MSc in Clinical Management of Pain

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ASSIGNMENT #1

Part 1: Management of CRPS in Children

STUDENT NAME & ID NUMBER:

George Boghozian, S1122670

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**INTRODUCTION**

Complex Regional Pain Syndrome (CRPS) is characterized by severe, non-dermatomal, chronic pain, vasomotor/sudomotor changes, and a movement disorder. It can be preceded by a noxious event, though the presentation is often disproportionate to the inciting injury. It has been recognized, studied and reported in adults for many years. However, it is a less acknowledged disorder in children, probably due to under-diagnosis [1]. CRPS has two types:

1. CRPS I (also called reflex sympathetic dystrophy or RSD): No nerve damage exists with this type.
2. CRPS II (also called causalgia): This produces similar symptoms after a nerve injury has occurred.

Although CRPS is an unusual diagnosis in general paediatrics, it is well recognised by paediatric rheumatologists [2]. Children have the greatest chance of achieving remission and therefore, left untreated in children, the symptoms of CRPS can become chronic, spread to other parts of the body, and persist for years. The earlier it is recognized and treated, the better the prognosis [1].

The presentation and response to treatment of CRPS differs in children as compared with the adult form. The purpose of this paper is to re-consider the case of Mrs. Walsh from a paediatric perspective (a 10-year-old child) and to highlight these differences.

**DEMOGRAPHICS**

A population based study [3] did not find any patients under age 10 and only three patients, one male and two females, in the 10 to 19 age group. There are only a few case reports of children in the preschool years [4,5]. Even though brachial plexus injury during delivery is common and can lead to longstanding motor weakness, neonates with Erb’s palsy do not generally develop pain in the extremity [8]. Children with diabetes have highest incidence of CRPS as compared to non-diabetic children [6]. Girls are affected 4-6 times more often than boys [7]. Whereas adults frequently present with symptoms in their upper extremities, in children, lower limbs are affected 5 times more often than upper ones [8].
**PATHOPHYSIOLOGY & ETIOLOGY**

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 [9]. Although pathophysiology of CRPS is poorly understood, many features, particularly the neurologic abnormalities, suggest both peripheral and central nervous system involvement [9]. Figure 1 [10] shows a simplified diagrammatic view of pathogenesis of CRPS.

![Figure 1: Mechanism of Complex Regional Pain Syndrome](image-url)
While a pathophysiological mechanism for CRPS remains unknown, number of etiologies have been proposed: a peripheral small fiber neuropathy, an exaggerated regional inflammation (neurogenic inflammatory pain involving small nerve fibers), and autonomic (sympathetic) dysfunction [11-14]. 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 [9,15]. Historically, psychological factors have been invoked to explain the genesis and persistence of CRPS in children, but the evidence is not compelling [8]. Cruz et al. (2011) suggest that children with CRPS-I may be at elevated risk for multiple pain-related symptoms, emotional distress, and attentional difficulties [16].

**DIAGNOSIS**

The diagnosis of CRPS in children is similar to adults. It remains a clinical one based on appropriate findings in the history and physical examination [8,17]. IASP diagnostic criteria [18] for CRPS are:

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.

Although there are no laboratory tests that can absolutely confirm or exclude diagnosis of CRPS, sensory changes, most commonly cold allodynia, have been documented using quantitative sensory testing (QST) [19]. The main role of radiological investigations is to rule out other underlying pathology [8].

**TREATMENT**

Similar to adults pediatric CRPS is also a biomedically multifaceted syndrome, frequently associated with psychosocial components that are additional pivotal diagnostic features (and thus, treatment targets) [20]. As such, any unimodal approach usually fails, and multiple modalities/disciplines are required to optimize pain management, functional rehabilitation, and quality of life [21,22].
CRPS in children has been widely held to be intrinsically different from that in adults due to a perception that children may have a milder course or a better response to conservative treatments [8]. Sporadic early reports of children with RSD (reflex sympathetic dystrophy, as it was known at the time) suggested spontaneous resolution [23-25]. This led to the suggestion that no treatment should be performed for children with CRPS. The rationale was that all treatments carry risks and side effects, and for a self-limited disease these should be avoided. Other authors used treatment strategies very similar to those used in adults, including sympathetic blocks, antidepressants, vasodilators, steroids, and so forth, generally with complete resolution of the disease [26]. Between these two extremes was a group of authors recommending conservative treatment consisting primarily of physical therapy (PT) either with [27, 28] or without [29] concomitant use of transcutaneous electrical nerve stimulation (TENS).

**Physical Therapy:** Active rehabilitation exercises tend to be the primary mode of treatment in children, with the vast majority recovering and managing relapses with this treatment modality. Limited treatment outcome studies [2, 29, 30] indicate that children generally respond well to PT, particularly aqua therapy (exercise in water) component of it [31]. It is thought that exercise works by inhibiting the sympathetic stimulation, increasing circulation, and releasing endorphins [30, 32]. TENS is one of the best non-invasive physical modalities for management of pain, and it has been described in CRPS case reports [33, 34].

**Psychological/Behavioural Therapy:** Psychological treatments including biofeedback, relaxation methods, self-hypnosis, and distraction techniques have been found to be very beneficial and even a necessary part of management for this age group [35, 36].

**Pharmacotherapy:** If needed, medications with a proven track record in neuropathic pain management can be added, such as the tricyclic antidepressants (nortriptyline or amitriptyline) or anticonvulsants (gabapentin and others) [8]. The use of corticosteroids in children has shown very little benefit and, because of the potential side effects, should be avoided in this population [37].
**Procedural Interventions:** Some studies report that sympathetic blocks are rarely effective in children, while others have found that they can be very successful. An epidural block to break the cycle of pain and allow for initiation of PT is sometimes required in severe cases [2, 37].

**CONCLUSION**

The general perception that children have a milder course may relate to the potentially greater willingness of children to actively participate in appropriately targeted treatment rather than to innate differences in the disease process itself [8]. Effective treatments of children with CRPS have shown to be much different than those successful in adults. In adults, aggressive pharmacologic intervention has been the standard. In children, it appears that the more conservative approach is often adequate, and therefore pharmacotherapy and invasive procedural interventions are rarely necessary. Clinical judgment dictates the extent of medication or interventional therapy added to the treatment to facilitate rehabilitation [8].

Therefore, the treatment plan for a 10-year-old kid (compared to 55-year-old Kay Walsh) would mainly focus on a combined PT and psychological/behavioural approaches without use of pain medications and anti-inflammatory agents. As immobilization, disuse, and fear of movement are common features of CRPS, the reversal of this operant movement phobia (“kinesiophobia”) provides an example of the need for coordinated co-treatment of modalities; in this example, PT and psychological/behavioural therapies [20]. This can be achieved by educating the patient about concept of “hurt vs. harm”, and by demonstrating to them that the consequences of movement are not always totally negative and/or painful.

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**References:**


