after an uneventful lower segment caesarean section. She had a tachycardia, an inflamed wound, and some vulval swelling. Blood was taken for culture and intravenous cephaloridine and metronidazole were started. She deteriorated rapidly and became pale and cyanosed with an unrecordable blood pressure. Her abdomen was distended and there were no bowel sounds. She was resuscitated with intravenous fluids, oxygen, and hydrocortisone (100 mg), and transferred to the Western Infirmary, Glasgow.

On arrival her wound was tender, dusky, and warm, with blistering of the skin. She again became shocked with a blood pressure of 90/60 mm Hg, intensive vasoconstriction, and oliguria. Her core temperature was 39·5°C. The cephaloridine was stopped, and cefotaxime and benzylpenicillin added to the metronidazole. At subsequent laparotomy the internal organs were found to be intact and no intra-abdominal collections identified. A total of 3·5 l of fluid was given together with further hydrocortisone (100 mg). In the intensive therapy unit she was given a further 3 l of fluid and over the next eight hours she regained normal haemodynamic values.

Pus collected from the wound at laparotomy contained numerous polymorphs and chaining Gram positive cocci. Intravenous benzylpenicillin (2 MU four hourly) was continued. Subsequently a *Streptococcus pyogenes* (β haemolytic Lancefield group A) was isolated. She was extubated and haemodynamically stable within 24 hours but remained pyrexial with cellulitis around her wound. Within 48 hours this covered her lower abdomen and extended down her left thigh. The benzylpenicillin was increased to 4 MU four hourly. Over the next six days her temperature returned to normal, and the cellulitis resolved. An area of necrosis below the wound subsequently required grafting.

This case shows the severity of the haemodynamic disturbance associated with *Str pyogenes*. Appreciation of this act should lead to immediate and aggressive resuscitation with fluids, oxygen, and benzylpenicillin. Myocardial depression is not specific to streptococcal infection and may be observed in many patients in whom shock is severe and prolonged.[1]

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Wilson's disease

SIR,—Dr David Parkes suggests that there may be between 250 and 1500 cases of Wilson's disease in Britain and that 1000 of these cases may be unrecognised (21 April, p 1180). In their recent monograph on the disease Dr I H Scheinberg and Dr I Sternlieb[1] summarised evidence for the prevalence of the disease and hence the gene frequency in populations where cosanguinity has not biased the results.[1] With increasing awareness of the disease and its pleomorphic manifestations together with increasingly sophisticated diagnostic techniques the rate of accurate diagnosis has increased.

In 1953 Bearn calculated an incidence of one in a million with a gene frequency of one in 500 of the population.[2] By 1968 the estimate had risen to one in 200 000 with a carrier rate of one in 224.[3] By 1979 the figure had reached 30 in a million with a gene frequency of 0·53%,[4] which led me to comment that if this figure were to be transposed to this country "there may be as many as 1600 cases of Wilson's disease in Britain. As the majority of these are probably not correctly diagnosed they will therefore die of their disease; this suggests that our diagnostic failures amount to something approaching a minor medical disaster."[5] This statement suggested some scepticism of the figures.

East Anglia, however, may well be ideal for estimating the incidence of the disease in Britain. We can count accurately diagnosed cases and need not rely on statistics gathered from several centres or from death certificates. The population is stable and largely rural, and the university hospital at Cambridge drains most of the population—apart from where Essex verges on London.

The area considered is greater than the administrative East Anglian region of Cambridgeshire, Norfolk, and Suffolk. The population under consideration was in the 1981 census 3 376 981 and has probably not varied much subsequently. Twenty four patients are known, and the diagnosis has been confirmed in all cases. I have broken down the patients into the cohorts used in the 1981 census figures, which is necessary as the disease does not present in the youngest age group and could not be expected in the older age groups (table). Patients over 45 would probably have been missed because of diagnostic shortcomings.

Taking the peak incidence for presentation as the two cohorts 5-15 and 16-24 we find 20 patients in a population of 998 720, and there were 23 patients in the age groups 16-24 and 25-34 among 933 878 people. Both these sets of figures are not so far from the 29 in a million of the East Germany survey.[4] Some patients may have died or may still remain undiagnosed, while some will have been referred elsewhere. Thus the figures are an underestimate. No consanguinity was recorded to bias the figures.

If the incidence of Wilson's disease for East Anglia is translated to the whole country there must indeed be more than 1000 patients, most of whom presumably remain undiagnosed. It would be interesting to know how many could be brought to light by a detailed survey in paediatric, neurological, and gastroenterological clinics among patients aged 5 to 25 years. In addition a search of psychiatric clinics for a slightly older age group might produce a few additional cases.

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Age distribution of cases of Wilson's disease in East Anglia

Age	0-4	5-15	16-24	25-34	35-44	Total
Population	207 584	557 084	441 672	492 206	421 743	3 376 981
No of patients (age in 1981)	0	1	9	13	1	24
No of patients (age at onset)	0	10	10	4	0	24

SIR,—Dr D Parkes emphasised the importance of diagnosing Wilson's disease in view of the greatly improved prognosis with correct treatment (21 April, p 1180). We draw attention to a mode of presentation he did not discuss.

A previously well 14 year old Indian girl with no relevant family history presented with severe acute haemolysis (haemoglobin concentration 6·39 g/dl, reticulocytes 14%). Examination showed severe jaundice but no stigmata of chronic liver disease. There was no neuropsychiatric disturbance. A peripheral blood film showed basophilic stippling, anisocytosis, and occasional erythroblasts. Bilirubin concentration was 40 μmol/l (2·3 mg/100 ml) (normal < 15 μmol/l (0·9 mg/100 ml)), alanine transaminase concentration was 124 IU/l (normal 2-20 IU/l), and serum albumin concentration was 25 g/l (normal 37-53 g/l). Drug induced haemolysis, viral infection, malaria, autoimmune disease, and red cell enzymopathy were excluded. Wilson's disease was diagnosed by finding bilateral Kayser-Fleischer rings and urinary copper excretion of 5·1 and 28·3 μmol/24 hours (0·3 and 1·7 mg/24 hours) (normal < 1 μmol 0·06 mg/24 hours). Serum copper and caeruloplasmin concentrations were unexpectedly normal. This was ascribed to increased release during hepatic necrosis. One further haemolytic crisis has occurred. Two asymptomatic siblings were shown to have Wilson's disease.

Acute recurrent haemolysis is a recognised feature of Wilson's disease but an uncommon mode of presentation.[1] It may be due to the toxic oxidant effect of the massive release of copper from the liver.[2] Wilson's disease should be considered in patients presenting with acute haemolysis, particularly if liver dysfunction is present.

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Medical education

SIR,—It is true that undergraduate medical training instructs rather than educates and both creates and rewards tunnel vision. It is probably true that new medical graduates are less equipped than ever before to deal with the problems in acute medicine. I doubt, however, that Dr J Horder and others' proposal for yet another period of structured professional training with formal assessment is the solution (19 May, p 1507).

My preregistration year is almost over. Most of the problems which confuse and haunt me could never be expressed in any textbook. I have mended oscilloscopes, advised male homosexuals on their technique, hitch hiked back from peripheral clinics when strikes affected transport, rummaged through patients' lockers in search of alcohol, and defended myself against physical and phychological assault. The skills which I am coming to acquire are so intangible that they disappear when I try to define them. The best and probably the only way to gain confidence and skill is to be thrown in at the deep end. One can no longer, however, simply "go and look it up" after the event. Today's medical problems demand a delicate blend of intellectual, emotional, technical, and interpersonal skills. Dr Horder and others have recognised this, but I find their proposed solution inappropriate.

From the shop floor, I see two obstacles to