Sir, We completely agree with Dr E N Harris that carrying out clotting assays to detect lupus anticoagulant is more difficult than an enzyme linked immunosorbent assay (ELISA) for anticardiolipin antibodies. Specialised laboratories are necessary, coagulation tests are time consuming, and improper preparation and handling of plasma samples strongly influence the results of lupus anticoagulant assays.

The goal of our article was not to propose the determination of lupus anticoagulant only and to sweep aside the anticardiolipin antibody determination but to emphasise that testing anticardiolipin antibodies alone is not enough to identify patients with increased risk of thrombotic complications. From the values we found for sensitivity and specificity with lupus anticoagulant and anticardiolipin assays we conclude that in patients with systemic lupus erythematosus (SLE) the lupus anticoagulant assays correlate better with the presence of a history of thrombotic complications. We did not claim that the anticardiolipin antibody ELISA is not a useful predictor for thrombosis, fatal loss, or thrombocytopenia, but that lupus anticoagulant tests are better predictors. Confirmation of our results by studies in other laboratories is necessary, however. Therefore we endorse the last sentence of Dr Harris's letter completely that 'with the knowledge we have today both tests have to be performed'.

As mentioned by Dr Harris our studies differ with respect to the percentage of lupus patients included and the selection of patients. We evaluated 111 consecutively seen lupus patients who were unselected apart from the fact that they were seen at a university hospital. Dr Harris selected on the availability of clinical notes 121 patients (76% with SLE) from a total of 300 patients with various autoimmune diseases (60% with SLE) who had been screened for IgG anticardiolipin antibody levels.

In our paper we incorrectly used the term exchange of sera when we referred to the freeze dried samples which were kindly provided by Dr Harris for participation in the International Anticardiolipin Standardisation Workshop (April 1986). Although we introduced some modifications, our assay appeared to be valid. The limits we gave in units for low, medium, and high levels were obtained by transformation of the known anticardiolipin antibody concentrations (in μg/ml) in the provided samples.

We agree that apart from standardisation of the anticardiolipin antibody ELISA an international consensus with respect to the definition of the lupus anticoagulant is also urgently needed. Unless we can be sure that only results that are obtained with valid assays performed on proper samples are presented, methodological differences remain a possible cause for conflicting reports. Therefore, we welcome and strongly support the tremendous efforts made by Dr Harris and coworkers to reach international uniformity.

Cyclosporin for dermatomyositis?

Sir, Cyclosporin was successfully used to treat adult dermatomyositis by Zabel et al. More recently, this drug was used to treat a patient with severe adult dermatomyositis with good effect. Here we present a case of severe adult dermatomyositis unresponsive to both cyclosporin and conventional treatment. In addition to redressing the balance, our case highlights other important aspects of this interesting disease.

A 67 year old woman was admitted for investigation of weakness and malaise of four months' duration. She had a heliotrope periorbital rash, marked proximal muscle weakness and wasting, Gottron's papules, and nail fold vasculitis. She was mildly anaemic, with raised inflammatory indices (Fig. 1). Electromyography indicated myositis. Left quadriceps muscle biopsy showed perifascicular fibre damage and a perivascular lymphocytic infiltrate. Serum creatine phosphokinase (CPK) was normal, as it is in 4-18% of patients with dermatomyositis at presentation, but lactate dehydrogenase was mildly raised, and she met the criteria for 'definite' dermatomyositis. Antinuclear antibody titre was weakly positive (1/40) as was smooth muscle antibody, but rheumatoid factor and anti-Jo-1-antibody were absent from her serum. Complement factors C3 and C4 were normal.

Prednisolone and azathioprine had a favourable but temporary effect after two months, when her steroid requirement fell, but she relapsed after an infection and subsequently failed to respond to increased corticosteroid

![Fig. 1 Clinical course of patient with adult dermatomyositis. ESR=erythrocyte sedimentation rate; CPK=creatine phosphokinase; CRP=C reactive protein.](http://ard.bmj.com/)

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and pulse methylprednisolone. In addition, she developed physical, spirometric, and radiological signs of interstitial lung disease.

Cyclosporin was added to her regimen at 7.5 mg/kg a day orally, resulting in trough concentrations within the therapeutic range (100–250 ng/ml). The rash and muscle weakness worsened, however, inflammatory indices remained high, and she died from respiratory failure secondary to rapidly progressive interstitial lung disease (Fig. 2) despite four weeks' treatment with cyclosporin.

Cyclosporin may be a logical choice for dermatomyositis as it inhibits interleukin 2 production, thus blocking the development of cytotoxic T lymphocytes and the proliferation of T helper cells; T cell mediated myolysis is thought to play an important part in dermatomyositis. Nevertheless, the reports of dramatic successes with cyclosporin need to be tempered by its failure to help with our patient.

It has been suggested that dermatomyositis without an increase in CPK is a poor prognostic sign. Of 38 patients seen at the University of Michigan, seven had a normal CPK, five of whom died within a year—three from an associated malignancy and two from severe interstitial lung disease. Interestingly, of Takizawa's 14 cases, nine developed interstitial lung disease, which was rapidly fatal in six cases. All these patients had low or normal CPK concentrations. The subgroup of patients with dermatomyositis and normal CPK concentrations, therefore, appears to be more likely to develop severe interstitial lung disease and to be more resistant to available treatment.

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Fig. 2  Chest x ray of patient with dermatomyositis and interstitial lung disease.

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