



Reflex Sympathetic Dystrophy

Discussion paper prepared for

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Prepared by:

Dr. J.F.R. Fleming

Professor Emeritus, Division of Neurosurgery
University of Toronto
The Toronto Western Hospital

Dr. J.F. Ross Fleming graduated from the University of Toronto Medical School in 1947. He did post-graduate training in neurosurgery at the University of Toronto, at the University of Michigan and at Oxford, England, from 1947 to 1956. He became a Fellow in neurosurgery in 1956. He holds the rank of Professor Emeritus in the Division of Neurosurgery, Department of Surgery, at the University of Toronto. His clinical and research interests were in neurosurgery. He has published widely in that area. He practiced at the Toronto Western Hospital as the Head of the Division of Neurosurgery from 1965 to 1984 and as staff in the Division of Neurosurgery from 1956 to 1996. Dr. Fleming was involved at the Tribunal as an assessor from 1988 to 1992, as a counsellor from 1993 to 1997 and as Chair of the medical counsellors group from 1998 to 2006.

This medical discussion paper will be useful to those seeking general information about the medical issue involved. It is intended to provide a broad and general overview of a medical topic that is frequently considered in Tribunal appeals.

Each medical discussion paper is written by a recognized expert in the field, who has been recommended by the Tribunal's medical counsellors. Each author is asked to present a balanced view of the current medical knowledge on the topic. Discussion papers are not peer reviewed. They are written to be understood by lay individuals.

Discussion papers do not necessarily represent the views of the Tribunal. A vice-chair or panel may consider and rely on the medical information provided in the discussion paper, but the Tribunal is not bound by an opinion expressed in a discussion paper in any particular case. Every Tribunal decision must be based on the facts of the particular appeal. Tribunal adjudicators recognize that It is always open to the parties to an appeal to rely on or to distinguish a medical discussion paper, and to challenge it with alternative evidence: see *Kamara v. Ontario (Workplace Safety and Insurance Appeals Tribunal)* [2009] O.J. No. 2080 (Ont Div Court).

REFLEX SYMPATHETIC DYSTROPHY

There are a number of syndromes in which pain in an arm and hand, or in a leg and foot, following injury, is disproportionate in its severity and duration to the initiating injury and is inappropriate to the initiating injury in its distribution. The term *reflex sympathetic dystrophy (RSD)* describes a prolonged complex exaggerated painful response to a limb injury in the absence of a nerve injury. The term *causalgia* describes prolonged severe limb pain in individuals who have sustained an injury to a major nerve in a limb.

Rather than being a description of the clinical symptoms, the term RSD implies presumed mechanisms of pain production and maintenance (i.e. "reflex" and "sympathetic"). Traditionally it has been believed that the sympathetic nervous system plays a key role in the production and/or maintenance of the pain. However, whether the sympathetic nervous system plays a primary or secondary role in initiating and/or maintaining the pain, or even no role at all, is not clearly established and is still disputed. The term "*sympathetically maintained pain*" has been used in the past, but its use is to be discouraged, as it may not accurately describe the role of the sympathetic nervous system in this syndrome.

There is considerable controversy and misunderstanding about the clinical definition of RSD and there is a lack of clear scientific understanding about its patho-physiology. The term RSD has been rather indiscriminately used, and may have lost much of its usefulness as a clinical designation. Therefore the International Association for the Study of Pain (IASP) recommends the use of the umbrella term "*Complex Regional Pain Syndrome*" (CRPS). This term requires the presence of regional pain and sensory changes following an injury, together with associated regional findings such as abnormal skin colour, temperature change, abnormal sweating and/or tissue swelling, and the combination of these findings exceeds their expected magnitude in response to the known physical damage during and following the injury. IASP proposes two types of CRPS, type I corresponding to RSD without a definable nerve injury, and type II correspondings to causalgia, where a definable nerve lesion is present (see Table 1).

In RSD the pain is out of all proportion to that expected from the injury or initiating noxious event, which is normally fully recoverable. Both *spontaneous* and *evoked* pain occur. Spontaneous pain is pain that is constantly present, even though the limb is not being touched or moved. Evoked pain ("allodynia" and "hyperpathia") is defined as exaggerated pain

in response to a sensory stimulus that would not normally be painful, such as touching the skin, deep pressure or moving a joint. The quality of the pain varies, and is often described as burning, aching or throbbing. The pain may be so severe that any contact with the affected part, or any movement whether active or passive, is extremely painful. The patient's protection of the limb and reluctance to allow any movement may lead to prolonged immobilization of the limb, with resultant muscle wasting, joint stiffness and loss of function.

Motor changes are usually present. There is almost always some degree of loss of function of the limb, muscle weakness and wasting are common, and occasionally there may be abnormal movements such as local tremor or dystonia.

The sympathetic nervous system is part of the *autonomic nervous system*. The sympathetic nerves to the limbs are separate from, and operate independently from the motor nerves to muscles and the sensory nerves to skin and joints. Sympathetic nerves regulate the calibre of blood vessels, and thus the local blood flow, temperature and colour of the skin, and the sweat glands which determine the dryness or moisture of the skin. In RSD autonomic changes are present at some stage in the course of the illness. These changes may include abnormalities of skin colour, temperature or sweating, and there is often some tissue edema (swelling). In the early stages the skin tends to be warm, and later it tends to become cold, pale and/or cyanotic, although the pattern of such colour and temperature changes is not consistent. Much later, "trophic" changes may occur, the skin may become thin and shiny, or thick and flaky, there may be hair loss in the limb, and the nails may become thickened. All these changes may be mediated by abnormal activity of the sympathetic nerves resulting from hypersensitivity of sympathetic nerve endings and receptors to circulating chemicals, or they may be secondary to inflammation in the early stages, and to immobilization and disuse of the limb later on.

Osteoporosis (decreased calcification) may occur in the bones of the affected limb, especially distally, and is probably secondary to prolonged immobilization of the limb. (Severe osteoporosis in RSD has been called "Sudeck's atrophy").

Diagnostic tests. There are no diagnostic tests to confirm the diagnosis of RSD. The diagnosis can only be made by thoughtful and careful analysis of the history and clinical picture, together with radiologic and laboratory data, by an experienced clinician who has examined the patient and has followed his/her progress. Some commonly used diagnostic tests include radionuclide

three phase bone scan, electro-physiologic tests, and the response or lack thereof to sympathetic blockade.

Bone Scan. The "three phase bone scan" (TPBS) is often abnormal in RSD, but a normal TPBS does not rule out RSD. An abnormal TPBS in a patient with the clinical picture described above, tends to support a diagnosis of RSD.

Electrodiagnostic tests. EMG and measurement of nerve conduction velocity are useful to confirm or exclude a significant nerve injury in patients with suspected RSD, but have no other value in the diagnosis of RSD. As a word of caution, it must be pointed out that currently available refined electro-physiologic methods are capable of detecting very subtle **subclinical** local nerve dysfunction (i.e. not detectable by clinical examination of the limb, and probably of no clinical significance), which unfortunately can be misconstrued as an authentic basis for the clinical symptoms.

Sympathetic Block. Blocking (temporarily interrupting) the sympathetic nerve supply to the affected limb by the injection of local anaesthetic has in the past been extremely common in the management of patients with RSD. Temporary pain relief may occur following sympathetic blockade, but long term results have generally been poor.

For the upper extremity, sympathetic block is achieved by injecting local anaesthetic into the front of the neck above the clavicle ("stellate ganglion block"); a satisfactory block should be followed by increased temperature in the arm and hand, with drooping of the eyelid on the same side as the block, for a few hours. For the lower extremity, the injection is made though the back beside the lumbar spine ("lumbar paravertebral sympathetic block"), and should be followed by increased temperature of the leg and foot for a few hours. Surgical division of these nerves has sometimes been done to permanently interrupt the sympathetic nerve supply to the limb, but long term results have generally been poor. Temporary chemical blockade of the sympathetic nerves can be achieved by drugs that paralyze sympathetic connections and receptors (guanethidine, clonidine or phentolamine). Unfortunately, in spite of claims that have been made about the benefits of various types of sympathetic blockade in RSD, there is no reliable and consistent scientific evidence of their effectiveness; there are no placebo controlled trials, there has been little uniformity or consistency in the clinical picture of patients reported as having RSD, and our understanding of the role of the sympathetic nervous system in initiating or maintaining the symptoms is incomplete. Not all patients respond to sympathetic blockade. Although sympathetic blockade may bring temporary pain relief in some

patients, it must be stressed that temporary relief following block does not necessarily confirm the diagnosis of RSD. Conversely, failure of sympathetic block to relieve the symptoms does not rule out the diagnosis of RSD.

Incidence. **Pain, tenderness, stiffness and swelling are very common after fractures and other injuries to limbs, and have a fairly predictable time of healing. RSD probably occurs in about 1 - 2% of all people who have sustained a fracture, and causalgia (after a major peripheral nerve injury) occurs in a slightly higher percentage.**

Natural history. The symptoms of RSD are often extremely persistent. They may gradually remit over time. Patients are commonly treated with physiotherapy, medication, various forms of sympathetic blockade and counselling, which may or may not influence the course of the illness.

Diagnosis. RSD is unilateral, affecting one limb only. It is a complex pain syndrome in a limb following an initiating noxious event. Its occurrence or severity is not necessarily related to the severity of the initial injury. RSD consists of pain and related sensory abnormalities, abnormal blood flow and sweating, abnormalities in the motor system and changes in superficial and deep tissues. Essential to the diagnosis is that the pain be disproportionate to the initiating cause in severity, duration and distribution, and that there are or have been some vascular and/or sudomotor (sweating) changes. Abnormality on a three phase bone scan tends to support the diagnosis, but a normal three phase bone scan does not rule out the diagnosis. There may be temporary relief of pain following sympathetic nerve blockade, but a response by no means confirms the diagnosis. Lack of pain relief following sympathetic nerve blockade does not rule out the diagnosis.

The problem in defining the diagnosis of RSD is to determine when the pain may be judged to be disproportionate in severity, duration and distribution to the initiating event, and to determine whether the swelling, vasomotor or sudomotor (sweating) changes that commonly occur after all injuries have become disproportionate to the initiating event. At what point can these changes be said to be excessive and indicative of RSD? As there are no diagnostic tests for this condition, even the experienced clinician faces a difficult diagnostic challenge. The clinician will need to rely on very careful clinical evaluation of the history and physical findings during the course of the illness, the radiologic and laboratory data, and thoughtful clinical reasoning which should consider and exclude the various conditions listed in the next two paragraphs.

Differential Diagnosis. RSD must be distinguished from an abnormally long healing time following the initial injury, from unexpected local complications of the injury, from unsuspected and unrelated pathology in the limb, from an exaggerated amount of pain during the recovery process in an individual who is particularly sensitive to pain, and from psychogenic magnification of the symptoms. RSD must also be distinguished from *neuropathic* pain, which is the continuing unpleasant pain that commonly accompanies nerve injuries, peripheral neuritis and certain other neurological conditions.

Patients with RSD may develop secondary psychological or even psychiatric disturbances in response to their pain, such as loss of function, fear, and depression, that may accompany and aggravate their symptoms. Nevertheless primary psychological or psychiatric conditions, including conversion reaction, factitious production of symptoms or malingering may present with symptoms that can closely resemble RSD, and they must always be considered as possible mechanisms for prolonged pain and disability.

Table 1

Complex regional pain syndrome (CRPS)*

Reflex sympathetic dystrophy (CRPS Type I)

Develops after an initiating noxious event

Spontaneous pain and/or allodynia/hyperalgesia occurs, is disproportionate to the inciting event in severity, duration and distribution. and is beyond the territory of a single peripheral nerve.

There is or has been evidence of edema, skin blood flow abnormality (i.e. temperature and/or colour change), or abnormal sudomotor (sweat gland) activity in the region of the pain since the inciting event.

This diagnosis is excluded by the existence of conditions that would otherwise account for the degree of pain and dysfunction.

Causalgia (CRPS Type II)

Develops after a nerve injury.

Spontaneous pain and/or allodynia/hyperalgesia occurs, is disproportionate to the inciting event in severity and duration, and is not necessarily limited to the territory of the injured nerve. There is or has been evidence of edema, skin blood flow abnormality, or abnormal sudomotor (sweat gland) activity in the region of the pain since the inciting event.

This diagnosis is excluded by the existence of conditions that would otherwise account for the degree of pain and dysfunction.

* Adapted from Stanton-Hicks M, Janig W et al (1995), and Boas RA (1996).

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