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, dyspneoa, and marked digital clubbing. His haemogram showed a polycythaemia of 9 200 000 RBC/mm^3 (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-acetyle-3MT (Ac3MT) and related vanillactic 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 microscopical 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