Recent developments in the understanding of paediatric musculoskeletal pain syndromes

Chronic idiopathic musculoskeletal pain syndromes in children are regarded as very rare by most rheumatologists and paediatricians. This is somewhat surprising as similar syndromes are among the most commonly diagnosed conditions in adult rheumatology clinics. In addition, over 15% of apparently normal school children report musculoskeletal pain in health questionnaires. The proportion of children in whom the pain becomes chronic (in the absence of a known cause) is unclear, but it may be much greater than previously realised. Of the 2959 children entered into the British Paediatric Rheumatology Group’s diagnostic index to date, about 10% had a chronic idiopathic musculoskeletal pain syndrome (Symmons D P M, personal communication). It has been suggested that failure to recognise the characteristic clinical features of these conditions may contribute to their chronicity. Recently, there have been several advances in the understanding of chronic idiopathic musculoskeletal pain syndromes in children, which may clarify some of the issues surrounding their diagnosis, epidemiology, and treatment.

Pain in the absence of an organic cause is a difficult symptom to label. None of the traditional names, including hysterical disorder, pain amplification, psychogenic disease, abnormal illness behaviour, and supratentorial dysfunction, are universally appropriate. They appear judgmental and even perjorative, implying perhaps that the problems are psychological, when in fact their cause is unknown. In addition, terms coined in reports of adults, such as reflex sympathetic dystrophy (RSD) and fibromyalgia, are not entirely satisfactory as their diagnosis requires the presence of several clinical features which when present may be subtle, variable, intermittent, and even absent. Can RSD be diagnosed if the child’s painful limb looks and feels normal on examination, or fibromyalgia be considered if no hyperalgesic trigger points are found? The answer is probably no, but recently Malleson and colleagues proposed that these specific conditions are only the more severe manifestations of a broader spectrum of disease in children. They put forward criteria for the diagnosis of localised and diffuse idiopathic pain syndromes in children (table 1), and also noted that both disorders share a number of clinical features, raising the possibility that they have a similar pathogenesis.

### Table 1 Criteria for the diagnosis of idiopathic musculoskeletal pain syndromes in children. (Modified from ref 4.)

<table>
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<tr>
<th>Localised idiopathic pain syndrome</th>
<th>Diffuse idiopathic pain syndrome</th>
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<tr>
<td>1 Pain localised to all or part of one limb persisting for more than one week despite medically directed treatment, or more than one month without treatment.</td>
<td>1 Diffuse/widespread pain affecting at least three areas of the body, persisting for more than three months.</td>
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<td>2 (for both localised and diffuse syndromes) Absence of other aetiologic factors that might reasonably explain the finding (for example, trauma, tumour, sepsis, chronic inflammation, osteochondritis, metabolic or dietary deficiency, etc)</td>
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**Reflex sympathetic dystrophy** Both of the above, and all of:

1 Localised tenderness
2 Altered skin colour or temperature, or both
3 Soft tissue swelling or atrophy

**Fibromyalgia** (adapted from Yunus and Masi) Both of the above, and:

1 At least five tender points: areas of exaggerated tenderness elicited on moderately firm fingertip palpation of pairs of specific anatomic sites: (a) upper border of trapezius (b) lower part of sternocleidomastoid (c) lateral part of pectoralis major (d) mid-supraspinatus (e) lateral aspect of elbow (f) upper outer quadrant gluteal region (g) greater trochanteric region (h) medial fat pad of knee
2 At least three additional features: (a) chronic anxiety or tension (b) fatigue (c) poor sleep (d) chronic headaches (e) irritable bowel syndrome (f) subjective soft tissue swelling (g) numbness Modulation of pain: (b) by physical activities (b) by weather factors (b) by stress/anxiety

Four tender points also satisfy the criteria provided that a patient has four of the above 10 features.

**Localised idiopathic pain syndromes**

The presenting symptom of a localised idiopathic musculoskeletal pain syndrome is usually inability to use the affected limb because of pain; the leg(s) being more commonly affected than the arm(s). Many descriptors are assigned to the accompanying pain, ranging from dull, burning and aching to sharp, shooting, and stabbing. Most paediatric patients are girls during their early adolescent years, but boys can also be affected, and patients as young as 7 years have been reported. There may be a past history of other chronic pain such as headaches or abdominal pain, and in many cases an illness or event which precedes the onset of limb pain is identified. When an affected limb is examined the patient will usually exhibit a characteristic response: the ‘grimace and gasp’ or ‘wince and withdrawal’. Complaints of severe pain to light touch are common and can be termed allodynia (painful response to non-noxious stimulus), or hyperpathia (a condition of excessive suffering). The initial episode can last for months to years, and recurrence occurs in about half of the patients. In Malleson’s study 41 children fulfilled the criteria for the diagnosis of localised idiopathic pain syndrome, of whom 24 had definite RSD. The exact relation between these conditions has yet to be clarified, but it seems likely that untreated or inadequately treated localised idiopathic pain would tend to progress to RSD.

The diagnosis of RSD is based on the presence of features of a localised idiopathic pain syndrome and additional features of dysautonomia. An affected limb will often have a pallid or purple mottled appearance which can be accentuated by placing the limb in a dependent position. Reduced skin temperature, slow capillary filling, and soft tissue swelling are sometimes prominent. ‘Tache cerebrale’, the appearance of an erythematous line 15–30 seconds after the skin of an affected area has been stroked by a blunt object, has been reported. Peripheral pulses and perspiration are variably affected. In its most severe form, the affected limb is held in a bizarre posture, and can become virtually fixed in that position with muscle wasting...
and soft tissue atrophy. Sometimes, despite extraordinary deformity, the patient can appear seemingly indifferent to the problem ('la belle indifférence').

Diffuse idiopathic pain syndromes

The usual presenting complaints in this group of conditions are generalised, symmetrical, musculoskeletal aches and pains, fatigueability, and physical deconditioning. Most children with this problem are significantly incapacitated by the pain, and have sleeping difficulties which may be contributory.

Fibromyalgia is the most easily recognised diffuse pain syndrome, but again it probably represents only the more severe end of a spectrum of disease. Malleson et al described 40 children with diffuse idiopathic pain syndrome, of whom 35 fulfilled diagnostic criteria for fibromyalgia. Yunus and Masi have documented complaints of pain and stiffness at the knees in two thirds of children with fibromyalgia, and other sites less commonly, including ankles, elbows, wrists, cervical spine, trapezius, fingers, thighs, and feet. The back is painful in at least a third of cases. Complaints are found in younger children, but there are no objective joint effusions or soft tissue swelling. Most affected children describe early morning stiffness of several hours' duration; this is an important if confusing feature, as it is usually equated with inflammatory arthropathy. Abnormalities of sleep are almost universal and, in particular, waking in the morning feeling tired (non-restorative sleep) is common.

Physical examination may disclose some evidence of anxiety and occasionally depression. The most characteristic findings are the presence of multiple symmetrical tender points, usually of soft tissues around the upper and lower limb girdles (table 1). The pressures required to elicit soft tissue tenderness in children have recently been defined. The multiplicity of the tender points is an important feature; one or two tender points are not uncommonly found in normal patients. In primary fibromyalgia there is no evidence of arthritis or other definite organic disease, but secondary fibromyalgia has been noted in children with defined organic diseases such as juvenile chronic arthritis.

The cause of the pain in fibromyalgia is unknown, but aberrant central pain mechanisms have been proposed. In this issue Gedalia and colleagues have raised the possibility that peripheral disorders may also be important (pages 494–6). In a study of 338 schoolchildren they found 21 who fulfilled criteria for fibromyalgia, of whom 17 (81%) were hypermobile. Only 11% of the children without fibromyalgia were hypermobile. Conversely, 1.4% of non-hypermobile children had fibromyalgia compared with 40% of those with hypermobility. The interpretation of their findings is complicated by the fact that hypermobility of the joints in children is a common clinical finding and is often asymptomatic. If found in your area children (15% of 6–10 year olds) more often than older children (8% of 11–14 year olds), and in girls more often than boys. In total, 7–12% of school age children fulfil criteria for the definition of generalised joint hypermobility (table 2). Recurrent episodes of joint pain occur in about 40% of children with hypermobility; over twice as frequently as found in non-hypermobile children. The basis of joint pain in isolated hypermobility is also not clear, but may involve mild synovitis from repetitive subclinical trauma or increased muscle tension secondary to postural imbalance, or both. The consequences of hypermobility are also unresolved, but may include osteoarthritis, and associated features of mitral valve prolapse, varicose veins, and uterine prolapse. It is interesting to note that the traditional treatment of hypermobility by improving physical condition and increasing muscle bulk has also been used with some success in fibromyalgia.

Overlap between syndromes

There is clinical evidence that local and diffuse pain syndromes overlap; about 10% of children with one syndrome will also have distinct episodes of the other. In addition, internal or external stress is found in most patients with either condition, though it is often hidden and not asyntomacknowledged by the patient or the parents. The most commonly recorded stressors include high or over achievement in academia or sports, learning difficulties, single parent families, and sexual abuse. Enmeshment (an inappropriately close relationship between child and parent) and separation anxiety are commonly found in localised pain syndromes and, anecdotally, are prominent in diffuse pain as well.

The treatment of the conditions is also similar. It is vital for the doctor and other health care workers to have empathy with the child; failure to give credence to the pain will almost certainly exacerbate both local and diffuse syndromes. Physical rehabilitation and particularly the restoration of normal limb mobility are important. Psychological intervention to identify and deal with potential sources of stress may be useful. Coping techniques have been advocated; but techniques such as progressive muscular relaxation and guided imagery require controlled evaluation.

Finally, there appears to be considerable overlap between idiopathic musculoskeletal pain syndromes and other paediatric conditions, such as chronic fatigue syndrome, postviral fatigue, or myalgic encephalomyelitis. Walford et al recently reported 12 cases of paediatric chronic fatigue syndrome, defined by the predominant presence of idiopathic fatigue persisting for more than six months. All had the sleep disturbance typical of fibromyalgia and 11 also complained of musculoskeletal pain. Unfortunately, the patients were not examined for the presence of hyperalgesic trigger points (McCluskey DR, personal communication).

Conclusions

Chronic musculoskeletal pain syndromes in children are probably more common than has been appreciated. Confusion over the terminology of the conditions has undoubtedly contributed to their underrecognition; the terms fibromyalgia and reflex sympathetic dystrophy may be too exclusive for use in children, and ‘specialty dependent’ labelling (for example, fibromyalgia v chronic fatigue syndrome) almost certainly occurs. A true indication of the epidemiology of these problems will require a unified approach to their classification. Recently, criteria for the classification of two broad syndromes (localised and diffuse idiopathic pain syndromes) have been proposed, and a relationship between hypermobility and fibromyalgia has been noted.
What will be the impact of this information? It is likely that more children will be labelled as having a pain syndrome, but the importance of labelling alone should certainly be open to question. Does it make a difference if these conditions are diagnosed or not? It is not yet possible to answer this question scientifically, but the answer will hopefully be yes. Chronic musculoskeletal pain in children, even in the absence of an organic cause, can result in significant physical incapacity, loss of schooling, and social disruption. The problems may be exacerbated through an unsympathetic approach, excessive laboratory investigations, diagnostic uncertainty, and inappropriate treatment. New approaches to diagnosis and enhanced understanding of pathogenesis, may eventually decrease the morbidity and improve the outlook for children with chronic musculoskeletal pain.

T R SOUTHWOOD


Pedanius Dioscorides (ca AD 40 to 90) was born in Asia Minor and was educated in the Greek medical centres. He is best known as a military physician in the Roman armies under Emperor Nero. During his travels he studied many plants and minerals. He wrote de Materia Medica, which had a profound influence on medicine for about 15 centuries. As the sale of drugs in Rome deteriorated into a racket he tried to classify over 600 plants and offered sound medical prescriptions in his ("Herbal"). He mentions willow (salicylates) and ephemeron (colchicine) for gout.

He is depicted on a stamp commemorating the second congress of union of Arab physicians in 1965. This stamp is based on a 13th century Arabian manuscript. The doctor on the right, a herbalist, anachronistically in Muslim garb, holds a mandrake plant as he explains to a student its virtues as an analgesic-anaesthetic.

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Ann Rheum Dis 1993 52: 490-492
doi: 10.1136/ard.52.7.490

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