An Overview of Amplified Musculoskeletal Pain Syndromes

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ABSTRACT. Children may have a wide variety of amplified musculoskeletal pain syndromes that may or may not be associated with overt autonomic signs and may be diffuse or localized to one body part. It is most common in pre- to adolescent girls. Hallmarks of the diagnosis include increasing pain over time, allodynia, an incongruent affect, disproportional dysfunction, and the absence of other causes. Psychological distress within the child or family is apparent in most, but not all, since it also is associated with injury or illness. Once the diagnosis is established, all medicines and testing are stopped. A sympathetically driven pain model is used to explain the pain to make it understandable. Treatment is an intense exercise program; ours is 5 hours daily. We focus on functional aerobic training specifically using the involved body part such as sports related drills, running, play activities, and swimming. Allodynia is treated with desensitization such as towel rubbing. A psychological evaluation is done and specific psychotherapy is recommended if indicated. The average duration of the daily program is 2 weeks with a 1 hour home program being done for another 2 to 8 weeks. After one month roughly 80% of the children have no pain and are fully functional, another 15% are fully functional with mild or recurrent pain; 5% are not better. Significant relapses are infrequent; 15% require retreatment. Five to 10% of the children will develop a different symptom of psychological distress. At 5 years, 90% are doing well. (J Rheumatol 2000;Suppl 58:44–48)

Key Indexing Terms:
REFLEX NEUROVASCULAR DYSTROPHY FIBROMYALGIA PHYSICAL THERAPY
CHILDREN MUSCULOSKELETAL PAIN SYMPATHETICALLY MEDIATED PAIN

A child in intense pain with an amplified musculoskeletal (MSK) pain syndrome can challenge all the diagnostic and therapeutic skills of the physician. Additionally, the duration and degree of pain and frustrations at failed diagnoses and treatments distress the child and family. I will outline my diagnostic approach, treatment, and outcome based on treating over 500 such children[1].

Pain is a subjective experience and the degree to which it is felt and to which it affects the child's life (such as activities of daily living, seeking medical care, school attendance, sports participation, and play) is individual. The report of pain should be taken at face value[2]. The amount of noxious stimuli does not correlate to the degree of pain; thus, it is imperative not to be judgmental, even if the quality and quantity of pain and pain behaviors are incongruent.

Children with amplified pain can present with a variety of symptoms and signs. The established nomenclature artificially divides these conditions into separate groups, whereas it is my experience that children with amplified MSK pain are on a continuum in which specific subsets may overlap (Figure 1)[3-6]. Nevertheless, the various subsets include those with overt autonomic signs (consistent with complex regional pain syndrome types I and II[2]), those without autonomic signs (with either continuous or intermittent pain) (consistent with psychogenic pain, localized or diffuse idiopathic pain[3,5]), those with painful points (at sites associated with fibromyalgia in adults[7]), and those who are hypervigilant. An individual child may present with a single subset, but there are many children in whom these subsets coexist (such as a hand with autonomic signs and a foot without) or have a different subset during a subsequent relapse. However, there are consistent features to the presentation, examination, treatment, and, perhaps, etiology.

The etiology of amplified MSK pain in children is unknown but most episodes seem to be causally related to injury, illness, or psychological distress, although age and hormonal and genetic factors may play a role. In most children, psychological distress is usually significant either as a cause or effect of the pain and dysfunction, although high quality controlled studies are lacking. Commonly there is a history of minor trauma, whereas many adults with complex regional pain syndrome have preceding major trauma; this is unusual in children. In our clinic, the most common illness associated with amplified pain syndromes is arthritis.

The clinical presentation of children with amplified MSK pain is remarkably consistent (Table 1). Most are female (80%) and tend to be of higher socioeconomic status. The mean age of onset is 12 to 13 years; it is unusual under the age of 8 years.

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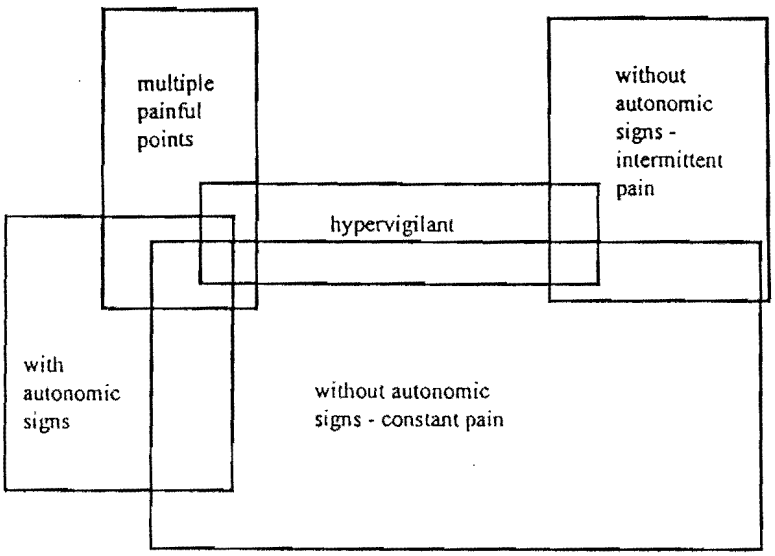


Figure 1. The overlap of the various pain amplification syndromes in children. Reproduced with permission from: Sherry DD. Pain syndromes. In: Isenberg DA, Miller JJ II, editors. Adolescent rheumatology. London: Martin Dunitz; 1999[1].

Table 1. Clues to childhood amplified pain from the history and examination.

Preadolescent to adolescent age (mean age 12 years)
Girl (80%)
Increasing pain after minor or no trauma
Marked disability
Crawls around house or up stairs
Allodynia (unable to bear light touch, clothing, or bedcovers)
May have symptoms of autonomic changes (any)
Cold
Color changes: purple, blue, or gray
Clammy
Edema
Worse or no better with splint or cast
Unsuccessful prior therapies
High level athlete, dancer
Typical personality
Mature beyond years
Excels at school and extracurricular activities
Perfectionistic
Pleaser (meets the needs of others at her own expense)
Role model for chronic pain or a similar pain in family or friends
Recent major life event (may be multiple)
Moving
Change of school, friends
Divorce
Change in nuclear family
Mother acts as spokesperson
Incongruent affect for amount of pain reported
La belle indifference about pain and disability
Compliant when requested to use limb
Autonomic signs, especially after use
Allodynia with a variable border
Pain is not restricted to a dermatome or peripheral nerve
Otherwise a normal neurological examination

A characteristic pattern emerges during the history and examination. Initially, most think the pain started as the result of minor trauma that may not even be remembered ("maybe someone stepped on my foot"). Many will have a history of autonomic signs (such as being cold or blue), but these signs may be transient. Allodynia (pain generated by normally nonpainful stimuli) is manifest by the inability to clothe or bathe the painful area and can be quite extreme — "the breeze of someone walking by hurts." During the examination one is frequently struck by the interdependency, or enmeshment, between the patient and parent (usually mother, but can be the father) in many of the families. The parent will speak for the child even when the child is directly addressed. The child is usually very cheerful, even when reporting severe pain (up to 10 out of 10 pain) and has *la belle indifference* about both the pain and dysfunction it causes. The amount of dysfunction can also be out of proportion to what one would expect. These children may have missed months of school because it hurts too much in the morning to go. Frequently they are unable to do even light chores, sometimes even feed themselves. Having to crawl around the house or up stairs is common, whereas crawling is exceeding rare in children with inflammatory joint or bone disease. Frequently, children with amplified MSK pain are overly mature and accomplished in school and in extracurricular activities, and are described by their parents as perfectionistic, empathetic, and pleasers.

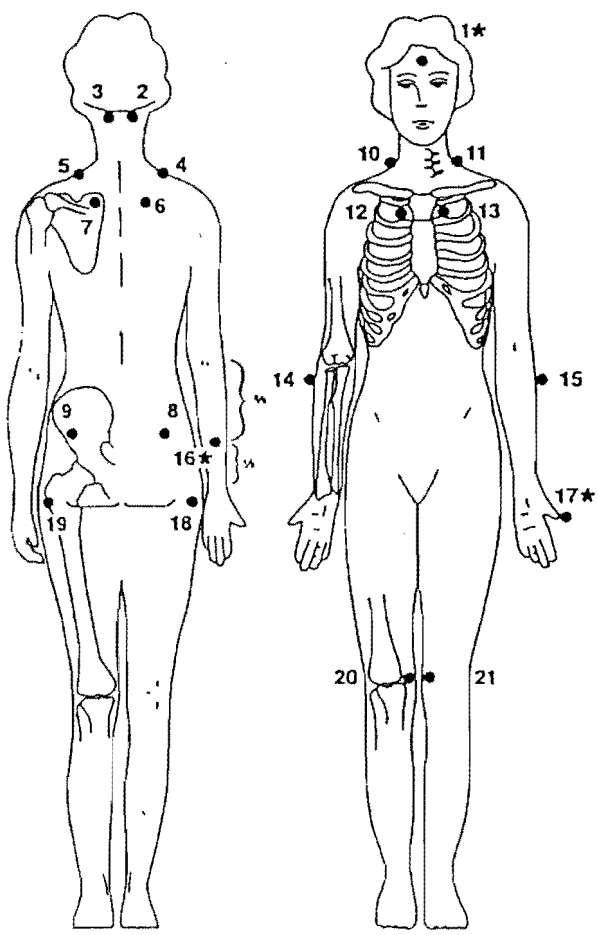
The examination varies somewhat between the different patterns of amplified pain. Children with overt autonomic dysfunction generally have continuous pain in a limb (more commonly the lower extremity), usually can recall exactly when it began, and some will display marked pain behavior such as crying or screaming with pain. Children without overt autonomic dysfunction are less sure of the exact onset, are more likely to have multiple sites involved including centrally located sites such as the back, chest, or jaw, a

longer duration of pain, and can have intermittent pain and dysfunction (usually following strenuous physical activities). Children with painful points (in both the sites described in fibromyalgia and overlying muscle bellies) are more likely to be depressed and fatigued. Children who are hypervigilant seem to pay too much attention to regular body sensations and interpret these sensations as painful. The pain can be fleeting, lasting only a few seconds, or last several hours (especially after minor trauma). Hypervigilant children are usually quite anxious, fearing that these pains are indicative of a serious disease. Children with any form of amplified pain may, in addition, be hypervigilant.

The review of systems may uncover conversion symptoms. The most frequent symptom is numbness, but paralysis, inability to walk due to dizziness or knees giving way, or changes in organs of sense, such as blindness, may be reported. A role model for chronic pain or pain similar to the child's pain may be evident from the family history. The social history frequently reveals recent major life events such as moving, school changes, school stress, changes in the nuclear family, or illness or deaths in the family or close family friends.

Notable elements of the examination include: (1) the absence of an underlying disease (although I have seen pain amplification in addition to organic diseases such as arthritis, enthesitis, infection, muscular dystrophy, and cerebral palsy); (2) a normal neurological examination (paying special attention to sensory tests); and (3) ascertaining allodynia. Allodynia is present if, when lightly touching the skin or gently pinching a fold of skin, pain is reported. The border of where the allodynia begins can vary greatly on repeat testing. The majority of children will be strikingly compliant; children nonambulatory due to foot pain will stand on the foot and even walk when requested to do so. If signs of autonomic dysfunction are not initially present they may appear after the child uses the limb. When testing for painful points (Figure 2), control points such as the forehead, mid-forearm, shin, and thumbnail should also be tested. In some children, even touching the hair is painful. The children with widespread pain seem, to me, to be different from those with fibromyalgic painful points only; they are usually younger, less depressed and fatigued, and respond more quickly to treatment.

Throughout the examination, even when the child reports



MANUAL TENDER POINT SURVEY

Patient Instructions

Read to the patient the statement: *"Various areas of your body will be examined for pain. Please say Yes or No if there is any pain when I press a specific point."*

Explain the scale by reading to the patient: *"I want you to rate the intensity of the pain on a scale from 0 to 10. 0 is no pain and 10 is the worst pain that you have ever experienced."*

After testing survey site 9, the patients should be reminded of the meaning of the pain scale to reinforce their understanding of the range.

Survey and Control* Sites

SEATED	Right	Left
Mid-Forehead (*):	1.___★	
Occiput: Suboccipital muscle insertions	2.___	3.___
Trapezius: Midpoint of upper border	4.___	5.___
Supraspinatus: Above medial border of scapular spine	6.___	7.___
Gluteal: Upper outer quadrant of buttocks	8.___	9.___
Low Cervical: Anterior aspect of intertransverse space of C5-7	10.___	11.___
2nd Rib: 2nd costochondral junction	12.___	13.___
Lateral Epicondyle: 2 cm distal to epicondyle	14.___	15.___
Dorsum R Forearm (*): Junction of proximal ⅓ & distal ⅔	16.___★	
L Thumbnail (*):		17.___★
SIDE		
Greater Trochanter: Posterior to trochanteric prominence	18.___	19.___
SUPINE		
Knee: Medial fat pad proximal to the joint line	20.___	21.___

Positive Survey Sites _____ Total Survey Site Scores (SS) _____

Positive Control Sites _____ Total Control Site Scores (CS) _____

Fibromyalgia Intensity Score ($^{SS}/_{18}$) _____

Control Intensity Score ($^{CS}/_{3}$) _____

Date _____ Examiner _____ Patient _____

Figure 2. Location of the painful and control points in children with one form of amplified musculoskeletal pain. From Okifuji, *et al*[7], reproduced with permission.

7. Okifuji A, Turk DC, Sinclair JD, Starz TW, Marcus DA. A standardized manual tender point survey. I. Development and determination of a threshold point for the identification of positive tender points in fibromyalgia syndrome. J Rheumatol 1997; 24:377-83.

severe pain, she usually has a markedly incongruent affect, smiling while using the limb. Although this gives the observer a sense that the pain is not real or as intense as reported, it is imperative to reassure the child that you understand how much it hurts and you need to see how much she can do. The pain is real.

Blood tests, if done, are always normal unless there is an additional underlying condition. Radiographs are normal or reveal osteoporosis due to disuse. Technetium bone scans are usually normal or show decreased blood flow on the blood pool phase and decreased uptake on the delayed images[8]. It is rare to see the spotty increased uptake pattern typical of adult disease.

Once the diagnosis is made, it is imperative to stop further medical investigations and drug treatment, fully explain the condition, and begin exercise therapy.

A model, based on sympathetically mediated pain amplification, is used to explain to the child and family why the pain is so intense (Figure 3). Although there is no consensus regarding the pathophysiology of these conditions, this model provides a tangible framework for the child to understand the pain, reinforces that the pain is real, and allows discussion of the possible etiologic factors including trauma, illness, and psychological stress. Additionally, using it we introduce our therapeutic plan.

To avoid emotionally laden terms, I call all forms of amplified pain reflex neurovascular dystrophy. This avoids the word sympathetic that suggests that the child is just looking for sympathy. These families, more than families of children with other rheumatic diseases, search for all available information and using the name reflex neurovascular dystrophy it is hard to find much on the Internet. The information on the Internet dealing with reflex sympathetic dystrophy and fibromyalgia is mostly about adult disease and, for these families, can be alarming and misleading.

We treat these children with aggressive, one-on-one, functionally focused intense aerobic exercise therapy, generally 5 to 6 hours a day, daily (Table 2). All aids, including crutches, are discontinued the first day unless the child is too weak to ambulate safely. It is important that the exercise therapy be one-on-one so that the therapist can modify the exercises minute by minute to gain maximum function. Group therapy can deteriorate into children competing to see who hurts the most. Function is the focus of therapy so, for example, if the child could go up and down 2 flights of stairs in 55 seconds that morning, she would have to beat 55 seconds in the afternoon, even if it meant multiple attempts. A skilled therapist functions as a coach using the child's innate compliance and competitiveness to motivate and improve function and reward each improvement, however small.

The allodynia is treated directly with desensitization with towel and lotion rubs, massage, and weight bearing. Wearing appropriate clothing, socks, and shoes is part of the desensitization process.

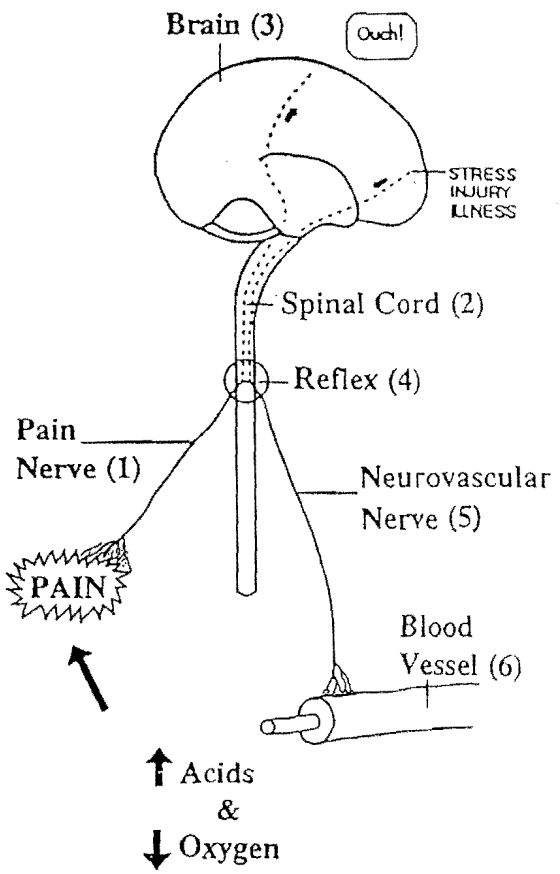


Figure 3. A model to explain the etiology and pathogenesis of pain amplification to the families and children. From Sherry[1], reproduced with permission. A painful signal goes from the pain nerves (1) up the spinal cord (2) to the brain (3), where the signal is interpreted as painful. In this condition, there is an abnormal short circuit in the spinal cord (4) to the neurovascular nerves (5) that go to the blood vessels (6). These nerves make the blood vessels to the bone, muscle, and skin get smaller, decreasing the amount of oxygen these tissues receive. The lack of oxygen and build-up of acids causes intense pain. The pain then goes back up the pain nerves, to the spinal cord, across the abnormal reflex, and back to the neurovascular nerves. This vicious cycle goes on and on, amplifying the pain. Illness, injury, or psychological stress can cause the abnormal reflex in the spinal cord. Reproduced with permission from: Sherry DD. Pain syndromes. In: Isenberg DA, Miller JJ II, editors. Adolescent rheumatology. London: Martin Dunitz; 1999[1].

It is not common for the pain to change location as the exercise therapy progresses. Most children have quite sore muscles during the second and third day but once they work through this pain they start to improve. Our average duration of therapy is 2 weeks. Some children develop an emotional dependency on the team and may, just as they are getting ready to graduate to a home program, have a relapse. Seeing them back for a couple of days a week for exercise therapy for a week or two usually sufficiently weans them to a home program and gets them back into school. The home program takes 40 to 60 minutes to complete and is in addition to usual childhood activities. This program is a continuation of the exercises, stretches, and desensitization done

Table 2. Typical exercise therapy for childhood amplified pain.

Morning:

1 hour occupational therapy for timed activities, functional activities to simulate normal chores such as wall washing, sweeping, carrying, upper extremity strengthening (especially in those with upper extremity pain) and weight bearing on the painful body part. Desensitization of the regions of allodynia are treated with contrast baths, towel rubs (from 30 to 120 s), and lotion massage (children eventually do the desensitization themselves).

1 hour physical therapy for endurance and strengthening exercises including stationary bicycling, treadmill walking and jogging, mini-trampoline jumping, and rope skipping. Physical education and sport-specific activities are simulated.

1 hour water aerobics and weight bearing activities in therapy pool.

Afternoon:

1 hour occupational therapy as in the morning with reevaluation of the child's functional mobility and timed activities. Goals advanced as appropriate.

1 hour physical therapy as in the morning with reevaluation of the child's functional mobility and timed activities. Goals advanced as appropriate.

1 hour family swim (not therapy supervised) for play and relaxation in the therapy pool.

Evening:

An evening home program consists of 30 to 40 min of activities to be done independently and compliance checked the next day by the therapist. If the patient experiences increased pain during the night, they are to repeat this program. Recreational activities are encouraged.

Schoolwork is generally put on hold and make-up work done once the child returns to school.

during the daily program and is generally those activities that either need further improvement or are the more challenging. Gradually, over a month or two, the home program is entirely replaced by normal activities.

Once they understand that the pain hurts but is not associated with damage, some children can be treated exclusively with a home program. However, it hurts too much for most to exercise on their own. Inpatient therapy is indicated if outpatient therapy fails, or the child has uncontrollable pain at night after exercising, or if the amount of dysfunction is too great.

Legions of other therapies have been used, most with much less success and a relatively high rate of relapse[1,9].

It is my experience that psychological factors play a role in most, but not all, of these children, either as a cause or consequence of the pain and dysfunction. Therefore, an initial psychological evaluation including school performance is our standard practice. If significant findings are discovered an appropriate referral is made. This can include family therapy, marriage therapy, individual counseling, help at school, or a change in academic expectations.

A cohesive team of committed, nonjudgmental occupational and physical therapists along with psychologists, social workers, and schoolteacher is indispensable. Especially manipulative children try to play one team member off another, necessitating good team communication.

The majority of children regain full function in one to three weeks; the pain gradually diminishes over a month or so. Occasionally the pain abruptly stops. About 80% of children regain full function and are pain-free within the first month; 15% are functional but have mild or intermittent pain. They may continue to improve over time. About 5% of children are no better and are referred for psychotherapy. In these children we maintain their level of function by seeing them intermittently for exercise therapy and continuing their home program.

In our experience, about 15% of children will experience a significant relapse. This is usually within the first 6 months after exercise therapy and may be associated with trauma. Relapses are usually successfully treated again, but may bespeak the presence of significant psychological distress. About an equal number of children report minor relapses that they are able to exercise out themselves. Over time, 5 to 10% of children will develop a conversion disorder, an eating disorder, or another manifestation of psychological distress, such as suicide attempt. Our 5 year outcome is excellent; 90% are pain-free and fully functional.

Pain amplification syndromes are frequently encountered in the pediatric rheumatology office and may assume one of several distinct patterns. Once recognized, education, intense exercise treatment, and desensitization can begin. This allows the child to gracefully become well and enables her, if necessary, to self-treat recurrent episodes. Although treatment is arduous for both the child and the treatment team, the outcome is quite gratifying.

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