Paediatric Pathology

gross deformities are not compatible with the concept of developmental arrest. The malformation appears to be due to an increased caudal extension of the cerebellum entirely as a consequence of normal growth in the presence of an early hydrocephalus.


In a postmortem series of 3 small-for-dates infants the brains were examined by naked eye and by light microscopy, and compared with controls. In 9 instances disorders of the cortical convolutional pattern such as agryria, pachygyria, and microgyria were found. Minor convolutional anomalies occurred in a further 5 infants (and also in 2 of the controls). The principal microscopic lesions observed included failure of neuroblast migration—both in cerebrum and cerebellum—as well as poor stratification of the cerebral cortex with retarded differentiation of ganglion cells. The cerebellar cortex showed impaired regression of the embryonal granular layer, reduced inner granular layer, and absence or ectopia of Purkinje cells; the latter abnormality, however, is significant only after eight months of gestation.

These anomalies of the brain were correlated with disordered placental morphology. Fetal dysmaturity, in general, was found to be associated with disturbed early embryonal development of chorionic villi, which may be due to maternal hormonal imbalance or, alternatively, to failing immune tolerance between mother and fetus. In addition to qualitative inferiority of the placental villi, there may also be a quantitative defect, due either to smallness of the placenta or to circulatory damage, causing loss of functional villi.

Primary hepatic cancer in childhood. D. Sinnamon, P. E. Campbell, and J. H. Colebatch. Department of Pathology, Royal Children's Hospital, Flemington Road, Parkville, Victoria 3052, Australia.

During the period January 1950 to June 1972, there were 2 children with primary malignant liver tumours at the Royal Children's Hospital, Melbourne, representing 0.08/1000 hospital admissions and 2% of all childhood malignancies (excluding leukaemia) seen at this institution. There were 16 hepatoblastomas and 4 hepatocellular carcinomas.

Hepatoblastomas. Age from 4 months to 10 years (75% under 3 years). The commonest presenting sign was abdominal distension and the most useful preoperative investigations were hepatic angiography and liver scan. There have been 4 long-term survivors following partial hepatectomy, chemotherapy (mostly mitomycin C and vincristine), and postoperative radiotherapy. Microscopically, the tumours arose in the right lobe and 4 in the left. There were no cirrhotic livers. Histologically 13/16 had fetal cells, in 6 they were the dominant cell type and in another one the only cell type. 14/16 had embryonal cells, 3 being the dominant cell type. 4/16 had anaplastic cell areas, 1 being the dominant cell type. 2/16 had areas resembling 'adult' hepatocarcinoma. 2/16 consisted predominantly of rhabdomyoblastic cells. In 9/16 osteoid was found. In 3 cases the histological pattern was truly mixed with fetal embryonal and stromal components represented about equally.

Hepatocarcinomas. All were advanced when diagnosed and all 4 children died. Symptomatology was varied and in none was abdominal swelling the initial complaint. Macroscopically these tumours were multiple and variegated. One arose in a cirrhotic liver. Microscopically they appeared 'carcinomatous' with cords and trabeculae of cells separated by sinusoids.

CSF in acute childhood leukaemia: cytocentrifuge studies. D. I. K. Evans. Department of Pathology, The Royal Manchester Children's Hospital, Pendlebury, Manchester M27 1HA.

The cytocentrifuge enables a satisfactory cytological preparation to be made when the cells in the CSF are normal in number or only slightly increased. A technique has been developed and the results of its use are described in 114 consecutive samples from 50 children with acute leukaemia, with and without involvement of the CNS. Analysis of the results shows that 30% of the samples with a normal cell count contained leukaemic cells when examined by cytocentrifuge; and only 74% of the samples with a raised count were found to contain leukaemic cells. It also appears that changes in the levels of protein and glucose in the CSF of leukaemic patients are not directly related to the presence of leukaemic cells, but are the result of changes in the cell count from whatever cause.

Screening for abnormal haemoglobins in the immigrant community. D. N. Raine and J. M. Pepper. Department of Clinical Chemistry, The Children's Hospital, Birmingham B16 8ET.

Following the advice that those at risk for sickle cell disease should be examined before any procedure that might precipitate a sickle cell crisis is undertaken, a number of centres have introduced a variety of means whereby these patients can be tested. Experience with the Sriver method of screening neonates for amino acids, in which blood is collected in capillary tubes, led to the consideration of a similar approach to the haemoglobinopathies.

The system involves direct testing by starch gel electrophoresis of blood collected via school clinics. The method allows 100 specimens to be processed daily. Data obtained by other methods in other laboratories is also used. A punched card file and magnetic tapes store is maintained in which identification is recorded by surname in words and phonetic code, forename, day and month of birth, and National Health Service number (more than 70% of NHS numbers are being obtained). The method of analysis, centre of testing, date of specimen, and result are also recorded. Fortran programs (IBM 1440 computer) allow the card to be decoded and the data printed, a new subject to be matched against the data bank, the Soundex code to be
automatically generated, and certain statistics which monitor the study to be recovered. Some 12,000 names have so far been filed.

Two cases of neuroblastoma with marrow findings mimicking acute leukaemia. T. E. Parry. Department of Pathology, Llandough Hospital, Penarth, Glamorgan CF6 1XX.

Two cases of neuroblastoma with multiple bone metastases in which marrow biopsy on first hospital admission presented findings indistinguishable from those of acute leukaemia occurred in a boy of 7 and a girl of 3 years, in both of whom the correct diagnosis was established at necropsy after an illness of some weeks. The features which may enable the two conditions to be differentiated during life are the absence of malignant cells from the peripheral circulation and the characteristic clumping of cells in the marrow with the formation of syncytiial masses and of pseudorosettes in neuroblastoma. Though well documented (Pinay, Mallarmé, and Ross, 1950; Gaffuey, Hansman, and Fetterman, 1959), the difficulty this differential diagnosis may present receives little attention in the standard textbooks of haematology.

REFERENCES


Diagnosis of immune deficiency states. J. Huber. Department of Pathology, Hospital for Sick Children, 555 University Avenue, Toronto 101, Ontario, Canada.

Significance of ‘focal sclerosis’ in lipid nephrosis. J. D. Elema. Department of Pathology, University of Groningen, Holland.

Renal focal sclerosis is characterized by the presence in some glomeruli of adhesions of Bowman’s capsule with thickening of the mesangial matrix and capillary eosiophilic sclerosis in the absence of a glomerular proliferative lesion. Immunofluorescence shows the presence of IgM and C3, and the increase in mesangial matrix and deposits of heterogenous material has been confirmed by electron microscopy. There is a definite relation between the histopathology and proteinuria in the nephritic syndrome, though in some patients increased protein excretion has been shown to precede the development of focal sclerosis.

A comparison may be drawn with the focal glomerulosclerosis sometimes observed in aging rats. The light and electronmicroscopical features and immunohistology are similar to human focal sclerosis and these changes may also be shown to be preceded by increased protein excretion. It is suggested that focal sclerosis in nephrotic patients resistant to therapy is the result rather than the cause of persistent proteinuria.


Without measuring the intrathoracic air pressure by means of a manometer, followed by embedding the suspected lung tissue in nitrocellulose, many cases of ectopic air in the thorax will escape detection. This is unfortunate because the finding of air ‘in the wrong place’ can help to establish the cause of death. In the present study, interstitial lung emphysema occurred in 7% of all the babies who died in the first week of life, and it is concluded that this precedes most of the cases of mediastinal emphysema and pneumothorax seen in the newborn.

A sudden rise in intrathoracic pressure applies a shearing force to the alveolar margins in contact with the periartrial sheaths which may then rupture, allowing alveolar air to enter the interstitium.

Predisposing diseases in the newborn were hyaline membrane disease, intracranial haemorrhage, renal dysgenesis, and adrenal hypoplasia.

Forced respiration in an attempt to recover from anoxia is thought to be the commonest cause, but in this study of 100 cases of interstitial pulmonary emphysema in the newborn, mouth-to-mouth respiration as a cause of the lung pneumatosis was seen in only 4%, which is much lower than expected.

Necrosis of vocal cords in infants. G. J. Cullity and J. L. Emery. Department of Pathology, Children’s Hospital, Western Bank, Sheffield S10 2TH.

Vocal cord lesions in infants have been reported at necropsy in association with sudden unexpected death (SUD) by 4 different groups of observers in the last 20 years. The lesions are generally described as deep-seated foci of necrosis or inflammation in the true vocal cord. Some have described them in up to 90% of cases of unexplained SUD, and most believe them to be pathognomonic. The aetiology and pathogenesis are unknown. Similar ulcerated lesions were described in 1962 by Osborn and Flett in newborn infants dying with respiratory distress syndrome and believed by them to result from vocal cord spasm. We have studied the vocal cords in an unspecified group of 117 infants and children, comprising 35 cases of SUD by history, 55 hospital deaths in infants and children, and 7 stillbirths. The lesions seen have been classified histologically into 7 groups. When allowance is made for the effects of intubation, the same pattern of lesions is present in all but the stillbirth group. This argues against a specific pathogenetic mechanism confined to the cot death situation. The lesions certainly occur before death, and suggest the existence of a laryngeal disorder which merits further study.

REFERENCE