Spondyloarthropathy in childhood: a review[1]

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The classical descriptions of ankylosing spondylitis, including the proposed criteria for diagnosis in population studies (Bennett & Wood 1968), have required reappraisal as knowledge has accrued, notably that related to the familial and racial aspects of the disorder, and more recently the clear correlation with HLA B27 (Brewerton *et al.* 1973, Schlosstein *et al.* 1973, Bywaters 1980). The concept of a spectrum of seronegative peripheral and spinal arthritic syndromes with sacroiliitis and familial incidence has slowly emerged (Moll *et al.* 1974).

In childhood, classic ankylosing spondylitis has been described (Edstrom *et al.* 1960, Ladd *et al.* 1971), but many children present with a peripheral arthropathy and only later develop sacroiliitis and/or ankylosing spondylitis (Ansell 1980, Schaller 1977). In a study on the natural history of juvenile chronic polyarthritis, the follow-up criteria used to delineate juvenile ankylosing spondylitis were sacroiliitis of ankylosing spondylitic type which could be unilateral or bilateral, with or without back limitation, and radiological change in the spine. Using these criteria a subgroup emerged characterized by a lower limb arthropathy affecting boys aged 9 and upwards (Ansell & Wood 1976). Subsequent studies on a group of patients so identified showed that 90% carried HLA B27 (Ansell *et al.* 1977). This paper will review current thoughts on spondyloarthropathy commencing before 16 years of age.

Clinical features

The age of onset is about ten years, with a wide range; boys are affected six times as frequently as girls (Figure 1). In our experience about 80% present with a peripheral arthropathy, the joints most frequently affected being hips and knees, followed by ankles. Approximately one-quarter give a history of pain in the heel, either due to plantar fasciitis or bursitis around the Achilles tendon, while involvement, usually asymmetrical, of interphalangeal joints of toes or metatarsophalangeal joints is not infrequent. Upper limb involvement is seen in less than 10% in the first few months of the disease. Very occasionally the presenting feature is pain in the neck, often accompanied by a monarthritis affecting particularly a knee or ankle. Atlanto-axial subluxation can be seen at this stage or later (Reid & Hill 1978). Recurrent irritable hip syndrome, either in the same hip or alternating from one hip to the other, the presence of persistent discomfort around a hip or buttock, particularly if associated with loss of movement, should alert one to the possibility of spondylitis. This mode of presentation is particularly common in teenage males, and there may occasionally be complaints of discomfort in the chest due to manubriosternal joint or costochondral junction involvement.

Schaller (1979) suggests that approximately half the cases of spondylitis of childhood begin with pain around the hip girdle, and the other half with peripheral arthropathy, the latter group being the younger patients. This distinction, however, is not always clear cut as some young patients may present with acute or chronic hip pain, while some teenagers present with persistent ankle and foot involvement, which can be followed fairly quickly by asymmetrical erosive change.

At presentation, sacroiliitis is very unusual but gradually develops over the years, the mean duration to definitive sacroiliac change being 6.5 years (Ansell 1980, Figure 2). Back limitation may take many years longer to develop and has been detected as late as 30 years

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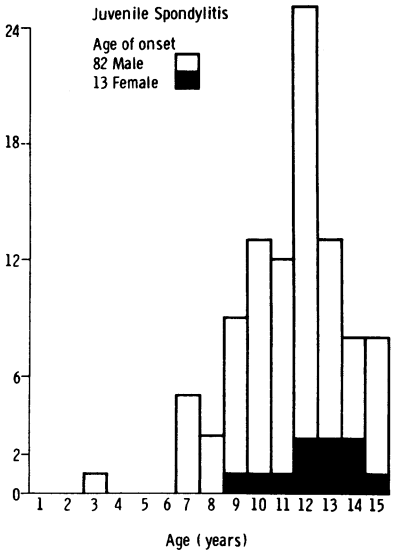

Figure 1. Age and sex distribution of peripheral joint arthritis in 95 patients who ultimately developed sacroiliitis

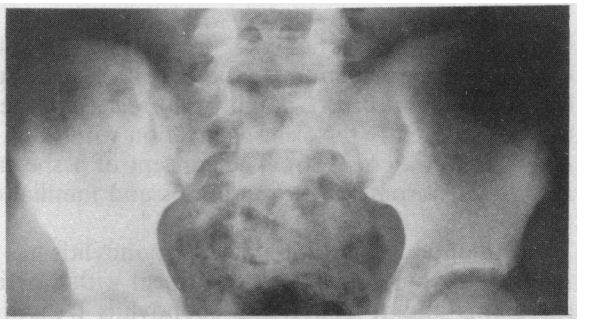

Figure 2. Very early sacroiliitis in a boy aged 12 who had presented with recurrent irritable syndrome two and a half years earlier

after the onset of peripheral arthropathy; in our experience it has been remarkable how infrequently the patient made a complaint of back pain at the time limitation was first noted. Typical radiological changes are ultimately seen and do not differ from those of adults at onset of ankylosing spondylitis although they develop much later (Riley *et al.* 1971).

The erythrocyte sedimentation rate, measured by the Westegren method, is usually raised except when there is only a single joint affected. In the majority the haemoglobin is normal, but persistence of prolonged peripheral arthropathy is associated with some fall in haemoglobin. Immunoglobulins, particularly IgG, are usually raised, but only in those patients with widespread polyarthritis. IgM rheumatoid factor is absent, as are antinuclear antibodies. HLA B27 is usually present. In the acute phase the synovial fluid frequently shows a polymorphonuclearcytosis, high protein content and increased complement activation. Histologically, the synovial membrane tends to resemble that seen in rheumatoid arthritis but with a specific feature that vascular granulation tissue can develop away from the cartilage periphery, erupting out from the middle of the joint instead of spreading from the margins (Bywaters 1976). Diagnosis is difficult because of the lack of specific haematological features and the fact that the sacroiliac joint interpretation is extremely difficult in the teenage period (Jacobs 1963). Similarly, epiphyseal development interferes with simple scanning techniques as epiphyses tend to cause an increase in uptake; newer methods with computerized tomography may be more useful.

The course of the disease is somewhat variable. In 50% the peripheral arthropathy remits and sacroiliitis, followed by minor back changes, develops; a further small proportion of patients in whom the peripheral arthropathy remits will go on to more classic ankylosing spondylitis. Approximately one-third of the patients slowly develop further incapacity due to recurrent episodes of peripheral arthropathy, increasing hip problems or progressive changes throughout the whole spine. The most serious functional problem results from limitation of hip movement, particularly if the patient develops stiffening in the lumbar spine. Radiologically, a number of patterns of hip change have been noted (Ansell & Kent 1977) but complete bony fusion is rare; the most common type is illustrated in Figure 3. Acute iridocyclitis punctuates the course, but in our series has never been the presenting feature; by 15 years from the onset it had occurred in 27% (Ansell 1980). Very occasionally the late

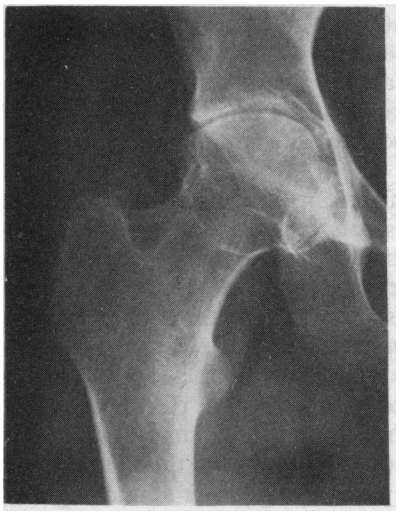

Figure 3. Joint space narrowing with minor erosive change and change in shape of femoral head with early ruff formation in a patient of 18 who had presented with a stiff neck aged 11 years

development of amyloidosis has been noted; this has been particularly in those who have had a severe peripheral arthropathy associated with a high sedimentation rate. Aortic incompetence has also been noted to develop in a proportion and may be present before obvious spinal changes have occurred (Kean *et al.* 1980).

Related disorders

Our long-term study suggests that some 6% of children presenting in this way ultimately develop psoriasis and 8% a bowel disorder, regional enteritis more usually than ulcerative colitis. Psoriatic arthritis in childhood is still not well defined (Lambert *et al.* 1976). It tends to commence around the age of 10 and a number of patterns similar to those seen in adults has been noted. Back symptoms occur late, and in general there appears little difference in psoriatic spondylitis from the description of juvenile spondylitis already given. The presence of nail pitting in the subject, a family history of psoriatic arthritis or psoriatic spondylitis, should alert one to its possibility. The incidence of HLA B27 in the small group we have as yet studied does not seem to be as high in the juvenile psoriatic spondylitics as in the uncomplicated spondylitics. There is considerable overlap with bowel disorders; thus in our psoriatic group we have seen regional enteritis and ulcerative colitis develop, supporting the concept that the spondarthritides in the juvenile behave in a similar manner to those in the adult (Wright & Moll 1976).

In inflammatory bowel disease two types of arthritis occur, namely 'intestinal peripheral arthritis' and 'spondylitis'. The former is characterized by episodes of synovitis, usually affecting knees and ankles, which may or may not be associated with exacerbations of the bowel disease. There is no apparent correlation between the arthritis and the severity or type of bowel disease (Lindsley 1977). In this group, other manifestations, particularly skin lesions such as erythema nodosum, pyoderma gangrenosum, and mucous membrane ulcers, are not uncommon. Spondylitis also presents as a peripheral arthropathy but affecting the lower limbs, and can be very difficult to differentiate. The incidence of hip involvement, however, is much higher than in intestinal synovitis and the joint involvement in the spondylitic subgroup does not seem to have the same transient course. Thus an adolescent presenting with a lower limb arthropathy, perhaps associated with anaemia, persistently positive occult bloods,

growth failure, or vague abdominal pain, should immediately be investigated for underlying bowel disorder, particularly regional enteritis. This is one of the few occasions when it may be justified to consider performing tissue typing to try to establish which pattern of arthritis is present, as intestinal synovitis will improve as the bowel state does but the same is not true of the spondylitic variety. At times the arthropathy may appear to antedate the bowel disorder by many years.

During childhood infective diarrhoea, which can be from any organism – salmonella, shigella, yersinia, campylobacter – is the commonest cause of Reiter's syndrome, although with sexual activity beginning earlier one can expect to see some sexually-acquired cases in the early teens. As the episode of diarrhoea may have been minimal it can easily be missed unless a careful enquiry is made in all cases of acute arthritis. The typical triad of urethritis, conjunctivitis and arthritis tends to develop within about ten days, but it can be anything from three to twenty days after the initial incident. There is no consistency in the order in which symptoms develop. In small children it is often difficult to spot a urethral discharge, but a history of dysuria may be obtained. Mucocutaneous lesions are not uncommon. These are mild and have to be searched for; they include erythematous ulceration on the hard palate, scattered pustules on the glans penis, and ulceration of the penis. A high fever mimicking that of systemic juvenile chronic arthritis has been recorded, but the typical maculopapular rash does not occur. Jacobs (1977) uses the term 'Incomplete Reiter's Syndrome' for children who present with polyarthritis and a history of diarrhoea or other suggested features such as conjunctivitis or mucocutaneous lesions. There is considerable overlap between this concept and that of 'reactive arthritis', this term having been coined by Aho *et al.* (1975) to describe arthritis following dysenteric infections, particularly *Yersinia enterocolitica* which is associated with HLA B27. Although there are juvenile patients described in his series, relatively little attention has so far been paid to this form of arthritis in childhood which overlaps so considerably with incomplete Reiter's syndrome following dysentery. It is very likely that in the past these children were just regarded as atypical juvenile chronic arthritics, as they certainly have a wide variety of features. In the young child, the arthritis may be a mild episode affecting two or three joints or a severe polyarthritis, associated with a high fever. In older children, particularly teenagers, there is asymmetrical migratory involvement more closely resembling that of adult disease, and particularly in older boys the arthritis can be severe and persistent with residual deformities developing, particularly in the feet, which differ in no way from that of adult Reiter's syndrome. Sacroiliitis can occur relatively early in these older patients and some will ultimately go on to ankylosing spondylitis.

Although arthritis and asymptomatic sacroiliitis have been described in familial Mediterranean fever, this does not appear to be related to HLA B27 (Lehman *et al.* 1978). As yet, the development of spondylitis and its relationship to the family of disorders just discussed, is uncertain.

Conclusions

Some 15–20% of children with seronegative chronic arthritis may well belong to one of the categories of spondyloarthropathies described here. The patterns of illness are only gradually being elucidated, but suspicion should arise, particularly in teenagers who present with a lower limb arthropathy or have one of the known associated features of the spondyloarthropathies, either personally or in their family.

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