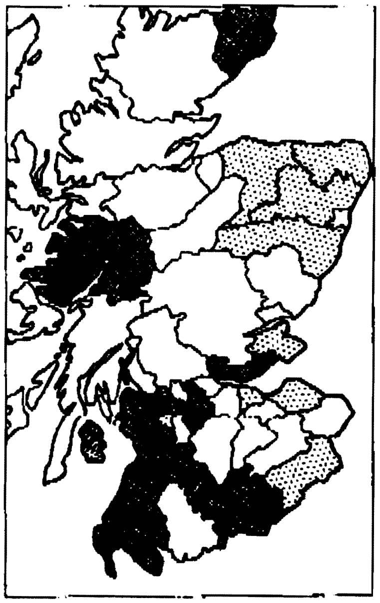
982.



IHD in Scotland 1979–83.

Stippled areas – low SMR.
Shaded areas – high SMR.

Scotland, and the highest (200) was in Whithorn in the south of the country; for 1969–73, the lowest SMR (29) was in Inverary (central west) and the highest (192) was in Doune, in central Scotland; for 1979–83 the lowest SMR (77) was in Eastwood (central west) and the highest (127) was in Cumnock and Doon Valley, in the south-west of the country.

The geographical distribution of mortality from IHD has been remarkably consistent since at least 1959, with high SMRs found frequently in the west and south of Scotland, and low SMRs found predominantly in the north and east (figure). This distribution by latitude within Scotland, therefore, does not follow that shown on the international scale.

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LIVER TRANSPLANTATION IN CYSTIC FIBROSIS

SIR,—Hitherto hepatobiliary complications in cystic fibrosis (CF) have been uncommon, probably because patients with CF have until lately not lived long enough for these problems to manifest. However, as treatment for the pulmonary manifestations of CF have increased life expectancy so hepatobiliary complications have been noted more often.[1,2] Liver transplantation is now the treatment of choice for end-stage liver disease, and more patients with CF and liver failure will be referred for transplantation. The results in nine CF patients so treated are reported here.

In the eight years ending on Dec 31, 1988, the 1788 liver transplants done at this centre on 1377 patients included 11 in 9 patients with CF (table). 5 patients were adults and 4 were children. All had end-stage liver disease. In one child (case D) CF was diagnosed after liver transplantation. The preoperative diagnosis had been biliary atresia but when pulmonary problems persisted sweat tests were done and were diagnostic of CF. Postoperative management did not differ much from that for other liver transplant recipients, except for the special attention paid to pulmonary care and the need for prolonged intravenous cyclosporin (because of sluggish gastrointestinal function) and for high doses of oral cyclosporin. Supplemental pancreatic enzymes were given postoperatively once oral feedings were restarted.

POST TRANSPLANT STATUS OF 9 CASES REVIEWED

—	FVC/FEV, (both %)		Current status
	Preop	Postop	
Case			
A (23/M)	72/81	109/119	Alive 45 mo, working full time
B (30/F)	62/50	59/43	Died 27 mo
C (24/M)	105/114	110/105	Alive 36 mo, working full time
D (27 mo/F)	..	..	Alive 28 mo, in kindergarten
E (41 mo/M)	..	..	Alive 12 mo, in nursery school
F (21/M)	77/74	..	Died 2 wk
G (8/M)	50/38	109/106	Alive 7 mo, in school
H (13/F)	81/68	67/61	Alive 2 mo, recovering from Nissen fundoplication
J (18/F)	61/50	81/70	Alive 20 mo, in school

7 patients are well after 2 months to 4 years with normal liver function (table). In patient F hepatic artery thrombosis caused loss of his first liver. Retransplantation was unsuccessful. The second liver functioned well, but the patient died of pseudomonas pneumonia and respiratory failure. Another patient with normal liver function died 27 months after transplantation from an unrecognised closed-loop obstruction and gangrenous bowel. This patient (B) also had Friedreich's ataxia and mild mental retardation.

The 3 adult survivors have normal liver function and are working full time or back in school (patient J). The 4 children, including 1 who required a second liver transplant because of hepatic artery thrombosis (E), also have normal liver function. Patient H has required a Nissen fundoplication for oesophageal reflux which resulted from sclerotherapy and an oesophageal stricture.

Postoperative sweat tests were done in 2 patients and showed no change. Pulmonary function tests were done in six patients before and after transplantation. Forced vital capacity (FVC) and forced expiratory volume in 1 s (FEV_1) was improved in patients A, G, and J, essentially unchanged in patients B and C, and slightly decreased in patient H.

Examination of excised livers in all 9 cases revealed cirrhosis, bileduct proliferation, and chronic inflammatory cell infiltrate. Eosinophilic concretions pathognomonic of CF liver disease were present in 3 specimens, and the findings in the other livers were consistent with the diagnosis of CF.[3,4]

Although the limiting factor in patient survival with CF is usually respiratory disease, our patients had only mild-to-moderate pulmonary disease while progressive liver disease had reached end stages. In such patients, palliative measures (eg, portocaval shunt or sclerotherapy) have been used.

In the only detailed case-report of liver transplantation in CF the patient died of infection after 45 days.[5] Our series shows that good results are possible. The improvement in FVC can be explained by the removal of ascites, but there is no clear explanation for the improvement in FEV_1. It is hard to believe that any somatic correction of the inborn error occurred since sweat test results were unaltered in patients tested postoperatively.

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