

Comparisons of phenobarbitone, magnesium sulphate, and calcium gluconate in treatment of neonatal hypocalcaemic convulsions. T. Turner introduced by F. Cockburn. Paediatric Department, Simpson Memorial Maternity Pavilion, Royal Infirmary, Edinburgh.

Randomized treatment with calcium gluconate (Ca) 10 ml 10% solution with each feed (22 infants), phenobarbitone (Phe) 7.5 mg 6-hourly orally or intramuscularly (19 infants), or magnesium sulphate (Mg) 50% solution 0.2 ml/kg intramuscularly on two or three occasions at 12-hourly intervals (14 infants) was given to 55 artificially-fed term infants having convulsions associated with hypocalcaemia on the 4th to 7th day of life. Treatment was discontinued after 48 hours in each of the three groups.

Pretreatment plasma calcium concentrations were -2 SD below the mean for breast-fed infants in all groups but were significantly greater in the Mg-treated group when compared with the Ca-treated group ($P = 0.005$). There were no other significant biochemical differences found before treatment, and the number of pretreatment convulsions did not differ between the groups.

There were no significant differences between the mean numbers of convulsions in the first 48 hours after the start of treatment (4.85 ± 5.9 SD in the Mg-treated group, 9.36 ± 9.3 SD in the Ca-treated group, and 9.36 ± 10.9 SD in the Phe-treated group). The number of infants still convulsing 48 hours after the start of Mg therapy was significantly lower than the number convulsing after Ca therapy (Mg:Ca $P < 0.05$; Mg:Phe $P > 0.1$; Ca:Phe $P > 0.3$).

After 48 hours' treatment the following biochemical differences were noted. Post-treatment plasma calcium concentrations were significantly greater in the Mg-treated group (Mg:Ca $P < 0.005$; Mg:Phe $P < 0.01$). Post-treatment plasma phosphate values were significantly lower in the Phe-treated group (Phe:Mg $P < 0.01$; Phe:Ca $P < 0.025$). Post-treatment plasma magnesium concentrations were significantly greater in the Mg-treated group (Mg:Ca $P < 0.001$; Mg:Phe $P < 0.001$).

Although prevention of convulsions associated with neonatal hypocalcaemia is better than cure, treatment of the established case with intramuscular magnesium sulphate is more effective than treatment with oral calcium gluconate, and probably more effective than treatment with phenobarbitone.

Secretin cells in childhood coeliac disease. Mary A. Rossiter and Susan Van Noorden introduced by B. A. Wharton. Queen Elizabeth Hospital for Children, London E.2, and the Royal Postgraduate Medical School, London W.12.

Although there has been considerable investigation of the absorptive and immunological functions of the small intestine in coeliac disease, less attention has been paid to the activity of the hormone-secreting 'Apud' cells.

Jejunal biopsy specimens from 17 children with untreated coeliac disease and 22 children with normal intestinal mucosa were examined for the presence of 'S'

cells. These cells are thought to produce secretin, and can be identified by immunofluorescence and by electron microscopy, if they contain hormone (Polak *et al.*, 1971). The results are shown in the Table. Most of the jejunal

TABLE
Occurrence of 'S' cells (by immunofluorescence) in jejunal biopsies from children with coeliac disease

Jejunal morphology	No. of jejunal biopsies containing 'S' cells		
	Large no. of cells	Few cells	No cells
Subtotal villous atrophy due to coeliac disease	10	2	5
'Normal'	1	5	16

$P < 0.001$, χ^2 test.

biopsies from children with coeliac disease contained 'S' cells, whereas no cells were seen in the majority of the 'normals'.

The increase in hormone-containing cells in cases of coeliac disease was also shown by ultrastructural and cytochemical techniques, which appeared to show actual hyperplasia of the 'S' cells.

It seems that secretin (and possibly other intestinal hormones, too) is not released normally from the Apud cells in childhood coeliac disease. This would affect intraluminal digestion and the metabolic rate of food substances after absorption.

REFERENCE

Polak, J. M., Bloom, S., Coulling, I., and Pearse, A. G. E. (1971). Immunofluorescent localization of secretin in the canine duodenum. *Gut*, 12, 605.

Limitations of xylose tolerance test as screening procedure for coeliac disease. S. P. Lamasadusuriya, S. Packer, and J. T. Harries. The Hospital for Sick Children, Great Ormond Street, and Institute of Child Health, London W.C.1.

The xylose tolerance test is widely used as a screening test for coeliac disease, and a recent communication (C. J. Rolles, B.P.A. Annual Meeting, 1973) reported that a modified version of the test (blood xylose concentration 1 hour after the oral load) was extremely useful in predicting patients who were subsequently proven to have coeliac disease.

We have compared urinary excretion (5- and 24-hour collections) and blood concentrations (1- and 3-hour samples) of xylose after an oral load (0.4 g/kg of D-xylose) in 47 children who underwent jejunal biopsy for suspected coeliac disease. The children fell into one of 3 groups. (A) Normal biopsy (27 patients); (B) abnormal biopsy due to coeliac disease (19 patients); (C) abnormal biopsy due to other causes (5 patients).

5- and 24-hour urinary xylose excretion, and 3-hour blood xylose levels were of no value in discriminating between the 3 groups.

The mean and (range) of 1-hour blood xylose levels (mg/100 ml) were 37.3 (22.5-54.5) group A, 21.5

(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