The effect of immunoglobulin on Vβ expression. Therefore the demonstration of Vβ changes after administration of immunoglobulin early in the disease may be difficult. In the UK, Kawasaki disease is unfortunately still often diagnosed late out of season, and treatment with immunoglobulin may therefore be delayed beyond the tenth day of illness. This has enabled us to study patients referred up to 21 days after disease onset before the administration of immunoglobulin.

In contrast to Leung et al we have found a variety of different staphylococcal toxins in throat and nose swab culture supernatants from children with Kawasaki disease and their relatives.2 In addition we have found activity suggesting the presence of one or more novel superantigen toxin or toxins. We propose that Kawasaki disease is either caused by more than one toxin or is caused by a novel superantigen toxin.

We disagree that we may have misled readers to conclude that superantigen involvement in Kawasaki disease is a proved fact. We concluded that our data 'supports the hypothesis that a superantigen is involved in the pathogenesis of Kawasaki disease'; a hypothesis that remains to be tested in further studies.

Burkholderia cepacia and ΔF508 homozygosity in cystic fibrosis

EDITOR—Colonisation by Pseudomonas aeruginosa has long been recognised as a common trait in cystic fibrosis. Lately also Burkholderia cepacia, formerly known as Pseudomonas cepacia, has emerged in cystic fibrosis as a significant, although not so widespread, pathogen.1 As it has been suggested that homozygotes for ΔF508, the commonest cystic fibrosis mutation, could be more often and more severely colonised by P aeruginosa,2 we decided to evaluate the correlation between ΔF508 homozygosity and B cepacia colonisation in cystic fibrosis.

All patients attending the Verona Cystic Fibrosis Centre between November 1991 and November 1994 were examined for B cepacia airway infection, and most of them genetically tested for ΔF508. B cepacia colonisation was considered chronic after at least two positive cultures a year for one or more years, or three consecutive positive cultures over a four to 12 month period.4 Fourteen out of the 40 (35%) chronic and 84 out of 202 (42%) non-colonised patients were homoyzotes for ΔF508. A significant difference in frequencies was shown between subjects chronically colonised by B cepacia who were homozygous for ΔF508 and patients chronically colonised carrying other genotypes (2×2 contingency table analysed by Fisher's exact test; p value=0.0123; odds ratio=2.468; confidence interval=1.236 to 4.927). Clinical evaluation of patients showed in ΔF508 homozygotes no signs of a more severe pulmonary disease, which could have explained the different colonisation rates, and the open comparison of two investigations showed no significant difference (forced expiratory volume in one second p value=0.8985; x ray score p value=0.7277). Furthermore we could not find any difference in early colonisation by B cepacia in ΔF508 homozygotes: there is no significant age difference at B cepacia first isolation in the two genotype groups (two tailed p value=0.2876).

Our data show that homozygous patients show a higher prevalence of B cepacia chronic colonisation, and have more than double the chance of colonisation, compared with those carrying different genotypes. Surely environ- mental factors, such as housing conditions, diet, have an impact on the colonisation rate, but apparently genotype is involved as well. If further studies in different populations and environments confirm these results, this new marker for chronic colonisation by B cepacia could be extensively included in studies on the genotype/phenotype correlation, considering also mutations less frequent than ΔF508.

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Birth weight in phenylketonuria

EDITOR—The report by Verkerk et al of relatively low birth weight in Dutch infants with phenylketonuria1 agrees well with our earlier finding of low birth weights in phenyl- ketonuria in Ireland and west Scotland.2 In agreement with other workers in the USA,3 the UK,4 and Poland,5 we found that in our total sample of 62 infants with phenylketonuria and 53 unaffected siblings, and within families, there was no significant difference between infants with phenylketonuria and their unaffected siblings either in their unadjusted birth weights or in their birth weights adjusted for factors that affect this quantity. Moreover, the 115 birth weights in the combined sample lay on a normal distribution curve with no evidence of a bimodal or trimodal distribution. However, in agreement with other workers in the USA,4 our data showed that birth weights of infants with phenylketonuria and their siblings (p≤0.02) for the adjusted birth weights the difference between the means was 121 g less than the mean for a randomly selected sample of 819 control infants in the same hospitals in the same years as the infants with phenylketonuria and their siblings. This difference could be explained that the mean birth weight of infants with phenylketonuria and their unaffected siblings was significantly lower than the mean birth weight of the control group. Furthermore, we are not in agreement with this theory.3 The study by Saugstad found that mean birth weight of 49 infants with phenylketonuria was 356 g lower than mean birth weight of their healthy siblings. After adjustment for differences in gestational age, the discrepancy in birth weight even increased to 530 g. I am therefore not yet convinced by the explanation offered by Woolf and Crockett. Perhaps a quantitative review of the literature (meta-analysis) may provide more information on this subject.


Diagnosis of mycobacterial lymphadenopathy

EDITOR—Clark and colleagues recently reviewed 17 cases of non-tuberculous mycobacterium (NTM) lymphadenopathy.1 As they conclude, it is important to differentiate between mycobacterium tuberculosis from NTM infection, although this is not always possible on clinical grounds alone. If mycobacterial infection is suspected, then definitive species group identification is essential, as the surgically resected material may be for further analysis. If no NTM and M tuberculosis are very different.

Skin testing is a useful first line investiga- tion of infective lymphadenopathy.2 Mycobacterium avium purified protein deriv- ative is available commercially in Australia (but not in the UK). Cross reactivity between this and the human tuberculosis test is common. However, if a NTM is the causative organism then the M avium hypersensitivity reaction is usually significantly larger than that caused by...
the human Mantoux.2 If skin testing is equivocal, or in cases of infection involving potentially vulnerable sites (such as the mastoid), when adjutant antimycobacterial drug treatment is given, then it may be helpful to have the surgery designed by Brown at el.1

Oscillations between (mean overnight temperature oscillations is a consistent characteristic of early infancy, occurring both in health and illness.

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Compliance with growth hormone treatment— are they getting it?

EDITOR—We previously reported that only 48-9% of our patients treated with recombinant human growth hormone (rhGH) complied in all aspects.

1 We identified patient education and rhGH reconstitution as the major contributory factors, and, as a consequence, offer patients a choice of rhGH preparation appropriate to their needs and a hospital based clinical nurse specialist to train them in its use at home. We have now administered the same questionnaire to a new group of patients.

Patients attending over a two month period were asked to complete a questionnaire if they were receiving rhGH. The questionnaire designed to assess level of understanding and compliance with treatment was accepted by 177 patients. Altogether 105 (59%) (group 1) had started treatment before the change in policy; 64 (36%) (group 2) had been trained by a clinical nurse specialist at home. Eighty one per cent of patients in group 2 had a good, 10% an adequate, and 9% a poor understanding of the therapeutic regimen compared with 50%, 34%, 15% respectively before (p<0.01). Patients in group 1, who had started rhGH before the change in policy failed to improve their understanding of the therapeutic regimen despite being seen at regular intervals at hospital visits by a clinical nurse specialist.

Compliance was assessed by questions designed to uncover the number of missed injections during a three month period. Fifty eight per cent of patients in group 1 complied with all aspects of their treatment, which was not significantly different from our previous experience; 84% of patients in group 2 complied with all aspects of their treatment (p<0.001).

Compliance in children prescribed rhGH treatment has improved considerably. Initial training of the patient and family at home appears to be the most important element in achieving compliance.

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Morbidity from excessive intake of high energy fluids: the 'squash drinking syndrome'

EDITOR—Following the article by Hourihan and Rolles on the 'squash drinking syndrome'1 we would like to take the opportunity to remind readers that excessive squash drinking can rarely be associated with serious side effects than failure to thrive.2 Recently a 22 month old girl presented here with a generalised afebrile convulsion and hyponaesthesia. She had previously been recognised elsewhere as failing to thrive, with her weight lying below the third centile. Her weight at presentation here was 8-7 kg. On questioning she was found to be drinking approximately two litres of squash a day, and at night slept with a large jug of juice at the bedside.

Investigation revealed a serum sodium concentration of 114 mmol/l, potassium 4-0 mmol/l, urea 2-9 mmol/l, creatinine 54 mmol/l, glucose 5-2 mmol/l, and calcium 2-34 mmol/l with a simultaneous urinary sodium of 19 mmol/l and urinary osmolality of 128 mmol/kg. Serum sodium rose to normal concentrations simply with fluid restriction to normal fluid requirements of around one litre a day. A water deprivation test subsequently revealed normal renal concentrating ability excluding diabetes insipidus as a cause for her polydipsia. The parents were advised to restrict squash consumption.

There have been no further fits on follow up over one year. Squash consumption has varied, but a normal serum sodium has been maintained. However, weight gain has been better at those times when squash consumption has been less excessive.

We agree with Hourihan and Rolles that excessive squash consumption is an important cause of failure to thrive. Additionally the possibility of water intoxication, with all its complications, should be considered if squash consumption is excessive.

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1 Hourihan J0B, Rolles CJ. Morbidity from excessive intake of high energy fluids: the 'squash drinking syndrome'. Arch Dis Child 1995; 72: 141-3.

The art of communication with children

EDITOR— The need to communicate well with children and their parents is fundamental to paediatric practice. Most of us see our own children, in our role as a doctor, but rarely do we get an opportunity to join them as normal adults with whom they can play and frankly discuss their problems. One way I learnt to understand children was to spend some weekends camping with the Woodcraft Folk, a recognised educational charity for children and young people. These camps are organised as