

(2.5–40.0) in group B, and 32.1 (12.25–50.0) in group C. 7 patients in group B and 4 patients in group C had blood xylose levels which fell within the normal range. The early effects of either gluten withdrawal or gluten challenge on sequential 1-hour blood xylose levels have so far been unhelpful in assessing gluten-induced changes.

Thus, a considerable proportion (37%) of children with untreated coeliac disease were found to have 1-hour blood xylose levels which fell within the normal range, indicating that the test is of limited value. Our findings emphasize the importance of intestinal biopsy in the diagnosis of coeliac disease.

Sodium/potassium ATPase activity in coeliac disease. F. Carswell and R. Lindsay. Department of Child Health, Royal Hospital for Sick Children, Bristol.

There is slow passage of barium through the small bowel in coeliac disease and this has been directly correlated with the intracellular potassium concentration. Increased sodium loss into the bowel lumen has been reported in both treated and untreated coeliac patients. These effects could arise because of increased sodium/potassium ATPase activity in cells in coeliac disease. Accordingly, we have examined the concentrations of sodium and potassium and the sodium/potassium ATPase activity of erythrocytes of patients with coeliac disease.

In 9 patients with untreated coeliac disease, the red blood cells contained a lower concentration of sodium and a higher concentration of potassium and there was significantly more ATPase activity in the red blood cells than in 12 control patients. In 20 coeliac patients who had been on a gluten-free diet, there were no significant differences from the normal values. Coeliac patients who had been on the gluten-free diet for less than a year had significantly higher sodium/potassium ATPase activity than patients who had been on the diet for more than 1 year.

We are currently testing the hypothesis that coeliac plasma contains a factor which increases the sodium/potassium ATPase activity of the red blood cells.

Physiological aspects and treatment of severe chronic constipation. J. O. N. Lawson and G. S. Clayden introduced by J. Scopes. St. Thomas's Hospital, London.

46 children presenting with a history of severe chronic constipation were investigated in order to exclude the diagnosis of ultra short segment Hirschsprung's disease. All had had previous unsuccessful medical treatment. Barium enema on unprepared bowel showed a megarectum, but could not exclude a very short aganglionic segment.

As part of the investigations, anorectal tonometry was employed and a characteristic trace obtained for those with 'congenital constipation' which was different from that seen in Hirschsprung's disease. Ultra short segment Hirschsprung's disease was found in 5 cases.

Physiological characteristics of 'congenital constipa-

tion' were defined as an anorectal pressure trace showing exaggerated rhythmical activity with normal inhibition responses to rectal distension but only occurring at larger volumes than normal and with greatly reduced subjective sensation of the wish to defaecate. The usual history was of constipation with overflow, from birth in 70%. In this study, 60% fell into the 'congenital constipation' group.

Children in this group responded to a vigorous anal dilatation under general anaesthetic; 42% had complete remission of symptoms and a further 38% were greatly improved.

Atypical phenylketonuria accompanied by a severe progressive neurological illness unresponsive to dietary treatment. Isabel Smith introduced by June Lloyd. Institute of Child Health, London.

Phenylketonuria has several biochemical variants and atypical phenylketonuria is a term used for those in which blood phenylalanine levels are below 20 mg/100 ml on a normal diet. Affected children are often of normal intelligence without treatment. 3 children (2 of them sibs) were reported with atypical phenylketonuria who have severe progressive neurological illness unresponsive to a low phenylalanine diet. The biochemical findings (blood phenylalanine of 7–15 mg/100 ml on normal diet, moderate phenylpyruvicaciduria, and failure to show a rise of tyrosine with a phenylalanine load) indicate a persistent defect in the hydroxylation of phenylalanine, though not of the type present in the classical disease. In one patient liver biopsy showed normal phenylalanine hydroxylase activity.

All 3 children had a similar clinical course. Feeding difficulties and choking attacks were progressive from the neonatal period, developmental delay was obvious by 5 months, and by 1 year a characteristic neurological picture was present. All voluntary movements and social awareness were lost by 18 months, and 2 of the children died at 2 and 6½ years. Apart from raised phenylalanine levels, no other biochemical abnormalities were found. All 3 patients were treated with a low phenylalanine diet, one from the neonatal period, the other 2 (sibs) from 5 months. Despite satisfactory physical growth and good control of blood phenylalanine, there was no effect on the clinical course.

It is suggested that these patients have a specific variant of phenylketonuria in which the impairment of hydroxylation of phenylalanine and the mechanism of mental retardation are different from that of the classical disease.

Problems encountered in design of diets for treatment of protein energy malnutrition. P. S. E. G. Harland* and J. Mason introduced by P. Dunn.† M.R.C. Child Nutrition Unit, Kampala, Uganda.

The diet previously recommended by the M.R.C. Child Nutrition Unit for treating protein-calorie malnutrition in Uganda was a low sodium and low lactose diet based on calcium caseinate, which provided around