OUTCOMES OF A SIMPLE TREATMENT FOR COMPLEX REGIONAL PAIN SYNDROME TYPE I IN CHILDREN

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ABSTRACT

Background: Chronic Regional Pain Syndrome type I (CRPSI) in children is a disorder of unknown etiology. No standard diagnostic criteria or treatment exists. Published treatment protocols are often time and resource intensive. Nonetheless, CRPSI is not rare and can be disabling. This reports the results of a simple and inexpensive treatment protocol involving no medicines, nerve blockades, physical therapy resources or referrals to pain specialists. The patient is instructed in a self-administered massage and mobilization program. The diagnosis required allodynia (pain on light touch of the skin) and signs or the history of signs of autonomic dysfunction.

Methods: A chart review of patient coded for "reflex sympathetic dystrophy" or 'autonomic dysfunction" was performed yielding a cohort of eighty-three patients treated by a common protocol. Most patients were identified in the last 15 years. Most patients with this CRPSI were doubtless coded simply as "foot pain" or "knee pain", etc and were not identified in this search. Charts were reviewed for patient demographics and outcomes. A subset of patients filled out the Pediatric Outcomes Data Collection Instrument (PODCI) giving a validated pre-treatment disability measure.

Results: The cohort characteristics were similar to prior reports with respect to age, gender, location, and history of trauma. Of the 26 patients who completed the PODCI before treatment the Pain/ Comfort Core Scale score mean was 20.81(0-63). The Global Functioning Scale score mean was 52.11(27-83.5). Eighty-nine percent of 51 patients who attended clinic until their outcome was definite had no or minimal residual pain. Treatment averaged 2.2 visits per patient, typically over a six-week period.

Conclusions: A simple, inexpensive protocol can be effective in treating CRPSI in children. The protocol is risk free, inexpensive to families and conservative of physician and physical therapy resources.

Level of Evidence: Therapeutic Level IV.

INTRODUCTION

Chronic Regional Pain Syndrome (CRPS) in children, like that in adults, is presently divided in Types I and II. CRPS I (previously called reflex sympathetic dystrophy) has no direct injury to peripheral nerves. CRPS II (commonly called causalgia) has clear evidence of peripheral nerve injury.

Little progress has been made in understanding and treating Chronic Regional Pain Syndrome Type I (CRPSI) in children over the last 30 years, since the first reports of this disorder appeared in the orthopaedic literature[1-6]. The etiology remains unknown and the optimal treatment is unclear. Average length of time from symptom onset to diagnosis continues to be measured in months[3,4,7,8,9]. Many unnecessary diagnostic investigations are performed prior to diagnosis. This limited progress has occurred in spite of significant attempts by the International Association for the Study of Pain (IASP) to define diagnostic criteria for use in research and clinical diagnosis[10]. The most recent consensus definition for clinical diagnosis in 2004 from the IASP is summarized as follows:

1) Continued pain that is out of proportion to the inciting event.
2) A history of three of the four of the following symptoms or signs:
   - Sensory: Hyperesthesia or allodynia.
   - Vasomotor: Temperature asymmetry or skin color changes or skin color asymmetry.
   - Sudomotor/edema: edema or sweating changes or sweating asymmetry.
   - Motor/ Trophic: decreased range of motion or motor dysfunction or trophic changes.

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3) Presence of at least one of the following signs in two or more of the following categories:
   Sensory: hyperalgesia or allodynia.
   Vasomotor: Temperature asymmetry or skin color changes or skin color asymmetry.
   Sudomotor/edema: edema or sweating changes or sweating asymmetry.
   Motor/ Trophic: decreased range of motion or motor dysfunction or trophic changes.

The diagnosis is excluded by the existence of conditions that would otherwise account of the degree of pain and dysfunction.

The criteria used in this study for the diagnosis of CRPS I predated the various consensus recommendations but have similar elements. The criteria used by the senior author are 1) presence of allodynia or dysesthesia; 2) signs of autonomic dysfunction or a history of signs of autonomic dysfunction; and, 3) an absence of pathological process to explain the pain.

The natural history of CRPS I in children is generally reported to be more benign than the adult version in that most patients are eventually relieved of their symptoms without suffering permanent atrophy or joint contracture[2,4,5,7,11,12,13]. However, a 2009 study investigating the quality of life of forty-two adults who had childhood CRPS I found that 52% of the patients had pain in the affected limb at a mean 12 year follow up[8].

A consensus is developing that the primary effective treatment is intensive physical therapy, although "intensive therapy" has been poorly defined and its efficacy has not established in a rigorous fashion[2,5,6,7,11,13,14,15,16]. Most authors feel that behavioral counseling of the patient and/or family of some sort is critical to the expedient and effective resolution of symptoms[2,6,7,11,13,14,15,16,17]. Most treatment programs are quite intensive in terms of family and medical practitioner commitments of time and resources.

This is a case series report of patients with CRPSI evaluated and treated by a program developed in 1987 by the senior author. A series of 5 cases treated by this method was reported in 1989[1]. This report is a much larger series of patients treated in the same manner over the last 25 years. We attempted to assess the pre-intervention disability in these children and the effectiveness of treatment using the Pediatric Outcomes Data Collection Instrument (PODCI), which is a validated pediatric orthopaedic outcomes instrument.

The treatment program consists of a patient directed program of mobilization and massage after a detailed description of the problem and validation of the patient's pain is accomplished during the initial clinic visit. There is no external physical therapy involved. The parents are not involved. It usually requires 1-2 visits at three-week intervals until sufficient symptom resolution has occurred so that no further visits are necessary. The underlying philosophy of the treatment program is that RSD in children is a "mind (brain)-body" problem. It is, therefore, best addressed by having the patient take responsibility for his/her own body. Any external interventions are seen by the body as invasions that tend to entrench rather than relieve the symptoms and signs of CRPSI. The written instruction sent home with the patients are in Table I.

Table I.

Treatment Plan for Reflex Sympathetic Dystrophy

1) Massage 3-5 times/day
   2 minutes firmly
   1 minute lightly
   2 minutes firmly
2) Focus on the body part that is cousing you pain and talk to it while massaging.
3) Always keep it moving, never let it be still.
4) Use it! Walk, run, jump and play as soon as possible.

**Remember, it will hurt worse at first but it will get better!

METHODS

To identify patients, the patient billing database was searched for patients treated by the senior author and given the diagnosis of CRPSI in the time from 1987-April 2010. ICD9 codes for Autonomic Nerve Disorder NEC(337.9) and Reflex sympathetic dystrophy (upper and lower limb, 337.21, 337.22) were used to search the patient database. The search identified 83 patients who met the inclusion criteria. All patients were 18 years old or less and they met the senior author's criteria for the diagnosis of CRPS I. These criteria were: 1. Presence of allodynia or dysesthesia; 2. Signs of autonomic dysfunction or a history of signs of autonomic dysfunction; and, 3. An absence of pathological process to explain the pain. The records of these 83 patients were then reviewed further to evaluate demographic data, presenting symptoms and outcomes. PODCI scores were assessed when available.

RESULTS

Eighty-three patients with the diagnosis of CRPSI were identified. The age at the time of diagnosis ranged from 6-17 years (mean 12.5). Sixty-eight were female and 15 male. The mean time of presentation from onset of symptoms was 7.0 months (range 3 days to 4 years). An onset associated with a minor traumatic injury was identified in 46 patients (55%). Thirty-one patients (37%) had no precipitating injury. Five patients had recent surgery, and one patient gave a history of a prior stress fracture. The

Table 2. Number of patients found through record search by time period.

1980's	3 patients
1990's	10 patients
2000-2004	23 patients
2005-2009	20 patients
2010-2013	27 patients

lower extremity alone was involved in 80 patients, upper extremity alone in 2 patients, and both upper and lower extremities involved in 1 patient. Of patients with lower extremity symptoms, the foot or ankle was involved in 61 patients and the knee was affected in 17 patients. One upper extremity case involved the forearm with the other 2 cases involved the hand and wrist.

All patients were noted to have allodynia or dysesthesia on examination in the clinic as this was required for diagnosis Tache cérébrale was only positive if seen in clinic. Physical exam findings of other sings of "autonomic" dysfunction were positive if noted to be present on examination or positive by history. Tache cérébrale was positive in 40 patients, negative in 3, and not mentioned in 40. Swelling was positive in 50, negative in 19, and not mentioned in 14. Temperature change was positive in 42, negative in 17, and not mentioned in 24. Color change was positive in 58, negative in 10, and not mentioned in 15. Range of motion was normal in 45, decreased in 17, and not mentioned in 21. Of the 81 patients with lower extremity involvement, 49 were walking without assistance at the time of clinic evaluation, 27 were unable to walk without crutches, and in 5 walking status was not mentioned.

Prior to presenting for evaluation 15 (18% of the patients) patients had bone scans. Five showed increased uptake, one showed decreased uptake, and nine were normal scans. Thirty-eight patients (46% of study group) had an MRI prior to diagnosis. Five MRI's had abnormalities including one osteochondral defect; one nonspecific edema in the calcaneus, medial cuneiform, and tibialis anterior insertion; two possible stress fractures; and one enhancement within the cuboid. Six patients had a CT scan, with all being negative. Eighty-one of the patients (98%) had radiographs with four showing abnormal findings including two osteochondral defects and two fractures. None of these radiographic findings were believed to correlate with the patients' symptoms. Additional evaluations included Neurology consults (1), Rheumatology consults (2), Pain clinic consult (2), ECG (1),Vascular studies (2), and EMG/NCV's (1).

Prior to evaluation at our clinic, 40 patients had received treatment that involved casting or fracture boot immobilization. Twenty-two patients had undergone physical therapy. Fifteen patients were started on NSAID's. Eight had TENS units trials. Three had arthroscopy. Two patients each had local injections, narcotics, oral steroids, acupuncture and lumbar sympathetic blocks. One patient each had a Bier block, amitriptyline, and ultrasound treatments. Four patients had been treated with gabapentin.

Pediatric Outcomes Data Collection Instrument (PODCI) was administered to 26 patients at the initial visit of which 24 forms were complete. The two patients with incomplete PODCI's were missing the Transfer and Basic Mobility Core Scale score. The mean Upper Extremity and Physical Function Core Scale score was 92.96 (87-100) in those patients with lower extremity complaints. The one patient with upper extremity complains had a score of 50. The Transfer and Basic Mobility Core Scale mean was 64.63 (21-95). The Sports and Physical Functioning Core Scale score mean was 26.35 (0-77). The Pain/Comfort Core Scale score mean was 20.81(0-63). The Happiness Core Scale score mean was 66.15 (10-100). The Global Functioning Scale score mean was 52.11(27-83.5).

Of the 83 patients initially diagnosed with RSD, 62 (75%) attended their scheduled return appointment in three to four weeks after the initial visit. Improvement was noted in 55 of 62 (89%) of returning patients with 18 patients (29%) reporting complete resolution of pain. Seven patients (11%) reported no relief. Three of the unimproved patients admitted to not following the suggested protocol.

Ultimately, only 51 of 83 patients were followed until symptoms had resolved or the treatment failed. Of the entire cohort 51patients (62%) had complete resolution of symptoms (47 patients) or minimal pain with no limitations (four patients). Six patients (7%) had continued, limiting pain. Twenty-six patients ((31%) have uncertain outcomes because they failed follow-up after one or two clinic visits. Of the patients followed-up with certainty, 89% had good outcomes with no limitation and no or minimal pain and 11% failed treatment. Of the failures, one patients showed initial improvement followed by multiple recurrences and was referred to rheumatology with resolution of symptoms 6 months later; one patient showed no improvement and was referred to anesthesia pain clinic and was treated with regional IV sympathetic blockade, TENS unit, tricyclic antidepressants, and calcium channel blockade without relief and last documentation noted 6 years later to have continued intermittent problems; four patients continued to have symptoms at their last visit and were subsequently lost to follow-up.

Four successfully treated patients (10%) suffered a recurrence of symptoms that was treated in the same

Table 3.Presence of signs of CRPS I (all could be by history except Allodynia and Tache Cerebrale)

	YES	NO	NOT MENTIONED
Allodynia	100%		
Tache Cérébrale	48%	4%	48%
Swelling	60%	23%	17%
Temperature Changes	51%	20%	29%
Color Changes	70%	12%	18%
Decreased ROM	21%	54%	25%

manner either at a clinic visit or recommended by phone. In one patient the recurrence was successfully treated with the same method followed by another recurrence in the opposite extremity to be treated in the same manner without further follow-up. The remaining three cases were instructed to treat as before without any follow-up on their outcomes.

For the entire cohort, there was an average of only 2.2 follow-up visits per patient (range 0-6 visits) and the average duration of treatment was 6.25 weeks (range 0-40 weeks)

DISCUSSION

Patients presented in this report displayed similar demographic characteristics to other studies of children and adolescents with CRPS I[2,7,9,11,13,14,15,18]. The patients were predominantly female (female:male, 4.5:1) and the lower extremity was most frequently involved (98%). This extremely high percentage of lower extremity patients may be due to the fact that children's upper extremity problems are seen by hand surgeons at our institution. Prior to the onset of symptoms, patients suffered minor trauma (55%) or no trauma (37%).

The typically recommended treatment for CRPSI in children is intensive physical therapy coupled with individual and/or family psychological therapy. In 1992 Wilder reported on seventy patients treated with multiple modalities. Over one-half of the patients received combined treatment with physical therapy (91%), NSAIDS (71%), transcutaneous electrical nerve stimulation (87%), psychological therapy (63%), tricyclic anti-depressants (59%), and sympathetic block (53%). With a median 3 year follow up period, 46% of these patients had persistent symptoms and less than half had returned to sports.[2] In 1999, Sherry, et al. reported outcomes of 103 children, one-half of whom were followed for more than 2 years, who were treated by a daily program of 4 hours of aerobic exercises, 1-2 hours of hydrotherapy, and desensitization[11]. Seventy-seven percent were referred for psychological counseling. Ninety-two percent became pain free with this program and 88% remained symptom free at more than 2 years. Lee et al. in 2002 randomized 28 children to weekly physical therapy versus trice weekly physical therapy, each program lasting 6 weeks.[14] The specific therapy sessions were not standardized and were very variable. Both groups underwent cognitive-behaviors therapy in addition. Ten of the 28 patients were reclassified as CRPS II but were continued in the study. Outcomes were not different between patients with one versus three physical therapy sessions per week. Ten of the 28 patients failed to obtain relief, either initially or on recurrence and required lumbar sympathetic and continuous lumbar epidural infusions. In 2007 Low et al. described 20 children treated with intensive physical therapy, psychological assessment and intervention (mostly cognitive behavior therapy), with most receiving amitriptyline or gabapentin[7]. All but 2 had resolution of symptoms with a mean time of 15 weeks.

The value of both physical therapy and psychological therapy has been questioned in recent years. A 2012 systematic review concluded that the evidence for the effectiveness of physiotherapy children with CRPS I is based on a small volume of poor-to-fair quality evidence.[18] Rarely is the physical therapy prescribed standardized or even described in most reports. The authors further noted that all studies combined other treatment interventions such as medications and psychological treatment, as well as the physiotherapy. The authors concluded that it was not possible to determine whether physiotherapy alone was effective in treating this population. A 2013 study assessed whether children and adolescents with CRPS I had more underlying psychological problems than children with other types of chronic pain[20]. They compared 101 children and adolescents with CRPS I with 103 children with chronic abdominal pain, 291 children with chronic headaches, and 119 with chronic backache. The overall psychological functioning of CRPS I patients was not different from children suffering these other types of chronic pain. This argues against a primary underlying psychological cause for CRPS I, but does not suggest that addressing psychological stress might not be important in treating the disorder.

While treatment of CRPS I with intensive physical therapy, psychological therapies and ad hoc adjunctive medicines have been shown to be effective, they can be costly and disruptive to families due to the time commitment and expense involved. Our protocol was effective in 89% of the patients who attended follow-up clinic in 6 weeks or less. The program is designed and required to be performed by the patient without involvement of any other services or resources.

The report is the first of our knowledge to present PODCI scores in patients diagnosed with CRPSI. The

Table 4. Final Outcome

Complete resolution of symptoms:	47 (57%)
Return to full activity with minor pain:	4 (5%)
Total good outcomes:	62 %
No F/U after 1st or 2nd appointment:	26
Uncertain outcomes:	(31%)
Continued pain:	6
Poor Outcomes:	(7%)

poor scores for transfer and mobility (65), sports and physical functioning (26), pain (21), happiness (66) and global function (52) documents the debilitated condition the patients are in at time of presentation. Although our PODCI data is limited, it shows that patients with this disorder are severely physically disabled and provides a standard for comparison for future studies. The PODCI forms were completed by less than half of the patients in the study. There were no PODCI scores available from the return visits to clinic.

There are several limitations to this current report. The retrospective nature of data collection depends upon the accuracy of data in the patient's record. Many signs or symptoms of CRPS I were not mentioned as either being present or absent. This cohort is certainly only a fraction of the CRPS I patients seen at our institution. Most CRPSI patients would be coded as "foot pain" or "knee pain" as this is a simpler diagnosis to find than "autonomic nerve dysfunction" and "reflex sympathetic dystrophy" which were used to identify this cohort. The number of patients coded with the nonspecific "pain" in a region of the body is so great as to be prohibitive in the amount of time required to find CRPS I patients coded as such. From January through June of 2013, the senior author personally coded all CRPS I patients. Eleven patients presented with CRPS I during that 6-month period. This suggest that we identified somewhat less than 1/3 of CRPS I patients from 2000 to 2013. There is no reason to suspect that the patients identified were different from those not identified except that is seems likely that more difficult patients who returned more frequently were more likely to be coded as "reflex sympathetic dystrophy" than those who were seen once or twice with a chief complaint of "foot pain". A significant further limitation is that thirty-one percent of the patients failed to return for follow up after the initial or second clinic visit and their symptom status is not known. It is possible that some did well and chose not to return. It is also possible that if the protocol did not work, the patients chose not to return and/or found other treatment.

Strengths of the paper are a well-defined protocol for treatment. A fairly large cohort for this disorder was reviewed. The senior author's practice location is very limited in other option for pediatric orthopaedic care, suggesting that many or most of the patients lost to followup did not seek care elsewhere.

The data presented in this report shows that a patient-directed program for treatment of CRPSI can be effective. If so, the reason that this approach works is unclear. It is possible that CRPS I is, in fact, a "mind-body" problem that the protocol addresses. It is also possible that this is just a "poor man's" physical therapy program. Furthermore, it is possible that the senior author's strong confidence in the effectiveness of the protocol is communicated to the patient and is of itself, therapeutic. Finally, if the protocol itself is therapeutic, it is unclear which elements of the protocol are critical, e.g. mobilization vs massage vs talking to the painful limb during massage vs constant motion. Nonetheless, we have found this to be an effective and resource conservative treatment.

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