joints due to overgrowth of the epiphyses; later features include chronic meningitis (often with mental retardation) and lesions in the eye, of uveitis, keratitis, and papilloedema.[11] The difference in prognosis makes it important that this syndrome should be differentiated from systemic juvenile arthritis.

A polyarthritic onset of juvenile chronic arthritis occurs in girls more commonly than boys and as young as 5 to 6 months of age. This will need to be differentiated from several rare causes of swollen joints. Farber's disease (characterised by a deficiency in acid ceramidase) tends to occur in the first few months of life, causing enlargement of the joints, subcutaneous nodules, and appreciable hoarseness.[12] The mucopolysaccharidoses, particularly the Hunter-Scheie syndrome, may well present as flexion deformity of the fingers and stiffening of the joints.[13] The Negro child of 6 months to 2 years with diffuse symmetrical, extremely tender, warm swelling of the hands or feet or both, associated with a fever and peripheral leucocytosis, should be suspected of having hand-foot syndrome of sickle cell disease. The presence of bruising in a frightened child who resists examination should make one suspect a battered child, while dislike of handling and apparent enlargement of the joints are occasionally seen in infants of Asian extraction with rickets. Neuroblastoma may present with synovitis, but this is usually mild, the pain arising from bony infiltration. Similarly, in leukaemia, though the joints may be swollen, the bony tenderness is out of all proportion to the degree of swelling, as is the overall disturbance in general health and low haemoglobin value.[5]

The pauciarticular onset of juvenile arthritis is characterised by fewer than five joints being affected in the first three months of the illness, and there is little disturbance of general health. Young girls appear at particular risk for the development of chronic uveitis and positive test results for antinuclear antibodies. Such cases may need to be distinguished from sarcoidosis, which, particularly in young children, may present with rashes developing into subcutaneous nodules, chronic uveitis, and painless, non-deforming arthritis, with boggy soft tissue swelling affecting particularly the wrists and knees, but also other joints. Although bone changes are seen in roughly half of these patients, pulmonary lesions are relatively uncommon.[14] The presence of uveitis in a child with a single swollen joint suggests juvenile chronic arthritis and in its absence other causes must be excluded. One of the more difficult is an infection which has been partly controlled with antibiotics given perhaps for an intercurrent infection elsewhere. Tuberculosis—which used to be the most common cause of chronic monarticular arthritis in a knee, hip, or wrist—is now extremely rare in white children, though it is occasionally seen in Asian children in Britain. Thorn synovitis in Britain is usually due to a rose thorn or wood splinter rather than a palm thorn.[15] Intercurrent swelling of a knee, often associated with minimal trauma, may result from a haemangioma of the synovial membrane.[16] The bloodstained aspirate will need to be differentiated from other causes of haemarthrosis, such as haemophilia. When a child with a very low factor VIII value (1-5%) starts to walk haemarthroses occur, the knee and ankle being particularly common sites.[17]

Clinical assessment calls for a good history to ascertain information about the child's general health, preceding events including upper respiratory and gastrointestinal infections as well as previous illness; the family history may be important. Examination should be conducted with all the child's clothes removed at some stage, when the general condition should be noted and when evidence of lymphadenopathy, hepatosplenomegaly, rashes, and local wasting may be assessed adequately. Diagnosis rests on clinical acumen supported by appropriate laboratory investigations.

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Blood counts and economics

When as a trainee pathologist I asked, "What is a full blood count?" I received the wise reply, "A full blood count is as much as is appropriate to that particular patient." Today's automated blood counters, however, have made us take red cell indices for granted and expect an accurate white cell count and platelet count. Some laboratories present clinicians with data from such apparently meaningless measurements as red cell distribution width and plateletcrit. Even more expensive machines provide automatic white cell differential counts done not on the 100 cells usual when examining a blood film but on 10 000.

No one questions the value of automated blood counters in terms of efficiency, accuracy, and reproducibility. The availability of an accurate mean cell volume has transformed the practice of haematology, and no laboratory could possibly cope with the numbers of requests received nowadays without the speed that these machines bring. So with all this extra information is there still a need for the haematologist to examine the blood film? And what is the place of the differential count?

The first question is easily answered. Examination of a blood film gives immeasurably more information than an automated

cell counter and should always be undertaken when the automated count is abnormal—and always by someone experienced enough to recognise what he is seeing rather than by a junior medical laboratory scientific officer likely to punch out a differential count and miss important morphological changes. A blood film should also be examined even if the automated count is normal if there is a good clinical reason—suspected glandular fever is a good example. A differential count should form part of that examination only when such numbers need to be known: when monitoring the effect of chemotherapy, for example, or looking for the toxic effect of drugs. We need, however, to be cautious of the spurious security in numbers that takes no account of variation due to sex,[1] smoking,[2] diurnal changes,[1] and random error in counting.[3] The value of the differential count in establishing the presence of infection when the total white cell count is normal is much less certain. Only five of 106 patients with acute appendicitis studied by Raftery had normal white cell counts with abnormal differentials, whereas four had completely normal blood counts.[4]

Possibly clinicians may request differential counts for less than adequate reasons, such as buying time or simple curiosity,[5] and some laboratories perform them on all blood counts as routine. Such activities are known to epidemiologists as "case findings" and are not particularly profitable.[6] Rich *et al* uncovered no clinically inapparent disease from 475 differential counts done for this reason.[7] Although they are not expensive—depending on who does them, differential counts cost between 25 and 50 pence each—a lot are done. An average sized laboratory might spend £15 000 a year on case finding. In the context of health service economics it is the little things that cost a lot.[8]

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Urticarial vasculitis

Urticarial vasculitis is a diagnosis applied in recent years to patients with urticarial weals which on histological examination have shown leucocytoclastic vasculitis (venulitis).[1-5] The general picture is one of individual weals which tend to persist—many last over 24 hours. Initially these weals were said not to itch, but this is a variable feature. Occasionally purpura may be seen in the centre of the weal, and rarely lesions resembling erythema multiforme may appear. In most cases there are no clinical features that will clearly differentiate vasculitis from ordinary chronic urticaria—angio-oedema and weals at the site of pressure occur in both. Arthralgia is more likely to occur in the patients with vasculitis, and less common features include abdominal pain, fever, lymphadenopathy, and rarely glomerulonephritis.

The histological picture varies but usually shows leucocytoclasis, fibrinoid deposits in and around the vessel walls, and a dense perivascular cell infiltrate, often with neutrophils predominating and present in the vessel walls. There may also be extravasation of erythrocytes and swelling of vascular endothelial cells. Direct immunofluorescence may show deposition of immunoglobulins, complement, and fibrin, but with no distinct pattern. Serological investigations show a raised erythrocyte sedimentation rate in many cases. Hypocomplementaemia is present in less than half the cases, and in a few the reduction of C1q is due to 7 S C1q precipitins. Many patients show evidence of circulating immune complexes.

One unresolved question is the relation of urticarial vasculitis to systemic lupus erythematosus, which has vasculitis as one of its histological features. In some series of patients with systemic lupus erythematosus up to 5-10% have had urticaria; indeed, O'Loughlin *et al*[6] recorded urticaria in 12 out of 54 patients with systemic lupus erythematosus. As with urticarial vasculitis, the weals may last longer than those of ordinary chronic urticaria and purpura may be present, but clinically urticarial vasculitis, chronic urticaria, and urticaria associated with systemic lupus erythematosus may be impossible to distinguish. Other manifestations of systemic lupus erythematosus may be present, including facial erythema, discoid plaques, livedo reticularis, and lesions of the nail folds. Cerebral, retinal, and renal lesions may also occur. Serological changes of systemic lupus erythematosus are usually present, and direct immunofluorescence is likely to show granular deposits in clinically normal as well as lesional skin. Considerable overlap between systemic lupus erythematosus and urticarial vasculitis seems the most likely explanation of these findings.

Perhaps the most interesting aspect of urticarial vasculitis lies in its relation to ordinary chronic urticaria. In a series of patients with chronic urticaria fully investigated some 5-10% have been shown to have leucocytoclastic vasculitis (this includes the 1-2% with systemic lupus erythematosus), and less than half have shown complement abnormalities.

Patients with severe urticarial vasculitis show more obvious clinical manifestations of the condition, and are more likely to have serological and histopathological changes than those with milder disease. The important question is, therefore, at what point does urticarial vasculitis merge with ordinary chronic urticaria?

Phanuphak *et al*[7] investigated 42 consecutive patients with chronic urticaria. Although their criteria for the histological diagnosis of vasculitis differed from those of other workers, they recorded 22 with vasculitis and a further eight with minor histological changes of oedema in the vessel wall and perivascular infiltrates suggesting a minor degree of the same condition. Monroe *et al*[8] investigated 45 patients with chronic urticaria, whom they divided into three groups. The first nine showed leucocytoclastic vasculitis with fibrinoid deposits, the second group of 15 a dense perivascular infiltrate of lymphocytes and eosinophils, and the third group of 21 showed sparse lymphocytic perivascular infiltrate. Direct immunofluorescence showed deposition in blood vessels of immunoglobulins and complement or fibrin, or both, in 33% of the first group, 13% of the second group, and 9% of the third. Similarly serological investigations showed an increased incidence of circulating immune complexes in the first two groups, and in these groups