

Paediatric Research Society

16th Meeting, the University of Birmingham, 17 and 18 April 1970

Abstracts of Papers

Parameters of Connective Tissue Metabolism Metabolism in the Neonatal Period. Brian Wharton and Charles Pennock (*Department of Child Health, University of Bristol; and Department of Chemical Pathology, United Bristol Hospitals*). Two parameters of connective tissue metabolism, viz. urinary hydroxyproline and urinary glycosaminoglycans, have been studied in newborn babies.

A characteristic pattern is discernible, which is probably related to the rate of growth. The precise clinical value of these observations is yet to be determined, but it seems probable that they will provide a useful assessment of 'chemical growth' in the newborn.

Preliminary Studies in Nervous Conduction through Myelomeningocele Lesions.* Alistair Blair (*introduced by Peter Dunn*) (*Department of Child Health, Bristol University*). Using the summation technique pioneered by Dawson (1947), somatosensory-evoked potentials were used in a group of children with myelomeningoceles and in a control group of young adults, to study how practical such a method might be in establishing the exact level of the sensory lesion in cases of gross cord damage. In only 5 out of 11 such attempts was a definite level established, but all of these corresponded with clinical neurological findings. The conclusion is drawn that, though with further refinement the method could be more reliable, at present its clinical use is limited.

REFERENCE

Dawson, G. D. (1947). Cerebral responses to electrical stimulation of peripheral nerve in man. *Journal of Neurology, Neurosurgery and psychiatry*, 10, 137.

Minimal Rates of Oxygen Consumption in Small-for-dates Newborn Babies. O. N. Bhakoo (*introduced by J. W. Scopes*) (*Hammersmith Hospital, London*). Scopes and Ahmed (1966) showed that the small-for-dates baby, unlike the normal term baby, has no rise in resting metabolic rate in the first 3 days of life. They found an abrupt rise in such babies on the fourth day. This study has now been repeated in the same Unit with the same techniques with quite different findings—namely that the small-for-dates baby now behaves as does the term baby. An important difference in the care of these babies between 1964–65 and 1969 is the much more liberal early feeding regimen used now.

*From work done during tenure of Wellcome-Swedish Research Fellowship.

A possible explanation of these findings is that, in 1965, inadequate food reserves were limiting basal metabolic rates.

REFERENCE

Scopes, J. W., and Ahmed (1966). Minimal rates of oxygen consumption in sick and premature newborn infants. *Archives of Disease in Childhood*, 41, 407.

Sequelae of Neonatal Jaundice. P. E. Culley, J. E. Powell, J. A. H. Waterhouse, and B. S. B. Wood (*introduced by J. Insley*) (*New Birmingham Maternity Hospital, and Department of Social Medicine, Birmingham University*). 381 newborn infants falling into 3 groups—non-haemolytic jaundice, haemolytic jaundice, and non-jaundiced controls—have been reassessed in the sixth year of life as regards neurological, audiological, and psychological function. Neurological handicap was concentrated among the infants of low birthweight, and was not related to jaundice apart from one case of athetoid cerebral palsy with deafness. No other cases of perceptive deafness were discovered. Intelligence testing on the Stanford-Binet scale showed no relation between depth of jaundice and IQ.

The findings support the majority of reports in the literature, that reduction in intelligence does not occur in non-haemolytic jaundiced babies with serum bilirubin levels below 20 mg./100 ml.; in haemolytic jaundice slight doubt remains. There is no indication for changing present standards for exchange transfusion.

Histopathological Studies in Post-streptococcal Glomerulonephritis. E. F. Glasgow (*introduced by R. H. R. White*) (*Department of Pathology, the Children's Hospital and Birmingham University*). In post-streptococcal glomerulonephritis, the glomeruli on renal biopsy present histological appearances which reflect the time at which biopsy is done. Among the characteristics of early biopsies are hypercellularity of the swollen tufts, polymorphonuclear infiltration, and a reduction in the number of patent capillary loops. On electron microscopy discrete, electron-dense aggregations may be seen within the basement membrane of the loops and also as subepithelial 'humps'; they are seen only in early biopsies, and the latter are said to be pathognomonic of post-streptococcal glomerulonephritis. Usually they have their bases applied to the basement membrane, while the remainder of the hump is completely separated from the urinary space by cytoplasm of the podocyte. Occasionally they appear to be entirely surrounded by podocyte cytoplasm. On ultra-thin sections of glutaraldehyde-fixed material, stained with a modification of

Movat's (1961) method, these humps can be visualized using both optical and electron microscopical methods. They can also be demonstrated on renal biopsy material prepared for light microscopy using minor modifications of a chromotrop-silver methenamine method (T. Ehrenreich and T. Espinosa, 1967, personal communication). These appearances were observed in renal biopsy specimens from 6 out of 7 children diagnosed as having acute post-streptococcal glomerulonephritis, but with atypical clinical features, and in one child who had had the initial streptococcal infection 17 months before biopsy. Humps were also identified in renal biopsy and necropsy material from a child with rapidly progressive glomerulonephritis, a condition in which they have not previously been described.

REFERENCE

Movat, H. Z. (1961). Silver impregnation methods for electro-microscopy. *American Journal of Clinical Pathology*, **35**, 528.

Water Excretion in Infants and Children with Obstructive Uropathy. C. Chantler and T. M. Barratt (*Institute of Child Health, London W.C.1*). Dilatation of the urinary tract may be due either to anatomical obstruction or to dysplasia; it is sometimes difficult to distinguish between these by radiological techniques and to decide whether significant obstruction requiring surgery is present. The ideal pre-operative assessment would predict the improvement in renal function to be obtained from surgical intervention.

Studies of experimental unilateral hydronephrosis in dogs (Suki *et al.*, 1966) indicate that a smaller fraction of filtered water (V/GFR : V = urine flow rate, GFR = glomerular filtration rate) is excreted during water diuresis by an acutely obstructed kidney than by its control kidney. In contrast, V/GFR is increased in chronic obstruction. It might be expected that those kidneys with the functional characteristics of the acutely obstructed model would show the greatest increase in GFR after relief of obstruction.

GFR was estimated from the plasma disappearance of ^{51}Cr -EDTA (Chantler *et al.*, 1969) before and after surgery in children with presumed obstructive uropathy. Fractional water excretion during pre-operative water diuresis was measured as the plasma/urine creatinine ratio (P_c/U_c):

$$V/GFR = V \div \frac{U_c V}{P_c} = P_c/U_c$$

Fourteen children have been satisfactorily examined to date: the largest fractional increases in GFR after surgery have been seen in the three children who had values of P_c/U_c during water diuresis lower than healthy adults. The other 11 had values higher than those of healthy adults.

REFERENCES

- Chantler, C., Garnett, E. S., Parsons, V., and Veall, N. (1969). Glomerular filtration rate measurement in man by the single injection method using ^{51}Cr -EDTA. *Clinical Science*, **37**, 169.
Suki, W., Eknayan, G., Rector, F. C., Jr., and Seldin, D. W. (1966). Patterns of nephron perfusion in acute and chronic hydro-nephrosis. *Journal of Clinical Investigation*, **45**, 122-131.

Adrenocortical Atrophy and Diffuse Cerebral Sclerosis: Biochemical Studies. Constance C. Forsyth (*Department of Child Health, University of Dundee*). A boy, diagnosed as having Addison's disease due to idiopathic atrophy of the adrenal glands at the age of 7 years, developed the first evidence of what was originally thought to be Schilder's disease at 8 years and 10 months. He died at 9 years and 11 months. There are 12 fully documented reports in the literature of boys with the combination of adrenocortical atrophy and diffuse cerebral sclerosis. The studies of adrenal function during life and of the biochemistry of the brain at necropsy were discussed in relation to theories of causation and the likely mode of inheritance of the condition.

Enzyme Studies of the Testis in Myotonic Dystrophy. William Hamilton and R. G. Mitchell (*Department of Child Health, University of Aberdeen*). Myotonic muscular dystrophy is characterized by familial incidence, inheritance as a dominant trait with variable penetrance, myopathy mainly involving the distal musculature of the extremities and the cranial muscles, cataracts, bony lesions, and endocrine defects such as a low BMR, progressive testicular atrophy, and low urinary 17-oxosteroids.

The testes show tubular degeneration with sclerosis and hyalinization, but Leydig cells may be increased. Testicular biopsies from two affected brothers have been studied. The specimens were incubated as described by Hamilton, Grant, and Wilson (1970). These authors observed that fetal testis synthesized testosterone preferentially from dehydroepiandrosterone before the 12th week of gestation but after that time, preferentially from androstenedione. Undescended testes behaved *in vitro* like the later fetal testes.

The corresponding enzymic activity of testes from these two boys is set out in the Table:

TABLE
Conversion to Testosterone (dpm) as % of Substrate Used

	From Androstenedione	From Dehydroepiandrosterone
Elder 13 years	3123 dpm (0.06%)	15,558 (0.23%)
Younger 11 years	17,549 dpm (0.94%)	5951 (0.10%)

Since the conversion of androstenedione to testosterone is by 17β -reductase, the findings might indicate that as the disease progresses there is progressive failure of 17β -reductase. Also dehydroepiandrosterone was utilized more by the older testis than by the younger testis. The enzyme in this reaction is 3β -hydroxysteroid dehydrogenase which is also the more active enzyme in the early fetal testis. Its persistence then as the