

Complex Regional Pain Syndrome

Overview

Complex Regional Pain Syndrome (CRPS) is an uncommon chronic pain condition that can affect the arms, legs, hands or feet of a patient, following an injury. Rarely, Complex Regional Pain Syndrome affects other parts of the body.

Complex Regional Pain Syndrome accounts for approximately 2% to 5% of adult pain clinic patients and up to 20% of paediatric pain clinic patients. The prevalence in Australia is unknown however, we do know that in adults, females are three times more likely to develop the condition than males.



Little is known regarding the cause and prognosis of Complex Regional Pain Syndrome. The severity of the patient's symptoms varies greatly, as does the duration of the condition. There is no simple cure and rather treatment is aimed at restoring function and movement to the affected limb.

Causes

The causes of Complex Regional Pain Syndrome are not very well understood. It is thought that Complex Regional Pain Syndrome is caused by an injury to, or an abnormality of, the peripheral and central nervous system. Many cases occur following a forceful trauma to the limb for example a crushing injury, penetrating injury, fracture or amputation. There is limited data to show that there may be a genetic connection and occasionally Complex Regional Pain Syndrome can develop without any known injury.



There are two types of Complex Regional Pain Syndrome. Patients who have not had a confirmed nerve injury are classified as having Complex Regional Pain Syndrome type 1 which was previously known as reflex sympathetic dystrophy syndrome. Patients who have had a specific nerve injury are classified as having Complex Regional Pain Syndrome type 2, previously known as causalgia. About 90% of people with Complex Regional Pain Syndrome have type 1.

Diagnosis and Symptoms

There is no specific diagnostic investigation for Complex Regional Pain Syndrome, rather the diagnosis is based on the clinical examination, and the patient's history and presenting symptoms. Investigations are often performed as a way of excluding other diagnoses.

Over the years the diagnostic criteria for this condition has changed, and in 2003 the International Association for the Study of Pain (IASP) attempted to provide a more helpful and robust diagnostic framework called the Budapest Criteria which, in the last decade, has become the most commonly used criteria for the diagnosis of Complex Regional Pain Syndrome.



International Association for the Study of Pain (IASP)– Budapest Criteria

1. Continuing pain, which is disproportionate to any inciting event.
2. Must report at least one symptom in each of the following four categories:
 - Sensory: Reports of hyperalgesia and/or allodynia.
 - Vasomotor: Reports of temperature asymmetry and/or skin colour changes and/or skin colour asymmetry.
 - Sudomotor/Oedema: Reports of oedema and/or sweating changes and/or sweating asymmetry.
 - Motor/Trophic: Reports of decreased range of motion and/or motor dysfunction (weakness, tremor and dystonia) and/or trophic changes (hair, nails and skin).
3. Must display at least one sign at the time of evaluation in all of the following categories:
 - Sensory: Evidence of hyperalgesia (to pinprick) and/or allodynia (to light touch and/or temperature sensation and/or deep somatic pressure and/or joint movement).
 - Vasomotor: Evidence of temperature asymmetry and/or skin colour changes and/or skin colour asymmetry.
 - Sudomotor/Oedema: Evidence of oedema and/or sweating changes and/or sweating asymmetry.
 - Motor/Trophic: Evidence of decreased active range of motion and/or motor dysfunction (weakness, tremor and dystonia) and/or trophic changes (hair, nails and skin).
4. There is no other diagnosis that better explains the signs and symptoms.

Treatment

Because Complex Regional Pain Syndrome is not very well understood, effective treatment options are limited. However, treatment is aimed at restoring function and movement to the limb and may involve physical rehabilitation and physiotherapy, as well as analgesic and anti-inflammatory medication. As with any chronic pain condition, psychological intervention may also be useful in helping a patient adjust to their pain.

Prognosis

Some cases are mild and eventually go away, whereas others are more severe and the patient may never recover. In 2016, a longitudinal study found that within the first year, 70% of patients with Complex Regional Pain Syndrome improved, especially in the function of the extremity and the visible symptoms (oedema, skin colour and sweating). However, 25% of the patients still fulfilled the Budapest Criteria and only 5% were without complaints.

Glossary

Allodynia	When pain is caused by something that does not usually cause pain, like a sheet or pulling on a sock.
Dystonia	Involuntary movement contractions that cause twisting, repetitive and patterned movements, as well as abnormal postures.
Hyperalgesia	Abnormally increased sensitivity to pain.
Oedema	Fluid retention, swelling.
Sudomotor	Describes anything that stimulates the sweat glands
Trophic	Abnormalities in the area of pain that include primarily wasting away of the skin, tissues, or muscles, thinning of the bones and changes in how the hair or nails grow.
Vasomotor	Refers to actions upon a blood vessel which alter its diameter leading to changes in temperature.

LexiMed Consultants

⌘ **Dr Leigh Atkinson**
Neurosurgeon and Pain
Medicine Physician

⌘ **Dr David Shooter**
Orthopaedic Surgeon

⌘ **Dr Malcolm Wright**
Physician

Reference:

<https://www.painmanagement.org.au/>
<https://www.betterhealth.vic.gov.au/health/conditionsandtreatments/complex-regional-pain-syndrome-crps>
<https://www.mayoclinic.org/diseases-conditions/complex-regional-pain-syndrome/symptoms-causes/syc-20371151>
<https://brainfoundation.org.au/disorders/dystonia/>
<https://www.ncbi.nlm.nih.gov/medgen/866865>
<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5741324/>
<https://academic.oup.com/bjaed/article/17/8/262/3957912>
<https://www.nps.org.au/australian-prescriber/articles/complex-regional-pain-syndrome#r1>
Beerthuisen A, Stronks DL, Van't Spijker A, Yaksh A, Hanraets BM, Klein J, Huygen FJ: Demographic and medical parameters in the development of complex regional pain syndrome type 1 (CRPS1): prospective study on 596 patients with a fracture. Pain. 2012 Jun; 153(6):1187-92.