normal haemoglobin and in the anaemic patients reported here and elsewhere[3] argues against a causative role for glucagon in the anaemia of end-stage renal disease.

We are grateful to Dr Nina C Mendoza for allowing us to study patients under her care.

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Shwachman's syndrome and acute lymphoblastic leukaemia

The association of hypoplasia of the exocrine pancreas and bone marrow dysfunction has been recognised since 1964.[1] A wide range of haematological disorders has been described, but we report a new association—acute lymphoblastic leukaemia.

Case report

A 14-year-old boy presented with a two-month history of recurrent chest infections. Despite antibiotic treatment he had deteriorated with increasing weight loss, pallor, and breathlessness. His medical history was unremarkable with no symptoms suggestive of a bowel disorder, although his sister had died aged 9 years of suspected coeliac disease. On admission he was pale and small for his age; his abdomen was protruberant with ascites; and his ankles swollen. He had generalised lymphadenopathy and gross hepatosplenomegaly. The results of initial investigations showed haemoglobin 5·2 g/dl, total white cell count 525×10^9/l with neutrophils 2% and blast cells 98%. Plasma concentration of sodium was 134 mmol/l (134 mEq/l), potassium 3·4 mmol/l (3·4 mEq/l), urea 43 mmol/l (257 mg/100 ml), uric acid 0·3 mmol/l (5·0 mg/100 ml), bilirubin 11 μmol/l (0·6 mg/100 ml). Serum albumin was 38 g/l and globulins 27 g/l; immunoglobulin concentrations were normal. Chest *x*-ray films showed chronic infective changes but no mediastinal widening. Bone marrow aspirate was hypercellular because of a dense infiltrate of blast cells which showed "block" periodic acid Shiff positivity but no myeloperoxidase activity. The abnormal cells also reacted with an antibody raised against leukaemic lymphoblasts.

Acute lymphoblastic leukaemia was diagnosed and he was entered into the appropriate MRC trial. Initial treatment was with intravenous vincristine 1·5 mg weekly and oral prednisolone 40 mg daily. The primitive cells disappeared from the peripheral blood within six days. During the ensuing pancytopenia he developed epistaxis and gastrointestinal bleeding necessitating frequent transfusions of red cells and platelets. He also developed a severe klebsiella pneumonia, which persisted despite appropriate antibiotic treatment. Diuretics caused a rapid disappearance of the peripheral oedema but the ascites increased. In the later stages of his illness he also developed persistent offensive diarrhoea. He died six weeks after treatment was started, still in a stage of profound pancytopenia. Bone marrow examination five days before death showed severe hypoplasia of the normal marrow tissue but no evidence of active leukaemia.

Postmortem examination showed consolidation of both lungs and evidence of massive gastrointestinal haemorrhage. The liver and spleen were enlarged, the liver histologically showing increased fibrosis. There was no sign of residual leukaemia in the liver or any other organ examined. The most striking histological abnormality was the total absence of exocrine pancreatic tissue, the islet cells being surrounded by fatty tissue with a few dilated ducts.

Comment

Though rare, hypoplasia of the exocrine pancreas is said to be the most common pancreatic disorder of childhood once cystic fibrosis has been excluded.[2] Haematological abnormalities described in association with this condition include anaemia, neutropenia, and thrombocytopenia, and bone marrow examinations have shown hypoplasia or a maturation arrest in one or more cell lines.[1 2] Only two cases have been described in association with leukaemia: one monocytic[3] and the other myeloblastic.[4] The association with lymphoblastic leukaemia is more difficult to explain. Lymphoreticular malignancy in patients with immunoglobulin deficiency is well recognised and some cases of Shwachman's syndrome have recently been described with low immunoglobulin concentrations.[5] In our case, however, the immunoglobulin concentrations were normal.

Shwachman's syndrome is often familial[1 2] and interestingly our patient's sister had died with a malabsorption syndrome. There was no evidence from the biopsy specimen of coeliac disease and she failed to respond to a gluten-free diet, so probably she also had pancreatic hypoplasia.

Requests for reprints should be addressed to Dr J S Lilleyman.

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Deaths in cold water

Most efforts to reduce the heavy loss of life in water accidents are directed to improving the ability to swim, but a recent Home Office report of the Working Party on Water Safety gives further emphasis to the fact that many of the people who die can swim. Of those whose swimming ability was known, most of those in the 15- to 35-year-old age group who died could swim, while most of those who died in the sea at any age were either good or very good swimmers. It is still not always realised that even summer waters around Britain are cold enough to kill during prolonged immersion, and that in winter and spring the high viscosity and other effects of water near 0°C can cause excellent swimmers to drown in as little as 90 seconds.[1] Advice about simple precautions to be taken in cold water, which can prevent or greatly reduce these hazards, was widely distributed by water sport clubs and schools after a conference organised by the Central Council for Physical Recreation in 1971. We have now analysed the Registrar General's Statistical Reviews to see whether this action was followed by any major changes in the accident rates.

Statistical analysis

The table shows the deaths in different age groups during the four years up to and the four years after 1971. The deaths are those registered as definitely accidental and due to sinking of boats, falling from boats, and other drownings and submersions (categories E830, E832, and E910) for England and Wales only. They show that the number of deaths fell significantly ($P < 0.001$ by the χ^2 test), in the 5- to 14-year-old age group only. The total deaths in the group were 549 in 1968-71 (18·3/million) and 438 in 1972-5 (13·9/million). This represents a reduction of 111 deaths or a little more if the

Accidental deaths in water

Age	1968–71		1972–5	
	Total deaths	Deaths/ million	Total deaths	Deaths/ million
0–4	400	24·7	337	23·1
5–14	549*	18·3	438*	13·9
15–44	879	11·5	864	11·3
45–64	346	7·3	345	7·4
65+	233	9·3	268	9·9

*Figures for 1968-71 differ from those for 1972-5 $P < 0.001$.