

mental, non-proliferative sclerosing lesions ('focal glomerulosclerosis') in 12 instances. Its recognition is important since it is a progressive condition showing almost total lack of response to therapy. The fully developed lesion is characterized by partly and completely sclerosed glomeruli as well as normal glomeruli, with tubular atrophy and interstitial fibrosis. Though early lesions may not be distinguishable on light microscopy from minimal changes, this latter condition can usually be ruled out on clinical grounds, and by the appearance of localized thinning as well as thickening of the capillary basement membrane, on electron microscopy.

We have now observed the lesion in 18 children, aged 2 months to 14 years at onset; 13 were girls. Thirteen had the nephrotic syndrome, 3 a mixed nephritic-nephrotic presentation, and 2 symptomless proteinuria. Thirteen had haematuria and 8 hypertension. Proteinuria selectivity was impaired in 16 out of 17 cases; serum  $\beta$ 1c-globulin levels were normal. One child out of 16 responded to corticosteroids and is still in remission. None responded to either cyclophosphamide or azathioprine. Three have died, one is on dialysis and three have renal insufficiency. The remaining 10 have persistent proteinuria.

## REFERENCE

White, R. H. R., Glasgow, E. F., and Mills, R. J. (1970). Clinicopathological study of nephrotic syndrome in childhood. *Lancet*, **1**, 1353.

P. M. DUNN (Bristol). 'Congenital Dislocation of the Hips and Congenital Renal Anomalies.'

Since Potter's first report in 1946 there have been many publications concerned with the presence at birth of various facial and musculoskeletal deformities in babies with congenital renal anomalies.

Clinicopathological and statistical studies made by the writer over a 10-year period strongly support the widely held view that these various deformities occur as the result of pressure because of oligohydramnios due to fetal oliguria or anuria.

Among the cases studied were 12 infants that were also noted to have congenital dislocation of the hips (CDH) at birth. All these infants died soon after birth. Postmortem examination revealed a wide range of renal and urinary tract malformation. Dissection of the hip-joints confirmed the clinical diagnosis in every case and displayed a spectrum of pathology which illustrates the progression from mild hip-joint instability to the 'late' CDH changes normally associated with cases that remain untreated for two or more years.

This association between anomalies of the kidneys and urinary tract and CDH does not appear to have been noted in previous reports. Its importance lies particularly on the light it throws on the aetiology of congenital dislocation of the hips.

W. HAMILTON (Glasgow). 'Re-appraisal of Salt-Losing and Non-Salt-Losing Variants of C21-Hydroxylase Deficiency.' To be published elsewhere.

C. B. MODELL introduced by PROFESSOR L. B. STRANG (London). 'Management of Thalassaemia Major.'

Information was obtained from: (1) a population study of Greek Cypriots in London; (2) a long-term study of 25 patients with thalassaemia-major (including red cell survival and iron excretion measurements), and (3) biochemical studies of Hb synthesis.

The population study yielded an estimate of 15% for frequency of the  $\beta$ -thalassaemia gene. The clinical and biochemical investigations indicate that most of the effects of the disease can be explained by the combination of anaemia, excessive iron loading, and an overactive but ineffective bone marrow. A high transfusion policy, in combination with the vigorous use of iron-chelating agents can make thalassaemia-major a disease with a good prognosis, and lead to avoidance of most of the complications.

C. B. S. WOOD introduced by PROFESSOR N. R. BUTLER (Bristol). 'Serum IgE Concentration in Asthma and its Clinical Significance.' To be published.

P. T. BRAY (Cardiff). 'Review of Histiocytosis-X.'

The paper reviews the clinical, pathological, and radiological features of 32 cases currently included in the diagnostic class of 'Histiocytosis-X'. The material comprises examples of Letterer-Siwe disease, Hand-Schüller-Christian syndrome, eosinophilic granuloma, and various atypical instances.

The effects of treatment by surgery, radiotherapy, steroids, and chemotherapy are discussed. Follow-up findings are also given, extending up to 17 years from diagnosis. Arguments are presented for and against the validity of the overall concept of 'Histiocytosis-X', together with indications for active therapy, and guides to prognosis.

J. M. TANNER (London). 'Isolated Growth Hormone Deficiency; Differential Diagnosis and Treatment with Human Growth Hormone.' To be published.

H. B. VALMAN introduced by DR. R. J. K. BROWN (London). 'Plasma Amino Acids in Small Infants on Various Protein Intakes.' To be published.

NINA A. J. CARSON introduced by PROFESSOR I. J. CARRÉ (Belfast). 'Diagnosis and Management of Hyperphenylalaninaemia.'

Mass screening of newborn infants by the use of the Guthrie microbiological inhibition assay technique on blood spots has only recently been generally adopted in centres throughout Great Britain. Experience of diagnosis and treatment of this disorder may be limited in some of these centres due to the relatively small number of infants screened.

In any mass screening survey there must be, in addition to the collecting and testing of specimens, facilities for confirming abnormal results and monitoring treatment. Close collaboration between the people involved in these various procedures is essential. The organization of such a team is described and the results of screen-