7 BICARBONATE REMOVAL DURING ACETATE HEMODIALYSIS AND ITS POSSIBLE USE IN RESPIRATORY FAILURE OF THE NEWBORN: ANIMAL STUDIES ON EXTRACORPOREAL BICARBONATE/CO_2-REMOVAL (ecBICCO_2R) AND APNEIC OXYGENATION

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Extracorporeal techniques for respiratory support in the newborn are feasible and established as neonatal extracorporeal membrane oxygenation (ECMO). It has been shown however, that even in severly damaged lungs, sufficient oxygenation (apneic oxygenation) can be maintained by only removing CO_2 with an artificial lung, a technique called extracorporeal CO_2-removal (ECCO_2R).

During routine acetate hemodialysis in 22 patients with end stage renal disease an overall CO_2-removal of 79.1 +/- 15.1 ml was measured, at least one third of the entire metabolic CO_2-production. For total metabolic CO_2-removal, acetate dialysate (22mmol/l) was modified with phosphate buffer (2mmol/l), lactate (5mmol/l) and sodium hydroxide (7mmol/l) to compensate for the bicarbonate loss. In 6 sheep, apneic oxygenation could be achieved with blood flow rates as low as 10-15 ml/kg/min for 4-6 hours.

These data suggest that a hemodialysis procedure for bicarbonate and CO_2elimination (ecBICCO_2R) is an efficient method for CO_2-removal necessitating much lower blood flow rates than other techniques presently used. An other advantage of this technique is the maintainence of acid-base, electrolyte and fluid balance as well as renal replacement in multiple organ failure.

8 RETINYL PALMITATE (Ret palm) IN RAT CONCEPTUS IS ASSOCIATED WITH VITAMIN A (vit A) EMBRYOPATHY.

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The pathogenesis of vit A embryopathy is unclear. This study was performed in order to gain insight into possible mechanisms involved. Vit A (Aquasol A) 50,000 U or saline were administered intragastrically to pregnant (day 9; day 0= sperm detected) rats. Vit A treatment of 22 pregnant rats was associated with 65.8% of abnormal fetuses examined on day 20. Retinol (R) and R metabolites were analyzed by HPLC in rat conceptuses on days 10 and 11 following the same treatments. While no R or R metabolites were detected in control conceptuses, traces of R and a large peak (with RT corresponding to Ret palm) were observed in homogenate extracts of conceptuses at both ages examined. Treatment of these extracts with 1M ethanolic KOH resulted in a single large R peak and disappearance of the Ret palm peak. It has been suggested that retinyl esters are related to vit A toxicity. Contrary to the well regulated passage of vit A across the placenta at lower doses, following very large doses to the pregnant mother vit A reaches abnormally high concentrations and at an earlier age in conceptus. Ret palm may play a role in the pathogenesis of experimental vit A embryopathy.

9 DERMATOGLYPHICS IN CHILDREN WITH ECTODERMAL ANHIDROTIC DYSPLASIA.

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Dermatoglyphic studies were carried out in 10 patients (8 boys and 2 girls) with ectodermal anhidrotic dysplasia, and compared with 200 healthy controls.

Severe malformations of dermal ridges among index cases made quantitative analysis inaccesible and therefore only qualitative dermatoglyphic parameters were examined. The most significant dermatoglyphic patterns of hands and feet which differentiated the patients from the controls were as follows:

1. significantly higher frequency of arches on the fingertips (boys: 31.4% vs 2.4%; girls: 15% vs 6%),
2. higher frequency of arches on the toetips (boys: 45.7% vs 11.7%; girls: 30% vs 16.4%),
3. absence of whorls on the finger and toetips,
4. increased incidence of double or ulnar loops in the hypothenar area (boys: 78.5% vs 13.0%; girls: 100% vs 15.5%),
5. significant increase in the incidence of Sydney line on the hands (boys: 56.2% vs 2.0%; girls: 50% vs 6%),
6. severe ridges malformations revealing a characteristic appearance of "ridge hypoplasia" or "ridge dissociation".

10 LYSINURIC PROTEIN INTOLERANCE (LPI): A POSSIBLE DEFECT IN DIAMINO ACID TRANSPORT IN PULMONARY ALVEOLAR EPITHELIUM.

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LPI is autosomal recessive disease with a defect in diamino acid transport in renal, hepatic and intestinal cells. There has been 3 cases of acute respiratory failure among 32 LPI cases, age 3 to 46 years; two of them died. Therefore, amino acids in bronchoalveolar lavage (BAL) and plasma were studied in 4 cases of LPI and 9 controls. Amino acid concentrations in the alveolar fluid were calculated (J Appl Phys 60:532, 1986). In LPI concentration of arginine in alveolar fluid was 19.9 (range 11.3-30.0) and in plasma 36.4 (15.9-56.9) umol/l; healthy controls 6.2 (2.6-15.2) and 121 (99-142) umol/l, respectively. Distribution of amino acids in alveolar fluid/ plasma (mean, range, umol/umol) was as follows:

*	Arginine	Lysine	Leucine	Glycine
1 (n=3)	0.57(.53-.65)	.22(.20-.26)	.14(.12-.17)	0.23(.18-.27)
2+ (n=3)	0.06(.02-.11)	.10(.04-.19)	.20(.12-.36)	0.35(.19-.53)
3+ (n=3)	0.15(.07-.27)	.31(.24-.37)	.27(.15-.48)	0.65(.20-.76)
4+ (n=3)	1.06(.80-1.5)	.90(.36-1.8)	.87(.78-.98)	1.40(.72-2.0)

* asymptomatic LPI,+ No LPI: 3.Lung injury,4.Respiratory failure.

Surfactant was normal in asymptomatic cases. - We propose that in LPI a defect in basolateral diamino acid transport in alveolar epithelium predisposes to acute respiratory failure.

11 MEMBRANE TOXICITY OF DUCHENNE MUSCULAR DYSTROPHY (DMD) SERUM IS LIPOPROTEIN-BOUND

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A toxic serum factor isolated from sera of DMD patients could be shown to increase the membrane fluidity of DMD and control lymphocytes [Hübner C et al. (1987) Pediatr Res 22:488-492]. We have recently found that isolated DMD lipoproteins have the same effect. Fluorescence anisotropy of diphenylhexatriene in peripheral blood lymphocytes (PBL) was significantly decreased when PBL were incubated for 4 h in DMD lipoproteins as compared to 4 h incubation in control lipoproteins (r=0.168±0.007, n=10 vs. r=0.182 ±0.012, n=9; p<0.02, Mann-Whitney test). Similar results could be obtained after incubation of PBL in the isolated LDL fraction of DMD lipoproteins versus control LDL (r=0.190±0.011, n=9 vs. r=0.206±0.007, n=7; p<0.02). Therefore, the LDL fraction seems to be mainly responsible for the increase of PBL membrane fluidity in DMD patients.

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12 L-CARNITINE REPLACEMENT THERAPY IN CHRONIC VALPROATE TREATMENT.

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Valproate (VPA) is known to cause carnitine (C) deficiency. 10 children receiving chronic VPA treatment were given equimolar C (1.2 mg C/mg VPA) concomitantly for 14 days. The plasma level of β-hydroxybutyrate was lower in VPA treated children than in the control subjects (31.8±7.4 vs 90.0±21.4 nmol/ml, means±SEM, p<0.05), which remained unchanged after the C treatment (29.7±7.1), showing that the C was not able in itself to improve the plasma ketone level. The plasma level of FFA, triglycerides and cholesterol remained unaffected by C treatment, the level of HDL cholesterol decreased in the supplemented group. The daily excreted total N was not affected by C treatment (6.0 ±0.5, 7.3±0.3 and 7.3±1.0 g/day; day 0, 14 and control subjects) with no changes of excreted urea and ammonia suggesting, that the organism does not utilize proteins (and/or amino acids) as alternative fuels instead of ketone bodies during VPA treatment.