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Complex Regional Pain Syndromes

- I. Know the definition of complex regional pain syndrome (CRPS) and be aware of the inciting traumatologic events (Stanton-Hicks et al. 1995).
 - A. CRPS type I (reflex sympathetic dystrophy). Minor injuries or fracture of a limb precede the onset of symptoms.
 - B. CRPS type II (causalgia) develops after injury to a major peripheral nerve.
- II. Know the common clinical characteristics of CRPS (Veldman et al. 1993; Birklein et al. 2000; Baron and Wasner 2001; Wasner et al. 2003, Fördereuther et al. 2004).
 - A. Know the common sensory signs and symptoms associated with CRPS:
 1. Continuous burning pain in the distal part of the affected extremity.
 2. Pain is disproportionate in intensity to the inciting event and usually increases when the extremity is in a dependent position.
 3. Stimulus-evoked pains include mechanical and thermal allodynia and/or hyperalgesia, and deep somatic allodynia (pain due to touching the joints and movement of joints).
 4. Sensory abnormalities are most pronounced distally, and have no consistent spatial relationship to individual nerve territories or to the site of the inciting lesion.
 - B. Know the common autonomic abnormalities associated with CRPS:
 1. Swelling of the distal extremity especially in the acute phase
 2. Hyper- or hypohidrosis
 3. Vasodilatation or vasoconstriction
 4. Changes in skin temperature
 - C. Know the common trophic changes associated with CRPS:
 1. Abnormal nail growth
 2. Increased or decreased hair growth
 3. Fibrosis
 4. Thin, glossy skin
 5. Osteoporosis
 - D. Know the common motor abnormalities associated with CRPS:
 1. Weakness
 2. Coordination deficits
 3. Tremor
 4. Dystonia
 5. Neglect-like symptoms or symptoms of disturbed body perception of the affected extremity
 - E. Know that sympathetically maintained pain (SMP) may occur in CRPS (Baron et al. 1999; Ali et al. 2000; Baron et al. 2002).
 1. Be aware that this pain component may be relieved by specific sympatholytic procedures (e.g., sympathetic blocks).

2. Know that SMP is defined to be a symptom or the underlying mechanism in a subset of patients with CRPS and not a clinical entity. The positive effect of a sympathetic blockade is not essential for the diagnosis of CRPS.
- F. Understand that CRPS evolves through different stages, although the duration of each stage may be variable.
- III. Know the proposed pathophysiological mechanisms of CRPS (Baron et al. 1999; Birklein et al. 2001; Huygen et al. 2002; Juottonen et al. 2002; Jänig and Baron 2003; Maihofner et al. 2003).
- A. In the periphery:
1. Know the abnormal characteristics of primary afferents after lesion in CRPS-II (e.g., spontaneous discharge, sensitization, ectopic mechanosensitivity, or acquired responsiveness to norepinephrine).
 2. Understand the proposed role in spontaneous discharge of sodium channel accumulation in the terminal membrane.
 3. Understand the proposed role of acquired norepinephrine responsiveness in intact C nociceptor terminals whose axons travel in a damaged nerve.
 4. Be aware of the potential role of an inflammatory reaction and neurogenic inflammation in the acute phase of CRPS.
- B. For the central nervous system:
1. Understand the proposed role of *N*-methyl-D-aspartate (NMDA)-receptor mediated hyperexcitability in the spinal cord dorsal horn.
 2. Understand how spinal cord neuron hyperexcitability can account for pain that shows a distally generalized distribution.
 3. Understand that a persistent source of nociceptor drive may dynamically maintain hyperexcitability in central neurons.
 4. Realize that the somatosensory cortical maps are changed in S1.
 5. Understand that an unilateral inhibition of central sympathetic vasoconstrictor activity is involved in vascular abnormalities.
 6. Realize that motor abnormalities must be generated in the central nervous system.
- IV. Know the proposed diagnostic procedures in CRPS (Galer et al. 1998; Allen et al. 1999; Bruehl et al. 1999; Harden et al. 1999; Oerlemans et al. 1999; Sieweke et al. 1999; van de Beek et al. 2002; Wasner et al. 2002, 2003).
- A. Realize that CRPS diagnosis is based upon clinical criteria and that there is so far no gold standard nor any objective diagnostic tool.
- B. Be aware that CRPS may be potentially overdiagnosed.
- C. Know the categories of clinical signs and symptoms that are important for diagnosis, know the interpretation of these diagnostic criteria for clinical use and the sensitivity and specificity (sensitivity 0.85, specificity 0.60).
1. Positive sensory abnormalities: spontaneous pain, mechanical hyperalgesia, thermal hyperalgesia, deep somatic hyperalgesia.
 2. Vascular abnormalities: vasodilation, vasoconstriction, skin temperature asymmetries, skin color changes.
 3. Edema, sweating abnormalities: swelling, hyperhidrosis, hypohidrosis.
 4. Motor, trophic changes: motor weakness, tremor, dystonia, coordination deficits, nail changes, hair changes, skin atrophy, joint stiffness, soft tissue changes.
 5. Interpretation. One or more symptoms of three or more categories each AND one or more signs of two or more categories each.

- E. Know the diagnostic tests which may aid the diagnosis of CRPS.
 1. Understand the roles that X-ray, three-phase bone scan, quantitative sensory testing (QST), autonomic testing, and thermography play in CRPS diagnosis.
- V. Know the therapeutic interventions applied in CRPS (Kingery 1997; Price et al. 1998; Stanton-Hicks et al. 1998, 2002; Kemler et al. 2000; van Hilten et al. 2000; Perez et al. 2001; Forouzanfar et al. 2002; Moseley 2004).
 - A. Know the basic guidelines in the treatment of CRPS.
 1. Realize that treatment should be immediate after diagnosis.
 2. Realize that even a suspected CRPS should be evaluated by a pain specialists to start adequate therapy immediately.
 3. Have knowledge of a multidisciplinary approach to the patient with CRPS, and understand that the treatment is most importantly directed toward restoration of full function of the affected area.
 4. Know that severity of the disease determines the therapeutic regime and that reduction of pain is the precondition with which all other interventions must comply.
 5. Realize that the intensity of therapy (in particular physiotherapy) should be adapted to the severity of the disease. It should be gentle and below pain threshold and must not exacerbate the pain since every painful stimulus may worsen the syndrome.
 - B. Know pharmacological approaches typically used for neuropathic pain of all types.
 1. Know specific agents for which data from controlled clinical trials have established efficacy in CRPS (e.g., gabapentin, corticosteroids, Ca-modulating drugs, free radical scavengers).
 2. Know agents for which substantial anecdotal evidence exists for efficacy in CRPS (e.g., antidepressants, anticonvulsants, opioids, capsaicin, lidocaine).
 3. Know drugs typically used in the treatment of sympathetically mediated pain and be aware of the limited data in support of their use (e.g., phenoxybenzamine, prazosin, guanethidine, reserpine, and clonidine).
 - C. Know the role of interventional approaches in the management of CRPS.
 1. Be aware of the indications, techniques, and potential complications of sympathetic blockade in CRPS.
 2. Know the different types of local sympathetic blocks.
 3. Know the indications, advantages and disadvantages, and drugs and techniques employed in regional intravenous blockade.
 4. Understand the limited role of surgical sympathectomy in patients who respond to temporary sympathetic blocks.
 5. Be aware of the methods employed to document effective regional sympathetic block (e.g., thermography and laser Doppler).
 6. Be aware of the existence, possible causes, and treatment of post-sympathectomy pain.
 - D. Be aware of neurostimulatory approaches to the management of CRPS.
 1. Be aware of the variability in response to transcutaneous electrical nerve stimulation and the multiple methods that can be employed.
 2. Understand the limited role played by percutaneous electrical nerve stimulation.
 3. Be aware of the data supporting the value of dorsal column stimulation.
 4. Be aware of the data supporting motor cortex stimulation.
 5. Be aware of the data supporting and refuting the value of deep brain stimulation.
 - E. Be aware of the response of pain to clonidine and refractory dystonia to intrathecal baclofen.
 - F. Be aware of the data supporting and the data refuting the value of acupuncture.

- G. Understand the fundamental importance of physical and occupational therapy in treatment for CRPS.
- H. Know the utility of psychiatric and psychological approaches to management of CRPS.

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