

chapter
4

**ARE THERE THREE STAGES IN
REFLEX SYMPATHETIC DYSTROPHY?**

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ABSTRACT

Objective: Clinically, reflex sympathetic dystrophy (RSD) is considered to develop according to three consecutive stages, starting with a warm skin temperature and edema, passing through a second stage with vasomotor lability and ending with a cold skin temperature and tissue atrophy. This staging however has never been documented in a large series of patients.

Patients: 177 consecutive RSD patients coming to our attention, were analyzed for signs and symptoms according to these three stages.

Results: Only 25 patients (14%) fitted into the classical staging, that is starting with a warm and ending with a cold skin temperature. 119 patients (87%) had warm RSD from the start and never developed stage I and/or III, while 24 patients (14%) had cold RSD from the start.

Discussion: Patients with primarily cold RSD have a much higher chance of developing severe late complaints, resulting in late referral, while an important number of patients with primary warm RSD, heal without major complaints. The population of a RSD clinic is therefore dominated by late cases with primarily cold RSD. Therefore, the three consecutive stages classically described in RSD, are only rarely seen. Probably the concept of three stages developed as the result of a patient selection bias.

INTRODUCTION

Reflex Sympathetic Dystrophy (RSD) is considered to develop according to three consecutive phases. This staging was introduced in 1938 by Sudeck¹ and more in detail by Maurer in 1940². In German literature, this staging was soon further modified and discussed^{3,4}. In English literature, the classical staging was first described by Steinbrocker in 1948⁵ and ever since duplicated - slightly modified - in many reports (table 4.1). In summary, it is generally accepted that RSD starts with an acute or warm phase in which pain, edema, a red and warm skin are prominent, that the second or dystrophic phase is characterized by vasomotor lability with alternating skin temperature and skin color, regression of edema, onset of patchy bone atrophy, hyperhidrosis or hypohidrosis, together with changes in nail growth and hair growth, and that the third, atrophic or cold phase is characterized by a cold and pale-blue shiny skin without edema, contractures and atrophy of all tissues.

Unfortunately, within the classical descriptions, there are conflicting statements concerning the occurrence of hyperhidrosis in phase 1 or 2, the occurrence of patchy bone atrophy in phase 1 or 2, or the presence of pain in phase 3. All authors agree as to the triphasic changes in skin temperature and that edema is present in acute cases only. However, Shumacker reported that skin temperature was cold from the start in 27 of 90 RSD patients⁶. Maurer, who supported the classical staging, described 9 patients, who 3 years after onset of RSD still showed all signs and symptoms of phase 2 without atrophy². Others stated that patients can cure in each phase and that RSD therefore does not have to go through each phase⁴. Steinbrocker, who reported a time-related staging first in 1948, reviewed the syndrome in 1968 and stated that the evolution into three stages may be seen on average, but is not a general rule. Some patients reach phase 3 within 4 to 6 months after onset, while others take a much larger time to reach phase 3^{5,7}.

Table 4.1 Classical staging of RSD

first stage	second stage	third stage
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Sudeck 1938¹

'Umbau'
warm, edema, cyanosis,
atrophy of muscles, no
trophic disturbances,

'Übergang'

'Atrophie', >3 months
cold, cyanosis, atrophy,
nails rippled pain
spontaneous and with

pain spontaneous and during exercise, hyperhidrosis, patchy osteoporosis

exercise, diffuse osteoporosis

Maurer 1940²

'physiologisothen reaktiven Umbau'. 2-3 months

warm, edema, limited range of motion, paresis with hypotony, hyperhidrosis, hypertrichosis, atrophy of muscles, no or patchy osteoporosis, normal sensibility, pain during exercise

'Dystrophie'. >3 months after onset cold, cyanosis, shiny skin, edema, (irreversible) limited range of motion, hypohidrosis, nails rippled, atrophy of muscles without hypotony, pain at rest and during exercises, patchy or diffuse bone atrophy, no sensory disturbances

'atrophie' > 9-12 months after onset cold, pale-blue, normal or shiny skin, no edema, paresis, irreversible limited range of motion, no pain during exercise, normal sweat secretion, normal hair growth, normal nail growth

Steinbrocker 1948⁵

lasting 3-6 months pain, warm, red/dusky pink, edema, limited range of motion, paresis, hyperreflexy, no osteoporosis

lasting 3-6 months warm/cold, no edema, fingers stiffen, atrophy of subcutaneous tissues and muscles, patchy bone atrophy

lasting many months cold, atrophy of all tissues, contractures of tendons, hypertrichosis, nodular fasciitis of palmar fascia, diffuse osteoporosis

When analyzing the literature, we could not find reports in which this staging is documented. Still, today this staging is used by many physicians and is even used to guide treatment^{2, 8, 9}. In a previous study¹⁰, we reported signs and symptoms of RSD in patients at the time of first consultation at our department.

When relating signs and symptoms to the duration of RSD, we could not confirm the classical staging. The natural history of RSD is unknown. As it is unethical to follow up patients without treatment, we therefore were unable to register the natural history in a prospective study. We analyzed all patients treated by our department, in order to find out whether three successive stages of RSD could be identified.

PATIENTS AND METHODS

All patients with a suspected diagnosis of RSD and coming to our attention were analyzed for signs and symptoms. RSD has not been clearly defined in literature. The criteria for diagnosis are summarized in table 4.2 and are discussed in a previous study¹⁰. Most patients were referred from other hospitals or were seen only once for second opinion or therapeutic advice. Prognosis in referred patients is probably worse than average because patients who favorably responded to treatment are not referred. Referred patients therefore may have other characteristics than the total population of RSD patients. Therefore only those patients were studied, which were coming from our own district, and treated in our institution.

Table 4.2 Diagnostic criteria for RSD

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

All patients were treated according a protocol including scavengers of oxygen radicals. This protocol is discussed elsewhere^{11, 12, 13, 14}. Skin temperature was carefully monitored during treatment. When skin temperature was cold, we treated patients with vasodilators or sympathetic blockade, in order to increase blood flow. This medication was continued up to the moment it could be stopped without recurrence of the cold skin temperature. If such medication was necessary for optimizing skin temperature these patients were considered as cold RSD.

RESULTS

From november 1984 to december 1991, 177 patients fulfilling the entry criteria were studied. 138 were female (78%) and 39 male (22%). Age varied between 13 and 83 years (median 46 years). In 121 patients (58%), RSD affected the upper extremity, in 56 patients (32%) the lower extremity. In 152 patients (86%), RSD followed trauma, in 11(6%) operation, in 6(3%) various other precipiting factors

and in 8 patients (5%) RSD developed spontaneously.

Skin temperature

In 144 patients (81%) skin temperature at onset of RSD - primary temperature - was warm, in 24(14%) cold, in 4(2%) the same as the contralateral extremity, and in 5(3%) primary temperature was unknown to the patient. In 25 patients with a primary warm RSD, skin temperature became cold in the course of the disease. In 119 patients with a primary warm RSD, skin temperature never became cold. From these 119 patients, 60 were cured at dismissal, but 59 patients still had complaints 3 months to 5 years (median 9 months) after onset of RSD. When skin temperature was cold at the time of examination, a treatment aimed at increasing blood flow was started in all cases. Therefore we can not tell whether primary cold RSD became warm because of the natural evolution of RSD or due to treatment.

All patients told us that the skin temperature varied during the day and/or the course of the disease. Skin temperature changed after exercise or after painful stimuli, changes in surrounding temperature, in dependency, during emotions or stress or exposure to sunlight. A warm skin temperature became warmer or turned to cold and a cold skin temperature became colder or turned to warm. These changes were invariably present at onset of the disease, during the course and, if not cured, at dismissal.

Table 4.3 Skin temperature at onset at RSD - primary skin tern temperature.

primary skin temperature	n	%
warm	119	67
became cold later	25	14
cold	24	14
no difference	4	2
unknown	5	3
Total	177	100

Edema

Edema was present in the acute phase in 146 patients (82%). At dismissal, 12 patients suffered from edema (13%).

Other signs and symptoms, such as hyperhidrosis or hypohidrosis, differences in nail growth and atrophy of tissues were not scored at dismissal so no data are available in this respect. Also unknown is whether patients developed patchy or diffuse osteoporosis because no radiographs were made during follow up.

DISCUSSION

The classical evolution of RSD into three consecutive stages could not be confirmed in the present study. In 14% of the patients, RSD started with a cold extremity, while a warm extremity persisted in 50% of the late cases. The alterations in skin temperature and skin color, classically described in the second stage of RSD, occurred in all three phases after exercise, painful stimuli, dependency or changes in surrounding temperature. In only 25 patients (14%) the disease developed according to the classical staging, that is warm at onset and ending with a cold extremity. In a previous study, we found that RSD patients with a primary cold skin temperature had a poor prognosis¹¹. Most patients with primary warm RSD heal without major complaints. Patients with primarily cold RSD have a much higher chance of developing severe late complaints, resulting in late referral. The population of a RSD clinic is therefore dominated by late cases with primarily cold RSD. Thus, the development of RSD according to three consecutive stages is not a uniformly present characteristic of RSD but seems to be the result of a patient selection bias.

As in this study the classical three stages of RSD could not clearly and regularly be found, and as the classical staging has no therapeutic or prognostic consequences, we suggest to drop this concept. Because the skin temperature at onset of RSD has prognostic value and therapeutic consequences¹¹, we suggest to identify two groups of RSD patients according to the skin temperature at onset of RSD: primarily cold RSD and primarily warm RSD.

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