Original Research

REFLEX SYMPATHETIC DYSTROPHY IN CHILDREN: AN ORTHOPEDIC PERSPECTIVE

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ABSTRACT

To design diagnostic criteria for reflex sympathetic dystrophy (RSD) and to initiate a prospective treatment protocol, we reviewed our experience with 49 episodes of RSD in 36 children. There were 24 females and 12 males; mean age at diagnosis was 13.4 years (range: 8 to 19); mean time from pain onset to correct diagnosis was 9.2 months (range: 1 to 53). Lower extremity involvement predominated. Pain was “severe” in 61%, and skin color changes, swelling, hyperesthesia, abnormal skin temperatures, muscle weakness, and decreased range of motion were all present in at least 75% of cases. Osteopenia was observed in 15 of 38 radiographs; of 24 bone scans, 7 were normal, 11 showed increased uptake, and 6 demonstrated decreased uptake. Of the 23 children who had psychological evaluations, 83% revealed some type of significant emotional dysfunction. Analgesic and antiinflammatory medications were not helpful, nor were local injections or regional blockades effective. An inpatient diagnostic and rehabilitation program for treating chronic pain, including orthopedics, rheumatology, psychology, and twice-daily physical therapy was most likely to lead to resumption of age-appropriate activities. Despite extensive physiological testing, physician, parent, and/or patient reluctance to accept absence of a primary organic disease was common. We present diagnostic criteria for pediatric RSD.

Evaluation and management of chronic pain disorders in children is a frequent occurrence for the pediatric orthopedist. However, the pain nomenclature is complex and may vary over a wide range of clinical expressions. A good example is the reflex sympathetic dystrophy (RSD) syndrome, which is also called shoulder-hand syndrome, causalgia, reflex neurovascular dystrophy, or Sudeck’s atrophy. The earliest comprehensive description was recorded in 1864 by Mitchell, who observed patients with RSD following civil war wounds with associated damage to major peripheral nerves. Today, these patients would be classified as having major causalgia, a specific subcategory of RSD.

The major clinical features of RSD include localized dependent edema, altered skin temperature (coldness or warmth), increased or decreased sweating, hyperesthesia or hypoaesthesia, decreased range of motion in affected joints, and dystrophic skin changes. Several of these findings may be present at diagnosis, although few patients will exhibit all of them simultaneously.
Until recently, the literature focused mainly on RSD in adults.2,3 RSD in children is described infrequently; only eight cases were reported prior to 1978.2,5-7 Since then, several series have focused on differences in presentation and treatment responses between children and adults.6-15 Children less often have major antecedent surgery, may have a larger psychological component, and often appear to respond to physical therapy.8,14,15

The presenting symptoms of RSD lead it to cross a wide spectrum of clinical disciplines. Primary care physicians often refer children with RSD when early intervention fails to achieve resolution. Because there is often a history of minor trauma, an orthopedic surgeon may become involved. Since there is a presumption of acute trauma, it may take several visits before it becomes apparent that the child’s level of discomfort is inconsistent with the degree of trauma sustained.

Because the physical findings and severity of RSD can be quite variable, evaluation of treatment alternatives is very difficult. Further, there is no unifying causal theory that explains the signs and symptoms of RSD. Treatment modalities have proliferated because no single strategy provides remission in a majority of patients; thus, few have been studied by standardized research designs.

Interventions for reflex sympathetic dystrophy have included physical therapy, splinting and immobilization, transcutaneous electrical nerve stimulation (TENS), local anesthetic and/or steroid injections, intravenous or regional perfusion of steroid compounds, oral corticosteroids, sympathetic blockade using both injections and permanent surgical interruption, and intravenous regional sympathetic blockade using various medications.3,7,8,16-20 Where controls have been included in treatment protocols of RSD, a significant portion of nontreated patients demonstrate improvement, and therefore the efficacy of all interventions must remain in question. Given the possibility of treatment side effects, it is important to determine which children require aggressive management.

Psychological disturbances tend to be frequent in children with RSD.6-14,15 It is difficult to determine which children have preexisting emotional difficulties that might predispose them to developing RSD versus those who manifest psychological changes in response to their chronic pain and impairment. Adolescents with RSD may be particularly difficult to assess because they often demonstrate situational adjustment reactions and/or altered coping mechanisms.

Our study goals were to develop baseline data from which we could design diagnostic criteria for pediatric patients with RSD and to initiate a prospective treatment protocol to assist orthopedists and pediatricians in the earlier diagnosis and management of this complex disorder.

**MATERIALS AND METHODS**

The records of children with a discharge diagnosis of RSD from 1940 through June 1989 were reviewed at the authors’ institution by a pediatric orthopedist, physical therapist, and rheumatologist. The minimum diagnostic criteria used to diagnose RSD are listed in the Table. Diagnosis of RSD was infrequent before 1950, becoming more common with each successive decade. A 60-item data collection record was developed and included demographics, mode of onset, number of prior physicians seen, medications and surgical interventions employed, duration of hospitalization, and treatment modalities utilized.

**RESULTS**

Forty-nine episodes of RSD were identified in 36 children. The age at disease onset ranged from 8 to 19 years (mean: 13.4); there were 24 females and 12 males. The mean time from disease onset to correct diagnosis was 9.2 months (range: 1 week to 53 months; median, 4 months). A possible initiating traumatic event was recalled by 31 children and was believed by the patient or parents to be responsible for the development of RSD. These included: 7 vehicular accidents, 15 simple lower or upper extremity sprains, 5 sports-related injuries, and 4 post-surgery or fracture. Lower extremity involvement predominated: 16 children had episodes primarily involving the ankle region, 12 had symptoms affecting the knee, 8 had wrist and hand symptoms, 2 patients had shoulder involvements, and 2 had symptoms about the hip (Fig 1). One child had involvement of the left index finger only. The average patient saw
1.6 subspecialty physicians prior to referral (range: 1 to 3), with five patients having seen multiple physicians, including orthopedists, neurologists, neurosurgeons, and physiatrists, in decreasing order of frequency.

At presentation, 22 children (61%) believed their RSD pain was "severe," and 13 (36%) rated it as moderate. Thirty-five of 36 patients had skin color changes, including dependent rubor, cyanosis, and mottling. Ten children had continuous swelling, whereas 16 had intermittent swelling that was usually activity related; 10 patients had no significant swelling. Twenty-eight patients (78%) had hyperesthesia, whereas five (14%) were believed to have normal skin sensitivity. Four children (11%) had increased skin temperature, 12 (33%) decreased, and 6 (17%) had variable skin temperatures at different times of observation. Active range of motion was normal in 6 patients (17%), minimal to moderately limited in 14 (39%), and severely limited in 12 children (33%) (Fig 2).

Osteopenia was noted in 15 of 38 radiographs (40%) from 15 children; the remaining 23 children had normal radiographs. Bone scans were obtained in 24 patients: 7 (29%) were normal, 11 (46%) had increased uptake, and 6 (25%) demonstrated decreased uptake. Of 23 children in whom a detailed psychological assessment was performed, 19 (83%) demonstrated some type of significant emotional dysfunction. Evidence of stressful situations, such as physical or sexual abuse or divorce of parents, was often documented.

Resolution of RSD was defined by complete resumption of age-appropriate physical activities, including school attendance, physical education, and extra-curricular activities. The most common form of management was intensive physical therapy. Multiple modalities were employed, including progressive weight-bearing, passive and/or active range of motion exercises, progressive ambulation, heat and/or cold applications, pressure desensitization, short-term splitting, transcutaneous electrical nerve stimulation, and biofeedback. No single physical therapy modality, or combination thereof, appeared clearly superior.

Numerous analgesics and/or antiinflammatory medications were also given, both prior to referral and during hospitalization. Patients and/or parents often noted subjective improvement, but this was usually not corroborated by independent observation or functional assessment. Eight children received local injections (22%), most often at the ankle or knee. Only seven patients (19%) received regional sympathetic blockade. Generally, neither injection nor blockade was effective by itself.

The average time to initial recovery of RSD was 9 months (range: 1 to 36 months) in the 25 cases where resolution could be clearly documented. In 12 children there were one or more recurrences; many of these were at the initially
involved site, although a few developed symptoms at another location. Eleven patients had continued limitations of physical activity, due to their initial RSD episode, at their most recent follow-up visit.

**CASE EXAMPLES**

**Case 1.** A 15-year-old girl presented on 12/88 with a complaint of pain with intermittent numbness and paresthesias of 18 months' duration involving her right arm, hand, and shoulder. She also complained of weakness and decreased sensation in the fourth and fifth digits of the right upper extremity. She had sustained an injury 18 months earlier, which was described as a depression of the right shoulder with a forced deviation of the neck to the left. She was referred to the neurology service for a possible brachial plexus injury. Examination revealed decreased range of motion of the shoulder and mild weakness in hand grip, but was otherwise normal. There were no findings to suggest a neurovascular sympathetic nervous system dysfunction at initial presentation.

Electromyograms (EMGs) revealed a questionable, mild decrease in conduction velocity consistent with a possible mild brachial plexus injury in the "upper roots." On 2/89 the patient presented with a complaint of "blotchiness, sweating, and swelling" of the right upper extremity. At that time findings consistent with sympathetic nervous system involvement were first noted. The skin had a mottled appearance and there was mild soft puffiness edema noted about the hand and elbow region. Over the ensuing 2 months the patient's condition steadily deteriorated. The range of motion of the shoulder, elbow, wrist, and hand continued to decrease in spite of aggressive outpatient physical therapy. She was admitted to the hospital on 4/24/89, and over the next 4 weeks underwent evaluation by orthopedics, rheumatology, neurology, and psychology. The initial psychological evaluation revealed that the patient was an "overachiever." She had extensive after-school work and recreational involvements with various sports activities. There were no apparent serious emotional disturbances. Therapy consisted primarily of behavior modification techniques, TENS treatment, vigorous range of motion and desensitization therapy, and minor anti-inflammatory medications. The patient was discharged after 4 weeks of inpatient treatment with modest improvement. During the next 30 days the patient seemed to improve significantly, with markedly increased range of motion of the elbow and wrist; and less improvement in shoulder stiffness.

The patient sustained a fall in 9/89 which aggravated the feeling of decreased sensation in the fourth and fifth digits of the right hand. A repeat EMG was performed, which was interpreted as being "equivocal" for mild residual upper nerve root damage in the brachial plexus. The patient was given therapeutic doses of lorazepam (Ativan) and carbamazepine (Tegretol) without documented improvement. A therapeutic trial of amitriptyline (Elavil) failed to show any improvement. In 6/89 the patient called to schedule an interview with the child psychologist who had initially evaluated her during her inpatient stay. At that time the patient discussed an episode of childhood sexual abuse which had occurred approximately 4 years prior. It was obvious that this was a source of significant psychological disturbance. A long and intense series of psychological interviews and evaluations proceeded throughout the course of the patient's continued medical care. Significant psychological stress was attributed to this episode. The situation apparently resulted in criminal charges against the abuser and a court trial was scheduled. This situation understandably caused marked increase in the patient's psychological disturbance and caused serious behavioral changes and alterations in the family environment.

During this time and over the next 6 to 8 months the patient's upper extremity symptoms increased remarkably and her functional situation deteriorated. For weeks the patient's right upper extremity was essentially of no functional benefit, and was carried in a sling against medical advice. In 12/89 the patient had improved significantly, although no major changes in her treatment had occurred. She continued to receive outpatient physical therapy modalities on a twice-weekly basis. No further medications were prescribed. In 1/90 the patient had a vascular headache episode, and after further extensive evaluation migraine headaches were diagnosed. The patient was initially treated with cyproheptadine (Periactin); however, this caused no improvement and therefore she was treated with propranolol (Inderal). This affected some significant improvement in the frequency of her migraine headache episodes.

In 2/90 the patient had approximately 60% use of the right upper extremity. During this time the criminal proceedings against the alleged child abuser were concluded and the patient's psychological counseling frequencies decreased. She sustained significant improvement in the functional use of the right upper extremity. On a trip to Europe in 7/90 the patient fell, sustaining an injury to the right knee, with clinical findings of swelling and diffuse tenderness about the medial aspect of the knee. She was placed on crutches, but on returning to the United States was noted on outpatient evaluation to have significantly altered functional use of the right lower extremity. To forestall development of RUL in the right lower extremity, the patient was admitted for a 1-week course of physical therapy and
intense knee rehabilitation. This was successful, and over the next 2 months the patient's right knee symptoms completely resolved. The right upper extremity symptoms had significantly improved, although intermittent discomfort and a persistent subjective decreased sensation of the fourth and fifth digits were noted by the patient.

At the time of her last evaluation the patient's right upper extremity was nearly normal in its function, with some slight decrease in active abduction of the shoulder. She continued to have intermittent complaints of subjective decreased sensation in the fourth and fifth digits of the right hand.

**Case 2.** A 12-year-old girl was well until "her brother pushed her into a bathroom cabinet," and she bumped her left knee. There were two emergency room visits for severe pain and "swelling." Hip and knee radiographs were normal. She was then seen by an adult orthopedist who placed her in a cylinder cast for 2 weeks. Following cast removal, the girl exhibited stiffness and severe pain, was non-weight bearing, and required crutches. At 3½ weeks post-injury, the patient was referred to us for evaluation.

There was marked wasting of the left quadriceps, a 10° flexion contracture, and only 5° of total motion. She also exhibited bluish discoloration, hyperesthesia, and decreased temperature below the knee. Radiographs of the leg revealed moderate osteopenia. Twice-daily physical therapy and a psychology consultation were ordered. The mother was noted to have a chronic pain syndrome; the patient appeared to get secondary gain from complaints of pain.

Following a 6-week hospitalization for intensive physical therapy, pool walking, nightly use of a continuous passive motion machine, psychology therapy, and behavior modification, the patient was minimally ambulatory. The left leg still exhibited mottling, decreased temperature, skin hypersensitivity, muscle atrophy, and a range of motion from 5° to 80°. Numerous frustrations were voiced by the family regarding both insufficient and conflicting information from physicians, nurses, and therapists concerning the etiology and ideal treatment for RSD in children. Both parents were employed in the health care field at another local hospital, and had complicated the treatment program by recurrently seeking independent opinions and advice from physicians inexperienced with RSD. This had resulted in increasing parental anger and confusion and may have contributed to the slow course of recovery.

A program of weekly education/counseling meetings with both parents and the patient was instituted. The mother's work schedule was modified to permit her to be home when her daughter arrived after school. A personality conflict with one teacher was resolved. Paternal demands for "straight A" school performance were modified. The father became less confrontational, demanding, and perfectionistic. The child was permitted to participate in the decision about whether to attend parochial or public school. The entire family became more relaxed and able to achieve compromises. Thereafter, the RSD resolved very rapidly, as did the chronic complaints of headache. School attendance and performance improved. A reduced frequency of physical therapy was ordered. The patient was fully recovered 5 months after hospital discharge. There have been no recurrences in over 2 years.

**Discussion**

Reflex sympathetic dystrophy is less common in children than in adults, but new awareness of the problem is emerging.8,21,22 Our study of 49 episodes in 36 children confirms the major delays in diagnosis previously suggested by others.8 Many patients saw multiple physicians before RSD was recognized. Often, expensive and/or lengthy evaluations for organic disease were done while attempting to explain an ill-defined syndrome of pain and limited function. Once diagnosed, the children entered a comprehensive rehabilitation program, often under the care of an orthopedic surgeon. Usually, simultaneous efforts were continued to exclude the possibility of organic disease.

The term RSD originally derived from descriptions of adults whose symptoms were relieved by diagnostic or therapeutic sympathetic blockade. But whether the RSD symptom complex is caused by dysfunction of the sympathetic system or increased sympathetic tone due to altered pain responses remains controversial. Excellent reviews of RSD in adults are available by Schutzer and Gossling1 and Schwartzman and McEllan.19

Most of the current proposed classification systems for RSD were developed in response to evaluations of adult patients. Many of these revolve around the time phase of the patient's RSD and are rather arbitrary. Certainly, specific physical findings (such as permanent skin pigmentation changes) are related to the duration and severity of patient symptoms, and perhaps these classification systems will be of some benefit in the management and evaluation of adult patients. It was clear on completing our review of our patients that there were no subcategories of patient involvement. Adult classification systems have included such subdivisions as minor and major causalgia, which traditionally have been useful in distinguishing relatively minor involvement due to nerve injury from major dysfunction due to injury of major nerves. Minor causalgia is generally understood to include RSD symptoms related to an injury.
(usually by blunt trauma) to a minor peripheral sensory nerve in an extremity. Major causalgia is a term reserved for symptoms caused by the complete severance (usually by penetrating trauma such as a gunshot wound or knife injury) of a major mixed motor and sensory nerve of the upper or lower extremity. Since we dealt exclusively with pediatric patients, and since we are not located in a major urban setting prone to injuries due to major penetrating trauma, we had no instances of RSD due to major causalgia.

Three prerequisites for developing RSD in adults have been suggested: 1) a painful lesion, 2) an abnormal autonomic reflex, and 3) an inherent tendency or constitutional predisposition that renders the patient susceptible to developing RSD.\textsuperscript{1,13} Such patients may also be sympathetic hyperreactors, who can be identified by a history of increased sweating of the palms, poor cold tolerance, and emotional lability. Additionally, they may have “inadequate personalities” noted on psychological testing. A typical patient with RSD may demonstrate a dependent personality, low pain threshold, insecurity, fearfulness, instability, and chronic complaining, and may seek to blame others for their condition.\textsuperscript{3,13} These patients often do not comply with advice, appear to sabotage treatment programs, avoid responsibility, and thus adopt a “sick” role. Children with RSD often exhibit variations of these behaviors.\textsuperscript{14}

The value of the bone scan has not been established in children. Recent reports have observed increased, decreased, and normal radiotracer uptake in children with RSD.\textsuperscript{23} These variations appeared to depend on the duration of illness prior to the bone scan. Although we found no value in planning treatment, the scan was helpful early in the search for unrecognized organic disease.

On completion of our review, we were able to reach a conclusion that minimal diagnostic criteria for pediatric RSD should include pain out of proportion to the inciting event and documented dysfunction of the neurovascular system as outlined in the Table.

We place treatment emphasis on rehabilitation, physical modalities to achieve pain relief, and functional improvement. Local injections of a long-acting anesthetic and/or corticosteroids were only occasionally used to assist in regaining lost joint range or function. Often, injections had more diagnostic than therapeutic value, and assisted the localization of symptoms. In contrast to adults with RSD, sympathetic blockade and/or medications rarely were effective, although many children received acetaminophen or aspirin for pain.

Intensive physical therapy appeared to be the most effective and least invasive treatment for children with RSD, and complications were extremely rare. In addition, the close, supportive relationship that developed among therapist, child, and parents appeared to be very beneficial. Hospitalization for management of RSD is often necessary. This allows intensive focus on behavior modification, which can be extremely difficult to achieve on an outpatient basis. The initial costs of inpatient care are high, but still quite small in comparison with the costs of life-long disability or life-long inappropriate utilization of medical services. Methods may eventually be developed to effectively treat children with RSD as outpatients, but this is presently difficult, as management expertise in RSD is not commonly available. Thus, referral to a tertiary care center is often needed.

In a typical case, we would propose that the patient be scheduled for an elective admission for an initial period of 2 weeks. A psychology evaluation of both patient and family should occur within the first 2 days. A very rigid schedule of twice-daily physical therapy sessions should be developed. Any further testing designed to rule out organic dysfunction (ie, bone scans, EMGs) should be scheduled so as to avoid canceling any physical therapy sessions. The patient should continue with all regular school work assignments while in the hospital. A specific schedule for “homework” in the evening hours is important. Beneficial behaviors (working “hard” in PT) are rewarded by extra evening privileges, visitors, etc. The use of injections and medications must be individualized. During the treatment period, the treatment team must meet regularly to discuss the progress of the patient in physical therapy and to discuss information developed by the psychologist. The team must avoid generating conflicting opinions concerning the etiology of the RSD and the recommended treatment. It is helpful for one member of the team to be the primary source of information for parents and patient.

The use of unnecessary protective devices (ie, crutches, splints, slings) is definitely contraindicated. The patient must be made full weight bearing as soon as possible. Specific physical therapy modalities are beyond the scope of this article, but will often include pool exercise, desensitization techniques, passive and active range of motion exercise, bicycle therapy, biofeedback techniques, and occasionally transcutaneous nerve stimulation.

In working with families of children with RSD, it is often necessary to move slowly from an organic disease model to a rehabilitation model in order not to alienate patients and parents, many of whom have difficulty under-
stiling that psychological factors and learned pain behaviors play important roles in the continuation of symptoms.14 Many children and their parents are reluctant to admit that ineffective stress coping mechanisms, or other psychological factors, play important roles in pain perception and maintenance of RSD. In children, secondary gain may take the form of significant time away from school-related stresses, or in focusing attention on a difficult or stressful family situation. Especially with parental conflict, the child with a medical illness can artificially bind the parents together in their common quest for a solution.

Although psychological factors play important roles in children with RSD, psychological interventions have not been consistently effective. It is difficult to develop a significant working relationship among psychologist, child, and family during a short hospitalization, particularly if the initial diagnostic and therapeutic mind set is intensely physiologic. Much work in both pediatric and adult RSD has focused on finding explanatory or causal psychiatric diagnoses. In our patients, symptom resolution, which includes the reversal of true secondary physiologic changes, has resulted from improved problem solving and coping mechanisms. Thus, counseling from a psychiatrist, psychologist, primary physician, or another health care worker enables the child and family to find more effective solutions to specific difficulties.

Psychological assessment may help to understand disease initiating or maintaining factors and will commonly identify areas of secondary gain. However, many patients and/or parents are reluctant to accept the absence of a primary organic disease, that symptoms can be mediated by emotional factors, and that these symptoms can be eliminated by altering behaviors. In these circumstances, impatient evaluation and management for pediatric RSD is often the most cost-effective approach.

In summary, RSD is a challenging pediatric diagnosis, and the frequency of initiating emotional factors is becoming more commonly recognized. A multidisciplinary diagnostic and treatment approach utilizing orthopedics, rheumatology, physical therapy, and psychology can achieve good results in the majority of children.

REFERENCES

EDITORIAL DISCUSSION
ORTHOPOEDICS: Does the age of the patient determine the specific treatment program?
Stanton et al: The majority of our patients presented in the early adolescent age range. It
would certainly be true that a patient presenting at the age of 5 or 6 would be treated in a somewhat different fashion from an adolescent patient. Adolescent patients are much more likely to have developed significant aberrations related to psychological stress coping mechanisms and would likely benefit from more intensive psychological assessment and support than would a younger patient. In general, we found that younger patients have far fewer identifiable secondary gains related to perpetuation of their symptoms, and in general their improvement is easier to effect in a more timely fashion. Younger patients respond more predictably to "fun" activities, especially water therapy (swimming pool, physical therapy). Younger patients are less likely to tolerate the "annoying" aspects related to TENS and are especially resistant to any type of therapeutic or diagnostic injection therapy.

**ORTHOPEDICS:** What are the three most important diagnostic criteria in RSD?

**Stanton et al:** By far the most important diagnostic finding in RSD is pain out of proportion to the inciting event. Pain in the absence of objective physical findings is another important diagnostic finding. Direct evidence of altered neurovascular function as manifested by three or more of the listed findings in the Table constitute the minimum diagnostic criteria for pediatric RSD.

**ORTHOPEDICS:** How does RSD differ in adults and children?

**Stanton et al:** In adults, RSD predominantly affects the upper extremities. Children are much more likely to show lower extremity involvement, especially about the knee and ankle. The treating physician is much more likely to suspect RSD in an adult patient; therefore, diagnosis in children is characteristically much more delayed. Psychological stresses play a very important part in perpetuating symptoms in children. Although the psychological situation is important in children and adults, the types of stresses that affect the child's life are much different from those typically seen with the adult patient. A child psychologist is very important in the evaluation and management of these patients.