1255 SYSTEMIC BLOOD PRESSURE RESPONSE TO CONTINUOUS POSITIVE AIRWAY PRESSURE (CPAP) IN INFANTS WITH HYALINE MEMBRANE DISEASE (HMD).

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The response of aortic blood pressure (BP) to the initiation of CPAP was studied in 8 prematures with birth weights 0.8-1.95 kg and gestational ages 28-33 weeks. CPAP (2.5-6.5 cm water) was started within the first 4 hours of life for the treatment of severe HMD. Instantaneous heart rate, aortic BP, respirations and rectal and toe temperatures were recorded continuously. Readings were obtained prior to and at 2 min. intervals for 20 min. during CPAP. Arterial blood gases were obtained before and while on CPAP. Using paired t test each variable was compared before CPAP and at 10 and 20 min. intervals after the onset of CPAP. The only significant change was a sustained elevation of systolic and diastolic BP after 10 and 20 min. ($p<0.05$) without a significant change in baseline heart rate. Five of 7 infants showed no change or an increase in toe temperature during this period suggesting that peripheral vasoconstriction was not responsible for the rise in BP. The increase in BP therefore should be the result of an increase in stroke volume, and with no change in heart rate should result in an increased cardiac output. The absence of change in heart rate in response to the increase in BP differs from the baroreceptor reflex described in normal preterm infants. This sustained increase in BP with initiation of CPAP appears to be a reflex not previously identified in the distressed preterm infant.

1256 CONGENITAL PULMONARY LYMPHANGIECTASIS OCCURRING IN SIBLINGS: POSSIBLE GENETIC IMPLICATIONS.

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Congenital pulmonary lymphangiectasis (CPL) has been a recognized pathological condition in the newborn for nearly a century. However, all previously reported cases have been sporadic. We report here two female siblings with CPL.

The children were born two years apart to normal parents. Family history and both pregnancies were unremarkable. Both, grossly normal at birth, developed bilateral pneumothorax, did not respond to treatment, and died within 24 hours of birth.

At autopsy, gross examination of the lungs revealed subpleural cystic areas containing clear fluid in all lobes. All other organs from both children appeared normal. Histological examination in both cases confirmed the diagnosis of CPL.

CPL has long been considered non-familial. However, its relative rarity may lead to underdiagnosis of CPL in distressed newborns, and thus, to underestimation of familial occurrence. This first reported instance of CPL occurring in sibs should stimulate interest in the accurate diagnosis and further search for a possible genetic component in this disorder.

1257 PULMONARY FUNCTION IN SHORT STATURE

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We studied 18 children with short stature (sst) ages 7 to 19 years without lung diseases on 26 occasions. Four groups of sst, based on overall pulmonary function were identified. First, the primordial and genetic sst had pulmonary functions appropriate for their heights. Second, a case of constitutional sst, Noonan's syndrome, mixed gonadal dysgenesis and two cases each of mosaic Turner's and renal sst had increased static lung volumes (except in Noonan's, who had normal volumes), and moderate to marked increases in flow rates (PEF, MMEF, FEV_1). Specific conductance, SGaw (normal 0.199 ± 0.047) ranged from 0.225 (Noonan's) to 0.293 (constitutional). These findings suggest an exaggeration of non-isotropic patterns of alveolar and airway growth, altering the interdependence of mechanical forces in the lungs. Third, two cases of post-operative craniopharyngioma had decreased static lung volumes, flow rates and SGaw. In both cases $^{D}LCOsb$ values were lower than predicted. Fourth, four cases of idiopathic growth hormone deficiency had low static lung volumes and lower than predicted $^{D}LCOsb$. The flow rates were within normal limits. Despite human growth hormone administration of 3 to 6 months' duration, there was no evidence of lung growth exceeding that of the height.

1258 THE EFFECT OF DIFFERENT HEAD POSITIONS ON THE WORK OF BREATHING IN VERY LOW BIRTHWEIGHT INFANTS WITH ENDOTRACHEAL TUBES.

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We measured mechanics of breathing of 3 very low birth weight infants (820, 850, 940 g) who required positive pressure ventilation (2-10 breaths/min) and distending pressure. We put a pneumotachograph onto the endotracheal tube and an esophageal balloon in the mid esophagus. Each infant was studied 3 times (29-66 days). Seven studies were satisfactory. Measurements were made with the infant's head in 5 different positions, to the right, left, flexed, neutral and extended. Respiratory flow and pressure were recorded and analyzed by a computer programmed to calculate respiratory rate, tidal volume, lung compliance and work of breathing, breath by breath. Each head position was maintained for approximately 1 minute and analysis made of a period of regular breathing. At least 10 (mean = 17) breaths were analyzed. There was a 2 to 8 fold difference in the work of breathing (per liter ventilation), between the different head positions in 6 of 7 studies. The average work in the position of least and greatest work was 2280 g. cm/L/min (S.D. $\pm$1240) and 6680 g. cm/L/min (S.D. $\pm$2980). The position corresponding to least work varied. Roentgenograms showed that the endotracheal tube position changed with different head positions with the tube tip moving up and down the trachea, into the main bronchi and against the wall of the trachea. We conclude that routine changes in the head position of these infants cause marked changes in the work of breathing which might prolong respiratory insufficiency.

1259 CHANGE IN EXTRAVASCULAR LUNG WATER WITH GROWTH IN RATS.

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Although previous studies have demonstrated that total lung water decreases with growth, the possibility that this may have resulted solely from changes in pulmonary blood water (PBW) has not been excluded. Since extravascular lung water (ELW) may be the more important determinant of lung function, we measured ELW at 2, 8, 14, 22, 28, 180 and 365 days in a total of 66 rats. Following intravenous injection of ^{51}Cr tagged red blood cells, radioactivity as well as wet and dry weight of samples of blood and lung were measured to determine ELW and PBW.

Age Days	ELW g/g dry bloodless lung	% ELW in lung	PBW g/g dry blood	% Water in blood
2	4.68 (SD 0.46)	82.3	8.63 (SD 0.57)	88.2
28	3.87 (SD 0.25)	79.4	5.15 (SD 0.40)	83.7
180	3.60 (SD 0.25)	78.1	4.05 (SD 0.44)	80.0
365	3.61 (SD 0.23)	78.1	4.32 (SD 0.43)	80.1

Regression analysis revealed a significant and rapid fall in ELW from 2 to 28 days (slope = -0.028) and a slower but significant decline subsequently through 180 days (slope= -0.002) $p<0.01$. We conclude that there is a decline in both ELW and PBW with growth throughout the first 6 months of life in the rat.

1260 FLOW-VOLUME RELATIONSHIPS IN INFANTS WITH HYALINE MEMBRANE DISEASE.

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Flow-volume ($\dot{V}/V$) loops and pressure-volume loops (P/V) were obtained during the performance of crying vital capacity (CVC) maneuvers in 31 infants ranging in birth weight from 960 to 2750 grams and in gestational age from 26 to 40 weeks. Eighteen of the 31 infants were diagnosed as having respiratory distress due to hyaline membrane disease (HMD) on clinical and radiological criteria. As reported in previous investigations, CVC was reduced in infants with HMD. Peak expiratory flow rate (PEFR) was not significantly related to CVC ($r=.16$). When CVC was standardized for body size a significant relationship ($r=.25, p<.025$) was found. Peak inspiratory flow rate (PIFR) demonstrated higher correlations with CVC ($r=.70, p<.001$) and CVC per cm of body length ($r=.64, p<.001$). Similar patterns of $\dot{V}/V$ loops were found in normal and HMD infants with HMD infants demonstrating smaller loops due to low flows and loss of lung volume due to disease. Their loops returned to normal with recovery. Expiratory flow was 25% of inspiratory flow in normal infants and 33% of inspiratory flow in HMD infants. During performance of CVC both HMD and non-distressed premature infants may generate pressures as high as 30 cm H_2O during expiration. Taken in conjunction with the significant reduction in PEFR/PIFR when compared with normal children and adults, this suggests that airway compliance in premature infants is high, and that airway closure during CVC may limit CVC and cause air-trapping during normal breathing.