



The idiopathic musculoskeletal pain syndromes in childhood

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Pain is a ubiquitous experience and musculoskeletal pain in children is common [1]. Acute musculoskeletal pain is frequent and the cause of the pain is usually obvious. Chronic pain often proves to be a difficult diagnostic and therapeutic challenge and, by definition, there are no obvious or overt noxious stimuli in children with idiopathic musculoskeletal pain to explain the pain. Because pain is subjective [2], the pain exists, it is real to the children, and it can cause much suffering and disability to the individual and the family, often resulting in multiple visits to many different health care providers in search of diagnosis and treatment. Thus the role of the rheumatologist is a critical one, initially ruling out the multitude of mechanical, inflammatory, neoplastic, and genetic conditions that can cause chronic pain, then—once a diagnosis of idiopathic pain is established—initiating treatment, including working with the child and family to understand and cope effectively with pain.

Pain pathophysiology

Acute pain in response to a noxious stimulus arises from nociceptors comprised of either the thinly myelinated A δ fibers or by slow-conducting, non-myelinated C fibers [3]. The former are responsible for well localized, immediate pain and the later more poorly localized, burning pain. These transmit their signal by way of neuropeptides including substance P and calcitonin gene-related peptide within several spinal segments in the spinal cord. Here the signals ascend

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by way of the spinothalamic, spinoreticular, and spinomesencephalic tracts that terminate in the thalamus, reticular formation, and periaqueductal gray area of the midbrain. The sensory cortex then integrates the sensory message with ongoing cognitive functions and organizes the conscious behavior manifest in response to the stimulus.

At all anatomic levels there exist modulating mechanisms that can help limit the pain experience, or, when gone awry, might amplify this experience. The pain most children experience resolves relatively quickly and does not develop into a chronic pain syndrome. There are a number of children who do develop chronic pain that is unremitting over long periods, however. The exact mechanisms leading to and sustaining this pain are not known. It seems certain that the pain is not due to ongoing activity of nociceptors in the periphery but is due to dysfunction of the more central nociception system (spinal cord and cerebral cortex). In many cases this dysfunction involves the sympathetic system [4,5].

Idiopathic musculoskeletal pain in children

Chronic musculoskeletal pain syndromes seen by rheumatologists often have a variety of manifestations. The terms used are multiple and confusing [6]. For the purpose of this article the authors will use the terms proposed by Malleon et al, since they are descriptive and underscore the fact that the etiology is unknown (Table 1) [7]. The authors acknowledge that there are several subsets within both those with diffuse and localized idiopathic pain subsets. For example, fibromyalgia would fall into the diffuse group and complex regional pain syndrome type I (or reflex sympathetic dystrophy) into the localized group. Additionally, there are those with intermittent pain that do not fulfill the duration criterion, but should be included because of the recurrent nature of their pain over months. Once a diagnosis of idiopathic musculoskeletal pain is made, evaluation is the same and treatment is similar regardless of the terminology used and the particular subset of pain the child is experiencing [8].

Table 1

The definition of the idiopathic musculoskeletal pain syndromes in children

Diffuse idiopathic musculoskeletal pain

These children have both:

1. Generalized musculoskeletal aching at three or more sites for at least 3 months
2. Exclusion of other diseases that could reasonably explain the symptoms

Localized idiopathic musculoskeletal pain

These children have all three:

1. Pain localized to one limb persisting
 - a. 1 week with medically-directed treatment or
 - b. 1 month without medically-directed treatment
 2. Absence of prior trauma that could reasonably explain the symptoms
 3. Exclusion of other diseases that could reasonably explain the symptoms
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Epidemiology

Although musculoskeletal pain in children is common, there are no specific incidence and prevalence data for the idiopathic musculoskeletal pain syndromes. Between 5% and 8% of children presenting to North American pediatric rheumatology centers have idiopathic musculoskeletal pain, however, and it is the subjective opinion of many pediatric rheumatologists that the number of children with these syndromes is rising [9,10].

The average age of onset is 12 years, and the authors are particularly cautious about making a diagnosis in patients aged <7 years, although the authors have seen children as young as 3 years old who have chronic idiopathic musculoskeletal pain [11,12]. Girls outnumber boys by four to one, and the vast majority reported patients are Caucasian.

Etiology

As the name implies, the etiology is unknown. There are important differences between children and adults with these pain syndromes. For example, children with complex regional pain syndrome type I have more frequent lower extremity involvement, lack of significant prior trauma, lack the initial phase of redness, edema and warmth, have decreased uptake on bone scintigraphy, are more frequently have significant psychologic distress, and have a good outcome with exercise therapy alone than adults [12–15].

Generally, significant trauma such as fracture is not a frequent feature of any idiopathic musculoskeletal pain syndromes in children [13,16]. Psychologic distress is much more of an overt factor in children and their families compared to adults [13,17–21]. Hormonal and genetic factors might also play a role because the ailment is much more common in girls than boys; it is also occasionally seen in siblings [22,23]. Other factors hypothesized to be important in the pathophysiology of idiopathic musculoskeletal pain (both in children and adults) include increased sympathetic nervous system activity, increased α -adrenoceptor responsiveness, disturbed sleep patterns, abnormal serotonin metabolism, and hypothalamic–pituitary–adrenal axis dysfunction [5,24]. It is probable that the cause of idiopathic musculoskeletal pain is multifactorial, with both “intrinsic” factors (low pain thresholds, female sex, hypermobility, intrinsic coping strategies) and “extrinsic” factors (previous pain experiences, social deprivation, physical or sexual abuse, parental modeling of chronic pain behaviors, sleep disturbance, decreased fitness) all potentially contributing to the pain [25]. The relative importance of these factors differs between individuals, but different factors can also change in their relative importance as the pain persists. For example, once the pain starts it can lead to disturbed sleep, which itself exacerbates the pain; the disruption of the pain can affect family functioning, particularly if the family is already stressed. Whether a child with idiopathic musculoskeletal pain develops inappropriate pain behaviors, pain-associated

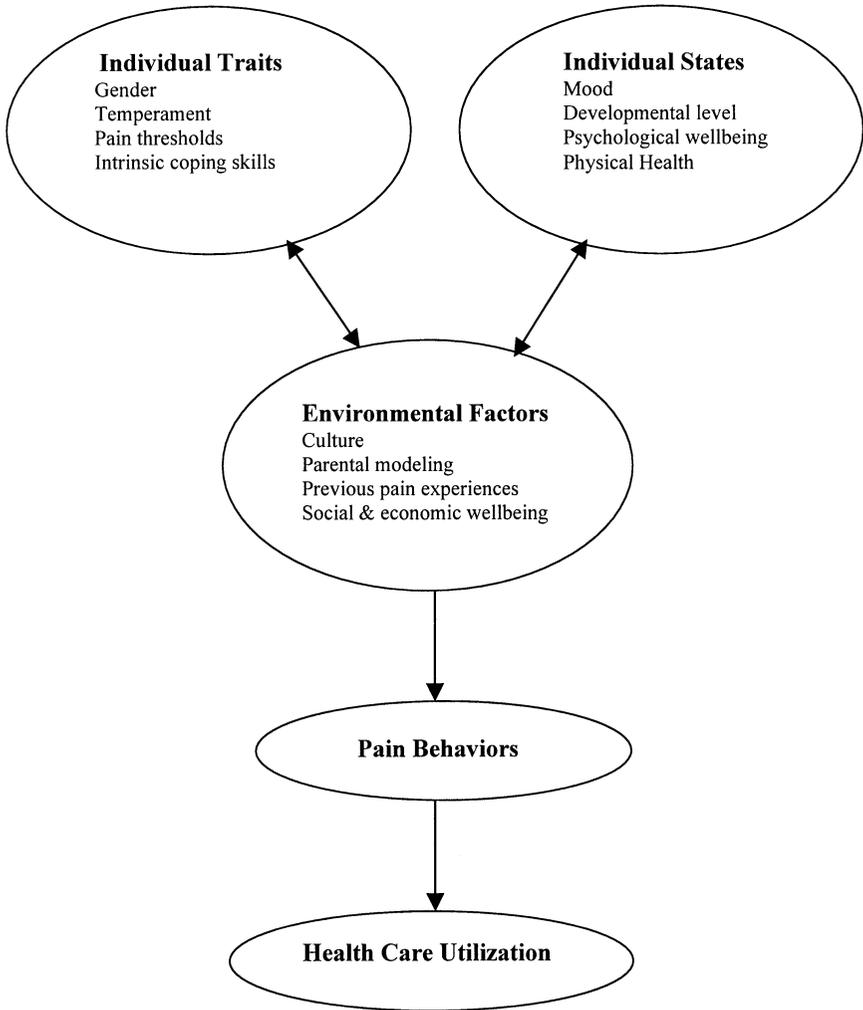


Fig. 1. Factors influencing pain behaviors and health care utilization.

disability, and seeks health care interventions are dependent on a number of interacting factors (Fig. 1).

Clinical findings

Clinical manifestations vary between those with diffuse versus localized idiopathic musculoskeletal pain, although there are many aspects of the history and physical examination that are very similar. Features that are common include age of onset in early adolescence, female sex, prolonged history of pain that is

unresponsive to therapy, increasing pain over time that generally is rated extremely high (a 9 or 10 out of 10), allodynia (pain generated by a nonpainful stimuli such as light touch), marked functional disability (including missing an inordinate amount of school), and conversion symptoms (such as paralysis) [7,11,12].

Features that differ include the frequent occurrence of autonomic symptoms and signs such as coolness, cyanosis, or increased perspiration in children who have the localized form of idiopathic musculoskeletal pain (complex regional pain syndrome type I) but not the diffuse form. In the localized variant the pain is usually peripheral, affecting a single limb. Occasionally a child will have a localized painful area on the chest or back, whereas children with diffuse idiopathic musculoskeletal pain might have total body pain or poorly localized aching with or without the tender points whose presence is deemed necessary for the formal diagnosis of juvenile fibromyalgia (a term that is eschewed by the authors and many pediatric rheumatologists) [7,8,26,27]. Children who have diffuse pain more frequently report nonrestorative sleep and depressive symptoms than do children with localized idiopathic musculoskeletal pain [8,27].

Physical examination also reveals many similarities between children with localized and diffuse idiopathic musculoskeletal pain. One of the more striking features is the incongruent affect most children manifest when reporting severe pain [8]. The vast majority of these children appear happy, calm, and even smile when experiencing 10 out of 10 pain. Additionally, they have a blithe unconcern about the disability they have, the so-called *la belle indifférence*. Most children will have allodynia. Allodynia is tested for by lightly touching the skin or gently pinching a fold of skin. The border of the allodynia frequently varies over time.

The most notable difference seen between the various idiopathic musculoskeletal pain syndromes during the physical examination is that of the autonomic signs in a subset of children who have localized idiopathic musculoskeletal pain [2]. Coolness of the limb is the most frequent finding, followed by cyanosis then increased perspiration. These signs might not be initially present but become manifest after the child has used the involved limb during the examination process. The development of dystrophic skin and subcutaneous changes is much less common in children compared to adults.

Children who have diffuse idiopathic musculoskeletal pain might or might not have the typical painful trigger points seen in adults with fibromyalgia [26]. Some children have no or only a few tender points, but frequently there is widespread tenderness with the child reporting pain when the examiner applies digital pressure over most any part of the body, including control points such as the clavicle and forehead. These children might be labeled as having fibromyalgia as they fulfill the criteria of widespread pain with the presence of 18 of 18 painful fibromyalgic points. It is not clear that this label is particularly helpful, however, because there is no good evidence that such children are different in any way from those who have diffuse pain without the requisite number of tender points.

Chronic back pain in childhood frequently portends a serious illness and requires careful investigation; however, in adolescence back pain is frequently

not due to serious pathology [8,11,28]. Most of these patients have other signs of nonorganic back pain such as axial loading, distracted straight leg raising, passive rotation, overreaction, or allodynia [29]. The axial loading test is positive when back pain is reported while the examiner exerts downward pressure on the top of a standing patient's head; neck pain is not a positive test by itself. A positive distracted straight leg-raising test is present when the patient reports back pain when the hip is flexed while the patient is supine, but not sitting. The passive rotation test is positive when the patient reports back pain when passively rotated at the ankles and knees keeping the pelvis, back, and shoulders in the same plane. Overreaction is defined as excessive wincing, shaking, screaming, or collapsing with pain. The definition of excessive is subjective and varies based on age, mental status, fear, and cultural background.

The psychologic and psychosocial aspects obtained by both the history and observation during the examination are noteworthy. A common finding is a mother whose emotional life is overly wrapped up and dependent on the daughter, the so-called enmeshed mother [30]. The mother will speak for the child even when directly inquiring the daughter about her feelings and thoughts. In addition, these children are frequently highly motivated and driven to achieve. The onset of pain sometimes seems to be associated with increasing demands of school in a child who has overachieved by dint of hard work, but who actually has unrecognized learning difficulties. Such children not only generally do very well in school, but they are also involved in multiple extracurricular activities such as music, dance, sports, and volunteer work. Their parents describe them as perfectionistic and pleasers, and usually worry more than the parents think they should. They are sometimes concerned about others to the point of sacrificing their own emotional needs to meet others needs and expectations. Rarely (about 10% of the time) symptoms and signs of depression will be present [30]. These findings, along with the incongruent affect, do not necessarily indicate major psychopathology but often lead others to accuse the child of making up the pain or dismiss the pain as purely psychologic. It should be stressed that apparently enmeshed families and perfectionist children can and do have organic disease, so the presence of these traits do not, by themselves, make a diagnosis of idiopathic musculoskeletal pain. Similarly, the presence of fibromyalgia tender points does not exclude the presence of organic disease.

A common finding is the increased distress manifested both by the child and the parent when the physician imparts the good news that no serious illness appears to be present. This, and the refusal to accept that psychosocial factors can play any kind of role in the degree of pain or extent of the functional disability, is often striking.

Laboratory findings

Overall, the authors attempt to limit performing a large number of investigations in children who are suspected on clinical grounds as having idiopathic

musculoskeletal pain. Most children have had numerous tests, procedures, and therapeutic trials—all to no avail—by the time they are referred to the rheumatologist. Performing further tests suggest to the child and parents that the diagnosis is in question, and tends to delay acceptance of the diagnosis and the institution of appropriate treatment. Having said that, however, the diagnosis must be absolutely clear before treatment can commence. The authors have, for example, seen idiopathic musculoskeletal pain as a consequence of another illness causing pain, such as arthritis.

If there is a suspicion of either an underlying cause (see differential diagnosis below) or a concurrent illness, a few basic and focused blood tests are usually all that is needed. Laboratory studies are normal for the vast majority of children with idiopathic musculoskeletal pain. A few children with idiopathic musculoskeletal pain will have a positive antinuclear antibody (ANA) test. These are generally of no diagnostic significance; a significant minority of healthy children will have a positive ANA test if it is performed. Unless there are symptoms or signs suggestive of an underlying disease, no further investigation is indicated [31].

Plain radiographic studies are normal except when disuse osteoporosis is present. In children who have localized idiopathic musculoskeletal pain it is rare to see the spotty osteoporosis that is described in adults [32]. Technetium radionuclide scintigraphy is the study most likely to be abnormal, and children usually have decreased uptake rather than the spotty increased uptake seen in adults with complex regional pain syndrome type I [14,15]. The bone scan might show increased uptake or be normal, however, and these changes do not seem to be related to whether or not there is any clinical evidence of autonomic dysfunction [11]. The main value of a bone scan is to exclude a focal abnormality such as a stress fracture or osteoid osteoma that is mimicking or even precipitating idiopathic musculoskeletal pain. Magnetic resonance images are normal or will show soft tissue edema children who have swelling as part of their idiopathic musculoskeletal pain.

Differential diagnosis

Although there are many positive historical and physical features to idiopathic musculoskeletal pain syndromes, other causes must be excluded. Different possible diagnoses that must be considered are briefly considered below (Table 2).

Spondyloarthropathy

The majority of children referred as having a possible idiopathic musculoskeletal pain syndrome who have another diagnosis will probably have a spondyloarthropathy. Enthesitis is frequently not specifically looked for, nor the symptoms of spondyloarthropathy appreciated. Children who have back pain as a manifestation of spondyloarthropathy are also frequently thought to have

Table 2

Differential diagnosis in children with idiopathic musculoskeletal pain

Spondyloarthropathy
Hypermobility
Leukemia
Spinal cord tumors
Myofascial pain
Chronic recurrent multifocal osteomyelitis
Primary Raynaud phenomena
Fabry disease
Erythromelalgia
Pernio
Chronic compartment syndrome
Progressive diaphyseal dysplasia
Peripheral mononeuropathy
Idiopathic juvenile osteoporosis

idiopathic musculoskeletal pain. This is due in part to the failure of ibuprofen, acetaminophen, or other over-the-counter medications to help. Children with spondyloarthropathies frequently respond to proper anti-inflammatory doses of nonsteroidal anti-inflammatory medications and are helped by regular doses of analgesics, whereas these medications are almost always of minimal benefit in children who have idiopathic musculoskeletal pain. Careful attention to subtle symptoms of inflammation, the finer points of the musculoskeletal examination, and judicious therapeutic trials will help establish this clinical diagnosis.

Hypermobility

Hypermobility is a common cause of musculoskeletal pains in children. There is some evidence that the frequency of hypermobility is increased in children with diffuse idiopathic musculoskeletal pain, perhaps acting as a recurrent trigger for musculoskeletal pain in the predisposed individual [33]. Most of these children are younger—generally preschool to elementary school age. The pain is often most severe towards the end of the day and the parents have commonly associated the pain with specific activities such as swinging on monkey bars or gymnastics. Although the pains usually affect the legs, hand and wrist pain can occur in children with hypermobile finger joints who are doing extensive writing at school. Nocturnal pain can occur, but unlike the pain associated with arthritis or malignancy it is not present the next morning and is not associated with complaints of stiffness or bony tenderness. Massage of the painful area usually helps the pain. The diagnosis is established by the typical history and evidence of hypermobility on examination [34].

Leukemia

Leukemia can present with bone pain and marked dysfunction due to the amount of pain. Usually these children look sick. They are wan, anorexic, lethar-

gic, frequently febrile, and can have episodic and migratory arthralgia or even overt synovitis. Nighttime pain is common on examination, as is bone pain. Blood counts are usually abnormal, as is the erythrocyte sedimentation rate.

Spinal cord tumors

Spinal cord tumors might grow slowly and cause pain and discomfort for months to years. The quality of the pain is different; it is often described as being of low, steady intensity rather than the maximal pain described by children with idiopathic musculoskeletal pain. Allodynia and an incongruent affect are unusual. An abnormal neurologic examination is the key to making the correct diagnosis. Most tumors are apparent on magnetic resonance imaging.

Myofascial pain

Myofascial pain can present as an idiopathic musculoskeletal pain, but it is much more limited because it arises from sustained contraction of a muscle, especially in the head, jaw, and upper back. The diagnosis can usually be confirmed by reproduction of the pain when the muscle is palpated.

Chronic recurrent multifocal osteomyelitis

Chronic recurrent multifocal osteomyelitis is a fairly common condition seen in both younger children and adolescents due to chronic, noninfectious inflammation in the metaphyses close to the physes of multiple bones. The tibia is most commonly affected, although almost any bone can be involved. There is bony tenderness over the affected sites. Lytic lesions (Brodie's abscesses) can be seen on plain radiographs. If the diagnosis is suspected, bone scintigraphy is indicated to locate other subtle lesions. The pain usually responds to nonsteroidal anti-inflammatory medications or, if necessary, to corticosteroids.

Primary Raynaud phenomena

Primary Raynaud phenomena can occur in childhood. Pain, numbness, or tingling can be experienced with the episode. The episodes are usually short-lived and can be reproduced with a cold challenge. Vibration or emotional upset stimulates an attack in children only rarely. The coldness and color change can be mistaken for localized idiopathic musculoskeletal pain (complex regional pain syndrome type I).

Fabry disease

Fabry disease is an X-linked recessive condition that causes a deficiency of ceramide trihexoside α -galactosidase. Clinically, it usually leads to episodic, excruciating burning pain in the hands and feet. Symptoms usually begin in adolescence. The diagnosis is suggested by the presence of bluish maculo-

papular hyperkeratotic lesions about the perineum and an elevated erythrocyte sedimentation rate.

Erythromelalgia

Erythromelalgia is a painful condition that causes intense pain, redness, and swelling of the hands and feet. It can be familial or indicative of an underlying myelodysplasia (the latter is usually response to aspirin while the former is not). Affected children almost uniformly want to apply cold to affected extremities and will often refuse to remove their feet from ice water.

Pernio

Pernio, or chilblains, causes a burning pain associated with violaceous papules, usually on the fingers and toes. It is a consequence of prior cold injury, usually prolonged exposure to wet cold such as wet socks while playing in the snow followed by rapid rewarming. It is episodic and recurs with cold exposure.

Chronic compartment syndrome

Compartment syndromes arise from increased pressure within the closed muscle compartment that is caused by exercising. The increased pressure leads to ischemia and pain. Classically, it occurs in athletes, who develop symptoms in the calf while running. The symptoms begin after a critical amount of exercise has occurred, for example, after running 4 miles, and will recur each time the critical level for that individual has been achieved.

Progressive diaphyseal dysplasia

Progressive diaphyseal dysplasia begins in adolescence and is marked by severe leg pain, fatigue, headaches, weight loss, weakness, and an abnormal, waddling gait. Plain radiographs that show cortical thickening and sclerosis of the diaphysis of the long bones confirm the diagnosis.

Peripheral mononeuropathy

Peripheral mononeuropathy can be associated with severe burning pain in the distribution of the involved peripheral nerves. A mononeuropathy can occur following an injury or an infection. A particular variant is acute neuralgic amyotrophy of the shoulder, a neuropathy of the brachial plexus that leads to pain and muscle wasting of the shoulder. Sensory neuropathies can be associated with the same kind of burning pain and heat described with erythromelalgia.

Idiopathic juvenile osteoporosis

Idiopathic juvenile osteoporosis is a relatively unusual condition seen in adolescents who present commonly with lower limb pain due to pathologic

micro-stress fractures secondary to decreased bone density. Plain radiographs reveal severe osteoporosis; a bone scan might be necessary to demonstrate the microfracture.

Thyroid disease

Both hypothyroidism and hyperthyroidism have been associated with widespread musculoskeletal pains. Associated symptoms and signs of thyroid dysfunction are usually clinically manifest.

Vitamin D deficiency

Rickets due to nutritional vitamin D deficiency is a rare cause of limb pain in the developed world, but should be considered in dark-skinned adolescents who live in Northern climates and are undergoing a rapid growth spurt. Genetic forms of rickets usually present with the early development of bony deformities, with or without limb pain, so they are not commonly confused with idiopathic musculoskeletal pain.

General assessment of disease activity

There are two major variables to be considered in assessing the degree of illness in children with idiopathic musculoskeletal pain: pain and dysfunction. These two factors are relatively independent of each other. Children can report extreme pain and be fully functional or have only minor degrees of pain complaints but be bedridden and even unable to feed themselves.

Pain is subjective; it is best measured by self-report [35–39]. The most common method is with either a verbal or a visual analogue scale [36]. This can be as simple as having the child rate the pain from zero to ten or as complex as picking descriptors from lists and coloring body parts with different colors to represent where and how much each body part hurts.

Functional measures depend on what body part is involved and any coexisting conditions. For example, the authors have seen children with idiopathic musculoskeletal pain who also have a variety of illness such as arthritis, muscular dystrophy, or cerebral palsy. Physical and occupational therapists will usually take into account speed, endurance, and quality of movement in assessing function.

Psychologic assessment is indicated for most of these children and their families because by the time they present to the rheumatologist significant psychologic dysfunction is present. Although it is not necessarily true that these syndromes are psychologic in cause, the psychologic toll that this condition has already wrought on the child and family is severe. In some seemingly intact families there might be marked underlying and unmet psychologic needs, and in other families that seem more dysfunctional there are remarkable strengths. The spectrum of psychopathology in those needing treatment is large, ranging from anxiety and poor coping to major personality disorders and severe depression.

Treatment

The fact that there is no proven therapy is evident in the plethora of widely disparate treatments reported in the literature. Many of these reports describe either a single child or a small group of children with idiopathic musculoskeletal pain; therefore, recommendations based on these reports cannot be made with any confidence. The larger series of treatment focus on a single method. No direct comparisons between methods are made, therefore no therapy recommendation can be authoritative.

The twin goals of treatment are restoring function and relieving pain; less than accomplishing both is not satisfactory. Having stated this, pain is subjective and not amenable to specific treatment; therefore, in some children one has to accept restoration to full function as the only attainable goal. In patients who experience psychologic distress, psychotherapy aimed at the specific abnormal psychologic problem is helpful and can help even those who do not attain full relief from their pain to cope [40,41].

The majority of data on the therapy of idiopathic musculoskeletal pain specifically deal with complex regional pain syndrome type I. Therapies reported to be of benefit, in the order of success, include exercise therapy, transcutaneous electrical nerve stimulation, and sympathetic blocks [11–13,23,42–50]. Other treatments that have been suggested include glucocorticoids, tricyclic antidepressants, anticonvulsants, opioids, sympathectomy, biofeedback, behavioral modification, and other forms of psychotherapy [32,40,50].

In the authors' opinion the most successful treatment is aggressive exercise therapy with or without psychotherapy. Bernstein et al were the first to report on a large number of patients when they treated 23 children who had complex regional pain syndrome type I (called reflex neurovascular dystrophy) with exercise alone [13]. Long-term results on 20 patients revealed 12 patients without pain or dysfunction; five had occasional pain without physical signs, two had moderate discomfort, and one had recurrent complex regional pain syndrome type I. Recently, Sherry et al reported on 103 children with complex regional pain syndrome type I treated with intense exercise therapy and psychotherapy (when indicated) with 92% of patients having full resolution of their pain [12]. Long-term outcome on 49 of these children revealed that after 5 years 88% were free of pain and dysfunction. This exercise therapy consisted of 5 hours per day (weekdays) for an average of 2 weeks. The activities were functionally directed (stairs, running, wall washing) and done one-on-one with physical and occupational therapists, who encouraged both speed and quality of movement. Allodynia was treated by desensitization with towel and lotion rubs and wearing appropriate clothing and shoes. For a more specific discussion of this exercise therapy see Schanberg et al [8].

The largest report using a combination of multiple physical and medical treatments, including sympathetic blocks and sympathectomy, was by Wilder et al. Seventy children with complex regional pain syndrome type I were treated, and 46% resolved their pain [50]. No long-term outcomes were reported.

There are few reports of therapy of localized idiopathic musculoskeletal pain other than complex regional pain syndrome type I. The largest series, that of Sherry, et al, reported on 100 children with musculoskeletal pain, over half of whom were classified as having localized idiopathic musculoskeletal pain [11]. These children were treated with exercise therapy, and most did well (78% of the entire group resolved their pain and 97% become fully functional). They received the same exercise therapy outlined above for children with complex regional pain syndrome type I.

The treatment of diffuse idiopathic musculoskeletal pain in children is similar to that in adults, and is not very satisfactory. Adult studies have advocated a wide range of therapies; however, most long-term studies in adults with fibromyalgia suggest that the best outcomes are associated with education, aerobic exercise, low-dose tricyclic antidepressants, and nonsteroidal anti-inflammatory agents [51]. One study reported cyclobenzaprine helped 11 of 15 children with fibromyalgia; however, in another study of 33 children, only three patients would recommend it to other children with fibromyalgia [52,53]. Psychologic support is also generally recommended [54,55].

If one accepts the fact that idiopathic musculoskeletal pain is multifactorial, it follows that no single treatment is likely to be effective in all patients. An interdisciplinary approach aimed at helping the child and family cope with the pain, reversing inappropriate pain behaviors, and minimizing pain-associated disability seem to offer the best chance of obtaining an optimal outcome. Evidence is accruing that cognitive behavioral therapy of this kind is effective for children with a variety of chronic pains and pain-associated disabilities [56–58].

Course and prognosis

There are no reports of the natural history of children with idiopathic musculoskeletal pain. Some of the children presenting to the rheumatologist have had symptoms for years, and others have episodes of what historically sounds like idiopathic musculoskeletal pain that has resolved, only to recur. One study of 15 schoolchildren found to have criteria for fibromyalgia were examined 30 months later; 11 (73%) were asymptomatic [26]. Another study reported that 92% of children diagnosed in a pediatric rheumatology center with fibromyalgia still were symptomatic with significant pain a mean of 33 months after diagnosis [59].

Children with complex regional pain syndrome type I treated with intense exercise therapy, as noted above, have one of the best outcomes reported [12,13]. Multiple factors might account for this, both physiologic (resetting the sympathetic tone, endorphins) and psychologic (empowering the child to take control of the illness, hope), but all are speculative at best. The long-term outcome is unknown, but at 5 years most patients were still without pain. The long-term outcome of children with complex regional pain syndrome type I treated with

sympathetic blocks and a combination of other medications such as antidepressants, opioids, and anticonvulsants has not been reported.

Overall, children with diffuse pain are more likely to be resistant to treatment and are at a higher risk for relapse. The authors have not found that age, sex, or duration of symptoms before diagnosis predict who will or will not respond to exercise therapy or who is more likely to suffer a recurrence [8]. Children with one form of idiopathic musculoskeletal pain might have a different form upon recurrence. It is likely that children who have recurrent disease are more likely to have significant underlying psychopathology than those who have a single short-lived episode.

Other unfavorable outcomes besides recurrence include the development of other painful conditions such as headache and abdominal pain with or without irritable bowel syndrome. Some children with idiopathic musculoskeletal pain develop other psychologic diseases such as conversion symptoms, eating disorders, or panic attacks, or they attempt suicide [8]. Because controlled studies have not been done it is unknown if these problems occur at a greater frequency than is observed in the general population.

Summary

Idiopathic musculoskeletal pain syndromes in children have a variety of manifestations; they can be diffuse or well localized, constant or intermittent, with or without autonomic symptoms and signs, completely incapacitating or not limiting activities, and they can tax the physician's diagnostic skill. A careful history and examination is usually all that is needed to make a diagnosis, although the differential diagnosis is large and might require laboratory and radiographic investigation. Pain and functional assessment help track the progress with therapy. Intense exercise therapy is associated with the best outcome. Psychologic issues should be evaluated to determine if further psychologic intervention is indicated. The medium-term outcome is probably good for most of these children, but the long-term prognosis is unknown. One must be aware that other manifestations of psychologic problems might emerge. By the time these children and their families see the rheumatologist they are desperate and can be frustrating to work with due to their difficulty in accepting any kind of psychologic element to the pain and its associated disability. Nevertheless, it is rewarding to help the children understand and work through their pain so they can resume normal lives.

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