Non-inflammatory musculoskeletal disorders in childhood

J A Sills

Many children have musculoskeletal pain. These symptoms are relatively common, occurring in 4–30% of children in a non-clinic setting, whereas Allen found that 1.6% of children attending the children's emergency/ambulatory department at the Royal Children's Hospital in Melbourne presented with a non-traumatic muscle or joint problem. The British Paediatric Rheumatology Group (BPRG) has reported that of 4948 cases on their register, 40% of children had a diagnosis of juvenile chronic arthritis, 24% had mechanical/orthopaedic disorders, and 29% a diagnosis of 'other', which would include many with pain syndromes. It has been reported that the three most common recurrent pains in childhood are head pain (15–20%), abdominal pain (10–15%), and limb pain (15%). These figures therefore put musculoskeletal pain as among the most common forms of pain in childhood and this frequency (15 000/100 000 children) contrasts with the incidence of juvenile chronic arthritis of 14–16 per 100 000 children under 16 (0.015%).

Rosenberg, in his review from his clinic over a seven year period, noted over 100 different disorders presenting with musculoskeletal symptoms. Thus the high incidence of such symptoms in the general paediatric population is reflected in the BPRG figures, which show that paediatricians and rheumatologists with an interest in rheumatological disorders of childhood have a significant number of referrals that are not due to truly inflammatory rheumatological disorders.

Differential diagnosis

The problem for clinicians when faced with such symptom complexes as abdominal pain and headache is that the pathology may range from the serious and ultimately fatal to the benign with a good prognosis. There is a similarity with musculoskeletal symptoms, as within this presentation there may be pathology ranging from malignancy (local or disseminated) through chronic arthritis or another connective tissue disease which may evolve into a chronic disease, to a benign disorder with an excellent prognosis. Although a potentially manageable organic pathology can result in a good quality of life, some children may develop a chronic pain syndrome unassociated with any obvious cause, but which may produce significant short and long term morbidity. Hence the evaluation of children presenting with musculoskeletal symptoms and their families may be difficult, time consuming, and frustrating. Just as it is important to recognise those children with an organic basis for their symptoms so that they can receive appropriate treatment and management, so too is it important to recognise those children in whom there is no organic basis to explain their symptoms so that measures can be taken to facilitate their rehabilitation and to prevent them from being subjected to unnecessary and potentially harmful investigations and multiple referrals to different centres.

The differential diagnosis of musculoskeletal symptoms is wide. The table of Malleson and Southwood reflects the diversity of potential pathology as well as being a useful aide memoire. In addition, it has to be recognised that chronic fatigue syndrome may have to be considered and that some children's musculoskeletal symptoms are truly hysterical.

History

As in any presentation of any symptom, the history is of prime importance. The precise nature of the presenting disorder should be clarified. Are the symptoms constant or intermittent? Is there a relation with exercise or with other events? Do the symptoms occur at night and is there recovery with full muscle activity the next morning? (Nocturnal symptoms that persist into the next day are potentially pathological.) Are the symptoms localised to one joint or limb or are they more generalised? Evidence for systemic disease, as well as details of the past history, immunisation status, family history and dynamics—for example, position of the child in the family and significant life events—should be sought. An assessment of current health status and any details of foreign travel or travel in the UK should be included. Sporting activities and the ambitions within the sports may also be important.

It is also useful to establish the previous management of the problem—for example, other professionals involved, their specialty and status—tests that have been carried out, diagnoses offered or excluded, or both, and
Discrepancies
Muscle wasting/weakness/neurological deficit
Muscles spasm (can be overcome by passive movement?)

Joints (including back): normal position/posture (bizarre?), asymmetry, normal landmarks, visible, ranges of movement active/passive (any hypermobility? Table 2), evidence of inflammation—warmth, redness, pain tenderness—swelling—effusion (type of fluid), soft tissue, skin, bone, limb length discrepancy

Muscle wasting/weakness/neurological deficit

Table 1  Differential diagnosis of musculoskeletal pain (after Malleson and Southwood*)

- Avascular necrosis and degenerative disorders
  - Perthes’ disease
  - Osteochondritis, Osgood-Schlatter disease, Scheuermann’s disease
  - Slipped upper femoral epiphysis
  - Chondromalacia patellae
  - Hypermobility
  - Reactive arthritis: poststreptococcal, postenteric, postviral
  - Trauma: accidental and non-accidental
  - Haematological: leukaemia, neuroblastoma, lymphoma, haemophilia, haemoglobinopathy
  - Rickets: hypophosphataemic rickets, other metabolic and endocrine disorders (diabetes, hypo/hyperthyroidism)
  - Infection: acute—septic arthritis, osteomyelitis; chronic—tuberculosis, Brodie’s abscess
  - Tumour: cartilage, bone, muscle, benign (osseous ostitis, pigmented villonodular synovitis)
  - Idiopathic pain syndromes: localised and generalised
  - Systemic connective tissue disease: systemic lupus erythematosus, vasculitis (including Takayasu’s arteritis), Kawasaki disease, dermatomyositis, polyarteritis nodosa, scleroderma, overlap syndromes

Table 2  Physical examination: important observations

- Skin: rash, vasculitic lesions, café au lait spots, haemangiomas, psoriasis, poorly healed scars
- Nail and nail bed changes
- Joints (including back): normal position/posture (bizarre?), asymmetry, normal landmarks, visible, ranges of movement active/passive (any hypermobility? Table 2), evidence of inflammation—warmth, redness, pain tenderness—swelling—effusion (type of fluid), soft tissue, skin, bone, limb length discrepancy
- Muscle spasm (can be overcome by passive movement?)
- Muscle wasting/weakness/neurological deficit
- Discrepancies

Due to problems of modesty, especially in teenagers, so there may have to be some compromise.

Useful observations are those of gait (walk and run), hopping, toe and heel walk, getting up from squatting and cross legged sit, looking up to the ceiling and over shoulders, hands above head, scratching small of back, holding arms out stretched, pronation, supination, ‘saying prayers’, and flexing fingers. Useful areas of functional assessment are undressing/dressing, buttons, laces, and writing. The examination should include growth and general examination and specific examination of locomotor system and central nervous system.

Before starting the formal examination, however, there may be information from previous observations which may direct the sequence and emphasis of the formal examination. In the examination of the joints it may not be possible to follow a strict routine, particularly if the child is young, fractious, or uncooperative, and examination of the area of symptoms may be wise at first. In older children the affected limb or joint may be left to last. None the less, the points listed in table 2 should be considered during the examination.

It is important to assess whether there are discrepancies in signs—for example, it is not uncommon for a child with hip or leg pain to have a significant restriction of movement of the hip joint when the joint is being formally examined with the child lying supine. When the child does not realise that a particular joint is being examined, however, they may then show a much more normal range of movement—for example, when asked to sit up so that the back can be examined the child may sit up and lean forward to facilitate this without realising that it is the hip that is being covertly examined.

Evaluation
After the history taking and examination it may be possible to formulate a diagnosis, or at least a differential diagnosis, and to have categorised the problem into one of a number of differentials, the most important of which are inflammatory, infection, orthopaedic disorders, and malignancy (Table 3).

It is important to recognise hypermobility (Table 4) as this is a common problem. The symptoms often occur after exercise and they may occur at night as well as during the day, but on the day after the night time symptoms the child is usually back to normal. The hypermobility can be shown and indeed once the diagnosis is suspected, examination of the parents with specific questioning over similar symptoms in their own childhood may reveal evidence of the disorder in one or other parent and sometimes it becomes apparent that one or more siblings may also have the disorder. In severe cases, with or without a family history, the possibility of an underlying structural connective tissue disorder should be considered (for example Ehlers-Danlos syndrome). Examination of the symptoms relating to the clinical findings should provide reassurance because the prognosis is ultimately good, and although there is no cure there is at least a diagnosis and
Table 3  Practical differential diagnosis

- Inflammation (joints, muscles, skin, soft tissue etc)
- Infection (acute/chronic infection of bones and joints, for example Brodie's abscess/tuberculosis)
- Orthopaedic (including trauma)
- Neoplasia (rarely)
- Hypermobility (table 4): benign, rarely; structural/genetic disorders of bone, muscle or connective tissue (for example Marfan's syndrome, Ehlers-Danlos syndrome, multiple epiphyseal dysplasia)
- Fatigue syndrome
- Pain syndrome
- Supratentorial/unclear/combination

Table 4  Features of hypermobility

<table>
<thead>
<tr>
<th>Feature</th>
<th>No of points for each feature</th>
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<tbody>
<tr>
<td>Hyperextension &gt;10° of knees</td>
<td>1 (for each)</td>
</tr>
<tr>
<td>Hyperextension &gt;10° of elbows</td>
<td>1 (for each)</td>
</tr>
<tr>
<td>Thumbs apposed onto forearm</td>
<td>1 (for each)</td>
</tr>
<tr>
<td>Little fingers hyperextended parallel with forearms</td>
<td>1 (for each)</td>
</tr>
<tr>
<td>Excessive dorsiflexion of the ankle</td>
<td>1 (for each)</td>
</tr>
<tr>
<td>Ability to put palms flat on floor without flexing knees</td>
<td>1</td>
</tr>
<tr>
<td>Ability to put ankles behind neck</td>
<td>1</td>
</tr>
<tr>
<td>Ability to 'W-sit'</td>
<td>1</td>
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</tbody>
</table>

Table 5  Useful investigations

- Haematology: full blood count, sedimentation rate or plasma viscosity
- Biochemistry: C reactive protein, urea, creatinine, electrolytes, calcium phosphate, creatine phosphokinase, liver function tests
- Immunology: immunoglobulins, antinuclear antibodies, rheumatoid factor, HLA B27 and other tissue types
- Radiology: plain radiographs (in the early stages may show soft tissue swelling only), ultrasound, computed tomography, magnetic resonance imaging and radioisotope scanning
- Other (occasionally): biopsy sample of bone marrow, skin, synovial tissue (arthroscopically)

an explanation for the symptoms, which may prove reassuring for parents who may have been worried for some time.

A score of four or more is consistent with generalised hypermobility. Symptoms and hypermobility may, however, be localised to the knees or another joint. It is also important to be aware that transient effusion may occur in joints in hypermobility16 and that true juvenile chronic arthritis can occur in children with hypermobility. In contrast, hypomobility may occasionally be the problem and cause symptoms with exercise.

It may be possible at this stage to reach a confident diagnosis which can be discussed with the parents and child. Tests may be indicated clinically to clarify the diagnosis, to reassure the parents, child and clinician, as in many instances there is an expectation that there will be ‘tests’, but it is important to emphasise that there is no diagnostic blood test for ‘arthritis’ (table 5). It is important, however, not to over investigate as this can sometimes compound the problem. On the other hand, there may be indications to repeat investigations later.

If it is not possible to reach a diagnosis at the first consultation, further assessment and management may have to be delayed until the results of investigations are available when the situation can be reviewed, but it may be helpful to arrange a physiotherapy assessment in the interim period. It is also important to emphasise that the clinical features should be under constant review because symptoms and signs can evolve over time and what may initially appear to be a simple organic or non-organic disorder can evolve into a complex problem after a few weeks or months. Malignancy is notorious for behaving in this way.

Management

An explanation of the symptoms is important in management. Even if the diagnosis is not absolutely clear and the main differentials including arthritis have been excluded, an important statement/diagnosis for the child and parents may be ‘not arthritis’. Many parents will come to clinic desperately worried about the possibility of arthritis and with preconceptions based on experience of rheumatoid disease in adults. Hence reassurance that their child does not have any evidence of arthritis may be a great relief, and even if the diagnosis is arthritis it is important to emphasise that arthritis in children is different from rheumatoid disease in adults.

If the diagnosis is of an inflammatory disease, then appropriate specific management will be initiated, as would occur if there was evidence of infection. Similarly, if the diagnosis is of an orthopaedic disorder, then there may be referral on to that specialty, as would be the case if there was obvious or suspected malignancy. If the symptom is pain and the main differentials are excluded (infection, orthopaedic disorders, and malignancy), and no other pathology is found, then the management is of the pain/fatigue syndromes.

Localised pain syndromes

In adolescents and in athletic children who are growing rapidly, pain may be localised to the anterior part of the knee where the patella, the patellar tendon, femoral condyles, and quadriceps muscle form an important unit. Pain in this area can be due to a number of causes, including a patellar tracking abnormality, chondromalacia patellae (abnormality of the cartilage underlying the patella), recurrent subluxation of the patella, and osteochondritis dissecans. These latter two disorders should be diagnosed from the history and radiographs. Physiotherapy and medial patellar strapping may help in anterior knee pain.17

Back pain, which is common in adolescence (8% of 14–15 year olds),18 particularly if the pain is localised in the paravertebral muscles, may respond to physiotherapy once other organic treatable causes have been excluded. Again, it is important to be aware that unrecognised organic causes may evolve with the passage of time and reassessment may be necessary.

After an injury which may be relatively trivial, after stress, or for no immediately apparent reason, some children, but more often girls early in their second decade, will overprotect a joint by splinting it in one position (C Murray, A Cohen, T Perkins, J E Davidson, J A Sills, 1996, unpublished data). This can cause changes in blood flow to the limb and produce secondary changes, which may then lead on to cause the recognised pain syndrome of reflex sympathetic dystrophy or algodystrophy.19 20 In extreme cases where the disorder leads on to produce a fixed deformity, Sudeck’s atrophy is the result. The foot, but not uncommonly the
hand, will be held in a fixed position, the limb may be blue or red, it may be swollen, and it is often colder than the contralateral limb. There may be severe burning pain in the limb (causalgia) and pain on touch or movement, and these secondary features cause further concern. Often the disorder has been present for some time, the child may have seen several doctors of different disciplines, and it is not uncommon for the disorder to have been considered to be a soft tissue injury and for the limb to have been immobilised by slings or plaster of Paris, and weight bearing and activity proscribed. The cause of the disorder has been thought to be due to an abnormal reflex in the spinal cord between the pain pathways and the neurovascular nerves, which seems to set up an amplification of pain, but more recently this traditional explanation has been challenged.

It is important to recognise this problem, which is commonly seen by those in paediatric rheumatology, but which does not appear in paediatric textbooks, as it can cause long term morbidity and loss of school time.

It is also important to try to glean any history to suggest an underlying problem—for example, over performance in academic and sports fields, learning difficulties, or problems at home or at school such as bullying or even sexual abuse. Physiotherapists skilled in the management of this disorder can usually achieve rehabilitation without the need to recourse to further treatment such as guanethidine nerve blocks or the use of transcutaneous nerve stimulation. It is not uncommon for the disorder to reappear, often in another limb, if the child has further stress. In many instances an underlying cause may not be found and persistence in attempting to find a cause can retard progress and thus a pragmatic approach may be the wisest course. In most instances referral to child and adolescent psychiatry services should be considered.

Chronic diffuse pain is often associated with tender trigger points, typically on the upper border of trapezius, and has been termed fibromyalgia. It usually responds to physiotherapy. There may, however, be somatic symptoms such as anxiety, tension, irritable bowel, headaches, and sleep disturbance, and these symptoms may well be part of the presentation in a child whose main problem is one of fatigue. Chronic fatigue syndromes can therefore also present with musculoskeletal symptoms and, as in fibromyalgia and other pain syndromes, physiotherapy is one important element in the rehabilitation of such children.

Even with comprehensive clinical evaluation, investigation, and observation there is still a group of children who do not have an identifiable pathology or pain syndrome, who are well, with no physical signs, no muscle weakness, and no underlying neurological disorder. Often these children are of primary school age and have predominantly night time symptoms, which are often improved by massage of the painful area. By exclusion, these children can be regarded as having nocturnal idiopathic pain syndrome and this can be discussed and conceptualised with the parents as ‘growing pains’.

There may be some children in whom it becomes clear that the problem is not organic. There is often a discrepancy between symptoms and the degree of disability and signs: they may have shown discrepancies during examination, the symptoms may not have been constant during periods of absence, often becoming worse when their parents visited or when one of the medical team was in the vicinity, or when they were being formally reviewed. Covert observation by nursing, medical, and other staff and by the physiotherapists is helpful in clarifying the problems with such children and empowering the clinicians to make referrals on to child and adolescent psychiatry services. Additional useful information about the child and the family may be obtained through the general practitioner and often the child’s teacher. In some instances, however, physical therapy, in particular physiotherapy, can succeed in improving such children by providing them with a face saving route to recovery or ‘a rope to climb down’ (D Jones, personal communication).

On occasions the symptom complex may be truly hysterical—that is, prominent somatic symptoms which indicate a disturbance of the central nervous system or limb dysfunction, but where investigation by a specialist unit has not established any anatomical or physiological basis for the disorder. Such symptoms may include those given in table 6.

In addition, there may be ‘belle indifférence’ to the problem and perhaps evidence of an ‘ally’: someone who believes in and colludes with the symptoms. Early referral of these disorders after basic exclusions of organic pathology is best, as over investigation may compound the problem. It is important, however, to recognise that it is not uncommon for organic symptoms to be considered to be hysterical in the early stages. Early discussion with psychiatric colleagues to plan a strategy and the appropriateness of referral is wise.

The role of physiotherapists in the management of children with pain syndromes and hysterical symptoms should not be underestimated. Through their ability to build up a relation with the child and the parents they may well be able to tease out significant unresolved problems which have a direct bearing on the symptom complex, and which would not come to light in the course of a routine consultation. Thus the physiotherapists can provide valuable additional information relevant to the clinical symptoms which may reinforce a view that the child’s symptoms do not have an organic basis and they do therefore require formal referral to child and adolescent psychiatry services.

This review cannot be totally comprehensive and cannot include every differential, but it is hoped that it provides a practical basis for clarifying the diagnoses of children presenting with these symptoms, preventing their over investigation and facilitating their early recognition, management, and rehabilitation.

<table>
<thead>
<tr>
<th>Table 6 Symptoms suggesting non-organic pathology</th>
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<tbody>
<tr>
<td>Abnormal gait</td>
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<tr>
<td>Paralysis</td>
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<td>Fits</td>
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<td>Torticollis</td>
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<td>Vomiting</td>
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<td>Incoordination</td>
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<td>Regression</td>
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<td>Amnesia</td>
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<td>Aphonia</td>
</tr>
<tr>
<td>Visual disturbances</td>
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<tr>
<td>Pain</td>
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</tbody>
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I acknowledge the help and support I have received from my physiotherapy colleagues, Tessa Perkins and Alison Cohen, over the years. They have taught me much and without their expertise, dedication, and intuition many children would not have recovered from both organic and non-organic musculoskeletal disorders.