## Scottish Paediatric Society

At the Summer Meeting held at Raigmore Hospital, Inverness, on Friday 30 May 1980, the President, Dr W H Galloway, was in the chair.

Post-perinatal infant mortality in Glasgow (1979). G C Arneil, A Harvie, A A M Gibson, W J A Patrick, H T McIntosh, and H Brooke. Royal Hospital for Sick Children, Glasgow.

A detailed study of post-perinatal deaths occurring in infants aged 7-365 days in 1979 has been conducted in the Greater Glasgow Health Board Area. The purpose was to establish the causes of such deaths and to classify them. The possible effects of improved perinatal care in postponing deaths beyond the 7th day of life, thus increasing post-perinatal mortality, were also examined. The study, carried out with the co-operation of the Procurator-Fiscal, was divided into several parts—a review of maternity and infant case records, an interview with the parents, and correlation with necropsy findings. The 77 deaths could be classified into three groups: 38 determined at the time of birth ('late perinatal' deaths); 3 caused by accident or known illness; and 36 presenting as cot deaths (sudden infant death syndrome). There were 13 339 live births, giving a post-perinatal infant mortality rate of 5.8 per 1000 live births from a total infant mortality of 13.3. Those post-perinatal deaths determined by prematurity or developmental abnormality were analysed together with the perinatal deaths for the area to give a more accurate record of total 'perinatally determined' mortality than had been possible previously.

Renal dialysis and transplantation in childhood—7 years' experience. A V Murphy, C S Nelson, and J McDonald. Royal Hospital for Sick Children, Glasgow.

In 1972 facilities became available at the Royal Hospital for Sick Children, Glasgow for the treatment of children with end-stage renal disease, and 28 patients (13 girls and 15 boys) have been treated. Their age range was from 3 to 15 years. 16 (57%) had been referred from other centres. Initial treatment in 25 was by intermittent haemodialysis, 12 families being subsequently successfully trained for home haemodialysis. One patient was treated by

intermittent peritoneal dialysis, and 2 by continuous ambulatory peritoneal dialysis (CAPD). Seven transplants had been performed, 2 from live donors. Of the 28 patients taken on since the start of the programme, 7 were being managed on home haemodialysis, 6 on hospital haemodialysis, and 2 on CAPD; 3 had functioning renal transplants. Four had been transferred to other units, and 6 had died. Of 17 school-age patients, 13 were attending school with attendance records above 70%, 2 had poor attendance records, and 2 were attending an adolescent guidance centre.

Gluten challenge in coeliac disease in the west of Scotland. R B Thomson, J F B Dossetor, and A A M Gibson. Royal Hospital for Sick Children, Glasgow.

A gluten challenge was given to 36 children taking a gluten-free diet since early childhood. A presumptive diagnosis of coeliac disease had been made on evidence which, in 33 cases, included a single histological examination of the jejunal mucosa. Before the challenge it was established that the ieiunal mucosal architecture was normal on glutenfree diet. Each patient then ate at least 5 g gluten daily for a period of between 6 months and 2 years before a repeat jejunal biopsy was performed. The mean age of onset of challenge was 13.1 years. Histological relapse has occurred in 22 (61%) patients. Only 3 of these complained of symptoms, and only 5 gained weight poorly during challenge. None of the 5 patients whose original diagnosis of coeliac disease was based on doubtful histological evidence (partial villous atrophy) has yet relapsed. 14 (39%) patients did not relapse, but only 2 of these had been assessed histologically after 2 years on a gluten-containing diet. Repeat histology after 2 years on the other 12 patients is awaited before a comment can be made on the value of routine gluten challenge in this population.

**Benign familial stiff baby syndrome.** J B P Stephenson. Royal Hospital for Sick Children, Glasgow.

Extreme rigidity in the newborn suggests serious central nervous system dysfunction. A new condition inherited as an autosomal dominant was described in which stiffness at birth disappears during infancy without sequelae. One family is known with 6 affected individuals out of the 10 members of two generations. Three had been admitted as neonates, each to a different special care baby nursery. All had a stiff trunk and limbs with back arching and adducted hips, suggesting tetraplegia. All had been sufficiently irritable to be given sedation; shaking episodes in one had first been presumed to be epileptic seizures. Improvement had occurred throughout the first months of life. Rigidity on handling persisted longer than stiffness and abnormal posture at rest but, examined at 2 years of age, the

children appeared to be normal. This syndrome resembles benign familial neonatal convulsions in five characteristics—namely autosomal dominant inheritance, neonatal presentation, resemblance to serious neurological disease, improvement over months, and complete recovery. In both conditions there is evidence of uncontrolled central nervous system excitation, and the suggestion is that delayed maturation of an inhibitory neuro-transmitter system may be responsible. Irrespective of the mechanism of its production, recognition of the disorder allows a confident prognosis.

## **British Paediatric Association**

## Annual meetings

1021	7-11	Anril	Vork	University
1701	7-11	AUIII	IOIK	University

1982 20-24 April Aviemore Centre, Scotland

1983 12-16 April York University

1984 10-14 April York University

1985 16–20 April York University