1200

HYPERVISCOSITY SYNDROME WITH COR PULMONALE SECONDARY TO CYSTIC FIBROSIS. Stella B. Kontras and Gordon A. Young, Department of Pediatrics, Ohio State University College of Medicine and Children's Hospital Research Foundation, Columbus, Ohio.

An 18 year old boy with cystic fibrosis complicated by cor pulmonale was found to have an elevated hemoglobin of 21.9 gms.% and a hematocrit of 72%. The studies of whole blood viscosity by the Wells-Brookfield microviscometer showed elevated viscosity at all shear rates. Because of the increasing plethora, headaches and lethargy unexplained by any other cause, it was elected to treat the hyperviscosity by phlebotomy and plasmanate infusion. Phlebotomies were done on 3 alternate days with withdrawal of 340 cc. blood each time and infusion of an equivalent amount of plasmanate. Bed rest and oxygen were administered after the treatment. The patient improved symptomatically and the plethora was decreased. The initial viscosity of 35.2 centipoise (cp.) at shear rate of 11.5 sec.$^{-1}$ was reduced to 23 cp. (normal is 12 ± 2 cp.). The hematocrit was reduced from 74% to 65.5%. Although O_2 tension did not change but remained at PO_2 of 45 mm., there was symptomatic improvement including decreased cyanosis. It is theorized that this was due to improved blood flow after reduction of blood viscosity and subsequent improved tissue perfusion. Hyperviscosity as a late complication of cystic fibrosis is presumed to be due to hypoxic stimulation of erythropoiesis and erythropoietin studies are in progress.

1201

THE ROLE OF STEROIDS IN EARLY BRONCHOPULMONARY DYSPLASIA (BPD) Lloyd I. Kramer, Christopher Hultzen Georgetown University Medical School, Georgetown University Hospital, Department of Pediatrics, Washington, D. C. (Spon. by P.A. Jose)

Eleven premature infants with hyaline membrane disease, whose gestational ages were 27-36 weeks, required ventilatory support and supplemental oxygen. Their clinical course indicated irreversible lung damage. Due to advancing stage II BPD on Xray and continued oxygen and ventilator dependence, they were started on a course of Dexamethasone (Decadron, MSD). Within forty eight hours, each patient showed a clinical response with increasing pulmonary compliance, a diminishing oxygen and ventilator requirement and a halt in the progression of the disease on Xray. A double blind study is now being set up to evaluate the usefulness of steroids in BPD. Our present evidence indicates prompt clinical improvement in lung disease, but with associated risk and side effects including hypertension, susceptibility to sepsis, gastric ulcers, cushinoid syndrome and difficulties in weaning which complicate management. It appears that in the inflammatory phase of developing BPD, Decadron is a useful, but not benign, new approach to this life threatening lung disease.

1202

A LOWER PHARYNGEAL "SPHINCTER" PRODUCING SLEEP APNEA IN CHILDREN:DIAGNOSIS AND TREATMENT. R.E. Kravath, C.P. Pollak, B. Borowiecki, C.B. Croft, (Spon. by L. Finberg), Albert Einstein College of Medicine, Montefiore Hospital and Medical Center, Departments of Pediatrics, Neurology and Surgery.

Knowledge of the clinical significance, diagnosis and therapy of sleep apnea in children is still incomplete. We have seen three children, 2, 5 and 6 years of age, with obstructive sleep apnea documented by polygraphic recording who required tracheostomy for relief of incapacitating and life threatening obstructive sleep apnea. These children did not improve on passage of a nasopharyngeal tube as do those with nasopharyngeal airway obstruction associated with enlarged tonsils and adenoids. On fiberoptic endoscopy under light anestheisa the site of obstruction was noted not to be in the nasopharynx, but in the oro and hypopharynx. In one patient airway obstruction was related to glossoptosis. In two patients a sphincter like closing of the lower pharyngeal walls was observed that was synchronous with inspiration. We will show a motion picture that demonstrates this phenomenon both aurally and visually. These patients represent 10% of all children referred for sleep apnea during the past 1½ years. This lower pharyngeal site of airway obstruction has also been noted in adults with obstructive sleep apnea who also respond well to tracheostomy.

Although tracheostomy may seem an extreme measure for this still obscure and poorly understood entity we think it is indicated for selected patients at this time.

1203

NEUROGENIC MEDIATION OF AUGMENTED SURFACTANT SECRETION AT BIRTH. Edward E. Lawson and Pearl S. Huang (Spon. by H.W. Taeusch, Jr.) Harvard Medical School, Children's Hospital Med. Ctr., Dept. of Pediatrics, Boston, MA.

The observation that alveolar lavage surfactant increases at birth following air or nitrogen breathing suggests that lung expansion affects net surfactant secretion (Ped Res 11:574, 1977). To study the mechanism of secretion we treated 13 pregnant rabbit does at 30 days gestation with repeated infusions of atropine (total, 2.4mg IV) or propranolol (total, 3-10mg IV) over a ½ hour period immediately prior to sacrifice. The pups were delivered by hysterotomy and divided into two groups. One group was sacrificed at birth without breathing. The remaining pups were sacrificed after 30 minutes hyperventilation induced by hypoxia (10-15% O_2) or anoxia. Following sacrifice, alveolar surfactant was recovered by saline lavage and quantitatively estimated on a surface-tension balance (Science 169:603, 1970).

SURFACTANT - mg.gm dry lung	Control(n=9)	Atropine(6)	Propranolol(7)
Non-breathing	1.79±.29	1.77±.29	1.09±.24
Breathing	2.59±.40	1.89±.25	1.90±.45
Paired t-test (p)	<.003	NS	<.03

mean±SE

These results provide additional support for the hypothesis that pulmonary surfactant secretion is stimulated following the onset of gas ventilation in newborns. We conclude that augmented surfactant secretion at birth is mediated by a neural mechanism. Furthermore, since the secretion is blocked by atropine and not propranolol we suggest secretion is under cholinergic control.

1204

DECREASED ARTERIAL OXYGEN TENSION ACCOMPANYING ASYNCHRONOUS CHEST WALL MOVEMENT IN ACTIVE SLEEP. Richard J.Martin,Albert Okken,Dov Rubin,Dept.Ped, CWRU,Cleveland, Ohio (Spon.by M. Klaus).

In the neonate a decrease in lung volume accompanies the asynchronous(paradoxical)chest wall movement seen during active sleep. We sought to determine whether this would result in a decrease in arterial PO2 in view of the higher incidence of apnea in active as compared to quiet sleep. 10 healthy term infants were studied at a mean age of 39 hours(range 15-56 hrs) during both active (A.S.)and quiet sleep(Q.S.).We monitored transcutaneous PO2 continuously and recorded the timing and synchrony of chest wall movement during the respiratory cycle by means of mercury strain gauges placed over the upper chest wall and abdomen.Sleep state was determined from observation of body movement, respiratory variation,and electro-oculograms.

Strikingly in all 10 infants PO2 was consistently lower and more variable during A.S.(see table). During A.S. the upper chest wall moved asynchronously with respect to the abdomen 80% of the time.This never occurred during Q.S. These findings reveal a marked instability of arterial PO2 accompanying asynchronous movement of the upper chest wall in the healthy term neonate during A.S. The lower and more variable PO2 may be due to regional ventilation/perfusion inequalities. Apneic episodes (>5 sec)occurred in 5 of the infants,only during A.S.These findings may have important implications in our understanding of the higher incidence of apnea during A.S. in both preterm and term infants.

PO2:	Mean	Coeff.Var.
Q.S.	77.0mmHg	1.7%
A.S.	70.5mmHg	6.1%
	p<.001	p<.01

1205

LUNG MATURITY AND THE PATENT DUCTUS ARTERIOSUS: WHY VENTILATE THE PDA? T. Allen Merritt, Jack Jacob, Thomas A. Clarke, Louis Gluck, Univ. of Calif. San Diego, Dept. of Pediatrics, La Jolla, Ca.

Alterations in tracheal aspirate phospholipids (TAPL) during recovery from respiratory distress syndrome (RDS) previously have been defined (Ped. Res. 10:984, 1976), the appearance of phosphatidylglycerol (PGL) on day 5 indicating lung maturity. In 11 preterm infants (B.W. 1302±357 gm; G.A. 30.6±2 wks) with sequential TAPL, 6 infants followed a characteristic maturational time course while in 5 infants PGL appeared on day 2 indicating "early" lung maturity. In these 5 infants hemodynamically significant PDA (bounding pulses, murmur, hyperactive precordium; LA/Ao >.9 pulmonary plethora) occurred at 2.2±.4 days compared to 2.4±3 days in infants showing lung maturation on day 5. Both groups had similar oxygen requirements & duration of ventilation (93.8 hr. vs 118.8 hr.) to maintain Pao_2 >50 torr & acid-base equilibrium inspite of biochemical evidence of lung maturity in the former. Evidence of pulmonary maturation in infants with pulmonary hyperperfusion due to a large left-to-right ductal shunt suggests that continued & increasing ventilatory requirements are due to ductal shunting rather than surfactant deficiency. Sequential TAPL in infants with RDS offers a basis for predicting ventilatory requirements during the course of RDS whereby maturing TAPL may suggest the timing for ductal closure; this deteriorating pulmonary function is owing to pulmonary edema rather than immature surfactant.

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