(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 constipation' 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

Arch Dis Child: first published as 10.1136/adc.49.3.245-c on 1 March 1974. Downloaded from http://adc.bmj.com/ on October 18, 2021 by guest. Protected by copyright.

100 cal/kg per day. Calcium caseinate was expensive and difficult to get and despite the reduced sodium intake, cardiac failure during recovery was a frequent problem. Furthermore, in the light of more recent knowledge of energy requirements for catch-up growth, the energy content of the previous diet was suboptimal.

Consequently, a number of diets were designed with varying amounts of sodium and energy. These were used to treat a series of malnourished children who were then closely followed clinically and biochemically during recovery. In particular, changes in estimated blood volume and in urinary excretion of sodium were measured. It was found that the children who subsequently developed cardiac failure had a significantly low sodium urinary excretion during the initial 5 day collection. This low sodium excretion was associated with large increases in estimated blood volume during the time when oedema was being lost.

An inexpensive diet was designed containing dried skim milk reinforced with energy derived mainly from fat. This was not associated with an increased incidence of cardiac failure when given at a concentration of 100 kcal/kg. After the loss of oedema the diet was given at a rate of 150 kcals/kg with good results.

Present address: *Department of Child Health, University of Bristol.
†Dunn Nutritional Laboratory, Cambridge.

L-Glutamine therapy in Leigh's encephalomyelopathy. A. Moosa and E. A. Hughes. Department of Child Health, Hammersmith Hospital, London.

The diagnosis of Leigh's encephalomyelopathy was suspected during life in a small-for-dates infant with severe neurological abnormalities, persistent lactic acidosis, and hyperalaninaemia. L-Glutamine therapy produced biochemical improvement but the infant died after smallpox vaccination. Necropsy confirmed the diagnosis and detailed enzymatic analysis of the liver revealed a deficiency of pyruvate carboxylase. The significance of this finding in relation to diagnosis and possible therapy was discussed.

Rat model for study of growth in renal failure. C. Chantler, R. D. Adelman, R. C. MacDonell, and M. A. Holliday. Department of Paediatrics, University of California, San Francisco, and Department of Paediatrics, Guy's Hospital, London.

A rat model has been developed to study growth in uraemia. The model allows growth to be observed over short periods and will enable the effects of manipulations in diet and treatment on growth to be examined within a controlled experimental design. The growth and food intake of male and female rats rendered uraemic by 80% nephrectomy was observed between 40 and 70 days of age. The uraemic rats gained significantly less weight and consumed significantly fewer calories than control sham operated rats; tail growth was also less in the uraemic rats. The effect of lowering blood urea by reducing protein intake was studied; no improvement in growth or calorie intake was observed even though blood urea levels were comparable with those found in control rats. Uraemic rats were gavaged with corn oil and improvement in growth was noted. This model should prove useful for studying other factors which have been implicated in the growth failure of uraemia.

Growth and dietary intake of children with chronic renal insufficiency. P. R. Betts and G. Magrath introduced by R. H. R. White. The Children's Hospital, Birmingham.

Growth retardation in children with chronic renal failure is of increasing importance now that replacement treatment is possible by haemodialysis or transplantation. The relations between growth, dietary intake, and degree of renal insufficiency have been studied in 30 children as part of a larger project. The children have been divided into groups A and B.

Group A. 20 children in whom the onset of renal insufficiency occurred in early infancy as a result of either congenital defects or other causes. The majority of these children were on or below the 3rd centile for height irrespective of creatinine clearance. However, they grew steadily along their centile unless they developed renal osteodystrophy. Marked pubertal delay occurred in 2 children.

Group B. 10 children with normal renal function in early infancy but who developed a progressive nephropathy during childhood. Growth retardation in these children was not so marked. They grew normally along their centile except for those with a rapidly progressive nephropathy whose height velocity fell abnormally. Puberty progressed satisfactorily in 3 children.

There was significant reduction in the calorie, protein, and vitamin D intake of these children from that recommended for their age, and of calorie intake compared with children of their own height. It appears that the onset of renal insufficiency in early infancy has a more deleterious effect on growth than that in later years. The reduction in calorie intake in these children may, in part, be responsible for their growth retardation.

Growth hormone secretion during sleep in short children: a continuous sampling study. P. M. Howse, P. H. W. Rayner, J. W. Williams, and B. T. Rudd. Institute of Child Health, University of Birmingham, and Department of Clinical Endocrinology, United Birmingham Hospitals.

A continuous sampling technique has been used to investigate growth hormone (GH) secretion during nocturnal sleep and after insulin-stimulated hypoglycaemia (ISH) in 8 small, normal children (group A) and 6 children with clinical evidence of GH deficiency (group B). In group A there was good correlation between GH response to sleep and ISH. Sleep GH secretion, but not ISH-GH response increased with age. All group B children had low nocturnal GH secretion; ISH-GH secretion was similarly low in 4, but normal in 2 children. A relation has been shown between impaired nocturnal GH secretion, irrespective of ISH-GH response and clinical features of GH deficiency. Sleep-related GH secretion may be an important component of normal growth.

Arch Dis Child: first published as 10.1136/adc.49.3.245-c on 1 March 1974. Downloaded from http://adc.bmj.com/ on October 18, 2021 by guest. Protected by copyright.