

COMPLEX REGIONAL PAIN SYNDROME (CRPS)

What is it?

Complex regional pain syndrome (CRPS) is a chronic and progressive neuro-inflammatory disorder characterized by pain disproportionate to the severity of the actual injury or stimulus. The pain cannot be explained by the initial trauma. CRPS most often affects the arms, legs, hands, or feet. The cause of CRPS is not fully understood but research supports a dysfunctional interaction between the central and peripheral nervous systems and/or an inappropriate inflammatory response.

There are a few differences in pediatric presentation of CRPS compared to adults. A majority of pediatric patients are girls between the ages of 8-16 years with lower limbs most commonly affected, compared to the upper extremities of adults. Children with CRPS generally have intact cognitive function but they can experience a higher rate of somatic symptoms and emotional distress.

CRPS can be classified into 2 different types. CRPS type I is more common and develops after an identifiable event such as a minor trauma, but without any significant nerve damage. In contrast, type II CRPS reflects the presence of a nerve injury, like that from a penetrating trauma or surgical intervention. CRPS can be further divided into stages. Some researchers argue these stages are not sequential and should be used more commonly as guidelines. People can experience symptoms of multiple stages concurrently, briefly, or not at all.

What are the symptoms?

The main feature of CRPS is amplified and unrelenting pain in response to a stimulus or touch. Pain often intensifies over time and can spread throughout the affected region. In rare cases, pain can spread to the opposite extremity. Other signs and symptoms of CRPS include:

- Continuous, deep, aching pain
- Burning, shooting, stabbing, or stinging pain
- Excessive sensitivity to pain
- Pain from stimuli not normally painful (touch of shirt, wind on face)
- Sensitivity to temperature
- Abnormal swelling
- Joint stiffness, limited range of motion
- Muscle spasms, tremors
- Muscle weakness
- Changes in skin temperature
- Change in skin texture (shiny, thin)
- Change in skin color (blotchy, blue, purple, pale, red)
- Excessive sweating in the affected area
- Changes in hair and nail growth
- Insomnia

What causes it?

In many pediatric cases, CRPS is triggered by a clear history of minor trauma or injury. This includes injuries such as a sprain, twist, dislocation, fracture, soft tissue injury (burn, bruise), or surgical procedure. Even minor medical procedures have been a trigger. In other cases, no actual injury was identified. Pain may start in a small area like a finger or toe and then spread to affect an entire limb. In rare cases, pain can even travel to an opposite extremity. No specific gene has been identified but rare family clusters of CRPS have been reported.

What is the treatment?

While there is no cure for CRPS, remission is possible. Early recognition and timing of diagnosis can affect a favorable prognosis and restoration of function. Treatment is directed at the contributing factors and a multidisciplinary management approach can include a combination of pharmacotherapy, physical therapy, and psychotherapy aimed at improving quality of life. The goal of rehabilitation is to improve function and quality of life; physical, emotional, and social.



Kennedy Krieger Institute

The Specialized Health Needs Interagency Collaboration (SHNIC) program is a collaborative partnership between the Kennedy Krieger Institute and the Maryland State Department of Education.

Suggested school accommodations

Like many chronic pain conditions, CRPS is a developmental health concern that can interfere with daily functioning. These students are at an increased risk for missing school, withdrawing from activities and peers, and experiencing difficulties with both memory and concentration. Stress and anxiety can play a role in inducing or perpetuating symptoms of CRPS. Absence from school is common as pain is unpredictable and medical appointments are necessary. Children also miss school because the classroom setting can create stress related to learning difficulties or over-achieving tendencies. Supporting students with this condition in the school require educators and parents/guardian to work as a team. Some accommodations to consider for a 504/IEP could include:

- Modified school day
- Plan for absences and make-up work
- Modify the classroom and school environment to avoid high traffic and bumping into student (changing of classes, dismissal time)
- Avoid mobility stressors (classroom on different levels, distance between classrooms, crowded halls)
- Designated parking spot or drop off close to school entry
- Ergonomic classroom assessment (set up of work area, chairs)
- Adapted/modified PE or recess
- Modifications for school uniform due to sensitivity (if necessary)
- Breaks or rest period
- Safe, quiet space
- Copies of notes and lectures
- Oral versus written testing
- Use of technology for lecture, writing
- Extra set of books for home
- Emotional support
- Peer support system
- Educating peers (with child and family permission)
- Staff education/training as appropriate
- Emergency Evacuation Plan (EEP)

Specific health issues for Individualized Healthcare Plan

- Diagnosis including co-occurring diagnoses
- Current medication list including PRN medications, note side effects
- Review of current treatments
- Identify potential triggers for pain reaction (touch, light, sound, smell)
- Physical limitations or restrictions
- Use of heating pad
- Note environmental and classroom temperature parameters
- Communicate with school staff, parents/guardian, and provider any changes or concerns about the disease
- Emergency Care Plan(s) (ECP) related to medical needs in the school setting and staff education/training as appropriate for each

Resources & Manuals

Reflex Sympathetic Dystrophy Syndrome Association (RSDSA)

<https://rdsd.org/>

National Institute of Health-CRPS Factsheet

<https://www.ninds.nih.gov/Disorders/Patient-Caregiver-Education/Fact-Sheets/Complex-Regional-Pain-Syndrome-Fact-Sheet>

Kennedy Krieger Institute– Pain Rehabilitation Program

<https://www.kennedykrieger.org/patient-care/centers-and-programs/pain-rehabilitation-clinic-outpatient>