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Trials and Tribulations: Interventional Management for Recurrent Complex Regional Pain Syndrome in Children

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Stem Case and Key Questions Content

A 15 year old female presents to your pediatric pain clinic from out of state for second opinion regarding reflex sympathetic dystrophy (RSD) diagnosed by her local orthopedic surgeon several years ago. Conventional over-the-counter analgesics were initially of modest benefit but no longer provide relief; a brief course of acetaminophen-hydrocodone was discontinued by the patient secondary to perceived excessive somnolence threatening her straight-A average. The patient is an avid runner, and wishes to participate in her upcoming state high school cross-country championships next month. The patient is brought to clinic by her father, a well-educated business executive; her mother is at home undergoing chemotherapy for recurrent breast cancer, any discussion of which makes the patient tearful.

KEY QUESTIONS: What is reflex sympathetic dystrophy (RSD), and how does it differ from complex regional pain syndrome (CRPS)? What are the current Budapest criteria for formal diagnosis of CRPS? Are these criteria an improvement over earlier criteria? How does the presentation of CRPS in children differ from that in adults? What signs and symptoms are most helpful in making the diagnosis? How should the patient who does not meet formal criteria for diagnosis be classified and managed?

The patient is a pale, thin white female wearing her school uniform and running shoes who speaks very quietly and makes little eye contact. She walks into clinic and moves about the exam room without apparent difficulty, but reports 10/10 pain in her legs below the knees. She is able to remove her socks and shoes readily, but winces and reports extreme discomfort with any examination or manipulation of her legs. Her feet are cool and slightly mottled without sudomotor changes, atrophy, or muscle weakness. She is without other neurologic deficits.

KEY QUESTIONS: What else do you need to know? Should anti-neuropathic analgesics be tried at this point? Anti-convulsant, anti-depressant, or both? Which ones? Should the patient be in formal mental health care? Occupational/physical therapy? Should these be prerequisites for further care in your clinic? Is interventional management reasonable at this point, given the chronicity of her condition?

The patient eventually admits to seeing a counselor and occasionally a psychiatrist for feeling sad, but is reluctant to describe herself as being depressed; she is currently taking a selective serotonin reuptake inhibitor (SSRI), and declines other anti-depressant for fear of untoward side

effects. She agrees to continue formal mental health care, but declines formal occupational/physical therapy, feeling she works far harder in cross-country. Over several months, sequential trials of gabapentin and pregabalin prove ineffective; both medications are associated with significant perceived somnolence. The patient finishes the cross-country season, but returns to your clinic several weeks later with worsening symptomatology. Her mother has recently suffered a setback in her oncologic care.

KEY QUESTIONS: Is interventional management more or less appropriate at this point? Are there predictors of positive or negative response? If she is interested in interventional management, what block should you perform? Lumbar epidural? Selective lumbar sympathetic block? Unilateral or bilateral? Local anesthetic, steroid, alpha-blocker, adjuvants? How many blocks? How often?

Three successive technically successful bilateral lumbar sympathetic blocks each provide 12-24 hours of partial analgesia, following which her symptomatology returns unchanged. She continues to see her counselor and psychiatrist, who have continued her SSRI; the patient continues to decline other anti-neuropathic anti-depressants and anti-convulsants. She continues to decline formal occupational/physical therapy. The family has researched several multi-disciplinary in-patient pediatric pain programs at various nationally prominent children's hospitals, but is reluctant to travel such distance, preferring to continue care in your clinic. After numerous requests from the patient and her family, you eventually agree to schedule her for tunneled lumbar epidural catheter placement, and to admit her to the pediatric hospitalist service at your facility.

KEY QUESTIONS: Why a tunneled lumbar epidural catheter? Why admit her to the pediatric hospitalist service? While she is in the holding area awaiting her procedure, her insurance declines authorization for the admission: what now? After obtaining insurance approval, you place a technically successful tunneled lumbar epidural catheter. What will you administer? Who will provide coverage of the epidural? What other services should see her while she is an in-patient? How long should she stay?

Continuous epidural infusion of local anesthetic and clonidine without opioid or demand dose is initiated with prompt onset of significant but incomplete analgesia. The patient is admitted for five days, and is evaluated by the pediatric hospitalist service, occupational/physical therapy, adolescent psychology, and adolescent psychiatry. She receives several hours daily of occupational/physical therapy, as well as counseling and reinforcement of established psychiatric care. Her father arranges to have an exercise bicycle brought to the patient's room, and times the sessions he insists she do above and beyond her occupational/physical therapy; he remains with the patient throughout her hospitalization. The mother is again experiencing a setback in her oncologic care, and remains at home out-of-state. The epidural is removed and the patient discharged Friday afternoon: she still reports significant albeit incomplete analgesia, and is given detailed instructions for further rehabilitation services and mental health care. The father telephones your clinic 90 minutes later while still on the interstate driving home to report the patient's pain has returned to its previous level, and asks if he should bring her back to your facility.

KEY QUESTIONS: What went wrong? Should you have used a different epidural infusion? A different regional anesthetic technique? Kept the patient in the hospital longer? Managed the

family differently? Never blocked or admitted her at all? Is there any benefit to further interventional management? Could this have been predicted? What is the likely long-term prognosis?

Model Discussion Content

Complex regional pain syndrome (CRPS) is a disorder characterized by characteristic pain in an area of the body (usually a limb, although CRPS has been described in the face as well as other regions) as well as symptoms of autonomic dysfunction. CRPS may occur after a localized process (burn, infection, sprain, fracture, surgery), in the setting of a more generalized condition not seemingly anatomically related to the affected area (pregnancy, stroke, myocardial infarction), or without apparent underlying etiology.

Two subtypes of CRPS have been identified: type 1 and type 2. CRPS type 1, previously known as reflex sympathetic dystrophy (RSD), denotes patients without identifiable major nerve injury. Type 1 comprises approximately 90% of patients with CRPS, occurs more commonly in females, and appears to have increased incidence in Caucasians.[1] CRPS type 2, previously known as causalgia, refers to patients with identical symptomatology in the setting of an identifiable major nerve injury. First described after battlefield amputations in the American Civil War, CRPS type 2 has no apparent gender or ethnic predilection.

The first description of a pediatric patient with CRPS was published in 1971. Pediatric CRPS appears to differ from CRPS in adults in several ways, although the underlying pathophysiology is thought to be generally similar. Pediatric patients are more likely to have type 1, and accordingly less likely to report an identified inciting event. The lower extremity is more commonly affected in children, and skin temperature tends to be cooler. Neurologic manifestations are less common in children, whereas psychological issues play a more prominent role. Perhaps because of the more functional nature of pediatric CRPS, children tend to be more responsive to occupational/physical therapy and mental health care, and less often require anti-neuropathic medication or interventional management. It is difficult accurately to determine the incidence of CRPS in children. Although there seems to be a progressive increase in reported cases, this could be due to improved recognition of the entity, particularly among primary providers. Pediatric CRPS is far more common in girls (90% of cases), with a mean age at diagnosis of 13 years.[2]

Children with CRPS are often described as having an incongruent affect, “la belle indifference”: they will appear cheerful while endorsing a pain score of 10/10. Psychological disorders are commonly associated with CRPS in children, although it is unclear whether such conditions are causal, secondary, or both. Conversion symptoms including non-epileptic seizures, blindness, paralysis, and muscle spasms have been described in children with CRPS.

The International Association for the Study of Pain (IASP) has developed criteria for formal diagnosis of CRPS. CRPS type 1 was initially categorized as having four components: a history of inciting event or immobilization; continued pain, allodynia, or hyperalgesia out of proportion to the inciting event; evidence of edema, skin blood flow changes, or abnormal sudomotor activity in the region of pain; exclusion of other conditions that would explain observed symptomatology. In CRPS type 2 the inciting event included an identifiable major nerve injury, although pain was not necessarily limited to the distribution of the injured nerve. These initial criteria proved

cumbersome and unwieldy in clinical practice; four groups of symptomatology commonly noted in CRPS became the basis for the Budapest Research criteria currently used for formal diagnosis of CRPS. Budapest criteria include:

1. Continuing pain disproportionate to any inciting event
2. Report of at least one symptom from each of the following four categories:
a. Sensory (hyperesthesia, allodynia)
b. Vasomotor (temperature asymmetry, skin color change)
c. Sudomotor/edema (sweating, edema)
d. Motor/trophic (weakness, decreased range of motion, changes in hair, nails, or skin)
3. Display of at least one sign from at least 2 of 4 of the above categories
4. Exclusion of other diagnosis explaining symptoms

Report of at least one symptom from at least 3 of 4 of the above categories is sufficient for clinical diagnosis. Differential diagnoses could include trauma, inflammatory conditions such as juvenile idiopathic arthritis or osteomyelitis, and malignancy. There are no laboratory or imaging studies required for diagnosis of CRPS, although such tests may help exclude other diagnoses. Once a formal diagnosis of CRPS is made, it generally thought prudent to limit further diagnostic evaluations.

Occupational/physical therapy and mental health care are the mainstays of treatment for pediatric CRPS, with the primary goal of functional restoration rather than pain reliefs per se. Patients often need to develop skills for maintaining function in the context of continued pain. Pharmacologic therapy with anti-neuropathic analgesics including antidepressants and anticonvulsants has been used, as have various regional anesthetic techniques including particularly selective sympathetic blockade. Some pediatric practitioners advocate for physical and behavioral rehabilitation without medication or interventional management:[3] pharmacologic therapy and interventional management have not been rigorously evaluated for management of CRPS in children, and are not without potential side effects and procedural risks. There is general agreement early initiation of rehabilitative therapy probably increases likelihood of timely functional recovery, with or without pharmacologic agents and interventional procedures.

Although pediatric CRPS often responds well to occupational/physical therapy and mental health care, some patients do not respond to such conservative treatments, and recurrence of symptomatology is not uncommon. Continuous peripheral nerve blocks have been used for analgesia and to facilitate intense rehabilitation.[4-7] Spinal cord stimulators have been used with some success for severe refractory CRPS [2] when other measures have failed. Some patients fail all efforts at interventional management, making ongoing management highly challenging.

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