PULMONARY HYPERTENSION (PH) AND CONGENITAL DIAPHRAGMATIC HERNIA (CDH). J. J. Pomerance, C.J. Richardson, D. Collins, K.W. Travis, and S.W. Turner, UCSD Sch. of Med., Dept. Ped and Surg. and Div. Ped. Card. and Anesth. Childrens' Health Cent. San Diego (Intr. Louis Gluck)

Despite rapid diagnosis, modern surgical technique and intensive pre- and post-operative care, survival of infants with CDH who present within the first 24 hours of life is poor. Some authors suggest that the degree of lung hypoplasia is often the determinant in survival. In our experience almost all infants with CDH can be adequately ventilated following surgery, yet many of these infants subsequently die. After surgery for CDH, 4 infants had blood sampled from pre- and postductal sites. All infants demonstrated R→L shunting suggesting PH. A 5th infant who became progressively hypoxic following surgery underwent cardiac catheterization. No congenital cardiac lesions were found. However, PH with R→L shunting through the foramen ovale and ductus arteriosus (DA) were demonstrated. In addition left pulmonary venous desaturation was present. Both tolazoline (TZL) and chlorpromazine (CPZ) were injected directly into the main pulmonary artery (MPA). Pulmonary artery pressure (PAP) dropped, systemic arterial saturation rose, and pulmonary circulation improved. It was unclear whether the improvement was fortuitous. Subsequently PAP rose and repeat doses of TZL and CPZ administered into the MPA caused little response. 14 hours later the infant died. PH complicating CDH probably occurs frequently. Establishment of pulmonary normotension may be necessary for survival.

Correlation of blood gas, lactate, and illness in newborn. R.C. Rosan, R.L. Beach, S.N. McEwan, & Y. Chen. Cardinal Glennon Mem. Hosp. for Children, Dept. of Pathology, St. Louis, Mo. 63104

We hypothesized that "true" blood lactate (LA) is an index of respiratory need superior to $Pa0_2$, $PaC0_2$, or pH. Our thesis required a simple, rapid, sensitive LA test. We used a new automated, enzymatic 2-wavelength, true kinetic, low-drift test; recovery c. 90%, precision c. ±5%, volume 5-20 ul, linear dynamic range c. 0.01-2.7 A, time c. 5 min. Normals: 1.7 meq/l ± 0.06 (term 3 d.) 2.0 ± 0.06 (cord). Normal neonatal, young, and adult guinea pigs' pH, gases and LA correlated poorly with or without stress. In sick newborns, r values were also poor, e.g. LA vs pH =0.05, LA/$Pa0_2$ vs pH =0.04. When plots of LA/$Pa0_2$ or LA/pH vs time suggested clinical utility, we constructed by blind technic an arbitrary multifactorial retrospective clinical grade system (CG) from histories, so that each infant received a total grade every 6 hrs. Thus, thousands of CG points provided parameters for 129 LA analyses during respiratory distress. A superior correlation appeared for LA vs total CG 24 hrs later: r=0.7, F=55, better than for gases or pH. These are non-parametric data obtained without human experimentation from patients who clinically required gas studies.

PERIPHERAL BLOOD FLOW DURING CONTINUOUS POSITIVE AIRWAY PRESSURE BREATHING (CPAP) IN TERM NEONATES. W.F. Powers and P.R. Swyer. The Research Institute of The Hospital for Sick Children and the Department of Paediatrics, University of Toronto, Toronto, Canada.

CPAP in adults impairs venous return, and decreases cardiac output and peripheral perfusion. These circulatory changes are generally demonstrable with pressures greater than those employed for neonatal respiratory conditions. We have measured peripheral blood flow in spontaneously breathing infants with CPAP levels that are used clinically.

We studied eight term newborns < one week of age, whose cardio-respiratory systems were normal. CPAP was administered by face mask. Intra-esophageal pressure (IEP - balloon catheter) and leg blood flow ($\dot{Q}_1$ - venous occlusion plethysmography) were measured before and during CPAP. Studies were carried out in the infants' incubators at least two hours after feeding.

The mean increase in peak expiratory IEP was 2.14 ± 1.0 cm H_20 compared to the mean applied CPAP of 7.93 ± 1.0 cm H_20. This 74% attenuation is similar to that reported in infants with RDS treated by CPAP. $\dot{Q}_1$ at atmospheric pressure 8.1 ± 3.0 ml/100 ml/min, and during CPAP was 7.6 ± 3.6 ml/100 ml/min, an insignificant change (paired T = 0.58, p > 0.50).

We conclude that moderate levels of CPAP, similar to levels employed clinically, do not impair leg blood flow in term newborns.

EFFECT OF CYSTIC FIBROSIS SERUM ON NASAL MUCOCILIARY CLEARANCE. Carol M. Rossman, Myrna B. Dolovich, Jerry Dolovich, William M. Wilson and Michael T. Newhouse. (Intr. by R.P. Bryce Larke). McMaster University, St. Joseph's Hospital, Departments of Medicine and Pediatrics, Hamilton, Canada.

Studies were performed to detect a possible in vivo effect of the ciliary dyskinesia factor of cystic fibrosis (CF) serum. Following application of serum to the nasal mucosa of CF patients and normal subjects, nasal mucociliary clearance (MCC) was assessed by a radioisotope technique. Removal of a radioactive droplet applied to the mucocutaneous junction of the nose was quantified by means of an Anger scintillation camera interfaced to a data storage and retrieval system. Autologous serum applied to the nasal mucosa failed to inhibit MCC in normals or in CF patients, nor did CF serum inhibit MCC in normals. By contrast, an inflammatory response induced by locally applied anti-serum to human IgE retarded MCC in CF patients but not in normals: it is presumed that the local effect of anti-IgE on MCC in CF was the result of an exudation of the serum dyskinesia factor. Possibly, recurrent or persistent pulmonary infection in CF relates to a local exudation of this factor, hindering effective clearance of secretions.

Supported by C.F. Foundation, MRC (Canada), Ont. Thoracic Soc., Can. TB & RD Soc.

HISTOGENESIS AND RESIDUAL PULMONARY CHANGES IN PNEUMOCYSTIS CARINII PNEUMONITIS (PCP). Robert A. Price, Walter T. Hughes and Shyamal K. Sanyal, St. Jude Children's Research Hospital, Memphis, Tennessee.

Little information is available concerning the histopathogenesis of PCP and the residual pulmonary structural alterations associated with this disease. Six hundred lungs obtained at autopsy from children dying at SJCRH, therefore, were examined to determine (1) the incidence of active and latent P. carinii infection, (2) the histogenesis of PCP and (3) residual pulmonary alterations in patients who recovered from PCP and subsequently died from other causes.

P. carinii was found in 7% of patients at time of death and was causally related in only 4%. The following sequence of events in the histopathogenesis of PCP was found: (1) an initial asymptomatic phase during which cyst forms of P. carinii are located within alveolar cell cytoplasm, (2) a clinically symptomatic phase during which time parasitic replication is at a maximum and (3) a final phase in which P. carinii is destroyed by the host. Although P. carinii cyst forms are confined to alveolar lumen, histopathologic features are found in both alveolar lumen and septae. Late structural alterations, including collagen deposition in alveolar septae, were demonstrated in a number of patients who had clinically documented PCP. We conclude that P. carinii produces a characteristic pulmonary alveolopathy and that a significiant number of patients develop residual histopathological changes which may result in long-term compromise of pulmonary functions.

ACID-BASE PROFILE AND BLOOD GAS CHANGES IN PNEUMOCYSTIS CARINII PNEUMONITIS (PCP). Shyamal K. Sanyal, Walter T. Hughes and Scott Harris, St. Jude Children's Res. Hosp., Memphis, TN.

A prospective study was designed to determine serial changes in blood gas pattern and acid-base profile in 23 children with PCP. Arterial blood samples were obtained throughout acute and convalescent stage, patients breathing i) room air; ii) 100% oxygen for 15 minutes and analyzed to determine pH, PaO_2, $PaCO_2$, bicarbonate, A-aDO_2 gradient and amount of venous admixture. (R-L shunt).

Initial mean values for pH, PaO_2, $PaCO_2$, bicarbonate, A-aDO_2 gradient and R-L shunt were 7.49 ± 0.056, 48.2 ± 16.8, 25.2 ± 3.4, 368.2 ± 108.1 and 12.4% ± 1.8, respectively and were significantly abnormal ($P<0.001$). Hypoxia was present in 21 and respiratory alkalosis in 20 (uncompensated, 17, compensated, 13). Hypoxia was related to respiratory rate ($r = +0.536$, $p<0.01$).

All but one patient became more hypoxic and needed increases in inspiratory oxygen concentration (FIO_2). Sixteen patients survived (2 with respirator). Survivors without respirator did not need $FIO_2 > 55\%$ and showed improvement within 3.8 ± 0.5 days. Each fatal case developed progressively severe hypoxia, A-aDO_2 gradient and R-L shunt and needed significantly higher FIO_2 ($p<0.001$). At a time when clinical recovery was apparent, hypoxia was still present in 9, as were respiratory alkalosis in 7, abnormal A-aDO_2 gradient and R-L shunt in 12.

We conclude that PCP produces potentially lethal alterations in blood gas and acid base profile and that these have prognostic significance. Equally important, some of these changes may persist even at the time of clinical recovery and thus, suggest residual compromise of pulmonary functions produced by the disease.