyses of spinal cord stimulation do support a beneficial effect on pain, particularly allodynia [31,32]. While there have been few reports of SCS being used in the pediatric group, there are numerous clinical reports of transtcutaneous electrical stimulation being used in children since the mid-70s [33,34]. All of the children in these reports did respond to this modality and were described as remaining in remission.

The authors concluded that as a minimally invasive technique, SCS in adults has been successful in the treatment of neuropathic pain. Its use in children as demonstrated in this case series provides support for its recommendation where all other modalities and management of CRPS have failed.

3. The study from Harvard referred to earlier is a valuable contribution to the literature. The authors investigated the sensory and cutaneous abnormalities found in children and adolescents with CRPS. Using quantitative sensory testing (QST) and an extensive neurological examination they were able to follow patterns of sensory dysfunction and those pain descriptors used by children and adolescents. While acknowledging the well-described features in other studies, they were able to contrast these with the physical findings that are reported in a number of citations. While acknowledging a similar diversity of signs and symptoms in the adult population, a similar constellation of clinical features is also observed in children. However, the authors noted that there was a significant distinction between what children reported and what physicians observed at least in terms of the frequency of neurovascular, atrophic, and pseudomotor symptoms in pediatric CRPS patients [24]. In 76% of children with CRPS, mechanical allodynia, static allodynia, and allodynia to punctate temporal summation were found in contrast to the adult in which mechanical allodynia occurred in 70 to 100% of patients [22,23,35]. Also of note was a report of 16.7% heat and 33% cold allodynia occurred less in children in comparison with the adult. Heat hyperalgesia in the adult ranged between 14 and 55% of cases. Of interest is the fact that the authors observed a wide range in the thresholds for cold, warm, and vibration sensations that were not significantly different with controlled values. They noted that these findings are similar to those in adults with post-traumatic neuropathies and CRPS.

Sixty-two percent of the children exhibited both mechanical dynamic, static allodynia and allodynia to punctate temporal summation. Allodynia to both light stroke and threshold- strength punctate stimuli is most likely mediated by A-beta fibers and is maintained by the sensitized state of WDR neurons induced by injury afferent input or chronically sensitized C-fibers. Although in some neuropathic pain states after traumatic or ischemic injury, mechano-insensitive C-fibers are sensitized. This may induce primary sensitization of C-fibers, mechanical static allodynia/hyperalgesia and/or increased neural input necessary to maintain central sensitization and mechanical dynamic allodynia. The authors note that while both peripheral and central sensitization contributed to mechanical dynamic and static allodynia in their patients, the results of QST studies in adults with CRPS I, like their own study, and in their study, suggest that abnormal hyperexcitable sensory patterns have a role in central sensitization [36,37–40]. Multi-variate analyses in which mechanical dynamic allodynia is present and absent are more predictive of cold and heat allodynia than a number of other clinical variables such as age, sex or duration of symptoms. Of interest is that 24% of their patients did not exhibit mechanically evoked cutaneous pain. The lack of pain in these circumstances does not exclude the possibility sensitization in deeper tissues involving nociceptors of muscles and joints. Furthermore, at the time that this study was carried out, many of their patients might have been in a phase of spontaneous remission. The distinction between hyperalgesia in deep tissues might have been overlooked due to the study design, which was limited to cutaneous testing. Certainly, the signs of deep tissue C-fiber desensitization could have contributed to spontaneous pain in the absence of evoked cutaneous pain.

The authors point out that the study was also limited by the degree of testing feasible in children and adolescents. Obviously serial testing would have improved the sensitivity of their methods and reducibility of a particular sensory dysfunction. The authors are to be commended on undertaking a difficult study but one that demonstrates quite clearly that QST can be used to test most children with
CRPS and is sensitive enough to catch the changes in mechanical and thermal sensibilities. This is also the first study to quantify changes in sensitivity of childhood CRPS.

4. In a retrospective analysis of 78 children over a period of 24 years, Tan and colleagues compared the clinical characteristics with those of 840 adults during the same time period [4]. The age of the children ranged between 5 and 16 years and 16 to 96 years in the adult population. One upper extremity was affected in 23.3% of children, one lower extremity in 72.6% and in both lower extremities in 4%. In contrast, the upper extremity of adults was more frequently involved 60.8%. In most children CRPS resulted from an ankle sprain in 15.4%. See Table 1.

The main complaints in both groups were pain, a difference in skin color, and skin temperature. In the pediatric population, however, the affected extremity was invariably cold (71.8%). The authors emphasized the significant clinical differences between adults and children with CRPS I. In Table 2 these observations in children and adults are shown.

### Table 1  Clinical characteristics of adult and children with complex regional pain syndrome I

<table>
<thead>
<tr>
<th></th>
<th>Adults, % (n/n_ev)</th>
<th>Children, % (n/n_ev)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gender</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Female</td>
<td>74.9 (712/951)</td>
<td>85.9 (67/78)</td>
</tr>
<tr>
<td>Male</td>
<td>25.1 (239/951)</td>
<td>14.1 (11/78)</td>
</tr>
<tr>
<td>Median age in years (range)</td>
<td>43.8 (16–96)</td>
<td>13 (5/16)</td>
</tr>
<tr>
<td>Upper/lower</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Upper extremity involved</td>
<td>60.8 (578/951)</td>
<td>23.3 (17/73)</td>
</tr>
<tr>
<td>Lower extremity involved</td>
<td>39.2 (373/951)</td>
<td>72.6 (53/73)</td>
</tr>
<tr>
<td>Upper and lower extremity involved</td>
<td>0</td>
<td>4.1 (3/73)</td>
</tr>
<tr>
<td>Left/right</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Right</td>
<td>51.5 (402/780)</td>
<td>47.4 (37/78)</td>
</tr>
<tr>
<td>Left</td>
<td>48.5 (378/780)</td>
<td>48.7 (38/78)</td>
</tr>
<tr>
<td>Bilateral</td>
<td>0</td>
<td>3.8 (3/78)</td>
</tr>
<tr>
<td>History of trauma</td>
<td></td>
<td></td>
</tr>
<tr>
<td>None</td>
<td>10.6 (98/926)</td>
<td>8.3 (6/72)</td>
</tr>
<tr>
<td>Mild (contusion, sprain/strain)</td>
<td>32.2 (298/926)</td>
<td>62.5 (45/72)</td>
</tr>
<tr>
<td>Severe (fracture, post-surgical)</td>
<td>57.2 (530/926)</td>
<td>29.2 (21/72)</td>
</tr>
</tbody>
</table>

Table 2  Signs and symptoms of children and adults with complex regional pain syndrome I (modified from Tan et al. [4])

<table>
<thead>
<tr>
<th>Signs and Symptoms</th>
<th>Adults, % (n/n_ev)</th>
<th>Children, % (n/n_ev)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Inflammatory</td>
<td>99.9 (950/951)</td>
<td>97.4 (76/78)</td>
</tr>
<tr>
<td>Pain</td>
<td>93.3 (887/951)</td>
<td>82.1 (64/78)</td>
</tr>
<tr>
<td>Difference in skin color</td>
<td>77.5 (737/951)</td>
<td>39.7 (31/78)</td>
</tr>
<tr>
<td>Edema</td>
<td>90.9 (864/951)</td>
<td>87.2 (68/78)</td>
</tr>
<tr>
<td>Difference in skin temperature</td>
<td>44.9 (427/951)</td>
<td>71.8 (56/78)</td>
</tr>
<tr>
<td>Unexplainable limited range of motion</td>
<td>90.1 (857/951)</td>
<td>62.8 (49/78)</td>
</tr>
<tr>
<td>Increase of complaints after exercise</td>
<td>82.3 (763/927)</td>
<td>70.5 (55/78)</td>
</tr>
<tr>
<td>Neurological</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Hypesthesia</td>
<td>75.4 (674/894)</td>
<td>46.2 (36/78)</td>
</tr>
<tr>
<td>Hyperpa</td>
<td>80.8 (705/873)</td>
<td>52.6 (41/78)</td>
</tr>
<tr>
<td>Dyscoordination</td>
<td>47.3 (365/771)</td>
<td>23.4 (18/77)</td>
</tr>
<tr>
<td>Tremor</td>
<td>43.6 (371/850)</td>
<td>22.1 (17/77)</td>
</tr>
<tr>
<td>Involuntary movements</td>
<td>28.5 (212/744)</td>
<td>23.1 (18/78)</td>
</tr>
<tr>
<td>Skeletal muscle spasm</td>
<td>21.7 (185/851)</td>
<td>21.4 (6/28)</td>
</tr>
<tr>
<td>Paresis</td>
<td>93.2 (670/719)</td>
<td>48.7 (38/78)</td>
</tr>
<tr>
<td>Pseudoparalysis</td>
<td>17.5 (152/867)</td>
<td>18.2 (14/77)</td>
</tr>
<tr>
<td>Myoclonus</td>
<td>8.1 (51/630)</td>
<td>14.1 (11/78)</td>
</tr>
<tr>
<td>Atrophy</td>
<td>40.2 (347/864)</td>
<td>7.8 (6/77)</td>
</tr>
<tr>
<td>Skin</td>
<td>26.1 (216/827)</td>
<td>11.7 (9/77)</td>
</tr>
<tr>
<td>Nails</td>
<td>25.9 (200/771)</td>
<td>6.5 (5/77)</td>
</tr>
<tr>
<td>Subcutaneous tissue</td>
<td>45.8 (374/816)</td>
<td>32.5 (25/77)</td>
</tr>
<tr>
<td>Skeletal muscle</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Sympathetic signs and symptoms</td>
<td>42.3 (343/810)</td>
<td>23.4 (18/77)</td>
</tr>
<tr>
<td>Abnormal sweating</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Complications</td>
<td>5.5 (24/440)</td>
<td>1.3 (1/77)</td>
</tr>
<tr>
<td>Chronic infection</td>
<td>47.2 (178/377)</td>
<td>6.5 (5/77)</td>
</tr>
</tbody>
</table>

n_ev = number of patients evaluated.

5. Low and co-workers describe the characteristics and diagnosis of 20 children who were diagnosed with CRPS over a 4-year period [3]. The results corroborate those of other investigators that pediatric CRPS involves primarily girls (90%) in later childhood and adolescents. The lower limbs are affected in 85% in which case it is mostly the foot. Minor trauma was generally responsible for the onset of the syndrome in 80% of their cases.

With approval of the hospital ethics committee, medical records were used to obtain demographic data, information regarding clinical presentation, causes of symptoms in children and adolescents and details of...
of cases. A corollary to this is that those children who are
delay in making a diagnosis with other adolescents was encouraged by attend-
they are less likely to have a normal scan or show hyperperfusion. The authors
100% of children with CRPS using the IASP diagnostic criteria and immediate
diagnosing pediatric CRPS. By the same token, normal bone scan does not exclude CRPS. As a prognostic aid,
remission, indicating a successful treatment outcome. In the forgoing papers
experience at the Cleveland Clinic, that early recognition of
unrecognized (10.6 vs 21.5 weeks, respectively). Late
diagnosis was also associated with a higher rate of hospitalization and included two patients in the series who
were lost to follow-up. Both patients were a symptomatic at 2 years. The authors note that bone scans in the adult
tend to show a diffuse hyperperfusion; this pattern is uncommon in pediatric CRPS. Children are more likely to
have a normal scan or show hyperperfusion. The authors conclude that diffusely abnormal findings may be helpful in
diagnosing pediatric CRPS. By the same token, normal bone scan does not exclude CRPS. As a prognostic aid,
a bone scan exhibiting hyperperfusion may be associated with more rapid symptomatic improvement. The authors
noted that 55% of their children were regarded as high achievers on their psychological profile. No pre-existing
psychiatric order was evident in any of the patients, but a recurring theme already mentioned is the coexistence of
family dysfunction, non-verbalization of feelings, and a lack of self-assertiveness. The authors observe that while significant
gains have been made regarding the awareness of CRPS in children, there remains widespread ignorance of
its prevalence. Treatment of pediatric CRPS should always be associated with a good prognosis.

Conclusion

It is clear from the literature presented and our own experience at the Cleveland Clinic, that early recognition of
CRPS using the IASP diagnostic criteria and immediate institution of a physiotherapeutic/behavioral algorithm is essential to a successful outcome. In the forgoing papers
and those by Sherry, Lee, and Wilder, early mobilization with behavioral support can provide dramatic reversal of
the presenting signs and symptoms [12,13,29]. The speed of onset and salutary response to these measures under-
score what I have described as the plasticity of this syndrome in the adolescent and child. In fact given early
diagnosis and treatment, almost 100% of children should achieve a complete resolution of their syndrome notwithstanding the possibility that 20 to 30% will experience an
exacerbation of their symptoms. Therapy that is directed toward increasing function by both occupational and

| Physical therapy | 1 | 17 weeks | 64 weeks | 0 | 0 |
| Physical therapy and medication | 1 | 9 days | 15 weeks | 0 | 0 |
| Physical therapy and psychological | 5 | 17.6 (2–4 weeks) | 16.7 (4–25 weeks) | 0 | 1 |
| Physical therapy, psychological and medications | 13 | 12.7 (2 days to 41 weeks) | 11 (3 days to 26 weeks) | 4 | 7 |

Pharmacologic agents used were paracetamol, nonsteroidal antiinflammatory drugs and/or codeine. Adjuvant
therapy included tricyclic antidepressants (amitriptyline) or anticonvulsants (gabapentin). Only those patients who
have showed no improvement during their outpatient therapy or who had an exacerbation of their symptoms
were admitted. Admission was only used to gain therapeutic control and to provide more intensive exercise
therapy. A typical schedule consisted of attending school, hydrotherapy, physical therapy and gym sessions. Social-
ization with other adolescents was encouraged by attending groups and independence was fostered by giving the
children skills in areas such as self care.

The age of onset varied between 8 and 18 years. 80% of children reported a traumatic episode. Of interest is the fact that 4 children (20%) recalled no precipitating event and 3 of these reported waking up in the morning with their symptoms.

The presenting signs were decreased range of motion and allodynia in 100% of children, skin color changes in 90%,
and temperature asymmetry and edema in 85%.

The authors emphasize that the delay in making a diagnosis of pediatric CRPS is still occurring in a large number of cases. A corollary to this is that those children who are
diagnosed early (less than 3 months) will achieve an early remission compared with those whose condition is at first unrecognised (10.6 vs 21.5 weeks, respectively). Late
diagnosis was also associated with a higher rate of hospitalization and included two patients in the series who
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diagnosis and treatment, almost 100% of children should achieve a complete resolution of their syndrome notwithstanding the possibility that 20 to 30% will experience an
exacerbation of their symptoms. Therapy that is directed toward increasing function by both occupational and
physical therapeutic measures will result in the reestablishment of function in the affected limb. Behavioral measures and desensitization are both helpful to alleviate the fear of movement to allow physical measures to become effective. Although most children responded to purely physical and behavioral measures when utilized in a coordinated fashion, there are those cases in which allodynia, either cutaneous or deep is so severe no effective physical measures can be applied. It is in this situation that one must accept the fact that some form of analgesia or interventional measure should be employed to facilitate therapy and maintain momentum toward a complete remission. To withhold such help from the few whose symptoms are so severe, will not only jeopardize their recovery, but may cause disenchantment with the medical community, which should be otherwise able to resolve their condition. These children may then proceed to develop irreversible secondary and degenerative changes that involve not only integumentary tissues, but also peripheral neuropathology and vasculopathy in the affected extremity or body region. Although there is an approximate 10% incidence of extension of CRPS to involve another extremity in the adult, this migratory incidence is not well documented in children. There is anecdotal evidence that delayed treatment in the adult may enhance the possibility of CRPS being expressed at a new site, it would seem prudent not to delay all possible measures to contain and achieve a clinical resolution of the syndrome in children. A comprehensive coordinated multidisciplinary approach for the management of CRPS in the adolescent and child will provide the best results in the majority of children.

Acknowledgment

Acknowledgments to Gerald A. Banez, PhD, Program Director, Cleveland Clinic Pediatric Pain Rehabilitation Program, for providing additional descriptive information about the program and preliminary outcomes data.

References


