showed increased AChE activity. 7 patients ultimately proved to be cases of Hirschsprung's disease; all but one of the rectal biopsies showed an increased AChE activity in the mucosa, and all biopsies showed an increased activity in the submucosa.

From the investigation of resected bowel segments it was shown that the strongest activity occurred in the most distal part of the aganglionic bowel and that the proximal extension of the enhanced activity into the aganglionic segment increased with age. Rectal suction biopsies stained for AChE activity therefore seem to be a valuable diagnostic aid for investigation of constipation. False negative results are possible in young infants, but can be avoided by taking the biopsy immediately proximal to the pectinate line.

Large-scale investigation of 2 phenylketonuria screening methods and factors affecting blood phenylalanine levels in the newborn. J. B. Holton. Department of Pathology, Southmead Hospital, Bristol BS10 5NB.

Bloods taken from 22,365 babies between the 5th and 20th day of life were analysed for phenylalanine by the Guthrie inhibition assay and by an automated fluorimetric method (Hill *et al.*, 1965). 6 phenylketonurics were found and both methods were equally effective in detecting the disease. The fluorimetric method showed a more consistent pattern of rising phenylalanine levels before treatment. Using an acceptable upper limit of mean $+2$ SDs, the fluorimetric method gave more 'false positive' results. Two-thirds of these high levels were shown by a thin layer chromatogram to be associated with tyrosinaemia. The distribution of blood phenylalanine by both methods was non-Gaussian, and a small shift in the accepted upper limit would eliminate many 'false positives' without significantly altering the detection of phenylketonuria.

An attempt has been made to identify in normal infants some factors which influence blood phenylalanine in the newborn period. The level is raised at day 5 then remains constant up to day 12. After the 6th day of life the phenylalanine level of boys tends to be higher than girls. Babies with a birthweight below 2.5 kg have significantly higher phenylalanine levels than heavier babies, presumably due to immaturity of liver enzymes. However, very low birthweight babies do not show as high levels as those between 2.0 to 2.5 kg, perhaps because of poorer feeding. A big difference was observed between babies born in rural areas and those from urban districts. The reason for the lower phenylalanine level in rural areas is unknown.

REFERENCE

- Hill, J. B., Summer, G. K., Pender, M., and Roszel, N. O. (1965). An automated procedure for blood phenylalanine. *Clinical Chemistry*, 11, 541.

Quantitative study of vesical ganglia in children with neurospinal dysraphism. M. Forbes. The Congenital Anomalies Research Unit, University of Sheffield, Thornbury Annexe, Sheffield 10. This project was undertaken to determine if neurogenic dysfunction

of the bladder in children with spina bifida and meningocele was in any way related to the population of vesical neurones present.

Two quantitative histological studies were performed. In the first, 10 whole bladders (3 normal and 7 spina bifida) were serially sectioned at 10μ . Every 50th section was counted and the total number of ganglion cells calculated per block of bladder from the fundus to the urethra. The results showed an overall reduction in the number of ganglion cells in the bladders of spina bifida children, but there was a particularly striking depletion in the region of the trigone.

In the second study, a single midtrigone block was taken from 100 bladders (50 normal and 50 spina bifida) and again the total number of ganglion cells estimated per block. The mean values for ganglion cell counts related to age showed a marked reduction in the spina bifida cases as compared with the controls.

The possible pathogenesis of these findings was discussed.

Brain swelling in the newborn: artefact, development, or pathology? J. Pryse-Davies. Bernhard Baron Memorial Research Laboratories, Queen Charlotte's Maternity Hospital, Goldhawk Road, London W.6.

Brain swelling was studied prospectively in 183 perinatal deaths of 20 to 42 weeks' gestation. Cerebral flattening in 67 babies obviously correlated with maturity, a factor difficult to exclude in any other analysis; there was also an apparent association with birth asphyxia and intrauterine growth retardation.

Macroscopically there was a progressive series of findings to suggest genuine brain compression. Cerebellar herniation of varying degree was found in 22 cases and slight herniation of uncus gyri in 30: both features occurred in 16 brains. Such herniation phenomena were only found in association with cerebral flattening, which was not always marked. Reduced cisternal CSF in 46 cases and skull moulding in 21 usually correlated with other features of cerebral compression, but also occurred without brain swelling; 18 swollen brains showed no confirmatory evidence of compression. Prolonged body storage and survival time were probably related to an increased incidence of cerebral flattening but not to herniation phenomena.

Babies showing cerebellar herniation have been described in detail, as this was considered the main indication of pathological brain swelling. This finding was associated with death before (3 cases), during (10), and after labour (9). Apart from gross trauma or a softened macerated skull, moulding alone was thought unlikely to initiate herniation. However, constriction of a swollen brain during intrapartum hypoxia might increase the lethal potential of compression and exaggerate the appearances in the dead fetus. In 9 babies cerebellar coning was probably unrelated to skull pressure and due to hypoxia or intracranial bleeding occurring after delivery.

Histological evidence of nerve cell damage showed no consistent relation to brain swelling. Purkinje cell