used for our patients, may cure some boys. Diagnosis of residual disease in the testis by routine biopsy before completion of chemotherapy may permit detection of those boys who require this treatment and perhaps improve the overall outlook for boys with leukaemia.

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Decreased serum tuftsin concentrations in sickle cell disease

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SUMMARY The serum concentrations of the phagocytosis-stimulating peptide, tuftsin, were determined by radioimmunoassay in 21 patients with sickle cell disease and in 12 healthy controls. The mean serum tuftsin concentration was significantly lower in patients with haemoglobin SS disease (154.3 ± 35.1 ng/ml; 308.6 ± 70.2 nmol/l, P<0.01) and in patients with haemoglobin SC and CC disease (180.9 ± 42.7 ng/ml; 361.8 ± 85.4 nmol/l, P<0.05) than in healthy controls (228.7 ± 46.7 ng/ml; 457.4 ± 93.4 nmol/l). Tuftsin deficiency is an indicator of splenic hypofunction and may contribute to the increased susceptibility of patients with sickle cell disease to severe infection.

Najjar and his associates described a cytophilic gammaglobulin, leucokinin, that stimulates phagocytosis by polymorphonuclear leucocytes and macrophages. The full biological activity of leucokinin resides in a tetrapeptide (Thr-Lys-Pro-Arg) named tuftsin, which is covalently bound to the heavy chain of the gammaglobulin.1-5 It is believed that the spleen is actively involved in either the generation or the release, or both, of the peptide from its carrier heavy chain as serum tuftsin concentrations are very low after elective splenectomy, and in functional asplenia due to infiltrative disease or infarction of that organ.6,7 Functional asplenia contributes to the increased susceptibility of patients with sickle cell disease (SCD) to severe infections.6 We have, therefore measured the serum tuftsin concentrations in patients with SCD and compared the values with those found in healthy controls.

Patients and methods

21 patients (14 with homozygous SS disease, 5 with haemoglobin SS disease, and 2 with haemoglobin CC disease) and 12 healthy age-matched controls were studied; the patients and controls were aged between 3 and 21 years. The patients were neither infected nor in crisis at the time or during the 6-month period preceding blood collection, and none of them had been transfused in the 6 months before the study. In 7 of the patients with haemoglobin SS disease,
Howell-Jolly bodies were observed on peripheral blood smears. Consent was obtained from patients or parents; the sera were collected in St Petersburg, Florida, stored at −20°C, and shipped frozen to Israel for assay. Serum tuftsin was determined by radioimmunoassay as previously described.5

Results

The mean (±SD) serum tuftsin concentration was significantly lower (Figure) in patients with haemoglobin SS disease (154.3 ± 35.1 ng/ml, P < 0.01) and in patients with haemoglobin SC and haemoglobin CC disease (180.9 ± 42.7 ng/ml, P < 0.05) than in healthy controls (228.7 ± 46.7 ng/ml). The mean (±SD) serum tuftsin concentration in haemoglobin SS disease patients with Howell-Jolly bodies in their peripheral blood smears was 147 ± 44.3 ng/ml (294.0 ± 88.6 nmol/l).

Discussion

Overwhelming infections contribute significantly to mortality and morbidity in patients with SCD.7 Although several abnormalities of host defence have been shown in SCD patients,7–8 functional and eventually anatomic asplenia is likely to be an important factor in the pathogenesis of the increased susceptibility of these patients to systemic infections.6–7 The present data indicate that measurement of serum tuftsin levels may well be of value as an indicator of the presence of a defective splenic function in SCD patients. Indeed, the lowest mean serum tuftsin concentration was recorded in patients with haemoglobin SS disease. In half of these patients, Howell-Jolly bodies, a reliable indicator of the presence of splenic hypofunction,6 were observed on peripheral blood smears. Our findings are in good agreement with those of Najjar who measured serum tuftsin activity in SCD patients using the polymorphonuclear leucocytes-bacteria phagocytosis system.4 Severe infections often occur in patients with familial tuftsin deficiency.1 4 It is likely, therefore, that tuftsin deficiency, in addition to its importance as an indicator of splenic hypofunction, impairs the phagocytic function of the blood polymorphonuclear leucocytes and of macrophages in SCD patients and contributes to the increased susceptibility of these patients to severe infections.

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