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 β_{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 β -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-

ing the newborn population in Northern Ireland (birth rate 33,000 per year) for hyperphenylalaninaemia with the 'Guthrie' technique over a two-year period are presented and compared with our previous results of ferric chloride testing of urine.

It is now recognized that not all patients with hyperphenylalaninaemia are instances of classical PKU. The importance in distinguishing between the two groups of patients lies in the fact that a phenylalanine restricted diet given unnecessarily will in itself cause failure to thrive and perhaps even brain damage. Distinction has been attempted between classical PKU and its hyperphenylalaninaemia variants; the incidence of the two forms and our experience of their management are recorded.

I. SMITH introduced by PROFESSOR O. H. WOLFF (London). 'The Natural History of Phenylketonuria.'

The natural history of phenylketonuria diagnosed by screening in infancy is obscured by the effect of early dietary treatment. It is, however, known that a 'normal' IQ may occur in untreated phenylketonuria. Cases of phenylketonuria referred to hospital on account of mental retardation do not represent an unbiased sample. A study of sib pairs among a series of 176 consecutive cases allowed us to avoid this bias, and enabled an estimate to be made of the prevalence of a 'normal' IQ in the untreated condition.

The series provided a total of 38 sib pairs. In 18 of these the index case was diagnosed because of mental retardation, and the sib was discovered by chemical screening of the family at the time of diagnosis of the index case. The IQ range of the index cases was 20-55 with a mean of 37, and the IQ range of the sib cases was 20-100 with a mean of 49, 3 out of the 18 siblings having IQs above 80. In this small group of 18 cases, in which the chemical diagnosis was made *not* because of mental retardation but because of the discovery of an affected sib, 1 in 6 of the children had an IQ within the 'normal' range.

A. H. CAMERON, J. H. EDWARDS, and I. MYSKOVA (Birmingham). 'Placentation and Fetal Growth in Twins.' This paper described an analysis of 1000 twin pairs born in Birmingham. The birthweight and morbidity were related to the different types of placenta. The two main types of vascular communication within the placenta and their effects on the fetus were described.

D. J. GIRLING introduced by DR. J. S. WIGGLESWORTH (London). 'Pulmonary Fibroplasia in Newborn Babies Treated with Oxygen and Artificial Ventilation.'

Pulmonary fibroplasia developing in babies subjected to intensive care for respiratory problems in the neonatal period has been ascribed variously to O_2 toxicity, ventilator therapy, or the natural progression of severe hyaline membrane disease. In order to assess the relative importance of these factors a clinicopathological study has been performed on all babies who died in the Neonatal Unit of the Hammersmith Hospital from

November 1965 to October 1970, who received oxygen therapy and who lived for 48 hours or more.

Of the 81 babies studied 39 had hyaline membrane disease and 64 received artificial ventilation. Unequivocal changes of pulmonary fibroplasia were present in 23 cases.

The main findings were as follows:

1. Of babies treated only with less than 60% O_2 , none developed pulmonary fibroplasia.
2. Of babies treated with over 80% O_2 for more than 105 hours, all developed pulmonary fibroplasia.
3. Pulmonary fibroplasia was seen in a few babies who had less than 48 hours artificial ventilation and was absent in several subjected to prolonged ventilation.
4. The most severe histological changes of pulmonary fibroplasia were almost invariably associated with hyaline membrane disease.
5. Gestational age had no effect on the development of pulmonary fibroplasia.

The implications of these findings for O_2 therapy of the newborn were discussed.

M. A. LE DUNE introduced by DR. I. RILEY (Glasgow). 'Insulin and Glucose Estimations in Hypoglycaemic Infants.' To be published.

H. STEINER introduced by DR. G. NELIGAN (Newcastle). 'The Quality of Survivors of Very Severe Perinatal Problems.' To be published elsewhere.

B. J. N. Z. DANESH introduced by DR. C. H. M. WALKER (Dundee). 'Influence of Acid Citrate in Stored Blood on the Level of Electrolytes in Plasma.' Citrate intoxication resulting from multiple transfusions of citrated blood is a well known hazard. Convulsions and cardiac arrest leading to death have been reported in neonates receiving exchange transfusions for erythroblastosis fetalis. These dangers are greater in the presence of hepatic and renal dysfunction, hypothermia, and when there is mechanical obstruction of hepatic circulation during cardiac surgery.

To gain more understanding of the influence of acid citrate on the level of electrolytes in plasma, studies were performed using semipermeable membranes and the technique of equilibrium dialysis.

It was found that citrate not only decreased the ionized fraction of plasma calcium and magnesium by forming citrate salts, but it also appears to combine with the protein bound fraction which, in the absence of citrate, would dissociate to take the place of ionized fraction which has become complexed with citrate. This changes the normal pattern and the total value of these electrolytes present in the blood.

In conditions where citrate intoxication is an impending danger, rapid transfusions of citrated blood should be avoided.

B. O'CONNELL introduced by PROFESSOR K. W. CROSS (London). K. G. Foster, E. N. Hey, and B. O'Connell, 'Sweat Function in Babies with Defects of Central Nervous System.' (*Archives of Disease in Childhood*, 46, 444.)