Hydrops fetalis. M. J. Becker. Department of Morbid Anatomy, Wilhelmina Gasthuis, Amsterdam.

Hydrops fetalis is characterized by an abnormal accumulation of serous fluid in the tissues and body cavities, and infants who show this condition are usually either stillborn or die soon after birth. Hydrops fetalis is not a symptom of a specific disorder, but can be caused by various diseases. In a series of 9 necropsies on hydropic infants various conditions were diagnosed as underlying the hydrops fetalis. Blood group incompatibility occurred in only 2 cases. Hydrops fetalis in the remaining 7 cases was due to syphilis, viral infection, cystic lung disease, premature closure of the ductus arteriosus with hypoplasia of the lungs, aneurysm of the umbilical artery, angiomyxoma of the umbilical cord, and a chorangioma of the placenta. Although blood-group incompatibility is the most frequent cause of hydrops fetalis, it does not account for the majority of cases.

Double inlet right ventricle. J. N. Cox, Institut Universitaire de Pathologie, 40, Boulevard de la Cluse, 1211 Genève 4, Switzerland.

A 16-year-old Moroccan boy was admitted for an intense cyanosed condition of long duration, dyspnoea, and marked digital clubbing. His haemogram showed a polycythaemia of 9 200 000 RBC/mm³ (Hb 20.3 g/dl and haematocrit 76%). Haemodynamic and angiocardiographic studies suggested the possibility of a double outlet right ventricle associated with pulmonary stenosis and hypoplastic left ventricle. However, the patient died shortly after surgery. The heart weighed 350 g at post mortem. The right ventricle was hypertrophied, dilated, and communicated by means of an interventricular defect with hypoplastic left ventricle from which the aorta took its origin. The pulmonary trunk was to the right and somewhat behind the aorta and arose from the right ventricle. Its cusps were thickened and rigid. The ductus arteriosus was patent. The atrium was dilated, and the tricuspid valve was well formed and large. The left atrium was somewhat smaller, received the pulmonary veins but had a defect in its lower portion. Behind the posterior leaflet of the tricuspid valve, and hidden by it, was the smaller mitral valve with its chordae tendineae anchoring it into the right ventricle. The superior edges of the posterior leaflet of the tricuspid and the anterior of the mitral valves were fused together forming a ridge dividing the interauricular opening almost into two portions; the upper part communicating with the right ventricle by way of the tricuspid orifice and the lower by way of the mitral valve.

Gonadoblastoma in familial XY pure gonadal dysgenesis. A. A. M. Gibson and M. A. Ferguson-Smith. Department of Pathology, Royal Hospital for Sick Children, Yorkhill, Glasgow G3 8SJ.

Visceral neuromatosis. A. H. Cameron. Department of Pathology, Children's Hospital, Ladywood Middleway, Birmingham B16 8ET.

Significance of 3-methoxytyrosine and its metabolites in urine of patients with sympathetic nervous system tumours. L. Penchansky, B. H. Landing, K. N. F. Shaw, and D. J. Taylor. Children's Hospital of Los Angeles and University of Southern California School of Medicine.

Excretion of 3-methoxytyrosine (3MT) and its metabolites has been associated with malignancy in neuroblastoma and allied tumours, but published series are small. Pathological features of tumours were compared to chromatographic findings in preoperative urine specimens for 52 children with adrenal medullary or sympathetic nervous system tumours. At 3MT levels above 30 mg/g urinary creatinine, N-acetyl-3MT (Ac3MT) and related vanillic acid (VLA) were often also excreted. Patients fell into 3 groups. (1) 9 patients with 3MT present, with homovanillic acid (HVA) levels slightly higher than vanilmandelic acid (VMA). 6 had adrenal primary site, 5 were male and 8 have died. (2) 7 patients with 3MT present, with HVA no higher than VMA. 6 had adrenal primary, only 2 were male and 5 have died. (3) 36 patients with no detectable 3MT. 13 had adrenal (6 alive), 20 non-adrenal (15 alive) and 3 uncertain primary site (none alive); 21 were female. Determination of 3MT and its metabolites Ac3MT and VLA appears to have prognostic value in this group of tumours (82% mortality for groups 1 and 2 against 41% for group 3) but certain patterns seem to indicate greater malignancy than others, and those patients with high 3MT and high HVA relative to VMA seem to have very primitive tumours (89% mortality against 71% for patients with high 3MT but with VMA as high as or higher than HVA, or against 54% for entire series).

Sarcomatous chest wall tumour with good prognosis. A. Ahmed, A. J. Barson, and A. M. MacDonald. Department of Pathology, University of Manchester M13 9PT; and Department of Pathology, Royal Hospital for Sick Children, Glasgow G3 8SJ.

Two intrathoracic tumours of similar light microscopic appearance were found in girls of 8 and 14 years of age. In both cases the ribs were involved; the tumour had a fleshy haemorrhagic cut surface and histologically was composed of round anaplastic cells with numerous vascular spaces. After surgical resection one patient was given radiotherapy and the other chemotherapy, and both are alive 2 and 1 years later, respectively. The differential diagnosis for both tumours lay between a vasoformative sarcoma or a Ewing's sarcoma.

The question was resolved by examination of the tumour from one patient by electron microscopy. The cells were characterized by prominent cytoplasmic organelles and a variable number of pinocytotic vesicles. Occasional tight junctions were observed but no desmosomes were present. These morphological features were consistent with an endothelial cell type and suggested a diagnosis of haemangioendothelioma. A diagnosis of Ewing's sarcoma was excluded because ultrastructurally the cells were poor in cytoplasmic organelles and showed an absence of pinocytotic vesicles.

and frequent desmosomes. The good prognosis of these tumours tends to support a diagnosis of haemangioendothelioma rather than Ewing's sarcoma.

Necropsy findings in a case of Lesch-Nyhan syndrome. V. Mahnovski, S. Dozic, D. Vulovic, B. Marjanovic, and G. Tasic. Institute of Mothers and Child Health of SR Serbia, 8, Radoja Dakica, 11070 Novi Beograd, Yugoslavia.

A case of self-mutilation in an infant was first apparent at the end of the second year of age and resulted in death at 3 years 8 months. During life he was physically and mentally retarded with polydipsia, polyuria, choreo-athetosis, and spasticity of the legs. EEG showed diffuse moderate cortical damage. Serum uric acid was >30 mg/100 ml and urinary uric acid was 50–130 mg/kg per 24 h. Renal function was impaired, but no calculi or haematuria was observed. At necropsy there were numerous excoriations and scars in the perioral and infraorbital regions and over both hands. The fingers and toes were plump and short and of similar size. The brain weight was slightly reduced (960 g, normal 1154 g) and there were moderate focal degenerative changes. The kidneys showed interstitial nephritis and focal glomerulosclerosis. There were numerous granulomata containing uric acid crystals, predominantly in the medulla. These granulomata were not seen in other organs though they have been described in the liver, spleen, and bone marrow. Additional findings were focal degenerative changes in some skeletal muscles, chronic enteritis with hyperplasia of mucosecretory glands, prominent pancreatic periductular fibrosis, and lamellation of the adventitial tissue of the periaortal blood vessels. There was also focal chronic pneumonitis and diffuse chronic bronchitis.

Oedema of umbilical cord and respiratory distress in the newborn. J. M. Scott and J. B. S. Coulter. Departments of Pathology and Neonatal Paediatrics, Glasgow Royal Maternity Hospital, Rottenrow, Glasgow G4 0NA.

Oedema of the umbilical cord (defined as visible oedema in a cord with a minimal cross-sectional area of 1.3 cm) was found in 11.5% of deliveries. It was seen more frequently in certain complications of pregnancy such as abruptio placentae, maternal diabetes, macerated intrauterine death, and conditions such as prematurity, Rhesus isoimmunization, respiratory distress syndrome, and transient respiratory distress. There was a higher incidence in infants delivered by caesarean section. There was no association between cord oedema and either fetal distress or neonatal asphyxia, nor any correlation with maternal hypertension or oedema.

Some factors involved in the production of oedema included low osmotic pressure, raised hydrostatic pressure in the placenta and umbilical cord, or an increase in total water in the fetoplacental unit. It is suggested that oedema of the cord may reflect similar changes in the lungs which prenatally predispose an

infant, whose pathway for production of surfactant is immature, to develop respiratory distress syndrome, and the mature infant to develop transient respiratory distress.

Pathogenetic implications of the lesion complex of hyaline membrane disease. D. R. Shanklin. Laboratory of Pathology, Chicago Lying-in Hospital, Chicago, Illinois 60637.

The elements of the lesion can be divided into principal and ancillary. The principal elements are (a) partial collapse with centrilobular air space distension ('air bronchogram'); (b) vascular congestion, especially in capillaries and venules; (c) pulmonary oedema and lymphatic dilatation; (d) membranes. Ancillary elements include (e) necrosis of bronchiolar epithelium, especially in early cases and in very small fetuses; (f) focal haemorrhage, both interstitial and in air spaces; (g) polymorphonuclear leucocytosis, especially at about 20 to 30 hours; (h) later macrophagic response; (i) swelling of interstitium with possible increased cellularity and increased matrix.

These changes represent phases in a classic form of injury, accommodation, and repair. The stability of the lung and alterations of permeability which are so striking provide evidence for disruption of the expected mechanisms for integration of perfusion-ventilation interaction. Disturbance of the ventilatory action could arise either from initiatory or from regulatory phenomena. Changes in permeability must mean profound injury to the usual vascular defenses, and the full range of factors that have to do with that integrity. These points suggest either a multifactorial aetiology and pathogenesis, or a sufficiently diverse agent or event to promote a wide range of physicochemical and physiological changes. The promptness of onset of clinically observable disease and the occasional severity of lesions in short-lived infants speak for the importance of events surrounding the onset of breathing, and the lack of development of defences in the prematurely born.

Wilson-Mikity syndrome. O. Braun. Niederoesterreichisches Landeskrankenhaus, Pathologisches Institut, A-2340 Modling bei Wien, Austria.

A preterm infant with a birthweight of 1350 g and length 38 cm lived for 80 days, in spite of progressively increasing respiratory distress and cyanosis. Oxygen was given from birth but could not control hypoxia and acidosis. Radiologically, increased striation in the perihilar fields was observed and at the age of 5 weeks the x-ray picture was typical of Wilson-Mikity syndrome. At necropsy the lungs were firm and dark red, with numerous small emphysematous bullae in the subpleural tissue. There was no histological evidence of infection either in the present case or in published reports. Special emphasis was given to the changes found in the fibrous scaffolding of the lungs. Collagenous fibres were increased, especially in the thickened septa and alveolar walls. Elastic fibres were irregularly distributed and in some areas at least were split, rolled up, and protruding into the alveolar lumen. Argrophil fibres were similarly fragmented. Conversion of