Correction of Severe Foot and Ankle Contracture Due to CRPS Using External Fixation and Pain Management: Report of a Pediatric Case

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Complex regional pain syndrome–induced dystonia is a severe deformity that can affect the lower extremities and hinder ambulation. Although a number of conservative treatments have been described for this condition, we are not aware of any publications describing the use of surgical intervention for the treatment of this condition. In this report, we describe the case of a pediatric patient with a severe lower extremity deformity in conjunction with chronic pain syndrome. A concerted, interdisciplinary treatment approach was undertaken for the management of this patient, and this included contributions from a foot and ankle surgeon, a pain specialist, an internist, a physical therapist, and a psychiatrist. The primary goal of the treatment strategy was to recreate a plantigrade, weight-bearing lower extremity, while controlling pain. To this end, gradual correction of deformity was achieved using an external fixator and, by 6 months after the surgery, the patient was for the first time in years ambulating on the realigned lower extremity. After 3 years of follow-up, she maintained an activity level that was equal to that which she enjoyed before the injury. Level of Clinical Evidence: 4 (The Journal of Foot & Ankle Surgery 47(5):434–440, 2008)

Key Words: botulinum, complex regional pain syndrome, CRPS, deformity correction, dystonia, equinovarus, external fixation

Complex regional pain syndrome (CRPS) Type I, previously known as reflex sympathetic dystrophy (RSD), is a chronic pain disorder of the extremities. The clinical signs and symptoms may include (but are not limited to) burning pain, edema, hyperpathia, allodynia, trophic changes, and functional impairment. The diagnostic criteria for CRPS Types I and II are purely clinical, as indicated by Figure 1 (1), and can be missed even by experienced clinicians. The median age for patients diagnosed with CRPS in the United States was reported to be 41.8 (range 18–71) years, whereas the mean age at time of the inciting injury was 37.7 (range

Complex Regional Pain Syndrome Type I

- The presence of an initiating noxious event, or a cause of immobilization
- Continuing pain, allodynia, or hyperalgesia with which the pain is disproportionate to any inciting event.
- Evidence at some time of edema, changes in skin blood flow (skin color changes, skin temperature changes more than 1.1°C difference from the homologous body part), or abnormal sudomotor activity in the region of pain
- This diagnosis is excluded by the existence of conditions that would otherwise account for the degree of pain and dysfunction

Complex Regional Pain Syndrome Type II

- The presence of continuing pain, allodynia, or hyperalgesia after a nerve injury, not necessarily limited to the distribution of the injured nerve
- Evidence at some time of edema, changes in skin blood flow (skin color changes, skin temperature changes more than 1.1°C), or abnormal sudomotor activity in the region of pain
- This diagnosis is excluded by the existence of conditions that would otherwise account for the degree of pain and dysfunction.

FIGURE 1 Differentiation of CRPS Types I and II according to Merskey and Bogduk[1].

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Financial Disclosure: None reported.

Conflict of Interest: None reported.

1067-2516/08/4705-0002$34.00/0

doi:10.1053/j.jfas.2008.05.010

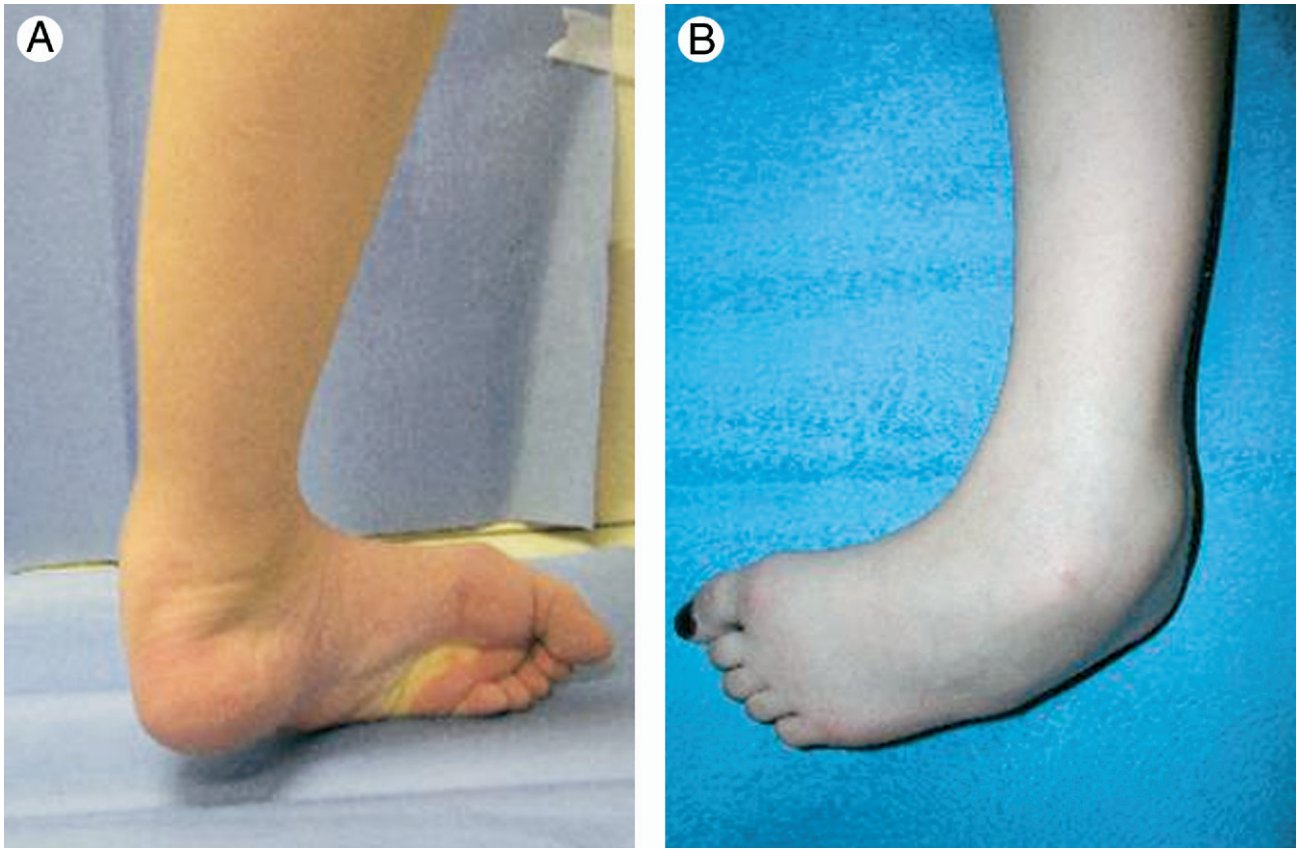

FIGURE 2 Clinical presentation shows the triplanar equinovarus position of the foot and ankle. A. Posteromedial view. B. Anterolateral view.

14–64) years, and the female to male ratio was 2.3:1 (2–4). One of the debilitating symptoms of CRPS is dystonia. In the lower extremities, an equinovarus deformity of the foot and ankle is the most common manifestation of CRPS-induced musculoskeletal contracture (5). Because of the progressive nature of this symptom, immediate treatment is usually necessary to prevent the development of severe deformity. Treatment options for CRPS-induced dystonia include serial casting, aggressive physical therapy, sympathetic ganglion blocks, and adjunctive botulinum toxin injections (6–16). Following a systematic review of the Medline, Excerpta Medica, and the Cumulative Index of Nursing and Allied Health databases, we were unable to identify a reference to the use of gradual correction of the deformity by means of surgical intervention. The patient underwent tendon lengthening, botulinum toxin injections, nerve releases, external fixation application, and spinal cord stimulator implantation. The purpose of this report is to describe the case of a pediatric patient with CRPS-induced dystonia, who was treated surgically with the use of external fixation in order to gradually correct a severe equinovarus contracture. This article also discusses the importance of a multidisciplinary approach to this disease process, including perioperative pain management provided by pain medicine specialists, as well as contributions from a physical therapist, neurosurgeon, and psychiatrist.

Case Report

A 14-year-old girl presented to The Foot and Ankle Institute of Western Pennsylvania on a referral from a pain management specialist at the Western Pennsylvania Hospital. The patient developed a progressive equinovarus deformity of the left lower extremity after she sustained an ankle sprain while diving into third base while playing softball. Within 4 months, she had developed severe equinovarus deformity and was unable to ambulate with weight on her left lower extremity. During an approximately 1.5-year period before presentation to our practice, the patient had undergone serial casting, physical therapy that included contrast baths, and she used an ankle-foot orthosis, under the guidance of the previous treating physician. She had also been manipulated under general anesthesia and casted in an effort to reduce the deformity, however this caused such a severe exacerbation of her pain that she had to be admitted into intensive care and the cast removed. Because of the severe nature of the pain syndrome and the resultant equi-

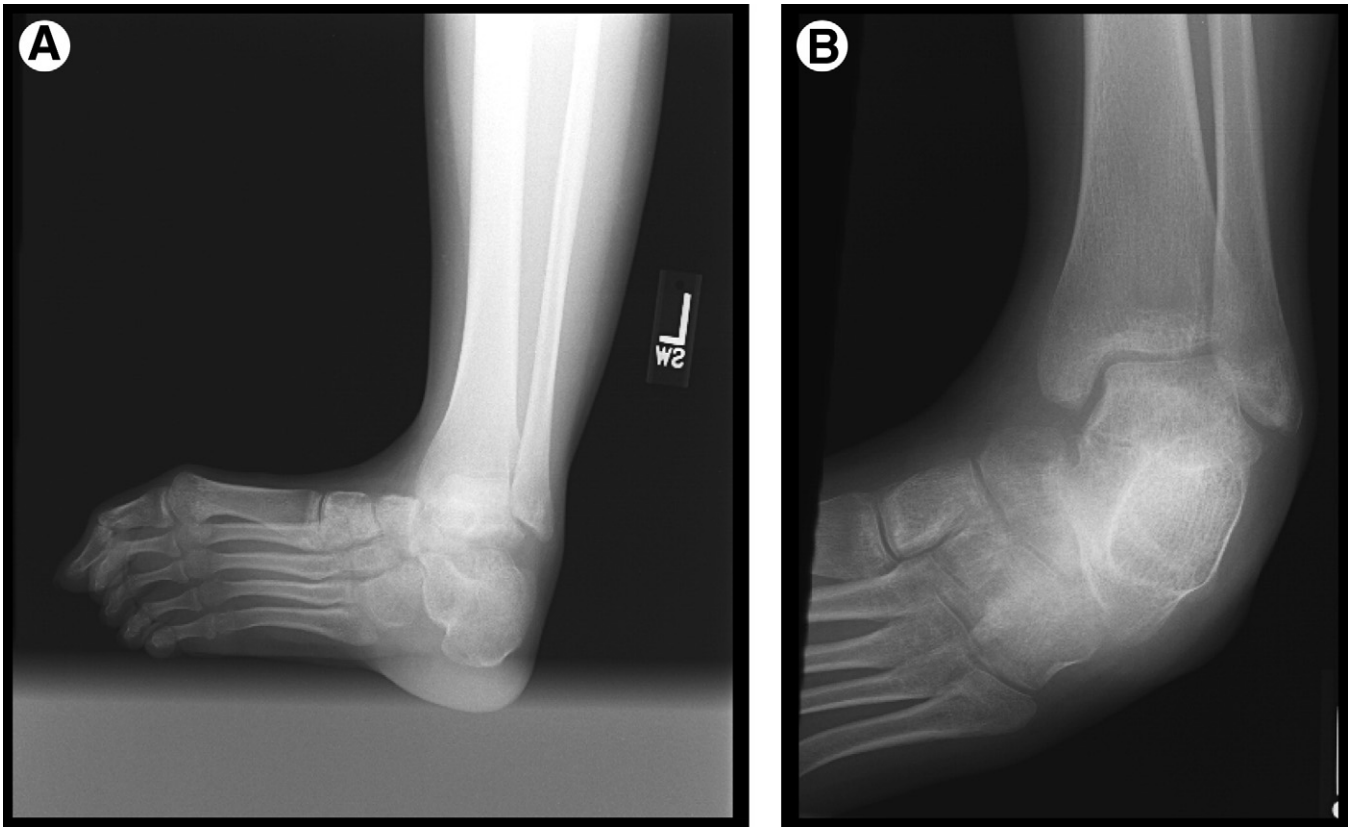

FIGURE 3 Radiographs of the left ankle demonstrate the severity of the deformity and multiple joints involved in the contracture. A. Anterior view. B. Mortise view.

novarus deformity, serious consideration was given to below-the-knee amputation. At the time of the referral to our practice, the patient was under the care of a physiatrist, neurologist, psychiatrist, pain management specialist, and a physical therapist. Moreover, she was receiving lumbar sympathetic ganglion blocks under fluoroscopic guidance, and these resulted in limited periods of partial pain relief.

The physical examination revealed a left lower extremity that was considerably cooler than the contralateral side, and the skin on the left side demonstrated diminished digital hair growth and atrophy. She also exhibited allodynia and hyperesthesia involving the left foot, ankle, and leg. Musculoskeletal examination demonstrated a rigid equinovarus contracture involving the mid and hindfoot, as well as the ankle, with little to no range of motion due to antalgic guarding. Furthermore, the posterior muscle group overpowered the anterior and lateral leg muscles (Figure 2). Radiographic images supported the clinical findings with evidence of an equinovarus deformity of the hindfoot and ankle and an adducted position of the forefoot (Figure 3). Additional nerve conduction velocity and electromyographic studies showed no evidence of any other neurological conditions.

Surgical treatment was carefully planned with cooperation between the foot and ankle surgeon, pain specialist, internal medicine physician, physical therapist, and the psychiatrist. The patient was admitted the day before surgery and had an indwelling epidural catheter placed, through which ropivacaine and fentanyl were administered under the guidance of the pain medicine service. This was maintained throughout the patient's hospital course. Left lower extremity reconstructive surgery was undertaken the following day.

Because of the nature of the contracture and associated dysfunction of the anterior and lateral muscle groups, common peroneal nerve and posterior tibial nerve decompressions were performed. Additionally, botulinum toxin was injected into all of the posterior leg muscles in the region or their respective neuromuscular junctions, in an attempt to relieve spasticity and to assist in the subsequent gradual correction of the deformity. The gastrosoleus contracture was the most severe, and the Achilles tendon was lengthened by means of an open technique, although the ankle was left in a position of slight equinus in an effort to avoid placing excessive traction on the musculature of the deep posterior compartment of the leg. An Ilizarov apparatus

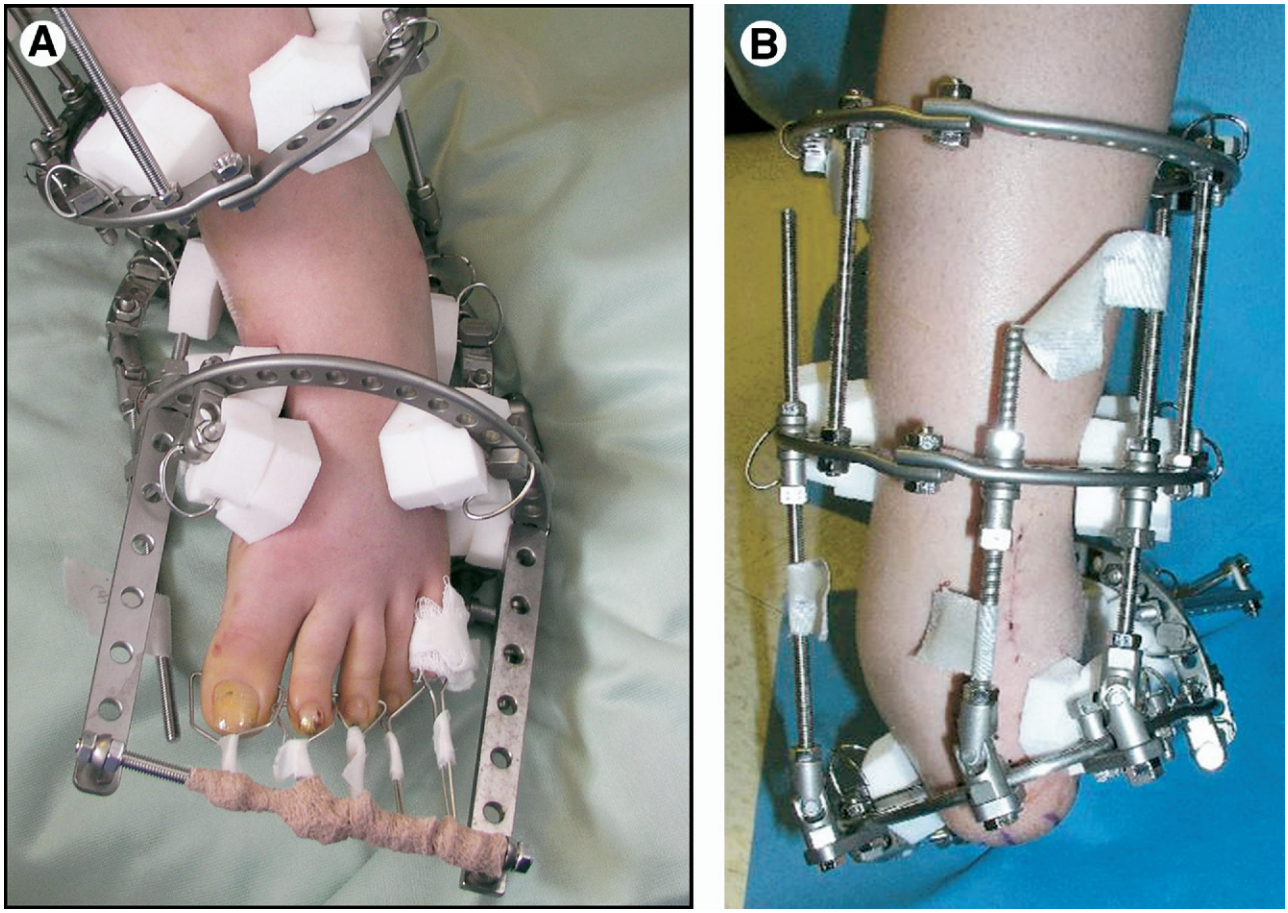

FIGURE 4 Immediate postoperative views of the left lower extremity demonstrate the frame designed to addres the severity of the deformity and the multiple segments that needed to be addressed during the correction. Note that use of pin fixation of the digits helped to prevent contractures distally during the correction. A. Anterior view. B. Posterior view.

consisting of a distal tibial block, a half ring at the calcaneus, and a 5/8 ring over the forefoot, was then applied. These were held with the appropriate smooth wire fixation, and then tensioned utilizing Ilizarov tensioning principles. The rings were connected with universal hinges and lengthening rods in an unconstrained fashion so that slow, postoperative adjustments could be made to correct the deformity (Figure 4).

The patient's early postoperative course included continuous infusion of the epidural block, and daily adjustment of the external fixation frame to correct the equinovarus deformity. Immediately following application of the external fixation frame, the pain management specialists continued to administer anesthesia throught the epidural. With the adequate resolve of the acute post-operative pain, a neurosurgeon implanted a spinal stimulator in an effort to provide long lasting pain relief. Satisfactory correction was gradually achieved over the next 6 weeks at the following rate: medial posterior = 8 mm/day, central posterior = 5 mm/day, lateral posterior = 3 mm/day, lateral foot = 2 mm/day, and medial foot = 5 mm/day. The lateral foot distraction was performed to prevent crushing the cuboid. After the deformity was fully corrected, the frame was left static for an additional 6 weeks, at the end of which the frame was removed in the operating room and additional botulinum toxin injections were administered to all of the posterior leg muscles. In total, the frame was on for 3 months. Upon removal of the external fixator, the patient was casted in the corrected position for an additional 2 weeks. The patient began aggressive physical therapy, including ankle range of motion, leg muscle strengthening, and stretching, once the cast was removed. For 6 months, she also used an ankle-foot orthosis that blocked plantarflexion and aided in the prevention of recurrent contracture. The remainder of the postoperative course proceeded without incident, and after more than 3 years of postoperative follow-up, she was ambulating well with no pain in the extremity (Figure 5). She also continued to perform physical therapy modalities in an attempt to prevent recurrent ankle and foot contractures, and she kept biannual visits with her pain specialist for neuromodulation therapy, and with our service for surveillance of her left lower extremity.

FIGURE 5 Follow-up clincial non weightbearing views demonstrate the plantigrade position of the foot and the realignment of the digits. A. Anterior view. B. Medial view.

Discussion

Dystonia is a well-documented clinical entity that may hinder ambulation and, particularly in pediatric patients with CRPS, may lead to progressive equinovarus deformity despite adequate pain control. Although a wide range of treatment options exist for the nonsurgical management of musculoskeletal contracture (6–15), including serial casting, physical therapy/manipulation, and injection of botulinum toxin, these methods may not be adequate for some severe deformities. Moreover, acute surgical correction of these deformities can be problematic because of perioperative pain issues that can be very difficult to manage. Gradual correction using an external fixation device, combined with adjunctive procedures such as isolated tendon lengthening and/or botulinum injections, may enable the establishment of a plantigrade foot in a less acutely traumatic fashion and, as such, may be less stressful to the patient and minimize the risk of exacerbation of the pain syndrome (17–29).

Furthermore, the use of prophylactic neurovascular releases can also be useful adjunct procedures to the surgical correction of severe deformity of the lower extremity. The rationale behind this concept hinges on the idea that prophylactic removal of any anatomical structures that may lead to strangulation or impingement of neighboring anatomical structures will decrease the likelihood of subsequent neurological damage and/or ischemia as the deformity is gradually corrected. This concept is particularly applicable to the posterior tibial nerve and its branches in the tarsal tunnel, and the common peroneal nerve near the head of the fibula, in association with the correction of a wide range of lower extremity deformities (20–23). In the case described in this report, we implemented this concept by means of prophylactic release of the neurological components of the tarsal tunnel.

Injection of botulinum toxin is a common adjunct intervention used for the treatment of skeletal muscle spasticity (12–16). We used this treatment in an effort to alleviate contractures of the posterior leg musculature, which appeared to be overwhelming the other muscular compartments of the leg. Botulinum toxin, however, does not usually cause complete resolution of all spasticity, and this was the case in our patient. As such, we felt that an open lengthening of the Achilles tendon would be a beneficial adjunct to the relief of posterior leg contracture in this particular patient. To avoid placing a sudden and excessive tensile load on the musculature of the deep posterior compartment of the leg, a process that could potentially exacerbate CRPS (27), the Achilles tendon was left in slight equinus. It was felt that the small amount of residual equinus would be adequately corrected gradually by means of adjustment of the external fixation frame.

The external fixation was mounted in the standard block construct with smooth wires. However, the segments were

connected with universal hinges and lengthening rods so that gradual correction could be obtained over a period of 6 weeks, after which the device was maintained static for an additional 6 weeks. Overall, the frame was left on for approximately 3 months in the patient we described. In general, when correcting a contracture, the external fixator is left on for a total of twice the amount of time it takes to correct the deformity. Typically, the first half of this period of external fixation is used to obtain the desired position of correction, and the second half is used to allow the tissues to adapt to the new alignment. In certain cases, it may be desirable to slightly overcorrect the extremity, however this was not the case with the patient described in this report. As an example of this time sequence, if it would take 2 weeks to correct a deformity, then the fixator would be left in place for a total of 4 weeks. If it takes 10 weeks to obtain the corrected position, then the fixator would be left in place for and additional 10 weeks (24–28). As a general rule, deformity correction without osteotomy and/or arthrodesis conveys a high rate of recurrence. For this reason, we feel that postcorrectional maintenance using appropriate orthoses and bracing, as well as ongoing physical and occupational therapy, is just as important as the surgical correction itself. For the patient described in this report, aggressive physical therapy was employed for almost a full year following removal of the external fixation.

Although the epidural block achieved pain relief throughout the perioperative period, our patient ultimately required long-term management with the use of a spinal cord stimulator. The successful use of spinal column stimulation in patients with CRPS is well documented (30, 31). In the case described in this report, it was not until after the neurosurgeon implanted the spinal column stimulator that the patient was capable of undergoing manipulation, physical therapy, and rehabilitation. Moreover, as was the case with our patient, ongoing physical therapy and the periodic use of lumbar sympathetic ganglion blockade can also be helpful in the postoperative phase.

The importance of a multidisciplinary approach to this complex clinical problem cannot be overstated. Pain management by pain specialists, surgery by a qualified foot and ankle surgeon, aggressive physical therapy, and evaluation of the psychiatric health of the patient, need to be coordinated to obtain therapeutic success (29, 32–34). This concept is supported by Singh et al (33), who reported an increase in functional outcome in CRPS patients who had simultaneous treatment by the pain management physician, psychologist, physical therapist, and occupational therapist. Additionally, we feel that strong family support is also important, especially in pediatric patients suffering with dystonia secondary to CRPS. Our experience with the patient described in this report leads us to believe that a thorough understanding of the complexity of the disorder, as well as an appreciation of the treatment plan, on the part of the parents is essential to achieving a successful outcome.

In conclusion, this case demonstrated that CRPS-induced dystonia could be treated using an external fixation device for gradual correction. However, perioperative pain management, physical therapy, and the expertise of other medical specialists was also important in regard to this patient's successful outcome. We believe that the successful management of this type of patient requires constant communication with providers from other medical disciplines, and that such treatment is best undertaken at a tertiary care facility that has the capacity to provide the appropriate level of expertise.

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