chapter 9

MULTIPLE REFLEX SYMPATHETIC DYSTROPHY
Which patients are at risk for developing a recurrence of reflex sympathetic dystrophy in the same or another limb?

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ABSTRACT

Many aspects of bilateral presentation or recurrence of reflex sympathetic dystrophy (RSD) are unknown.

Method: For this reason 1183 consecutive patients with RSD were analyzed.

Results: In 10 patients RSD started in symmetrical limbs. In 34 patients RSD recurred in the same limb after a period of no or few complaints and in 78 patients RSD recurred in one or more limbs other than the first limb. Compared to 1065 patients with RSD without these features, these patients were younger of age ($p = 0.00003$) and RSD started more frequently with a cold skin temperature ($p = 0.02$). Patients did not differ in gender or primary localization of RSD. Involvement of a second limb concerned in 47% the symmetrical limb. Recurrences were in 53% of spontaneous origin and often characterized by few signs and symptoms. The incidence of a recurrence was 1.8% per patient/year. No measures are known to prevent recurrence.

Conclusion: Reflex sympathetic dystrophy may recur in the same or in another limb, though only in a minority of patients. Recurrences occur especially in younger patients and in the symmetrical limb. Diagnosis of a recurrence is difficult, for often the recurrence occurs spontaneously and presents with few signs and symptoms.

INTRODUCTION

Reflex sympathetic dystrophy (RSD) is an abnormal reaction of the body to trauma and one of the most frequent complications after surgery to extremities. RSD is characterized by pain, edema, vasomotor changes, loss of function and increase of these signs and symptoms after exercise. Several other signs and symptoms, such as neurologic disturbances, hyperhidrosis and atrophy may also occur [29]. The signs and symptoms are localized in the periphery of a limb. In the upper limb there may be concomitant complaints of the shoulder, known as the shoulder-hand syndrome[24 28] Some authors report patients with RSD localized around a knee [18], hip [1],in the thoracic wall [10] or in the face [27], though these presentations are rare. Recurrence of RSD after a period of no or few complaints or localization in more than one limb has been reported in French literature concerning algodystrophy [1 7 9 20 23], though their diagnostic criteria are different from what is called reflex sympathetic dystrophy in Anglo-Saxon literature. In English literature, these features have been the subject of a few case reports or - when presented in series of patients - as a matter of secondary importance. Still many patients ask us, when they are discharged from further therapy - cured or not cured - "Can it recur?"

Both aspects- localization of RSD in multiple limbs and recurrence of RSD in the same or another limb - are the subject of this report.

PATIENTS AND METHODS

In november 1984 an outpatient clinic for RSD patients was instituted by the department of surgery of the University Hospital Nijmegen. Since then, we have seen approximately 1500 patients - mostly referred from other departments or hospitals - with a presumed or suspected diagnosis of RSD.

RSD has not been clearly defined in literature. The criteria for diagnosis are:

1. 4 or 5 of following symptoms:
   - unexplained diffuse pain
   - difference in skin color in relation to the healthy symmetrical limb
   - diffuse edema
   - abnormal skin temperature in relation to the healthy symmetrical limb
   - limited active range of motion

2. Above signs and symptoms increase after using the affected limb
3. Above signs and symptoms are present in an area much larger than the area of primary injury or operation and including the area distally of the primary injury

These selection criteria approximate those, utilized in other studies concerning RSD [2 6 15 19] and are discussed in a previous report [29].

Special attention was paid to signs and symptoms, localization and etiology of RSD. It no luxating events, even minor trauma, could be remembered by the patients, the RSD was considered to be spontaneous in origin. Skin temperature at onset of RSD was called primary temperature. Statistical analysis was performed by the Chi-square-test and the Kruskal-Wallis test.

RESULTS

During the study period (november 1984 to april 1994) 1183 patients fitted into the above criteria for RSD. 1065 patients were seen with a single episode of RSD in one limb (further called single RSD). In 118 patients (10%), history or follow-up revealed presentation of RSD in two limbs or a recurrence of RSD in the same or another limb (further called multiple RSD) (table 9.1).

Table 9.1 Patients with bilateral presentation or recurrence of RSD (multiple RSD). Numbers of patients and interval between first and last appearance of RSD. Two patients are counted twice. They developed a recurrence in the same limb after a period of no complaints, and later they developed a recurrence in another limb.

	n	interval
bilateral presentation	10	-
recurrence in same limb	34	3 months 20 years
2 limbs	64	2 weeks - 15 years
3 limbs	8	10 months -

		9 years
4 limbs	4	2.5 years- 14 years

The group with single RSD consisted of 267 male (25%) and 798 female (75%) patients (table 9.2). Age varied from 4 to 84 years (median 41 years). RSD was localized in the upper limb in 635 patients (60%), and in 430 patients (40%) in the lower limb. In 105 patients (10%) RSD was of spontaneous origin (table 9.3).

The group of 118 patients with multiple RSD consisted of 23 male (20%) and 95 female (80%) patients (table 9.2). Age - at time of onset of RSD - varied from 9 to 71 years (median 35 years). The first localization of RSD was in the upper limb in 58 patients (49%) and in the lower limb in 60 patients (51%). In 72 of all 136 recurrences (53%), the recurrence or the affection of another limb developed spontaneously (table 9.3).

Comparing the patients with single versus multiple RSD, no difference was found as to gender or the first localization of RSD (table 9.2). Patients with multiple RSD were younger of age (Kruskal-Wallis: p = 0.00003).

Table 9.2 Differences between patients with RSD in one limb (single RSD) versus patients with bilateral or recurrent 850 (multiple RSD).

	single RSD	multiple RSD
n	1065	118
gender; M : F	1:3	1;4¶
age; median (range)	41(4-84) years	35 (9-71) *
primary cold #	38%	52%
primary site upper limb	60%	49%

* patients with multiple RSD were younger of age (P = 0.000031)
• skin temperature of the affected limb at time of onset of RSD. More often cold in multiple RSD (P = 0.021)
¶ differences between single and multiple RSD not significant

940 of the RSD patients could remember which difference in skin temperature existed between the diseased and the healthy symmetrical limb at the time complaints started (warm, cold or same temperature: primary temperature). In the multiple group, 49 out of 94 patients (52%) told us the skin temperature was colder, in contrast to 317 out of 846(38%) in the single group (p = 0.02; Chi-square, Yates corrected).

In 10 patients, RSD started simultaneously in 2 limbs; these were always symmetrical limbs. In 5 of these patients RSD developed after bilateral trauma, in 2 after bilaterally performed surgery' and in 3 patients RSD occurred spontaneously in both limbs. In 34 patients, RSD recurred in the same limb after a period of no or few complaints. Time between first and second appearance varied from 3 months to 20 years (median 2.7 years). Patients with a recurrence occurring in the same limb did not differ from patients with a recurrence in another limb, as to gender, etiology of primary' RSD, primary temperature or affected extremity, but patients with a recurrence in the same limb were younger of age (Kruskal-Wallis: p=0.01). In 64 patients RSD recurred in a second limb. When RSD recurred in a second limb this concerned the symmetrical limb in 30 of 64 patients (47%); in 34 patients (53%) primary' RSD and recurrence concerned one upper and one lower limb; 18 times at the same side (hemiplegic distribution), 16 times at opposite sides. 8 patients suffered from RSD in three limbs. Because of intractable pain and total incapacitation, one of these patients committed suicide. 4 patients suffered from RSD in all 4 limbs.

In 2 of these patients, RSD recurred in the first limb after a period of no or few complaints while some time later they developed RSD in a second limb.

In most cases recurrences started with diffuse pain in the limb without any obvious signs or symptoms. Later, and sometimes only after muscular exercise, typical signs and symptoms occurred which, enabled us to make the diagnosis RSD. For this reason recurrences were often diagnosed with some delay.

Table 9.3 Etiology of RSD

	single RSD		multiple RSD			
			first		recurrence	
trauma	711	67%	66	56%	44	32%
surgery	195	18%	23	20%	16	12%
spontaneous	105	10%	19	16%	72	53%
others	54	5%	10	8%	4	3%

Recurrences were seen in 10% of our patients. When the time period of analysis is taken into account - 110 days to 24.8 years, median 5.1 years-, the incidence of a recurrence per patient/year at risk was 1.8%.

DISCUSSION

Recurrence in the same limb

Recurrences of RSD in the same limb have been reported by Evans [8]. He mentioned exacerbations of RSD after surgery or infection but did not present any details concerning the development of the syndrome. In French literature several studies report recurrences. As mentioned before, we must make some reservations because diagnostic criteria are not the same in French and Anglo-Saxon countries. Acquaviva et al [1] reported 32 patients with a recurrence of RSD in their presentation of 765 patients (4%) and Gougeon et al [9] reported 41 recurrences in 573 patients (7%); both studies did not report the duration of follow-up.

Bilateral RSD

Bilateral presentation of RSD has been reported before. Livingstone [17] reported "mirror images" in 35 patients with bilateral RSD, as the affection was always located in symmetrical areas of symmetrical extremities. Other studies on bilateral RSD found no such mirror images [11 14 19 21 25]. In the present series, no mirror images were seen but bilateral presentation was found in 10 patients and recurrences concerned the symmetrical limb in 47% of cases.

Complaints in the symmetrical limb without clinical signs and symptoms of RSD have been reported before. Kozin et al [14] reported 11 patients with a shoulder-hand syndrome. One of their patients showed clinical signs and symptoms of RSD in both arms and another patient in one arm and one leg. In 5 patients they found an increased number of painful joints in the symmetrical limb without other signs and symptoms of RSD. Kurvers et al [16]reported increased red blood cell velocity (RBCV) in patients with warm RSD. 74% of these patients showed increased RBCV in the symmetrical limb without signs and symptoms of RSD. In cold RSD they found decreased RBCV, and in 30% of these patients RBCV in the symmetrical limb was decreased without signs and symptoms of RSD. Such measurements in other than the symmetrical limbs have - by our knowledge - not been performed.

Bhatia et al [5] reported spreading to other limbs in 7 of 18 patients with both RSD and dystonia. Secondary involvement concerned the symmetrical limb in 3 and the homolateral limb in 4 patients.

Localization of RSD in 3 or even 4 extremities is extremely rare and known from case reports only [3 4 22] .

In 53% of our patients, no luxating factor could be related to the recurrence of RSD. Acquaviva et al [1] reported 32 recurrences in 765 patients with RSD, without a luxating factor in more than 50% of the patients. Gougeon et al [9] also reported a high incidence of spontaneous origin in recurrences but did not present details. The high frequency of spontaneous recurrence of RSD, and the onset of recurrent RSD at a younger age, suggests that these patients are predisposed for developing RSD.

No other reports on the incidence of recurrent RSD in the same or another limb could be found. The incidence of RSD is estimated at 5-15% after all injuries [25] Once a patient has developed RSD, the incidence of recurrence of RSD is 1,8%per year. In this study 10% of the patients developed a recurrence of RSD and in the future more patients will probably develop another episode of RSD. From this study we conclude that young patients in which RSD started with a cold skin temperature, have the highest chances for developing a recurrence.

Preventive measures for recurrences of RSD are unknown. Many physicians expect trauma or surgery will reactivate the syndrome, though this hypothesis has never been proven in a prospective study. As the incidence of recurrence is low and as more than 50% of the recurrences of RSD are of spontaneous origin, we do not advise our patients to take any special measures for preventing trauma.

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