I. History
II. Diagnosis
III. Current (revised) diagnostic criteria and tests
IV. Physical findings specific to CRPS and differential diagnosis
V. Treatment of CRPS:
   A. Medical
   B. Interventional
   C. Interdisciplinary Rehabilitation
   D. Emerging Approaches (e.g., HBOT)
CRPS

• Definition:
  A syndrome of burning pain, hyperesthesia, swelling, hyperhidrosis, and trophic changes. Partial forms are more common than the full-blown syndrome.

• Other Names/Variants:
  Sympathetically maintained pain, reflex sympathetic dystrophy, Sudeck’s atrophy, shoulder-hand syndrome.
Early History of CRPS

• In the 17th Century, Ambroise Paré presented the earliest description of CRPS as severe burning pain following peripheral nerve injury. Paré, a surgeon, treated King Charles IX for smallpox by inducing bleeding with a lancet applied to the arm. After this treatment, the king suffered from persistent pain, muscle contracture, and inability to flex or extend his arm.

• In 1864, Mitchell coined the term causalgia, which means burning pain, to describe persistent symptoms following gunshot wounds to peripheral nerves during the American Civil War.
History of CRPS (20th century)

• In 1900, Sudeck described radiographic spotty osteopenia (Sudeck’s atrophy)

• In 1916, Leriche focused on the sympathetic nervous system.

• In 1943, Livingston expanded the Leriche vicious circle theory that includes the following:
  • Abnormally firing, self-sustaining loops in the dorsal horn
  • Provoked by a small irritation focus in small nerve endings of major nerve trunks
  • Activating central projecting fibers, giving rise to pain.
History of CRPS (20th century), cont.

- In 1946, Evans used the term RSD, believing that sympathetic hyperactivity is involved somehow in the abnormal activity in the periphery.

- In 1993, the International Association for the Study of Pain (IASP) held a Special Consensus Conference addressing diagnosis and terminology (endorsing the term CRPS).

- In 1995, Paice wrote that, even after 130 years, there was still no general agreement on what to call RSD/CRPS, what causes it, or how best to treat it.

- In 2003, the closed Budapest Conference proposed new criteria, with greater specificity, that were subsequently validated and adopted by the American Academy of Pain Medicine in 2013.
A Diagnostic Dilemma

- There is no established test to provide a definitive diagnosis of CRPS.
- The diagnosis of CRPS is primarily clinical, based on history and physical exam.
- X-rays, 3-phase bone scans, and sympathetic blockade are all used to help confirm diagnosis, but not to establish it.
- The IASP criteria from 1993, while sensitive, were found to be insufficiently specific. They have since been replaced by the Budapest criteria, adopted in 2013.
- The new criteria require signs across a greater number of objective categories; which moreover need to be present at the time of evaluation and not simply reported to have been present at some time.
Rectifying Distortions

• “I haven’t even had an MRI”

“Well, Bob, it looks like a paper cut, but just to be sure let’s do lots of tests.”
IASP Diagnostic Criteria for CRPS (1993)

1. The presence of an initiating noxious event, or a cause of immobilization. *(not essential for the diagnosis to be made)*

2. Ongoing pain, allodynia, or hyperalgesia in which the pain is disproportionate to the inciting event.

3. There is or has been evidence of edema, changes in skin blood flow, or abnormal sudomotor activity in the region of pain.

4. This diagnosis is excluded by the existence of other conditions that would otherwise account for the degree of pain and dysfunction.
1. Continuing pain which is disproportionate to any inciting event.

2. Must report at least one symptom in *three of the four* following categories:

   A. **Sensory:** Reports of hyperesthesia and/or allodynia

   B. **Vasomotor:** Reports of temperature asymmetry and/or skin color changes and/or skin color asymmetry

   C. **Sudomotor/Edema:** Reports of sudomotor edema and/or sweating changes and/or sweating asymmetry

   D. **Motor/Trophic:** Reports of decreased range of motion and/or motor dysfunction (weakness, tremor, dystonia) and/or trophic changes (hair, nail, skin)
Budapest Criteria for Diagnosis of CRPS (2013) (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 temperature sensation and/or deep somatic pressure and/or joint movement)

**Vasomotor:** Evidence of temperature asymmetry (>1°C) 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.
Implications of The New Budapest Criteria

- While there are still no imaging studies or diagnostic tests that establish the diagnosis of CRPS, the more specific guidelines for clinical examination result in fewer people being diagnosed incorrectly with CRPS.

- One important aim of the more specific criteria was to reduce the potential morbidity (and even mortality) associated with inappropriate therapies, such as adverse reactions to medications and unnecessary invasive treatments.

- As a result, patients can be guided toward more appropriate, less invasive interventions.
Possible Predisposing Factors

• Demographic/Familial
  • CRPS is three times more frequent in females than males
  • Genetic predisposition has been hypothesized

• Behavioral/psychological
  • Cigarette smoking-- Some studies have indicated that cigarette smoking was strikingly present in patients and is statistically linked to CRPS. This may be involved in its pathology by enhancing sympathetic activity, vasoconstriction, or by some other unknown neurotransmitter-related mechanism.
  • Significant psychiatric co-morbidities are often noted, particularly anxiety, depression, PTSD, and character disorders. Many report a history of childhood trauma. Substance abuse issues may be present as well.
Difficult as it was to cut down on her caffeine consumption, Marcie strictly observed her self-imposed limit of one cup of coffee a day.
Differential Diagnosis

• Among others, CRPS must be distinguished from other conditions that can mimic the appearance of CRPS:
  • Acute Nerve Injury
  • Cellulitis
  • Septic Arthritis
  • Upper or Lower Extremity Occlusive Disease
  • Malingering / Secondary Gain
Malingering

“Our case is up next. By the way, you look great!”
Secondary Gain

“I told you! I can’t do the laundry or use the stove! The allergy medicine I’m on says not to operate heavy equipment.”
Assessment Of Potential Malingering

- Observed inconsistencies in symptom presentation
- Observed inconsistencies between observed symptoms and reports of dysfunction
- History of substance abuse or character disorder
- Potential fabrication of symptoms: use of tourniquet, shaving, other means of fabricating apparent objective signs
- Evidence from other medical records, reports of other clinicians (one of the benefits of interdisciplinary care)
Fig. 59-3  In first stage of reflex sympathetic dystrophy, swelling is usually soft and puffy with redness over joints. (From Lankford LL and Thompson JE: Reflex sympathetic dystrophy, upper and lower extremity: diagnosis and management. In American Academy of Orthopaedic Surgeons: Instructional course lectures, vol 26, St Louis, 1977, The CV Mosby Co.)
Fig. 59-6 Stage III is characterized by atrophy of skin and subcutaneous tissue, which produces a glossy appearance of skin. (From Lankford LL and Thompson JE: Reflex sympathetic dystrophy, upper and lower extremity: diagnosis and management. In American Academy of Orthopaedic Surgeons: Instructional course lectures, vol 26, St Louis, 1977, The CV Mosby Co.)
Diagnosis

A diagnosis of reflex sympathetic dystrophy is based on the following criteria: (1) history of recent or remote accidental or iatrogenic trauma or disease; (2) presence of persistent pain that is burning, aching, and/or throbbing in character; (3) one or more of the following: (a) vasomotor and/or sudomotor disturbances; (b) trophic changes, edema of the limb, sensitivity to cold, muscle weakness or atrophy, or trophic changes; and (4) relief of pain and modification of signs after regional sympathetic blockade. As previously mentioned in connection with causalgia, the fourth criterion is considered by most writers on the subject as one of the most important diagnostic features of reflex sympathetic dystrophy. While the “typical” case of reflex sympathetic dystrophy can be diagnosed without difficulty, many subtle cases of grade 3 RSD present only one or two of the symptoms and signs, thus simulating other...

Fig. 11-8. Roentgenograms of the hands of MB. A. Moderate degree of osteoporosis in the left hand 3 months after injury. B. More severe and more diffuse osteoporosis of the left hand and a normal right hand, 15 months after injury.
Fig. 11-4. Thermograms of a patient with severe reflex sympathetic dystrophy, for which a surgical sympathectomy was carried out. Sympathectomy did not relieve the symptoms. **A.** Thermogram of the patient after surgery. The left forearm was cold (blue), which is consistent with persistent reflex sympathetic dystrophy. **B.** Thermogram after a cervicothoracic sympathetic block was carried out with a local anesthetic. Note the warming effect over the left forearm and digits (purple, red, and yellow) compared to the preblock thermogram as well as the crossover effect of warming of the right forearm. The results of the sympathetic interruption with a local anesthetic proved that the surgical sympathectomy was incomplete. Thermograms courtesy of Dr. Pierre LeRoy.
Treatment

• The most important factor in the effective treatment of CRPS is the early recognition and treatment.

• The effectiveness of treatment is limited once the patient has reached the chronic fibrotic stage. Certainly, the incidence and severity of CRPS can be greatly reduced by initiating prophylactic measures, including immediate and aggressive mobilization of the involved extremity with passive and then active range-of-motion exercises.

• Similarly, in patients with established CRPS, physical and occupational therapy are key components of any therapeutic regimen.
Treatment Plan
(1st line therapeutics)

• PT and/or OT with Prednisone or Sympathetic block or Bier Block

• +/- NSAIDS, tricyclic antidepressants, Neurontin
Physical/Occupational Therapy

- Range Of Motion exercises
- Desensitization
- Strengthening
- TENS
- Physical modalities
- Biofeedback
- Graded motor imagery
- Mirror therapy
Medications

- Gabapentin (Neurontin)
- NSAIDs
- Prednisone
- Calcium Channel Blockers
- Alpha blockers
- Clonidine
- Capsaicin
- Beta blockers
- Calcitonin
- Tricyclic antidepressants
- SSRIs
Sympathetic Blockade

• Stellate Ganglion Blocks
• Lumbar Paravertebral Sympathetic Blocks
• Differential
Fig. 59-14  Stellate ganglion of sympathetic chain is located opposite seventh cervical vertebra, which is palpated less than 2 cm inferior to Chassaignac’s tubercle and can be found when sternocleidomastoid muscle and great vessels are retracted laterally. (From Lankford LL and Thompson JE: Reflex sympathetic dystrophy, upper and lower extremity: diagnosis and management. In American Academy of Orthopaedic Surgeons: Instructional course lectures, vol 26, St Louis, 1977, The CV Mosby Co.)
Bier Block

• Technique of regional anesthesia
• Lidocaine
• Guanethidine
• Reserpine
Post-acute Treatment: An Interdisciplinary Team Approach Is Essential

“The best hope of helping our patients is the adoption of a systematic, stable, empathetic and, above all, interdisciplinary approach that addresses those symptoms… The interdisciplinary approach for treating patients with CRPS remains the most pragmatic, helpful, and cost-effective therapeutic approach available today.” Harden et al., 2013

“A carefully selected combination of therapies—including medications, interventions, rehabilitation therapies, and psychological treatment approaches in the context of a functional restoration model of care—provides the best hope for treating CRPS.”

Feinberg and Feinberg (2010), published in Practical Pain Management, and cited in the website of the Reflex Sympathetic Dystrophy Syndrome Association
Comprehensive Rehabilitation

- Exercise
- Psychological intervention
- Medical management
- Smoking cessation
- Vocational retraining
Other Interventional/Surgical Approaches

• Catheter With Continuous Infusion
• Spinal Cord Stimulator
• Surgical Sympathectomy
• Ketamine Infusions
Continuous Infusion

- Risk of infection
- Catheter placement
- Physical limitations
Spinal Cord Stimulation

- Nondestructive
- Temporary stimulator on a trial basis
- Expensive
Surgical Sympathectomy

• Destructive procedure
• Frequent recurrences
• Laparoscopic technique
Emerging Treatment Options: Hyperbaric Oxygen Treatment (HBOT)

- The rationale for HBOT is fully consistent with what is known about the pathophysiology of CRPS. HBOT:
  - causes vasoconstriction,
  - decreases edema, and
  - increases the partial pressure of oxygen in the tissues
  - stimulates the activity of depressed osteoblasts
  - decreases the formation of fibrosis tissue.

- Moreover, HBOT has fairly minimal associated risks, especially at pressures under 3.0ATA (three times the “normal” atmospheric pressure).
In the case of CRPS, it is thought that HBOT supersaturates tissues that have been deprived of oxygen because of the swelling of a limb. Saturation levels of oxygen in blood and tissues increase 10 to 20 times while in the chamber.

Further, HBOT has a tendency to constrict vessels by about 15 percent, which causes a decrease in swelling from the edema present in most people with CRPS.

Various published case reports and one randomized clinical trial have indicated improvements in pain, swelling, and range of motion.