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Acute febrile mucocutaneous lymph node syndrome (Kawasaki disease)

Sir,

We would like to report the following case. A two-year-old girl was referred to the dermatology clinic after presenting with a seven-day history of sore throat, fever and a skin rash. A five-day course of cloxacillin had had no effect on the pyrexia. Examination showed an irritable, ill child with a pyrexia of 39°C and there was a blotchy erythematous rash on her trunk and legs, with areas of desquamation. Her fingertips were red with peeling back of the skin towards the base of the fingers. There was left cervical lymphadenopathy and conjunctival injection. Examination revealed no other abnormalities. The diagnosis was not clear at this stage and she was admitted to hospital for further evaluation.

Over the next few days she remained highly pyrexial despite penicillin V therapy. Extensive crusting and ulceration of the tongue and lips developed which necessitated feeding via a naso-gastric tube. Significant haematological findings included neutrophil leucocytosis, thrombocytosis and raised erythrocyte sedimentation rate (ESR). Both electrocardiogram and chest radiograph were within normal limits. Cultures from multiple sites revealed no pathogens; blood and urine cultures were sterile and serological tests were negative. At this stage a diagnosis of Kawasaki disease was made. Detailed questioning of the mother about events prior to the onset of symptoms revealed that the house had been extensively 'spring-cleaned' during the preceding week.

Soluble aspirin was commenced and there was a dramatic clinical response. The pyrexia resolved within 24 hours and the mouth ulceration healed within four days. The skin rash and desquamation had cleared within a fortnight. The soluble aspirin was stopped after four weeks, the platelet count and ESR having settled to within normal limits. At this time also, a distinct transverse furrow was noticed on all the fingernails (Beau's lines). At follow-up over a two-year period she has remained well apart from developing allergic rhinitis, eczema and mild asthma.

Our patient presented with a perplexing clinical picture which did not really correspond with any of the common febrile illnesses. However, it did fit with all of the established principal features of Kawasaki disease[1,2] which are fever, conjunctivitis, mouth ulceration, inflammation of palms and soles, exanthemata and cervical lymphadenopathy. Kawasaki disease is most common in children between six months and four years of age. It occurs most commonly in the late winter and spring. Cardiac involvement occurs in 20% of patients with Kawasaki disease, with ensuing fatality in 1–2%. Fortunately there was no evidence of cardiac involvement in this patient. Another feature is thrombocytosis which may increase the risk of thrombosis. The most useful treatment is aspirin which has several beneficial effects. Corticosteroids appear to be contraindicated.

The cause of Kawasaki disease remains unknown. Clustering of cases has suggested a common infectious agent though none has been consistently isolated. Immunological tests during the acute phase of the illness have indicated exposure to the house dust mite which suggests that the disease may be due to a hypersensitivity to the mite itself or to some organism carried by the mite.[3] Our patient would probably have been exposed to high concentrations of house dust mite during the 'spring-cleaning' of her house just before the onset of her illness and so this could have been the precipitating event.

We present this case to make this disease more widely known among general practitioners since they spend a significant amount of their time in dealing with febrile illness in young children. The differential diagnosis includes scarlet fever, staphylococcal scalded skin syndrome (toxic epidermal necrolysis), Stevens-Johnson syndrome and Reiter's syndrome. Because of the potentially fatal outcome of Kawasaki disease, early recognition and prompt treatment are essential.

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The treatable canary

Sir,

The efficacy of doctors' management and treatment of patients depends on another part of their job — the reaching of a diagnosis. The importance of making a correct diagnosis is never more obvious than when a incorrect one has been made or an important one has been missed. The least that a patient expects of their doctor is that he or she will know what, if anything is wrong with them. Doctors are not omniscient, yet along with their other tasks they are required to find a cause for a multitude of complaints and queries. How best can they do this, correctly, consistently and promptly?

Underpinning the teaching and practice of making the diagnosis are the almost absurd-sounding maxims that 'common things occur most commonly'; and that 'the bird on the wire is more likely to be a sparrow than a canary'. But there is no absurdity here. These maxims, though imperfect, are valuable devices bringing a logical sequence out of the multiplicity and even chaos of symptomatology. For each symptom and sign, a huge list of possible causative diseases may be compiled. It is impossible to try and exclude by thorough investigations each and every disease until the correct one is alighted upon. Over-investigation is costly, meddlesome and does a disservice to the patient. The matching of symptoms and signs to the frequency of occurrence of each disease — its commonness — usually leads speedily to a correct diagnosis.

Thus, most patients with headaches do not need computerized tomography scans. Their headaches are understandable using simple means and have a common, benign cause. Most children with a fever and red pharynx need neither a blood test nor a bone marrow examination. Their upper respiratory infection is common, benign and self-limiting. The problem is that brain tumours and leukaemia do occur, and provided they are diagnosed at a sufficiently early stage they can be treatable and curable. Yet they are often missed until it is too late. Review of the history in these cases may reveal their con-

dition could have been diagnosed earlier, had the doctor thought of it. The main reason such conditions are missed is because they are uncommon.

Such is the importance placed upon first selecting the commonest diseases that it becomes imprinted upon the doctor's mind. The quickest way to run into trouble in under- or post-graduate examinations is to state a less common disease before a common one. Commonness, however, can become too easy a way of evaluating symptoms. Picking the commonest disease first can become a thought-precluding habit.

It is not enough for medical minds and memories to have, for each symptom and sign, just a list of diseases in descending order of frequency of occurrence. Alongside there must be a list of treatable diseases. Indeed, there is a powerful case to be made for diagnostic thinking to be guided by treatability — the most common treatable disease being at the top and the first to be thought of and excluded.

Untreatable disease is best diagnosed early or inappropriate management may make matters worse. The consequences, however, of missing untreatable disease are generally less dramatic than missing treatable disease. With this in mind, should doctors relegate self-limiting or untreatable disease to second place for their time and interest? The answer, in terms of the care and management which all illness demands, is a categorical no. But the answer in terms of diagnosis, because it will lead to more curable disease being promptly identified, is an equally categorical yes.

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Surveillance programmes for sudden infant death

Sir,

A number of health authorities are introducing surveillance programmes for sudden infant deaths based on the Sheffield study,[1] in which a risk score system was devised by the analysis of maternal and infant factors and the circumstances of the birth. It was designed so that 15% of infants were deemed to be at 'high risk' and the remainder at 'low risk' of sudden infant death. By increasing health visitor care to 'high risk' infants it is hoped to decrease the incidence of sudden infant deaths.

No one can criticize the laudable aim of trying to tackle the problem of sudden unexpected infant death, but it is premature, and possibly unethical, to introduce increased health visitor surveillance to some infants and to tell parents that their child is at high risk of suddenly dying. 'High risk' is a relative term and in the Sheffield study is less than 2% even at the highest score. Telling parents that their child is at high risk certainly causes marked parental anxiety[2] (personal observation) and unless a proven decrease in sudden infant deaths occurs as a result of increased health visitor surveillance we should not give parents false expectations or cause unnecessary anxiety.

All new methods of care should be subject to careful, scientific evaluation before they are adopted on a wide scale. Such evaluation does not appear to have been the case with the sudden infant death surveillance programmes in operation in the UK at the present time. In the Sheffield study it is claimed that by increasing the number of visits by their health visitors from the standard three to nine (in the 'high risk' group) there has been a marked decrease in the incidence of sudden infant deaths in Sheffield. In addition the authors claim that the reduction in deaths is numerically similar to the number of lives saved by treating cancers in children.[1] These claims cannot be sustained from the data available.[3]

The reasons[4] why control groups were not being continued past the first year of the Sheffield study have been criticized.[5] The Sheffield team extrapolated their observed data to estimate expected mortality had the number of health visitor contacts not been increased in the 'high risk' group. Recommendations are being based on this dubious approach and on the assumption[1] that there is a log linear relationship between risk of death and risk score. This has not been confirmed in an Irish population.[6]

In evaluating surveillance programmes, it is essential to compare increased health visitor contacts with a standard number of health visitor contacts in both 'high risk' and 'low risk' groups within the same population, over the same period of time, and with a sufficient population size. Until such studies are made, there is no case for introducing[7] or continuing birth scoring systems for sudden infant deaths.

These expensive and unproven exercises should cease until there is adequate funding to ensure that there is no decreased health visitor provision for the elderly or for clinical programmes of proven cost-effectiveness.

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Buying and selling practices

Sir,

We are a group of five principals currently in the process of building a new health centre in inner London at a cost of around £500 000.

Because there is effectively a moratorium on all new health centre building by district health authorites we have had to finance the cost of this ourselves, primarily through the General Practice Finance Corporation. We are not keen to lease back to the GPFC as we feel the future of this particular quango cannot be guaranteed in the future.

These financial arrangements will mean that in about 10 years time, any partner leaving will have to be paid a substantial sum for their share of the value of the building. Similarly any incoming partner will have to contribute a very substantial sum of money to buy in. The longer the period from construction, the greater the sums of money involved. In short, buying and selling of practices is still a major consideration for incoming or leaving partners.

We would be interested to hear from any general practitioner or partnership who have devised a means of getting round these problems either by clauses in the partnership agreement or by the setting up of some kind of charitable trust, company or other device. Our aim would be to end buying in or selling out.

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