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 antigen stimulation.

Ondine's curse: a pathogenetic 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 14 months 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

Proceedings: Mural bronchitis in childhood.

C Sinclair-Smith, F Dinsdale and J L Emery

Arch Dis Child 1975 50: 667
doi: 10.1136/adc.50.8.667

Updated information and services can be found at:
http://adc.bmj.com/content/50/8/667.1.citation

These include:

Email alerting service
Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

Notes

To request permissions go to:
http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:
http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:
http://group.bmj.com/subscribe/