Pulmonary reticulosis. J. I. Gibson. Department of Pathology, University of Aberdeen Medical School, Foresterhill, Aberdeen.

A case is presented of an 'abnormal individual', a female aged 4 months, the result of a first cousin mating. The main features clinically were extreme dolichocephaly, arachnodactyly, and susceptibility to infection. There was no chromosomal abnormality.

The pathological features were dominated by an infiltration present in the lungs thought at first to be extramedullary haemopoiesis. The liver and spleen were not enlarged and though a few of the primitive cells could be found easily in each organ, there was not the pronounced infiltration observed in the lungs. The bone marrow was considered to be acute reticulo-endotheliosis, Letterer-Siwe disease.

Cerebral rhabdomyosarcoma. H. B. Marsden. Department of Pathology, Royal Manchester Children's Hospital, Pendlebury, Manchester M27 1HA.

Striated muscle fibres may be observed in 3 primary tumours of the CNS, namely medulloblastomas, teratomas, and rhabdomyosarcomas. The medulloblastoma has been regarded as a variant of medulloblastoma, though the possibility that this is a malignant teratoid tumour has been considered. Teratomata are typically seen in the pineal region but have also been reported in association with spina bifida. Rhabdomyosarcomas have, with few exceptions, been confined to the cerebellum. A case of a tumour of the pineal region in a 10-year-old boy was presented which showed extension to the cerebellum and seeding to the ventricular system and meninges. In the extrapineal region the microscopical picture was that of a rhabdomyosarcoma, but at the primary site a nodule of cartilage was also noted.

The origin of mesodermal tumours of the CNS was discussed. The presence of cartilage in the tumour presented was not conclusive of a teratoid origin. Cartilage has been encountered in glial tumours, principally ependymomas, and it is possible that the mesodermal tumours arise from ectomesenchyme of the neural crest.

Malacoplakia in childhood. C. Sinclair-Smith, L. B. Khan, and S. Cywes. Departments of Pathology and Surgery, Red Cross Children's Hospital, University of Cape Town, South Africa.

A case of malacoplakia involving the adrenal gland and colon of a 6-week-old infant was presented. The adrenal gland was the site of a previous haemorrhage and the bulk of the malacoplakia cells occurred in this region. The colon was adherent to the adrenal gland and a perforation was present in the segment of bowel involved

by the disease. Ultrastructurally, the cytoplasm of the malacoplakia cells contained granular inclusions of varying electron density, distinctive 'fingerprint bodies', and laminated mineralized inclusions. The latter corresponded to the Michaelis-Gutmann bodies seen by light microscopy. In a review of published reports in English, only 8 other cases of malacoplakia occurring in children under the age of 13 years were found out of a total of 164 cases. The ages in the series ranged from 15 months to 9 years. The bladder was involved in 4 cases, bladder and ureter in 1, both kidneys in 1, descending colon, mesocolon, and regional lymph nodes in 1, and colon and terminal ileum in 1. 4 patients died with malacoplakia, but only in 1 case could death be directly attributed to the disease.

Spontaneous rupture of the aorta in a 13-year-old boy. J. N. Cox and E. Jacot-des-Combes. Institut, Universitaire de Pathologie, 40 Boulevard de la Cluse, 1211 Geneve 4, Switzerland.

A 13-year-old boy was admitted to hospital with abdominal pains which began while he was playing football. There was no apparent trauma. He was kept under observation but suddenly collapsed and died the second night after hospitalization. At necropsy there were 5 transverse tears, complete and incomplete, of the thoracic and abdominal aorta, and mediastinal and retroperitoneal haematomas as well as a bilateral haemothorax. There was no perceptible aneurysm nor coarctation of the aorta. Histology showed dissection into the outer third of the aortic wall in the vicinity of the torn areas. Outside these zones there were focal medial degenerative changes of the elastic and smooth muscle fibres with the formation of clefts and cyst-like spaces in a condition termed cystic medial degeneration of the aorta. In some places hyaline connective tissue was seen replacing, to some extent, the normal aortic structure. There was anteroposterior flattening of the crystalline lens (platyphakia) with degenerative vacuolization of the posterior surface (lenticonus), but no cataract.

Cystic medial degeneration is probably due to developmental defects of the connective and elastic tissue of the aorta. It is commonly associated with coarctation of the aorta or other anomalies of the cardiovascular system. It is also associated with certain hereditary conditions such as Marfan's syndrome or the Ehlers-Danlos syndrome. In the case presented, inflammation of, as well as trauma to, the aorta can be excluded, while degeneration of the media remains the basic lesion. In the absence of a family history and stigmata of Marfan's syndrome this diagnosis is not supported, however, it is possible that we are dealing with a *forme fruste* of this syndrome.