UPPER AIRWAY OBSTRUCTION AND HEART FAILURE IN PATIENTS WITH HEINER'S SYNDROME, Thomas F. Boat, Victor Whitman, Stephen H. Polmar, Carl F. Doershuk, Robert C. Stern and LeRoy W. Matthews; Case Western Reserve Univ. Sch. of Med., Rainbow Babies and Childrens Hosp., Dept. of Pediatrics, Cleveland.

Two conditions, right heart failure secondary to upper airway obstruction and pulmonary hemosiderosis with serum precipitins to cow's milk (Heiner's Syndrome), have been simultaneously documented in 3 children. A total of 6 patients with Heiner's Syndrome were found by screening for multiple precipitins in 160 consecutive children with idiopathic chronic lung disease. All 6 were black; 3 were infants and 3 were 1-4 years of age. All had normal or elevated serum immunoglobulins, normal sweat chlorides, and AA or AS hemoglobin. None were clearly predisposed to aspiration. All displayed typical manifestations of Heiner's Syndrome; recurrent pulmonary infiltrates (6/6), hemosiderin-laden macrophages (5/6), intermittent wheezing (4/6), eosinophilia (4/6), iron deficiency (4/6), and failure to thrive (4/6). The 3 older patients also manifested chronic rhinitis, large adenoids, evidence of right heart failure, hypercapnia and acidosis during sleep, and pulmonary/systemic resistance ratios of 1/2 to 1/3 with resultant mean pulmonary artery pressures from 35-45 mmHg. Symptomatic therapy, a cow's milk-free diet, and adenoidectomy when indicated led to improvement in all patients. Heiner's Syndrome and upper airway obstruction with pulmonary hypertension may be two stages, early and delayed, of the same process. Early dietary intervention may prevent cardiac complications and CO_2 narcosis in children with this process.

DEVELOPMENT OF A PRACTICAL DISPOSABLE CPAP HEAD BAG, Paul A. Byrne, M.D., B. Kent Garlinghouse. St. Louis University Sch. of Med., Cardinal Glennon Memorial Hosp., Dept. of Ped., St. Louis, Missouri. Introduced by: Arthur E. McElfresh, M.D.

A practical disposable head bag has been developed for delivering Continuous Positive Airway Pressure (CPAP), obviating the need for the insertion of an endotracheal tube or a nasal piece.

The head bag itself is made of transparent polyvinyl chloride film allowing visualization of the face and head of the infant. A vinyl press - closure zipper is incorporated in the head bag so that with a simple separation of the zip lock, access to the nose and mouth for suctioning is possible.

Air-oxygen mixture then passes to the head compartment. The pressure throughout the system, both in the neck collar and in the head bag, are essentially the same.

15 infants have been treated. 3 infants developed a pneumothorax. 3 are considered failures because it was necessary to intubate and ventilate these infants. 2 infants developed a macular papular rash on the neck resembling erythema toxicum. Smear demonstrated eosinophilia and bacterial culture was negative.

EARLY PULMONARY FUNCTION STUDIES OF INFANTS WITH BRONCHO-PULMONARY DYSPLASIA (BPD): P.A. Branca, J.H.Chen, G.Polgar, A.B.DuBois, and M.Delivoria-Papadopoulos. Depts.of Ped. and Physiol.,Univ. of Pennsylvania,School of Medicine, Phila.,Pa.

In 3 infants (birthweight 920,1720 and 2030 g) with clinical and radiologic BPD, minute volume ($\dot{V}_E$), lung compliance C_L (ml/cm H_2O/kg), pulmonary resistance R_L (cm H_2O/L/sec) and blood gases were measured repeatedly during the first eight weeks of life. All had severe respiratory distress syndrome at birth and they were treated with high O_2 and assisted ventilation for varying periods of time. Two infants remained O_2 dependent until death at 4 and 11 wks. of age, and the survivor until 4 months of age. The mean of the measurements in each infant was significantly below normal for C_L/kg, 0.3, 0.4, and 1.1 respectively and significantly increased for R_L at mid-inspiration 107, 81 and 52, at mid-expiration 190, 218, 247, and mean R_L 143, 158, 109 respectively. $\dot{V}_E$ was increased (720 ml/min). Arterial blood gases showed hypoxia and hypercarbia reflecting the infants' clinical condition. A large portion of the abnormalities could be explained by an extreme loss of lung volume (atelectasis) and lung fibrosis. This was confirmed by necropsy in 2 cases. The two most severely ill infants had persistently low lung compliance and succumbed due to intractable lung damage.

OUTPUT OF THE FETAL RESPIRATORY CENTRE IN-UTERO. Victor Chernick and Andrija Bahoric. Dept. Pediat., Univ. of Manitoba, Winnipeg, Canada.

Dawes, using indirect methods described the presence of fetal respiratory movements up to 40% of the time with a frequency of 1 to 4 Hz. We undertook to monitor fetal respiratory centre output in-utero more directly by simultaneous recording of tracheal pressure & phrenic & diaphragmatic electrical activity in awake sheep. One week was allowed for recovery following surgery. Four fetuses were studied from 132 days to 145 days gestation and a total of 29.4 hours of recording analyzed in detail. Phrenic nerve activity was found in all fetuses and was coupled with diaphragmatic activity and tracheal pressure changes. In contrast to newborn and adult animals, fetal phrenic nerve activity was not phasic but consisted of episodic bursts interspersed by quiescent periods lasting up to 30 minutes. There were two types of bursts: a) a short brief burst with an average duration of 267 msec, b) a more prolonged burst with an average duration of 1.9 sec. Type A burst occurred 1313 times and type B 462 times during 29.4 hours; thus fetal respiration occurred during a total of 20.3 minutes or approximately 1% of the time. Continuous respiration was seen on 2 occasions just prior to fetal death. These studies indicate that normally the fetal respiratory centre during late fetal life is only minimally active in-utero. Continuous intrauterine respiration is probably associated with fetal asphyxia. Supported by MRC (Canada) & Children's Hospital Research Fdn.

THE CONTROL OF RESPIRATORY FREQUENCY IN PREMATURE INFANTS. M.H. Bryan, A.C. Bryan. Dept. of Pediatrics, Univ. of Toronto and the Research Inst., The Hosp. for Sick Children, Toronto, Canada (Intr. by P.R. Swyer).

There are two components to respiratory frequency in the premature infant: a phasic component related to the Hering Breuer reflex and a tonic component set by the bulbo-pontine centres. We have previously shown that the phasic component is related to gestational age.* We have now attempted to quantitate the bulbo-pontine output by measuring the inspiratory duration and pressure developed during airway occlusion at FRC. We have studied 10 infants under 1400g., all with periodic breathing, and correlated the results with the blood gases. There is a significant ($p<0.01$) correlation between inspiratory duration ($1/T_{Iocc}$) and the rate of change of pressure (dP/dt) with pCO_2 ($1/T_{Iocc} = 0.09\ pCO_2 - 1.66$). There is no correlation with the pO_2. These results suggest that the bulbo-pontine output is primarily determined by pCO_2. However, because inspiratory duration becomes so short, the maximum pressure developed falls with rising pCO_2. This makes the respiratory system very vulnerable to respiratory loads. This vulnerability is lost in older premature infants as they develop large pressures when breathing against an obstruction.

* Pediat. Res. 7, 291, 1973.

PYRIDOXINE TREATMENT OF BRONCHIAL ASTHMA. Platon J. Collipp, Sanford Goldzier, Nathan S. Weiss, Joseph Reiss, Yussef Soleymani, Richard Snyder. Dept. of Ped., Nassau County Med. Ctr., E. Meadow, N.Y. and Maimonides Med. Ctr., Brooklyn, N.Y.

76 children (age 2-16) with bronchial asthma received pyridoxine (200 mg) daily or placebo in a double-blind study. Each day their parents completed data sheets assessing their symptoms and other medications. Each month they were examined by their allergist and provided with new sheets and medication. Duration of the study was 5 months. There were no significant differences in the 2 groups at the outset, but by the second month the pyridoxine group had significantly ($p < 0.02$) less asthma attacks, and less bronchodilator medicine was being used. Significant differences in wheezing, tightness in chest, breathing difficulty and cough medicine were also seen. We had previously reported (SPR, 1972) significant abnormality in excretion of xanthurenic and kynurenic acid before and after oral tryptophane loading which returned toward normal with oral pyridoxine therapy. These and other studies suggest that asthmatic children have a relative increase in serotonin and decrease in epinephrine production which are at least partially reversible with pyridoxine therapy.