## 114 CARDIORESPIRATORY ASSOCIATIONS OF ACUTE BRONCHIOLITIS IN INFANCY

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The occurrence of apnoea during acute viral bronchiolitis in infancy has not been documented in relation to age and maturity of the infant and stage of the illness. We have recorded respiratory movements (chest and abdomen) and heart rate over 24 hours in 27 infants with acute viral bronchiolitis. There were 20 full-term and 7 pre-term infants studied at a mean age of 10.5 weeks (range 2-20) and 10.3 weeks (range 6-17) respectively. Sequential studies of 10 infants during the coryzal, bronchiolitic and recovery stages of illness showed the changing pattern of events. Of 9 infants (2 pre-term) studied in the coryzal stage both the pre-term infants showed abnormalities - 1 prolonged central apnoea >20 seconds without bradycardia and the other apparent obstructive apnoea with bradycardia (≤80 bpm for 10 seconds). During the acute bronchiolitic stage, 5 of 18 infants (2 term, 3 pre-term) had central apnoea > 20 seconds associated with bradycardia in one pre-term infant. Apparent obstructive apnoea with bradycardia in 7 (4 term, 3 preterm). Of 14 infants in recovery, 1 term infant had prolonged central apnoea and 1 other 'obstructive' apnoea with bradycardia. Apnoea was most severe during the acute stage for both the group and individual infants studied sequentially. All those with significant central apnoea were aged less than 10 weeks, 'obstructive' apnoea was seen up to 13 weeks.

Age, maturity and stage of illness influence the pattern of respiration in bronchiolitis.

## 115 EXTRALOBAR SEQUESTRATION OF THE LUNG IN CHILDREN

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Extralobar sequestration of the lung is defined as nonfunctioning lung tissue with no or nonluminous connection with the normal bronchial tree and is covered and surrounded by its own visceral pleura. This definition does not include blood supply from anomalous systemic artery, which will be shown to be absent in many cases. 38 patients with pulmonary sequestration were treated in 1961-84 at our hospital, 22 had intralobar sequestration, 15 extralobar and 1 patient had both types. Only the 16 patients with extralobar sequestrations are further analysed. The age of the patients varied from 21 days to 12 years (mean 3,61 yrs). The sequestration was on the left side in 14 patients and on the right side in three patients. There were only 7 left lower accessory lobes, the lesion was in 7 cases above the lung hilum and in the remaining 2 in the right lower pulmonary region. The most common symptoms were: dyspnea (9), pneumonia (5), cough attack (4), syanosis (3), asthmatic symptoms (2), funnelchest (2), abdominal symptoms (1), routine chest X-ray (1). All 16 patients were operated on and the accessory lobe was removed. Most of the patients were symtomatic and that indicates the necessity of surgical treatment. The clinical picture of extralobar sequestration varies considerably and many of the "typical" features, such as systemic arterial supply, left lower lobe localization and diaphragmatic defect are often absent.

## 116 A DEFINITIVE ANATOMICAL CLASSIFICATION OF PULMONARY SEQUESTRATION: REVIEW OF 16 CASES

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Sixteen cases of pulmonary sequestration treated at the Brompton Hospital and Great Ormond Street Hospital are presented. These demonstrate the wide range of anatomical features found in this condition together with the high incidence of associated congenital lesions - especially diaphragmatic and cardio-vascular anomalies. Excluding young infants with significant haemodynamic problems pre-operatively, our operative mortality for the series is nil. However a review of the literature involving similar patients reveals an unacceptably high mortality (up to 20% in some series). These are usually due to unexpected vascular complications occuring when surgery is performed without accurate pre-operative assessment. The current terminology of intralobar and extralobar sequestration fails to describe the lesion adequately and alert the surgeon to the full range of pathology present. We propose a more definitive clasification describing the essential anatomical features, namely, tracheo-bronchial connection, arterial supply and venous drainage of the affected area. It is hoped that this approach will encourage accurate pre-operative assessment thereby reducing the current unacceptable surgical mortality.

## 117 CHARACTERISATION OF FACTORS PRODUCED BY PSEUDOMONAS AERUGINOSA WHICH SLOW AND DISORGANISE BEATING OF HUMAN CILIA IN VITRO.

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In cystic fibrosis colonisation by Pseudomonas aeruginosa leads to clinical deterioration. We measured ciliary beat frequency of normal human cilia by a photometric technique. 8 culture filtrates of clinical isolates of Ps.aeruginosa produced a range of ciliary slowing (3-57%), dyskinesia and stasis. This activity did not correlate with filtrate protease, elastase or haemolytic activity, nor with other enzymes measured by plate methods. Other virulence factors (exotoxin A, lipopolysaccharide, alginic acid) did not slow cilia. However, filtrate pigment content (absorbance 400 nm) correlated (r=0.97) with ciliary slowing. Gel filtration of lyophilised filtrate gave a single peak of activity corresponding to pigment fractions. Pyocyanin pigment was extracted from an active strain with chloroform and crystallised with petroleum ether. 1-hydroxyphenazine was produced by acid precipitation from an alkaline solution of pyocyanin. When purified by reverse phase high performance liquid chromatography and characterised by UV spectra and mass spectrometry, both slowed cilia in a dose-dependent manner. 1-hydroxyphenazine caused immediate ciliary slowing, dyskinesia and stasis (50 μM). Pyocyanin caused gradual slowing but finally widespread stasis and epithelial disruption (1 μM). Colonisation by Ps.aeruginosa may be assisted by the paralysing effect of these factors on mucociliary clearance.

## 118 CILIARY DISEASES

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The immotile-cilia syndrome has been characterized as an inborn disease in which cilia (and sperm tails) are unable to move in a normal and effective manner (Afzelius, Science 193, 317, 1976). When studied with the electron microscope the ciliary axoneme was found to lack the dynein arms, i.e. the components that are responsible for converting the chemically bound energy into microtubule sliding and hence ciliary movements. Later other types of defects in the ciliary axoneme have been found, such as deficiencies in the so called spokes or in the nexin links. By now our clinical material includes 40 children (aged 0 - 18 years), 18 of which have situs inversus. (There are also 35 adult cases, 20 of which have situs inversus.) Of the forty children about half the cases have a markedly lowered number of dynein arms and in most cases the orientation also is poor. In others the number of dynein arms was only slightly decreased or the dynein arms were shorter than normally. Some had defective spokes or a disorganized axoneme which probably was caused by defective nexin links. In three cases no ciliated cells were seen. Two children had cilia with a normal ultrastructure and a third case had cilia that appeared normal except for the length that was twice the normal one. Evidently the immotile-cilia syndrome is a genetically heterogeneous disease although it is clinically homogeneous. These data can be compared with data from three cases suffering from cystic fibrosis and having cilia with a normal ultrastructure except for the ciliary orientation that in one case was poor.

## 119 CHRONOLOGY OF SYMPTOMS AND COMPLICATIONS OF IMMOTILE CILIA SYNDROME (ICS).

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Up to date most pubblished reports on ICS refer to clinical picture without any distinction between symptoms and complications observed when diagnosis is performed.

In a series of 12 ICS pts patterns and time of onset of symptoms and of organ system related complications have been established.

Nasal dropping and cough were prevalent from birth followed by bronchitis, recurrent otitis and lung infections.

Serous otitis and synusitis were the most early and frequent complications.

Nasal polyps, bronchiectasis, segmental/lobar atelectasis, hear loss and anosmia were less frequent and more late.

A detailed clinical feature is usefull for early diagnosis expecially in those pts without situs viscerum inversus.