WHAT IS REFLEX SYMPATHETIC DYSTROPHY ?

M. DRIESSENS[1], H. DIJS[1], G. VERHEYEN[1], P. BLOCKX[2]

In the literature there is no unanimity with respect to the diagnosis of reflex sympathetic dystrophy (RSD). Frequently, the diagnosis is established on mere clinical grounds. In our opinion, however, bone scintigraphy is of major importance for the diagnosis. Using this examination, true RSD can be clearly differentiated from other conditions which are incorrectly diagnosed and treated as RSD. If the bone scan is not suggestive of RSD, the clinical picture, radiological examination and vascular scan may lead to the correct diagnosis. This may be a pseudodystrophy, in which a hypovascularization is found right from the start, while in true RSD there is initially a hypervascularization. Other conditions which may be confused with RSD are causalgia, neurotic compulsive postures, hysterical conversion, malingering and even self-mutilation. In the spontaneous course of RSD three phases can be distinguished. Stage I is the warm or hypertrophic phase, stage II the cold or atrophic phase. Per definition the third phase corresponds to stabilization or, in rare instances, to healing. By means of the vascular scan the correct stage can be determined, and the results of treatment evaluated. Finally it should be noted that in children the condition is completely different from true RSD, as it concerns a pseudodystrophy or disuse-related dystrophy. This condition may also be seen in adults and adolescents, usually females. The bone scan is always negative. In this way bone scintigraphy constitutes the means to answer the question as to what RSD is and what it is not. An algorithm for the differential diagnosis is presented.

Keywords : Reflex sympathetic dystrophy ; bone scintigraphy ; pseudodystrophy ; children.
Mots-clés : Dystrophie réflexe sympathique ; scintigraphie osseuse ; pseudodystrophie ; enfants.

TERMINOLOGY

Reflex sympathetic dystrophy is a condition that has received numerous denominations during the course of history. This is because this pain syndrome is characterized by various clinical forms, precipitating factors, localizations, physiopathological hypotheses and diagnostic criteria. In the literature, there is still no unanimity with respect to the correct description of this disease, so that fundamental and comparative research remains difficult, and some clinical pictures are labeled as reflex sympathetic dystrophy while in fact they have nothing to do with it. In the future a classification will have to be made of the various syndromes which are currently reduced to the same denominator in the literature, but which are completely different from each other. It is difficult to accept that for example pseudodystrophy, resulting from disuse, is labeled as "reflex sympathetic dystrophy with decreased tracer uptake on bone scintigraphy" or, as it is termed in the French literature, "les formes froides". Scientifically it is difficult to imagine that the same condition would express itself in one patient with increased tracer uptake on bone scintigraphy and in another patient with a normal bone scintigraphy or decreased tracer uptake. Although reflex sym-

[1] University Hospital Antwerp, Department of Physical Medicine.
[2] University Hospital Antwerp, Department of Nuclear Medicine, Wilrijkstraat 10, B-2650 Edegem, Belgium.
Correspondence and reprints : M. Driessens.

pathetic dystrophy may express itself in the form of an incomplete and variable clinical picture, some syndromes which have one feature or other in common with reflex sympathetic dystrophy should still be regarded as separate conditions and accordingly be defined by different terms. If this is not done, different diseases are grouped together in one and the same patient population, and in this case it is not surprising that the results of fundamental or clinical research are often disappointing. The term reflex sympathetic dystrophy should therefore be reserved for the real Sudeck disease. "Reflex" and "sympathetic" refer to the pathophysiological mechanisms which are assumed to play a role in the disease, namely the autonomic sympathetic reflex loop. On the other hand, "dystrophy" refers to the trophic changes which occur not only in the bone, but also in the muscles and soft tissues and which frequently determine the clinical picture. A further advantage of the term reflex sympathetic dystrophy is that it does not refer to the localization of the condition, to the stage or to the precipitating factor. This cannot be said of many other current homonyms as is shown in table I. For purposes of convenience, the abbreviation RSD will be used from now on for the term reflex sympathetic dystrophy.

According to the evolution of the symptoms, various stages can be distinguished in the course of RSD : stage I, also called the hypertrophic phase or warm phase ; transition stage I-II, dystrophic phase ; stage II, atrophic phase or cold phase ; stage III, healing or stabilization.

In recent years, RSD has been associated with what is called "sympathetically mediated pain" (see below). This has led to much confusion in the literature, as not all syndromes that fulfill the criteria for sympathetically mediated pain are RSD, while the opposite does hold true.

PRECIPITATING FACTORS

Trauma and Surgery

The most frequent cause or precipitating factor of RSD is trauma (1). The most frequent localizations of traumatic RSD are the wrist and hand, the ankle and foot and the shoulder-hand syndrome. It usually follows distortions or fractures, but other trauma may also be involved as a precipitating factor (tendinous lesions, contusions, nerve lesions, ...). The risk of developing posttraumatic RSD is usually estimated at ± 1%, although some studies mention a higher percentage (up to 5%). In the literature figures of even more than 30% are reported after fractures of the wrist, but this is largely exaggerated and can be attributed to incorrect diagnoses. A high percentage of RSD,

Table I. — Common terms for R.S.D.

1. General terminology	2. Topographic reference
Acute bone atrophy	Shoulder-hand syndrome
Algodystrophy	Transient osteoporosis of the hip
Algodystrophies sympathiques	
Algoneurodystrophie décalcifiante	
Algoneurodystrophy	
Babinski-Froment syndrome	
Idiopathic neurodystrophic disorders	
Peripheral trophoneurosis	
Reflex-algodystrophy	3. Etiological reference
Reflex-dystrophy	Causalgia
Rhumatisme neurotrophique	Chronic traumatic oedema
Rhumatisme gardénalique	Drug induced neurotrophic disorders
Sudeck's atrophy	Leriche's posttraumatic osteoporosis
Sudeck-Leriche syndrome	Neurotrophic painful osteoporosis
Sympathetic reflex dystrophy	Ostéoporose douloureuse traumatique
Sudeck reaction	
Dystrophic reaction	

however, is seen after calcaneal fractures. This is one of the reasons why the recovery after calcaneal fractures is often disappointing. Also, RSD affecting one ray of the hand after localized trauma ("radial" RSD) is seen with increasing frequency, while numerous surgical interventions are also regarded as traumatic cause of RSD. Posttraumatic RSD constitutes 50 to 70% of the total number of cases of RSD (1, 23).

Neurological Conditions

The second most important cause of RSD is neurological lesions. One can distinguish central neurological conditions and peripheral nerve lesions. The latter usually falls under the posttraumatic form, in which case they involve causalgic syndromes. On the other hand, nontraumatic peripheral nerve lesions can also give rise to RSD (nerve root or plexus lesions, tumoral invasion, compression syndromes such as carpal tunnel syndrome -1,15). With respect to the central neurological conditions, the hemisyndrome after stroke or brain injury is usually involved. Most often this involves a shoulder-hand syndrome. After hemiplegia, distinction should always be made between an ordinary frozen shoulder, pseudodystrophy due to disuse and RSD of the shoulder-hand. To accomplish this, bone scintigraphy is indispensable. The vascular scan is used to determine the stage of RSD and the follow-up during treatment ; in pseudodystrophy it is used to determine the severity of the disuse and the hypovascularization.

Idiopathic or Primary Causes

RSD can occur spontaneously. Most frequently, either the shoulder or the wrist and hand are affected, although other localizations are possible. Depending on the author, they constitute 5 to 20% of the cases (1,15).

Other Precipitating Factors

Other less frequent causes of RSD involve various conditions which may affect the locomotor system such as infections, venous and arterial thrombosis, thoracic outlet syndrome, eczema, bursitis, gout,...etc. Also, a number of internal conditions may sometimes give rise to RSD. Among these are coronary artery disease, in which RSD of the shoulder occurs in 10 to 20% of the cases, or even a shoulder-hand syndrome may occur. The same can be said about other heart and pleuropulmonary conditions. A clear correlation also exists between the use of certain drugs and the occurrence of RSD (mainly RSD of the shoulder). Most frequently tuberculostatics are involved (mainly isoniazide), as well as barbiturates and antiepileptic drugs (24). Sometimes RSD is associated with hormonal factors. For example, in diabetic patients, RSD of the shoulder may occur. Thyroid hypofunction is also predisposing. During pregnancy, RSD of the hip is described, but is rare (8). Finally, RSD sometimes is attributed to metabolic factors, such as lipid disturbances (2).

Psychosocial Factors

These will be discussed below.

LOCALIZATIONS

RSD occurs most frequently at the wrist and hand, and at the ankle and foot (15). The shoulder-hand syndrome is also an important form. The other localizations occur less frequently. The limited forms of RSD require special attention ; among them are radial and parcellar RSD.These forms are frequently missed, although they are not so rare (11, 12, 25). In table II the possible localizations of RSD are listed. It should be noted that isolated RSD of the elbow is extremely rare and is found every now and then with tripolar involvement of the upper limb.

PATHOPHYSIOLOGY

With respect to the pathophysiology of RSD, numerous theories and hypotheses exist (fig. 1). The working hypothesis most widely accepted in the past was the local reflex theory of Leriche (26, 28). In this theory, it is assumed that an "excess" of afferent sympathetic impulses, originating from

Table II. — Topography

I. Isolated involvement (focal)
1. Upper extremities 1.1. R.S.D. of wrist and hand 1.2. R.S.D. of the shoulder 2. Lower extremities 2.1. R.S.D. of the knee 2.2. R.S.D. of ankle and foot 2.3. R.S.D. of the hip 3. Kümmell-Verneuil syndrome (spine)
II. Plurifocal involvement (unilateral)
1. Upper extremities 1.1. Shoulder-hand syndrome 1.2. Tripolar involvement of the shoulder, elbow and hand 2. Lower extremities 2.1. R.S.D. of the knee, ankle and foot 2.2. R.S.D. of hip and knee 2.3. Tripolar involvement of the hip, knee and ankle-foot
III. Bilateral involvement
1. Upper extremities 2. Lower extremities
IV. Combined involvement
V. Forms with limited localization
1. Partial R.S.D. 1.1. Radial : only 1 ray at the hand and foot 1.2. Zonal : e.g. a zone in the femur condyle 2. Parcellar R.S.D. 2.1. R.S.D. of the patella

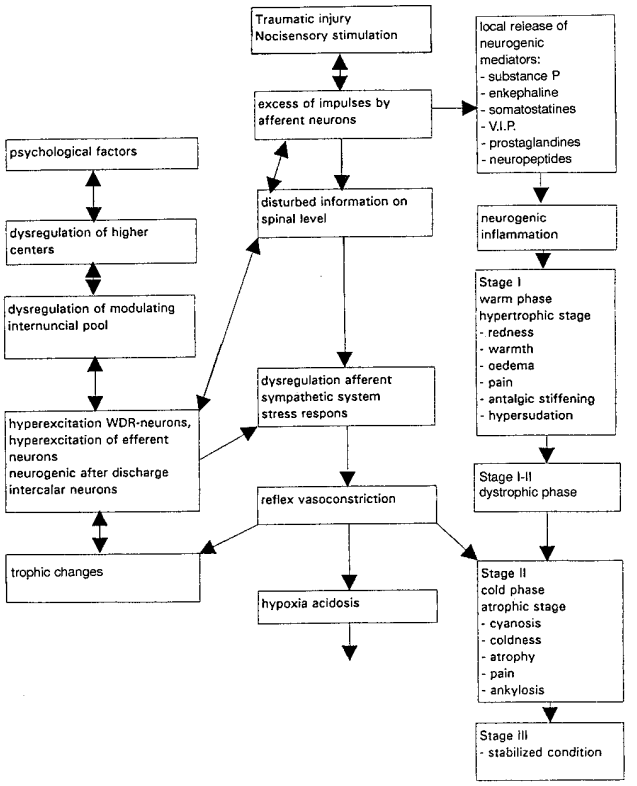


Fig. 1. — Physiopathology of R.S.D.

traumatized tissue, gives rise, through multiple connecting circuits, to a phenomenon of after-discharge and hyperexcitation of the efferent sympathetic fibers. This leads to an abnormal sensitivity of spinal nociceptive neurons of higher centers (29).

In the hypothesis of Roberts (34), specific classes of afferents are held responsible for chronic sympathetically-induced activation of spinal nociceptive receptors. In this hypothesis it is assumed that the primary pathology is to be found in the sensibilization of wide-dynamic-range (WDR)-neurons of the spinal cord which are disturbed by peripheral nociceptors. In their turn, efferent sympathetic neurons excite the peripheral sensory afferent nerve endings, which cause a tonic discharge of previously sensitized WDR-neurons and in this way effect a deregulation of the efferent sympathetic nervous system. In contrast to the reflex theory of Leriche, neither tissue damage nor nerve lesion is required in the hypothesis of Roberts. Sudden nocisensory stimulation evokes a stress response. By amplification of the stimulus-response relation the average activity at the spinal cord segment increases (4, 9). Pain perception and pain experience determine to a large extent the level of activity and the ability to select, and because of this they determine the degree of increase in orthosympathetic activity (4). In this way various psychological factors can act through this gate to give rise to a deregulation of higher centers by influencing endogenous pain modulating systems (beta-endorphines, cortisol).

Local release of various chemical mediators, especially neuropeptides and transmitters, gives rise to neurogenic inflammation. The list of neuropeptides has been growing over the last 25 years

of research (20, 33). Most studies concern bradykinin, histamine, substance P, encephalins, prostaglandins, leucotrienes, serotonin and vasoactive intestinal peptide. In the future, however, more studies are necessary to uncover the correct mechanism of these and more recently discovered mediators (cholecystokinin, neuropeptide Y, calcitonin-related peptide, somatostatin, angiotensin II — 3, 10, 35, 38, 39, 41, 42).

The pain problems in which the sympathetic nervous system is involved are characterized by one or more of the following symptoms : hyperpathy, pain of abnormally long duration, excessively intense pain, pain perception in a nonsegmental distribution, burning sensations, little tolerance for palpation or movement, and increased pain on the slightest weight bearing of the limb.

Not all forms of "sympathetically-mediated pain" fulfill the criteria of RSD : bone scintigraphy is negative in a number of cases ; blood flow is normal in other cases. The relationship between pain and the sympathetic nervous system is demonstrated by many clinical observations, but there is little scientific evidence. The pathophysiological mechanisms through which the sympathetic nervous system would influence pain perception are insufficiently known at the moment. The release of peptides may be responsible for the development of stage I, the warm or hypertrophic phase of RSD, characterized by a state of hyperemia and neurogenic inflammation. According to most authors, the disturbance of the autonomic nervous system mentioned above gives rise to the development of progressive hemodynamic changes. Due to a deregulation and hyperexcitation of the efferent sympathetic nerve fibers, there is a progressive evolution from increased to decreased blood flow. The latter is associated with stage II. In any case, the opposing vascular conditions in stage I and II have extremely important repercussions on the choice of treatment.

CLINICAL FEATURES

RSD is a syndrome characterized by the occurrence of one or more time-varying symptoms in one or more regions of the locomotor system. The clinical picture is variable and often incomplete with respect to the classic symptoms. In a large number of cases there are not even visible abnormalities, and the initial clinical findings only involve pain of abnormally long duration or progressively increasing pain in the area affected. This is particularly the case when large joints (shoulder, hip) are involved. Classically, three stages are distinguished in the course of RSD (21, 23, 28, 30).

Stage I : Hypertrophic stage

The most important factor is the phenomenon of pain. It usually involves a progressive increase in complaints, pain of abnormally long duration or an increase in complaints after an initial period of progressive improvement. Frequently, there is no correlation between the severity of the pain and that of the initial lesion. With the slightest suspicion, early RSD should be considered.

The pain may be gnawing, burning or stabbing. There may be an inflammatory pattern with pain at night and morning stiffness. However, there is also a typically mechanical pattern : usually the slightest loading or movement increases the pain. This is accompanied by progressive stiffening of the joints. On the other hand, there frequently exist inflammatory signs such as edema, warmth and redness (fig. 2). We call this a pseudoinflammatory syndrome, as laboratory examination does not show elevated inflammatory parameters. The

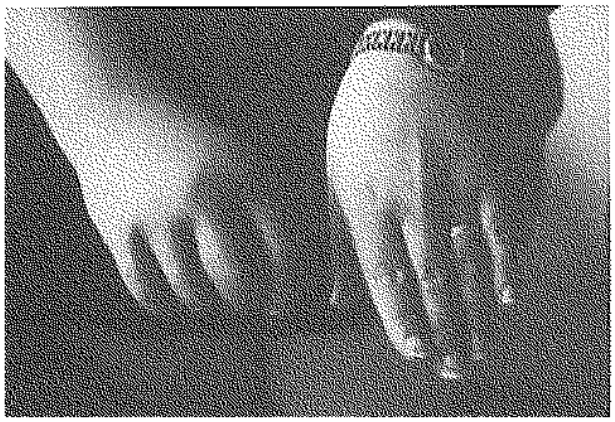


Fig. 2. — R.S.D., stage I, left hand.

problem in recognizing RSD is the fact that a noncomplicated posttraumatic condition frequently, although temporary, gives similar symptomatology, and because of this, the diagnosis of RSD may be established too late. The stage between stage I and II is called the transition stage I-II or dystrophic phase, in which the inflammatory signs gradually make way for trophic changes and cyanosis.

Stage II : Atrophic stage

This stage is characterized by persisting pain. The inflammatory pattern of stage I is replaced by opposite symptoms in the form of cyanosis, atrophy of the skin and sclerosis (fig. 3). Articular stiffening may increase to a true ankylosis. Contractures may appear, as do muscle tremors. Often there is hypersudation, and sometimes hypertrichosis. The transition, of course, takes place progressively and involves a variable amount of time (3 to 8 months).

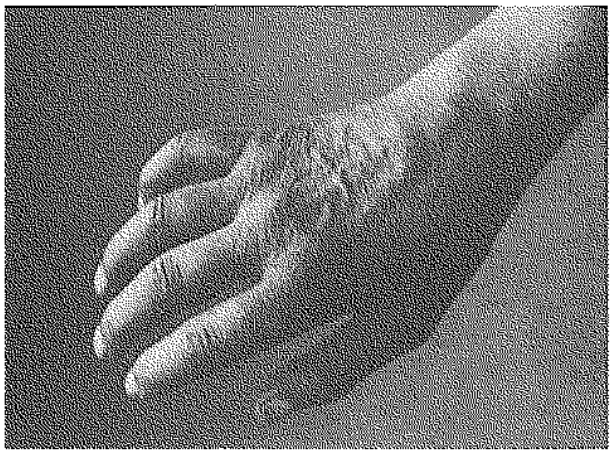


Fig. 3. — R.S.D., stage II.

Stage III : Stabilization

In due course (average 1 year), stabilization of the condition is seen. Rarely, spontaneous healing occurs. Usually, there remains atrophy of the skin with a thin, translucent character. Frequently, there is a cyanotic discoloration and coldness of the skin. The articular stiffening, established during stage I and II, persists if it has not been treated adequately (fig. 4). The pain usually decreases or disappears.

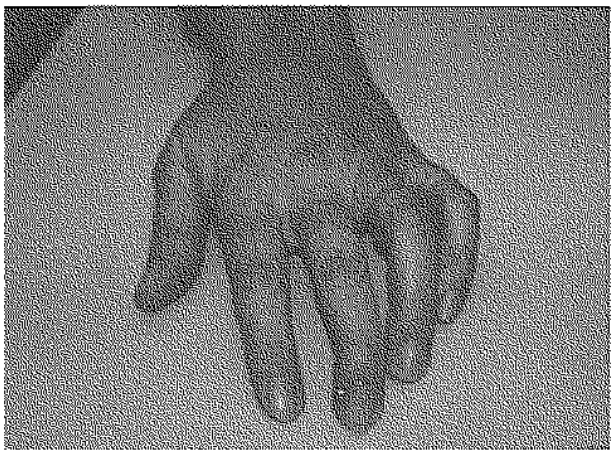


Fig. 4. — R.S.D., stage III (stabilization).

Evolution and Diagnosis

Clinically, it is impossible to establish the correct stage in an individual patient at a certain time. For this, the variation in clinical expression is too wide, while the symptoms may also overlap. For example, edema may persist in stage II, while cyanosis may already appear in stage I. There are large differences with respect to the severity of the condition and the duration of its course. There are mild forms and extremely disabling forms. Some forms evolve very rapidly, in which stage II is reached already after 2 or 3 months, while in other cases this stage is reached only after 8 months or sometimes after more than 1 year. A major problem is the fact that many other conditions may present with symptoms similar to those of RSD, stage I and/or II. Therefore, the application of strict diagnostic criteria is necessary. About this, however, there is still no consensus in the literature. Nevertheless, bone scintigraphy is undoubtedly the most important diagnostic means to distinguish RSD from other pain syndromes (causalgia, other sympathetically-mediated pain syndromes, disuse and pseudodystrophy, hysterical conversions). This distinction is essential for numerous reasons including adequate treat-

ment, fundamental research and clinical studies. The most frequently occurring mistake is to confuse RSD with pseudodystrophy (see below).

RADIOLOGICAL FINDINGS

The typical abnormality that occurs and becomes visible on x-ray examination is patchy osteoporosis of the affected parts of the skeleton (fig. 5). The affected side should always be compared with the healthy side, as the osteoporosis may be inconspicuous in the beginning (23, 36, 37). It should be noted that the radiological findings are not pathognomic for RSD. A similar patchy osteoporosis may be found in disuse and pseudodystrophy. Another feature of RSD is that the demarcation of the joints and the joint spaces always remains unaffected (except in the case of pre-existing traumatic lesions). The radiological examination may contribute to the diagnosis of RSD, but is not essential. In a large number of cases no abnormalities are found, especially in the early stages. It is not indicated to wait for the appearance of the radiological abnormalities, as this could cause an important delay in diagnosis and treatment. It is known that a cure can usually only be accomplished when treatment with calcitonin is started in a very early stage (14, 15). The radiological examination therefore gives typical, but not pathognomic findings in RSD and does not allow distinction to be made between the various stages of the disease, although this is essential in the correct choice of treatment.

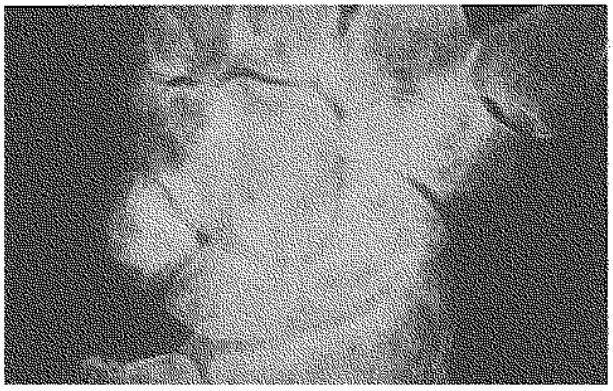


Fig. 5. — Patchy osteoporosis of the wrist.

IMPORTANCE OF BONE SCINTIGRAPHY

To establish the diagnosis of RSD, the performance of bone scintigraphy is an absolute necessity (15, 17, 22). Without this examination, the existence of RSD cannot be established with certainty. On the basis of clinical findings only, the diagnosis is impossible : the condition frequently presents with an incomplete clinical picture, so that the diagnosis may be missed ; on the other hand, many posttraumatic and other conditions show external features also found in RSD stage I or II. In view of the importance of early diagnosis and treatment, it should be considered severe negligence not to perform bone scintigraphy with the slightest suspicion of RSD. A missed or late diagnosis of RSD leads, in most cases, to a disabling process which becomes partly or completely irreversible in stage II, resulting in important lesions in the end. On the other hand many patients are wrongly labeled as "Sudeck" for complaints or symptoms that have nothing to do with RSD and therefore do not receive the appropriate treatment for their condition, with all the consequences. Futhermore, the label "Sudeck" becomes a diagnosed label for many patients who use it to gain attention, while for physicians it becomes a receptacle for all kinds of poorly-defined conditions, which are all approached and treated in the same way. The only result of this ineffective "group therapy" usually is the continuation of the problems of the patient concerned. Bone scintigraphy is the only method to distinguish between real RSD and a number of apparently similar conditions. Figure 6 shows an example of

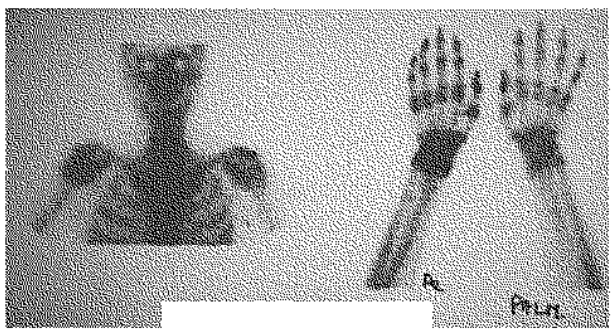


Fig. 6. — Bone scan of shoulder-hand syndrome.

the findings on bone scintigraphy in RSD (shoulder-hand syndrome). Typical is the increase in tracer uptake in the region affected. A special form is radial RSD, which is frequently overlooked or confused with posttraumatic arthritis. In this case also bone scintigraphy is the only examination to confirm the diagnosis (fig. 7).

To distinguish RSD from other pain syndromes, bone scintigraphy is by far the best means. In most cases the differentiation can be made without any problem. In some difficult cases, however, bone scintigraphy is not pathognomic, as in RSD of the shoulder. In table III guidelines are given for interpretation of the bone scintigraphy. Table IV shows an algorithm useful for differential diagnosis.

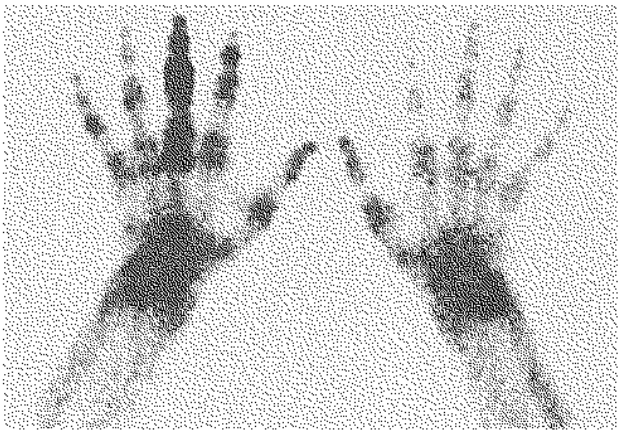
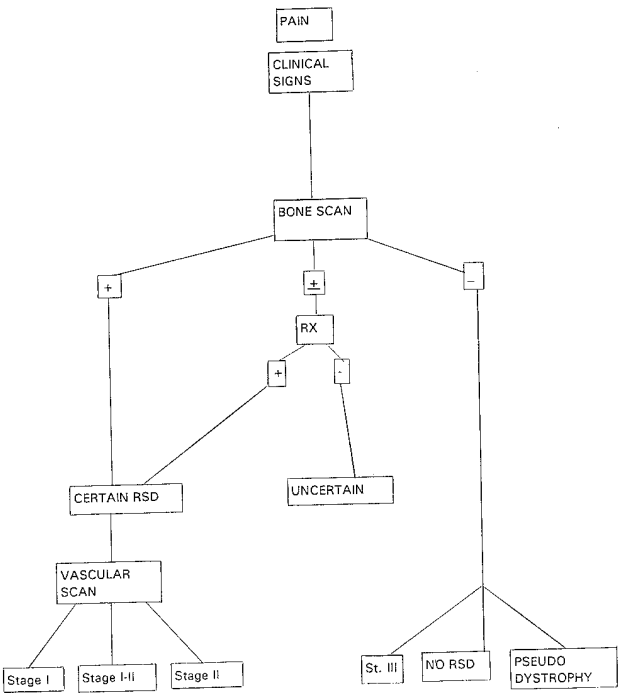


Fig. 7. — Clinical picture and bone scan in a patient with radial R.S.D. of the third ray.

Table III. — Differential diagnosis

1. Positive bone scan :		
– Fracture without R.S.D. :	localized hyperfixation at the fracture zone	
– Arthritis :	– rheumatic monoarthritis : – gout : – septic arthritis : – posttraumatic arthritis :	localized hyperfixation at the joint involved
– Arthrosis :	acute inflammatory increase of osteo-arthritis :	mostly 1 joint or multifocal and irregular localizations
– Aseptic necrosis :		focal
– Tumour : – benign : – malign :	 osteoid osteoma : primary metastases :	 very focal (hot spot) initially limited to 1 area of the bone focal or plurifocal myeloma : – sometimes negative – sometimes focal or plurifocal
– Vascular :		diffuse and slight hyperfixation with increased vascularization
2. Negative bone scan :		
Distortion (mainly wrist, ankle and knee) – Adhesive capsulitis (mainly shoulder), without R.S.D. – Arthrosis (sometimes slightly positive scan) – Myeloma – Phlebitis – Simulation — secondary illness gain — malingering – Psychosomatic – Causalgia – Sympathetically mediated pain syndromes (excluding R.S.D.)		
3. Hypofixation :		
– Disuse : disuse can lead to pseudodystrophy	hypofixation due to hypovascularization	

Table IV. — Algorithm for differential diagnosis



STAGING : VASCULAR SCINTIGRAPHY

Because of the different underlying hemodynamic conditions, it is of major importance to know the correct stage of RSD when the patient is seen for the first time, especially to establish the most adequate therapy. Also, it will allow some prediction with respect to the prognosis.

As was indicated previously, the correct stage cannot be determined with certainty on the basis of the clinical picture, x-ray examination or bone scintigraphy. The clinical symptoms are very variable and may overlap. For example, cyanosis may already be present in an early stage (stage I). On the other hand, edema may persist in stage II. Also, it should not be forgotten that the original lesions of an accident may mask the symptoms of RSD : for example, posttraumatic arthritis may result in a persistance of inflammatory features, although the RSD has evolved to stage II. The radiological features of RSD usually involve patchy osteoporosis, which, however, is seen both in stage I and in stage II. Bone scintigraphy remains positive during the complete evolution of the condition. It only becomes negative in stage III. For these reasons it is necessary to have a suitable and additional method in order to determine the correct stage. This can easily be accomplished by means of vascular scintigraphy (7, 17, 18). In this examination, labeled serum albumin is used. The choice of this product is determined by the fact that this tracer remains within the blood vessels. The product is injected intravenously and imaging is started immediately ; the affected side is compared with the healthy side. In this way, the local vascularization in the affected region can be determined in an objective way in comparison with the contralateral healthy side. The tracer counts are measured over 20 minutes. In this way one gets vascular curves, characterized by an inflow phase and an equilibrium phase. The inflow phase reflects the local blood flow ; the equilibrium phase is a measure for the local blood volume. When the curves of the affected region and the corresponding healthy side are similar in shape, it may be concluded that the vascularization is comparable and normal. When the curve is steeper and higher in the affected region, one may conclude that in this region hypervascularization exists. A slower and lower curve indicates hypovascularization.

Studies have shown that in RSD stage I there is always hypervascularization (15). The inflow curve shows an earlier start and a steeper gradient on the affected side, while the equilibrium phase is always higher (fig. 8). Therefore, in stage I the local blood flow and blood volume are increased. The opposite is found in stage II : decreased blood flow and blood volume (fig. 9). In stage III, normalization of the vascularization is sometimes found, but most of the time a more or less pronounced hypovascularization persists. By means of this technique, it is possible to determine the correct stage in each patient with RSD. When bone scintigraphy is positive and there is a state of hypervascularization, one may conclude that the patient is in stage I. In stage II, on the other hand, there is a positive bone scan and hypovascularization (fig. 10). In case of known RSD, it

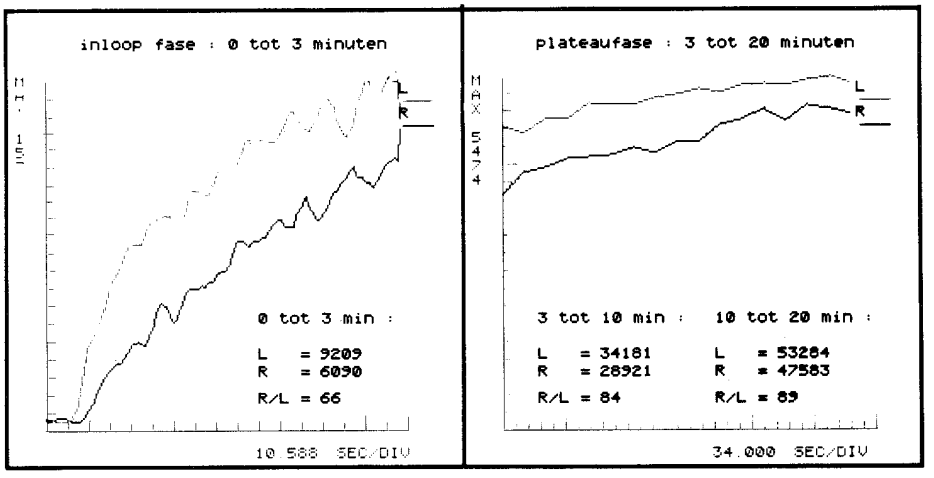


Fig. 8. — Vascular scan in R.S.D. stage I, illustrating hypervascularization in the affected area.

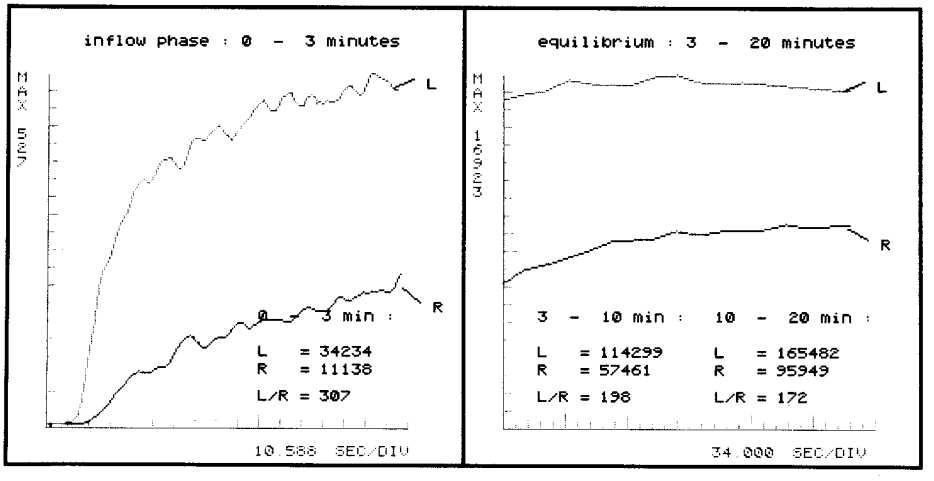


Fig. 9. — Vascular scan in R.S.D. stage II, illustrating hypovascularization.

may be concluded that the patient is in stage III (stabilization) when the bone scan has become negative and hypovascularization is found. A similar combination, however, is also seen in pseudodystrophy. During the transition from stage I to II (dystrophic phase), apart from a positive bone scan, vascular parameters with near normal values are found, as there is a progressive transition from the hyperemic to the atrophic and cyanotic condition (fig. 10). Usually the clinical picture shows a parallel evolution, but, as mentioned above, this is not always the case. For this reason it is essential to request a vascular scan for each patient with RSD.

Obviously, this is of importance to determine the correct treatment : in stage I the therapy of choice is the administration of calcitonin. In stage II, on the other hand, vasodilating drugs are indicated and by no means calcitonin, as the vascular situation is completely the opposite of stage I.

It should be noted that the early inflow curves obtained by means of a 3-phase bone scan cannot be used to determine the stage of RSD, as the "vascular" parameters may be influenced in an adverse way by the rapid uptake of the tracer used in the bone (5, 6, 15, 16). This could lead to incorrectly classifying a stage II as stage I.

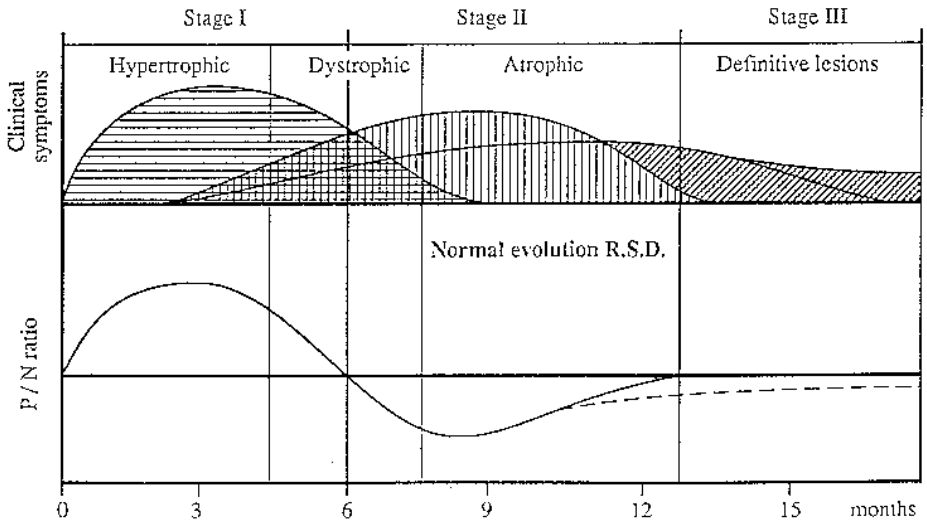


Fig. 10. — Evolution in clinical signs, bone scan and vascular scan during the spontaneous course of R.S.D.

PSEUDODYSTROPHY

In recent years it has become very clear that a strict distinction should be made between RSD and pseudodystrophy (18). These are completely different conditions, which, however, are still frequently placed under the same classification (12, 31). An essential difference between the syndromes is the fact that the diagnosis of RSD can only be made on the basis of a positive bone scan, whereas pseudodystrophy is characterized by either a normal bone scan or decreased tracer uptake compared to the healthy side. Furthermore, inflammatory signs, characteristic of RSD stage I, are never seen in pseudodystrophy. The condition is the result of disuse of the affected limb and is characterized clinically by progressive cyanosis, atrophy and severe function impairement. This leads to complete disuse and loss of function. The lower leg is especially affected, but the condition may also occur in the hand (fig. 11).

Clinically, the picture resembles that of RSD stage II, but there are essential differences which allow the differential diagnosis : in RSD stage II stiffening of the affected joints occurs systematically, evolving into ankylosis ; this is caused by retraction and sclerosis of the joint capsules, ligaments and tendons. This phenomenon is never seen in pseudodystrophy ; the articular status usually remains normal and in case of limited joint mobility, the cause is a contracture of some muscle groups resulting in a compulsive posture (fig. 12). Under general anesthesia, for example, the limited joint mobility in RSD cannot be released, while the opposite is true in pseudodystrophy. Of course, when the pseudodystrophy persists for a long period of time, secondary joint stiffening may occur. However, this condition is usually reversible, in contrast to the ankylosis seen in RSD. There is also a clear difference in patient population :

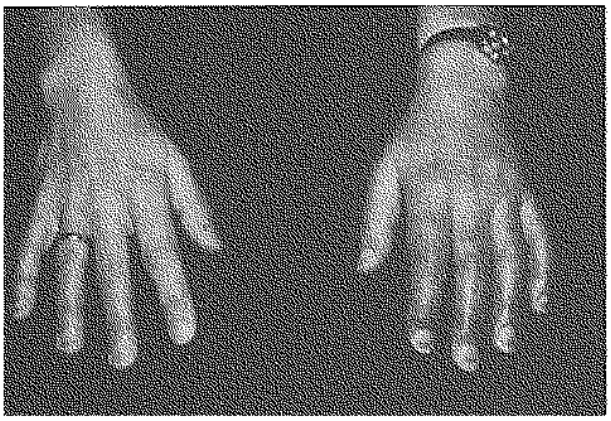


Fig. 11. — Pseudodystrophy at the left hand.

Fig. 12. — Pseudodystrophy with compulsive posture at the right hand. Note that the passive mobilization appeared to be perfectly normal.

RSD can affect the entire adult population without preference for age or gender, while pseudodystrophy typically occurs in children, adolescents and young adults, especially females. A further essential distinction with RSD is the fact that in pseudodystrophy a causal link with psychosocial problems is nearly always found. These are relational problems, whether or not associated with hysterical conversion. Very typical is the aberrant father-daughter relation with excessive concern and affection. In the child or young adult this leads, frequently after minor trauma, to somatization and loss of function. Factors such as narcissism and attention seeking also play a part.

One of the essential elements of treatment is therefore psychotherapy. It is best to conduct comprehensive psychometric testing first, but during the further psychotherapeutic support a suitable physician-patient relation must be established to uncover the underlying problems, to bring the patient to a clear understanding of his or her illness and to resolve the downward somatizing spiral and in this way alleviate the problems. This may require time-consuming discussions, and not infrequently the environment of the patient has to be involved in this approach. In many cases the underlying problem can be solved in this way, while in the meantime the physical lesions may be improved or cured by means of intensive rehabilitation and suitable medication. With respect to the latter, vasodilating drugs are indicated. More invasive dilating drugs (Ismelin blocks, Bier's blocks, sympathectomy), are to be avoided at all costs as they have no indication except in unusual cases. The hypovascularization is temporary and can be normalized completely by resolving the disuse, providing intensive support and full-time rehabilitation and temporarily using oral vasodilating drugs. Moreover, it is essential to avoid aggressive treatment (neurostimulator, morphine pump) in these patients. These methods have entirely different indications and may push patients with pseudodystrophy even further in their social isolation. Furthermore, they never effect a cure ; on the contrary, they have an even more disabling character for the patient concerned. Cases are even known in which an amputation was performed! The same remarks also apply to RSD (see below). Nevertheless, in some cases the underlying psychological problems are so severe and fundamental that almost no solution is possible. In this case sustained psychiatric and social support is preferred to the "palliative care" mentioned above. Apart from the oral vasodilating drugs, other medications may also have an indication during the rehabilitation process : e.g. analgesics when the pain syndrome is prominent, anti-inflammatory drugs when there are still sequelae of initial trauma, psychiatric drugs to be adjusted to individual needs. The accompanying personality structure and/or possible psychiatric components may vary widely : elements of depression, hysteria and even psychosis.

It is frequently indicated for medical specialists to consult the family physician because in this way unknown or hidden facts may sometimes be revealed. Conversations with parents, spouse or employer may also be very useful. On the other hand, there is frequently a correlation between the problems of the patient and his relationship with his environment, so that some resistance may be met in uncovering these problems. In many cases it is essential to remove the patient from his environment and to admit him to a specialized rehabilitation center. The treatment applied may involve full-time therapy including heat therapy, electrotherapy, hydrotherapy (alternating warm and cold baths, underwater massage and mobil-

ization), exercise therapy and gait rehabilitation. It is to be noted that the pain threshold may be exceeded during exercise and mobilisation physiotherapy. The use of crutches is decreased progressively. Occupational therapy (observation and treatment) is also indicated. One has to be careful with group therapy as this may lead to conspiracy among the patients with the same problems. On the other hand, it may also have a stimulating effect on the patient who sees his "fellow patient" in a more advanced stage of treatment. Hard confrontations with reality are best avoided (for example showing photographs to the patient in which his affected limb shows completely normal mobility under general anaesthesia), but may have a convincing effect in selected cases.

Nerve phenolization because of compulsive postures should be avoided as a spontaneous remission of the "pseudocontracture" can be achieved with the more adequate therapy mentioned above. In short, in many cases, a complete cure can be achieved.

PSYCHOSOCIAL FACTORS : CAUSE OR CONSEQUENCE ?

It is very difficult to rely on the literature with respect to the possible existence of psychosocial influences and/or consequences in the course of RSD, as in the past no distinction was made between real RSD and other syndromes with a comparable clinical picture (e.g. pseudodystrophy). For this reason numerous descriptions of psychiatric disorders and aberrant behavior are found, whether or not in combination with paranormal personality structures or psychosocial and relational problems. On the other hand, many cases are known in which no apparent psychological aberrations exist (32, 40).

In any case, RSD is hardly ever seen in ambitious and motivated patients (sportsmen). Furthermore, the condition is seen much more frequently in countries with a highly developed social security system (health insurance, sickness benefits). When the urge and need to perform are present, RSD seems to occur only rarely. On the other hand, the condition appears to occur more frequently in cases of excessive self-pity and in narcissistic persons. Also, secondary sickness benefits and retirement neurosis can sometimes not be denied. When the injured region is regarded by the patient himself as a tender part that has to be cherished and surrounded with all possible care, then the road is open for RSD. In such cases the essential criteria for successful rehabilitation and cure are not met due to the abnormal behavior of the patient (lack of cooperation and persistence). Without making any effort, such patients await the course of events meekly ; they are sometimes even proud of their RSD. For this reason, such patients should be treated early to show them the end of the tunnel and to provide the necessary therapeutic means. If this is not done, progression to stage II is difficult to stop, and at this stage a complete cure is rarely achieved.

Apart from the rehabilitation program, sufficient time should be made to build up a positive physician-patient relationship, which must allow the patient to have a new look at his problems. Frequently one is confronted with signs of lack of understanding, fear and loneliness. When an aberrant personality structure is present (and this certainly is the case in a higher percentage than in the common population), a psychiatrist should be involved. This may also be essential in the differential diagnosis. Too many patients have the label "Sudeck", while in fact they have nothing to do with the disease (see above). The psychoanalysis will be able to differentiate this problem from others including forms of hysteria and retirement neurosis. Some authors completely reject the existence of psychosocial causes and regard problems in this area as a consequence of the disease.

MEDICOLEGAL PROBLEMS

The cause-effect relationship between RSD and an accident can be accepted without doubt when the condition occurs in conjunction with the accident and at the site of the initial lesion. If there is recovery from the initial injury followed by a period without complaints and RSD develops thereafter, then the cause-effect relation should be denied. It becomes more difficult, when RSD occurs in the context of incomplete cure of the

initial trauma and complaints continue. Then it will have to be established to what extent the sequelae of the initial lesions were stabilized and after what period of time the RSD appeared. Any relationship between RSD and the accident can only be accepted when RSD appears within a relatively short time (a few weeks at the most). Another problem arises when RSD occurs in a region other than that of the initial trauma (e.g. RSD of the foot after injury to the knee). The judgement of this will at least in some cases be arbitrary.

Some authors argue that an accident is not always the cause of RSD, but frequently the reason that evokes an underlying RSD or a predisposition to RSD. Frequently, however, this involves a pseudodystrophy, which is usually insufficiently differentiated from RSD. In these cases the cause-effect relationship with the accident should be denied and the sequelae of both conditions should be separated. In any case, the different procedures with respect to accidents at work, personal accidents, and common law should always be taken into account. These may vary according to the insurance policy, legislation and domicile (19).

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SAMENVATTING

M. DRIESSENS, H. DIJS, G. VERHEYEN, P. BLOCKX. Wat is een reflexsympathische dystrofie ?.

In de literatuur bestaat er geen eensgezindheid voor de diagnose van reflexsympathische dystrofie (RSD). Dikwijls is de diagnose gebaseerd op klinische gronden. Volgens onze opinie echter is botscintigrafie belangrijk voor een duidelijke diagnose. Met dit onderzoek kan echte RSD worden gedifferentieerd van andere aandoeningen welke een RSD simuleren. Wanneer de botscintigrafie niet suggestief is, kan het klinisch beeld, radiologisch onderzoek en vasculaire scan leiden tot een correcte diagnose. Dit kan pseudodystrofie zijn met een hypovascularisatie in de eerste fase terwijl bij een echte dystrofie een hypervascularisatie aanwezig is. Andere toestanden zijn causalgie, neurotische compulsieve houdingen, hysterische conversie, automutatie en malingering. In het spontane verloop van RSD kunnen 3 fasen worden onderscheiden. Fase 1 is de warme hypertrofische fase, stadium 2 is de koude of atrofische fase. Per definitie is de derde fase een stabilisatie en in zeldzame gevallen heling. Door de vasculaire scan kan een correcte staging worden uitgevoerd. Het resultaat van de behandeling kan worden geëvalueerd.
Tenslotte moet er worden genoteerd dat bij kinderen de toestand verschillend is van echte RSD en het vaak een disuse gerelateerde dystrofie of een pseudodystrofie is. Deze toestand zien we soms bij adolescenten en volwassenen, meestal vrouwen. De botscan is dan altijd negatief.
Wij kunnen besluiten dat de botscintigrafie een belangrijk element is om de vraag van 'Wat is RSD', te beantwoorden.

RÉSUMÉ

M. DRIESSENS, H. DIJS, G. VERHEYEN, P. BLOCKX. A quoi correspond la dystrophie réflexe sympathique ?

La littérature n'est pas unanime en ce qui concerne le diagnostic de la dystrophie réflexe sympathique. Fréquemment, le diagnostic est établi sur base de critères cliniques mal définis. Nous pensons que la

scintigraphie osseuse est particulièrement importante pour poser le diagnostic. Selon ce critère, la dystrophie sympathique vraie peut clairement être différenciée d'autres affections pour lesquelles un diagnostic incorrect de dystrophie réflexe sympathique est posé, avec instauration d'un traitement inapproprié. Si la scintigraphie osseuse n'est pas suggestive de dystrophie réflexe sympathique, le diagnostic clinique et radiologique et les scintigraphies vasculaires peuvent permettre de poser le diagnostic correct, par exemple de pseudodystrophie, avec hypovascularisation d'emblée, alors que dans la dystrophie sympathique réflexe vraie, existe initialement une hypervascularisation. D'autres affections qui peuvent être confondues avec la dystrophie réflexe sympathique sont la causalgie, les postures névrotiques compulsives, la conversion hystérique, la sinistrose et même l'automutilation. Dans l'évolution spontanée de la dystrophie réflexe sympathique, trois stades peuvent être distingués. Le stade I correspond à la phase chaude ou hypertrophique ; le stade II, à la phase froide ou atrophique. Par définition, la troisième phase correspond à la stabilisation, ou, rarement, à la guérison de l'affection. La scintigraphie osseuse permet de déterminer la stade précis de la maladie, et d'évaluer les résultats du traitement. Finalement, il faut noter que chez l'enfant, l'affection est totalement différente de la dystrophie réflexe sympathique de l'adulte, et correspond à une pseudodystrophie ou à une dystrophie d'immobilisation. Cette dernière affection peut également être observée chez les adultes et les adolescents, surtout chez les jeunes femmes. La scintigraphie osseuse est habituellement négative. Ainsi, la scintigraphie osseuse permet de répondre à la question de savoir si l'affection présentée par un patient correspond ou non à une dystrophie réflexe sympathique. Un algorithme de diagnostic différentiel est présenté.