Complex Regional Pain Syndrome (CRPS)

Jian Zhang
Registered physiotherapist

Definition of CRPS

- CRPS is a neurologic syndrome characterized by
  - Pain and skin hypersensitivity
  - Vasomotor skin changes
  - Sweat disturbance/ edema
  - Motor disturbances
  - Various degrees of trophic change

- CRPS often follows a musculoskeletal injury, surgery, or immobilization.

- Type I: *without* evidence of major nerve damage = reflex sympathetic dystrophy syndrome
- Type II: *with* evidence of major nerve damage = causalgia

Bogduk N. *Curr Opin Anesthesiol*. 2001;14:541-546.

Harden RN. and Bruehl SP. Introduction and diagnostic considerations. *Complex Regional Pain Syndrome: Treatment Guidelines.* RSDSA press. 2006:1-11

Epidemiology

- Affects between 200,000 and 1.2 million Americans
- Age: common in younger adults
  - Mean age at diagnosis: 42 years old
- Females Vs males: ~ 2 - 3 :1

Raja SN, Grabow TS. Complex Regional Pain Syndrome I (Reflex Sympathetic Dystrophy) Anesthesiology. 2002; 96(5):1254-1260.

Reflex Sympathetic Dystrophy Syndrome Association (RSDSA). www.rsds.org

Cause of CRPS

- Cause: unknown
- A new study shows that a reduction in small-diameter nerve fibers is evident in CRPS-I. This finding of nerve damage could provide a specific physical trait, that clinicians could use in the future to help diagnose and measure the natural history of CRPS.

Oaklander AL, Rissmiller JG, Gelman LB, Zheng L, Chang Y, Gott R. "Evidence of focal small-fiber axonal degeneration in complex regional pain syndrome-I (reflex sympathetic dystrophy)." Pain, February 2006, Vol. 120, pp. 235-243.

Diagnosis of CRPS

- Diagnosis
  - Based on clinical signs and symptoms
  - Currently, no diagnostics that objectively indicate absence or presence of CRPS
  - Defining characteristic: "continuing pain that is disproportionate to any inciting event"

The Updated Diagnostic Criteria for CRPS

- Revised CRPS Criteria proposed by the Budapest IASP Consensus Group in 2004
- Two versions of the diagnostic criteria:
  - Clinical version: maximize diagnostic sensitivity with adequate specificity
  - Research version: more equally balance optimal sensitivity and specificity

Harden RN. and Bruehl SP. Introduction and diagnostic considerations. *Complex Regional Pain Syndrome: Treatment Guidelines.* RSDSA press. 2006:1-11

Clinical Diagnostic Criteria for CRPS

1) **Continuing pain, which is disproportionate to any inciting event**
2) **Must report at least one symptom in *three of the four* following categories:**

- *Sensory:* Reports of hyperesthesia and/or allodynia
- *Vasomotor:* Reports of temperature asymmetry and/or skin color changes and/or asymmetry
- *Sudomotor/Edema:* Reports of edema and/or sweating changes and/or sweating asymmetry
- *Motor/Trophic:* Reports of decreased range of motion and/or motor dysfunction (weakness, tremor, dystonia) and/or trophic changes (hair, nail, skin)

Clinical Diagnostic Criteria for CRPS Cont.

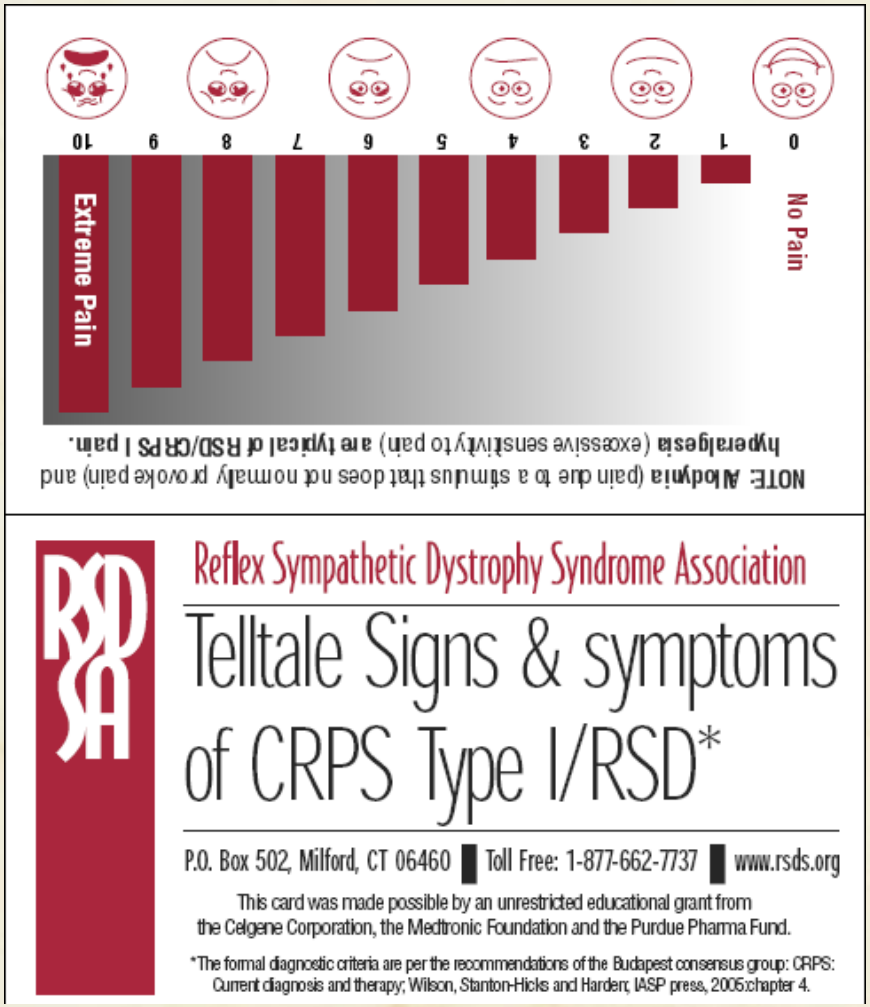
3) Must display at least one sign * at time of evaluation in *two or more* of the following categories:

- *Sensory:* Evidence of hyperalgesia (to pinprick) and/or allodynia (to light touch and/or deep somatic pressure and/or joint movement)
- *Vasomotor:* Evidence of temperature asymmetry and/or skin color changes and/or asymmetry
- *Sudomotor/Edema:* Evidence of edema and/or sweating changes and/or sweating asymmetry
- *Motor/Trophic:* Evidence of decreased range of motion and/or motor dysfunction (weakness, tremor, dystonia) and/or trophic changes (hair, nail, skin)

4) There is no other diagnosis that better explains the signs and symptoms

*A sign is counted only if it is observed at time of diagnosis.

Telltale Signs and Symptoms of CRPS Type I/RSD



Reflex Sympathetic Dystrophy Syndrome Association (RSDSA). www.rsds.org

CRPS I /RSD is a diagnostic consideration for patients who have pain (moderate to severe) that is disproportionate to any inciting event (sprain, fracture, etc.) and has some of the following characteristics:

- ✓ Pain is described as deep, aching, cold, burning, and/or increased skin sensitivity
- ✓ The presence of an initiating noxious event (sprain, fracture, etc.)
- ✓ Continuing pain (moderate to severe) associated with allodynia, or hyperalgesia.
- ✓ The pain is disproportionate to any inciting event.

- ✓ Abnormal swelling in the affected part
- ✓ Abnormal hair or nail growth
- ✓ Abnormal skin color changes
- ✓ Abnormal skin temperature (greater than 1°C assemtry)
- ✓ Abnormal sweating
- ✓ Limited range of movement, weakness, or other motor disorders (Paralysis, dystonia, etc)
- ✓ CRPS I /RSD is excluded by the existence of conditions that would otherwise account for the degree of pain and dysfunction

Swelling/Edema

http://www.singhealth.com.sg/Newsroom/Publications/Aescapulus/Complex+Regional+Pain+Syndrome.htm

Vasomotor Changes

Abnormal Sweating

Motor Disturbance

Psychological Distress

- CRPS is not a psychological syndrome, but people may develop psychological problems because of pain or when their complaints of pain are doubted
  - Anxiety
  - Depression
  - Fear
  - Anger

Raja SN, Grabow TS. Complex Regional Pain Syndrome I (Reflex Sympathetic Dystrophy) ***Anesthesiology*****. 2002; 96(5):1254-1260.**

Prognosis of CRPS

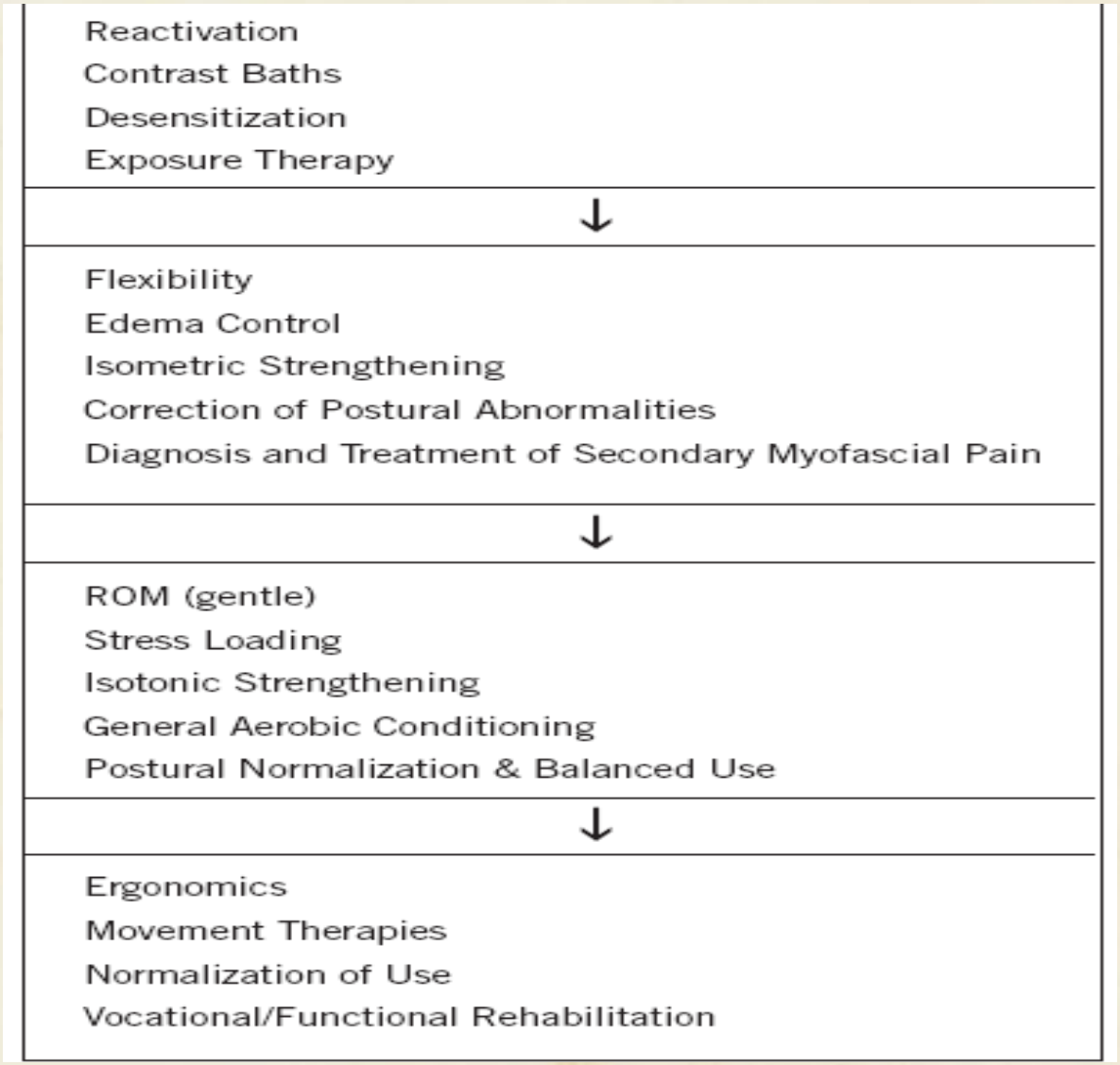
- Difficult to predict
- Earlier intervention may be more likely to be successful
- Some patients experience reduced symptoms or apparently full recovery (? No cure)
- Some patients get worse over time and experience significant disability
- May spread to other parts of the body

Raja SN, Grabow TS. Complex Regional Pain Syndrome I (Reflex Sympathetic Dystrophy) *Anesthesiology*. 2002; 96(5):1254-1260.

Treatment – Functional Restoration

- Currently, the ideal treatment methodology is a systematic interdisciplinary approach -- functional restoration
- Functional restoration emphasizes physical activity, desensitization and normalization of sympathetic tone in the affected limb, and involves a steady progression from the most gentle, least invasive interventions to the complete rehabilitation in all aspects of the patient's life.

A Stepwise, Functional Restoration Algorithm



.Harden RN, Swan M., Costa BR, et al. Interdisciplinary management. *Complex Regional Pain Syndrome: Treatment Guidelines.* RSDSA press. 2006:12-24..

Treatments of CRPS

- Medication
- **Physical therapy**
- **Occupational therapy**
- Psychological support
- Recreational therapy
- Vocational rehabilitation
- Sympathetic nerve blocks
- Spinal cord stimulation
- Surgery

Pharmacotherapy Guide

Symptoms/ Signs	Treatment
Mild-moderate pain	Simple analgesics/ nerve blocks
Severe/intractable pain	Opioids/ nerve blocks
Inflammation, edema	Steroids, NSAIDs
Depression, anxiety, insomnia	Sedatives, antidepressants, anxiolytics
Allodynia, hyperalgesia	Anticonvulsants, lidocaine patch
Osteopenia, immobility, trophic changes	Calcitonin, bisphophonates
Vasomotor disturbances	Calcium channel blockers, sympatholytics, and/or blocks

Harden RN. Pharmacotherapy. Complex Regional Pain Syndrome: Treatment Guidelines. RSDSA press. 2006:25-36 .

Interventional Pain Management

Minimally Invasive Therapies

- •Sympathetic / Somatic nerve blocks
- •IV Regional nerve blocks

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More Invasive Therapies

- •Epidural / Plexus Catheter Blocks
- •Neurostimulation
- •Intrathecal Drug Infusion

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Surgical Therapies

- •Sympathectomy
- •Motor Cortex Stimulation

Burton A. Interventional therapies. Complex Regional Pain Syndrome: Treatment Guidelines. RSDSA press. 2006:51-62.

PT and CRPS

- The goal of PT program is the gradual increase of strength and flexibility, principally through weight bearing.
- The goal is achieved through active or active-assisted means; it should encourage pacing and include rest breaks and relaxation techniques as well.

Reflex Sympathetic Dystrophy Syndrome Association (RSDSA). www.rsds.org

Harden RN, Swan M., Costa BR, et al. Interdisciplinary management. Complex Regional Pain Syndrome: Treatment Guidelines. RSDSA press. 2006:12-24..

Initial PT Session

- Patient education re: CRPS, the assessment and treatment process
- Listen to how they feel about themselves and their condition
- A thorough visual inspection of the affected area -- muscle atrophy, abnormal movement patterns, posture, edema and guarding
- Assessment: limited to gross assessment of AROM and functional movement, while noting restrictions in movement due to guarding or edema
- More formal measures for ROM, strength, coordination, and edema can be taken at a subsequent session.
- Help patients identify or reframe their goals, ie, less pain, improvement in function, the return of a more normal physical appearance

Reflex Sympathetic Dystrophy Syndrome Association (RSDSA). www.rsds.org

Devices for PT

- Upper extremity: foam rubber balls, spring-grip strengtheners
- Lower extremity: Swiss balls, foam rolls, and antigravity resistive equipment such as a Pilates reformer
- These devices help to gradually introduce a variety of weight-bearing/strengthening techniques.

Pilates Reformer

Mat Exercises

- Mat exercises provide strengthening of both the extremity and the postural muscles in a non-weight-bearing approach.
- Particularly valuable mat exercises include movement therapies such as the **Feldenkrais technique.**
- Feldenkrais teaches and encourages gentle, active motions within the patient's available range to increase body awareness and promote appropriate movement patterns.

Myofascial Pain Syndrome

- All patients with advanced CRPS will present with myofascial pain syndrome of the supporting joint.
- Aggressive treatment of this myofascial pain is a critical component of successful PT treatment.
- Hands-on techniques such as massage and myofascial release can sometimes offer effective relief from the myofascial pain.

TENS

- A TENS unit may be effective to decrease edema, sensitivity, and sympathetic response.

- The electrodes are placed over the peripheral nerve associated with the injury as this is where the majority of the sympathetics flow. Common placements are on the median and ulnar nerve distributions, for 30-45 min, 2-4 times /day depending on the patient's response.

- Active exercise can be performed while the TENS unit is being worn and may allow for increased tolerance to exercise.

Christine M. Kleinert Institute for Hand and Micro Surgery
http://www.cmki.org/LMHS/Chapters/35-ComplexRegionalPainSyndrome.htm

Other PT Modalities

- Electrostimulation modalities have demonstrated some efficacy
- Ultrasound therapy has been less effective
- Contrast baths: a controversial treatment option for CRPS
  - In mild cases of CRPS : can be beneficial to facilitate improved circulation in the affected extremity by alternating vasodilation with vasoconstriction.
  - In advanced cases: the immersion in the cold water may cause vasomotor changes.

(All physical therapies mentioned are level 3-4 evidence.)

Aquatic Therapy

- A good place to start therapeutic exercise
- Help decrease edema
- Introduce lower extremity weight bearing, especially useful for early restoration of functional activities such as walking
- Allow early participation in progressive PT, as nearly all exercises that are executed on land can be executed in the water
- Important to maintain water temperature, because excessively cold or hot water may temporarily exacerbate the CRPS.

Land-based Exercise Program

- The water activities need to be transitioned into a land-based exercise program to improve daily function.

Home Exercise Program (HEP)

- Initially, HEP focus on scrubbing/weight loading and desensitization techniques to engage the muscles in the affected area and manage pain
- HEP may be upgraded to increase range of active movement and strength.
- Functional activities: based on tasks that patients currently have difficulty performing, such as drinking from a cup with the affected hand or wearing a shoe on the affected foot. Do these for short periods of time frequently throughout the day to help incorporate the affected area back into routine activities.

Precautions of PT

- Inappropriately aggressive PT can trigger extreme pain, edema, distress, and fatigue, and may in turn exacerbate the inflammation and sympathetic symptoms of CRPS
- Use of assistive or ROM devices, prolonged application of ice, and inactivity may aggravate CRPS
- All PT must be executed within the patients' tolerance and never after a nerve block or with pronounced hypoesthesia
- Patient education:
  - Both exercising too much *and* exercising too little may trigger pain
  - To avoid physical stressors (ie, the stress of extended inactivity and bed rest on one extreme, and the stress of excessive exercise at the other).

OT and CRPS

- In general, OT should aim to normalize sensation, promote normal positioning, decrease muscle guarding, minimize edema, and increase functional use of the extremity in order to increase independence in all areas — work, leisure, and ADL.

Management of Edema

- Elevation
- Conventional TENS with gentle AROM
- Desensitization modalities
- Manual edema mobilization techniques such as manual lymph drainage, retrograde massage
- Specialized garments, bandaging, isotoner glove

Desensitization Techniques

- Superficial or subcutaneous desensitization techniques will aid in normalizing sensation to the affected area ie, rubbing with something soft, like silk, and progressing to rough or textured materials, like burlap

- Regular use of the affected limb during everyday tasks

Stress Loading Program

- A stress loading program initiates active movement and encourages compression of the affected joints.
- At first, stress loading may exacerbate symptoms in the extremity, but after several days, pain and swelling should decrease.
- The two components: Scrubbing and carrying
- Watson and Carlson performed a study of 52 patients with RSD solely using the stress loading program. With long-term follow-up they found 95% of patients returned to normal activities with this intervention.

Carlson LK, Watson HK. Treatment of reflex sympathetic dystrophy using the stress-loading program. J Hand Ther. 1988;1:149-154.

Scrubbing

- "Scrubbing" entails the back-and-forth movement of a weightbearing affected extremity.
- Patients use a scrub brush and assume a quadruped position for upper extremity involvement or an elevated sitting position for lower extremity involvement.
- Initially, the program consists of 3-5 minutes of scrubbing a wooden board followed by carrying activities in which the patient carries a 1 pound weight for 5-10 minutes.
- The duration of the activity and the amount of weight applied to the affected extremity are both gradually increased.

Carrying

- For upper extremity conditions, patients begin to carry a briefcase or purse in the affected hand with the arm extended. The amount of weight is the maximum tolerated by the patient. An initial weight generally ranges from 1 to 5 pounds. The weight should be carried throughout the day whenever the patient is walking or standing.
- A variety of weight-bearing techniques exist for lower extremity patients. Walking is a principal loading technique. Verbal and physical cueing can help ensure increased weight bearing during gait. Having the patient carry a weighted object on the affected side can also increase weight bearing.

Proprioceptive Neuromuscular Facilitation (PNF)

- Once the patient is actively involved in an edema management and stress loading program, functional restoration treatment can begin -- PNF.
- PNF patterns are spiral and diagonal combinations of motion that "permit maximum elongation of related muscle groups so that the stretch reflex can be elicited throughout the 'pattern'."
- These patterns, akin to normal movement patterns, simultaneously facilitate strength and balance as they increase the ability to perform ADL. PNF patterns promote "response of the neuromuscular mechanism through stimulation of the proprioceptors" and are usually well tolerated during the rehabilitation process.

Phillips ME. OT treatment for complex regional pain syndrome. OT Pract. August 20, 2001.

Voss D, Ionta M, Myers B. Proprioceptive neuromuscular facilitation. In: Voss D, Ionta M, Myers B, eds. Patterns and Techniques. 3rd ed. New York, NY: Harper & Row; 1985:xvii.

Functional Splinting

- In extreme CRPS cases, functional splinting may be required to encourage improved circulation/nutrition to the affected area as well as to promote more normal tissue length/positioning during rehabilitation.

Other Therapeutic Interventions

Hyperbaric Oxygen Therapy

- Hyperbaric oxygen therapy was assessed in a medium-sized RCT and produced a significant decrease in pain and edema versus "normal air" (level 2 evidence).
- Cost benefit considerations

Kiralp MZ, Yildiz S, Vural D, Keskin I, Ay H, Dursun H. Effectiveness of hyperbaric oxygen therapy in the treatment of complex regional pain syndrome. J Int Med Res. 2004;32:258-262.

Acupuncture

- Although acupuncture is mentioned in many treatment reviews, there is only one very small RCT in CRPS which failed to show a significant difference in outcomes, but this may be due to the small sample size.

Korpan MI, Dezu Y, Schneider B, Leitha T, Fialka-Moser V. Acupuncture in the treatment of posttraumatic pain syndrome. Acta Orthop Belg. 1999;65:197-201.

Chiropractic

- There is no research available supporting the use of chiropractic in CRPS.

Muir JM, Vernon H. Complex regional pain syndrome and chiropractic. J Manipul Physiol Ther. 2000;23:490-497.

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