Diagnosis and treatment of amplified musculoskeletal pain in children

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Amplified musculoskeletal pains in children frequently tax the diagnostic and therapeutic skills of the physician. Additionally, the long duration of symptoms and frustrations at failed diagnoses and treatments distress and perplex the child and family. This report will outline my experience with over 700 children with these conditions including my diagnostic approach, treatment, and outcome (1).

It is important to remember that pain is a subjective experience; the degree to which it is felt and to which it affects the child's life (activities of daily living, going to the physician, school attendance, sports participation, and play) is individual. The report of pain is almost always to 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.

Children with amplified pain can present with a variety of manifestations. The established nomenclature artificially divides these conditions into separate groups, whereas it is my experience that children with amplified musculoskeletal pain are on a continuum in which specific syndromes may overlap (Fig. 1) (3-6). Nevertheless, various manifestations include those with overt autonomic signs, those without autonomic signs (with either continuous or intermittent pain), those with painful points (at sites associated with fibromyalgia in adults), and those who are hypervigilant. An individual child's presentation may be quite distinct, but there are many children in whom these syndromes either co-exist (such as a foot with autonomic signs and a hand without) or occur sequentially during relapses. There are, however, consistent features to the presentation, physical examination, treatment, and, perhaps, etiology.

The etiology of amplified musculoskeletal pain in children seems to be causally related to injury, illness, or psychological distress although age, hormonal, and genetic factors may also play a role (5, 7, 8). Psychological distress is usually significant, either as a cause or effect in most children, although good controlled studies are lacking. Commonly there is a history of minor trauma unlike most adults who have preceding major trauma (which is rare in children).

The clinical presentation of children with amplified musculoskeletal pain is remarkably consistent and outlined in Table I (1, 9). Most are female (80%) and tend to be of higher socioeconomic status. The mean age of onset is 12 to 13 years and it is unusual under the age of 8 years.

As the physician obtains the medical history and examines the patient a characteristic pattern is seen. Initially, most

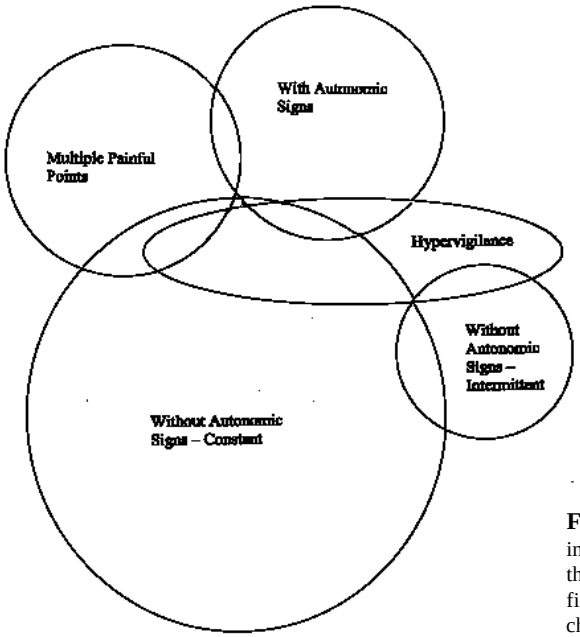

Fig. 1. Diagram showing the overlapping of the various pain amplification syndromes in children.

Table I. Clues to amplified pain from the history and physical examination.

Pre- to adolescent girl (80%)
Increasing pain after minor trauma
Marked disability
Crawls around house or up stairs
Unable to bear light touch, clothing, or bedcovers
May have symptoms of autonomic changes
Cold
Color changes: purple, blue, or gray
Clammy
Edema
Worse or no better with splint or cast
Failed prior therapy
High level athlete
Typical personality
Mature
Excels at school and extra-curricular activities
Perfectionistic
Pleaser
Role model for chronic pain or a similar pain
Recent major life event
Moving
Divorce
Change in nuclear family
Mother acts as spokesperson
Child is mature beyond years
Incongruent affect for amount of pain reported
La belle indifference towards disability & pain
Compliant regardless of reported disability & pain
Autonomic signs, especially after use
Allodynia with a variable border
Pain is not along a dermatome
Pain is not in the distribution of a peripheral nerve
Otherwise normal neurological examination

think the pain started as the result of minor trauma that they may not even recall ("maybe someone stepped on my foot"). Many have a history of autonomic signs, which may be transient (2). Allodynia (pain generated by normally non-painful stimuli) by history is manifest by the inability to clothe or bathe the painful area. It 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 the mother, but can be the father). The parent speaks for the child even when the child is directly addressed. The child is usually cheerful, even when reporting severe pain (up to 10 out of 10 pain) and has a *la belle indifference* about both the pain and dysfunction it causes. On the whole, these children are overly mature and accomplished in school and in extra-curricular activities and are described by their parents as perfectionistic, empathetic, and pleasers (8, 9).

A few variations in the examination are observed between the different patterns of amplified pain. Those with overt autonomic dysfunction 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. Those 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). Those with painful points described in fibromyalgia are more likely to be depressed and fatigued (10). Those who are hypervigilant pay too much attention to regular body sensations and interpret these sensations as painful. The pain can be fleeting, lasting only a second or two, or last several hours such as muscle soreness. They are quite anxious, fearing that these pains are indicative of a serious disease. Children with any form of amplified pain may also be hypervigilant.

The review of systems not uncommonly uncovers conversion symptoms (5). The most frequent symptom is numbness but these children can also manifest paralysis, a histrionic gait, or changes in organs of sense, such as blindness. Family history may reveal a role model for either chronic pain or disability. Social history frequently reveals recent major life events such as moving, changes in the nuclear family, family illness and deaths, or school stress.

Salient points of the physical examination include the absence of an underlying disease (although I have seen pain amplification in addition to organic diseases such as arthritis, enthesitis, infection, and cerebral palsy), a normal neurological examination (paying special attention to sensory tests), and ascertaining allodynia. Allodynia is present if pain is reported when lightly touching the skin or gently pinching a fold of skin. There can be a marked difference in the border of allodynia on repeat testing. Most children will be strikingly compliant; those who are non-ambulatory due to foot pain will walk repeatedly when requested to do so, surprising both the child and parent. It is important to check for signs of autonomic dysfunction, especially after exercising the limb. Painful points as seen in fibromyalgia should be tested for (11). Just how widespread the pain is, is ascertained by digital pressure over control points such as the forehead, shin, and thumbnail (even the hair in some patients is painful). It is not unusual for multiple sites or even the entire body to be involved.

Throughout the examination, even when the child reports severe pain, she usually has a markedly incongruent affect, smiling while she walks or does push-ups. 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, but you need to see just how much she can do.

Blood tests are always normal unless there is an underlying condition in addition to the amplified pain. Radiographs may show some osteoporosis depending on the duration of disuse. Technetium bone scans usually are normal or show decreased blood flow on the blood pool phase and decrease uptake on the delayed image (8). Rarely does a child have the spotty increased uptake pattern typical of adult disease.

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

I have developed a working model to explain the mechanism of amplified pain to the child (Fig. 2). Although there is no consensus regarding the pathophysiology of these conditions, the model is based on sympathetically mediated pain amplification. This model provides a tangible framework to understand the pain, reinforces that the pain is real, and allows discussion of the possible etiologic factors including trauma,illness, and psychological stress.

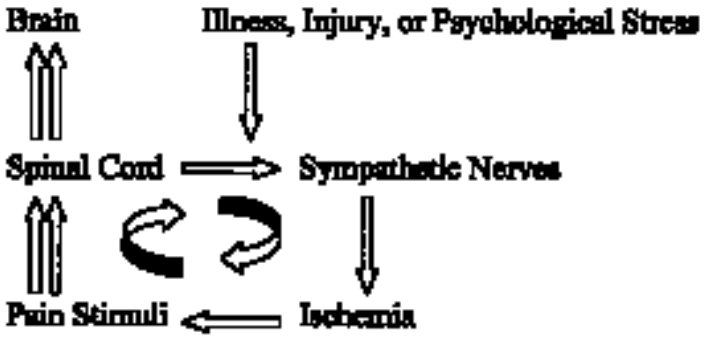

Fig. 2. Schematic of a model to explain the etiology and pathogenesis of pain amplification in children.
Pain signals travel to the spinal cord and up to the brain, where they are interpreted as unpleasant. In those with pain amplification, there is an abnormal short circuit in the spinal cord to the sympathetic nerves that,via vasoconstriction,lead to ischemia. This ischemia causes more pain signal to be generated. Thus, this viscous cycle amplifies the pain.

Additionally, with it we can introduce our therapeutic plan.

To avoid emotionally laden terms, I use the name reflex neurovascular dystrophy for all forms of amplified pain. The information on the internet using the more popular terms such as reflex sympathetic dystrophy or fibromyalgia is rarely applicable to children and can be alarming and misleading.

The treatment is aggressive, one-on-one, functionally focused intense aerobic exercise therapy for up to 6 hours a day, daily (Table II). We discontinue the use of all aids, including crutches, on the first day unless the child is too weak due to disuse. The exercise therapy is one-on-one for two reasons. First, the therapist can encourage full participation and modify the exercises minute by minute to gain maximum function. Second, group therapy can deteriorate into children competing to see who hurts the most. The focus in therapy is on function so if the child could do a task such as a 50-foot run in 12 seconds that morning, she would have to beat 12 seconds in the afternoon even if it meant multiple attempts. A skilled therapist works as a coach and uses the child's innate compliance and competitiveness to motivate and improve function.

In addition to exercises, the allodynia is treated directly with desensitization including towel and lotion rubs, massage and weight bearing. Wearing appropriate clothing and shoes is part of the desensitization process.

As the exercise therapy progresses, it is not uncommon for the pain to change location or for minor setbacks to occur. Most have quite sore muscles during the second and third day but once they work through this pain most will start to improve. The average duration is 2 weeks. Some children develop an emotional dependency on the hospital team and may, just as they are getting ready to graduate to a home program, have a minor 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. The home program usually takes 40 to 60 minutes to complete and is in addition to usual childhood activities. Gradually, over a month or two, the home program is replaced with normal childhood activities.

Initially, home exercise therapy can be successful in some highly motivated children but most children hurt too much to do it on their own. I have seen a couple of dozen children cure themselves once they understand that the

Table II. Typical exercise therapy for amplified pain in children.

Morning

1 hour occupational therapy for timed activities, functional activities to simulate normal chores such as wall washing, dishes, sweeping, 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 seconds to 2 minutes), and ice or lotion massage (children are encouraged to do the desensitization themselves).

1 hour physical therapy for endurance and strengthening exercises including stationary bicycling, treadmill jogging, mini-trampoline jumping and skipping rope. Every effort is made to simulate physical education or sport specific activities, especially those that reportedly cause pain.

1 hour water aerobics in therapy pool.

Afternoon

1 hour occupational therapy as in the morning with re-evaluation of the child's functional mobility and timed activities with advancement of the goals as appropriate.

1 hour physical therapy as in the morning with re-evaluation of the child's functional mobility and timed activities with advancement of the goals as appropriate.

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

Evening

An evening home program consisting of 30 to 40 minutes worth of activities is done independently and compliance checked the next day by the therapist. If the patient experiences increase 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.

pain hurts but is not associated with damage. Outpatient therapy is the rule but if it fails, or the amount of dysfunction too great, inpatient therapy is indicated.

Multiple other therapies have been used with much less success and a relatively high rate of relapse (1, 12). There is not space to discuss these here; the reader is referred to references 1 and 12.

It is my experience that psychological factors play a role in most children, either as a cause or consequence of the pain and dysfunction. Therefore, an initial psychological evaluation of the family and individual child including school performance is done. If significant findings are uncovered, appropriate referral is made. This can include family therapy, marriage therapy, individual counseling, or a change in academic expectations.

A cohesive team of committed nonjudgmental occupational and physical therapists along with psychologists, social workers, and school personnel working together facilitates the rapidity and completeness of recovery. Good intra-team communication aborts the attempts of especially manipulative children to play one team member off another.

Most children regain full function in 1 to 3 weeks and the pain gradually diminishes over a month or so. Occasionally the pain abruptly stops. Approximately 80% are fully functional and pain free in the first month and 15% are functional but have mild or intermittent pains (13). These may diminish further over time. About 5% are no better and are referred for psychotherapy. In these children we maintain their level of functioning by seeing them intermittently for exercise therapy and continuing their home program.

About 15% of children will experience a significant relapse, usually within the first 6 months after exercise therapy. Most relapses are successfully treated again but may bespeak the presence of psychological distress. There are a few additional children who report minor relapses that they are able to exercise out themselves. Relapses may occur in conjunction with injury or illness so it is important that recurrent amplified pain is recognized early so exercises can begin before the child develops significant disability or unnecessary medical investigations. Five to ten percent will develop a conversion disorder, an eating disorder, or another manifestation of psychological distress such as suicide attempt. Our 5-year outcome is excellent; 88% are pain free and fully functional (13).

Pain amplification syndromes are not uncommon in childhood and may assume one of several distinct patterns. Once recognized, intense exercise treatment and desensitization can begin. This allows for the child to gracefully become well and enables her to self-treat recurrent episodes if necessary. Although the treatment is arduous for both the child and treatment team, the outcome is quite gratifying.

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