The effect of immunoglobulin on Vβ repertoire is uncertain. 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 outside paediatric centres, 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 patients 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.

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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 earlier colonised by *P aeruginosa*,[2 3] 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%) chronically colonised and 84 out of the 469 (22%) non-colonised patients were homozygotes 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 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 an evident 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 ΔF508 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 environmental factors influence the colonisation rate, but apparently genotype is involved as well. If further studies in different populations and environments confirmed these results, the determination of 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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1 Anonymous. Pseudomonas cepacia – more than a harmless commensal? [Editorial.] *Lancet* 1992; **339:** 1385–6.
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Birth weight in phenylketonuria

EDITOR,—The report by Verkerk *et al* of relatively low birth weight in Dutch infants with phenylketonuria[1] agrees well with our earlier finding of low birth weights in phenylketonuria 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, the mean unadjusted birth weight of the combined sample was 121 g less than the mean for a randomly selected sample of 819 control infants born in the same hospitals in the same years as the infants with phenylketonuria and their siblings ($p \leq 0{\cdot}02$); for the adjusted birth weights the difference between the means was 107 g ($p < 0{\cdot}02$). We concluded that, as the reduction in birth weight was the same for both infants with phenylketonuria and their unaffected siblings, the lower birth weight was not related to the pathogenesis of phenylketonuria or to the fetal genotype. It appears that the reduction in birth weight was a reflection of the maternal genotype affecting the intrauterine environment and was a previously unknown effect of the phenylketonuria gene in single dose.

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1 Verkerk PH, van Spronsen FJ, Smit GPA, Sengers RCA. Impaired prenatal and postnatal growth in Dutch patients with phenylketonuria. *Arch Dis Child* 1994; **71:** 114–8.
2 Crockett DJ, Woolf LI, McBean MS, Woolf FM, Cahalane SF. Birth weight and pathogenesis in phenylketonuria. *Int J Neurosci* 1990; **54:** 259–66.
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Dr Verkerk comments:

Woolf and Crockett hypothesise that the reduced birth weight in phenylketonuria as found in some studies may be the result of maternal genotype affecting the intrauterine environment. This interesting hypothesis is based on their own findings and on a qualitative review of the literature. According to their theory no differences should be found in birth weight between infants with phenylketonuria and their healthy siblings. However, the findings of the first study on the relationship between phenylketonuria and birth weight are not in agreement with this theory.[1] The study by Saugstad found that mean birth weight of 49 infants with phenylketonuria was 356 g lower than mean birth weight of their 86 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.

1 Saugstad LF. Birthweights in children with phenylketonuria and in their siblings. *Lancet* 1972; i: 809–13.

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 *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 surgical management, and the antibiotic sensitivities, of NTM and *M tuberculosis* are very different.

Skin testing is a useful first line investigation of infective lymphadenopathy.[2 3] *Mycobacterium avian* purified protein derivative is available commercially in Australia (but not in the UK). Cross reactivity between this and the human Mantoux test is common. However, if a NTM is the causative organism, then the *M avian* hypersensitivity reaction is usually significantly larger than that caused by