53 LIVER TRANSPLANTATION FOR METABOLIC DISORDERS IN CHILDREN.

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Of 166 paediatric liver transplant recipients, 17(10.2%) had transplant for a metabolic disorder. Four had α -1-antitrypsin deficiency (mean age 9.1 y), 5 Wilson's disease (mean age 11.2 y), 2 tyrosinemia (1y & 4y), 1 type IV glycogenosis (1 y). Five had no liver insufficiency: three Crigler-Najjar syndromes (2.2, 3.5 & 7.5 y), 1 type I glycogenosis (7 y), 1 primary hyperoxaluria (10y, liver-kidney transplantation). Type IV glycogenosis patient died subsequently from related cardiac failure. Three patients died from causes non related to their original diseases. 1 year actuarial survival is 79.4%. Patients with tyrosinemia kept mild elevation of succinyl-acetonuria. Primary oxaluria patient keeps high oxaluria 2 months after transplantation. α -1-antitrypsin associated proteinuria disappeared in 1 patient. Liver transplantation corrects metabolic disorders of liver origin. Potential pathogenicity remains in other tissues for pluri-organ metabolic defects.

54 Pulmonary dynamics during high frequency ventilation, evaluated by digital pulmonary fluorography.

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We ventilated 6 NZ white rabbits (wt 3.0-3.8 kg) using high frequency ventilation (HFV) at combinations of frequencies (f) 1 to 8 Hz and mean airway pressures (P_{aw}) 4 to 12 cm H₂O. The changes in thoracic gas volume (TGV) during ventilation were evaluated by digital subtraction fluorography. Fluoroscopic video images of thorax in ap-projection were captured at 40 ms intervals during HFV trials, digitized and stored in computer's memory. Special software allowed analysis of intra-breath changes (at 40 ms intervals) in TGV over regions of interest covering the whole lung or projectional areas of the lungs. Using consecutive intra-breath digital subtractions, tidal volume (V_t)-related changes as well as expiration and inspiration dynamics could be evaluated. Changes in TGV at end-expiration ($-FRC$) were both f and P_{aw} dependent (ANOVA), showing f related air trapping especially at higher P_{aw} 's. V_t 's were f dependent at constant P_{aw} 's. Ventilation at applied f's and P_{aw} 's was mainly synchronous over the lungs.

Digital pulmonary fluoroscopy is a novel technique to monitor pulmonary dynamics during mechanical ventilation. Air leak in ventilator-lung interface does not effect the results. The low radiation dose makes the technique applicable also in newborns.

55 DEVELOPMENTAL CHANGES OF THE MYELIN/OLIGODENDROCYTE GLYCOPROTEIN (MOG): A BIOCHEMICAL AND IMMUNOCYTOCHEMICAL STUDY

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The myelin/oligodendrocyte glycoprotein (MOG) is exposed at the surface of myelin and oligodendrocyte processes. Using an immunofluorescent technique and antibodies to different oligodendrocyte and myelin components, we followed the expression of these markers in cryostat sections of rat optic nerves. Galactocerebroside and proteolipid protein were already present in newborn optic nerves. At 5 days postnatally, the myelin-associated glycoprotein was present in a few oligodendrocytes whereas MOG appeared later. By gel electrophoresis and immunoblotting, two closely migrating bands with apparent M_r of 26 and 28 kDa were identified. Treatment with N-glycanase or trifluoromethane sulfonic acid showed that carbohydrates represent ca 10% of MOG M_r and that carbohydrates were mainly linked by N-bonds. The concentration of both MOG components increased coordinately in mouse CNS during the most active phase of myelin deposition and, with a delay of 2-3 days, followed the increase of myelin and myelin basic protein. These results suggest that MOG is not involved in the early phase of myelin formation, but could play a role in terminating and maintaining the compact myelin sheath in the CNS.

56 CAN RESPIRATORY WATER LOSS AND OXYGEN CONSUMPTION BE MEASURED ACCURATELY DURING VENTILATOR AND CPAP TREATMENT.

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When newborn infants are mechanically ventilated or treated with CPAP, much of the expired air is lost to the environment, and is not returned to the ventilator circuit. A method for measurement of respiratory water loss (RWL) and oxygen consumption ($\dot{V}O_2$), that takes this into account, has been devised and assessed. The method is based on an open flow-through-system, where the air leaving the ventilator circuit is redirected to a soft Teflon funnel attached to the infant's face. The air evacuated from the funnel contains both the expired air in the ventilator circuit and the leakage round the endotracheal tube / CPAP-adaptor. Concentrations of water vapour, oxygen and carbon dioxide in the gas leaving the system, in the ventilator circuit and in the incubator are measured using a mass-spectrometer. Air flows and temperatures are also monitored. The system has been tested by depositing known amounts of 'expired' air into the system. The mean difference between given and recovered gas was for oxygen $+1.0 \pm 2.9$ % ($\pm$ SD, n=11), for carbon dioxide -0.9 ± 2.2 % ($\pm$ SD, n=11) and for water vapour 0.3 ± 3.7 % ($\pm$ SD, n=6).

Thus RWL and $\dot{V}O_2$ can be measured accurately during ventilator and CPAP treatment.

57 PREDICTION OF EXTRA-RENAL COMPLICATIONS AND RENAL OUTCOME IN HAEMOLYTIC URAEMIC SYNDROME.

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Since 1970 we have seen 105 children with diarrhoea-associated haemolytic uraemic syndrome (HUS). Only 6 cases were rhesus negative; significantly lower than expected ($p < 0.05$). Moreover this group were more likely to develop extrarenal complications ($p = 0.02$) and early death ($p < 0.02$). Further analysis was performed on the 78 cases since 1982.

Poor renal outcome was seen in 7 (11%) of the 64 with an adequate follow up period. 4 have hypertension, 1 was transplanted, 4 have chronic renal failure and 1 other has proteinuria. The use of FFP in 41% of cases did not reduce the incidence of renal sequelae or extra-renal involvement. We applied our data to a scoring system that was designed to predict poor renal outcome (Birmingham group, BPA, 1988). We found it to be a poor predictor of renal outcome (sensitivity 33%, specificity 83%). We found a strong relationship with the duration of anuria ($p < 0.0001$); anuria in excess of 10 days predicted poor outcome with a sensitivity of 86% and a specificity of 91%.

Severe extrarenal involvement occurred in 13 of the 78 children (17%) with multiple seizures (7), coma (4), hemiparesis (2), necrotising enterocolitis (4), diabetes mellitus (2) and oesophageal stricture (1). Two of this group died of acute cerebral disease. There was no correlation between the presence of extra-renal disease and subsequent renal outcome; 8 of the 11 survivors have no renal sequelae. The Birmingham score infact predicted severe extra-renal disease ($p < 0.003$) better than poor renal outcome. Other associated parameters were the trough platelet count, the plasma sodium at presentation and the duration of anuria.

58 PULMONARY ARTERY PRESSURE IN HYALINE MEMBRANE DISEASE (HMD).

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In the presence of Tricuspid Regurgitation (TR), Doppler ultrasound can accurately determine systolic pulmonary artery pressure (PAP) by application of the Bernoulli equation to the maximal TR jet velocity. 152 serial ultrasound scans were performed from birth to extubation on 20 neonates ventilated for HMD (27-39 weeks, 945-3020g). Maximal TR jet velocity was measured using continuous wave Doppler. Systemic blood pressure (BP) was measured directly by intra-arterial catheter or by Doppler sphygmomanometry. PAP was expressed as a percentage of systemic BP (PAP/BP%). Results were compared with values obtained from 17 healthy preterm neonates examined serially from birth to 72 hours.

Results: Maximal TR velocity was measurable on 68 occasions:

AGE (hours)	0-12	13-36	37-72	72-110	111-168
mean FiO ₂	48	69	61	47	37
HMD PAP/BP%	88 $\pm$ 5	87 $\pm$ 12	78 $\pm$ 21	69 $\pm$ 10	56 $\pm$ 17
NORMAL PAP/BP%	89 $\pm$ 20	53 $\pm$ 8	45 $\pm$ 9		

** $p < 0.001$, * $p < 0.005$

Serial estimations of PAP can be made in neonates with HMD due to the presence of TR. Pulmonary hypertension is a part of HMD which resolves as oxygen requirements fall.