

anti-*i*, were now readily detectable. Though the relation of transfer factor to the polyclonal gammopathy may be coincidental, it is possible that in severe combined immunodeficiency disease transfer factor may induce uncontrolled B cell proliferation.

Biochemical state of the vitreous humour of infants at necropsy. P. G. F. Swift, E. Worthy, and J. L. Emery. Department of Pathology, Children's Hospital, Western Bank, Sheffield S10 2TH. To be published in full in the *Archives*.

Hypernatraemia and uraemia in unexpected death in infancy. J. L. Emery, P. G. F. Swift, and E. Worthy. Department of Pathology, Children's Hospital, Western Bank, Sheffield S10 2TH. To be published in full in the *Archives*.

Paediatric dosage of gentamicin. T. A. McAllister and D. G. Young. Department of Pathology, Royal Hospital for Sick Children, Yorkhill, Glasgow C3.

Ototoxicity of the broad spectrum antibiotic gentamicin has been recognized for 10 years and it has been customary to use it cautiously to achieve blood levels of 2 to 10 $\mu\text{g/ml}$. Most sensitive bacteria have a minimum inhibitory concentration of less than 2 $\mu\text{g/ml}$ and ototoxicity is most unlikely below 10 $\mu\text{g/ml}$. Evidence of gentamicin nephrotoxicity is poor unless it is used in combination with cephalosporins or diuretics.

Therapeutic levels are achieved in adults with a dose of 0.8 mg/kg 3 times daily, but with this regimen results in children are disappointing. The drug is excreted by glomerular filtration, which is proportional to surface area and therefore more efficient in children, especially neonates.

We studied 61 peak serum assays in 30 children with severe or potentially severe sepsis given 2½ to 3 times the adult dose intramuscularly or intravenously. The children had a variety of surgical conditions, complicated by infection, and ranged in age from 3 days to 12 years. There was little difference in response in the different age groups. Apart from 3 'dampened responders', we concluded that children require a dose of at least 2 mg/kg 3 times daily for therapeutic efficacy. Neonates may require more, but all cases should be monitored by serum assays. These recommendations have been submitted to the Committee on the Safety of Medicines.

Adult type of polycystic disease of kidneys and liver presenting in childhood. B. G. Ockenden. Department of Pathology, North Staffordshire Royal Infirmary, Stoke on Trent.

Spina bifida and the value of the α -fetoprotein estimation in antenatal diagnosis. M. A. Ferguson-Smith, L. D. Allan, I. Donald, E. M. Sweet, and A. A. M. Gibson. Royal Hospital for Sick Children, Yorkhill, Glasgow C3.

Chromosome studies and the paediatric post-mortem. R. Sutherland, R. Bauld, and A. D. Bain. Department of Pathology, Royal Hospital for Sick Children, Sciennes Road, Edinburgh EH9 1LF.

Since October 1972 chromosome studies have been carried out on all stillbirths and infants coming to postmortem. The results of the study to date were presented and their significance discussed. The frequency of chromosome abnormalities in this group is between 5 and 10%. In view of the high incidence of chromosome abnormalities it is suggested that chromosome studies should be routinely carried out at the paediatric postmortem.

Rubinstein-Taybi syndrome. A. J. Barson. Department of Pathology, St. Mary's Hospital, Whitworth Park, Manchester M13 0JH.

The case is described of a growth-retarded infant with multiple congenital anomalies born at 33 weeks' gestation to a 30-year-old woman with 1 normal son, after a pregnancy complicated by hydramnios. The infant died on the 4th postnatal day with congestive heart failure, jaundice, and skin petechiae.

The infant had abnormally broad tips to the fingers, thumbs, and toes, an anomaly characteristic of the Rubinstein-Taybi syndrome. In addition he had a small maxilla and mandible with a high, arched, furrowed palate, a depressed nasal bridge with the septum below the alae, low-set abnormal ears, palmar simian creases, an atrial septal defect, a 4-lobed right lung, biliary atresia, bilateral hydronephroses, a large anterior fontanelle, deficient falx cerebri, and a Dandy-Walker type of malformation of the cerebellum. Histologically the thyroid and adrenal glands showed disordered differentiation.

These features are all compatible with the syndrome. As with previously reported cases, no chromosomal anomaly was shown.

Deformity of the lateral cerebellar lobes in children with meningocele. S. Variend. Department of Pathology, Children's Hospital, Western Bank, Sheffield S10 2TH.

A study was made of the gross structural deformity of the cerebellum from 100 children with meningocele. The fissural pattern of the superior surface was classified in a variety of forms indicating degrees of displacement of the major fissures.

The portion of the cerebellum that had herniated into and through the foramen magnum was also described and classified. When the changes in the cerebellar tail were correlated with those of the upper and posterior surfaces, it appeared obvious that the deformity of the cerebellum is essentially a displacement backwards over the superior surface and downwards over the posterior surface.

It has already been shown that the weights of these deformed cerebella are less than that of normal infants, thus opposing theories of tissue overgrowth. When minor degrees of deformed organs occur there is a resemblance to the early fetal cerebellum, but the more