In the homocystinuric patient, cystathionine synthetase is absent or abnormal, and this results in an increase in the concentration of the blood and urine homocystine and in an absence of cystine. As the reaction methionine $\rightleftharpoons$ homocysteine is reversible, methionine levels are also found to be raised in the serum. It is these findings that are used to establish a diagnosis.

Because pyridoxine is an essential co-factor in the enzymatic reaction of homocysteine to cystathionine, two groups of workers have given this vitamin in high dosage, and have reported biochemical success in some of their patients.

The response of 10 patients with homocystinuria, treated with oral pyridoxine and a normal mixed diet, is presented. In 5, the biochemical changes were normalized, i.e. homocystine concentrations in the serum have been much reduced, and cystine has appeared. In those cases in which the serum methionine concentration was raised, it has reverted to normal. The biochemical and therapeutic implications of these results were discussed.

M. M. SEGAL introduced by DR. JUNE K. LLOYD (London). 'Treatment of Familial Hypercholesterolaemia in Childhood.' Familial hypercholesterolaemia is associated with a high incidence of ischaemic heart disease in adult life. Treatment has therefore been instituted in affected children in an attempt to reduce this risk. Our experience with 12 heterozygous patients (aged from birth to 16 years) and one homozygous patient (aged 10 years) are presented. The homozygous patient developed tuberous and tendon xanthomata at the age of 18 months; her pretreatment serum cholesterol level was 1050 mg./100 ml. The heterozygous patients did not have xanthomata, but 2 had corneal arcus; their pretreatment serum cholesterol levels were 284-510 mg./100 ml. (mean 375).

Treatment reduced the serum cholesterol levels in all the patients. In the heterozygous children this was achieved by dietary means alone. The diet consisted of restriction of saturated fat and the addition of corn oil to provide polyunsaturated fat. A significant negative correlation was found between the percentage reduction in serum cholesterol and the amount of saturated fat allowed in the diet ($r = -0.80$; $p < 0.01$). In the homozygous child treatment had been previously unsuccessful, and cholestyramine and clofibrate were therefore used in addition to diet. A fall in serum cholesterol of about 400 mg./100 ml. was achieved, and this reduction has been maintained over a period of one year; though the levels remain high (589-675 mg./100 ml.), the xanthomata are regressing.

ALINA T. PIESOWICZ introduced by PROFESSOR O. H. WOLFF (London). 'Hyperprolinaemia.' A 6-month-old boy was found to have clinical and biochemical features of Type I hyperprolinaemia^{1,2}. Plasma proline was 25 mg./100 ml. There was high urinary excretion of proline, hydroxyproline, and glycine, with normal excretion of 4-pyrrolidine carboxylic acids. He had moderate mental retardation, EEG abnormalities, and dilated ureters on intravenous pyelography. In addition he had steatorrhoea, hypocalcaemic convulsions, generalized osteoporosis, and other skeletal changes, abnormalities which have not been previously described in hyperprolinaemia.

He was given a low proline diet (1.5 mg./kg./day). Plasma proline fell to 7 mg./100 ml. within 24 hours and remained around this level. After a month, proline intake was increased (63 mg./kg./day) without rise in plasma proline. Further studies including proline tolerance tests on parents, are proceeding in an attempt to find out whether this child has hyperprolinaemia or presents the heterozygous state, or has temporary immaturity of the enzyme system.

Proline is generally regarded as a non-essential amino acid, and it has been widely assumed that dietary treatment is unlikely to lead to a fall in plasma proline level. Low proline diet has been instituted in 2 other patients³, but results of treatment have not been previously described.

REFERENCES

- ¹ *New Engl. J. Med.* (1962). **267**, 51.
- ² *New Engl. J. Med.* (1965). **272**, 1243.
- ³ *Amer. J. Dis. Child.* (1967). **113**, 116.

A film on 'The PETO Experiment' was shown by Dr. G. G. Fagg.

THE WINDERMERE LECTURE. The Lecture was delivered by Professeur Maurice Lamy, Hôpital des Enfants Malades, Paris, on April 25; 'Immunologic Deficiency Disease in Childhood.'

Members and guests attended a Sherry Party at the Royal College of Physicians of Ireland on Wednesday evening, April 24, by invitation of the Irish Paediatric Association. The Association and its guests were entertained by the Minister for Health at a Reception on the evening of Thursday, April 25, at Iveagh House.

The Ulster Cup competition was held at Portmarnock Golf Club on Friday, April 26, and was won by Dr. K. R. Keay.

The Annual Dinner of the Association, attended by members, guests, and wives, was held on the evening of Friday, April 26, with Mr. Erskine Childers, Minister for Transport and Power and Posts and Telegraphs, as guest of honour.