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Limb Pain Syndromes

Version of 2016

3. Complex Regional Pain Syndrome Type 1 (Synonyms: Reflex Sympathetic Dystrophy, Localized Idiopathic Musculoskeletal Pain Syndrome)

3.1 What is it?

Extremely severe limb pain of unknown cause frequently associated with skin changes.

3.2 How common is it?

The frequency is unknown. It is more common in adolescents (average onset age is around 12 years) and in girls.

3.3 What are the main symptoms?

Usually, there is a long-term history of very intense limb pain that is unresponsive to different therapies and increases over time. Frequently, it results in the inability to use the affected limb.
Sensations that are painless to most people, such as light touch, may be extremely painful to affected children. This odd sensation is called "allodynia".
These symptoms interfere with the daily activities of affected children, who often miss many days at school.
Over time, a subset of children develops changes in skin colour (pallid or purple mottled appearance), temperature (usually reduced) or perspiration. Swelling of a limb may also be present. The child may sometimes keep the limb in unusual postures, refusing any movement.

3.4 How is it diagnosed?

Until a few years ago, these syndromes received different names, but today physicians refer to them as complex regional pain syndromes. Different criteria are used for the diagnosis of the disease.
The diagnosis is clinical, based on the features of the pain (severe, prolonged, limiting activity, unresponsive to therapy, presence of allodynia) and the physical examination.
The combination of complaints and clinical findings is quite characteristic. The diagnosis requires that other diseases that generally can be managed by primary care doctors, clinicians or paediatricians are ruled out before referral to a paediatric rheumatologist. Laboratory studies are standard. An MRI may show non-specific alterations of the bone, joints and muscles.

3.5 How can we treat it?

The best approach is an intensive physical exercise therapy programme supervised by physical and occupational therapists, with or without psychotherapy. Other treatments have been used, alone or in combination, including antidepressants, biofeedback, transcutaneous electrical nerve stimulation and behavioural modification – all without definitive results. Analgesics (pain-killers) are usually ineffective. Research is currently underway and in the future better treatments will hopefully arise as the causes are identified. The treatment is hard for all people involved: the children, the family and the treating team. Psychological intervention is usually necessary because of the stress produced by the disease. Difficulty on the part of the family in accepting the diagnosis and in complying with treatment recommendations are the major causes of treatment failure.

3.6 What is the prognosis?

This disease has a better prognosis in children than in adults. In addition, most children recover faster than adults. However, it takes time and the lapse to recovery varies widely from child to child. Early diagnosis and intervention lead to a better prognosis.

3.7 What about everyday life?

Children should be encouraged to maintain physical activities, regular school attendance and leisure time with their peers.