

1834 PULMONARY FUNCTION TESTS (PFT) IN JUVENILE RHEUMATOID ARTHRITIS. Madi Rao, Senih Filbrig, Edward Kong, Maria Ames, Jeffrey Caran and Phillip Steiner, SUNY, Downstate Medical Center, Department of Pediatrics, Brooklyn, N.Y.

As clinically obvious pulmonary disease and/or abnormal pulmonary function tests are very rare in children with JRA, we report our initial findings of a longitudinal study of pulmonary function tests (PFT) in 13 children with JRA. The age range is 6-16 years with m:f: 8:5

	Type of JRA	
	Polyarticular(9)	Fauciarticular(4)
Abn. Chest X-ray (Pl. EFF, PN, Fibrosis)	2	1
Abn. PFT	5*	2
Rest. dis.	5	2
Obst. dis.	1	0
Diffusion	3	2
Exercise induced bronchospasm	4	2

*A 6 year old child died of respiratory failure secondary to fulminant fibrosis

The abnormal PFT in 53.8% of the patients are not related to age, sex or the type of disease, but appear more frequent with increasing severity of JRA. It is early to comment on the relation to the type of drugs used. Exercise induced bronchospasm is most interesting. In two patients, the PFT did not improve in spite of excellent recovery from JRA.

1835 NUTRITIONAL AND PULMONARY ABNORMALITIES AT THE TIME OF DIAGNOSIS OF CYSTIC FIBROSIS IN INFANTS IDENTIFIED BY NEONATAL SCREENING. Mary C. Reardon, Keith Hammond, Frank J. Accurso, Edward R.B. McCabe, Ernest K. Cotton, and C. Michael Bowman (Spon. by Ronald W. Gotlin), University of Colorado School of Medicine, Department of Pediatrics, Denver.

The status of patients with cystic fibrosis (CF) prior to overt symptomatology is poorly described. Early identification of CF infants through neonatal screening using the immunoreactive trypsinogen assay followed by sweat testing allowed us to evaluate CF patients prior to onset of overt symptoms. Seventeen CF infants (mean age 5.7 weeks) without acknowledged symptoms were studied prior to medical intervention to determine their nutritional and pulmonary status. Eight infants decreased weight percentile from birth to diagnosis despite adequate caloric intake (mean intake 160 cal/kg/day). Triceps skin fold thicknesses were less than the 50th percentile for all infants measured. Three infants had significantly low serum albumin and/or total protein values. Even more strikingly, the group's mean level of prealbumin, a rapid-turnover protein, was significantly ($p<.05$) lower than that of age-matched normals. In addition, two of 12 infants became hypoxic during sleep, decreasing their transcutaneous P_{O_2} to less than 40 torr. Four infants without previously acknowledged lung disease had abnormal chest x-rays, showing hyperexpansion, linear markings of bronchial cuffing and infiltrates. We conclude that abnormalities of growth, protein metabolism and pulmonary function exist in infants with CF prior to the onset of marked clinical symptomatology. We speculate that some of these deficits may be reversible with medical intervention.

1836 RESPIRATORY PROBLEMS IN ACHONDROPLASIA. C. Reid, S. Metz, R. Meny, J. Phillips III, C. Francomano and R. Pyeritz. Depts of Peds and Med, Johns Hopkins Univ Sch of Med and Dept of Peds, Univ. of Maryland, Baltimore.

A significant number of infants and children with achondroplasia have respiratory complications. Suggested etiologies include: 1) upper airway obstruction, 2) thoracic cage deformity and 3) neurologic dysfunction. We prospectively evaluated 10 achondroplasts (6 males, 4 females, ages 4 mos-6 yrs) for respiratory problems. 2/10 were asymptomatic, 3/10 had mild symptoms (3 with tachypnea, 1 with pneumonia) and 5/10 had severe symptoms (2 with obstructive sleep apnea [OSA], 3 with episodic cyanosis while awake and/or recurrent pneumonia). Nasal airflow was compromised and hyperextended neck posture was assumed during sleep in 6/10. Chest circumference was <3rd percentile for age in 6/10. Asymmetric hyperreflexia was found in 3/10. Cor pulmonale was diagnosed by echocardiography in 6/10, including 1 with mild symptoms. Arterial hypoxemia ($P_{O_2}<75$) while awake was found in 4/10 and sleeping hypoxemia in 6/10. In the 2 hypoxemic infants evaluated, low tidal volumes were found. Polysomnograms confirmed OSA in 2/10 and mixed apnea in 1/10. In 2 hypoxemic infants, small chest circumference was the only clear etiology found. Cervicomedullary cord compression was diagnosed by CT and evoked potentials in 4/10 (2 with episodic cyanosis, 1 with OSA and 1 with tachypnea and pneumonia). Thus in only 3/8 with respiratory complications could a single etiology be implicated; 5/8 had at least two etiologies. We conclude that symptomatic patients require comprehensive evaluation to delineate all possible etiologies in order to clarify treatment.

1837 THE INSPIRATORY:EXPIRATORY TIME (I:E) DURING HIGH FREQUENCY JET VENTILATION (HFJV) OF AN RDS MODEL. Jerrilyn Johnston, Peter Richardson, and Jeffrey Carlstrom, (Spon. by H. Hill) Dept. of Peds., University of Utah Medical Center, Salt Lake City, UT.

HFJV, unlike high frequency oscillation, relies on passive elastic properties of the lung for exhalation. I:E can be adjusted to allow time for exhalation and prevent inadvertent PEEP. To determine which I:E affords maximal cardiopulmonary function we varied I:E from 1:1 to 1:5 at 600 BPM and measured Pa_{O_2} , Pa_{CO_2} , calculated alveolar-arterial O_2 differences ($AaDO_2$) and estimated systemic blood flow (\dot{Q}) using Fick's principle (assuming constant oxygen consumption) in surfactant depleted cats (lungs lavaged 6 times). Mean airway pressure (MAP) was held constant at 6 cm H_2O , inspiratory pressure was 9 to 10 cm H_2O , PEEP 4 cm H_2O , and FiO_2 1.0. Results (mean \pm SE) show highest Pa_{O_2} and Pa_{CO_2} and lowest $AaDO_2$ and \dot{Q} were at 1:1. Pa_{O_2} increased as ratios changed from 1:3 to 1:5 ($p<0.01$). It is interesting that oxygenation varied even though MAP, rate and FiO_2 were constant. \dot{Q} at 1:5 was less than 1:4 ($p<0.01$). We conclude: the combination of best oxygenation and ventilation with least cardiovascular effects was obtained when an I:E of 1:4 was used.

I:E	1:1	1:2	1:3	1:4	1:5
Pa_{O_2} (mmHg)	127 \pm 26	91 \pm 13*	93 \pm 16*	104 \pm 17	114 \pm 20
Pa_{CO_2} (mmHg)	41 \pm 6	36 \pm 5	34 \pm 3	30 \pm 3*	27 \pm 3*
$AaDO_2$ (mmHg)	345 \pm 44	385 \pm 42*	385 \pm 41*	378 \pm 42*	372 \pm 44*
\dot{Q} (% of 1:1)	100	159*	161*	154*	133

*different from values at 1:1 $p<0.0123$

1838 THE EFFECT OF PERIODIC BREATHING AND SLEEP STATE ON THE INCIDENCE AND "STRUCTURE" OF AUGMENTED BREATHS IN NEONATES. Jaya Bodani, Tazeem Aizad, Kathy Yorke, and Henrique Rigatto, Dept. of Peds., Univ. of Manitoba, Canada.

To determine the incidence and "structure" of augmented breaths (AB) in neonates, we studied 13 preterm infants (GA 31 \pm 0.4 wk; BW 1.56 \pm 0.09 kg; PNA 25 \pm 4 days) and 11 term infants (GA 40 \pm 0.3 wk; BW 3.4 \pm 0.14 kg; PNA 4 \pm 0.8 days). The incidence of AB was higher in periodic than in regular breathing both in preterm (0.914 \pm 0.07 vs 0.434 \pm 0.09 breaths/min; $p=0.0063$) and in term infants (0.803 \pm 0.09 vs 0.406 \pm 0.04 breaths/min; $p=0.0009$). In term infants the incidence was greater in active than in quiet sleep (0.79 \pm 0.12 vs 0.48 \pm 0.06 breaths/min; $p=0.02$). V_T of the AB was 18.9 \pm 0.71 ml as compared to 7.5 \pm 0.71 ($p<0.001$) during control in quiet sleep in preterm infants. The increased V_T was associated with an increase in T_I from 0.46 \pm 0.03 to 0.80 \pm 0.04 seconds ($p<0.001$), in V_T/T_I from 15.9 \pm 1.46 to 23.5 \pm 1.8 ml/sec ($p<0.001$) with no change in T_{tot} . Instantaneous ventilation increased from 0.327 \pm 0.041 to 0.666 \pm 0.073 L/min/kg ($p<0.001$). V_T of the first inspiratory component of the AB was greater than V_T of control breaths, but similar changes in V_T in other control periods was not associated with AB. Airway occlusion produced no pressure-on-the-top-of-a-pressure pattern but was followed by AB after occlusion was released. Results suggest, 1) ABs are highly correlated with prematurity, periodic breathing and active sleep; 2) their appearance is not dependent on chemical drive or volume alone. We speculate that both chemical drive and lung volume changes are important to induce augmented breaths in neonates.

1839 THE EFFECT OF SLEEP STATE AND CO_2 INHALATION ON THE CONTROL OF EXPIRATORY DURATION IN THE NEWBORN INFANT. Patrick Van Reempts, Guy Moriette, Don Cates, Kathy Yorke, and Henrique Rigatto, Dept. of Pediatrics, University of Manitoba, Winnipeg, Canada.

To determine the effect of sleep state and CO_2 inhalation on the control of expiratory time (T_e) in neonates we studied 9 preterm (BW 1990 \pm 92 g; GA 33 \pm 0.5 wks and PNA 12 \pm 3 days) and 9 term infants (BW 3420 \pm 211 g; GA 40 \pm 0.4 wks and PNA 4 \pm 1 days). We measured tidal volume (V_T), expiratory time (T_e), post-inspiratory diaphragmatic activity (Pi_{di}), transpulmonary pressure (P_L), expiratory flow (\dot{V}_{exp}) and expiratory pulmonary resistance (R_e) in both quiet and active sleep. After breathing 21% O_2 for 3 mins in each state, infants rebreathed from a bag containing 5% CO_2 in 40% O_2 for 2 to 3 minutes. We calculated R_e using P_L and \dot{V}_{exp} measured at 0.25 sec after the beginning of expiration and also at the point of maximum expiratory flow. A total of 1538 breaths were analyzed. \dot{V}_{exp} was similar in preterm and term infants (30.55 vs 29.32 ml/sec; $p=0.5$) and greater in active than in quiet sleep (33.7 vs 26.1; $p=0.02$). \dot{V}_{exp} was a highly correlated function of Pi_{di} , the lower the flow the greater the Pi_{di} ($r=0.84$; $p=0.001$; $n=815$). The effect of CO_2 was to decrease Pi_{di} ($p<0.01$). P_L and \dot{V}_{exp} correlated negatively with T_e in such a way that the R_e remained the same within the range of T_e studied. The observations suggest that the control of expiratory time in the neonate is greatly dependent on Pi_{di} and that R_e has no measurable role. This may be a handicap to small infants who cannot depend on expiratory resistance to stabilize T_e .