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


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 polydysplia, polyuria, choreoathetosis, 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 mucosseitary glands, prominent pancreatic peri-ductular fibrosis, and lamellation of the adventitial tissue of the periadrenal 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 placenta, 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 hyper tension 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 predisperse 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 centrolobular 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, accomodation, 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 p-rfusion-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 physiologic 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 prematurity born.


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. Argyrophil fibres were similarly fragmented.
collagenous fibres into elastin-like material is one of the characteristic features of this condition.

The Wilson-Mikity syndrome is considered to be due to disturbed pulmonary maturation, but it is the interstitial tissue rather than the alveoli that bears the brunt of the disorder. The syndrome is thus characterized not only by typical clinical and radiological features, but also by pathological changes, so that it is justifiably considered an entity.

Mural bronchitis in childhood. C. Sinclair-Smith, F. Dinsdale, and J. L. Emery. Department of Pathology, Children's Hospital, Western Bank, Sheffield S10 2TH.

The total involvement of bronchi and bronchioli in children by a wide zone of round cells has been termed mural bronchitis, and the similarity in composition of this lesion to lymphoreticular aggregates has been noted. Lymphoreticular aggregates are increased in incidence in sudden unexpected death in infancy and this study was performed to determine the relation of mural bronchitis to unexpected death in infancy, to a history of respiratory symptoms, and to determine its incidence.

A random series of 503 lungs from children of all ages and comprising a cross-section of almost all deaths in Sheffield was evaluated as to the presence or absence of mural bronchitis. 36 children showed the lesion. Mural bronchitis was found to be absent at birth and the frequency increased with age. No relation to unexpected death in infancy was found but there was a positive correlation between prominent lesions and a history of respiratory symptoms of long duration. The frequency of isolation of Haemophilus influenzae from children with the lesion was significantly higher than control cases. Lungs showing squamous metaplasia of the bronchial epithelium had a negative relation to the isolation of H. influenzae. Mural bronchitis appears, like lymphoreticular aggregates in the lung, to be a manifestation of nonspecific antigenic stimulation.

Ondine's curse: a pathogenic mechanism in pulmonary hypertension. H. S. Rosenberg and R. L. Williams. Departments of Pathology and Pediatrics, Texas Children's Hospital and Baylor College of Medicine, Houston, Texas.

In primary pulmonary hypertension (PPH), pulmonary arterial structural changes appear in the absence of a known stimulus, apparently the result of prolonged vasoconstriction. As a stimulus to pulmonary hypertension, hypoxaemia produces pulmonary vasoconstriction which is reflected anatomically in medial hypertrophy of pulmonary arteries. In another form of pulmonary hypertension without a well-defined stimulus, persistent fetal circulation (PFC) syndrome includes term, cyanotic infants with a right-to-left shunt due to raised pulmonary vascular resistance.

In a review of 14 children with PPH, the disease in one child resulted from chronic hypoxia of alveolar hypoventilation due to inflammation in the brain stem; 5 had the residual of PFC; the disease in the remaining 8 remains idiopathic. The patients ranged in age from 2 months to 6 years. 7 patients, including 5 in the first year of life, had medial hypertrophy of the small muscular arteries with no intimal disease. The infant with the clinical syndrome of Ondine's curse had an obscure illness at one month of age followed by lethargy, episodes of apnoea, and poor ventilation. By 4 months pulmonary and systemic arterial blood pressures were equal. Necropsy findings at age 1½ years included right ventricular hypertrophy, medial hypertrophy, and intimal sclerosis of the pulmonary arteries and central nervous system inflammation confined to the brain stem. Intimal muscular plaques, possibly unique for pulmonary hypertension due to hypoxia, were found only in the patient with Ondine's curse. The development of intimal plaques suggests a potential for more advanced arterial sclerosis.

Serum uracil + uridine levels in normal subjects. T. E Parry and J. A. Blackmore. Department of Pathology, Llandough Hospital, Penarth, Glamorgan CF6 1XX.

Serum uracil + uridine levels, expressed as uracil, have been measured on 144 normal subjects ranging in age from birth (cord blood) to the eighth decade, by a microbiological method using Streptococcus lactis (NCIB no. 10769) as test organism. The organism has a specific growth requirement for uracil or uridine but it does not respond to uridylic acid. The mean level of 22 μmol/l (0.25 mg/100 ml) in cord blood decreases to 15 μmol/l (0.17 mg/100 ml) in adults over the age of 20. There is no difference between the sexes. Uracil is of interest because (a) it is a constituent base of RNA, (b) it is the precursor of two of the bases thymine and cytosine that enter into the composition of DNA, and (c) under certain circumstances it has mutagenic properties. The last is dependent upon the existence of two tautomeric forms of uracil, the common keto and the rare enol form. The former pairs normally with adenine, but the latter can pair with guanine. The entry of uradil in its enol form into the DNA molecule to pair with guanine can result in a G = C → GA = T base transition in the DNA molecule. The molecular mechanisms involved as well as its possible bearing on somatic mutation were discussed.

Myelofibrosis in mongols. D. I. K. Evans. Department of Pathology, Royal Manchester Children's Hospital, Pendlebury, Nr. Manchester M27 1HA.

Since 1958 3 children with Down's syndrome and acute myelofibrosis have been seen at this hospital. Details of one case have been published (Hillmann and Forrester, 1968). Myelofibrosis is rare in children: 24 cases of primary disease have been reported. 3, including the case of Hillmann and Forrester, had Down's syndrome. There appears to be an increased incidence of acute myelofibrosis in children with Down's syndrome, which is a further example of the instability of the haemopoietic system in this disease.

REFERENCE